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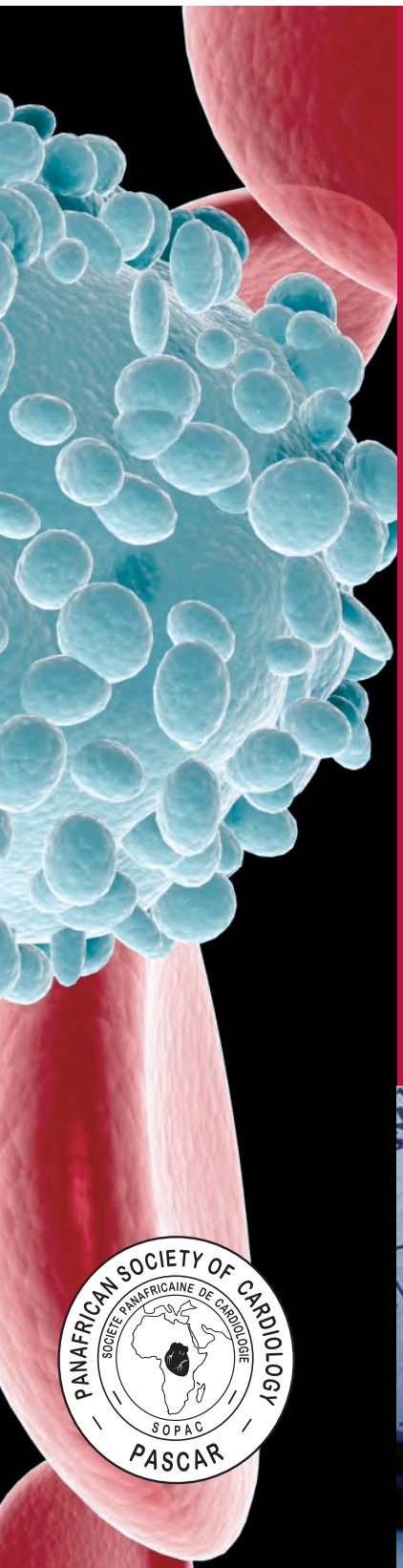

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References
1. Sever PS, Dahlof B, Poulter N, Wedel H, et al, for the ASCOT Investigators. Lancet. 2003;361:1149-58

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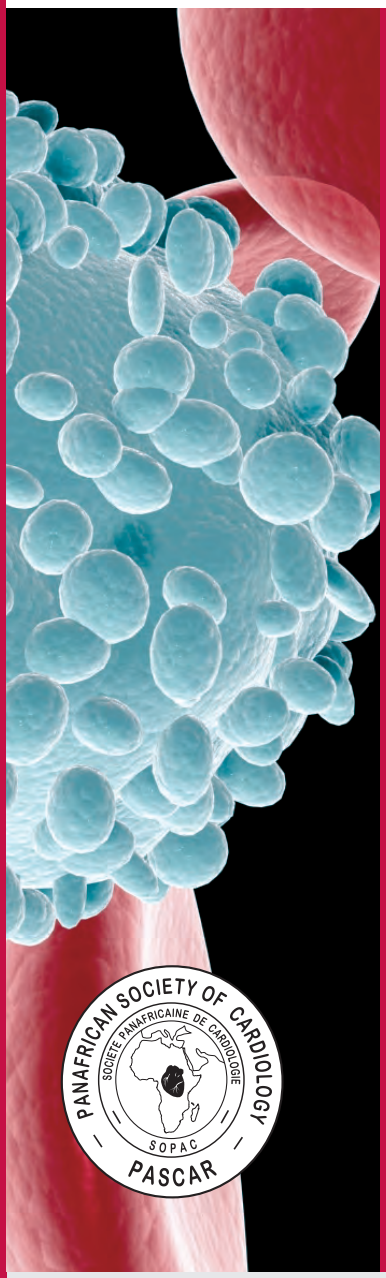

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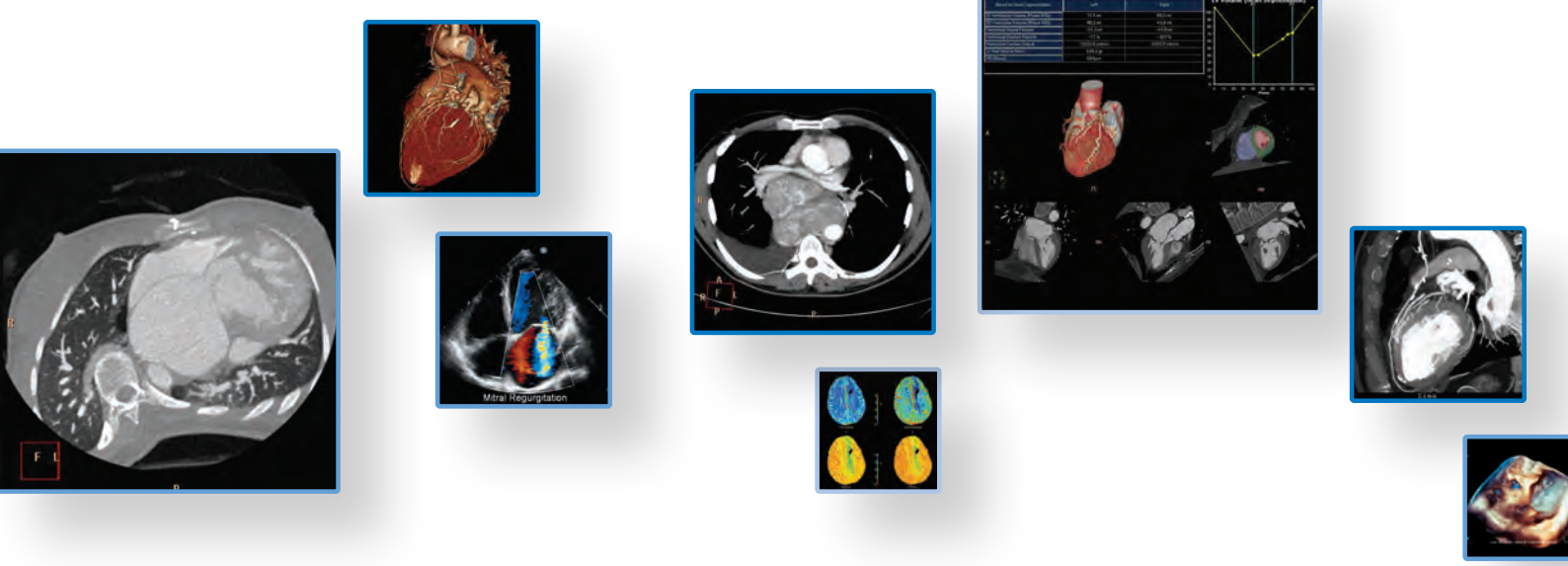

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Topic	*Multidimensional Imaging and its application in children and adults with congenital heart disease – where are we now?* Professor Dr Gerald Greil, Consultant Pediatric Cardiology and Director of the Congenital Cardiac MRI Imaging Service at Guy's and St. Thomas' Hospital/Evelina Children's Hospital, UK

Date	Thursday, 21 February 2013
Venue	Cape Town ICC – Room 2.4
Time	7:00 a.m. – 8:45 a.m.
Topic 1	*Structural Heart Disease Interventions – The growing wave of new therapies.* Thomas E. Fagan, Associate Professor of Pediatrics, Children's Hospital Colorado
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SURGERY, ANAESTHESIA AND INTENSIVE CARE

8: SYNERGIES IN OPEN-HEART SURGERY IN ZAMBIA IN 2011

John Musuku[1], Bruce Bvulani[1], Emmanuel Makasa[1], Munanga Mwandila[2], Harsh Singh[2], Wilson Mbewe[1], Slavetilana Kalininchecho[1], Dorothy Kavindele1, Evans Mulendele[1], Christopher Kangwa[3]
[1]University Teaching Hospital, Lusaka, Zambia
[2]Christchurch, New Zealand
[3]Cardiac Trust of Zambia, Zambia

Background: Currently, heart operations are not done in Zambia due to lack of infrastructure, equipment and skilled manpower. Most patients have heart operations abroad at very high costs. Even if the government sends patients abroad for these heart surgeries, it cannot afford the prohibitive costs for every needy Zambian. Although the Ministry of Health through the University Teaching Hospital *ad hoc* committee has the responsibility of ensuring that patients needing specialised treatment are given the opportunity either locally or abroad, there is a long and growing waiting list of patients needing cardiac surgery. Unfortunately most patient on the waiting list die before their turn for specialised heart surgery.
Objectives: To present the results of open-heart surgeries done in Zambia by the Mutima project.
Methods: Fifteen patients were selected for surgery after meeting the criteria; seven were operated on. Five patients had rheumatic heart disease (RHD), one had a PDA and another had an atrial myxoma. The median age group for the patients was 29 years and all were female.
Results: Seven female patients with age range of 14 to 43 years were successfully operated on with good results. Five patients had valve replacement for RHD, one had left atrial myoma excision and one a PDA ligation. Three patients received tissue valves and two had mechanical valves.
Conclusion: Visiting teams are a short-term solution but are not sustainable. They do help to clear the patient backlog on the waiting list and stimulate interest in local personnel through transfer of skills in cardiovascular medicine and surgery.

10: THE VARIATION IN PLASMA CORTISOL LEVELS IN RESPONSE TO ANAESTHETIC INDUCTION WITH ETOMIDATE OR KETAMINE IN CHILDREN UNDERGOING INTRA-CARDIAC REPAIR OF TETRALOGY OF FALLOT

Sandeep Chauhan, Anil Pandey, Sachin Talwar, R Lakshmi
All India Institute of Medical Sciences, New Delhi, India

Objective: To compare the effect of a single induction dose of etomidate or ketamine on plasma cortisol levels in children with tetralogy of Fallot (TOF) undergoing intra-cardiac repair on cardiopulmonary bypass (CPB).
Methods: This was a prospective, randomised trial performed at a tertiary-care hospital on 30 children with TOF undergoing intra-cardiac repair on CPB. After random allocation of the children to two groups, they received either etomidate or ketamine 0.2 mg/kg intravenously for anesthetic induction, along with fentanyl 2 mcg/kg and midazolam 100 mcg/kg. Anaesthesia was maintained with sevoflurane in air:oxygen. Serum cortisol was measured on three occasions: pre-operatively, at the end of surgery and 24 hours postoperatively.
Results: The two groups were comparable with regard to age: 18.86 ± 3.81 months in the etomidate group versus 17.93 ± 4.68 months in the ketamine group; bypass times: 60.87 ± 6.20 min in the etomidate versus 64.35 ± 5.06 min in the ketamine group. Baseline plasma cortisol (normal 5–25 mcg/dl) in the etomidate group (19.91 ± 3.51 mcg/dl) decreased significantly at the end of surgery (5.78 ± 2.0 mcg/dl) and rose to significantly higher than baseline values at 24 hours (27.31 ± 8.30 mcg/dl). The baseline cortisol levels in the ketamine group (20.91 ± 3.19 mcg/dl) increased significantly at the end of surgery (44.02 ± 5.49 mcg/dl) and remained significantly higher than baseline at 24 hours (45.93 ± 3.05 mcg/dl). Plasma cortisol levels in the etomidate group at the end of surgery and 24 hours postoperatively were significantly lower than in the ketamine group.
Conclusion: This study shows that etomidate is a suitable and safe agent for suppression of the increase in serum cortisol associated with the use of CPB in children with TOF undergoing intra-cardiac repair.

16: POST-OPERATIVE ICU COURSE OF INFANTS BELOW 2.2 KG UNDERGOING CARDIAC SURGERY

Akhter Mehmood, Sameh Ismail, Mohamed Kabbani, Riyadh Abu-sulaiman, Hani Najm
King AbdulAziz Cardiac Centre, King AbdulAziz Medical City, Riyadh, Saudi Arabia

Introduction: Infants with low body weight (LBW) are major challenges for postcardiac surgery care. We conducted this study to compare postoperative course and outcome of infants weighing 2.2 kg or less with a matching group of infants with normal body weight who underwent similar cardiac surgery.
Methods: We retrospectively reviewed all infants below 2.2 kg who underwent cardiac operations at our institution from January 2001 to March 2011. Cases with LBW (group A) were compared with the matching group (group B) of normal body-weight infants who had similar cardiac surgery and matching surgical risk category. We compared demographics, ICU parameters, complications and short-term outcome of both groups.
Results: Thirty-seven patients were included in group A and 39 in group B. Except for weight (2.13 ± 0.08 kg in group A vs 3.17 ± 0.2 kg in group B), there was no statistical difference in demographic data between the groups. Cardiac procedures included coarctation repair, arterial switch, VSD repair, tetralogy of Fallot repair, systemic-to-pulmonary shunt and Norwood procedures. Patients in group A had a statistically significant difference from group B in terms of bypass time ($p = 0.01$), duration of inotrops ($p = 0.01$), duration of mechanical ventilation ($p = 0.004$), number of re-intubations ($p = 0.015$), PCICU length of stay ($p = 0.007$) and mortality (13.5% in group A vs 0% in group B, $p = 0.02$).
Conclusion: Patients with LBW below 2.2 kg can go for cardiac surgery with overall satisfactory results but with increased risk of ICU morbidity and mortality.

37: S-100B PROTEIN AND PERI-OPERATIVE BRAIN INJURY IN CONGENITAL HEART DISEASE INFANTS AND CHILDREN UNDERGOING OPEN-HEART SURGERY USING CARDIOPULMONARY BYPASS

Omneya Ibrahim Youssef, Nevin Mamdouh Habeeb, Adel Al Ansary, Nermeen Helmy, Nadine Mamdouh Habeeb
Faculty of Medicine, Ain Shams University, Cairo, Egypt

Brain-derived S-100B protein has been shown to be a useful marker of brain injury. Neurodevelopmental problems in patients with congenital heart diseases (CHD) have become the focus of increasing concern.
Aim: To assess level of S-100B protein as a brain-damage marker in patients with CHD undergoing cardiosurgical procedures involving cardiopulmonary bypass (CPB) both pre- and postoperatively.
Methods: Fifteen patients (eight with cyanotic and seven with acyanotic heart disease) with a mean age of 4.8 ± 3.9 years, who were neurologically free of brain damage and had been admitted to the cardiosurgery department for procedures involving CPB, were enrolled in the study. They were compared to 15 healthy children as a control group. S-100B protein levels as well as heart rate, mean arterial blood pressure, haematocrit value, central venous pressure (CVP), PO_2 and PCO_2 were assessed before surgery, half an hour after CPB and 24 hours after surgery.
Results: S-100B protein levels were significantly elevated in patients

half an hour after CPB, and before and after surgery, but not in the controls, with the highest values half an hour after CPB ($p < 0.0001$, $p < 0.001$ and $p < 0.001$, respectively). S-100B protein levels were elevated in the cyanotic compared to the acyanotic group ($p < 0.001$). S-100B protein levels half an hour after CPB correlated positively with aortic clamping time, and negatively with body temperature ($p < 0.01$ and $p < 0.001$, respectively).
Conclusion: Patients with CHD are susceptible to subtle brain damage, which increases during surgical intervention, as evidenced by increased S-100B protein levels.

44: PERI-OPERATIVE ASSESSMENT OF PATIENTS WITH REPAIRED TETRALOGY OF FALLOT UNDERGOING PULMONARY VALVE REPLACEMENT

Xavier Iriart, Jean-Bernard Selly, Philippe Mauriat, Francois Roubertie, Jean-Benoit Thambo
Division of Paediatric and Congenital Cardiology, Bordeaux, France Bordeaux, France

Aim: Pulmonary valve replacement (PVR) is commonly performed in adults with repaired tetralogy of Fallot (TOF) to avoid late complications related to severe pulmonary regurgitation or residual RVOT obstruction. However, few data are available concerning peri-operative complications. The aim of this study was to evaluate the peri-operative complications and to determine predictive factors of the low-cardiac output syndrome (LCOS) in patients undergoing PVR.
Methods: Thirty patients with TOF who underwent PVR between 2008 and 2009 were retrospectively enrolled. LCOS was defined according to lactate level > 3 mmol/l, use of inotrope drug for more than 24 hours, and renal dysfunction. Mean age at valve surgery was 29.5 years (range: 6.5–56.5). Indications were RVOT stenosis (n = 4), severe pulmonary regurgitation (n = 25), and mixed lesions (n = 1). RVOT replacement was conducted with a beating heart using a normothermic CPB (mean time 77 ± 25 min) in 16 patients; 14 patients underwent additional surgery requiring aortic cross-clamp. In these patients, CPB mean time was 113 ± 21 min.
Results: Overall survival rate was 97% at 90 days. Post-operative complications were uncommon (ventricular tachycardia in 6%, mechanical ventilation over 24 hours in 6%, renal dysfunction in 10%) except for LCOS (46%). Prolonged duration of CPB over 80 min ($p < 0.01$) and aortic cross-clamp ($p = 0.03$) increased LCOS (OR 33, 95% CI: 3.18–342.2, $p < 0.01$). Surprisingly, age, right ventricular or left ventricular volume and function, and pre-operative additional lesion (tricuspid regurgitation, residual pulmonary stenosis) were not significatively predictive of peri-operative complications.
Conclusion: These data underline the major role of myocardial protection during PVR in TOF patients. Short-beating heart and normothermic CPB without aortic cross-clamping led to a decrease in LCOS. Additional surgical repair requiring aortic cross-clamping and longer time of CPB should be well balanced with the risk of peri-operative complications.

52: RETROSPECTIVE ANALYSIS OF SURGICAL TREATMENT AND LONG-TERM RESULTS IN CHILDREN WITH TAKAYASU'S ARTERITIS

Maria Elena Soto[1], Cuauhtémoc Vásquez[1], Eleazar Muruato-Ontiveros[1], Samuel Ramírez[1], Felipe Santibáñez[1], Nuria Illana Flores[2], Nilda Espinola Zavaleta[1,2], Rafael Bojalil[1], Rodolfo Barragan[1]
[1]National Institute of Cardiologia Ignacio Chavez, Mexico City, Mexico
[2]Medical Center ABC, Mexico City, Mexico

Background: Takayasu's arteritis (TA) affects the aorta and its major branches. The inflammatory activity is subclinical and there is no consensus on its evaluation. It leads to occlusion stenosis or aneurysm. Clinical manifestations depend of the location and extent of arterial damage, and whether it is reversible, affects the organs and surgical treatment is necessary.
Objective: To evaluate the evolution, presence or absence of inflammatory activity and survival of children and adolescents with TA who received surgical treatment in childhood.
Methods: This was a retrospective study from 1977 to 2006 of 222 cases of TA, classified according to the American College of Rheumatology criteria. Seventy-one received surgical treatment, 14 were children. Arterial damage was classified according to Hata, inflammation by Dabague-Reyes criteria, and type of surgery according to: organ sparing, for lesions with stenosis/occlusion (shunt/replacement), exclusion, and cardiac.
Results: Eleven females were enrolled, mean age was 13 ± 3 years. Symptoms were arterial lesions type I: 14%, III: 21%, IV: 29% and V: 36%; systemic hypertension 64%; dyspneoa 57%; headache 50%; angina pectoris 29%; dizziness 29%; syncope 29%; paresthesias 29%; abdominal pain 29%; and blurred vision 21%. Organ-sparing surgery was carried out in seven patients, bypass in three, exclusion in two, replacement in one and cardiac surgery in two. Inflammatory activity was found in 57%. Twelve survived and the median surgical survival time was 12 years (0–26). Two patients died: seven and five years old. Of those who survived: a woman who presented with terminal renal failure 10 years after surgery. In another two, surgeries were performed, and four cases had inflammatory activity at the time of surgery.
Conclusion: Surgery is safe in the long term. Cardiac and vascular surgery with critical lesions should be performed early. Surgical techniques depend on the affected organ and type of arterial damage. Medical treatment must be prior to, during and after surgery to maintain the remission of inflammatory activity, which requires a consensus on evaluation and thereby improves results in the surgical evolution and survival.

53: THE EFFECT OF MECHANICAL PERIODONTAL TREATMENT IN HYPERLIPIDAEMIA

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[1]Periodontology Department, Khartoum Dental Teaching Hospital, Khartoum, Sudan
[2]Cardiac Surgery Department, Ahmed Gasim Cardiac and Renal Transplant Centre, Sudan

Background: Periodontitis is a bacterial infection which has been classified as a local chronic inflammation. This and cardiovascular disease may share common risk factors.The study aimed to evaluate whether local non-surgical periodontal therapy may influence plasma lipid level in hyperlipidaemic patients with chronic periodontitis.
Methods: Thirty patients (11 female and 19 male, age 30–70 years) were randomly assigned to the study and control groups. Lipid profile, C-reactive protein (CRP) and dental variables were measured at baseline and at the end of the study in both groups.
Results: In the third month, there was a significant decrease in low-density lipoprotein (LDL), cholesterol and CRP levels in the study group compared with baseline values. Also the reduction in pocket depth, attachment loss, plaque index and gingival index were statistically significant in the study group.
Conclusion: The study indicated that periodontitis causes changes in total and LDL cholesterol and CRP levels, and local non-surgical periodontal treatment resulted in a significant decrease in these markers. These results suggest a potential effect of periodontitis-driven systemic inflammation on lipid metabolism.

56: RIGHT VENTRICULAR BYPASS: COMPLICATIONS AND SURVIVAL IN MID-TERM FOLLOW UP

Mariettaa Victoria Lafuente[1], Marisa Di Santo[1], Mariela Mouratian[1], Claudia Villalba[1], Alberto Sciegata[1], Ariel Saad[2], Juan Pablo Laura[1], Horacio Capelli[1]
[1]Hospital JP Garrahan, Argentina
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Background: Right ventricular (RV) bypass is a palliative procedure with a high incidence of complications.
Objective: To evaluate mid-term RV bypass events, analyse long-term mortality and compare the classic surgical technique with the extracardiac conduit Fontan procedure.
Methods: A total of 191 patients between 1987 and 2010 were analysed. They had either atrio-pulmonary (AP) or extracardiac conduit (EC) surgery, with mid-term follow up of 6.5 ± 5 years (1–20 years). They were subdivided according to the type of surgery into two groups: group I: AP, 39 patients with a median follow up of 14 years and group II: EC, 152 patients with follow up of four years.
Results: Fifty-seven per cent of patients (n =116) suffered from complications, including late events (n = 111), arrhythmias (n = 49), atrial flutter (n =13), thrombus (n = 31), cerebrovascular accidents (n = 4), protein-losing enteropathy (n = 9), subaortic stenosis (n = 7), therapeutic catheterisation (n = 43), re-operations (n = 20), reconversions (n = 6), mortality (n = 9), ventricular dysfunction (n = 21), and plastic bronchitis (n = 1). The late global mortality was 4.6% (n = 9). In the univariate analysis, mortality was associated with ventricular dysfunction (p = 0.0000), protein-losing enteropathy (p = 0.0000), atrial flutter (p = 0.0012), re-operations (p = 0.0006), subaortic stenosis (p = 0.0024), thrombus (p = 0.01) and AP surgical technique (p = 0.0004). Multivariable analysis revealed that mortality was related to ventricular dysfunction [p = 0.0003, OR = 27.7 (4.64–165.24)], AP technique [p = 0.0036, OR = 0.06 (0.01–0.40)] and protein-losing enteropathy [p = 0.01, OR = 9.31 (1.53–56.66)].
Conclusion: Patients with atrio-pulmonary surgery presented with a higher incidence of arrhythmias, thrombi, re-operations and mortality in comparison to those with extracardiac conduit surgery. Late mortality was associated with ventricular dysfunction, atrial flutter, protein-losing enteropathy, subaortic stenosis, re-operations, thrombi and atrio-pulmonary surgical technique. Mortality predictors were ventricular dysfunction, protein-losing entheropathy and atriopulmonar surgical technique.

66: A 100-CASE CLINICAL ANALYSIS OF RECONSTRUCTION OF THE PULMONARY VALVE WITH AUTOLOGOUS PERICARDIAL PATCH ON TETRALOGY OF FALLOT CHILDREN

Hong Zhou[1], Jing Lei[1], Hongfeng Gao[1], Bo Li[1], Liang Tao[2], Xiaodong Zhu[2], Guibao Huo[1]
[1]Xinjiang Hospital of Asia Heart Hospital, Xinjiang, China
[2]Wuhan Asia Heart Hospital, Wuhan, China

Objective: In order to reduce postoperative pulmonary insufficiency (PI), a transannular monocusp patch was implanted in 100 patients with severe tetralogy of Fallot (TOF) or double-outlet of the right ventricle (DORV), who had a hypoplastic pulmonary valve.
Methods: We have developed a method to reconstruct a triple-cusp pulmonary valve ring. The patient's pulmonary valve ring was divided into two parts, and a transannular monocusp patch was implanted as the third one. The results of follow up were obtained within one year of the correction, and were compared with all patients who had received a simple transannular pericardial patch. The mean maximal systolic pressure gradient between the right ventricle and the pulmonary artery were compared pre- and postoperation (73.95 ± 30.42 mmHg; 15.67 ± 10.62 mmHg), and the degree of PI was not significant. The available monocusp patches could prevent or reduce postoperative pulmonary insufficiency in patients with severe tetralogy of Fallot. There were no deaths and in half the patients, no low-heart output syndrome was observed.
Conclusions: Children with symptomatic hypoplastic pulmonary valve rings should be repaired early and mid-term follow-up results are good. Improved operative techniques and emphasis on pericardial pulmonary valves play an important role in increasing the outcome of surgical treatment.

79: 15-MINUTE REINFORCEMENT TEST RESTORES MURMUR-RECOGNITION SKILLS IN MEDICAL STUDENTS

John Finley[1], Rachel Caissie[1], Pam Nicol[2], Brian Hoyt[1]
[1]Dalhousie University, Canada
[2]University Western Australia

Background: Accurate recognition of murmurs is an important skill but is poorly performed by students and practitioners. Current teaching methods are ineffective. We have previously shown that murmur recognition can be rapidly taught to 90% accuracy with auditory training but it declines within two months without reinforcement. This study examined late reinforcement to restore skills.
Methods: Thirty-six Canadian second-year medical students (17 controls and 19 study group) were enrolled in the study. Students performed an online randomised test of 20 recordings, identifying murmurs as innocent or pathological, followed by a one-hour auditory training programme. The programme presents murmurs in groups of four, each to be identified as normal or abnormal. Difficulty increases through seven levels. Performance was scored with a 20-item test immediately, and after two months. Twenty-two students had re-testing one year later on 20 recordings and a mastery-style reinforcement programme: any student scoring less than 90% took another 20-item test, and if that test score was less than 90%, the student took a final 20-item test.
Results: With initial auditory training, the study group improved from 79.7 (45–100%) to 92.1% (70–100%) (p = 0.005) but after two months declined to 84.2% (65–100%) (p = 0.015), a non-significant increase over pre-test scores. Controls had no change over two months. The one-year follow-up test mean was 81% (55–100%), a significant decline from the two-month post-test. Only six students achieved the 90% level at this test, but after first and second reinforcement tests, an additional six and two students, respectively reached 90%. The mean final score achieved by all students was 90% (70–100%).
Conclusion: Murmur recognition is rapidly learned using auditory training but the skill declines within two months. Most students restored their skills with a brief reinforcement test one year later. The optimal timing is unknown.

82: SURGICAL RECONSTRUCTION OF PULMONARY STENOSIS WITH VENTRICULAR SEPTAL DEFECT AND MAJOR AORTO-PULMONARY COLLATERALS

Richard Mainwaring, Mohan Reddy, Frank Hanley
Stanford University School of Medicine, USA

Background: Pulmonary stenosis with ventricular septal defect and major aorto-pulmonary collaterals (PS/VSD/MAPCAs) is an extremely rare form of congenital heart defect. Although it has been assumed that PS/VSD/MAPCAs would be similar to pulmonary atresia (PA) with VSD/MAPCAs, there are currently no data to support this conjecture. The purpose of this study was to review our surgical experience with reconstruction of PS/VSD/MAPCAs.
Methods: This was a retrospective review of 25 patients who were born with PS/VSD/MAPCAs and underwent surgical reconstruction. All patients had pre-operative pulmonary angiography to define the central branch pulmonary arteries and MAPCAs. There were 14 females and 11 males and the median age at first surgery was four months.
Results: There was one operative mortality (4%) and no late mortalities in this cohort of 25 patients; 96% of the survivors have achieved complete repair. There were two distinct sub-groups: 11 patients demonstrated cyanosis and underwent an initial procedure to augment pulmonary blood flow (+ PBF). The remaining 14 patients formed the second group (– PBF).
Conclusions: Outcomes for PS/VSD/MAPCAs were excellent with low surgical mortality and high rates of complete repair. There were two identifiable sub-groups with distinctive differences. These results provide a prognostic outlook for patients with PS/VSD/MAPCAs, which can be compared and contrasted with PA/VSD/MAPCAs.

86: THE APPLICATION OF ALL-AUTOLOGOUS THREE-SINUS REPAIR (MODIFIED BROM OR MYERS TECHNIQUE) TO SUPRAVALVULAR PULMONARY STENOSIS

Toshiki Fujiyoshi, Koji Kagisaki, Takaya Hoashi, Isao Shiraishi, Hajime Ichikawa
National Cerebral and Cardiovascular Center, Oosaka, Japan

Background: Various surgical techniques have been proposed to repair supravalvular pulmonary stenosis (SVPS) in paediatric populations. Whereas growth potential should be promised, excessive expansion under the presence of undiminished high pulmonary arterial pressure should be avoided. We applied all-autologous three-sinus repair, the so-called modified Brom or Myers technique, to SVPS and examined mid-term outcomes.
Methods: Between March 2010 and March 2012, 15 patients (eight males, median age 12 months) with SVPS underwent all-autologous three-sinus repair. Thirteen patients (87%) had previously undergone pulmonary artery banding to treat high pulmonary vascular resistance. Two patients (13%) had Noonan syndrome associated with valvular pulmonary stenosis. Follow up was complete in all patients and the median follow-up period was 13.5 months (range: 1 month to 2.4 years). The data were presented as mean ± standard deviation (range).
Results: There was no mortality. The diameter of the stenotic part at the main pulmonary artery increased from 47.0 ± 14.1% (29.1–70.0) of the normal pulmonary artery diameter at pre-operative evaluation to 108.4 ± 31.7% (58.9–148.1) at six months, then 110.8 ± 15.0% (87.7–134.1) at one year after the operation. Estimated pressure gradient across the main pulmonary artery decreased from 76.2 ± 12.2 mmHg (57.8–108.2) at the pre-operative evaluation to 11.3 ± 12.0 mmHg (1.4–49.0) at six months, then 6.7 ± 5.5 mmHg (2.0–19.4) at one year after the operation. Only one patient showed more than 25 mmHg estimated pressure gradient across the main pulmonary artery, and after careful follow up, was found to have Noonan syndrome. No pulmonary insufficiency was observed.
Conclusions: Mid-term outcomes after supravalvular pulmonary stenosis by all-autologous three-sinus repair were acceptable. Although long-term follow up is mandatory, application of this technique may provide symmetry and growth of the reconstructed main pulmonary artery.

96: THE LONGEVEITY OF CLASSICAL BLALOCK-TAUSSIG SHUNT FOR PALLIATION OF TETRALOGY OF FALLOT: A CASE REPORT AND REVIEW OF THE LITERATURE

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Background: Tetralogy of Fallot (TOF) is a common cyanotic congenital heart disease. Total surgical correction may be preceded by a temporary palliative systemic–pulmonary shunt procedure. We describe here a patient with TOF who has survived on a classical Blalock-Taussig (BT) shunt for 31 years.
Methods: The medical record of a patient who had a BT shunt performed 31 years ago was reviewed. An electronic search was made on the PUBMED database of the National Library of Medicine using the search term 'classical Blalock-Taussig shunt'. The articles were reviewed and a manual search was performed to review other articles of interest found in their references. The review was to examine other reports on the longevity of the classical BT shunt in the management of TOF.
Results: The patient is a 34-year-old male diagnosed with TOF and he had a classical BT shunt in 1981 at the age of three years, with subsequent improvement in symptoms. He has been followed up since then. The last review on 5 September 2011 showed features suggestive of multi-infarct dementia and a mild left-sided cerebrovascular accident with a patent shunt. He has not been able to afford total correction. The search yielded 394 articles which were reviewed.
Conclusion: Although the use of the classical BT shunt for palliation of TOF is declining, it may still be useful in sub-Saharan Africa where access to standard cardiac care is difficult.

98: TAILOR-MADE BICUSPID PTFE VALVE IN RVOT RESECTION: EARLY EXPERIENCE

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Background: The reconstruction of RVOT using monocuspid or bicuspid valves has been shown to reduce the degree of early pulmonary regurgitation and help smooth the postoperative recovery. The long-term result with a bicuspid PTFE pulmonary valve is claimed to be better than with a monocuspid valve. The described method of reconstruction requires a lot of 'eyeballing'. We describe a standardised technique of construction of a bicuspid PTFE valve tailored to the anatomy of the patient, with good early results.
Methods: Twenty-five patients with a diagnosis of TOF had implantation of a bicuspid valve when they needed a transannular patch for RVOT reconstruction. An outflow tract 2 mm larger than the recommended size was used to design the valve. A piece of silk with a small clip as the marker of the required diameter was used to measured the width of the PTFE membrane needed, the 0.1-mm PTFE membrane was folded and the silk thread with marker was used to measure the width of the required bicuspid valve. The length of the PTFE membrane was also measured using a piece of sillk thread from the point of implantation of the valve to the tip of the ventriculotomy incision. The PTFE was sutured using 6-0 prolene suture to the outflow tract. The width of the pericardium needed to patch was measured using a silk thread to measure the required length and cutting the length of the posterior wall of the RVOT tract. A rectangular piece of autologous unfixed pericardium was used for the RVOT reconstruction.
Results: Two patients had moderate PR, the other 23 had trivial to mild PR early postoperatively and it remained the same at one week and at one month of follow up. There was no mortality in this series and all patients were discharged home after a median stay of two days in ICU and five days in hospital.
Conclusion: Fashioning of a bicuspid PTFE valve using tailored measurements provides gratifying early results. Long-term follow up is required to evaluate pulmonary competence in the long run.

99: CORRECTED TRANSPOSITION WITH BIVENTRICULAR OUTFLOW OBSTRUCTION MANAGEMENT BY RELIEF OF SUBAORTIC OBSTRUCTION

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Background: Congenitally corrected transposition of the great arteries (CCTGA) is more often associated with subpulmonary obstruction and very rarely, subaortic obstruction. We report a rare variant of CCTGA with both subaortic and subpulmonary obstructions with no intra-cardiac shunts.
Methods: This was a predominantly asymptomatic three-year-old child who presented with systolic ejection murmur grade 4/6 along the left parasternal border. Transthoracic echocardiography confirmed CCTGA, mitral valve accessory tissue attached to the left ventricular outflow tract (LVOT) causing moderate to severe LVOT obstruction (peak gradient 4 m/s peak). The mucle-bound right ventricle (RV) showed severe infundibular obstruction (peak gradient 50 mmHg). The subaortic obstruction was relieved, leaving the subpulmonary obstruction untouched.
Result: The child had an uneventful postoperative recovery. The LVOT gradient was now less than 8 mmHg, and the RVOT was

similar to the pre-operative value. The child was doing well at the two-year follow up.
Conclusion: We describe a rare presentation of a child with CCTGA and both subpulmonary and subaortic obstruction, relieving the latter before symptoms of systemic obstruction became an issue and deliberately leaving untouched subpulmonary obstruction as a natural PAB. This strategy has the following advantages: (1) leaving the subpulmonary obstruction keeps the LV prepared for a future double switch, (2) delays the onset of systemic AV valve regurgitation by supporting the septum, and (3) reduces the chances of heart block and rhythm disturbances. Whether this strategy is useful needs to be proved by long-term follow up of the child.

100: EXTENSION OF RVOT PATCH AFTER TOF REPAIR TECHNIQUE TO RETAIN COMPETENCE OF PREVIOUSLY PLACED BICUSPID VALVE

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Background: Limiting the length of ventriculotomy has been an accepted practice to reduce the long-term consequence of arrhythmia and right ventricular dysfunction following tetralogy of Fallot repair. Surgeons may increasingly find themselves in situations where extension of a previously placed right ventricular outflow (RVOT) patch with a mono/bicuspid valve may be necessary to relieve the RVOT obstruction. We describe a technique where extension of a previously placed patch can be done, retaining the competence of the valve.
Methods: Two patients who had had transannular patches with PTFE bicuspid valves, needed extension of the patch to relieve infundibular obstruction after TOF repair. The procedure was done on a beating heart with a single large RA cannula. The lower portion of the previously placed transannular patch along with the PTFE membrane was removed and fixed at the either end using an interrupted 6-0 prolene suture. The incision was extended as required to relieve the obstruction. The previously placed patch and membrane was cut transversely, and a new piece of bovine pericardium of required dimensions was used for the extension. The superior margin was sutured to the previous PTFE patch and the pericardial membrane, forming the neo-annulus of the bicuspid valve. The rest of the bovine pericardium was sutured to the ventriculotomy using a 6-0 prolene suture with a 13-mm curved needle.
Results: In both patients the RV pressure, which was suprasystemic, fell to 50% of that of the systemic values, with competent bicuspid valves and uneventful postoperative recovery.
Conclusion: Extension of previously placed competent transannular patches can be done, retaining the competence by creating a neo-annulus at the superior margin of the newly placed extension patch. This significantly hastens the postoperative recovery in spite of having a long ventriculotomy incision. Disruption of the previously working mono/bicuspid valve is not necessary for extending a previously placed transannular patch.

116: LEFT PULMONARY ARTERY PLASTY USING MAIN PULMONARY ARTERY TURNDOWN: A TECHNIQUE IN MANAGING LPA STENOSIS

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Background: Ten to 30% of patients undergoing single or biventricular repair for decreased pulmonary flow have some degree of left pulmonary artery narrowing, attributable to ductal tissue. Surgical relief of LPA stenosis is notoriously recurrent and a variety of approaches have been used, including stenting to tackle this problem. We propose a technique where, in the main pulmonary artery, tissue is used to plasty the left pulmonary artery origin.
Report: A three-year-old male child with bilateral SVC with right > left and unbalanced AV canal, PS with saturation of 65–70% was brought in for bilateral Glenn surgery. His CT angiogram showed severe LPA-origin stenosis with a well-developed main pulmonary artery. During surgery, the MPA stump was divided and the good MPA tissue was turned down on the LPA, akin to subclavian arery turndown for coarctation repair. The Glenn pressures postoperatively were 14-16 mmHg with a transpulmonary gradient of 7 mmHg. His saturations improved to high 80s and he was discharged after an uneventful postoperative course.
Results: The 18-month follow up of the child has shown good growth of the left pulmonary artery origin with no gradient across the Glenn anastomosis.
Conclusion: The MPA turndown technique can be a useful alternative to pericardial patch augmentation or stenting of the left pulmonary artery, especially in the management of single-ventricle patients with LPA-origin stenosis with well-developed MPA.

117: MANAGEMENT OF TOF WITH ABSENT PULMONARY VALVE WITH SYMMETRIC PLICATION OF PULMONARY ARTERIES AND INSERTION OF BICUSPID PTFE VALVE

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Background: Tetralogy of Fallot with absent pulmonary valve (TOF with APV) is a rare condition that can present in infancy, with severe respiratory symptoms and pulmonary anterior translocation. Use of a homograft valved conduit has been recommended to reduce airway compression. We propose a simpler technique of repair without the use of a homograft.
Methods: A 7-kg 10-month old child presented to us with a previous history of respiratory distress requiring ventilatory support. The child was was diagnosed with TOF with APV and was referred for surgery after weaning from ventilation. The child had branch PAs of 24 mm with a narrow annulus. The child underwent intra-cardiac repair with excision of a portion of the anterior wall of the right and left pulmonary arteries and *in situ* plication of the posterior wall over a 7 Hegar dilator. The angle of the branch of the PA with MPA was opened up and two separate patches were used to enlarge the confluence and RVOT. The MPA and RVOT were reconstructed with a bicuspid PTFE valve and autologous pericardium.
Results: The child had an uneventful recovery with no respiratory issues in the postoperative period. Echo showed normal-sized branches of the pulmonary arteries with a competent pulmonary valve.
Conclusion: Symmetric plication of the pulmonary arteries anteriorly and posteriorly can help tailor the size of the branch of the pulmonary arteries without distortion and without resorting to anterior translocation, which would need aortic transaction. *In situ* plication posteriorly may prevent bleeding, and a bicuspid PTFE valve may replace the homograft in RVOT reconstruction of TOF with APV.

118: PULMONARY ATRESIA WITH SEVERE BIFURCATION STENOSIS IN ADOLESCENCE: SURGICAL CONSIDERATION AND POSTOPERATIVE MANAGEMENT ISSUES

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Background: Management of severely cyanotic adolescents can prove both a surgical and postoperative challenge. We present our experience with a 16-year-old girl with saturations of 40%.
Methods: A 16-year-old girl from Bengal presented to us with severe cyanosis (saturation 30–40%) and severe symptoms of hyperviscos-

ity with functional class IV. Investigation revealed a diagnosis of tetralogy of Fallot with pulmonary atresia with right aortic arch. There were confluent branch pulmonary arteries with severe bifurcation stenosis, which was supplied by a posteriorly located stenotic duct. There were no MAPCAs. During surgery it was noted that there was severe peri-adventitial fibrosis of the branch pulmonary arteries, which was supplied by a stenotic duct. The size of the pulmonary arteries looked satisfactory at the hilum. Aorta and SVC transaction provided unhindered exposure to the branch PAs at the hilum, which was reconstructed with 15-mm Goretex tube. The VSD was closed with a PTFE patch and a bovine jugular vein was used to establish RV–PA continuity.
Results: The postoperative course of the child was complicated by bleeding (which was controlled using factor VIIa), reperfusion lung injury and renal failure (which was managed with appropriate ventilator settings and continuous veno-venous haemodialysis). The child made a gradual recovery and was discharged home at the end of the third week. Postoperative echo showed satisfactory surgical repair.
Conclusion: Management of severely cyanotic adolescents can prove both a surgical and postoperative challenge. Early institution of supportive therapy with appropriate multidisciplinary input may provide gratifying results. Whether an interim palliation with shunt would reduce the severity of postoperative complications remains debatable.

122: CROSS-BORDER TRAINING IN PAEDIATRIC ANAESTHESIA: CHALLENGES AND SOLUTIONS

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A number of countries have the basic infrastructure to perform paediatric cardiac surgery. However, they lack the expertise in various fields to perform complicated surgeries. KK Hospital for Women and Children in Singapore, in conjunction with the Singapore International Foundation has undertaken a three-year project to improve the outcomes of paediatric cardiac surgery at Children's Hospital No. 2 at Ho Chi Minh, Vietnam. The paediatric anaesthetic team involved in the project was faced with numerous challenges to fulfill the aims and outcomes of this project. The challenges faced by the team included the language barrier, non-availability of certain equipment, lack of trained manpower, non-availability of certain medications, and certain aspects of managing smaller children (< 5 kg) on pump. A total of three trips lasting five days each have been undertaken. Solutions have been found to all the challenges listed above and the project objectives and outcomes are being evaluated. The WHO's aim of 'Health for all by 2020' can only be met if such trainings are carried out on-site and by overcoming the challenges faced by the trainers.

124: MORPHOLOGY AND SURGERY IN ATRIO-VENTRICULAR SEPTAL DEFECT WITH LEFT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

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Background: Outcome after repair of AVSD may be unfavourably altered by co-existing left ventricular outflow tract obstruction (LVOTO).
Methods: The morphology of 73 heart specimens was examined and compared with the morphology and clinical data of operated patients with atrio-ventricular septal defect (AVSD). The aim of the study was to determine the prevalence of LVOTO and analyse the results of the surgical treatment of AVSD with LVOTO.
Results: LVOTO was found in nine (12.3%) of 73 specimens. It was caused by hypertrophy and anterior displacement of the antero-lateral papillary muscle (five hearts), septal hypertrophy (three), or fibromuscular membrane (one). In our clinical series, LVOTO was found in 19 (2.8%) of 675 patients with AVSD. It was caused by a fibromuscular membrane, septal hypertrophy, abnormal valvar attachments and fibromuscular strands, which occured in combination. LVOTO was present at the time of AVSD repair in eight patients and developed after repair in 11. Membrane excision (nine patients), myectomy (six), excision of abnormal valvar tissue (three) and valvotomy (one) were required. Correction of AVSD consisted of two- or one-patch repair and individually modified plasty of the AV valves. Survivors were examined by echo. There was one (5.3%) early and one (5.3%) late death. The causes of death were heart failure and mitral stenosis, respectively, and resulted in part from the presence of LVOTO in both of them. Two survivors required re-operation for LVOTO. Operated patients remain in a good condition without LVOTO at a mean of 6.5 ± 3.8 years after surgery.
Conclusions: The prevalence of LVOTO was 2.8%. Fibromuscular membrane, septal hypertrophy and abnormal valvar tissue represented the most common causes of LVOTO in the operated patients. Septal hypertrophy, oblique 'cleft', displastic valve and abnormal attachments in LVOT may contribute to LVOTO formation after surgery. LVOTO did not increase mortality but the long-term outcome and the re-operation rate may be influenced by progression of the LVOTO.

135: EXPERIENCE WITH RESECTION OF LARGE LEFT VENTRICULAR FIBROMAS

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Background: Large left ventricular (LV) fibromas are rare tumours with an unfavourable prognosis and high risk of ventricular fibrillation. There is not enough experience with their surgical treatment.
Methods: Since 2004 large LV fibromas have been seen in four children at the ages of 3.2, 1.7, 0.9 and 1.2 years, respectively. One child who was not operated on suffered from an attack of ventricular fibrillation, which resulted in severe neurological deficit. Three patients underwent surgical resection of the tumour. The preoperative diagnosis was set by echocardiography. All fibromas were similarly located at the lateral LV free wall from the base to the apex. The surgery was performed with CPB and blood cardioplegia. It consisted of total excision of the tumour and plication of the LV wall. All tumours were large (70 × 40 × 25 mm) with a similar appearance. They were ovoid, white and stiff. The thin external layer of the LV wall was longitudinally incised and the tumour was excised, leaving both layers intact. Meticulous inspection for bleeding and injury to the inner layer was necessary. The LV wall was plicated.
Results: Total excision without injury to the coronary arteries, atrio-ventricular (AV) valves or connective tissue was possible in all patients. The diagnosis was confirmed histologically. In two operated patients the postoperative course was uneventful. In one patient re-operation for bleeding of the LV wall was required on the first postoperative day. All operated patients survived and are now 8.2, 2.4 and 2.2 years, respectively after surgery, without arrhythmias and with good LV and MV function. The unoperated patient remains in an unfavourable neurological status, though the deficit is improving. In this child a cardioverter/defibrillator was implanted and resection of the tumour is being considered.
Conclusion: LV fibromas can be completely excised. The risk of postoperative complications and arrhythmias is low in surgically treated patients.

141: EXPERIENCE ON THE PRACTICAL USE OF LEVOSIMENDAN IN CHILDREN IN KUWAIT: EXPANDED REVIEW

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Introduction: Levosimendan is a relatively new calcium-sensitising drug that opens adenosine triphosphate-dependent potassium (K_{ATP}) channels with effects that increase myocardial contraction and cause vasodilatation. Its main potential advantages are the improvement of myocardial contractility without increasing oxygen requirements, reduction of ventricular preload, and an anti-stunning, anti-ischaemic effect by opening K_{ATP} channels. We present here a large group of paediatric patients treated with levosimendan.
Methods: This was a retrospective observational study in our paediatric intensive care unit, in which more than 80 patients received levosimendan. There were no adverse events attributable to levosimendan and no instances where the clinical condition worsened after administration. Arterial lactate levels decreased significantly following levosimendan administration during cardiopulmonary bypass for anticipated low cardiac output. In those with established low cardiac output, trends toward improved haemodynamics were seen, with heart rate reduction, an increase in mean blood pressure, a reduction in arterial lactate, and reduced conventional inotrope use.
Results: Levosimendan was safely and successfully used in a significant number of paediatric patients with established low-cardiac output state who demonstrated improved haemodynamics and tissue perfusion, with a tendency to reduced conventional inotrope usage. This warrants its evaluation as an inotrope in the paediatric population.
Conclusion: Levosimendan is the safe drug of choice that could revolutionise the outcome of cardiac surgery in children.

152: CASE REPORT: CARDIOGENIC SHOCK IN A POST-OPERATIVE NEONATE WITH CYANOTIC CONGENITAL HEART DISEASE

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Cardiogenic shock in a post-operative neonate with cyanotic congenital heart disease is a challenging emergency where the cause determines management. Postoperatively, iatrogenic complications can be overlooked, especially when invasive monitoring and haemodynamics become the focus of on-going assessment. We present a male neonate of 34 weeks' gestation with pulmonary atresia and ventricular septal defect who received a right modified Blalock-Taussig shunt on day 3 of life.
Results: Postoperatively he remained ventilator-dependent secondary to nosocomial pneumonia. On day 5 postoperatively he had increasing ventilatory requirements, with an acute deterioration due to cardiogenic shock, requiring aggressive resuscitation. Chest X-ray revealed progressive cardiomegaly. Urgent echocardiography confirmed the presence of a large pericardial effusion. Pericardiocentesis yielded 80 ml of lipid-rich fluid, with immediate clinical improvement. An echogenic focus close to the umbilical venous catheter (UVC) tip was also imaged near the Eustachian valve of the right atrium (RA).
Conclusion: We present a neonate with cyanotic congenital heart disease, post-surgery who presented with cardiac tamponade due to a pericardial effusion. This was a result of infusion from the UVC tip, which although appropriately positioned, had eroded into the pericardial cavity through the right atrium. The echogenic focus noted on echocardiography marked the site of erosion. Cardiac tamponade in a neonate post cardiac surgery can occur unrelated to the operative procedure and its complications. A UVC tip, although appropriately positioned, had perforated through the RA into the pericardial cavity and was the unexpected cause of death in this patient. Catheter iatrogenicity is an important cause of morbidity in postoperative cardiac neonates.

154: MONITORING OF OUTCOME AND PERFORMANCE IN CONGENITAL HEART SURGERY: COMPLETE AND VERIFIED SINGLE-INSTITUTION DATA, 16 YEARS OF PRACTICE

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Background: Analysis of surgical outcomes using raw mortality rate without risk adjustment is inadequate. The Aristotle basic score and EACTS-STS mortality score and categories have been developed based initially on experts' opinion, and finally on huge amounts of multi-continental data. These tools allow for monitoring of team performance over long time periods with regard to increase of complexity and risk of procedures.
Methods: Complete and verified data on 8 404 procedures performed between 1995 and 2010 in our institution were collected in our registry using International Nomenclature for Pediatric and Congenital Heart Surgery, and EACTS-STS database format; 6 992 procedures scored by Aristotle basic score and EACTS-STS mortality score and categories were included. Procedures with less than 25 occurrences were excluded. Hospital mortality, Aristotle basic score, mortality score and categories, and postoperative length of stay were used for measurement of team performance, defined as score × survival/100 and LOS coefficient [100 – LOS (days)/score]. The same calculations were performed using 8 STS benchmark procedures.
Results: Mean annual volume of procedures included in the analysis was 437 (388–510). Mean hospital mortality was 4.55% (3.08–6.81), mean Aristotle basic score was 5.89 (5.60–6.67), mortality score was 0.59 (0.49–0.79), mortality categories were 1.94 (1.76–2.25), and mean postoperative LOS was 14.4 days (10.2–25.2). Over 16 years, continuous increase in team performance followed the increasing complexity of procedures. Mortality score appeared the strongest predictor of death in logistic regression analysis [AUC 0.780 (0.77–0.789)] and was used for calculation of performance and LOS coefficient. Team performance increased from 0.47 in 1995 to 0.75 in 2010 ($p < 0.001$). LOS coefficient increased from 75.3 to 81.3 ($p = 0.85$).
Conclusions: Available quality measures allow for continuous assessment of the team performance in congenital heart surgery. Increasing complexity and risk of procedures is neutralised by accumulated growth of team experience, represented as team performance and LOS coefficient.

168: BUG BUSTERS: THEMED INITIATIVE FOR THE REDUCTION OF INFECTION RATES IN A CARDIAC INTENSIVE CARE UNIT

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Background: To reduce infection rates by raising awareness of infection risks and types, precautions, preventative measures and treatment options.
Methods: Through SWOT analysis we identified strengths, weakness, threats to infection prevention and opportunities for reducing infections in a cardiac intensive care unit (CICU). To promote infection awareness we created a 'BUG BUSTERS' themed acronym to prompt best practices (Bundles, Utility bins, Gadgets, Biopatches, Utilising housekeepers, Sterilising, Transformation website, Educating colleagues, Reducing errors, Stamping out infections), and recruited key staff to become BUG BUSTER champions. Staff and parent notice boards were updated, and hand hygiene, CVL and SSI compliance were audited monthly. CVL route-cause analysis highlighted femoral lines as a problem area. A new policy was implemented, introducing biopatches (antimicrobial dressing) to prevent catheter-related blood stream infections. We organised a study day and created bay leader checklists to disseminate best practices throughout the CICU, and created a BUG BUSTERS newsletter detailing audit results and reiterating ward policies and protocols. Infection-control teaching for new and existing staff in collaboration with educational facilitators highlighted inadequate understanding of infection types. In response we developed a BUG BUSTERS poster displaying viruses vs bacteria, including routes of transmission, isolation precautions, preventative and treatment methods.
Results: Hand hygiene compliance has improved from 93 to 100%

since October 2011. CVL audit compliance has improved from 86 to 100% since July 2011. SSI infections 30 days post cardiac surgery have reduced from 8 to 2% since November 2011

Conclusions: Our BUG BUSTERS themed initiative has led to increased awareness of infection control issues and contributed to a reduction in infection rates of children in CICU.

173: CONTROLLING OXYGENATION DURING INITIATION OF CARDIOPULMONARY BYPASS: EFFECT ON RENAL AND HEPATIC SYSTEMS IN CYANOTIC CHILDREN UNDERGOING CARDIAC SURGERY

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Objective: Cardiopulmonary bypass (CPB) initiated with high oxygen levels exposes cyanotic children to reoxygenation injury, which can affect multiple organ systems. Controlling oxygenation during initiation of cardiopulmonary bypass has been demonstrated to be associated with decreased myocardial injury. This study tested the effect of this strategy on the renal and hepatic systems.

Methods: Thirty-one cyanotic children were randomised to group A (intervention) and group B (hyperoxaemic). CPB was initiated with a fraction of inspired oxygen (FIO_2 0.21), and after one minute of full bypass, FIO_2 was increased at increments of 0.1 per minute to reach 0.6. In group B, CPB was initiated using $FIO_2 > 0.6$. Aortic cross-clamp and CPB time (minutes) was measured. Serum creatinine (mg/dl), aspartate aminotransferase (AST) (U/l) and alanine aminotransferase (ALT) (U/l) were measured pre-operatively (Pre-op) and on postoperative days (POD) 1 and 2.

Results: CPB time (group A median = 71.5, IQR = 64–100; group B median = 95.5, IQR = 58–145, $p = 0.71$), cross-clamp time (group A mean = 59.2, 95% CI = 47.6–70.8; group B mean = 66.57, 95% CI = 47.6–88.5, $p = 0.57$).

Serum creatinine [Pre-op (group A median = 0.6, IQR = 0.53–0.68; group B median = 0.6, IQR = 0.6–0.7, $p = 0.11$), POD-1 (group A median = 0.6, IQR = 0.5–0.76; group B median = 0.6, IQR = 0.5–0.75, $p = 0.54$), POD-2 (group A median = 0.5, IQR = 0.43–0.68; group B median = 0.6, IQR = 0.5–0.77, $p = 0.11$)].

AST [Pre-op (group A mean = 27.5, 95% CI = 24.91–30.09; group B mean = 31.31, 95% CI = 25–37.64, $p = 0.37$), POD-1 (group A median = 100, IQR = 83–132; group B median = 103, IQR = 74.75–146, $p = 0.8$), POD-2 (median = 66.5, IQR = 52–76.25; group B median = 82, IQR = 62–124.75, $p = 0.12$)].

ALT [Pre-op (group A median = 18.5, IQR = 15.25–19.75; group B median = 17.5, IQR = 13.25–22.50, $p = 0.84$) and POD-1 (group A median = 21, IQR = 19–23; group B median = 24.50, IQR = 19.5–34.5, $p = 0.19$] showed no significant difference.

ALT: POD-2 (group A median 19.5, IQR = 16–21; group B median = 25, IQR = 21–33, $p = 0.044$) was significantly lower in group A.

Conclusion: This study suggests possible decreased hepatic injury associated with this protocol.

184: IMPACT OF AFTERLOAD REDUCTION ON CEREBRAL TISSUE OXYGENATION AFTER THE NORWOOD PROCEDURE FOR HYPOPLASTIC LEFT HEART SYNDROME

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Background: Lower cerebral tissue oxygenation has been observed by near infrared spectroscopy (NIRS) after the Norwood procedure. Altered cerebral vascular resistance and pharmacological afterload reduction redirecting blood flow away from the cerebral circulation are possible mechanisms. We evaluated the impact of afterload reduction with Milrinone on cerebral (cSO_2) and somatic tissue oxygenation (sSO_2) in comparison to retrospective controls treated with sodium-nitroprussid and enoximone.

Methods: NIRS and routine intensive care monitoring data were recorded for 24 hours before and 48 hours after the Norwood procedure in 68 hypoplastic left heart syndrome (HLHS) patients (milrinone $n = 34$, control $n = 34$). Average values of the last four preoperative hours (baseline) and of the first and last four postoperative hours (early and late course) were calculated.

Results: Baseline, early and late postoperative cSO_2 values were 58 ± 7%, 52 ± 12% and 61 ± 7% for patients treated with milrinone and 58 ± 7%, 52 ± 9% and 60 ± 6% for controls; sSO_2 values were 58 ± 9%, 78 ± 8% and 69 ± 10% and 59 ± 8%, 76 ± 10% and 67 ± 9%, respectively. Baseline and postoperative NIRS values were not different between groups. cSO_2 was below 40% for 45 (0–720) minutes in patients treated with milrinone and for 50 (0–1 040) minutes in controls ($p = 1.00$). A lower haemoglobin level early after operation was associated with $cSO_2 < 40\%$ for more than 60 minutes (14.9 ± 1.7 vs 16.0 ± 1.3 g/dl, $p = 0.005$). cSO_2 correlated with pO_2 (r: 0.137, $p < 0.001$), with SaO_2 (r: 0.223, $p < 0.001$), and with SvO_2 (r: 0.404, $p < 0.001$). pCO_2 was weakly and negatively correlated with sSO_2 (r: –0.165, $p < 0.001$), but not with cSO_2.

Conclusions: Early after the Norwood procedure, cSO_2 was lowered with both strategies of afterload reduction and the wide difference between cSO_2 and sSO_2 indicates a mismatch between cerebral and splanchnic perfusion. Other strategies to improve cerebral tissue oxygenation after the Norwood procedure are needed.

187: TRUNCUS ARTERIOSUS COMMUNIS REPAIR WITH OR WITHOUT RIGHT VENTRICLE-TO-PULMONARY ARTERY CONDUIT: NO DIFFERENCE IN EARLY AND MID-TERM FOLLOW UP

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Background: Implantation of a conduit between the right ventricle and pulmonary artery is a part of the truncus arteriosus communis (TAC) repair in many centres. Repair without a conduit was implemented in our centre to decrease the re-intervention rate. The aim of the study was to compare early and mid-term results of conduit versus non-conduit repair of the TAC and to assess risk factors for mortality and prolonged hospital stay (> 30 days).

Methods: All patients who underwent two-ventricle repair of TAC between 1995 and 2012 were included in a retrospective cohort study and divided into conduit ($n = 12$) and non-conduit ($n = 20$) groups. The conduit was implanted at the discretion of the surgeon. Data are presented as median (range).

Results: The age at surgery was 28 days (6–466). The mortality rate was 21.8% ($n = 7$). There was no difference between the conduit and non-conduit groups in weight, age at surgery, classification according to aortic versus pulmonary dominance, and duration of intensive care, inotropic support, mechanical ventilation and mortality rate. Cardiopulmonary bypass time and cross-clamping time were longer in the conduit group: 154 minutes (120–513) versus 107 minutes (84–197), $p = 0.006$; and 91 minutes (48–160) versus 65 minutes (31–108), $p = 0.01$, respectively. Truncal valve repair with aortic conduit was required in two patients in the conduit group. Five patients required seven surgical re-interventions in the non-conduit group and one patient required an interventional procedure in the conduit group until hospital discharge ($p = 0.27$). One- and five-year freedom from right ventricle outflow tract re-intervention was 80 and 53% in the conduit group and 80 and 80% in the non-conduit group, respectively ($p = 0.16$). No risk factors for mortality were identified. The needs for mechanical ventilation at the time of surgery and for

surgical re-intervention were risk factors for prolonged hospital stay.
Conclusion: Truncus arteriosus communis repair with or without right ventricle-to-pulmonary artery conduit provided similar early and mid-term results.

209: CONGENITAL PORTAL VEIN ANOMALY AS A CAUSE OF SEVERE PULMONARY HYPERTENSION

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Background: Congenital absence of the portal vein (CAPV) as a consequence of aberrant venous development in the early embryonic period is an extremely rare condition, with only about 40 cases published. Due to the porto–systemic shunt, the mesenteric–splenic venous drainage bypasses the liver and drains directly into the systemic circulation.
Methods: Although considered rare, in our centre in a period of four years we detected three cases of CAPV due to portal vein agenesis with porto–systemic shunting presenting as severe porto–pulmonary hypertension (PPHT).
Results: Two young adults (34-year-old female, 36-year-old male) and a 34-month-old girl presented with pulmonary arterial hypertension (PAH). Right heart catheterisation confirmed severe PAH in all three patients (mean PAP: 65/85/47 mmHg and PVR 8/13 WU, 10.4 WU/m^2, respectively). Due to the clinical course and suspicion of PPHT, abdominal ultrasonography, MR and CT were performed and the diagnosis of CAPV with porto–systemic shunts to the inferior vena cava in all three cases was established, and the diagnosis of PPHT was confirmed. Due to the functional status (WHO II/III/II), specific therapy for PAH was initiated as follows: ambrisentan, sildenafil and bosentan. In two adult patients, treatment (over seven and three years) improved continually to functional status WHO I, and physical performance was highly significantly improved (6 MWT: + 128 m/+ 68 m). The follow up as well as therapy of the child was interrupted after one year due to incompliant parents and according to the GP's information, died three months later due to acute respiratory failure.
Conclusion: CAPV is a rare condition, with only a few cases presented. It is usually associated with cardiac, skeletal and visceral malformations. The association of CAPV with severe PPHT is extremely rare but may be more frequent than expected. However it often remains undiagnosed. The prognosis is dependant on early diagnosis, as well as management with PAH-specific treatment including an experienced PH centre.

215: MODIFIED EXPOSURE IN SURGICAL REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION: FIVE-YEAR EXPERIENCE

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Objective: Surgery for total anomalous pulmonary venous connection (TAPVC) requires either anastomosing pulmonary venous confluence (PVC) to the left atrium (LA) or intra-cardiac baffling. Limited convenience and lack of space during the creation of an anastomosis can potentially compromise the repair and outcomes. The aim of this study was to present our experience with a modified approach in the repair of TAPVC, which provides superior exposure and facilitates repair.
Methods: One-hundred and twenty patients with isolated TAPVC were operated on in the years 2000 to 2010. Conventional approaches to surgical exposure were used in 72 patients (group I). Starting from 2006, modified exposure was used in 48 consecutive patients (group II). For the modified approach, the right pleural cavity is widely opened, the heart is rotated around the axis of the caval veins into a pleural cavity and this maneouvre provides better exposure for creation of anastomosis.
Results: There were two early deaths (4.2%) in the modified-exposure group. Multifactorial analysis has shown the modified exposure to be among the factors responsible for improved outcomes. Follow up was 93.1% with no late deaths or re-interventions in either group. All survivors were asymptomatic at a median follow up of 31.7 months.
Conclusion: Anastomosing the LA to the PVC is a key component of TAPVC repair. Modified surgical approaches to repair may provide better exposure and superior outcomes. Long-term outcomes are good with all approaches.

216: DISTINCTIVE HAEMODYNAMICS IN THE IMMEDIATE POST-OPERATIVE PERIOD OF PATIENTS WITH A LONGER CARDIAC INTENSIVE CARE STAY POST TETRALOGY OF FALLOT REPAIR

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Background: The majority of tetralogy of Fallot (ToF) patients have up to two days' stay in the cardiac intensive care unit (CICU) while some stay longer. We undertook this study to investigate the differences in immediate postoperative (first four hours) variables in patients who had shorter compared to those who had a longer stay in CICU.
Methods: Patients who underwent ToF repair at the Aga Khan University, Pakistan between July 2006 and December 2011 were considered. Exclusion criteria were: mortality in the first 24 hours, pulmonary atresia, absent pulmonary valve syndrome, infection or other non-cardiac cause of prolonged CICU stay. Clinical parameters were compared between the shorter-stay group (SSG) (≤ 2 days) and the longer-stay group (LSG) (> 2 days). Continuous variables are presented as medians.
Results: Eighty-nine patients (LSG 57, SSG 32) were included. There was no difference in age at repair (in years) between the groups (LSG 5 vs SSG 6, $p = 0.07$). LSG had a lower pre-operative saturation (78%) compared to SSG (85%), $p = 0.04$. LSG had a significantly longer total bypass time (150 vs 137 min, $p = 0.02$). Averaged over the initial four post-operative hours, patients in the LSG had a significantly higher heart rate (136 vs 122 beats/min, $p = 0.04$), central venous pressures (CVP) (11 vs 9 mmHg, $p = 0.001$) and inotropic score (IS) (10 vs 8, $p = 0.01$). Averaged over the total CICU stay, the LSG had a significantly higher IS (10 vs 6, $p = 0.009$), CVP (9 vs 8 mmHg, $p = 0.01$), longer duration on inotropes (2 vs 1 days, $p = 0.001$) and mechanical ventilation (1 vs 0.5 days, $p = 0.001$) when compared to SSG.
Conclusion: Patients who ended up staying longer in the CICU had features that were distinctive in the immediate postoperative period. These clinical parameters can be used to predict patients who may need more support and longer CICU stay and thus help in parent counselling.

217: HEARTFELT: MOTHERS EXPERIENCES OF THEIR INFANTS FOLLOWING CARDIAC SURGERY

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Background: Many infants with congenital heart disease require major surgery within weeks of birth. Attachment theory and research

have emphasised the critical importance of the early mother–infant relationship for the general development and future well-being of the child.
MethodsA prospective, longitudinal, staged study was designed to explore the psychological well-being of mothers and babies and the nature of the developing mother–infant relationship following the infant's heart surgery. Complementary qualitative and quantitative research methods were used. Data were collected when the infant was two and nine months old. Mothers' perceptions, standardised measures of maternal distress, and observations of infant interaction with the researcher were compiled at each stage. Maternal distress was evaluated using the Edinburgh Post-Natal Depression Scale, State-Trait Anxiety Scale and Parenting Stress Index Short Form. Infants' well-being was tapped using a measure of social withdrawal (ADBB). In-depth interviews with the mother explored experiences of the diagnosis, infant's hospitalisation and treatment, and her perceptions of her developing relationship with her infant. The resulting narratives were subjected to thematic content analysis.
Results: The cardiac diagnoses varied from an isolated VSD to HLHS. Very high levels of maternal distress were found and almost half of the infants were socially withdrawn Maternal distress was associated with infant social withdrawal. Thematic analysis of the maternal interviews revealed shock and acute stress as central to mothers' experience. Also, mothers' perception of infant responsiveness appeared to affirm life, and recognition of these qualities in the infant may help to facilitate mothers' recovery.
Conclusion: Important issues need to be addressed to provide services for facilitating the well-being of both mother and infant over and above improving surgical outcomes. The research was funded by the NHMRC and carried out with the support of Monash University, MCRI, Monash Medical Centre and The Royal Children's Hospital.

218: ROLE OF FENESTRATION IN THE FONTAN CIRCULATION: HAEMODYNAMIC RESPONSES TO CARDIAC OUTPUT CHANGE

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Background: Although the fenestrated Fontan procedure was developed to assure haemodynamic stability during the acute postoperative phase, controversy exists whether fenestration for the chronic phase is beneficial and how fenestration works in response to homonymic changes. In this study, we investigated the haemodynamic behavior of the fenestrated Fontan circulation in response to increased and decreased cardiac output.
Methods: We studied 15 Fontan patients with fenestration. During cardiac catheterisation, haemodynamic changes in response to dobutamine infusion and atrial pacing were examined both before and after temporal occlusion of fenestration.
Results: The trends of changes in cardiac output (CO) and arterial oxygen saturation (SaO_2) by atrial pacing varied among patients. With the increase in cardiac output, fenestration predominantly provided an increased amount of CO. Because of increased CO, oxygen saturation was preserved during the dobutamine infusion. As for atrial pacing, ventricular relaxation could determine the CO response to increased heart rate (HR), irrespective of baseline pulmonary-to-systemic flow ratios (Qp/Qs) and pulmonary arterial resistance. Conversely, SaO_2 was determined by the change in Qp/Qs and change in central venous oxygen saturation. Interestingly, we observed a linear relationship between stroke volume (SV) and Qp/Qs, indicating that SV could determine Qp/Qs during HR variations in fenestrated Fontan. Furthermore, we found that patient age was an independent determinant of individual SV–Qp/Qs relationships by multiple regression analysis ($p = 0.013$).
Conclusions: The Qp/Qs can change with HR variations on the basis of the patient's specific SV–Qp/Qs relationship. Because this relationship varies with the patient's growth, and because oxygen saturation is strongly influenced by central venous oxygen saturation, fenestration showed beneficial and ideal activity, especially in younger patients. Fenestration would be active in preserving both oxygen saturation and preload corresponding to cardiac output variations, both in increasing and decreasing cardiac output.

228: PAEDIATRIC ECMO AT SMALL PAEDIATRIC CARDIAC CENTRES IN THE NORDIC COUNTRIES

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Background: Paediatric ECMO is a highly specialised treatment and experienced teams are necessary for an acceptable outcome. The five Nordic countries all have a confined background population, resulting in a restricted number of paediatric ECMO runs in each centre. In order to compare the performance of the Nordic centres with the rest of the world we compared data from a 10-year period with the ELSO register and with selected high-volume centres.
Methods: A MEDLINE and PUBMED search (2005–2012) was done to find results from larger single-centre studies. Data will be collected retrospectively and will comprise children < 19 years of age from the six Nordic centres Cumulative data from all the Nordic centres will comprise sufficient data to perform relevant statistics and evaluate the overall performance of minor ECMO centres.
Results: Mortality and secondary outcome as well as data on demographics, indications and complications will be presented from the Nordic centres. A comparison of indications, outcome and complications associated with ECMO will be performed between data from the Nordic centres and the ELSO register as well as selected single large-centre studies.
Conclusion: The analysis will provide insight into the performance of minor centres, and if specific ECMO runs or indications should be improved or referred for larger centres. Furthermore, the analysis will address whether there are certain risk factors associated with a poor outcome. As data analysis is still pending, no conclusions can be made at this point.

244: EARLY AND MID-TERM OUTCOMES OF A SUTURELESS TECHNIQUE FOR POSTOPERATIVE PULMONARY VENOUS STENOSIS

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Background: Sutureless *in situ* pericardium repair for the relief of pulmonary venous stenosis (PVS) after the repair of TAPVC was firstly reported by F Lacour-Gayet in 1996. This technique has been popular because of its potential to avoid recurrent stenosis. However, detailed clinical advantages remain unclear. We retrospectively reviewed our surgical experience and compared the outcome with that of conventional procedures.
Methods: For the relief of postoperative PVS after TAPVC repair, five patients underwent a conventional procedure such as orifice cutback or resection of a proliferated intima from 1999 to 2004 (group C, four males, median 93 days old, 3.6 kg). Thereafter seven patients with PVS were repaired with the sutureless technique from 2005 to 2011 (group S, five males, 119 days old, 3.4 kg). Patients with a single ventricle were excluded. There were no significant differences regarding patient characteristics. Follow-up examinations were completed in all patients.
Results: The cumulative survival rates at five years in groups C and S were 60 and 85.7%, respectively. Freedom from re-intervention for the pulmonary veins at five years in groups C and S were 60 and 85.7%, respectively. The rate of re-stenosis after relief of PVS were 100% (10/10) in group C and 31.6% (6/19) in group S ($p = 0.0088$).

In bilateral venous stenosis patients, survival was 0% (0/2) in group C and 83.3% (6/7) in group S ($p = 0.035$). Among three patients (one in group C and two in group S) who developed PVS in all four pulmonary veins, only one in group S survived.
Conclusion: PVS relief with a sutureless technique was superior to conventional procedures, particularly in more severe cases such as bilateral PVS or PVS in all four pulmonary veins. The sutureless technique for postoperative PVS could be applied aggressively, even at the initial repair for TAPVC, to decrease the risk of postoperative PVS.

248: CLINICAL ANALYSIS OF ORTHOSTATIC HYPERTENSION IN CHILDREN

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Objective: To study the clinical characteristics of orthostatic hypertension (OHT) in children.
Methods: A total of 96 OHT children who met the diagnostic criteria and clinical manifestations were recruited in the Department of Paediatrics, Peking University First Hospital. Age and gender distributions were noted. The duration of disease, frequencies of symptoms and the predisposing factors were recorded. The haemodynamic changes from supine to upright positions were also analysed.
Result: There were 50 boys and 46 girls in our study group. The mean age was 11.8 ± 2.7 years; 32 children were between six and 10 years old, accounting for 33.3% of all subjects, while 64 patients were from 11 to 17 years old, accounting for 66.7%. Duration of symptoms of OHT were less than one month in 22.9% children, from one month to one year in 51.1% of children and longer than one year in 26.0% children. The most common clinical manifestations were syncope and dizziness. The incidence of this were 70.8 and 46.9%, respectively. Other clinical manifestations included transitional amaurosis, nausea and/or vomiting, and pallor. These often occurred with positional changes (24.0%) and standing for long periods (57.3%). Other predisposing factors included exercise, emotional changes and environment. The baseline systolic and diastolic blood pressure was 103 ± 8 mmHg (1 mm Hg = 0.133 kPa) and 59 ± 6 mmHg, respectively. The standing systolic and diastolic blood pressure at 3 min was 113 ± 8 mmHg and 73 ± 6 mmHg and the differences were significant ($t = 27.674$, $p < 0.01$; $t = 17.936$, $p < 0.01$). The baseline heart rate in the supine position was 81 ± 11 beats/min and the maximum heart rate in the upright position was 113 ± 12 beats/min ($t = 33.092$, $p < 0.01$).
Conclusion: OHT is commonly seen in puberty in children. The main symptoms are syncope and dizziness. They were mostly induced by positional changes and long periods of standing.

260: LATE SURGICAL CORRECTION OF ANOMALOUS LEFT CORONARY ARTERY FROM PULMONARY TRUNK IN CHILDREN, USING AUTOGENOUS AORTIC AND PULMONARY ENDOTHELIALISED TUBE

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Background: Direct re-implantation of an anomalous left coronary artery into the aorta is the preferred surgical option for creating a dual coronary arterial system in patients in whom the anomalous artery originated from the pulmonary trunk.
Methods: Three patients presented with the origin of the left coronary artery from the left posterior pulmonary sinus, associated with moderated mitral valve insufficiency caused by left ventricular dysfunction. For surgical correction we used a trapdoor-like coronary artery elongation with autogenous aortic and pulmonary endothelialised tube to connect the left coronary ostium to the ascending aorta, avoiding the mitral valve intervention.
Results: There were no early or late deaths. All patients were in functional class I, with good biventricular function and a competent mitral valve at a median follow up of 94 months, ranging from 108 to 132 months. Postoperative computerised tomography of the aorta in our first patient showed good arterial flow without any distortion. A local and distal stenosis of the left pulmonary artery was observed and submitted for stent treatment.
Conclusions: The potential benefits of the trapdoor-like technique and its modifications allow for excellent operative exposure. The use of an autogenous endothelialised tube is a viable tissue capable of further growth, and this avoids injury to the aortic and pulmonary valvular apparatus or production of an obstruction within the right ventricular outflow tract.

264: CARDIAC EFFECT OF LOW- AND HIGH-DOSE IDEBENONE THERAPY IN FRIEDREICH'S ATAXIA PATIENTS

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Introduction: Cardiac involvement in Friedreich's ataxia (FRDA) is present in 63% of patients, mostly represented by hypertrophic cardiomyopathy. The use of antioxidants, such as idebenone, has shown promising results in improving cardiac hypertrophy parameters at low to intermediate doses. Higher doses of idebenone are suggested but the related long-term cardiac effects have not been studied, which was the objective of this study.
Methods: This was a prospective, non-controlled, comparative, open-label trial of a 12-month regimen of low versus high doses of idebenone between two cohorts with FRDA. Cardiac evaluation was performed before initiation of therapy, and at six, nine and 12 months of therapy.
Results: Significant left ventricular mass reduction from baseline was observed in both groups after six, nine and 12 months ($p \leq 0.05$). Systolic function parameters were within the normal range in all patients, with no significant differences between baseline and last follow up in either group. Diastolic function was marked by an improved mitral deceleration time in the high-idebenone group at the last follow up ($p = 0.029$), but not in the low-idebenone group ($p = 0.13$).
Conclusion: There were comparable effects of high- and low-idebenone therapy in terms of reduction of left ventricular hypertrophy parameters. Both therapeutic regimens seemed to preserve systolic cardiac function, with an advantage of the high-idebenone dose improving diastolic filling.

269: OUTCOMES OF EXTRACORPOREAL LIFE SUPPORT FOLLOWING CARDIAC SURGERY IN CHILDREN WHO FAIL TO WEAN FROM CARDIOPULMONARY BYPASS

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Background: Extracorporeal life support (ECLS) following paediatric cardiac surgery varies according to centre availability and philosophy. We assessed local outcome of patients receiving ECLS for failure to wean from cardiopulmonary bypass (CPB), with the aim of identifying factors associated with outcome.
Methods: Institutional databases from our tertiary referral centre identified children who received postoperative ECLS. Retrospective medical record review preceded standard statistical analyses, including factors associated with survival.
Results: Between 1 January 2002 and 1 January 2011, 65 children (median age 1.5 months) received ECLS for failure to wean from

CBP. Extracorporeal membrane oxygenation (ECMO) was used for 35 children (median one month) and centrifugal left ventricular assist device (LVAD) for 30 (median 2.8 months). Fourteen children (21%) were post-palliation for single-ventricle physiology. Eleven patients (17%) underwent surgical revision on ECLS, and 19 (29%) received multiple ECLS runs. Thirty-eight patients (58%) survived to hospital discharge. Survival was not associated with diagnosis, single-ventricle physiology, surgical revision, organ-specific complications or ECMO versus LVAD. Survival occurred after up to nine days of ECLS. From univariable analysis, older age (2.2 vs 0.2 months), lower arterial lactate at four hours (2.6 mmol/l vs 4.9 mmol/l), shorter ECLS (three vs six days), less patient or circuit complications, and single ECLS run were associated with survival (all $p < 0.05$). From multivariable regression models, prolonged ECLS ($p < 0.001$), elevated lactate four hours post-support ($p < 0.02$) and repeat ECLS ($p < 0.03$) were associated with hospital deaths.
Conclusion: Almost 60% of children receiving ECLS for failure to wean from CBP post cardiac surgery survived to hospital discharge. Inadequate support as represented by higher arterial lactate and multiple runs were associated with worse outcomes. Although prognosis worsens with prolonged ECLS, individual patients survived after up to nine days of support.

285: SYSTEMIC THROMBOLYSIS IN CHILDREN WITH LIFE- OR ORGAN-THREATENING THROMBOSIS AFTER CARDIAC SURGERY

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Background: The use of thrombolysis in the postoperative period is relatively contra-indicated because of risk for serious haemorrhagic complications. The aim of the study was to assess the efficacy and safety of thrombolysis and identify risk factors for major bleeding complications in children with intracardiac or major vessel thrombosis after cardiac surgery.
Methods: This retrospective study included children with clinically significant thrombi confirmed by sonography, angiography or CT scan, who where treated with recombinant tissue plasminogen activator (rtPA) in the postoperative period after cardiac surgery between 2000 and 2011. Data are presented as median (range).
Results: Fourteen patients at the age of 24 months (one month – 15 years) received 15 courses of systemic thrombolysis for intracardiac (six) or major vessel (nine) thrombosis. Thrombolysis was initiated on postoperative day nine (36 hours – 40 days after surgery). Duration of therapy was six hours (two hours – three days) with cumulative doses of rtPA of 2.7 mg/kg (0.3–18.2 mg/kg). Complete clot resolution, a partial effect, and no effect were achieved in nine (60%), four (26.7%), and two children (13.3%), respectively. Major bleeding required blood transfusion in five patients (33.3%) and surgical intervention in two (13.3%). One patient died of inferior vena cava thrombosis after a Fontan procedure. All-cause hospital mortality for the whole group was 35%. Higher international normalised ratio immediately after thrombolysis discontinuation was associated with haemorrhagic complications ($p = 0.01$). Other factors were not identified as predictors of outcome.
Conclusions: Thrombolysis can be used as a treatment modality for symptomatic thrombosis in children after cardiac surgery. The risk–benefit ratio should be assessed for each individual patient.

292: CONGENITAL CARDIAC SURGERY THROUGH THE MINIMALLY INVASIVE MID-AXILLARY RIGHT LATERAL MUSCLE-SPARING THORACOTOMY APPROACH IN INFANTS WEIGHTING 6 KG AND MORE

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Background: There is growing interest in minimally invasive techniques in cardiac surgery and a legitimate desire to adopt these approaches for minimal-weight and young patients. We used right lateral thoracotomy (RLT) to repair congenital heart defects (CHD) in infants weighting 6 kg and more.
Methods: Between May 2009 and June 2012, 384 patients from six months to 18 years underwent correction for CHD through RLT with the use of direct cannulation of the aorta, caval veins and cardioplegia. Of these, 59 patients were infants younger than one year (group 1). This approach was compared to median sternotomy done on 78 infants under one year (group 2).
Results: For CHD that could be approached through the right atrium (atrial septal defect, partial anomalous pulmonary venous drainage, atrial component of atrioventricular septal defect, ventricular septal defect), we operated through RLT in infants weighing at least 6 kg. Mean patient age was 0.82 ± 0.15 years (range 0.5–1.0) and 0.76 ± 0.16 years (range 0.5–1.0) in groups 1 and 2, respectively ($p > 0.05$); the degree of CHD was the same. Exposure to the intracardiac anatomy in the RLT group was good and there was no need for conversion to another approach. The mean duration of operation was 122.4 ± 32.2 min in group 1 and 142.8 ± 57.5 min in group 2 ($p < 0.05$). There was no operative or late mortality or major morbidity. The follow up was 1.7 ± 0.6 years. All patients in the RLT group had gratifying cosmetic results. There was no scoliosis or deformity of the chest or a breast.
Conclusions: RLT can be used as an alternative to median sternotomy for a wide range of CHD that could be approached through the right atrium in infants weighing at least 6 kg. RLT cosmetic results were much better than with standard median sternotomy.

293: LATERAL ATRIAL TUNNEL AND EXTRACARDIAC CONDUIT: COMPARISON OF EARLY RESULTS IN A SINGLE-CENTRE EXPERIENCE

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Background: After more than 40 years of history of Fontan operations (FO), two modifications are currently being used for cavopulmonary anastomosis: lateral atrial tunnel (LT) or extracardiac conduit (EC). The aim of the study was to compare the hospital outcomes of LT and EC at a single institution over the same period of time.
Methods: Between June 2007 and June 2012, a series of 149 consecutive children at a mean age of 3.6 ± 2.1 years underwent FO: 56 (37.6%) patients after hemi-Fontan operation underwent fenestrated LT and 91 (61.1%) patients after bidirectional Glenn anastomosis underwent EC; two (1.3%) patients underwent other variants of FO and were excluded from the study. The most common malformations were: hypoplastic left heart syndrome (55.1%), hypoplastic right heart syndrome (10.2%), double-inlet left ventricle (10.2%) and double-outlet right ventricle with hypoplastic left ventricle (8.8%). Haemodynamic, electrocardiographic and clinical peri-operative data were retrospectively analysed.
Results: The hospital mortality was 0%. There were no differences between the groups regarding age, weight, morphology of the single ventricle, pre-operative cardiac catheterisation values and postoperative intubation time (15.4 ± 28.2 vs 11.5 ± 17.6 h; $p = 0.313$). Children after EC tended to stay longer in hospital (18.4 ± 9.6 vs 15.5 ± 8.2 days; $p = 0.061$) and had significantly longer right pleural drainage for effusions (9.2 ± 7.1 vs 5.9 ± 5.0 days; $p < 0.01$). Patients after LT had more frequent junctional or ectopic atrial rhythm on the day of the operation ($p < 0.01$) and at discharge ($p = 0.016$).
Conclusions: Fenestrated lateral atrial tunnel seems to facilitate the early adaptation to Fontan physiology but in our experience this operative technique caused higher incidence of atrial rhythm disturbances, which can influence the late functional status of the patients.

320: MID-TERM OUTCOME OF EXTRACARDIAC FONTAN OPERATION USING CONTEGRA CONDUIT

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Background: Reports have shown increased risk of thrombotic occlusion of Fontan circulation with the use of a Contegra conduit. We intended to retrospectively compare the outcome of Fontan completed using a Contegra conduit with those using a Dacron tube.
Methods: Medical records, echocardiograms and catheterisation data of all patients undergoing Fontan completion from 2002 to 2010 were reviewed. The outcome of the Contegra group was compared with those of the Dacron tube group. All patients were anticoagulated using heparin in the immediate postoperative period and later with coumadin to maintain therapeutic INR. The primary outcome was the prevalence of thrombotic complications and survival in the two groups. Chi-square analysis was used to compare the categorical variables. Independent two samples *t*-test was used to compare the pre- and post-operative variables. Log-rank test was performed and Kaplan-Meier curves were generated to compare primary outcomes in the two groups.
Results: Seventy-six patients underwent the Fontan procedure, with Contegra conduit ($n = 47$) and Dacron tube ($n = 29$). The two groups were matched with regard to demographic variables, pre-operative haemodynamic data, intra-operative and postoperative outcomes. Within 30 days, thrombotic complications occurred in 6/47 (13%) in the Contegra and 3/29 (10%) in the Dacron groups ($p = 0.983$). Relative risk of thrombosis in the Contegra group was 0.949 (95% CI = 0.8–1.3). The mean follow up for the whole group was 87 months (Contegra = 70, Dacron = 95) ($p = 0.304$). Nine patients died: 7/47 in the Contegra and 2/28 in the Dacron groups ($p = 0.486$). The relative risk of dying in the Contegra group was 0.909 (95% CI = 0.8–1.1).
Conclusion: This is so far the largest series evaluating the outcome of extra-cardiac Fontan procedure using a Contegra conduit. Our results suggest that using a Contegra conduit for Fontan completion does not increase the risk of thrombotic complications or death compared to a Dacron tube.

335: PLASMA EXCHANGE FOR CARDIOGENIC SHOCK IN DILATED CARDIOMYOPATHY

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Background: Auto-immunity is suggested as one of the causes of dilated cardiomyopathy (DCM). The sera of many patients with DCM are positive for several antibodies directed against cardiac antigens. These antibodies play a role in the pathophysiology of cardiac dysfunction. We performed slow plasma exchange plus continuous haemodiafiltration (SPE+CHDF) to eliminate these antibodies for a DCM patient with cardiogenic shock, using extracorporeal life support (ECLS).
Case presentation: He was diagnosed with idiopathic DCM at two years of age. His heart failure became gradually worse. He began oral administration of carvedilol from six years of age, however, the heart failure worsened (NYHA III). Left ventricular ejection fraction (LVEF) was reduced to 28%. At the age of 13 years, he rapidly deteriorated and developed multiple organ failure. His antibeta-1 adrenergic receptor antibody titre and anti-muscarinic M2 receptor antibody titre were 80 times the background density on enzyme-linked immunosorbent assay. We planned to register for heart transplantation and performed ECLS using a combination of SPE+CHDF in order to rescue him. Daily PE with CHDF was performed for two days. PE was performed over eight hours, using 1.2 times the circulating plasma volume of fresh frozen plasma. After SPE+CHDF, his blood pressure and LVEF were dramatically improved. He could discontinue catecholamine infusion and end the ECLS in three days. He had no complications during SPE+CHDF.
Conclusion: A patient with DCM using ECLS for cardiogenic shock could be weaned from the ECLS by performing SPE+CHDF. In patients with DCM, SPE+CHDF treatment for elimination of antimyocardial antibody is very effective and useful for improving cardiac function. This therapy is a new strategy for helping patients recover from heart failure in DCM.

339: A 20-YEAR COMPARISON OF SIMPLE AND COMPLEX TAPVR

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Background: Total anomalous pulmonary venous return (TAPVR) frequently requires neonatal surgery. Two of the largest determinants of surgical timing and mortality in TAPVR patients are other complex cardiac lesions and/or pre- or postoperative pulmonary vein obstruction (PVO). Simple TAPVR refers to TAPVR associated with an ASD and/or PDA, and complex TAPVR refers to TAPVR associated with other complex cardiac lesions. In this study we compared our outcomes with simple and complex TAPVR with a focus on the influence of pulmonary vein obstruction on the outcomes.
Methods: Since 1966, 216 children have undergone simple and complex TAPVR repair at our institution. The first 105 were previously reported. This study reviews the most recent 111 patients from 1990 to 2011. The mean age was 5.2 months, ranging from one day to 16 years. Sixty-one of the patients had simple TAPVR and 50 (45%) had complex TAPVR.
Results: Early and late mortality was at 1.6 and 6.6%, respectively, in the simple TAPVR group and 14 and 18%, respectively, in the complex TAPVR patients. Pre-operative PVO was more frequent in the complex than in the simple TAPVR group (36 vs 23%). Re-intervention rate for postoperative PVO was also higher in the complex than in the simple TAPVR group (12 vs 7%). Pre-operative PVO occurred at a higher rate in mixed-type TAPVR patients and was lowest in cardiac-type TAPVR patients. Re-intervention for post-operative PVO was highest for the infracardiac TAPVR group and lowest in the supracardiac TAPVR group. Other risk factors for poor outcomes were low birth weights, young age, need for pre-operative ECMO, and single-ventricle physiology.
Conclusions: Outcomes for simple TAPVR were quite favorable. Complex TAPVR with and without pulmonary vein obstruction remains a vexing problem. Improving the management of PVO is the key to improving outcomes.

348: A COMPARISON OF BLALOCK-TAUSSIG SHUNTS WITH AND WITHOUT CLOSURE OF THE DUCTUS ARTERIOSUS IN NEONATES WITH PULMONARY ATRESIA AND INTACT VENTRICULAR SEPTUM

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Background: Neonates with pulmonary atresia and intact ventricular septum (PA IVS) who have a right ventricle that is deemed not suitable for decompression undergo single-ventricle palliation. Early survival after the modified Blalock-Taussig shunt (MBTs) for these infants is disproportionately low compared with other lesions. The aim of this report was to compare the results of closure versus non-closure of the patent ductus arteriosus (PDA) during MBTs surgery in neonates with PA IVS.
Methods: This retrospective study included neonates with PA IVS who underwent a single-ventricle pathway with primary MBTs through a sternotomy approach at a single institution between January 2000 and May 2012. Postoperative hospital mortality, need

for early re-intervention, time to extubation, maximum vasoactive inotropic score and length of hospital stay were studied as primary outcomes.
Results: Twenty neonates (median age five days; range 3–14 days) with PA IVS underwent a MBTs procedure (shunt size 3–3.5 mm). The PDA was closed surgically in 10 patients and left open in 10. Compared with patients in whom the PDA was left open, neonates with surgically closed PDA had a higher operative mortality (40 vs 0%, $p = 0.02$). A trend toward a higher vasoactive inotropic score in the group with a closed PDA was observed (17 vs 10.2, $p = 0.08$). The need for re-intervention and length of hospital stay did not differ between the two groups ($p = 0.63$ and $p = 0.59$, respectively). Higher diastolic arterial pressures and lower arterial oxygen saturation to fraction of inspired oxygen ratio ($SatO_2/FiO_2$) were observed in the group with a closed PDA during the first 24 postoperative hours.
Conclusions: In our limited retrospective cohort, PDA closure during MBTs in newborns with PA IVS was associated with increased hospital mortality.

360: MORBIDITY AND MORTALITY IN PAEDIATRIC HEART TRANSPLANTS: A 25-YEAR SINGLE-CENTRE EXPERIENCE

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Background: The outcomes of paediatric cardiac transplantation continue to improve. We retrospectively reviewed our outcomes over the last 25 years to determine the risk factors for poor outcomes in our patient population.
Methods: Since 1985, 126 heart transplants in 120 patients (69 males) have been performed. Median age at transplantation was 3.6 years (range: four days to 17.8 years). The primary indications for transplantation included CHD ($n = 61$), cardiomyopathy ($n = 58$), and re-transplantation ($n = 7$). Pre-operatively, 40% ($n = 51$) had previous surgical interventions. Pre-, peri- and postoperative data were analysed to identify risk factors.
Results: Early and late mortality were 7% ($n = 9$) and 39% ($n = 49$), respectively. Actuarial survival at one, five, 10 and 20 years was 86, 73, 62 and 46%, respectively. The diagnosis of CHD and transplantation prior to the year 2000 were independent risk factors for early mortality. At median follow up of 76.7 months (range 0–294 months), 17% ($n = 22$) required further cardiac intervention, including the 5% ($n = 6$) who needed subsequent re-transplantation. There were no risk factors associated with late mortality or need for further surgical intervention.
Conclusions: Paediatric patients transplanted for CHD and those transplanted prior to the year 2000 were independent risk factors for early mortality. Further review of the specific CHD diagnoses and immunosuppression management protocols will be compared to see if they affected outcomes.

387: MANAGEMENT OF WOUND INFECTIONS POST CARDIAC SURGERY IN PAEDIATRICS

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Background: The spectrum of sternal wound infections after cardiac surgery ranges from superficial infections to a deep sternal infection known as mediastinitis. Mediastinitis is an uncommon and clinically relevant source of postoperative morbidity and mortality in paediatric patients after cardiac surgery.
Methods: A retrospective observational study was carried out in our paediatric intensive care unit, in which more than 800 patients post cardiac surgery were assessed for risks of sternal wound infection (SWI). We identified all patients diagnosed with mediastinitis after cardiac surgery from January 2009 to June 2012. Staging of wound care using a standard protocol of antibiotics and selected dressings was done.
Results: Major risk for sternal wound infection was associated with delayed sternal closure. Chest wound infection developed in 40 of 800 (5%) children after median sternotomy or lateral thoracotomy. Superficial wound infection developed in 30 (3.751%) children and 10 (1.25%) had deep infection. Children with sternal wound infection were younger, had delayed sternal closure, longer periods of ventilation and inotropic support, and longer intensive care unit and total postoperative hospital stay. Staging of wound care proved effective and successful.
Conclusion: Infections continue to be a significant cause of morbidity in paediatric cardiac surgery patients. Knowledge of risk factors for infection could be useful in preventative and treatment strategies for these high-risk groups. Paediatric strategies differ from adult programmes. Standardised protocol, timely diagnosis, timely wound debridement and liberal use of specific antibiotic mixes with selected dressings can potentially minimise the morbidity and mortality rates in paediatric postoperative cardiac patients

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390: EVALUATION OF TWO-STAGE ARTERIAL SWITCH AS A TREATMENT STRATEGY IN THE MANAGEMENT OF DELAYED PRESENTATION OF TRANSPOSITION OF THE GREAT ARTERIES WITH A REGRESSED LEFT VENTRICLE

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Background: Management of transposition of the great arteries with intact ventricular septum (TGA/IVS) is currently an arterial-switch operation (ASO) performed in the first two weeks of life. Two-stage ASO is one form of treatment in infants with TGA presenting late.
Methods: From December 2009 to date, a total of eight patients with TGAIVS presented late and were not deemed suitable for immediate ASO due to left ventricular regression, and were selected for a two-stage ASO. Serial echocardiography was used to assess the increased thickness of the left ventricular (LV) posterior wall. A stage II ASO was done a few weeks later. A retrospective review of patient charts was done. Effects of variables such as age, BSA and time interval between two procedures on mortality rate were analysed. Data were formulated into a structured database, and statistical analyses were performed with the statistical package SPSS for Windows.
Results: Eight patients underwent stage I, which had an in-hospital mortality of 12.5% (1/8), while the interval mortality between both stages was also 12.5% (1/8). Two patients died from non-cardiac complications due to aspiration-related pulmonary sepsis. One patient had a failed stage I (12.5%) due to acute LV failure and had to undergo a Senning atrial switch as a salvage operation. One patient is currently waiting for stage II definitive repair after a successful stage I. The mean interval between the two stages was 3 ± 1 weeks. Four patients have undergone a successful stage II ASO with zero mortality. All patients had remarkably rapid recoveries and short hospital (6 ± 2 days) stay.
Conclusion: Early experience indicates that in a developing country such as Pakistan, a rapid two-stage arterial switch is an acceptable treatment option. Patients who survived stage I and the intervening period had excellent results with stage II.

397: TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION: MANAGEMENT AND OUTCOME, EXPERIENCE FROM CHILDREN'S HOSPITAL, LAHORE, PAKISTAN

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Background: We report on a retrospective analysis of the demographic, morphological and clinical profiles of patients along with results of operative repair for total anomalous pulmonary venous connection.
Methods: In the last five years, 65 patients (45 males, 20 females) underwent repair for total anomalous pulmonary venous connection. Ages ranged from two days to nine years (median six, mean 17 months) and 46 were under one year old. Weight ranged from 2.4 to 18 kg (median 5 kg). The anomalous connection was supracardiac in 35 (54%), cardiac in 18 (27%), infracardiac in seven (10%) and mixed in five (8%) patients. Twenty-five (38%) patients had obstructed drainage and 40 (61%) had moderate or severe pulmonary arterial hypertension; 21 patients (32%) had to be operated on as an emergency.
Results: Mortality was more in obstructed drainage patients (26%) compared with patients without obstruction (8%).The major causes of early death were weight < 10% (OR 1.1; 95% CI: 0.1–6.5, p = 0.009), obstruction (OR 9.8; 95% CI: 1.6–60, p = 0.006) and sepsis (OR 23.3; 95% CI: 3–177, p = 0.002). Follow up ranged from one to 45 months (median 24 months). There was one re-operation due to late pulmonary vein stenosis but the patient later died.
Conclusions: In a developing country such as Pakistan, mortality continues to be high in infants with total anomalous pulmonary venous connection. Weight < 10th percentile, obstruction, severe pulmonary arterial hypertension and postoperative sepsis appear to be the most important predictors of operative mortality.

408: MORTALITY OUTCOMES UP TO ONE YEAR FOLLOWING PAEDIATRIC CARDIAC SURGERY IN WESTERN AUSTRALIA (2001–2010)

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Background: Paediatric cardiac surgical outcomes have been traditionally described as 30-day or in-hospital mortality. There are limited reports of late deaths up to one year after surgery. This study aimed to determine the mortality outcomes for cardiac surgeries performed on patients from western Australia (WA) from 1 January 2001 to 31 December 2010 up to one year after the procedures, and to ascertain possible causes.
Methods: Data were obtained and cross-linked from the Department's customised surgical and clinical databases, and medical records were reviewed. Inclusion criteria: paediatric cardiac surgical cases operated on between 1 January 2001 and 31 December 2010 in WA and interstate; and deaths occurring up to one year post surgery. Thoracic, non-cardiac surgeries and pre-term neonates with PDA ligation were excluded.
Results: A total of 1 198 cardiac surgical procedures were performed on WA children, of which 128 surgeries were performed interstate; 30-day (early) and one-year (> 30 < 365 day/late) mortality rates were 2.3 and 1.4%, respectively. In-hospital mortality was 2.6%. The cause of the majority of late deaths (10/17) was cardiac but non-surgical related, with pulmonary hypertension and pulmonary venous obstruction being common. Two/17 late deaths appeared directly related to cardiac surgery. The remainder (5/17) were unrelated to the cardiac problem or unknown. Other high-risk factors included indigenous children living in remote locations, and trisomy 21 following AVSD repair.
Conclusions: The overall mortality rate at one year following all paediatric cardiac surgeries between 2001 and 2010 on WA children was 3.7%. The 30-day mortality rate was 2.3%. Late mortality up to one year added 1.4%. Some of the high-risk factors identified for late deaths are being approached by specific changes in management and follow up. Parents should be informed regarding the potential for ongoing risk of mortality in high-risk situations.

411: 29 YEARS OF FOLLOW UP IN THE DAMUS-KAYE-STANSEL PROCEDURE: ANASTOMOSIS REVISION AND NEED FOR VALVE REPAIR

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Background: While the Damus-Kaye-Stansel (DKS) procedure has been used for complex congenital heart disease and systemic outflow tract obstruction, limited data are published on late outcomes. This study examined the incidence of surgical intervention in patients with more than one year of follow up after DKS over 29 years.
Methods: This was a retrospective study on 54 patients who underwent DKS (not including Norwood) from 1983 to 2007. Patients were excluded if there was no follow up beyond one year postoperatively (three), or if death occurred within one year of surgery (14). Survival was calculated from the date of DKS to the last known follow up or time of death from 1984 to 2012.
Results: Thirty-seven patients met the study criteria, with a median follow up of 12.9 years (2–28 years). Diagnoses included double-inlet ventricle (16), double-outlet right ventricle variants (12), tricuspid atresia (six), and others (three). Seven patients (18.9%) experienced death over one year after DKS (4.6–24.2 years). Five deaths were sudden at ages 13 to 30 years. One death occurred due to sepsis complicating a MRSA-infected pseudoaneurysm of DKS. One death occurred awaiting transplant. The mean number of cardiac surgeries was three, median 3.5 for the 37 patients reviewed; 28 patiens (75.7%) obtained Fontan status. Eight patients (21.6%) required DKS anastomosis revision due to aneurysmal DKS connections in four patients and outflow tract stenosis in four; 15 patients (40.5%) required repair/replacement of a semilunar valve.
Conclusions: Our review demonstrates a high incidence of aneurysmal DKS connections, stenosis of the systemic outflow tract, valvular insufficiency and sudden death. These patients warrant close long-term surveillance with imaging, arrhythmia and heart-failure assessments.

443: HEART TRANSPLANTATION IN PATIENTS WITH SINGLE VENTRICLE: A SINGLE-CENTRE EXPERIENCE

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Background: In the 1990s, heart transplantation was a common first-line therapeutic option for many single-ventricle congenital heart diseases. The aim of this study was to present our centre's experience with heart transplantation in these patients.
Methods: A retrospective review of 105 children and adult patients with congenital heart malformation who underwent cardiac transplantation between 1988 and 2012 revealed 22 patients who were transplanted for a single right (n = 18) or left (n = 4) ventricle. Group 1 (n = 13) had not been operated on before transplantation or undergone preliminary palliative surgery. Group 2 (n = 9) had undergone a partial (n = 3) or total (n = 6) cavopulmonary anastomosis. Median age at transplantation was 0.2 years (5 days to 21.2 years) in group 1 and 15.2 years (2.4 to 34.6 years) in group 2.
Results: The median follow up in the study series was 8.3 years (3.5 to 18.1 years) and was complete in 100%. The overall mortality rate was 45% (n = 10) with a hospital mortality rate of 27% (n = 6). Three patients had to be re-transplanted. The five-year survival rates after transplantation were 69% in group 1 and 53% in group 2. Kaplan-Meier analysis showed no significant difference between the two groups in the log-rank test (p = 0.39) and an overall median survival of 11 years. For comparison, the five-year survival rate of all 105 patients was 82%.

Conclusions: The outcome after transplantation for patients with or without previous cavopulmonary anastomosis is comparable. Taking into account the higher median age at transplantation in the group with cavopulmonary anastomosis regarding life expectancy, it seems to be reasonable to do the palliative surgery instead of early transplantation.

446: OPERATING ROOM SAFETY IN DEVELOPING COUNTRIES: THE IMPORTANCE OF LANGUAGE-INDEPENDENT DIAGRAMS, CHECKLIST AND TIME-OUT

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Background: In July 2009, the International Children's Heart Foundation piloted a quality-improvement initiative implementing operation room time-out as a standard on all medical mission trips. Through this process, safety initiatives, standardised teaching and process improvement initiatives have been developed. We describe a qualitative evaluation of a multi-faceted OR safety and education initiative in a multi-national assistance programme.
Methods: Intra-operative nursing checklists were introduced in addition to the time-out process. OR volunteers were asked to note OR complications in a database sheet. Language-independent diagnostic documentation (heart diagrams) followed patients from pre-operative cardiology to the post-operative phase and were utilised in the OR as part of pre-operative time-out and for teaching.
Results: Time-out was performed on 86% of trips in 2012. Complications found and dealt with during pre-surgical time-out included: suboptimal antibiotic timing, blood availability, wrong procedure prepared for by local staff, wrong patient brought to OR, implant not on field, no definitive diagnosis/or understanding prior to incision, change of plan based on an in-OR echo review, and equipment issues/availability. Language-independent diagrams ensured the entire team agreed with and understood the plan of care. Although intended to ensure diagnostic echo accuracy and the right patent in the OR, the diagrams were also used in staff and family education.
Conclusions: Checklists are proven methods of improving patient safety. Compliance of performing and tracking time-out is greater where an ICHF OR staff member is present at the time of incision. Issues that arose were recognised and fixed in a timely fashion, usually due to the implementation of a 'standard' process prior to incision. Heart diagrams are an indispensible language-independent resource and aid in patient care when language barriers exist, for the safety of diagnosis and understanding.

447: CHALLENGES IN DIAGNOSIS AND MANAGEMENT OF PULMONARY ARTERIOVENOUS MALFORMATIONS IN A RESOURCE-POOR SETTING: A CASE REPORT

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Background: Pulmonary arterio-venous malformations (PAVMs) are rare vascular anomalies resulting in abnormal direct communication between pulmonary arteries and veins. Most PAVMs are congenital but acquired cases also occur. PAVMs result in right-to-left shunts and are an unusual cause of chronic cyanosis with consequent polycythemia. The direct connection between arteries and veins causes impairment of the normal filtering function of the lungs, with potential paradoxical embolism and systemic infections.
Methods: A young adult male with PAVMs and complications highlights the challenges in diagnosis and management in a resource-poor setting. His report of congenital cardiovascular malformations is presented.
Result: PC, an 18-year-old male has been on evaluation and management for polycythemia from chronic cyanosis since the age of eight years. Cyanosis was variable and noticed by his mother from the age of five years, for which she sought no treatment until he presented at seven years with multiple chronic discharging fistulae of the left thigh. He was treated for chronic bacterial osteomyelitis with some relief, but defaulted on follow up. Five years later (now aged 12 years), he was rushed to the emergency unit with a day's history of recurrent convulsions culminating in coma. CT scans showed left frontal lobe abscess suggestive of tuberculous abscess, CXR revealed vague opacities in the right middle lobe. He was commenced on anti-tuberculous therapy, regained consciousness and the fistulae healed. However due to persisting cyanosis and polycythemia, a chest MRI was done and revealed an abnormal connection between the right pulmonary artery and pulmonary veins of the right middle lobe. An interventional pulmonary angiogram could not be done due to lack of skills and facilities. His need for further intervention where facilities exist was hampered by financial constraints.
Conclusion: PAVMs with complications are challenging to diagnose and manage in resource-poor settings, resulting in chronic morbidity. Provision of skilled manpower and facilities in these centres is imperative to avert mortalities.

471: MENTORING PAEDIATRIC CARDIAC SURGERY IN DEVELOPING COUNTRIES: FIVE-YEAR PROGRESS REPORT AND EXIT STRATEGY

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Background: Establishment of stable cardiac surgical centres in developing countries includes training of personnel in the established foreign centres, visits of mission teams to do multispeciality on-site training, and donation of equipment and supplies. This usually suffices to form a sustainable programme, but some countries require further assistance. We report the results of a five-year mentoring project in one such country.
Methods: Following a 'scouting' trip with a small medical team, a surgeon relocated to work full-time with the local surgeon, mentoring him in surgical techniques, patient evaluation and care, programme administration, and working closely with other team members and with the local fund-raising organisation. After 4.5 years, a transition period was initiated in which the visiting surgeon began to spend progressively increasing amounts of time out of the country, leaving the programme in the hands of the local surgeon. The local database was examined for RACHS complexity, primary and assistant surgeons, mortality and fundraising. Statistical significance was defined as $p < 0.05$.
Results: From January 2007 to June 2012, 282 operations were performed with 21 deaths (mortality 7.4%). Case complexity increased over time (RACHS 2/3 cases 44% in 2011 vs 22% in 2007, $p < 0.001$). Overall mortality decreased to < 7.5% per year after the first year ($p < 0.01$). The number of operations performed by the local surgeon steadily increased ($p < 0.001$); all 2012 operations were done by the local surgeon. Local fund raising increased progressively from $10 909 in 2006 to $97 554 in 2010 ($p < 0.01$).
Conclusions: Placement of a 'mentoring' surgeon full time in a developing country can effectively promote the safe establishment of a sustainable congenital heart surgery programme when other methods of aid have failed. Good surgical results can be obtained and maintained while simultaneously increasing the case complexity and experience of the local surgeon.

475: INFLUENCE OF PULMONARY ARTERY SIZE ON EARLY OUTCOME AFTER THE FONTAN OPERATION

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Objective: Since Fontan stated his criteria for patient selection undergoing the total cavopulmonary anastomosis, small pulmonary arteries (PAs) are sometimes considered a contraindication to the operation. The aim of this study was to evaluate whether the size of the PAs is still one of the major impact factors on early outcome after the Fontan operation (FO).
Methods: Data of 146 patients (mean age of 3.6 ± 2.4 years, mean weight of 14.3 ± 6.9 kg) who underwent a modified FO at our clinic between 2007 and 2012 were retrospectively analysed with regard to the traditional McGoon ratio, Nakata index and modified indices (measuring the narrowest diameters) and with regard to the early postoperative course.
Results: Patients with a McGoon ratio ≤ 1.6 (modified ≤ 1.2) or a Nakata index < 150 mm²/m² (modified < 100 mm²/m²) were not at a higher risk of prolonged hospital stay [$p = 0.078$ (0.157) and $p = 0.220$ (0.178), respectively] or effusions [$p = 0.323$ (0.723) and $p = 0.289$ (0.703), respectively]. Children with persistent (> 14 days) effusions tended to have smaller PAs in comparison with other patients, but McGoon ratio and Nakata index did not differ significantly ($p = 0.220$ and $p = 0.069$, respectively). The need for interventional dilatation before FO did not adversely influence the time of mechanical ventilation ($p = 0.652$), ICU ($p = 0.778$) or hospital stay ($p = 0.130$) and pleural effusions ($p = 0.166$). Younger and smaller children tended to have smaller PAs, but younger age (< 24 months) and lower weight (< 12 kg) were not predictive of poor early postoperative outcome.
Conclusion: Small pulmonary arteries did not significantly affect the early postoperative period after FO. In our opinion there is no need to postpone the Fontan operation due to 'smaller' pulmonary arteries. The pre-Fontan palliative procedures to augment the size of PAs at the expense of ventricular overload are not recommended.

476: OUTCOMES OF INFLOW-OCCLUSION OPEN PULMONARY VALVULOTOMY PLUS CENTRAL SHUNT IN PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

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Background: Patients with pulmonary atresia with intact ventricular septum (PA-IVS) and tripartite right ventricle (RV) have a great opportunity to live with biventricular circulation. Direct-vision open pulmonary valvulotomy under inflow occlusion is safe and provides good pulmonary valve opening.
Methods: Between August 1999 and September 2010, 18 patients with PA-IVS underwent inflow-occlusion open pulmonary valvulotomy with a concomitant central shunt. All had tripartite RV with a tricuspid valve *Z*-score of –1.17 ± 0.99 (–0.08 to –3.5). The mean inflow occlusion time was 2.5 ± 0.4 minutes (1.6–3.0 minutes).
Results: The primary operation was successful in all patients. There was only one (5.6%) in-hospital death. At a median follow up of five years (2–13 years), survival was 100%. Of the remaining 17 survivors, 16 (94.12%) patients achieved biventricular circulation and one (5.88%) patient survived with one-and-a-half circulation. There were five (29.4%) survivors who needed percutaneous balloon pulmonary valvulotomy and all had good results. Four (23.5%) of the survivors needed surgical right ventricular outflow tract (RVOT) reconstruction. Bidirectional Glenn was performed concomitant with RVOT reconstruction in one patient.
Conclusions: Inflow-occlusion open pulmonary valvulotomy plus central shunt is a safe and precise opening of the pulmonary valve in PA-IVS patients with tripartite RV. Almost all patients achieved biventricular circulation with this technique.

487: CARDIAC STROKE VOLUME AND SYMPATHETIC–PARASYMPATHETIC MEASUREMENTS INCREASE THE SENSITIVITY AND SPECIFICITY OF TILT-TABLE TESTS (HUTT) IN CHILDREN AND ADOLESCENTS

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The head-up tilt-table test (HUTT) is the gold standard in evaluating autonomic dysfunction and syncope in children and adolescents. Limitations of the conventional HUTT, cycling blood pressure (BP) every one to two minutes, with heart rate (HR) correlated with patient symptoms results in low sensitivity and specificity. Investigators have evaluated more reliable and sensitive physiological parameters to increase the predictability of HUTT.
Methods: From May 2009 to May 2012 we performed 422 HUTT evaluations on children and adolescents. The first group of 152 patients had conventional HUTT, including HR, arm cuff BP and oxygen saturation recorded every minute for 10 minutes while supine, for 30 minutes while head up 70° and for 10 minutes with supine reposition while recording patient symptoms. The second group included 270 patients with HUTT using Task Force Monitor® with display and storage of continuous BP, HR and cardiac stroke volume (SV) by trans-thoracic impedance and calculated sympathetic and parasympathetic activity correlated with symptoms and signs. Median ages were 12.5 years and 13.2 years in groups one and two, respectively. Patients from both groups were referred by paediatric neurologists, cardiologists, gastroenterologists and rheumatologists with syncope (63%), dizziness (88%), light headedness and headaches (52%), chronic nausea and stomach pains (32%), chronic fatigue (42%), convulsions (6%), fibromyalgia (2%), palpitations and chest tightness (12%) and metabolic disorders (10%).
Results: A positive test was defined in group one as severe symptoms of syncope, blackout, vomiting, severe headache, excessive fatigue and tremors or convulsions accompanied by changes in HR (tachycardia, bradycardia) and/or blood pressure. In group two, similar symptoms were accompanied by significant changes in HR, BP, cardiac SV and sympathetic/parasympathetic activity. There was increased ability to correlate clinical manifestations with physiological abnormalities on HUTT in the second cohort of subjects and also an increased sensitivity of the test to determine whether there was orthostatic intolerance.

493: ULTRA-FAST TRACK ANAESTHESIA WITH EARLY EXTUBATION IN RESOURCE-LIMITED SETTINGS: RESULTS OF A LARGE INTERNATIONAL COHORT

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Background: Very early extubation [in the operating room (OR) or on ICU admission] has been routinely practiced by our group in countries with delayed access to cardiac care, older presentation, and limited resources. We describe the ventilation data for a sequential cohort of 2 300 children in a programme spanning 19 countries and 26 centres over 4.5 years.
Methods: The database of the International Children's Heart Foundation was analysed for the period January 2008 to May 2012. Outcomes of interest were ventilation times, re-intubation rates, mortality, by RACHS-1 complexity category, and age.
Results: Deaths in OR or ICU without extubation (64), incomplete data (190) and re-operation on same admission (217) were all excluded; 1 829 extubations were analysed with a median age/weight of 3.5 years/12 kg. Re-intubation rate and mortality decreased with decreasing duration of postoperative tracheal intubation and mechanical ventilation ($p = 0.005$, $R^2 = 0.89$, and $p = 0.002$, $R^2 = 0.93$, respectively). Median ventilation time was 1.5 hours, with

1 363 (73%) extubations in under four hours. Extubation in under one hour was possible in 42% overall, and by RACHS (R) category: R1 (70%), R2 (42%), R3 (29%) and R4 (19%). Median ventilation time increased with increasing RACHS category ($p < 0.05$): R1 ($n = 327$) 0.1 hour, R2 ($n = 827$) 1.5 hours, R3 (n = 466) 3 hours, R4 (n = 114) 5.9 hours. Ventilation times (re-intubation rates) were increased in the smallest children: < three months, 12 hours (12%); three months to one year, four hours (14%).
Conclusions: This is the largest such series analysed in children. Age and weight were typical of many developing world populations awaiting surgery. Mechanical ventilation need not be a standard part of postoperative cardiac surgical management; significant numbers in all age and RACHS groups can be safely extubated in OR or under one hour postoperatively in ICU.

506: IMPACT OF VSD ENLARGEMENT WITH D-TGA OR DORV FOR POSTOPERATIVE CARDIAC FUNCTION AND ARRHYTHMIA IN RASTELLI PROCEDURE

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Introduction: Restrictive VSD with TGA or DORV may cause LVOTO in the late period, and simultaneous VSD enlargement is mandatory in that case. However it may cause cardiac dysfunction and arrhythmia. We separated our patients who underwent Rastelli procedure into two groups; group A with VSD enlargement (46 cases) and group B without enlargement (28 cases). We evaluated the long-term outcomes.
Methods: Since 1983, Rastelli procedures have been performed in 74 patients: d-TGA (56 cases) or DORV (18 cases) in our institute; mean age at operation 7.0 years, mean body weight at operation 18.5 kg, mean follow-up period 13.7 years. Indication of VSD enlargement in our institute: existence of pressure gradient between the two ventricles, the VSD diameter below the diameter of aortic valve. Rhythms were assessed with ECG and Holter. Pacemaker implantaion or medication for anti-arrhythmia was searched. Postoperative LVEF, CVP and pressure gradient at intraventricular conduit were assessed in catheterisation.
Results: There were no early deaths and five late deaths in group A and two in B. Replacement of intraventricular conduit was performed in two cases in group A and two in B. With ECG, 42 cases in group A and 26 in B kept sinus rhythm. No significant PVC was recognised in either group. Pacemaker implantation was performed in three cases in group A and one in B. Need for anti-arrhythmic agency were two cases in group A and two in B. Mean LVEDV was 150.6% of *n* in group A and 144.8% of *n* in B. Mean LVEF were 57.0% in group A and 57.9% in B. There were statistically significant differences between the two groups.
Conclusion: VSD enlargement with the Rastelli procedure was performed safely without cardiac dysfunction and arrhythmia. VSD enlargement should be performed if necessary to prevent LVOTO in the late period.

513: CONGENITAL CARDIAC SURGERY THROUGH A MINIMALLY INVASIVE MID-AXILLARY RIGHT LATERAL MUSCLE-SPARING THORACOTOMY APPROACH: A THREE-YEAR EXPERIENCE IN 384 PATIENTS

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Background: Median sternotomy has been the conventional approach for correction of congenital heart defects (CHD) despite poor cosmetic results. Minimally invasive mid-axillary muscle-sparing right lateral thoracotomy (RLT) was assessed as an alternative procedure with better a cosmetic outcome.
Methods: Between May 2009 and June 2012, 384 patients aged from six month to 18 years underwent correction of CHD through RLT with the use of direct cannulation aorta, caval veins and cardioplegia (group 1). This approach was compared to median sternotomy on 135 patients (group 2) from the position of exposure to the intra-cardiac anatomy, postoperative period and cosmetic results.
Results: CHD that could be approached through the right atrium (atrial septal defect, partial anomalous pulmonary venous drainage, atrial component of atrioventricular septal defect, ventricular septal defect, mitral or tricuspid valve regurgitation) were operated on through RLT. Mean patient age was 6.0 ± 5.1 years (range 0.5–18) and 3.1 ± 6.3 years (range 0.2–18) in groups 1 and 2 ($p < 0.05$) and the degree of CHD was the same. Exposure to the intracardiac anatomy in the RLT group was good. There was no need for conversion to another approach. The mean operative time was 132.3 ± 36.2 min in group 1 and 151.4 ± 52.7 min in group 2 ($p < 0.05$), mean cardiopulmonary bypass time was 49.7 ± 27.3 and 58.5 ± 28.7 minutes ($p < 0.05$), respectively, and mean aortic cross-clamping time was 32.4 ± 19.1 and 36.7 ± 21.4 minutes, respectively ($p > 0.05$). The advantage of RLT included absence of any pericardial effusion. The follow up was 1.8 ± 0.8 years. Cosmetic results of RLT were good in all patients. There was no scoliosis, deformity of the chest or breast.
Conclusions: RLT is a safe and effective alternative to median sternotomy for correction of CHD that could be approached through the right atrium. Its cosmetic results were highly satisfactory.

524: ASSESSMENT OF RECOVERY IN CHILDREN ON BERLIN HEART EXCOR VENTRICULAR-ASSIST DEVICE SUPPORT

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Background: Ventricular assist device (VAD) support is increasingly used in paediatrics, mainly as a bridge to transplant, although use as a bridge to recovery is increasingly described. In 2007 we developed a protocol to assess recovery of ventricular function in children on Berlin Heart Excor® VAD support. We aimed to review the effectiveness of this protocol in assessing which patients could have the VAD safely removed.
Methods: Patients with myocarditis were initially assessed after two weeks of support, and with cardiomyopathy after four weeks. Testing involved echocardiographic and haemodynamic assessment over 90 minutes with the VAD paused, and measurements were taken at predetermined intervals. If haemodynamic stablility was maintained with fractional shortening > 25% and normal response to dobutamine, explantation was subsequently performed. A retrospective review was performed of patients assessed using this protocol.
Results: Ten of 55 (18%) supported patients from 2007 to 2011 showed recovery of ventricular function on this protocol and underwent explantation. Primary diagnoses were dilated cardiomyopathy ($n = 4$), hypertrophic cardiomyopathy ($n = 1$), post-transplant acute graft failure ($n = 1$), myocarditis ($n = 3$) and congenital heart disease ($n = 1$). Median age was 1.1 years (range 0.5–17 years) with median time on support 31 days (range 7–120 days). Three patients underwent early testing or 'forced' testing not in keeping with the protocol outlined, due to complications on support and the need to explant early if possible. There was one non-cardiac death post-explantation and two patients needed to go back on support (one after eight days and one had three VAD runs with two successful explantations each over a year apart). Both were subsequently transplanted.
Conclusions: Long-term support to recovery is achievable even in small children. A protocol that demonstrates normal cardiac output and ventricular function on echocardiography with a positive inotropic response may be used to predict which patients can successfully undergo VAD explantation.

531: GOING DOWN, GOING SLOW: ESMOLOL AS POTENT MYOCARDIAL PROTECTOR IN RESCUE CARDIAC EXTRACORPORAL MEMBRANE OXYGENATION

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Introduction: Cardiac failure or arrest post elective cardiac surgery in neonates and children are rare events. However their occurrence during a highly vulnerable period of myocardial recovery implies immediate expert support. Timing, efficiency of resuscitation and duration of cannulation for ECMO are crucial. Equally important is the subsequent cardiovascular management to optimise myocardial recovery. Besides volume unloading, optimal coronary perfusion has to be maintained to protect cardiomyocytes from oxidative stress. Beta-blockers combine cardioprotective mechanisms such as improved myocardial relaxation, coronary perfusion and also anti-oxidative activity.
Methods: Six patients required rescue ECMO post elective cardiac surgery. They were started on esmolol infusion as soon as they stabilised (full-flow ECMO ≥ 150 ml/kg/min). Serial transthoracic echocardiography was performed to assess myocardial contractility.
Results: Six patients (two male, four female), age 2.2 ± 4.1 years with single-ventricle physiology (n = 3), complex cyanotic heart disease (n = 2) and coronary anomaly (n = 1) all had myocardial stunning. ECMO was carried out at 8.8 ± 1.9 days, maximum dose esmolol was 106.7 ± 50.1 µg/kg/min, maximum heart rate (HR) prior to esmolol was 168.3 ± 11.7 beats/min (bpm), maximum heart rate during esmolol was 73.3 ± 8.2 bpm, fractional shortening (FS) prior to esmolol was 9.2 ± 4.9%, FS post esmolol was 33.3 ± 7.5%. Weaning of ECMO was successful in four patients.
Conclusions: In this small pilot study without case controls, all patients showed significantly improved myocardial contractility. Esmolol appears to provide cardioprotection for paediatric patients post cardiac failure/arrest requiring ECMO. Its combined anti-oxidative effect may support recovery of myocytes by increased gluthation peroxidise and superoxide dismutase activity.

540: INCREMENTAL RISK FACTORS FOR MORTALITY AND MORBIDITY AFTER INFANT HEART SURGERY IN THE DEVELOPING WORLD

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Background: We examined our institutional paediatric heart surgery database to determine the impact of low birth weight, malnutrition, need for pre-operative intensive care support or mechanical ventilation on the outcomes of 447 consecutive heart operations in children under two years.
Methods: Data on paediatric heart surgery (January 2010 to December 2011) were collected prospectively as a part of the International Quality Improvement Collaborative. Stepwise logistic regression analysis was performed and all pre-operative variables with $p < 0.05$ were allowed to enter into the model.
Results: There were 90 newborns (20.1%) and 359 (80.3%) were under one year old. Mean weight Z-score was −2.7 ± 1.7 at surgery and 112 (25%) had low birth weight. Prior to surgery, 59 (13.2%) needed intensive care, 44 (9.8%) were ventilated and 13 (2.9%) had positive blood cultures. Mortality (5.1%) was significantly associated with RACHS-I risk category (1, n = 10: 0%; 2, n = 237: 1.3%; 3, n = 148: 8.8%; 4 and above, n = 52: 13.5%; $p < 0.001$). After adjustment for RACHS-I category, pre-operative intensive care and pre-operative mechanical ventilation emerged as significant determinants of mortality, duration of mechanical ventilation and postoperative sepsis. Pre-operative sepsis was strongly associated with postoperative sepsis alone (odds ratio: 34.65; 95% CI, 7.35–163.4; $p < 0.001$). Low birth weight and malnutrition were not associated with any of the outcome measures. On stepwise logistic regression analysis pre-operative ICU stay and pre-operative ventilation emerged as significant predictors of outcome.
Conclusions: The need for pre-operative intensive care and mechanical ventilation were strongly associated with poor outcomes after infant cardiac surgery in this large single-centre experience from a developing country. It is worth exploring the utility of these additional variables in refining existing risk-adjusted scores for congenital heart surgery.

556: A PILOT PHASE I/II TRIAL OF ERYTHROPOIETIN NEUROPROTECTION IN NEONATAL CARDIAC SURGERY

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Background: Acute neurological injury and longer-term neurodevelopmental problems are common in neonates undergoing cardiac surgery, with up to 50% of patients affected. Erythropoietin (EPO) has anti-apoptotic, anti-inflammatory and anti-excitatory cell death properties, protecting the brain against cerebral injury in animal models and birth asphyxiated neonates. This phase I/II trial was designed to assess safety and indicate efficacy of EPO treatment for neonatal cardiac surgery.
Methods: This was a prospective, randomised, blinded, placebo-controlled trial of EPO vs normal saline control (US FDA IND 100011, NCT00513240). Neonates (< 30 days) scheduled for cardiac surgery with hypothermic CPB were enrolled. EPO doses were 1 000 units IV (or placebo equivalent) in three doses: (1) 12–24 hours pre-operatively; (2) immediately after CPB; (3) 24 hours after dose 2. Brain MRI was performed pre-operatively and seven days postoperatively. Primary outcome was Bayley scales of infant and toddler development III (BSID III) at 12 months of age.
Results: The study drug was given to 59 patients. Five patients had dural sino-venous thrombosis (two EPO, three placebo); six patients died before the age of 12 months (three EPO, three placebo); 11 patients declined 12-month follow up (seven EPO, four placebo, p = 0.48), leaving 42 patients with 12-month BSID III (79% of survivors). BSID scores were not different with EPO.
Conclusions: EPO treatment was not associated with a difference in 12-month BSID III. Complications, including major intracranial thrombosis, MRI brain injury and death were not more common with EPO treatment. An optimised study design, likely in a multicentre setting, is required to define the utility of EPO neuroprotection in neonatal cardiac surgery. We advocate such a study because of the many desirable properties of EPO for neuroprotection, and its demonstrated efficacy in other neonatal settings of cerebral injury.

558: THE ASSOCIATION OF VOLATILE ANAESTHETIC EXPOSURE WITH NEURODEVELOPMENTAL OUTCOMES AT AGE 12 MONTHS AFTER NEONATAL CARDIAC SURGERY

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Introduction: Volatile anaesthetic agents (VAA) cause neuro-apoptosis in neonatal animals, and human data demonstrate an association between early anaesthetic exposure and neurobehavioural problems. This study quantitated VAA exposure in the first 12 months in neonates undergoing cardiac surgery, to determine association with neurodevelopmental outcomes.

Methods: Ninety-three neonates (< 30 days) undergoing surgery with hypothermic CPB had prospective data collection for both cardiac and non-cardiac anaesthetics. End-tidal VAA was recorded every 15 minutes and inspired VAA concentration in the CPB sweep gas recorded every 10 minutes. Age-adjusted minimum alveolar concentration (MAC) hours were calculated. The primary outcome was the relationship between MAC hours of VAA, and cognitive, language and motor composite scores of the Bayley scales of infant development III (BSID) at age 12 months.
Results: Ninety-three patients had neonatal cardiac surgery, 10 of these patients died and 24 did not return for 12-month follow up, leaving 59 undergoing BSID (71% of survivors). After unadjusted linear regression analysis, higher MAC hours VAA exposure was associated with lower 12-month BSID cognitive score [Cog = 107.7 – (1.5 × MAC hours), $R = 0.33$, $R^2 = 0.11$, $p = 0.01$]. There was a trend towards lower language ($p = 0.18$) and motor ($p = 0.09$) scores with higher MAC hours. After backward stepwise multivariable analysis, increasing total MAC hours was associated with lower cognitive score ($p = 0.01$). Higher MAC hours were associated with lower language score ($p = 0.02$), but were not associated with lower motor score ($p = 0.49$).
Discussion: We demonstrated an association between increasing exposure to VAA, and lower BSID III scores at 12 months of age, independent of covariates including cardiac anatomy, CPB time, regional cerebral oxygen saturation and MRI brain injury. Each additional MAC hour of VAA exposure was associated with a 1.5-point decrease in cognitive score. Anaesthetic technique may be an important factor in adverse neurodevelopmental outcomes after neonatal cardiac surgery.

562: EARLY OUTCOMES FROM A NEW REGIONAL PROGRAMME FOR THE SURGICAL MANAGEMENT OF HLHS IN AUSTRALIA

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Introduction: Early survival and quality of outcome after surgery for hypoplastic left heart syndrome (HLHS) are influenced by patient-specific factors, the quality of surgery and peri-operative care. Some skills are common to the care of other complex neonatal presentations but integrating this expertise is a key challenge for new programmes. We began offering surgery for HLHS from 2006 and provided a regional service from January 2009, and we report early outcomes here.
Methods: Prospectively collected data for neonates with HLHS undergoing surgical palliation from January 2006 until June 2011 were analysed. Standard definitions of high- and standard-risk presentations were utilised.
Results: Thirty neonates underwent surgical palliation of HLHS with a modified Norwood procedure with an overall survival to stage II palliation of 80%; 46.7% of our patients were categorised as high risk, mostly on the basis of low birth weight. Survival to stage II palliation was 100% in standard-risk patients and 57.1% in the high-risk group. Survival at one year was 67% and comparable to the SVR study at 69%.
Conclusion: Outcomes for this new programme are comparable to reported outcomes. Excellent outcomes can be achieved in standard-risk patients. Outcomes in the high-risk group may be improved by alternative approaches and rigorous case selection. Providing information about local outcomes is an important element in counselling of families with antenatal diagnosis of HLHS.

564: TWO-VENTRICLE REPAIR FOR COMPLEX CONGENITAL HEART DEFECTS THAT ARE PALLIATED TOWARDS SINGLE-VENTRICLE REPAIR

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Background: Complex congenital heart defects that present earlier in life are sometimes channelled towards single-ventricle repair, because of anatomical or logistical challenges involved in two-ventricle correction. Given the long-term functional and survival advantage, we have been consciously exploring the feasibility of a biventricular repair when these patients present later for Fontan completion.
Methods: Since June 2009, 71 patients were referred for staged completion of Fontan. Following detailed evaluation that included 3D echo and MRI, 10 patients (group I: median age six years) were identified and later underwent complex biventricular repair with take down of Glenn shunt. Non-routable VSD, straddling tricuspid valve, requirement of complex repair using conduit at a young age, and ventricle being too small to support the systemic or pulmonary circulation were the reasons to defer biventricular repair initially. In two patients, the decision for palliation with Glenn shunt was taken on the surgical table during previous failed attempts at biventricular correction. Completion of extracardiac Fontan repair was done in 61 patients (group 2: median age seven years).
Results: Two-ventricle repair was accomplished in all 10 group 1 patients. The mean cardiopulmonary bypass time (362 ± 93 vs 155.9 ± 95.7 min, $p < 0.0001$) and mean ICU stay of 7 ± 3.6 vs 5.4 ± 3 days were longer in group 1 patients, but were not statistically significant. One patient developed complete heart block requiring permanent pacemaker. Late patch dehiscence occurred in another (awaiting repair). At a median follow up (15 months), there was no mortality among group 1 patients and all except one were symptom free. There were two early deaths (3.2%) in group 2.
Conclusion: Two-ventricular repair, although surgically challenging, should be considered in all patients with two functional ventricles who come for Fontan completion. Comprehensive pre-operative imaging and meticulous planning helps in identifying suitable candidates.

567: SHORT-TERM RESULTS OF TETRALOGY OF FALLOT REPAIR IN THE HOSPITAL INFANTIL DE MEXICO FEDERICO GAMEZ

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Background: Tetralogy of Fallot (TOF) accounts for 3.5% of all the patients with congenital heart defects. Complications associated with the repair vary according to surgical technique and patient-specific factors. The present study was conducted to investigate the short-term clinical outcomes of TOF repair and to identify risk factors associated with complications and death.
Methods: Between 2006 and December 2010 we retrospectively reviewed the outcomes of 78 patients operated for tetralogy of Fallot at the Hospital Infantil de Mexico Federico Gómez.
Results: Seventy-eight patients were operated on during the period of study. The mean age at diagnosis was 1.2 years; the mean age at surgery was 2.7 years. Fifteen patients had received a previous systemic–pulmonary fistula (19%). The type of surgery was infundibuloplasty in 44 patients (55.7%), with a transannular patch in 27 (34.2%), with a valved conduit in two (2.5%), with valve replacement in two (2.5%), with homograft in two (2.5%), and one patient underwent a monocusp valve formation (1.3%). The extracorporeal circulation mean time was 133 minutes and the aortic clamp of 79 minutes. The mechanical ventilation time was 2.29 days. The mean

time in intensive care unit was 5.39 days. Twenty-four patients had complications (31%); 15 required some type of re-intervention and five died before hospital discharge (6.3%). The main cause of death was infection. The length of hospital stay before surgery was a risk factor for infection and death.
Conclusions: In our country refererral for congenital heart surgery is delayed. Children often arrive with associated infections or acquire an infection in hospital awaiting corrective surgery. It is imperative to educate primary care physicians for an earlier referral and to improve our referral system to avoid long periods of hospitalisation before surgery.

587: THE PROCESS AND EXECUTION OF MULTI-INSTITUTIONAL CROSS-SECTIONAL FOLLOW-UP STUDIES: THE CONGENITAL HEART SURGEONS' SOCIETY DATA CENTRE EXPERIENCE

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Background: The Congenital Heart Surgeons' Society (CHSS) data center (DC) serves as the coordinating centre for multi-institutional, cross-sectional follow-up studies of children with complex and rare congenital heart defects (CHD). The success of our studies relies on our ability to follow patients long term. Recent trends show a decline in follow up. The DC has developed new strategies to improve long-term follow up.
Methods: Our annual cross-sectional follow-up process extends over a three-month period, which entails questionnaire mail-out, reminder calls and dedicated time to locate patients with wrong addresses. A variety of options exist for patients to complete follow-up forms including mail, e-mail or telephone. Locating patients with wrong addresses continues to be challenging despite the use of death registries, internet searches, Facebook and Twitter. New strategies to address these issues include creation of a one-page form sent to participating institutions verifying patient information as well as decreasing follow up to two months, and utilising the third month to solely address these issues. The DC team continuously explores additional ways of completing and submitting annual follow up including a secure website patients can log into that would adhere to current privacy laws.
Results: The implementation of the patient update form has been somewhat effective in obtaining current patient information essential to follow up. The use of social media has not been as successful as we had anticipated. The DC staff and members of the Research Institute at the Hospital for Sick Children continue to collaborate on developing a secure website for patients to complete online annual follow up.
Conclusions: Although these strategies have been somewhat effective in improving our follow up, we are continuously exploring other strategies to improve the success of our long-term follow up. To ensure the success of multi-institutional, cross-sectional follow-up studies, continuous evaluation and implementation of new strategies is critical.

590: EFFECTIVENESS OF CONTRAST-ENHANCED COMPUTED TOMOGRAPHY AS A SUPPORTIVE MEASURE FOR TREATMENT OF MEDIASTINITIS AFTER PAEDIATRIC CARDIAC SURGERY

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Objectives: Mediastinitis after cardiac surgery is a serious complication; however, there is no consensus on the treatment strategy of mediastinitis in paediatric cardiac surgery.
Methods: Paediatric patients who showed postoperative infectious signs (e.g. fever, wound swelling, redness, pain, draining pus and abnormal laboratory data) were evaluated using 320 multi-detector row contrast-enhanced computed tomography (CECT) for diagnosing mediastinits. We defined that a typical CECT image of mediastinitis showed fluid collection with ring enhancement behind and between the sternum. Aggressive debridement followed by primary sternal closure was performed when typical findings of CECT were confirmed.
Results: From January 2007, 533 paediatric patients underwent median sternotomy. Thirteen cases (2.4%) had CECT performed for evaluating mediastinitis. The typical findings were obtained in 12 of the 13 cases. In one case without typical findings, pus flowed out from infectious site before CECT. Aggressive debridement followed by primary sternal closure was performed in all cases. There were no operative and hospital deaths. Re-exploration was required in one case due to the placement of a polytetrafuoloethylene membrane after the initial debridement. In 14 debridements for the 13 cases, the median mechanical ventilation time was 16.0 hours (range 0–116.6), and length of intensive care unit stay after primary sternal closure was four days (range 1–21 days). The median postoperative hospital stay from primary sternal closure was 23 days (range 15–53 days).
Conclusion: Primary sternal closure is feasible surgical treatment for mediastinitis in paediatric patients. The CECT image is helpful for aggressive debridement due to precise detection of the inflammatory area of mediastinitis. CECT should be a useful supportive measure for improving the outcome of primary sternal closure.

603: CARDIOPULMONARY EXERCISE TESTING IN PATIENTS WITH PRIMARY PULMONARY HYPERTENSION

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Background: Cardiopulmonary exercise testing (CPX) has been widely used for assessing heart failure severity in children and adolescents with cardiac disease. However, little CPX-derived data on those with primary pulmonary hypertension (PPH) are available.
Aim: Our purpose was to clarify relationships between CPX-derived variables and haemodynamics obtained by cardiac catheterisation in children and adolescents with PPH.
Methods and Results: From 1995 to 2011, 18 PPH patients (age: 18.1 ± 6.2 years) underwent 48 CPXs and the results were compared with the haemodynamics. Ventilatory equivalent for carbon oxide production at peak exercise (peak VE/VCO_2) closely correlated with the pulmonary-to-systemic artery pressure and resistance ratios (r = 0.5, $p < 0.001$, and $r = 0.6$, $p < 0.001$, respectively). When patients were divided into two groups according to the peak VE/VCO_2, i.e. high VE/VCO_2 vs low VE/VCO_2, the high-VE/VCO_2 group showed significantly higher pulmonary arterial pressure and resistance, lower cardiac index, smaller left ventricular size, and shorter distance of six-minute walk ($p < 0.05$). In contrast, the other CPX-derived variables, including peak oxygen uptake or heart rate, did not correlate with the haemodynamics.
Conclusions: Peak VE/VCO_2 was the best CPX-derived variables for predicting haemodynamic severity in children and adolescents with PPH, indicating the importance of exercise ventilatory abnormality in PPH pathophysilogy.

605: HEART RATE VARIABILITY IN CHILDREN WITH VASOVAGAL SYNCOPE DURING ASYMPTOMATIC PERIODS

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Objective: To retrospectively analyse the heart rate variability parameters in children with vasovagal syncope (VVS) during asymptomatic periods.

Methods: Forty-five children with a typical history of VVS were enrolled as a study group, among them, 38 patients had a positive response to HUTT while seven had a negative test. Twenty healthy children with similar characteristics without syncope were matched as a control group. HRV was calculated over a 24-hour period for the time-domain indices and frequency-domain indices.
Results: The study group had lower time-domain indices of SDNN ($p < 0.01$), SDANN ($p < 0.05$), and higher frequency-domain indices of LF ($p < 0.05$) and LF/HF ($p < 0.01$) than the control group. There were no significant differences in time-domain and frequency-domain indices between HUTT+ VVS patients and HUTT– VVS patients. VVS patients with vasodepressor responses had lower SDNN ($p < 0.01$), SDANN ($p < 0.05$), rMSSD ($p < 0.05$) and PNN50 ($p < 0.05$) and higher frequency-domain indices of LF ($p < 0.05$) and LF/HF ($p < 0.01$) when compared with cardio-inhibitory response patients. The same occurred in the VD group, rMSSD ($p < 0.05$), PNN_{50} values ($p < 0.05$) were lower and LF ($p < 0.05$), LF/HF ($p < 0.01$) were higher when compared with mixed-response patients. In the CI and MX groups there were no significant differences in HRV values. Compared with the controls, rMSSD and PNN50 declined obviously in the VD group while LF increased significantly.
Conclusion: There were different changes in baseline autonomic activity in VVS children with different haemodynamic types during asymptomatic periods. Changes in RMSSD, PNN50 and LF parameters may be used as a predictor to identify vasodepressor type of VVS in clinical diagnosis.

607: SUCCESSFUL TREATMENT OF THERAPY-RESISTANT LEFT VENTRICULAR OUTFLOW OBSTRUCTION IN CHILDHOOD HYPERTROPHIC CARDIOMYOPATHY USING SHORT ATRIOVENTRICULAR DELAY DUAL-CHAMBER PACING WITH A LEFT VENTRICULAR EPICARDIAL ELECTRODE

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Background: In hypertrophic obstructive cardiomyopathy (HOCM), left ventricular outflow (LVOT) gradient is caused by the septum bulging into the LVOT, and systolic anterior movement of the anterior cusp of the mitral valve. Unrelieved LVOT obstruction is a risk factor for death. Primary treatment for gradient reduction is pharmacological, with beta-blockers, calcium antagonists and disopyramide. Optimal management of myectomy and drug-refractory LVOT gradient is unknown. In adults, short atrio-ventricular (AV) delay dual-chamber (DDD) right ventricular pacing has been used.

Results are modest, with some non-responders.
Methods: We report two consequtive paediatric patients with HOCM and therapy-resistant LVOT gradient after short AV-delay DDD pacing with an epicardial ventricular electrode placed at the left ventricle apex. Age at pacing was seven months and 16 years. Both had maximal pharmacotherapy with propranolol and disopyramide, and had been through surgical myectomy of LVOT without any significant relief of outflow obstruction. Optimal AV-delay velocity through the LVOT was measured by continuous-wave Doppler from the apical view. The AV delay producing the least gradient was chosen. The mitral inflow signal was then recorded and care was taken that the atria would not contract against a closed mitral valve. Both patients remained on their medical treatment during pacing.
Results: Before pacing, the peak gradient was 81 and 61 mmHg. AV delay producing the smallest gradient was 80 ms in both patients. After pacing, at 12 and 3.5 months, the gradients were 7 and 9 mmHg, respectively
Conclusions: Short-term results in our two paediatric patients who failed medical treatment and myectomy showed complete relief of the gradient. Pacing LV apex with short AV delay activates papillary muscles before the outflow septum. The mitral valve apparatus moves posteriorly and the LV apex has time to empty before the septum bulges into the outflow tract. Pacing from the LV apex seems to be more effective than RV pacing. It however requires thoracotomy.

610: AORTICO-LEFT VENTRICULAR TUNNEL: CASE REPORT AND LITERATURE REVIEW

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Background: ALVT is a rare congenital anomaly. Echocardiography can identify the ALVT and associated lesions. Catheterisation should be reserved for patients with unclear non-invasive findings or percutaneous closure. Neonatal surgery has been advocated in all patients due to long-term concern of aortic regurgitation (AR).
Objectives: The purpose of this case report was to review the literature on aortico-left ventricular tunnel (ALVT) and describe a case that was misdiagnosed as ventricular septal defect (VSD).
Methods: We describe an ALVT case that was misdiagnosed initially as a VSD. A two-year-old boy, presenting with heart failure and murmur was investigated by echocardiography that misdiagnosed a VSD. Clinical, echocardiographic and surgical case details as well as the literature were reviewed.
Case: Congestive heart failure and a murmur were diagnosed in the physical examination and furosemide, enalapril and digoxin were prescribed. Pre-operative echocardiogram revealed a perimembranous VSD of 4.6 mm with moderate repercussion, aortic dilatation and mild AR. The patient underwent surgery to occlude the VSD, which was not detected. A right sinus of Valsalva aneurysm was detected and aortotomy revealed an ALVT that was corrected by patch closure of the aortic end of the ALVT and a plasty of the sinus aneurysm. At six months' follow up, the patient was asymptomatic, without AR and distal tunnel flow in the interventricular perimembranous septum was observed.
Conclusion: Aortico-left ventricular tunnel is a rare cardiac malformation. The literature shows a good postoperative long-term outcome and surgical correction is recommended soon after diagnosis in symptomatic patients. In our case two-dimensional and pulsed colour-flow Doppler imaging did not establish the diagnosis. Three-dimensional or transoesophageal echocardiogram and more clinical suspicion was helpful in its diagnosis. This article is to remind us that sometimes the diagnosis of rare congenital heart diseases can be done in the operating room if not diagnosed pre-operatively.

614: SCIMITAR SYNDROME: SURGICAL DIRECT CONNECTION BETWEEN RIGHT PULMONARY ANOMALOUS VEIN AND LEFT ATRIUM

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Background: Scimitar syndrome is a rare congenital heart disease. Different surgical approaches have been described.
Objectives: The aim of this study was to review the case of a patient of scimitar syndrome who underwent surgery to directly connect the right pulmonary anomalous vein (RPAV) to the left atrium (LA).
Methods: A four-year-old girl presenting with pneumonia was investigated by means of computed tomography, which diagnosed RAPV connected to the inferior vena cava. Clinical, echocardiographic, computed tomography and surgical case details as well as the literature were reviewed. Her pre-operative echocardiogram revealed enlargement of the right atrium and right ventricle with an anomalous connection from the right pulmonary vein to the inferior vena cava. The patient was submitted to surgery and a direct connection between the right pulmonary anomalous vein and left atrium was done. At 10 months' follow up she was asymptomatic. Right phrenic nerve paralysis was observed.
Conclusion: In this case a direct connection between RAPV and LA was feasible because of the anatomical features.

620: SAFETY AND TOLERANCE OF ORAL ENOXIMONE IN PAEDIATRIC MYOCARDIAL FAILURE

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Background: Continuous intravenous administration of phosphodiesterase 3 inhibitors have proven to be a valuable and successful therapy in paediatric patients with acute or chronic myocardial dysfunction. Intravenous treatment is associated with risk of infection and considerable patient discomfort. There is currently no specific oral preparation available. Weaning from intravenous medication to oral angiotensin converting enzyme (ACE) inhibitors and beta-blockers can be challenging. We examined our experience using the intravenous preparation of enoximone as an oral medication in this setting.
Methods: We retrospectively reviewed hospital records of 33 patients receiving oral enoximone in a single tertiary paediatric cardiac centre between November 2005 and December 2011. Indications for oral enoximone were inability to wean from intravenous milrinone infusion and/or intolerance of ACE inhibitor therapy.
Results: Patient age at the start of oral enoximone was 0.5–191 months (median 8.5 months). Seven patients (21%) had left ventricular dysfunction due to myocarditis/cardiomyopathy and 26 (79%) had myocardial dysfunction complicating congenital heart disease, 25 (75%) following cardiac surgery. Of this latter group, 12 (48%) had left ventricular, nine (36%) had right ventricular and four (16%) had biventricular dysfunction. All patients received oral enoximone at 1 mg/kg three times a day. Enoximone was well tolerated at this dose without adverse haemodynamic effect. Due to the alkaline nature of this solution there were two (6%) patients with blood-stained gastric content aspirates when enoximone was administered without milk, which subsequently resolved when given with milk. No other adverse effects were encountered and the families tolerated well the inconvenience of using an intravenous preparation orally. Results of outcome data analysis will be presented.
Conclusions: Oral enoximone is a safe alternative to protracted intravenous treatment of severe myocardial failure in children. Based on our experience it is a well-tolerated and promising alternative when ACE inhibitors and beta-blockers are not tolerated.

622: THE NIKAIDOH SURGICAL PROCEDURE: INITIAL EXPERIENCE AND MID-TERM RESULTS

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The Nikaidoh procedure is a recent surgical approach to repair TGA and DORV with VSD and PS.
Objective: To review our experience using the Nikaidoh procedure.
Methods: Between 2005 and 2012,10 patients underwent a modified Nikaidoh procedure at our institution, median age: 2.9 years (CI 25–75% = 1–5.6) and median weight: 12.5 kg (CI 25–75% = 8.7–19). Two anatomical variants were identified: TGA with VSD and PS (seven patients) and DORV with PS (three). All had a VSD distant from the great arteries (inlet type, six; and muscular, four patients).
Results: The mean CPB time was 244 min (SD ± 50) and aortic cross-clamping was 181 min (DS ± 44). The median length of stay was 11 days (CI 25–75% = 8–45), MV six days (CI 25–75% = 4.5–22) and inotropic requirement was eight days (CI 25–75% = 5.5–23.5).
Early outcome: No mortality occurred; three patients suffered transient arrhythmias (one AVB, two JET). One patient developed infective endocarditis and needed a mitral valve and RV–PA homograft replacement. Six patients presented some degree of ventricular dysfunction controlled with medical treatment, except one who needed ventricular assistance. There was no obstruction to either RV or LV outflow tracts. None presented aortic insufficiency more than trivial or pulmonary incompetence more than moderate.
Mid-term results: The median follow up was 4.3 years (2–5.6 years). No deaths occurred. All patients were free from re-operation and re-intervention. All patients were in NYHA functional class I, with no arrhythmias, no LVOTO and a good biventricular function. The aortic valve was competent in 40%. In six patients the aortic insufficiency was mild. Regarding the RV–PA conduit, there was a mild stenosis in six patients and moderate in one. Mild pulmonary insufficiency was found in four and it was moderate in six patients.
Conclusions: Mid-term actuarial survival was 100% after the Nikaidoh procedure. This surgical technique provides complete freedom from significant insufficiency and left or right ventricular outflow tract obstruction. The modified Nikaidoh operation is a good surgical option in patientts with TGA, VSD and PS, and with DORV and PS, particularly when some anatomical features, such as the anatomical position of the VSD are considered inadequate for a Rastelli procedure. Its long-term benefits need to be evaluated with a larger number of patients and longer follow up.

626: ROLE FOR IMMUNE-MONITORING TO TAILOR INDUCTION PROPHYLAXIS IN PAEDIATRIC HEART RECIPIENTS

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Background: Rabbit antithymocyte globulin (rATG) is routinely used for induction, but is costly and associated with increased risk of infection and post-transplant lymphoproliferative disorder (PTLD).
Hypothesis: We hypothesised that CD_3 monitoring to tailor rATG induction would have less infection, reduced costs and similar rejection rate.
Methods: Heart transplant (Htx) recipients who received rATG between November 2006 and July 2011 were reviewed. We compared patients with CD_3 monitoring used to adjust medications (group 1, n = 16) vs group 2 (patients with standard rATG dosing). Absolute CD_3 count < 25 cells/mm^3 was used to adjust rATG in group 1. Outcomes included first-year incidence of infection and rejection, direct costs of rATG, and incidence of PTLD and all deaths. Statistics: Fisher's exact test, Wilcoxon rank sum and Wilcoxon sign rank test.
Results: Demographics and cardiac diagnoses were similar between the study and control cases. Compared to the controls, the study cases received fewer daily doses of rATG [median 3 (3–7) vs 4 (2–7), p = 0.005] and less total rATG (median 3.1 vs 7.4 mg/kg, $p < 0.001$). Compared to a fixed dosing regimen of 1 mg/kg/day × 5–7 days, the drug cost savings for study cases were significant (total cost $58 432 vs $93 627, $p = 0.001$), a 37% reduction in total cost. There was no difference in the incidence of infection (29 vs 44%), rejection (53 vs 31%), or mortality (6 vs 6%) during the first year following Htx. There was one late death and one late case of PTLD in group 2 (p = ns).
Conclusions: CD_3 monitoring to tailor rATG induction in Htx recipients is associated with reduced drug costs and similar rates of rejection and infection. Longer follow up will determine if there is extended benefit from tailored immunotherapy.

641: LIVOSEMENDAN VS MILRINONE IN PREVENTION AND TREATMENT OF LOW CARDIAC OUTPUT IN CHILDREN UNDERGOING CORRECTIVE OPEN-HEART SURGERY FOR CONGENITAL HEART DISEASE

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Background: Low-cardiac output (LCO) state is a well-known complication in children undergoing corrective open-heart surgery; it usually occurs eight to 10 hours after surgery. Milrinone has been shown to be superior to conventional ionotropes in prevention and treatment of LCOS in children. Recently, levosimendan, a Ca^{2+} sensi-

tiser, has shown its superiority in various clinical trials conducted in adult cardiac surgery. However its efficacy and safety in prevention of treatment of LCOS in children undergoing surgery for congenital heart disease has not been extensively studied.
Aim: In children under two years of age after corrective open-heart surgery for congenital heart disease to compare (1) the efficacy of livosemendan to milrinone for prevention and treatment of low-cardiac output state; and (2) the safety of livosemendan against that of milrinone.
Methods: This was a case–control study in which livosemendan was used in place of milrinone in children undergoing total correction for congenital heart disease. Both drugs were used prophylactically while coming off bypass, with other ionotropes as deemed necessary. The babies were monitored in the paediatric cardiac ICU over the next 72 hours for evidence of low-cardiac output state, in the form of heart rate, blood pressure, urine output, serum lactate levels, vasotropic ionotropic score, and ventricular function by echocardiography.
Results: The groups did not differ significantly in the development of LCOS and its recovery in the 72 hours. There were no serious adverse events or unexpected adverse drug reactions during the study.
Conclusion: Livosemendan is as safe and effective as milrinone in the prevention and treatment of LCOS in children undergoing total correction for congenital heart disease

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645: EARLY AND INTERMEDIATE-TERM OUTCOMES AFTER TRANSPLANTATION FOR RESTRICTIVE CARDIOMYOPATHY IN CHILDREN

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Background: Restrictive cardiomyopathy (RCM) has the worst outcome of all cardiomyopathies, with > 50% mortality within two years of diagnosis. Heart transplantation (Htx) in this patient population has acceptable early mortality, however no studies of intermediate-term outcomes exist. We hypothesised that RCM has excellent early and mid-term survival.
Methods: We reviewed our experience with paediatric Htx for RCM from May 1988 to August 2012. Clinical data, support while waiting, recipient and donor variables, and explant pathology were analysed. Outcomes were examined via Kaplan–Meier survival curves.
Results: Of 198 transplants in 189 patients during the study period, 17 (9%) had RCM. Median age at Htx was 7.7 (range 0.89–16.2) years. Most common presentation was tachypnoea/cough (11), FTT (three) and syncope (one). No patients had a dysmorphic syndrome; however, one was diagnosed with a desmin cardiomyopathy and another a mitochondrial disorder after Htx. Only two had familial cardiomyopathy. Between listing and Htx, five patients were ventilated, and two were on mechanical support; 11 patients were status 1 or 1a. Median time from listing to Htx was 16 days (range 1–81). Two had ABO-incompatible Htx at ages eight and 10 months. Thirty-day, one-year and five-year survival were 100, 100 and 73%, respectively, with seven deaths at 3.3 to 13.7 years post Htx. One patient was retransplanted 8.9 years post Htx (CAV) and died four years later of rejection. Other deaths were from rejection and/or sudden death (four), rejection/infection (one), CAV (one) and PTLD (one). Non-adherance was suspected in five of these patients.
Conclusions: Heart transplant is an effective therapy for children with RCM, providing excellent early and mid-term survival. Late mortality was primarily due to rejection.

646: PREVALENCE, ASSOCIATED FACTORS AND OUTCOMES OF ACUTE KIDNEY INJURY AMONG CHILDREN AND ADOLESCENTS UNDERGOING CARDIOPULMONARY BYPASS SURGERY IN NAIROBI, KENYA

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Background: AKI is a serious complication associated with cardiopulmonary bypass surgery. The development of AKI is associated with substantial morbidity and mortality. This study was done to determine the prevalence and risk factors of AKI, and outcome of children and adolescents with AKI following cardiopulmonary bypass surgery at Kenyatta National and Mater Hospitals.
Methods: This was a prospective cohort study of 89 participants from birth to 17 years recruited consecutively as they were admitted for cardiopulmonary bypass surgery. Creatinine measurements were obtained the week before surgery, at eight hourly intervals in the first 24 hours post operatively, and then at 48 hours. A decline in creatinine clearance of ≥ 25% from baseline was used as the threshold to define AKI. Kidney injury was classified according to the RIFLE system where patients are categorised as being at risk, having injury and complete failure.
Results: The overall prevalence of AKI was 37.1%. According to the RIFLE system, risk occurred in 34.8%, injury in 11.2%, and failure in 1.1% of patients. Patients with AKI were older, median age nine years (IQR 3–12), compared to four years (IQR 1.5–8) for those without AKI; 55% of AKI patients had moderate to severe malnutrition compared to 34% of patients without AKI (OR 2.00, $p = 0.17$). The median length of ICU stay in AKI patients was four days (IQR 2–6). The mortality rate in AKI vs non-AKI patients was 15.2 and 9%, respectively (OR 1.8; 95% CI: 0.5–6.8, $p = 0.37$). Up to 18.2% of AKI patients required dialysis compared to 5% of non-AKI patients (OR 3.9; 95% CI: 0.9–16.9, $p = 0.06$).
Conclusion: The prevalence of AKI following cardiopulmonary bypass surgery in children and adolescents at the KNH and Mater Hospitals was high (37.1%), and was found in older patients. Malnutrition was associated with a two-fold risk of developing AKI.

649: INVASIVE PULMONARY BLOOD FLOW RESTRICTION IN PRE-OPERATIVE MANAGEMENT OF PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME IN A SINGLE LOW-VOLUME INSTITUTION

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Background: For patients with HLHS it is optimal to be in a stable condition prior to undergoing the Norwood procedure. In the setting of limited resources, the Norwood procedure sometimes has to be postponed for several days, which may expose the patient to the negative effect of systemic hypoperfusion due to runoff into the pulmonary circuit. In order to balance pulmonary and systemic blood flow, different non-invasive measures can be used, but their effectiveness and durability of effect are limited, and invasive measures may be necessary to restrict pulmonary blood flow.
Methods: Short-term ligation of the right pulmonary artery (rPA) (in 2007) or bilateral banding of pulmonary arteries (PAs) (from 2008 onward) were used as methods of invasive pulmonary blood flow restriction (iPBFR). Indications for iPBFR were as follows: systemic hypoperfusion with lactate level > 5 mmol/l, and oliguria together with ineffectiveness of non-invasive measures to limit pulmonary blood flow. Nine out of 29 HLHS patients required invasive pulmonary blood flow restriction from 2005 to 2011. Three temporary rPA ligations and six bilateral bandings of PAs were performed. Blood gas and metabolite level analyses were performed on a regular basis before and after iPBFR.
Results: The patients' mean body weight was 3.1 ± 0.56 kg. iPBFR

was performed at 5.3 ± 0.7 day of life. Mean blood lactate level prior to iPBFR was 7.8 ± 1.9 mmol/l, and mean arterial oxygen saturation was 89.8 ± 7.6%. Mean blood lactate level after iPBFR was 1.8 ± 0.8 mmol/l, and mean arterial oxygen saturation was 77.8 ± 5.3%. One patient died 20 hours after the procedure due to progression of organ dysfunction. The rest of the patients remained stable and underwent the Norwood procedure at 5.6 ± 2.8 days after the initial palliation.
Conclusions: Invasive pulmonary blood flow restriction reduces systemic hypoperfusion, improving the metabolic status and stabilising patients' condition before the Norwood procedure.

652: LONG-TERM SURVIVAL IN PIGLETS AFTER SECTIONING AND RE-SUTURING VESSELS MIMICKING THE ARTERIAL-SWITCH PROCEDURE FOR TRANSPOSITION OF THE GREAT ARTERIES

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Background: Children born with transposition of the great arteries undergo an arterial-switch operation (ASO) in the neonatal period. The ASO sections the sympathetic nervous inflow along the aorta and coronary arteries. Experiences of long-term follow up after ASO indicate that the autonomic nervous system consequences of the ASO need to be addressed. We developed a protocol for cardiopulmonary bypass (CPB) surgery in piglets allowing long-term survival after sectioning and re-suturing vessels as in the ASO. Post-operative care without intensive care unit facilities was a challenge.
Methods: Female piglets of Yorkshire–Hampshire crossbreed, aged 8.5 weeks were operated. Anaesthesia combining paediatric anaesthetic ICU and veterinarian experience included pre-operative sedation with midazolam/ketamine, induction and maintenance with propofol and fentanyl in combination with isoflurane inhalation. Lidocaine was infused to prevent arrhythmias. Standards for neonatal monitoring were applied. CPB equipment: oxygenators, circuit and pumps, were adapted to the range of flow and venous drainage, and cross-matched pig blood was added to the prime. The surgical technique had to be adapted to the different position of the the piglet's heart and large vessels. The protocol was approved by the local animal research ethics committee. Animal welfare rules forbidding division of the manubrium in pigs that must be able to stand after the operation added surgical difficulties. Special rigging of the aortic arch was necessary for cannulation and access. In addition, fragile tissues resulted in suturing difficulties. Re-warming to body temperature of 38°C before extubating was essential.
Results: Fourteen out of 19 piglets that underwent the mimicked ASO survived for long-term follow up and later *in vivo* and *in vitro* analysis of the physiological consequences of sympathetic denervation of the heart caused by the sectioning of the large vessels and the re-implantation of the coronaries.
Conclusions: This model can be used to enhance the knowledge of the short- and long-term consequences of paediatric cardiac surgery.

656: VARIABILITY IN RESPONSE TO AMLODIPINE IN HYPERTENSIVE PAEDIATRIC CARDIAC TRANSPLANT RECIPIENTS

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Background: Fifty to 70% of paediatric cardiac transplant patients develop late-onset hypertension. Amlodipine, a calcium channel blocker, is the most commonly used first-line antihypertensive agent.
Objective: To analyse the efficacy of amlodipine as first-line therapy for hypertension in paediatric cardiac transplant recipients.
Methods: Paediatric cardiac transplant recipients prospectively enrolled through the heart centre biobank were studied. Twenty-four-hour, daytime and night time mean systolic and diastolic blood pressure (BP) were captured from serial ambulatory BP measurements. Hypertension was defined as being above the 95th percentile in BP for gender, age and height. Amlodipine dose and BP indexed to amlodipine dose (in mg/kg/mmHg × 100) was assessed.
Results: Of 124 heart transplant patients in the biobank, 53% were male (mean age, 11.8 years at enrollment; 74% were white, 11% Asian, 4% black, 10% other). Ninety-seven received amlodipine during follow up. Of 97 patients in whom ambulatory BPs were available, 21 were analysed. Mean dose of amlodipine was 0.165 mg/kg/day (range 0.054–0.355). The BP indexed to amlodipine dose was highly variable.
Conclusions: There was large variability in amlodipine dose requirements and dose-adjusted response to amlodipine in hypertensive paediatric transplant patients. Genetic testing is underway to determine the contribution of pharmacogenetic variation to the amlodipine response. This knowledge will help in optimising the choice and dose of first-line antihypertensive therapy in this cohort.

661: PULMONARY VALVE IMPLANTATION USING SELF-EXPANDING TISSUE VALVE

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Introduction: Significant pulmonary regurgitation is a common problem after surgical or percutaneous treatment of congenital cardiac defects such as tetralogy of Fallot, negatively affecting long-term prognosis and necessitating re-interventions. The bio-integral no-react injectable pulmonic (NRIP) valve allows pulmonary valve replacement with or without cardiopulmonary bypass (CPB), minimising the impact of surgery. The aim of the work was to describe our multi-institutional experience with the clinical use of this device.
Methods: Between January 2006 and June 2012, 35 symptomatic patients, mean age 20.0 ± 12.5 years, with severe pulmonary regurgitation and progressive right ventricular dysfunction after tetralogy, received NRIP in 11 different European institutions. All patients underwent magnetic resonance (MR) before and after the implant and transoesophageal 2D echocardiography during the surgical procedure and follow up.
Results: Valve insertion, delivery and placement were successful in all patients but two that required the repositioning of the same valve in CPB. Of these, five patients implanted the valve in CPB to allow repair of intracardiac defects. Early recovery was uneventful and all the patients were discharged home after a mean length of hospital stay of 6.3 ± 2.4 days. Intra-operatively, transoesophageal echocardiography was the unique tool to guide device positioning and verify early surgical results. In three patients echo documented a valve displacement after delivery and guided the repositioning. In the postoperative course, serial echocardiographic and MR studies documented right ventricle reverse remodelling and excluded later complications. The mean follow up was 3.8 years.
Conclusions: The NRIP valve allows safe and easy pulmonary valve replacement without CPB in selected cases. Its mode of implantation offers a less-invasive approach with less blood loss and shorter hospital stays. Transoesophageal echocardiography plays an important role in the intra-operative management and the MR permits an adequate selection of patients. Longer follow up is required to assess valve performance.

671: LOW-VOLUME BLOOD SAMPLING TUBES CAN BE USED FOR THROMBELASTOMETRY IN PAEDIATRIC PATIENTS

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Background: Minimising the volume of blood used for diagnostic procedures in children undergoing congenital heart surgery is desirable. We investigated to what extent the size and the type of sample tube affected the results of thrombelastometry (RoTEM®).
Methods: In 20 healthy individuals, we compared four sampling tubes (evacuated 0.109 M sodium-citrate plastic tubes): Venosafe® 1.8, 2.7 and 3.6 ml and BD Vacutainer® 1.8 ml. Using four parallel RoTEM® devices we studied clotting time, maximum velocity and maximum clot firmness in three assays: ExTEM®, InTEM® and FibTEM®.
Results: No difference was found in any of the RoTEM® parameters using four different tubes for blood sampling. Intra- and inter-individual variation was acceptable. When comparing the two low-volume tubes, we found that the technical design of the tube did not influence RoTEM® results.
Conclusion: RoTEM® results were not affected by the size and the type of the sampling tube. Therefore, to minimise the volume of blood drawn, smaller tubes of 1.8 ml are preferable in paediatric patients.

672: IDEBENONE IN FRIEDREICH ATAXIA: IMPROVED FUNCTION AND REDUCED MYOCARDIAL HYPERTHROPHY

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We present a patient at the age of nine years who suffers from Friedreich ataxia with hypertrophic cardiomyopathy. She was born after a normal pregnancy and delivery. She began walking at 13 months but with difficulty. At the age of three years she often complained of pain in the legs while walking and she kept falling. She sweated a lot and in every respiratory infection symptoms got worse. Friedreich ataxia was diagnosed and a cardiological examination was done. Ultrasound showed hypertrophic cardiomyopathy with depressed contractility and there were repolarisation disturbances in the electrocardiogram. We treated her with idebenon after which the cardiac muscle contractility was normalised.

Friedreich ataxia is an autosomal recessive multisystem progressive disease characterised by disturbances in walking, and extremity ataxia caused by deterioration of the rear column and spinocerebellar tracts in the spinal cord. Mutation of the gene for frataxin (FXN) causes the disease. In most patients, the disease affects the heart and in combination with neurological problems, significantly reduces the patient's abilities and often causes premature death, particularly in patients who have the disease before the age of 20 years. Hypertrophic cardiomyopathy with normal ejection fraction is the most frequent ultrasound finding in the heart in 75% of the cases.

Unfortunately, there is no cure for this disease and the function of fraxatin protein is unknown. The present studies show that frataxin protein is important in regulating the transfer of iron in mitochondria. Idebenone is a short-chain bensoquinon similar to coenzyme Q_{10}. It is a powerful antioxidant and electron carrier in the respiratory chain and helps the creation of ATP. The recent studies have shown that idebenone is effective in doses of 5–10 mg/kg/day in patients with FA and cardiomyopathy.

674: A NEWBORN WITH HIGH-OUTPUT CARDIAC FAILURE DUE TO A LARGE VEIN OF GALEN MALFORMATION

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Neonatal heart failure in newborns is often caused by asphyxial cardiomyopathy, left-sided obstructive lesions, large mixing cardiac defects and myocarditis. The most frequent haemodynamically significant extracardiac arterio–venous shunt found in newborns is a vein of Galen malformation. However, it is not a frequent cause of severe cardiac failure in infancy. Effective treatment has not yet been found.

We report on a patient with a large vein of Galen malformation who presented with high-output cardiac failure, pulmonary artery hypertension and respiratory distress soon after birth. During an emergency cardiac catheterisation, we diagnosed a large vein of Galen malformation. The cardiac status was improved after partial embolisation.

Nine months later the echocardiographic finding was normal and further therapy was discontinued. Unfortunately, as in similar cases, hypertensive hydrocephalus developed. A ventriculoperitoneal shunt was placed but there were major neurodevelopmental delays.

676: THE CASE OF NEONATAL LUPUS SYNDROME: TRANSIENT COMPLETE CONGENITAL AV BLOCK IN A NEWBORN

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The congenital heart block in neonatal lupus is a form of passively acquired autoimune disease in which maternal auto-antibodies to the intracellular ribonucleoproteins Ro (SS-A) and LA (SS-B), cross the placenta and injure the foetal heart. Studies show that AV block in neonatal lupus is irreversible. The aim was to present a patient who had spontaneous recovery from complete heart block.

The patient was born after a normal, controlled pregnancy. In the 36th week of gestational age she was delivered by emergency caesarean section because of foetal bradycardia in a cardiotocographic recording, BW 2 750 g, Apgar score 9/9. After the birth, electrocardiography results showed complete AV dissociation with atrial frequency around 150 bpm and ventricular around 55–60 bpm. Echocardiography showed normal heart structure and function.

Serology for autoimmune diseases was performed and showed that the newborn had maternal anti-Ro-SSA and anti-La-SSB antibodies in the circulation. Anamnesis of the mother showed that she had years of dermatological treatment due to skin lesions on her legs, and occasional pain in the joints. We advised her to have rheumatology tests done. During her stay, the newborn had no cyanotic crisis and no signs of loosing heart function. The pulse rate ranged from 65–90 bpm and ECG showed a persisting complete atrioventricular block.

At the age of three months the patient was re-admitted to the clinic to run control tests. During the 24-hour ECG, she was in sinus rhythm, with an average frequency of 133 bpm. The lowest frequency was recorded while sleeping, at 67 bpm. There were no episodes of complete AV block or other arrhythmias. There was spontaneous recovery of the cardiac conduction system.

690: A RETROSPECTIVE REVIEW OF PALLIATIVE SYSTEMIC-TO-ARTERIAL SHUNTS IN A RESOURCE-SCARCE AFRICAN ENVIRONMENT

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Background: Due to the lack of cardiac surgical programmes undertaking corrective surgery in the neonatal period in resource-scarce African countries, the vast majority of patients presenting with cyanosis in the newborn period are palliated by a systemic-to-pulmonary arterial shunt.
Methods: We reviewed the case records of all patients who underwent systemic-to-pulmonary arterial shunting over a seven-year

period (2005–2011) at a tertiary cardiac surgical centre with a referral base of 14 million people.
Results: Ninety-eight patients underwent systemic-to-pulmonary arterial shunting over a seven-year period, of which only 23 ultimately underwent correction of their primary cardiac lesion. A significant number of patients were lost to follow up or died due to diarrhoea and respiratory disease.
Conclusions: In a resource-scarce environment, the primary palliation of cyanosed neonates with systemic-to-pulmonary arterial shunts has a sub-optimal long-term result. Resources should be put into the development of centres specialising in early neonatal corrective surgery.

696: A RETROSPECTIVE REVIEW OF HIV-INFECTED PATIENTS UNDERGOING CONGENITAL CARDIAC SURGERY IN KWAZULU-NATAL, SOUTH AFRICA

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Background: South Africa has one of the highest prevalences of HIV infection in the world (30%), with the perinatal HIV infection rate in KwaZulu-Natal over 40%. HIV-exposed neonates and infants undergoing congenital cardiac surgery (either palliative of corrective) in sub-Saharan Africa remain an unstudied population. We have formulated institutional guidelines based on clinical experience in order to manage these patients.
Methods: We reviewed the case records of all HIV-exposed patients who underwent congenital cardiac surgery over a seven-year period (2005–2011) at a tertiary cardiac surgical centre with a referral base of 14 million people.
Results: No significant difference in in-hospital morbidity or mortality rate was noted in HIV-exposed patients undergoing congenital cardiac surgery, compared to non-exposed patients with similar risk stratification. However, the late mortality and long-term outcome following surgery remains unclear.
Conclusions: While HIV-exposed patients undergoing congenital cardiac surgery have an acceptable in-hospital morbidity and mortality rate, late mortality (particularly related to opportunistic infections) requires further evaluation. In addition, patient access to and compliance with highly active antiretroviral therapy (HAART) is variable. In undertaking congenital cardiac surgery in HIV-positive patients, a sound knowledge of drug interactions, immunology, risks of mother-to-child transmission (MTCT) and the manifestations of opportunistic infections is required in order to formulate institutional guidelines.

743: SUSTAINED BIVENTRICULAR PACING MAY IMPROVE CARDIAC INDEX, BLOOD PRESSURE AND CEREBRAL BLOOD FLOW IN INFANTS WITH ELECTRICAL DYSSYNCHRONY AFTER SURGERY FOR CONGENITAL HEART DISEASE

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Background: Cardiac index (CI) decreases in the initial postoperative period after congenital heart disease (CHD) surgery, exposing infants to adverse haemodynamics. Electrical dyssynchrony, manifested by prolonged QRS duration (QRSd), may be an important factor. We hypothesised that continuous, simultaneous left and right ventricular pacing (BiVp) after CHD surgery improves cardiac index and haemodynamics in the early postoperative period.
Methods: We prospectively recruited infants with biventricular CHD who were under four months of age and undergoing surgery on cardiopulmonary bypass. Infants were randomised, regardless of QRS duration, to receive standard of care or standard of care + BiVp for 48 postoperative hours or until extubation, if sooner. Infants randomised to BiVp received atrial, right ventricular and left ventricular leads. Continuous BiVp (atrial tracking) was initiated upon return to ICU. Haemodynamics were assessed at least every three hours for the first 24 hours and at least every six hours thereafter for up to 48 hours. CI was measured by Fick using mass spectroscopy for oxygen consumption. Near-infrared spectroscopy was used to measure cerebral saturation. Primary outcome was change in CI over the first 48 postoperative hours.
Results: Forty-two infants (21 controls, 21 Bivp; 3.8 ± 0.9 kg, 50% transposition of the great arteries) were randomised. Infants with prolonged baseline QRS (> 98th percentile) experienced decreased CI, which was prevented by BiVp. Controls with normal QRS durations for age had consistent improvement in CI in the first 48 hours, while those with prolonged QRS durations showed delayed CI recovery. BiVp tended to improve CI recovery [+0.118 (0.374) ml/min/m^2, p = NS], blood pressure [BIVP +8 (3) mmHg, p = 0.01] and cerebral blood flow in infants with wide QRS, but did not improve CI in infants with narrow QRS.
Discussion: Continuous BiVp may be useful to improve CI, blood pressure and cerebral flow in infants with electrical dyssynchrony after CHD surgery.

756: COMPLICATIONS AND OUTCOMES OF CHYLOTHORAX FOLLOWING CARDIAC SURGERY

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Background: To review the experience of post-cardiac surgery chylothorax at our institution over a six-year period, with emphasis on complications and mortality.
Methods: Episodes of postoperative chylothorax from 1 June 2005 to 31 May 2011 were identified from intensive care and cardiac surgical databases. Demographic, operative, haemodynamic, laboratory and outcome data were collected from the medical record and electronic databases. A multivariate model was constructed to define predictors of death.
Results: One hundred and sixteen episodes of chylothorax were confirmed, yielding an incidence of 2.2% in postoperative patients. The median age of patients was 57 days (range one day – 16 years). One-third of episodes occurred following one of the three stages of single-ventricle palliation. The highest specific incidence occurred following neonatal repair of anomalous pulmonary venous drainage (17.4% of such operations). Maximum daily chyle drainage (median 46 ml/kg/day, 2–617) was negatively correlated with age and positively with duration of chylothorax. Initial feeding was parenteral in 49.5%, and enteral in 50.5%. Patients treated with parenteral nutrition at any time had higher mean daily drainage ($p < 0.001$). Laboratory consequences of protein loss were common: lymphopaenia 92.2%; hypoalbuminaemia 87.1%; hypoproteinaemia 85.3%; hypogammaglobulinaemia 67.2%. Thirty-one per cent developed sepsis; these patients were younger and had lower immunoglobulin levels than those who did not develop sepsis. Median ICU stay was 10 days with a median hospital stay of 116 days. The mortality rate was 15.6%, with a median time to death from the day of surgery of 68 days (9–296). The lowest blood lymphocyte count and the lowest serum albumin were independent predictors of death.
Conclusions: Chylothorax is an infrequent complication of cardiac surgery but is associated with high morbidity and mortality. Minimum blood lymphocyte count and serum albumin are independent predictors of death in patients with this complication.

758: PERIODONTAL DISEASE IN CHILDREN WITH CONGENITAL HEART DISEASE AND ITS CORRELATION WITH LEVELS OF LDL CHOLESTEROL

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Hypothesis: The number of children and adults with congenital heart disease has increased in recent years, but there are few studies correlating changes in oral mucous and the mouth in this patient group. Moreover, recent research in adults has shown that a relationship exists between oral cavity diseases, especially periodontal infections and systemic diseases, including atherosclerosis. The aim of our study was to correlate the levels of LDL cholesterol in patients with congenital heart disease and periodontal disease.
Methods: We evaluated 33 patients with congenital heart disease (group I: average age of nine years and seven months) and compared with a control group of 28 patients without cardiac disease (group II: average age nine years and eight months). We analysed the clinical periodontal parameters of plaque index, clinical attachment level and bleeding on probing and blood level of LDL cholesterol.
Results: The LDL cholesterol levels were slightly higher in the control group (group II: 87.23 ± 23.94 mg/dl) than in the cardiac group (group I: 85.49 ± 19.83 mg/dl), with enough similarity between the groups regarding clinical attachment level (group I: 1.91 ± 0.31 mm, group II: 1.93 ± 0.28 mm). Only when there was an analysis of patients with LDL level higher than expected in theses groups, was the level of clinical attachment greater in group I (2.11 ± 0.31 mm) than group II (1.84 ± 0.32 mm).
Conclusions: Patients with congenital heart disease followed up in our service have improved plaque control compared to the control group, but the more periodontal disease is developed in this group of patients, the more the LDL cholesterol levels can be changed. As the percentage of hypercholesterolaemic patients was 24.59%, we believe that to better assess this correlation in paediatric patients, further studies should be performed.

761: SIMULTANEOUS REPAIR OF PECTUS EXCAVATUM AND CONGENITAL HEART DEFECTS: A GOOD SURGICAL OPTION OF A ONE-STAGE PROCEDURE

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Background: Simultaneous repair of pectus excavatum and congenital heart defect is highly efficient and reduces to one the number of operative procedures in children.
Methods: Case reports are presented of two patients operated on in a one-stage procedure consisting of a simultaneous pectus excavatum correction and congenital heart defect repair.
Results: Between 2007 and 2010, two patients (seven years old) received a simultaneous pectus excavatum repair with a modified Ravitch technique and congenital heart defect repair by a multidisciplinary team of cardiac and thoracic surgeons. Both patients received a transverse linear submammary incision. Deformed cartilages were sub-perichondrially bilaterally resected and the sternum was lifted anteriorly. Through a pericardiotomy the cardiac lesion was fixed. One patient received an aortic valve-sparing and ascending aorta replacement (Tirone David operation) for a severe aortic insufficiency and aortic anuloectasic disease. In the second patient an atrial septal defect was repaired with an autologous pericardial patch. After the cardiac defect repair, the sternum was positioned back to the original position, the pectoral muscle was fixed to the sternum, the xiphoid was fixed to the abdominal rectus and subcutaneous tissue, and the skin was closed in a regular way. A vacuum system and a pericardial tube were placed subcutaneously. Patients were ventilated for 13 hours (12–13) in order to avoid tension during the first hour of the post-operative period. The intensive care stay was 2.5 days (2–3). No complications or mortality were observed. Aesthetic results were good after 23 months of follow up, and the cardiac repair is still satisfactory.
Conclusions: Simultaneous repair of the pectus excavatum and congenital heart defect is a good option for treatment since it reduces to one the operative event, there is no increased risk of complications, the patients tolerated it well, and the short-term results were good.

773: THE KONNO AORTO-VENTRICULOPLASTY PROCEDURE IS AN EXCELLENT ALTERNATIVE TO RELIVE SEVERE LEFT OUTFLOW TRACT OBSTRUCTION AT ALL AGES

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Background: Aortic root enlargement (ARE) procedures are believed to allow implantation of larger valve prostheses; however, little evidence exists to support the specific efficacy of various techniques. The Konno procedure is the best technique used to enlarge the aortic root and increase the size of the aortic valve implanted.
Methods: This was a case series of six patients: three children and three adults from January 2009 to March 2012. Patients had aortic root enlargement surgery, 'Konno aortoventriculoplasty'. We present continuous variables in means or medians and SD or IQR, and categorical variables in absolute and relative frequencies; paried *t*-test was used to compare continuous pre- and postoperative haemodynamic variables.
Results: Median age was 19.5 years (3–56) and 57.1% (4/7) were women; 83.3% (5/6) of patients had symptoms at the diagnosis. All patients had a previous cardiac procedure. The underlying anatomical diagnoses were valve and subvalvar aortic stenosis in two, subaortic fibromuscular tunnel with moderate aortic stenosis in two, severe aortic stenosis and insuficiency in one patient and dehiscence of a previous Konno operation in one. Mean ejection fraction 65.7 ± 16.9%, aortic valve peak gradient mean was 74.2 ± 61.1 mmHg and mean aortic ring diameter was 17.7 ± 3.4 mm. Mean aortic clamp time was 107.6 ± 20.0 minutes; 83.3% (5/6) of patients received a new mechanic valve. There were no mortalities, one re-operation for bleeding, and one new complete AV block. Postoperative aortic valve peak gradient mean was 22.1 ± 7.3 mmHg ($p = 0.08$) and ejection fraction was 55.5 ± 15.3 (I = 0.08). At the follow up all patients were alive.
Conclusions: Konno aorto-ventriculoplasty is a safety operation. Excellent results may be achieved despite previous aortic root enlargement procedures, and it may even reduce the risk of re-operation in children by allowing placement of a larger prosthesis.

779: CONSTRUCTION AND VALIDATION OF AN INSTRUMENT TO DETERMINE THE DEGREE OF UNDERSTANDING OF POSTOPERATIVE CARE ON CAREGIVERS OF PAEDIATRIC PATIENTS UNDERGOING CARDIOVASCULAR SURGERY

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Introduction: The importance of postoperative care is widely described in the literature, however there is no published scale that measures the level of understanding of the information given by the nursing service to patients and caregivers in the postoperative period of cardiac surgery.
Objective: To develop and validate an instrument to measure the degree of understanding of postoperative care in cardiovascular surgery given by the nursing service of the Fundación Cardioinfantil to caregivers of paediatric patients undergoing cardiac surgery.
Methods: This was a cross-sectional study conducted between August and November 2011. The nursing team carried out the construction of the instrument based on the instructions of postoperative care following the guidelines of a self-care model of Dorothea Orem. The instrument has five categories: (1) appointment with a cardiovascular surgeon, (2) daily activities, (3) drug administration, (4) endocarditis prevention, (5) warning signs. Constructive validation was performed by 10 experts in cardiovascular medicine. We performed content validity and reliability through Cronbach's alpha test statistic, calculated in Stata 8.0.
Results: One hundred caregivers were evaluated and 92% (92/100)

were female. The instrument was applied by a nurse from the service after receiving information on postoperative care as service protocol. The instrument was applied 30 to 60 minutes after surgery. The average time of application was 4 ± 2.3 minutes. Content validity was 0.85, reliability 0.70.
Discussion: The construction and validation of an instrument to assess the degree of understanding of postoperative care in cardiovascular surgery found that educational interventions made by the nursing service provided clear concepts for achieving patient care at home.

780: IMPROVEMENTS IN RESULTS OF NEONATAL AND INFANT CARDIAC SURGERY: A JOURNEY AT A SMALL CENTRE

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Background: Mortality in congenital cardiac surgery (CCS) in centres worldwide is very low. Every new centre has to match the benchmark in quicker time. This requires a team effort. We describe our efforts to improve the results of CCS at our centre.
Methods: Our centre started CCS for the first time in central India. The surgeon carried out the roles of cardiologist, surgeon and intensivist. The anesthesiologist carried out the roles of anesthetist, perfusionist and intensivist. We operated on 350 patients with congenital heart diseases. In the initial period, we started with hypothermic perfusion, blood prime, cold-blood cardioplegia, conventional and modified ultrafiltration. We did intra-operative epicardial echocardiography to confirm proper surgical correction. Our intubation times, re-intubation rates and tracheostomy rates were high. After a visit to two centres, we modified our prime by increasing the albumin content and changing the modified ultrafiltration circuit. We introduced bubble CPAP (continuous positive airway pressure) as a step down from mechanical ventilation in neonates and infants less than 4 kg weight. We improved our neonatal nursing care with emphasis on hand hygeine.
Results: We present our results on the different types of surgery.
Conclusions: Our mortality rate for patients with weight < 8 kg was significantly higher but there was significant improvement in our results after modifications were done in our practice.

785: MODIFIED SINGLE-PATCH REPAIR OF COMPLETE ATRIO-VENTRICULAR SEPTAL DEFECT IS PERFORMED MORE EFFICIENTLY WITH NO DETRIMENTAL EFFECT ON LEFT VENTRICULAR OUTFLOW SIZE AND ATRIO-VENTRICULAR VALVE COAPTATION RESERVE

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Objectives: Concerns have been raised about left ventricular outflow tract (LVOT) narrowing and increased left atrio-ventricular valve regurgitation (LAVV) following atrio-ventricular septal defect (AVSD) repair with a modified single-patch technique. Therefore, we sought to compare the effects of modified single and two-patch surgical repair of complete AVSD on the LVOT diameter and the LAVV coaptation.
Methods: We retrospectively reviewed postoperative two-dimensional echocardiograms of all AVSD patients who underwent modified single- or two-patch repair between 2005 and 2011. We measured leaflet coaptation length and tenting height of the LAVV in the apical four-chamber view. The LVOT was measured in the long-axis view.
Results: Fifty-one patients underwent AVSD repair (single patch, n = 29, two-patch, n = 22) with 46 having adequate images for analysis. Five patients were re-operated after single-patch repair [three residual ventricular septal defect (VSD) and LAVV regurgitation, one residual VSD and right AVV regurgitation, one pacemaker implantation]. One patient after two-patch repair had re-operation for a residual VSD. The difference in re-operation rates was not significant (p = 0.22). Patient characteristics, LVOT and leaflet findings are summarised
Conclusions: The modified single-patch repair was performed with significantly shorter bypass and myocardial ischaemic time, without significant difference in post-operative LVOT diameter or LAVV leaflet coaptation length. Further investigation of re-operation rates may be warranted.

802: RISK OF PROCEDURAL SEDATION AND ANAESTHESIA IN CHILDREN WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION

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Background: Children with idiopathic pulmonary arterial hypertension (IPAH) are at high risk of complications while undergoing general anaesthesia (GA) or procedural sedation (PS). We aimed to determine the incidence of related complications.
Methods: A retrospective review was carried out on patients with IPAH undergoing GA or PS between 1980 and 2012, at a single tertiary paediatric centre. Data collected included measures of disease severity, clinical management and complications occurring within 30 days from GA or PS. Era of management (1980–1999 and 2000–2012) was based on availability of PAH specific therapies. Major complications were defined as need for cardiopulmonary resuscitation or death within 30 days.
Results: A total of 26 patients (15 current and 11 historic) underwent intervention with GA or PS. Of these, 11 patients, median age at diagnosis 11.2 years (IQR 5.6, 11.9) underwent 17 procedures at median age 9.6 years (IQR 7.5, 11.9) in the historical era. In the current era, 15 patients, median age 6.9 years (IQR 4.8, 11.3; p = 0.5) underwent 59 procedures at median age 11.3 years (IQR 7.26, 14.44; p = 0.45). Duration of follow up was limited in the historical control group to median 0.75 years (IQR 0.1, 2.77) compared to the current era of 5.1 years (IQR 3.1, 9.6; p = 0.006). Historic patients underwent an average of 1.55 procedures vs 3.93 for current patients (p = 0.01). Severity of PAH and WHO functional class were similar between the two eras. Major complications occurred in 3/59 (5%) procedures in the current era compared to 7/17 (41%) procedures in the historic era (p < 0.002). Nine of 10 patients with complications had features consistent with acutely elevated pulmonary arterial pressures. Three of the four deaths occurred under GA and one after PS.
Conclusions: Children with IPAH undergo more interventions using GA or PS in the current era. The risk of major complications has reduced significantly but remains unacceptably high in this vulnerable patient group.

805: EVALUATION OF NUTRITIONAL SUPPORT DELIVERY IN CRITICALLY ILL INFANTS AND CHILDREN ON EXTRA-CORPOREAL MEMBRANE OXYGENATION

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Background: Early nutritional support (NS) is an essential component of care in the paediatric intensive care unit (PICU), particularly for patients on extra-corporeal membrane oxygenation (ECMO), where considerable nutritional and metabolic burdens exist. Our study is the first to systematically audit NS within a paediatric ECMO population and proposes the hypothesis that NS is suboptimal in this patient group.
Methods: A retrospective audit was conducted in all patients receiving ECMO between October 2008 and August 2011 in our tertiary care PICU. Medical records were reviewed to collect data, including:

demographics, ECMO type, nutritional fluids, and hours to NS initiation. Protein and calorie delivery were calculated and compared to international consensus guidelines or age-specific recommendations where available.
Results: Fifty-five subjects were analysed, 42 (76%) cardiac and 13 (24%) respiratory ECMO patients. Five (9%) patients received no NS during their ECMO period, 85% of patients received NS within 24 hours of starting ECMO, with a mean time (± SD) to feed initiation of 8 ± 17 hours. In all age categories, recommended target calorie and protein requirements were not met. There was no significant difference in protein ($p = 0.4$) and calorie ($p = 0.08$) delivery within or between any age group. However, 'respiratory' ECMO patients received significantly more calories than the 'cardiac' group (44 ± 18 vs 29 ± 23 kcal/kg, $p = 0.04$). Furthermore, enteral nutrition (EN) delivered significantly less protein (0.55 ± 0.39 g/kg) than parenteral nutrition (PN) (1.28 ± 0.55 g/kg, $p = 0.02$) and combined PN and EN (1.13 ± 0.67 g/kg, $p \leq 0.01$).
Conclusion: We have shown that despite early feed initiation, the delivery of NS is suboptimal in paediatric patients on ECMO. Future research focusing on prioritising and optimising NS practice, with emphasis on EN as the preferred feeding route in this population should be considered.

818: AN INITIAL EXPERIENCE OF SURGICAL CORRECTION OF TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE IN A NEW PAEDIATRIC CARDIAC SURGERY CENTRE

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Background: A low mortality rate with surgery for total anamolous pulmonary venous drainage (TAPVD) is reported worldwide these days, but there has been controversy regarding ligation of the vertical vein in supracardiac and infracardiac TAPVD, and keeping a small PFO. We report our small initial series of 20 patients.
Methods: We operated on 20 patients for TAPVD from August 2009 to July 2012. A standard protocol of moderate hypothermia, blood prime, blood cardioplegia, and conventional and modified ultrafiltration was used in these surgeries. We ligated the vertical vein in three of the six supracardiac type of TAPVD patients and one infracardiac type of TAPVD patient. A small PFO was left in 17 patients.
Results: We present our results on the surgeries.
Conclusions: We preferred keeping a small PFO in our patients, especially those who had severe RV dysfunction due to late presentation and repeated chest infection. These patients had persistent right-to-left shunt across the PFO for a few postoperative days, which then reversed to left-to-right shunt. The vertical vein was ligated only if patients had stable haemodynamics post-operatively. Our high mortality might be due to lack of nitric oxide at our centre and also due to the lack of paediatric cardiologists and intensivists (SSD joined recently). The primary surgeon (SDK) performed echocardiography and managed the ICU as well.

857: A SUCCESSFUL BEATING-HEART SURGERY IN CONGENITAL HEART DISEASE IN INDONESIA

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Background: Reperfusion injury is a well-known phenomenon that can occur in cardioplegic techniques with cardiopulmonary bypass. Therefore great effort is made to prevent reperfusion injury. Beating-heart continous coronary perfusion (BHCCP) surgery is one of the alternative technique to improve an ischaemic reperfusion injury in open-heart surgery, in either paediatric or adult patients. It is divided into antegrade and retrograde perfusion. Keeping the heart beating results in less myocardial oedema and better myocardial function.
Methods: We report on eight patients with congenital heart disease (from December 2011 to June 2012). Five patients suffered from secundum ASD, one with VSD, and two patients with TOF. Pre-operative diagnoses were established by echocardiography and cardiac catheterisation if necessary. All the patients underwent BHCCP surgery. Echocardiographic examinations play a significant role in evaluating the heart immediately after surgical repair.
Results: All the patients who underwent correction using BHCCP techniques showed good outcomes such as reduced ventilator time (mean ± 10 hours) and length of stay in ICU (mean ± 2 days), and less inotropes. Our results indicate that BHCCP surgery is a safe and reliable technique for treatment of cardiac diseases and it is a good option in patients with poor LV function.
Conclusion: After the two-month follow up, all of our patients showed no significant complications. There were no mortalities, no neurological deficit, and only one patient with ToF showed a 2-mm residual VSD.

871: PRETREATMENT BEFORE CORONARY ARTERY BYPASS SURGERY IMPROVES POSTOPERATIVE OUTCOMES IN MODERATE CHRONIC OBSTRUCTIVE PULMONARY DISEASE PATIENTS

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Hypothesis: The aim of this study was to analyse the impact of different COPD stages on the early surgical outcomes in patients undergoing primary isolated non-emergency CABG.
Methods: According to the protocol applied by the Department of Pulmonary Diseases, two different treatment protocols were used before and after 2010. Before 2010 no treatment was applied to patients with moderate COPD before the CABG procedure. After 2010 the pretreatment protocol was begun. Due to this change in pre-CABG treatment protocol, we organised the study groups. Patients who underwent surgery between 2008 and 2010 formed group 1 (no pretreatment group, $n = 51$) and patients who underwent surgery between 2010 and 2012 formed group 2 (pretreatment group, $n = 53$). These two groups were compared according to the postoperative morbidity and mortality rates, retrospectively from medical reports.
Results: Mean age of the patients in both groups were 62.1 ± 7.6 and 64.5 ± 6.4 years, respectively. Mean Euroscores of the patients were 5.5 ± 2.3 and 5.9 ± 2.5, respectively in the two groups. Average number of grafts were 3.1 ± 1.0 and 2.9 ± 0.9. Mean extubation times were 8.52 ± 1.3 hours in group 1 and 7.41 ± 1.1 hours in group 2 ($p < 0.05$). The number of patient who needed pharmacological inotropic support were 12 in group 1 and five in group 2 ($p < 0.05$). Duration of hospital stay of the patients was also shorter in group 2 (9.29 days, $p < 0.05$). While there were seven patients who had pleural effusions requiring drainage in group 1, there were only two requiring drainage in group 2 ($p < 0.05$). There were no in-hospital or early mortalities in either group.
Conclusion: Pretreatment in moderate-risk COPD patients improved postoperative outcomes while decreasing the adverse events and complications. Therefore in patients undergoing elective CABG we recommend the use of medical treatment.

883: FLUID DYNAMICS AND FLOW PROFILES IN THE GREAT ARTERIES IN TGA PATIENTS AFTER ARTERIAL-SWITCH OPERATIONS WITH OR WITHOUT LECOMPTE MANOEUVRE ON LONG-TERM FOLLOW UP

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Background: The fluid dynamics (shear stress and vorticity) of the blood in the transposed great arteries in TGA patients after arterial-switch operations (ASO) with spiral course or anterior branching of the pulmonary arteries (Lecompte) is unknown. The aim of this study was a comprehensive assessment of blood flow profiles in the great arteries utilising advanced MRI techniques two decades after the ASO.
Methods: Twenty-four TGA patients (Lecompte: n = 12, 19.2 ± 3.9 years post ASO; non-Lecompte, spiral: n = 12, 24.1 ± 4.3 years post ASO) were studies with high-field MRI at 3 Tesla. All patients underwent a comprehensive cardiovascular MRI, including anatomical and functional cardiovascular evaluation. Additionally, 13 patients and five healthy volunteers received time-resolved 3D flow measurements (4D flow) using novel phase-contrast MR techniques (FOV 250–337 mm^2, venc 150 cm/s in three orthogonal directions, true spatial resolution: 2.5 mm^3 isotropic, temp. resol 35 ms, TR/TE 4.6/3.2; α 5–10°). Dedicated software was used for colour-coded 4D visualisation of blood flow profiles and streamlines (GT-Flow™, Gyrotools Inc, Zurich). Fluid dynamics were calculated with in-house customised software.
Results: In patients with a spiral course (non-Lecompte) of the great arteries, vorticity index and shear stress were more favourable compared to the Lecompte group (aorta: 234.51 ± 35 vs 289.36 ± 24 m^2/s, pulmonary artery: 72.54 ± 15 vs 93.53 ± 13 m^2/s; $p < 0.01$, respectively; aorta: 0.35 ± 0.14 vs 0.54 ± 0.21 N/m^2, pulmonary artery: 0.31 ± 0.09 vs 0.42 ± 0.17 N/m^2; $p < 0,01$). In both groups we found one patient with an occluded left coronary artery. A sinus valsalva aneurysm was present in the non-Lecompte group.
Conclusions: On long-term follow up, a spiral course of the great arteries in TGA patients post ASO showed more physiological blood flow dynamics compared to anterior branching of the pulmonary arteries (Lecompte). Therefore, in elegible patients, a spiral course should be considered.

899: EXTRACELLULAR MATRIX GRAFT FOR CARDIAC AND VASCULAR RECONSTRUCTIVE SURGERY: PRELIMINARY CLINICAL RESULTS

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Background: Porcine extracellular matrix graft (Cormatrix® ECM) has been widely used in clinical setting to repair the pericardium but no reports exist to date showing its use and efficacy in vascular and cardiac repair.
Aim: To evaluate Cormatrix® ECM material as a possible patch for clinical use in reconstructive cardiac surgery.
Methods: We reviewed the report of the last clinical and echocardiographic evaluation of 23 patients affected by Tof, PV stenosis isolated or in association with other CHD, AS, PAPVD, other CHD, operated on at our Institution since December 2009, in whom Cormatrix® ECM patch was used during reconstructive surgery. We included in our evaluation only patients in whom Cormatrix® ECM patch was employed for RVOTor PV/PAs reconstruction, with at least one echo examination after discharge home.
Results: Fourteen patients met the enrolment criteria: affected by ToF, 10 cases; DORV sub-pulmonary VSD + subAoS + AoCo, complete A-V septal defect, PS and RV hyoplasia, asplenia syndrome + single RV + PS, one case each. Median age at operation was 4.8 months (1–66). Cormatrix® ECM patch was used for RVOT reconstruction in ToF patients, PV reconstruction in two cases, PAs patch augmentation in two. At a mean follow-up of 15.3 months (2–24) severe RVOT stenosis requiring treatment was present in two ToF patients, one in association with signs of persistent inflammation and pulmonary haemorrhage requiring pneumonectomy. Absent or trivial pulmonary regurgitation was recorded in two ToF, mild in two, moderate in four, in one associated with moderate stenosis, the last four with RV dilatation. One patient each in the group of PV reconstruction and PAs augmentation presented severe recurrent stenosis, requiring haemodynamic manoeuvres.
Conclusion: On short-term follow up, the performance of the Cormatrix® ECM patch seemed suboptimal. Further clinical experience and longer follow up are needed to judge the adequacy of this material.

911: TREATMENT OF PULMONARY ARTERIAL HYPERTENSION IN PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Pulmonary arterial hypertension with Eisenmenger syndrome as its most advanced form is an important complication of congenital heart disease. In recent years advanced therapy for pulmonary arterial hypertension has been introduced. Efficacy and safety of the advanced therapy in our patients with pulmonary arterial hypertension associated with congenital heart disease was analysed.
Methods: We analysed results of advanced therapy in patients treated between November 2007 and December 2011. Clinical status, systemic oxygen saturation measured by systemic pulse oximetry, six-minute walk distance and laboratory parameters were assessed. Results at three, six, 12 and 24 months on the treatment were compared to baseline parameters.
Results: In the observed period, 23 patients were treated with PAH-specific therapy. As a first-line drug, bosentan was used in 19 and sildenafil in four patients. Due to clinical worsening a second- and a third-line drug had to be added on during a study period in four and one patient, respectively. Eighteen patients (78.3%) reported improvement in functional capacity. Two patients (8.6%) died. The mean six-minute walking distance significantly increased over time from 334.7 ± 87.7 m at baseline to 348.5 ± 89.1 m at three months (p = 0.002), 373.2 ± 74.4 m at six months (p = 0.005), 383.2 ± 62.3 m at 12 months (p = 0.017) and 396.3 ± 92.8 m at 24 months of treatment. No significant adverse events were reported.
Conclusions: Advanced therapy for pulmonary arterial hypertension was beneficial in patients with congenital heart disease. Significant improvement in exercise capacity was observed. The therapy is safe and no significant adverse effects were reported.

935: ADJUSTABLE BILATERAL PULMONARY ARTERY BANDING AIMING AT STAGED BIVENTRICULAR REPAIR FOR INFANTS WITH UNBALANCED VENTRICLES AND COARCTATION OF THE AORTA

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Background: Bilateral pulmonary artery banding (bPAB) is a surgical option for infants with unbalanced ventricles and coarctation of the aorta (CoA), who are borderline candidates for biventricular repair. However, it is not easy to know the optimal length of the bands in early infancy, because we have to consider their growth and changes in pulmonary vascular resistance during follow up. We applied a unique treatment strategy which consisted of adjustable bPAB using pliable ePTFE bands combined with staged percutaneous balloon dilation on the banding sites for three critical infants. We report one of the cases who had been successfully treated based on this strategy.
Case report: A newborn girl was diagnosed as having unbalanced complete atrio-ventricular defect (cAVSD), CoA and hypoplastic aortic arch, associated with 21-trisomy. The small LV volume and

the higher pulmonary vascular resistance related to chromosomal anomaly were difficult issues, to be discussed in the initial treatment. Subsequently, we performed bPAB using handmade banding tapes, made of double-layered ePTFE membrane, 0.1 mm in thickness, on the 10th day of life. The bands were tightly fixed using 7-0 polypropylene sutures. The lengths of the bands were 9.0 and 9.0 mm, respectively. As she gained weight under medical treatment with continuous lipo-prostaglandin E1 infusion, her saturation had gradually declined to 70%. Percutaneous balloon dilation was performed to increase pulmonary blood flow using 3.5- and 4.0-mm balloon catheters at 80 and 133 days old. At 169 days old, CoA repair was carried out via a left thoracotomy. She had repeatedly undergone balloon dilation until she reached a definitive operation. Catheter study at one year after bPAB showed the adequately increased LV volume. Thereafter she underwent biventricular repair with a successful outcome. No peripheral pulmonary arterial patch angioplasty was required.
Conclusion: Our adjustable bPAB strategy provides great benefits for treatment of borderline infants for biventricular repair.

939: SINGLE-PORT SUBXIPHOID APPROACH WITHOUT FEMORAL CANNULATION FOR ASD CLOSURE
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Aim: To present the technique and results of the single-port subxiphoid approach without femoral cannulation for ASD closure in children.
Methods: ASD closure can be performed through a mini-sternotomy or subxiphoid approach with a small skin incision. Femoral arterial cannulation is generally used to minimise the length of the incision. We present our experience with a small-incision (average 2 cm) subxiphoid approach for ASD closure without femoral cannulation. We established CPB with an aortic cannula and venous cannulation with RA and IVC cannulas. We used a reinforced arterial cannula, a right-angled reinforced venous cannula for IVC, and straight reinforced cannula for RA. Core cooling was done to 32°C. ASD closure was done under fibrillatory arrest. We used an autologous pericardial patch for ASD closure in the majority of patients. After de-airing the LA, the fibrillator was removed. An internal defibrillator was used if the heart did not pick up sinus rhythm spontaneously. After rewarming, the patient was weaned off CPB.
Results: We perfomed single-port subxiphoid ASD closure in 137 patients over three years from May 2009 to April 2012. In that time, 73 patients were female and 64 were males. The mean CPB time was 41 min. The mean fibrillatory arrest time was 10 min. In three patients, it was converted to full sternotomy. The mean length of the skin incision was 2 cm. The mean ventilation time was 83 min. All patients had a very short ICU and hospital stay.
Conclusion: The single-port subxiphoid ASD closure without femoral cannulation can be performed in all patients efficiently and safely and the technique is reproducible.

942: RECURRENT RESPIRATORY PAPILLOMATOSIS COMPLICATED BY INTRACARDIAC EXTENSION
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Intoduction: Intracardiac masses are rare in children. Normal structures and variants may mimic a cardiac mass. Abnormal cardiac masses commonly include tumours, thrombi and vegetations.
Case report: We present an unusual case of recurrent respiratory papillomatosis with malignant progression and intracardiac extension. An eight-year-old male presented with a chronic history of recurrent respiratory papillomatosis involving the larynx initially but which later spread distally to involve the trachea and bronchi. He subsequently developed bronchiectasis. He has had regular ENT consultations and required laser treatment for the papillomas; in addition he had a tracheostomy at a young age. His acute presentation was a threatened right upper limb due to right axillary artery and brachial artery occlusion. Echocardiography demonstrated a mass within the left atrium. Despite anticoagulation, antibiotics and antifungal treatment, this mass progressively enlarged and further proceeded to embolise to the distal aorta, resulting in an acutely threatened lower limb. Surgical resection of the intracardiac mass was undertaken after the second embolic event and at surgery the mass was noted to arise from a pulmonary vein. The final diagnosis of the intracardiac mass was made on histology, which showed a well-differentiated squamous cell carcinoma which had arisen in the context of the antecedent history of human papilloma virus-induced laryngeal and bronchopulmonary papillomatosis.
Conclusion: The above confirms that intracardiac tumours in the paediatric population are more likely to be metastatic. Even though echocardiography permits dynamic evaluation of intracardiac masses, allowing delineation of the anatomical extent and the physiological consequences of the mass, histology provides the definitive diagnosis.

949: CONGENITAL CARDIAC ANAESTHESIA DATABASE RESULTS 2010–2011
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The Congenital Cardiac Anesthesia Society (CCAS) has partnered with the Society of Thoracic Surgeons (STS) to include fields relevant to our speciality as part of the STS Congenital Surgery database. This cooperative effort started in January 2010. Since that time, participation has grown to include data from over 30 sites in the United States. The locations include representation from a wide range both geographically and in programme size. The Spring 2012 harvest, encompassing 1 January 2010 to 31 December 2011, includes data from 20 226 discrete anaesthetics; 13 796 of these cases were cardiac surgical (CPB, no CPB, support devices), 3 354 were from the Cardiac Catheter Lababoratory and 3 076 were thoracic procedures, minor procedures or non-CV/non-thoracic on CV patients requiring CV anaesthesia (such as G-tube placement or Ladd's procedure). Data are being harvested on a wide variety of anaesthesia topics such as airway and medication management, monitoring modalities and anaesthesia-related adverse events. The overall adverse event rate was 2.1% and ranged from relatively minor (line placement requiring more than one hour) to severe (cardiac arrest unrelated to surgical events). The overall adverse events are detailed in the presentation.

950: SURGICAL REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION IN EMERGING ECONOMIES: ARE GOOD OUTCOMES POSSIBLE *SANS* INHALED NITRIC OXIDE AND ECLS?
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Background: Peri-operative management of sick infants with total anomalous pulmonary venous connections (TAPVC) remains a challenge, especially in emerging economies where many patients present with unstable haemodynamics. Post-operative pulmonary hypertension (PH) and secondary low-cardiac output state (LCOS) may be refractory, needing expensive therapeutic modalities such as inhaled nitric oxide (iNO) and or mechanical support.
Objective: To evaluate the early outcome of TAPVC repair without recourse to mechanical support or inhaled nitric oxide.

Methods: A prospective observational study was carried out from 2001 until June 2012; 203 patients underwent re-routing of pulmonary veins for TAPVC during this period. The median age was 89 days (1 day – 34 years); 127/203 had varying degrees of obstruction at different levels (supracardiac: 76, cardiac: 25, infradiaphragmatic: 17, mixed: nine). Five of 203 were re-do surgeries for obstruction at the anastamotic site; 42/127 obstructed TAPVC presented with circulatory collapse needing pre-operative resuscitation. No patient was refused surgery. Management strategies included (1) urgent surgery irrespective of haemodynamic status, (2) quick and accurate surgery, (3) deferred sternal closure, (4) epicardial echo in all to confirm accuracy of repair. Peri-operative pulmonary hypertension and associated low cardiac output was managed pre-emptively using multiple simple, inexpensive conventional strategies (intra-operative ultrafiltration, fluid restriction, peritoneal dialysis, lung recruitment, milrinone, calcium infusion, corticosteroids in refractory cases and elective non-invasive ventilation after extubation) with limited use of catecholamines.
Results: In-hospital mortality was four out of 203 (1.97%). There were two late deaths after hospital discharge due to non-cardiac causes. Median ventilatory requirement was 52 hours and median length of stay 11.2 days.
Conclusion: Successful re-routing of TAPVC in sick infants with pulmonary hypertension and severe right ventricular dysfunction is feasible without ECLS or inhaled nitric oxide, with minimum morbidity.

953: COMPARISON OF THREE INOTROPIC STRATEGIES IN THE NEWBORN AFTER STUNNING OF THE RIGHT VENTRICLE

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The immature myocardium has significantly different beta-receptor kinetics, metabolism and enzyme activity from the adult. We therefore undertook a piglet study to investigate the efffect of three different inotropic strategies.
Methods: Twenty-eight piglets aged four days were prepared, to measure cardiac output (CO), and central venous and arterial pressures. Stunning of the right ventricle (RV) was induced by 10 cycles with ischaemia–reperfusion injury. Animals were randomised to one of three inotropic protocols or placebo: (1) AM: adrenaline: 0.09 $\mu g.kg^{-1}min^{-1}$ and milrinone: 50 $\mu g.kg^{-1}$ bolus and 0.4 $\mu g.kg^{-1}min^{-1}$, (2) DM: dopamine: 6 $\mu g.kg^{-1}min^{-1}$, milrinone: 50 $\mu g.kg^{-1}$ bolus and 0.4 $\mu g.kg^{-1}min^{-1}$), (3) Dob: dobutamine (8 $\mu g.kg^{-1}min^{-1}$), (4) saline (2 $ml.kg^{-1}h^{-1}$). One-way ANOVA with Tukey's multiple comparison test was used to test differences between groups.
Results: Cardiac output (CO) had decreased by 29% in the placebo group 60 min after RV stunning. CO was significantly higher in the AM- and DM-treated animals, compared to placebo ($p < 0.05$), whereas Dob-treated animals remained unchanged. CO in DM-treated animals was significantly higher compared to that in Dob-treated animals. MAP was maintained in the DM animals, but decreased by more than 35% in AM- and Dob-treated and placebo animals after RV stunning. MAP decreased after 180 min in AM- (–37%) and Dob- (–44%) treated groups, and to the same extent in the control group (–41%) (ns). In the DM group, MAP remained stable throughout the observation period (–5%), in contrast with the other intervention groups ($p < 0.001$). SVRI decreased 14% ($p = 0.1$) during I/R. During the observation period SVRI decreased further by approximately 46% ($p = 0.01$) with no difference between control and any intervention group.
Conclusions: Following I/R injury of the right ventricle, optimal haemodynamics in newborn piglets was significantly better maintained in the DM-treated animals compared to AM and Dob treatment.

956: MECHANISMS OF EXERCISE INTOLERANCE IN ADOLESCENTS WITH REPAIRED PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM: A CONGENITAL HEART SURGEONS' SOCIETY STUDY

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Background: Among patients with pulmonary atresia with intact ventricular septum (PAIVS) for whom the optimal repair type is unclear, there are some for whom selection of biventricular repair diminishes survival rate and may impair late functional outcomes. We sought to determine the late patterns of exercise intolerance and associated factors.
Methods: From 1987 to 1997, 448 neonates with PAIVS were enrolled on presentation; 79/271 survivors underwent exercise testing in a cross-sectional follow-up study. An expert reviewed blinded exercise test results and grouped patients by the mechanism of exercise intolerance. Groupings were then related to demographics, neonatal morphology and repair type.
Results: Study participants (median age 17.2 years) included 44 biventricular, 22 univentricular and 13 1.5-ventricle repairs. Mechanisms of exercise intolerance were: 18 (23%) reduced stroke volume, three (4%) chronotropic insufficiency and three (4%) desaturation, with one patient having all three mechanisms; 32 (41%) were unclassifiable due to submaximal effort or missing data, and 25 (32%) were normal. Exercise intolerance by any mechanism was associated with lower body mass index (BMI) *z*-score at testing (OR = 2.17, $p < 0.05$) and smaller birth weight (OR = 1.02, $p < 0.05$). Exercise intolerance showed a trend of association with larger initial right ventricular size (OR = 1.64, $p = 0.08$) and higher pre-operative left ventricular systolic pressure (OR = 1.07, $p = 0.09$). Lower BMI *z*-score at exercise testing (OR = 1.79, $p < 0.05$) and absence of coronary–cameral fistulae (OR = 13.28, $p < 0.05$) were associated with exercise intolerance due to reduced stroke volume. Repair type and initial tricuspid valve *z*-score had no association or interaction with exercise intolerance.
Conclusions: Late exercise intolerance after repair of PAIVS is common. Although not directly associated with repair type, exercise intolerance is associated with morphological surrogates for biventricular repair. Failure to augment stroke volume during exercise appears to equally affect patients with Fontan physiology and patient status post biventricular repair, perhaps due to ongoing right heart hypoplasia, non-compliance or impaired ventricular interaction.

962: RECONSTRUCTION OF THE RIGHT VENTRICULAR OUTFLOW TRACT WITH A TRANSANNULAR PATCH AND MONOCUSP POLYTETRAFLOURETHYLENE VALVE

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Background: Corrective surgery of a hypoplastic RVOT often requires transannular patching, followed by pulmonary incompetence, right ventricular dysfunction, arrhythmias, etc. Different techniques have often been tried to restore valve function, with disappointing results. More recently, use of a PTFE monocusp valve has been suggested as a better alternative with limited data so far on long-term results.
Methods: Between 2003 and 2011 a total of 58 patients (54 Fallot, two AVSD with Fallot, one PA,VSD and one PS) underwent RVOT reconstruction with PTFE monocusp and transannular patch; 25 had

previous shunt palliation. Data on postoperative complications, long-term outcome of valve function and frequency of re-operation were assessed. Data are expressed as mean and range.
Results: All patients survived surgery, one patient with AVSD and Fallot died two months after surgery due to sepsis. Age at operation was 9.9 months (one month – 11.1 years), median 5.3 months. After surgery the RV/LV pressure ratio was 0.52 (0.32–0.77). Postsurgical time on ventilator was 2.1 days (1–3). Stay in ICU was 2.9 days (1–10). Two patients developed junctional ectopic tachycardia. Six patients needed re-operation (10%), with implantation of valved conduits, either homograft (two) or Contegra (three), and one patient had enlargement of the RVOT with a new transannular patch. Freedom from re-operation was 96% at two years, 89% at five years and 72% at eight years. Retraction and stiffness of the monocusp was revealed in most re-operation cases. No signs of calcification or pulmonary embolisation and no risk of development of pulmonary stenosis at valvular level were observed.
Conclusions: The PTFE monocusp valve can safely be used in combination with a transannular PTFE patch, with low risk of developing pulmonary stenosis or incompetence in the early and mid-term postoperative period.

969: ANESTHETIC MANAGEMENT OF PLEUROPULMONARY BLASTOMA IN A CHILD: CASE REPORT

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Pleuropulmonary blastoma (PPB) is a rare malignancy of childhood arising from the lung or pleural cavity. Surgery is the mainstay of treatment with the aim of resecting the neoplasm completely. Anaesthesia management for surgical resection of PPB is challenging because of the risk of respiratory and cardiovascular collapse due to the diminished respiratory reserves, possible invasion into the mediastinal structures and likelihood of a concurrent lung infection. We report a case of a massive PPB in a three-year-old child and discuss an effective strategy of anaesthesia management for this high-risk surgical resection.

The three-year-old girl had mild respiratory distress on pre-anaesthetic examination. The chest X-ray showed diffuse opacification of the right hemithorax and significant mediastinal shift to the left. The laboratory tests were essentially within normal limits. The anesthetic technique involved induction with IV ketamine, gradual deepening with sevoflurane, intubation and manually assisted ventilation. The monitoring included arterial line, CVP, urine output and nasal temperature. IV fentanyl and morphine were used for analgesia. IV atracurium was given after surgical exposure was completed, and the child was connected to ventilation with pressure control mode.

There were many episodes of transient hypotension during surgical resection and manipulation, which were managed by co-coordinating with the surgical team to release traction/pressure on chest/lung and bolus administration of albumin and packed cells. The total blood loss was ~ 600 ml. The child was kept intubated at the end of surgery and was transferred to ICU. She was extubated 24 hours later and had BIPAP for the next 24 hours. The rest of her stay in hospital was uneventful and she was discharged from the hospital on the eighth postoperative day.

1004: TRICUSPID VALVE REPAIR IN PAEDIATRIC PATIENTS WITH EBSTEIN ANOMALY: THE MAYO CLINIC EXPERIENCE IN THE CURRENT ERA

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Background: Historically, tricuspid valve (TV) repair for paediatric patients with Ebstein anomaly (EA) has had varied results. Cone reconstruction (CR) has revolutionised TV repair since it is at the 'true' anatomical annulus. We report our recent experience with TV repair in EA patients < 21 years old.
Methods: Medical records were reviewed for all patients < 21 years with EA, having surgery at the Mayo Clinic from June 2007 to June 2012; 79 patients (41 males, 52%) had TV repair. Mean age = 10.0 ± 5.9 years (5 days – 20.8 years). Echo showed severe TR in 72 (91%) patients. Six patients had prior TV repair elsewhere.
Results: Initial CR was successful in 75 patients (95%). There was one early death (a 19-day-old). There were three (3.8%) early (before discharge) CR breakdowns. These patients had repeat surgery [re-repair (two), replacement (one)] prior to discharge; 77/79 (97%) patients were discharged with TV repair. Mean CPB was 107 ± 23 (51–162) min, cross-clamp time (CX) was 84 ± 17 (48–125) min. Length of hospitalisation was 6 ± 3 (3–16) days. Age, gender, CPB and CX times were not associated with early CR failure. Use of an annuloplasty ring correlated with successful initial CR (p = 0.01). There have been no early CR breakdowns since 2010. Follow up was available in 65 patients (83%). Mean follow up was 0.9 ± 1.3 years. Longest follow up was 5.1 years. There were no late mortalities or re-operations. Follow-up echo demonstrated: trivial/mild TR, 66/77 patients (90%); moderate, six (8%) patients; severe, two (2%) patients. Two patients had TV stenosis (mean gradients > 6 mmHg).
Conclusion: Of the paediatric patients with EA, 97% were discharged with TV repair. All CR failures presented before discharge. Since current TV repair results for EA and durability were excellent, intervention is recommended for younger patients.

1017: MID-TERM RESULTS AFTER AORTIC VALVE REPAIR

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Background: Aortic valve repair (AVR) is considered a good temporary solution as it offers reduction of regurgitation (AR) and stenosis (AS), and stabilisation of the ventricular dimensions until the patients grow older, at which time the full range of possible treatment options, including mechanical valve or the Ross procedure might be used. The aim of the study was to analyse mid-term outcomes of AVR.
Methods: From 2004 to June 2012, 167 AVRs were performed. Mean age at operation was 109 ± 88 months; 107 patients had predominantly AS, 17 had pure AR, and combined lesions were noted in 43 patients. The majority of patients (n = 134) had a primary repair (PR) and the remaining 33 had balloon dilation before surgery. Various surgical techniques were used including cusp extensions (CE) (n = 41), resulting in bicuspid (BC) (n = 51) or tricuspid (TC) (n = 116) morphology. In the TC group 46% (n = 53) had one cusp replaced (CR) while preserving two native cusps.
Results: There were two early deaths. Mean follow up was 48 ± 24 months.
Discussion: Valve repair is safe. The best results with reconstruction are obtained by tricuspidalisation of the aortic valve with a cusp-replacement technique and with primary repair. Trends for longer durability after AVR were noticed in older patients and those with AR, which might reflect the importance of the growth potential and quality of tissue.

1020: GOOD INTERMEDIATE TO LONG-TERM RESULTS OF THE SWITCHBACK ROSS OPERATION: A REPORT OF TWO CASES

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Background: NeAo valve regurgitation (NAVR) and neoAo root dilatation (NARD), albeit infrequent, are a matter of concern in late complications of the Jatene operation as they may progress over time and require surgical intervention. The switchback Ross operation (SBRO) was introduced by Hazekamp *et al.* in 1997 as an innovative technique to address these conditions when neoAo valve preservation is deemed unfeasible. Although the technique looked compelling as it brings the anatomical Ao root (pulmonary autograft) back to the LVOT, the single case published by the authors developed postoperative Ao valve dysfunction that required Ao valve replacement. This led to generalised skepticism towards the SBRO. We however decided to apply the SBRO and herein report the first two cases worldwide, with good medium to long-term results.
Methods: Case 1: A child with TGA + LVOT stenosis was submitted to the Jatene operation + LVOT obstruction resection at age eight years. Moderate NAVR was noticed postoperatively. In 2002, at age 16 years, he became symptomatic on moderate physical exertion. Significant NARD + severe NAVR + gross LV dilatation were diagnosed, and an SBRO was performed.

Case 2, on day 35 of life, had the Jatene operation and transpulmonary VSD closure for a Taussig-Bing heart. In 2007, at age five years, he was diagnosed with severe NAVR + sinus of Valvalsa aneurysm + subneoAo valve obstructive membrane + tight pulmonary anastomotic line stenosis. The SBRO + subneoAo membrane resection were carried out.
Results: Bypass and aortic cross-clamp times were, respectively, 300 and 202 min, and 235 and 164 min. Both patients are presently asymptomatic, with normal-sized left heart structures and only trace aortic valve insufficiency.
Conclusion: The SBRO, although challenging, needs renewed consideration as a possible definitive solution for NAVR and/or NARD after the Jatene operation.

1023: INTRA-UTERINE EXPOSURE TO MATERNAL DIABETES IS ASSOCIATED WITH INCREASED AORTIC STIFFNESS IN EARLY INFANCY

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Background: Infants of mothers with diabetes (IDM), the most common complication of pregnancy, have an increased risk of adult cardiovascular disease (CVD). Although the aetiology and timing of onset of the cardiovascular changes remain unclear, recent studies have shown that IDM have increased aortic intimal–medial thickness in early infancy, which may be an early feature of CVD. Experimental models also suggest the intra-uterine diabetic environment structurally and functionally alters the aorta of affected offspring. We sought to determine if there was increased aortic stiffness, a feature of CVD in adults with diabetes, in IDM.
Methods: Diabetic pregnancies were recruited prospectively to examine the role of diabetes in foetal cardiovascular programming. For this aspect of the study, their infants were evaluated at three to six weeks by echocardiography for assessment of aortic stiffness, and the findings were compared to those of healthy infants from uncomplicated pregnancies. The pulse-wave velocity (PWV) was calculated as $[D/(T_2–T_1)]$; where D was the distance of blood flow through the arch; T_1, the time measured from QRS to onset of ascending and T_2, onset of descending aortic systolic flow.
Results: Twenty-five maternal–infant pairs were assessed, including seven IDMs and 18 controls. No statistical difference was observed in age at examination, BSA and systolic blood pressure between IDMs and controls. Haemoglobin A_{1C} (HbA_{1c}) of the diabetic mothers during pregnancy ranged from six to 10.3 µg/dl (mean 7.1 ± 1.2). Aortic PWV were significantly higher among IDMs compared to controls (mean 5.6 ± 1.5 vs 3.7 ± 1.2 m/s respectively, $p = 0.008$). IDM PWV in this small cohort tended to correlate positively with maternal HbA_{1c} ($r = 0.59$, $p = 0.068$).
Conclusion: IDM have evidence of increased aortic stiffness in early infancy, which may relate to maternal glycaemic control. Whether the aortic stiffness persists later in life and contributes to adult CVD is not clear.

1027: CARDIOLOGY *IN SILICO*: USE OF AN ELECTRONIC MEDICAL RECORD AND ECG DATABASE IN AN INTEGRATED HEALTHCARE SYSTEM TO IDENTIFY INDIVIDUALS WITH UNDIAGNOSED NOONAN SYNDROME

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Background: Noonan syndrome (NS) is autosomal dominant, characterised by short stature, dysmorphology, pulmonic stenosis (PS) and hypertrophic cardiomyopathy (HCM). These and other features can be coded in an electronic medical record (EMR) using ICD-9. Left-axis deviation (LAD) on ECG, found in 1% of healthy children, is found in 50% of those with NS (independent of structural heart disease).
Aim: To use an EMR and ECG database to identify previously undiagnosed NS.
Methods: An EMR serving 954 650 individuals < 19 years old identified 73 patients with NS (ICD-9 759.89), prevalence 7.6/100 000. Of these 73, 60 (82%) had an ECG, and 32 (53%) revealed LAD. Then, to identify previously undiagnosed NS, we screened the EMR for a combination of commonly coded NS features (short stature, PS, etc.). Using the ECG database, we identified those with ECG LAD (–30° to –90°). Lists were cross referenced. Of the 73 individuals with known NS, seven (9.6%) would have been identified by the outlined *in silico* process.
Results: Two or more NS features and LAD on ECG were identified in 65 individuals (study group). These EMRs were reviewed by both a cardiologist and geneticist to identify those with additional documented features (e.g. hearing loss, ptosis) warranting further evaluation for NS. Of these, four carried a diagnosis of NS, one cardiofaciocutaneous syndrome (CFC), and one Kabuki syndrome. Of the remaining 59, 18 were felt to warrant further clinical evaluation. Of these 18, eight thus far have sufficient documented features to establish a clinical diagnosis of NS.
Conclusions: Using an EMR and ECG database, undiagnosed patients with NS can be identified. This is the first demonstration of an integrated healthcare system's EMR being used to identify previously undiagnosed rare genetic disorders. ECG may be of utility in evaluating individuals with short stature and other NS-associated clinical features.

1044: SURGICAL REPAIR OF VENTRICULAR SEPTAL DEFECTS IN CHILDREN WITH ELEVATED PULMONARY VASCULAR RESISTANCE: THE DOUBLE-PATCH TECHNIQUE

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Background: In developing countries, children with ventricular septal defects frequently have a delay in their treatment. The result of

this late presentation is often elevated pulmonary vascular resistance. Consequently, in those countries where paediatric cardiac surgery is performed, these children are considered too high a risk to undergo an operation because of postoperative pulmonary hypertension. We have pioneered the use of a fenestrated uni-directional double-patch closure technique in such children since 1996.
Methods: We reviewed our database and contacted all sites where the procedure has been performed to our knowledge. The database was searched for catheterisation data, pre- and postoperative echo data, extubation time, survival on discharge and use of sildenafil pre- or postoperatively. All values are expressed as mean ± standard deviation. A *p*-value of 0.05 was considered significant.
Results: A total of 219 patients underwent the double-patch operation. There were 139 who underwent isolated ventricular septal defect (VSD) closure and 80 who received the double patch as part of a more complex repair. Pre-operative pulmonary vascular resistance was 11.4 ± 2.7 wood units in the VSD group and 9.2 ± 3.1 in the complex group ($p < 0.02$). The ratio of pulmonary to aortic systolic pressures was 0.97 ± 0.08 in the isolated VSD and 0.91 ± 0.1 in the complex group ($p < 0.01$). The Qp/Qs in the VSD group was 1.4 ± 0.5, and 1.7 ± 0.9 (NS) in the complex group. Pre-operative saturation was 90 ± 4% in the VSD group and 85 ± 9% in the complex group ($p < 0.05$) and sildenafil use was not significantly different. Survival in the isolated VSD was 95.9 and 83.7% in the complex group ($p < 0.01$). Recent sildenafil use improved survival in both groups ($p < 0.05$).
Conclusion: Double-patch VSD closure provides operative intervention in developing countries, with reasonable mortality rates. Complex defects remain difficult to deal with.

1048: ARE EXTREMELY HIGH BNP (BRAIN NATRIURETIC PEPTIDE) LEVELS AN INDICATOR OF CATASTROPHIC OUTCOMES IN PAEDIATRIC PATIENTS?
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Objectives: BNP is known to be increased in patients with clinical heart failure and correlates with the severity. It is unknown if BNP has predictive value regarding prognosticating outcomes or mechanical circulatory support (MCS) requirements. Current literature hints towards this possibility. We attempted to quantify at which critical BNP level patients are likely to need MCS in a non-homogeneous paediatric population.
Methods: We carried out a single-centre retrospective review of all patients who had a BNP level drawn between 2009 and September 2011. Chart reviews of all patients with a value > 600 pg/ml were undertaken. Primary outcomes included: urgent/emergency surgical intervention, MCS and transfer for MCS, and sudden cardiac death. Fisher's exact test was used to predict likelihood of primary outcome at BNP levels > 1 500 pg/ml. Sensitivity, specificity, and positive and negative predictive values were calculated for BNP > 1 500 pg/ml.
Results: Eleven (surgical intervention = four, MCS = three, emergency transfer for MCS = two, sudden cardiac death = two) patients had the primary outcome, nine with a BNP level > 1 500 pg/ml compared to two patients who didn't have the primary outcome [one with neonatal hypertension and congestive heart failure (CHF) and one with diabetic keto-acidosis with CHF] (odds ratio = 209.25, 95% CI = 26.25–1 668.18, $p < 0.001$). BNP > 1 500 pg/ml had sensitivity and a positive predictive value of 82%, while specificity and a negative predictive value was 98%.
Conclusions: BNP levels > 1 500 pg/ml are predictive of poor outcomes and alert the clinician to the seriousness of the patient and potential need for MCS, even when other parameters of perfusion seem normal. We recommend following BNP values serially to assist in determining changes in cardiac function/reserve in selected patients, i.e. stable dilated cardiomyopathy versus viral myocarditis. As low levels of BNP showed good negative predictive value, low results should be reassuring.

1089: ASSESSMENT OF LEFT VENTRICULAR FUNCTION AND MITRAL VALVE REGURGITATION AFTER CREATION OF A DUAL CORONARY SYSTEM FOR ANOMALOUS LEFT CORONARY ARTERY ORIGINATING FROM THE PULMONARY ARTERY IN INFANTS
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Background: Anatomical correction seems to be an ideal method of surgical treatment of the anomalous left coronary artery arising from the pulmonary artery (ALCAPA) in infancy. The medium-term outcome was investigated for infants with the ALCAPA following creation of a dual coronary arterial circulation.
Methods: Between April 1995 and July 2012, 23 infants with a median age of four months underwent aortic re-implantation of the ALCAPA at our Department. Direct implantation of the anomalous coronary artery into the ascending aorta was feasible in 16 patients. A trapdoor flap method was used in five infants and a tubular extension technique in two patients. No infant underwent mitral valve repair at the time of the ALCAPA surgery. Left ventricular function and degree of mitral regurgitation were assessed during a 10-year follow up.
Results: Four patients died in early postoperative period. There were no independent predictors associated with postoperative mortality. During follow up, both early and late improvement of myocardial function was observed in all patients. There was only one improvement of severe mitral regurgitation in the postoperative period. Later, two children needed mitral valve replacement. There were no early or late re-operations of the re-implanted coronary arteries.
Conclusions: Anatomical correction is an effective method of surgical treatment of the ALCAPA in infants burdened with a low risk of re-operation due to coronary artery stenosis. There is a favorable potential for myocardial recovery within the first year of surgery. Primary mitral valve repair should be associated with coronary revascularisation in infants with severe mitral regurgitation.

1098: INITIAL PALLIATION OF COMPLEX INTERRUPTED AORTIC ARCH WITH A 'HYBRID' APPROACH
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Background: Successful neonatal surgical treatment of complex interrupted aortic arch (CIAA) remains challenging. This report constitutes our experience with the 'hybrid' approach for palliation of these patients prior to elective definitive repair to facilitate concomitant treatment of the associated lesions and to decrease morbidity.
Methods: This report is a single-institution, retrospective review of consecutive patients with CIAA treated by the 'hybrid' approach. Complex IAA is defined as weight ≤ 2.5 kg, multiple VSDs, multi-organ system failure and diminutive left ventricular outflow tract. All patients were palliated via median sternotomy with bilateral branch pulmonary arterial bands and ductal stenting. Left heart structural dimensions were measured pre-palliation and prior to surgical repair and are reported.
Results: From July 2007 to December 2010, eight patients underwent 'hybrid' palliation. Weights were 1.7 to 3.4 kg (mean of 2.6). Mean aortic valve annulus was 4.63 mm. Associated complexity was weight ≤ 2.5 kg ($n = 5$), multiple VSD ($n = 2$), shock with multi-organ failure ($n = 1$), and aorto-pulmonary window ($n = 1$). All patients were discharged home after 'hybrid' palliation. One patient died two weeks post palliation from necrotising enterocolitis. All other patients underwent successful biventricular definitive repair at a mean age of 4.4 months. One patient underwent trans-catheter device closure of a muscular VSD and another peri-ventricular device closure at the time of definitive biventricular repair.
Conclusions: 'Hybrid' palliation of CIAA with bilateral branch PA banding and ductal stenting is feasible and provides adequate palliation with excellent somatic growth. Although LV structures did not

'grow' relative to somatic growth, final repair was facilitated, especially in the presence of additional VSDs. This approach provides a safe alternative strategy in complex patients with IAA.

1111: RELATIONSHIP BETWEEN HEART DYSFUNCTION AND MANIFESTATION OF GASTRIC RESIDUAL OF NEONATE SEPSIS

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Background: Neonatal sepsis with diminished heart dysfunction is regarded as the main pathology of sepsis. The death rate is double in septic neonates with cardiovascular dysfunction. The myocardial dysfunction is defined as the diagnostic criteria for severe sepsis in the adult. The occurrence of sphlancnic and mesenteric hypoperfusion impact disorder of the digestive system manifests as gastric residue.
Objective: To analyse the relationship between gastric residue and heart dysfunction among neonates at risk of sepsis.
Methods: This cross-sectional study was conducted from January to October 2011 on neonates suspected of sepsis who were hospitalised at Neonatal HCU, Moewardi General Hospital, Surakarta. A sample was selected by quota sampling. Sepsis was assessed by clinical major–minor criteria. Gastric residue was defined when the volume of gastric aspiration four hours after feeding reached 20% for two days. Heart dysfunction was measured using two-dimensional Doppler echocardiography. A Chi-square test was performed to analyse the data using SPSS 17.0.
Results: Among 48 septic-risk neonates, we found 27 (56.3%) manifested gastric residue, 25 (64.1%) having heart dysfunction, of whom 17 (70.8%) had systolic function disorders. Neonates with impaired heart function, especially disorders of systolic function, were at risk of gastric residue (OR = 6.25; 95% CI: 1.14–34.29 and OR = 3.40; 95% CI: 1.03–11.26, respectively), and the results were significant. Neonates with gastric residue of milk were at risk of heart dysfunction compared with no gastric residue but the results were insignificant (OR = 8.00; 95% CI: 0.87 to 73.27).
Conclusion: There was a relationship between gastric residue and heart dysfunction among neonates at risk of sepsis. The presence of gastric residue could become a marker of heart dysfunction among septic-risk neonates.

1128: MULTIPLE VENTRICULAR SEPTAL DEFECTS: A NEW STRATEGY

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Objective: This was a multicentre, prospective study to evaluate a new strategy for infants with multiple ventricular septal defects (VSDs).
Methods: From 2004 to March 2011, 15 consecutive infants, mean age 3.6 months (9 days to 9 months), mean weight 4.2 kg (3.1–6.1 kg), with multiple VSDs underwent pulmonary artery banding (PAB) with adjustable FloWatch-PAB®. Associated cardiac anomalies were patent ductus arteriosus (nine), aortic coarctation (two), hypoplastic aortic arch (one) and left isomerism (one). Mean duration of preoperative mechanical ventilation was 22 days (0–240 days).
Results: There were no early or late deaths during a mean follow up of 45 months (12–89 months). FloWatch-PAB® adjustments were required in all patients, a mean of 4.7 times/patient (1–9) to tighten the PAB, and a mean of 0.8 times/patient (0–3) to release the PAB with the patient's growth. After a mean interval of 32 months (8–63 months) five/15 patients underwent re-operation: three/five PAB removal and closure of a remaining peri-membranous VSD, and two/five only PAB removal. All muscular multiple VSDs had closed in all five patients. PA reconstruction was never required. In six/10 of the remaining patients all muscular VSDs had already closed.
Conclusions: This reproducible new strategy with adjustable PAB simplifies the management of infants with multiple VSDs, providing the following advantages: (1) good results (0% mortality); (2) delayed surgery with high incidence (11/15 = 73%) of spontaneous closure of multiple muscular VSDs; (3) facilitated closure of residual peri-membranous VSD at older age and larger body weight; (4) PAB application and late removal remains the only procedure required for Swiss cheese multiple VSDs without associated peri-membranous unrestrictive VSD.

1129: CONCOMITANT ANTERIOR AND POSTERIOR TRICUSPID LEAFLET AUGMENTATION WITH A SINGLE PATCH: THE RESPECT OF THE GEOMETRY

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Objectives: Isolated tricuspid anterior leaflet extension is an option used to increase leaflet coaptation. Nevertheless the posterior leaflet and the commissure between septal and posterior leaflets could lead to residual valve regurgitation. We describe the augmentation of both the anterior and the posterior tricuspid leaflets with a single patch.
Methods: In the last year, four patients (mean age 37 years) presented with severe tricuspid regurgitation with leaflet tethering and coaptation deficit. Mean coaptation deficit was 19.5 mm. They received concomitant anterior and posterior leaflet augmentation with a single patch. In one patient an extracellular matrix patch was used while in the others an autologuous patch was implanted. The patch was trimmed using a 32-mm ring sizer. After detaching of the leaflets from the annulus, secondary chordae were removed. In all of them an anuloplasty was performed using a prosthetic ring. CE no 32.
Results: There were no deaths or major complications during in-hospital stay. No early residual regurgitation could be observed. After a mean follow up of eight months, all patients demonstrated absence of recurrent regurgitant jet. Mean gradient across the valve was 1.5 mmHg. After combined leaflet augmentation, the valve acts as a single-leaflet valve, abolishing the effect of the septal leaflet tethering.
Comments: Anterior leaflet augmentation to treat tricuspid valve regurgitation might induce geometric disarray between anterior and posterior, and posterior and septal valve leaflets. In the last year we introduced the anterior and posterior leaflet augmentation using a single patch, thus reducing the possible residual regurgitant jet in the commissure between the anterior and the posterior leaflet. The patch size was identical to the prosthetic ring in order to have a functional single-leaflet valve and abolishing the detrimental effect of the posterior-leaflet tethering. Immediate and early results are encouraging.

1130: CANNULATION OF THE BRACHIOCEPHALIC TRUNK AND ANTEGRADE CEREBRAL PERFUSION IN AORTIC ARCH REPAIR IN NEONATES

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Introduction: Cannulation of the brachiocephalic trunk (BCT) with the interposition of a goretex conduit allows easy antegrade cerebral perfusion (ACP) and possibly coronary artery perfusion during arch surgery. ACP is advocated as a factor protecting the abdominal organs, which are supposed to be perfused through the collaterals. We describe our series of aortic arch repair in neonates.
Methods: Between April 2005 and June 2012, 18 neonates presented with aortic arch hypoplasia (age 2–27 days, mean weight 3.39 kg). Eleven patients presented with an associated VSD: seven received pulmonary artery banding while five had concomitant VSD surgical closure. The BCT was cannulated through a Goretex conduit in all patients. The arch was clamped distally to the BCT, distal to the isthmus and then both the left carotid and left subclavian arteries were clamped. The arch was reconstructed under cardio-cerebral perfusion

with a beating heart and body temperature of 25°C. A short period of cardioplegic arrest was used to insert the patch into the ascending aorta. A homograft patch was used in all patients but two, in whom a porcine extracellular matrix patch was preferred. Mean cross clamp time in isolated arch repair was 21 min.
Results: One patient experienced a cardiac arrest in the first postoperative day, he was resuscitated and sustained with the ECMO but died on the fifth postoperative day. One patient who received pulmonary artery banding required further increase in the banding. None experienced neurological damage or new onset of seizures. None required further arch surgery after 38.6 months follow up (1–87 months).
Conclusions: BCT cannulation in neonates allows us to perform arch surgery under cardio-cerebral perfusion, thus reducing the cross-clamp time. The results of our series are encouraging and evidence that this technique is safe and effective.

1132: TELEMETRIC FLOWATCH PULMONARY ARTERY BANDING: SINGLE-CENTRE EXPERIENCE AND OUTCOMES

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Background: An assumed advantage of the FloWatch pulmonary artery band (PAB) is that it has a low incidence of pulmonary artery (PA) distortion and requirement for PA reconstruction after its removal. We describe our experience with FloWatch PAB with regard to pulmonary artery distortion needing patch reconstruction and admission stay in a large single-centre population.
Methods: We carried out nearly 10 years of retrospective analysis of all patients at our centre who underwent FloWatch PA banding to control pulmonary blood flow for initial single-ventricle or bi-ventricle palliation. In a total of 70 patients, the diagnosis was multiple VSDs in 30, complete AVSD in 10, and 30 with mixed complex congenital conditions.
Results: Seventy patients needed FloWatch PAB between December 2003 and June 2012; 19/70 (27%) had single-ventricle morphology and 51/70 (73%) biventricular morphology. Median age at the time of PAB was 88 (range 7–1 486) days and median weight was 4.2 (range 2.6–15.9) kg. There were seven deaths in our series, six were late deaths and were not associated with PA band. There was one early death; 34/70 (48%) had their band removed for the next stage of surgery and 36/70 (52%) still have the band in place; 23/34 (67%) did not have any PA distortion and did not require any patch enlargement. However, 11/34 (32%) had PA distortion needing patch enlargement. Of more concern, in two/34 (6%) patients the FloWatch was found to have eroded through the MPA at the time of its removal. The mean duration of PICU stay after PAB insertion was 5.8 days; 46% of patients were discharged within three days and 76% discharged within seven days.
Conclusion: Telemetric FloWatch PA banding does have undoubted advantages over conventional banding in terms of adjustability of pulmonary flow without re-operation and limited hospital stay. The majority of patients did not require any patch reconstruction of the pulmonary artery.

1149: ROOT REPLACEMENT WITH AORTIC VALVE SPARING IN PAEDIATRIC PATIENTS

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Background: Aortic root and ascending aortic aneurysm in children is a rare entity usually associated with connective tissue disorders. Aortic valve sparing with ascending aortic replacement (TD) is the ideal treatment in these patients
Objective: To evaluate the short- and medium-term results of valve-sparing aortic root replacement in paediatric patients.
Methods: This was an historic cohort between January 2006 and June 2010 and included patients under 15 years of age treated with aortic valve re-implantation for aneurysm or dissection. Postoperative clinical and echocardiographic follow up was performed.
Results: Four patients with annulo-aortic ectasia had Marfan syndrome and one had a type A dissection. Mean age was 10 years (7–14); 75% were male in functional class I (50%) and II (50%). There were no bicuspid valves. Two patients had aortic regurgitation 1+, and two had 3 to 4+. Aortic annulus was 23.7 mm (18–30), aortic root was 60.7 mm (40–90), three patients received TD IV (75%), one (25%) TD V. Aortic graft diameter was 28 mm (18–30); the TD V was done with 30/22-mm tubes. Complications: three patients presented with coagulopathy without re-operation for bleeding. Echocardiogram showed in two patients, residual 1+ regurgitation. At the end of follow up, there was no progression of aortic regurgitation, and function and ventricular dimensions were preserved. All patients are in functional class I and there were no re-operations or mortality.
Conclusions: Aortic root replacement with valve sparing is useful in the management of aortic root aneurysms and dissection in children, preserving ventricular geometry and function, with low postoperative morbidity and it eliminates the risk of chronic anticoagulation. Studies of long-term monitoring will allow statistical power to evaluate the durability of the anatomical and functional results.

1160: EXTENDED THORACIC AORTA REPAIR IN CHILDREN VIA LEFT THORACOTOMY WITH DEEP HYPOTHERMIC CIRCULATORY ARREST AND CERVICAL CANNULATION

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Background: Repair of the thoracic aorta via median sternotomy can be difficult due to the limited exposure, particularly in older patients or during re-do surgery. We are reporting a novel approach via left thoracotomy with cervical cannulation for CPB.
Methods: The patient is positioned supine and both RCCA and RIJV are cannulated as per V-A ECMO. CPB is commenced and the patient positioned to perform a left thoracotomy. The pericardium is opened and a vent inserted in the LA appendix. The aortic arch, head and neck vessels and the descending aorta are dissected while cooling systemically. At temperature, the circulation is arrested, the aorta cross clamped between the innominate and LCCA, the head and neck vessels occluded, and cardioplegia is delivered into the ascending aorta through the arterial cannula. Antegrade cerebral perfusion is commenced directly in the LCCA or via the arterial cannula clamping between the LCCA and the LSA. The repair is performed with extensive patch enlargement of the diseased aorta, after which systemic circulation is restarted and the patient is rewarmed, weaned off CPB and protamine is given. The thoracotomy is closed and the patient is repositioned supine for neck decannulation with primary vessel reconstruction.
Results: Between June 2011 and August 2012, three patients were operated. Median age and weight were, respectively, 14 months and 9.7 kg. Two patients had previous aortic surgery and presented with severe hypoplastic distal aortic arch and recoarctation, one of whom also had long descending aortic hypoplasia. The third patient had severe aortic coarctation with a long hypoplastic descending aorta. Perfusion times: (median): CPB = 209 min; aortic cross-clamp = 39 min; antegrade cerebral perfusion = 25 min; peripheral body ischaemia = 39 min, mean DHCA = 20 min. Ther was no hospital mortality. Postoperative echocardiography showed laminar flow in the descending aorta. No neurological symptoms were detected at follow up with US patency of the vessels.
Conclusions: This technique offers good cerebral and body perfusion during circulatory arrest and optimal exposure of the aortic arch and descending aorta.

1161: THE CIRCADIAN RYTHM OF BLOOD PRESSURE IN NORMOTENSIVE CHILDREN WITH A FAMILY HISTORY OF ESSENTIAL HYPERTENSION

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A family history of hypertension is a primary predictor of high blood pressure (BP). In children and adolescents, some changes in cardiovascular structure and function may be seen independent of the level of blood pressure and even before the blood pressure increases.
Aim: The aim of this study was to evaluate blood pressure circadian rhythms of normotensive children with a family history of essential hypertension and investigate the relationship between target-organ damage, future adult hypertension and this rhythm.
Methods: Fourteen healthy children (19 girls, 21 boys) with hypertensive parents (HP), and 20 controls (10 girls, 10 boys) with normotensive parents (NP) were recruited. Mean age was 14 ± 3.5 years in girls and 15 ± 4.5 years in boys (range 8–22 years). Age, gender and body mass index did not differ between the two groups.
Results: No difference was found in casual BP between the two groups. In contrast, during ambulatory blood pressure monitoring (ABPM), daytime systolic BP and systolic load were elevated in children with HP ($p < 0.05$). Also mean systolic and diastolic blood pressures were higher in children with HP compared to the control group but it was not statistically significant. Increased thickness of left ventricular posterior wall and left ventricular mass index have been observed in children with HP ($p < 0.05$). BP circadian rythm in the children with a family history of hypertension had more non-dipper status, especially starting from the age of 20 ($p < 0.05$). Non-dippers had significantly higher values of left ventricular mass index than dippers and also a positive correlation has been found between night systolic blood pressure and left ventricular mass index ($p < 0.001$).
Conclusion: Early changes in ambulatory BP parameters were present in healthy children of HP. The non-dipper normotensive children with a family history of hypertension are thought to have target-organ damage, especially after the age of 20 years, before clinical findings of onset of hypertension.

1172: RIGHT VENTRICULAR APICAL EXCLUSION AS A TREATMENT OF LARGE APICAL VENTRICULAR SEPTAL DEFECT

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Background: Optimal management of muscular apical ventricular septal defect (VSD) remains a challenge, especially when there is a large apical defect that is not appropriate to be closed with a device. Left apical ventriculotomy offers a good guarantee of correct closure but has several mid- and long-term complications such as left ventricular dysfunction, arrhythmias or apical aneurysms.
Methods: Two patients with a diagnosis of large muscular apical VSD, aged 13 and two months, respectively, were submitted to a technique of right ventricular apical exclusion. Two heterologous pericardial patches were used in each case, the first one sutured between the upper edge of the apical VSD and the septomarginal band and the second one between the septomarginal band and the free edge of the right ventricle (RV). The apex of the RV was then left on the left ventricle side and the initial apical VSD did not connect the two ventricles any more. This was performed through a right atriotomy and through the tricuspid valve.
Results: Both patients had a normal postoperative evolution and were discharged without any complicactions. There was no residual communication between the two ventricles. Normal RV functional parameters were found during postoperative control echocardiography. No echographic data indicating right ventricular dysfunction were demonstrated after extensive examination, including lower vena cava diameter respiratory changes, M-mode tricuspidal plane excursion (TAPSE), E' wave peak velocity on tricuspid annulus (tissue Doppler imaging mode), standard Doppler and bidimensional echocardiography.
Conclusion: This technique for correction of apical complex VSD offers very good initial results, avoiding the complications associated with left ventriculotomy or the necessity of a palliative approach before surgery on low-weight patients. The short-term follow up did not show any disturbance in RV function.

1207: SURGICAL MANAGEMENT OF PULMONARY ARTERY SLING

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Objectives: Pulmonary artery sling is frequently associated with tracheal and/or bronchial stenosis. A number of patients receive only re-implantation or relocation of the left pulmonary artery while other patients may require tracheoplasty for stenosis of the airway. We studied the clinical outcomes with or without tracheoplasty.
Methods: A total of 20 patients with a PA sling who received surgery in our institute were included in this study. We reviewed the clinical outcomes and the severity of trachea stenosis, and evaluated various surgical strategies.
Results: Among the 20 patients, all received left pulmonary artery (LPA) re-implantation, and tracheoplasty was performed in 12 (60%). Of the 12 patients who received LPA re-implantation and tracheoplasty, nine received slide tracheoplasty and eight survived. One patient died of cytomegalovirus infection despite a patent airway. Two patients had pericardial patch augmentation and both died. One patient received a resection and end-to-end anastomosis and survived. The patients without tracheoplasty all survived, but the trachea diameter remained stenotic in the follow-up period.
Conclusions: In this study cohort, approximately 60% of the patients with PA sling received tracheoplasty. Slide tracheoplasty provided acceptably good results for patients with PA sling and tracheal stenosis.

1215: USE OF INHALED TREPROSTINIL IN VENTILATOR-DEPENDENT INFANTS

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Background: Inhaled treprostinil is a tricyclic benzidine prostacyclin analog used in adults with pulmonary hypertension and NYHA FC III and IV symptoms. We present two infants with pulmonary artery hypertension treated with inhaled treprostinil.
Results: SB was a 10-month-old male with repaired right-sided CDH s/p tracheostomy who was ventilator and iNO dependent. Chest CT at five months revealed small areas of scar and air trapping in the left lung with overall normal aeration. His right lung was hypoplastic with changes of CLD and air trapping. Serial echocardiograms demonstrated elevated PAP. By eight months, despite treatment with sildenafil, SB developed right ventricular (RV) dysfunction by echocardiogram with iso-systemic PAP. Because of the high potential for V:Q mismatch with systemic therapy, SB was treated with

inhaled iloprost 2.5 mcg q2h. Over the next week SB was weaned off iNO and his ventilator support was decreased by 50%. Cardiac catheterisation revealed mPAP 40 mmHg and PVRi 6.58 units/m² with normal RV function. He was transitioned to inhaled treprostinil three breaths q6h in an effort to provide an acceptable regimen for a step-down unit. SB tolerated the transition without systemic side effects or bronchospasm. Echocardiogram at 10 months reveals mild elevation of PAP and normal RV function.

NH was a two-month-old with partial monosomy 10q26, partial trisomy 10p12.2, PDA and PPHN, who developed suprasystemic PAP and RV dysfunction on sildenafil and iNO. At one month he was started on iloprost 2.5 mcg q2h after severe hypotension with milrinone, and subsequently transitioned to inhaled treprostinil three breaths q6h. At two months he underwent cardiac catheterisation with mPAP of 49 mmHg and PVRi 5.6 units/m² with normal RV function. NH's PDA was uneventfully closed. He continues to tolerate inhaled treprostinil without side effects.

Conculsions: Inhaled treprostinil may be safely and effectively administered to ventilator-dependent infants.

1219: COMPARISON OF RESULTS OF AUTOLOGOUS VERSUS HOMOLOGOUS BLOOD TRANSFUSION IN OPEN-HEART SURGERY

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Background: In this randomised, controlled study we investigated the effects of autologous and allogenic blood transfusion in adult on-pump cardiac surgery.

Methods: Three hundred and four patients who underwent surgery between January 2009 and April 2012 were randomised into two groups. The A group (n = 164) consisted of patients who received autologous blood transfusions, and the B group (n = 160) patients received allogenic blood transfusions. All patients underwent on-pump cardiac surgery via a sternotomy. The time to extubation, chest tube drainage volume, postoperative red blood cell counts, amount of blood transfused, postoperative temperature, and postoperative haematocrit level were recorded in the intensive care unit on the 14th postoperative day.

Results: Intra-operative bleeding and fluid resuscitation were similar in the two groups, however, there were significant decreases in postoperative blood loss, extubation period, postoperative red cell counts, and fever in group A compared with group B. Drainage volume in group A was 397 ± 77.6 ml with a mean haematocrit value of 29.3 ± 3.5%. There were no deaths. No patients needed re-exploration for bleeding. There were no postoperative complications; 64 patients (40%) did not receive any blood or blood products and 24 patients (7%) needed only one unit of homologous red cell transfusion in the retransfusion group, whereas 40 patients (26%) needed one unit, and three patients received two units of homologous red cell transfusion in control group B (p = 0.008). At discharge, the mean haematocrit value was statistically higher in group A compared with group B.

Conclusion: The use of autologous blood in patients having undergone open-heart surgery not only attenuated side effects and complications of transfusion but also positively affected the postoperative recovery process. Therefore, it can be considered to be an easy, effective and cheap technique.

1227: ROUTINE CLOSURE OF VENTRICULAR SEPTAL DEFECT THROUGH LEFT VENTRICLE OUTFLOW TRACT DURING ARTERIAL-SWITCH OPERATION FOR D-TRANSPOSITION OF THE GREAT ARTERIES/VENTRICULAR SEPTAL DEFECT

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Background: Arterial-switch operation (ASO) and VSD closure for d-TGA-VSD is performed worldwide with excellent surgical results. Traditionally, VSD closure has been proposed via a trans-tricuspid or trans-ventricular (in Taussig–Bing anomaly) approach. We present our experience with VSD closure through the left ventricular outflow tract (LVOT) as an easy and reproducible technique.

Objective: To study safety and feasibility of VSD closure via LVOT during ASO.

Methods: Operations were performed with continuous, moderately hypothermic CPB cardioplegia. Having the great arteries divided, the VSD was closed through the LVOT with a PTFE patch and running suture. The ASO then continued in the usual manner. A double-clam-shell device was applied to close additional apical VSDs. Results were evaluated with intra-operative TEE.

Results: Neonates (9 SD, 4.8 days) undergoing ASO (n = 102) had their VSDs closed (38) via LVOT. Aortic/pulmonary diameter ratio was 0.64. Morphological malalignment: VSD with (21 patients) and without (five) sutures anchored superficially on remnants of the peri-membranous septum and postero-inferior muscular rim; Taussig–Bing anomaly (nine patients); three additional apical VSDs were repaired. Coexisting LVOTO (eight), neo-aortic valvotomy/plasty (four), and aberrant MV chords (three) were addressed from the same approach. TEE showed no significant residual shunt. All patients had normal AV conduction postoperatively. No surgical morbidity/mortality occurred.

Conclusions: Larger neo-aortic/LVOT diameter in d-TGA-VSD offers a safe and reproducible technique for VSD closure during ASO. This approach obviates restricted trans-tricuspid visibility and/or the need of ventriculotomy (Taussig–Bing anomaly). Associated LVOT anomalies can be simultaneously addressed. Concerns of injuring the AV conduction can readily be avoided by appreciating the morphology and anchoring superficial sutures on the fibrous remnants of the perimembranous septum and/or applying a suture-less flap extension of the patch in the area of the penetrating bundle.

1235: PREVALENCE AND CORRELATES OF LEFT VENTRICULAR HYPERTROPHY IN THE PAEDIATRIC HYPERTENSIVE POPULATION

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Background: Hypertension (HTN) is common in childhood, yet the diagnosis is frequently delayed. Prior literature shows that up to 35% of children diagnosed with HTN have left ventricular hypertrophy (LVH) at presentation. Therefore an echocardiogram (echo) is recommended at the initial evaluation. In recent years, however, awareness of paediatric HTN has improved. We hypothesised that the current prevalence of LVH in paediatric hypertensive patients is lower than previously reported.

Methods: A single-site retrospective chart review was performed from 1 July 2009 to 20 February 2012. Charts with ICD-9 code of hypertension were identified in the echo server and appointment database. Patients were considered at risk for LVH if ECG voltages (S in V1, R in V6) were ≥ 98%, or if LVMI was > 95 g/m² (females) or 115 g/m² (males) as per American Society of Echocardiography (ASE) guidelines.

Results: Children (140, age 3–17 years) with untreated HTN were identified; 33 were excluded due to incomplete data. Mean age was 13.6 (SD 3.2) years; 78.5% of the cohort were male; 51.4% were obese, 33.6% had stage 1 HTN and 47.7% had stage 2 HTN. Mean LVMI was 79.6 (SD 20.7) g/m², and 4.7% (95% CI: 0.6%, 8.7%) met ASE LVH criteria. LVMI correlated positively with age (r = 0.43, p < 0.001), SBP (r = 0.29, p = 0.001), black vs white race (r = 0.24, p = 0.009), male gender (r = 0.30, p < 0.001); 18.7% (95% CI: 11.2%, 26.2%) of patients demonstrated ECG LVH evidence. ECG LVH indicators did not correlate with LVMI (r = 0.1, p = 0.32) or any other variables (all r < |0.10|). ECG voltages and LVMI showed poor agreement (kappa = 2.1, p = 0.93).

Conclusion: In our single-institution cohort of 140 patients, LVH incidence was < 5% in untreated hypertensive children, which was

significantly lower than prior literature reports. Our findings also suggest that ECG LVH criteria do not correlate with LVH findings by echo. We suggest that ECG may be redundant for initial evaluation of hypertension.

1237: THE $ScvO_2$/LACTATE RATIO FOLLOWING SURGERY FOR HYPOPLASTIC LEFT HEART SYNDROME

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Introduction: Mortality following stage I hypoplastic left heart syndrome (HLHS) surgical repair is variable and may be as high as 25%. Postoperative low cardiac output is a significant risk factor for mortality and has been shown to be associated with a low central venous oxygen saturation ($ScvO_2$) and high lactate level. In cases where a $ScvO_2$/lactate ratio is < 5, major postoperative adverse events have previously been reported in a heterogenous group of patients.
Hypothesis: A $ScvO_2$/lactate ratio < 5 is a predictor of major adverse postoperative events following stage I HLHS repair.
Methods: Following IRB approval, 37 infants having undergone HLHS stage I repair at one institution from 2004 to 2009 were examined retrospectively. Outcomes for subjects with a $ScvO_2$/lactate ratio ≥ 5 or < 5 were compared using a Mann-Whitney *U*-test and Chi-square test. A two-tailed *p*-value < 0.05 was considered significant. Analyses were performed using Stata 12.0 (College Station, TX).
Results: Overall 30-day mortality was 19% (7/37). Six/36 infants had a $ScvO_2$/lactate ratio < 5 within 24 hours ofg surgery. One of seven deaths (14.3%) occurred following hospital discharge. ECMO was required in three/37 infants (8%) and one/three survived to hospital discharge. Prolonged mechanical ventilation (> 14 days) was necessary in 10/37 (27%). The $ScvO_2$/lactate ratio was not significantly correlated with overall mortality ($p = 0.6$), need for ECMO ($p = 0.7$), hospital LOS ($p = 0.3$), and ICU LOS ($p = 0.3$). However, a $ScvO_2$/lactate ratio < 5 was associated with need for prolonged mechanical ventilation (83 vs 25%, $p = 0.01$).
Conclusion: A $ScvO_2$/lactate ratio < 5 was not significantly associated with outcomes following stage I repair for HLHS except for the need for prolonged mechanical ventilation. Further studies of markers of postoperative low-cardiac output syndrome are needed in this patient population.

1243: DEVELOPMENT OF A NOVEL CONGENITAL CARDIAC ANAESTHESIA DATABASE

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The Congenital Cardiac Anesthesia Society (CCAS) partnered with the Society of Thoracic Surgeons' Congenital Heart Surgery database (STSCHSD) beginning on 1 January 2010. The CCAS chose to collaborate with the STSCHSD because of the interconnected nature of our patient populations and the multiple data fields of interest to both groups. This combined dataset would minimise the duplication of efforts in entering data such as patient demographics, diagnoses and procedures for those cases occurring in the operating room. In addition, for the first time, the STSCHSD was opened up to procedures occurring outside the operating room on patients with congenital cardiac lesions undergoing non-surgical procedures such as cardiac catheterisations, radiological procedures (cardiac MRI, CT and interventional radiology), and non-cardiac operations on patients requiring cardiovascular anaesthesia because of their underlying physiology. Anaesthesia is one of the common denominators in the care of these patients throughout their hospitalisations and it has been well established that this subset of patients experiences cardiac arrests at a far higher rate than comparably aged children undergoing similar procedures without congenital heart defects. By the Spring 2012 data harvest, representing 1 January 2010 to 31 December 2011, 30 institutions from a wide geographic and programme size in the United States had submitted data on over 20 000 discrete anesthetics. Data submitted included information on patient demographics, pre-operative medications, intra-operative anaesthetic management and monitoring and anesthesia-related adverse events. As this data set grows it can serve as a model for other anaesthesia populations as well as act as an enormous 'data sink' that can be mined for both care- and outcomes analysis.

1258: UTILITY OF ARGININE VASOPRESSIN IN NEONATES WITH PERSISTENT PULMONARY HYPERTENSION OF THE NEWBORN RETRIEVED FOR EXTRA-CORPOREAL MEMBRANE OXYGENATION

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Background: Our institution uses arginine vasopressin (0.06–0.09 U/kg/h) in stabilisation of PPHN infants with high inotropic requirements (≥ 0.1 mcg/kg/min of adrenalin) for potentiation of adrenalin and sparing pulmonary vascular resistance. This application of vasopressin is unpublished.
Methods: A retrospective chart review was done of neonatal ECMO retrieval deom January 2010 to August 2012. Inclusion criteria: echo-proven PPHN, structurally normal heart, vasopressin used for transport, complete dataset at pickup and on return (before ECMO). Exclusion criteria: congenital diaphragmatic hernia. Paired *t*-test (two-tailed) was used to analyse data. Group characteristics: 14 term neonates, five girls, aged one to four days, mean weight 3.8 kg. Ten had meconium aspiration, eight proven or suspected sepsis, six received hydrocortisone, all inhaled nitric oxide. Transport times were two to four hours (mean 2 h 50 min), 11 were given ECMO (seven veno-arterial, four veno-venous). All survived back-transfer to the referring unit at a median of six days.
Conclusions: Despite small numbers, retrospective design and patients as own controls, this data suggests vasopressin improves blood pressure and reduces vasoactive inotrope score in PPHN. A larger prospective study is warranted.

1263: EFFECT OF CARDIOPULMONARY BYPASS ON NERVE CONDUCTION VELOCITES IN INFANTS

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Objectives: To report on the effect of cardiopulmonary bypass (CPB)-induced systemic inflammatory response syndrome (SIRS) on nerve conduction velocities (NCV) in infants undergoing elective cardiac surgery.
Methods: This was a pilot study with prospective recruitment of six infants admitted to hospital requiring elective cardiac surgery under bypass. Non-syndromic infants aged between three and 12 months who were to undergo elective ventricular septal defect or tetralogy of Fallot repair were consented for study at the pre-admission clinic. On the morning of surgery, an initial nerve conduction study (NCS) was performed under anaesthesia before administration of any muscle relaxants. After surgery, patients were shifted to the paediatric intensive care unit where a second study was performed 24 hours after the initial study if infants remained intubated for 24 hours or longer.
Preliminary results: Critical illness polyneuropathy (CIPN) and critical illness myopathy are commonly reported in the intensive care setting. SIRS, induced by various mechanisms, is a known risk factor for the development of both. Our aim was to study whether SIRS induced by CPB altered NCV in any detectable way. Five of six patients underwent a second NCS 24 hours after the first study. None had any clinical change in their neurological examination or NCV as assessed by a paediatric neurologist proficient in performing this

procedure in young infants.
Conclusions: Preliminary findings suggest that NCV abnormalities are undetectable in the early period after CPB. Further studies are required to investigate early features of CIPN in ill children in the intensive care unit.

1275: SCIMITAR SYNDROME: A 20-YEAR SINGLE-INSTITUTION EXPERIENCE

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Objectives: To review our experience in the management of scimitar syndrome over the last 20 years.
Methods: Clinical records, echocardiographic images, catheterisation data, operative reports and follow-up information were recorded and reviewed. The patients were divided into infant (< 1 year) and adult (> 1 year) forms. Those who had complete repair of the anomalous vein were subdivided into baffling or re-implantation of the anomalous scimitar pulmonary vein to the back of the left atrium.
Results: Between 1986 and 2006, 36 patients presented with scimitar syndrome. There were 20 females and 16 males (mean age 29 months), with 22 patients in the infant group. Higher proportions of infants had right lung hypoplasia and four had primary right pneumonectomy as sole therapy with good outcome. Thirty patients had systemic collateral supply to the right lung; 20 had coil embolisation. Coil embolisation was the only intervention in five patients. Surgical repair of the scimitar vein was accomplished in 21 patients with 10% mortality. No major differences were encountered in the incidence of pulmonary hypertension, early mortality and late survival of the two age groups. Also no differences in early mortality, late outcome and rates of obstruction between different methods of surgical repair were seen.
Conclusion: Good, comparable results were achievable in infants and older children with scimitar syndrome with an aggressive approach comprising liberal coil embolisation of collateral vessels and early surgical repair of the anomalous pulmonary vein. Primary pneumonectomy may be an option in selected patients. Both baffling and re-implantation techniques provided similar outcomes.

1281: BEATING-HEART AORTIC ARCH REPAIR WITH MILD HYPOTHERMIA: OUR EXPERIENCE WITH THREE CASES

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Introduction: Aortic arch repair surgery can be performed by various techniques. Selective antegrade cerebral perfusion with continuous coronary perfusion at mild hypothermia is discussed here.
Methods: From March 2009 to date, three patients have been operated on with this technique.
Case 1: A nine-year-old child with supravalcular aortic stenosis of diffuse variety involving aortic arch and arch vessels. Case 2: An eight-year-old male child with Shone's complex, hypoplastic aortic arch, and severe coarctation of the aorta. Case 3: An eleven-year-old male child with hypoplastic arch, severe coarctation of the aorta, single ventricle PDA, and severe PAH.
Result: All three patients had innominate artery cannulation for antegrade cerebral perfusion and ascending aortic cannulation for coronary perfusion. Total mean CPB time was 85 min. Arch isolation time was 40 min and mean temperature was 28°C. All patients recovered well without any neurological complications and hospital stay was eight days. At the two-year follow up, all patients were doing well.
Conclusion: Aortic arch repair can be done safely with a selective antegrade cerebral perfusion technique.

1282: BROM'S TECHNIQUE FOR SUPRAVALVULAR AORTIC STENOSIS: OUR EXPERIENCE

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Introduction: Supravalvular aortic stenosis is a rare form of left ventricular outflow obstruction, commonly seen in William's syndrome with a diffuse variety of localised narrowing at the sino-tubular junction. This was a study of the management of supravalvular aortic stenosis with Brom's three-patch enlargement technique.
Methods: We operated on a total of four patients with supravalvular aortic stenosis, two with a localised variety and one with a diffuse variety involving the ascending aorta and aortic arch vessel origin. Mean age of patients was 8.4 years. Three patient had William's syndrome while one had Shone's complex component (parachute mitral valve with non-significant gradient). All four patients with supravalvular aortic stenosis were repaired with Brom's technique (individual patch augmentation of each sinus). In patients with the diffuse variety along with Brom's technique, the ascending aorta and origin of the aortic arch vessels were enlarged with a pericardial patch. Mean CPB time was 190 minutes, aortic cross-clamp time was 116 min. There was no hospital mortality. Mean ICU stay was 2.5 days, hospital stay was 7.6 days. Discharge echo on the seventh postoperative day suggested no gradient at the sino-tubular junction. Three patients had their three-year follow up without any complications.
Conclusion: Brom's technique is a good, simple and safe technique for supravalvular aortic stenosis, with a good surgical outcome.

1283: CASE REPORT: EXTENSIVE ARCH REPAIR WITH PA BANDING FOR DORV, LARGE VSD, SINGLE VENTRICLE

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Case report: A six-year-old boy presented with frequent lower respiratory tract infections since early infancy, needing frequent hospitalisations. Clinically, the child had cyanosis, clubbing, precordial bulge, Harrison's sulcus indicating a high pulmonary blood flow situation with a loud P2, soft pansystolic murmur and a mid-diastolic flow murmur at the apex. Clinically he appeared operable. Echo examination revealed DORV, large inlet VSD, subaortic extension, apical Swiss cheese defects, straddling tricuspid valve, normally related great arteries with hugely dilated, confluent branch pulmonary arteries, a hypoplastic aortic arch with interruption, and PDA supplying the descending aorta. CT angiogram revealed a hypoplastic transverse arch with post subclavian, preductal coarctation of the aorta. The child underwent extensive arch repair with coarctoplasty and PA banding. Postoperatively, the child was extubated within 24 hours. He was discharged within 10 days. At the one-year follow up, the PA band gradient was 70 mmHg with room air saturation at 84%.

1285: CASE REPORT 2: MODIFIED GLENN ANASTOMOSIS IN A CASE OF PAPVC TO SVC AND SINGLE VENTRICLE

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Introduction: Many procedures have been reported for the repair of partial anomalous pulmonary venous connection (PAPVC) with return to the high superior vena cava (SVC). Total cavo-pulmonary connection (TCPC) has been a common procedure but anatomical complexity of the pulmonary veins and systemic venous return in an asplenic heart makes the definitive repair more difficult. The PAPVC repair concomitant with Glenn anastomosis and the shift of the inferior vena cava (IVC) orifice has seldom been reported. We present a novel technique, in which the pulmonary artery (PA) translocation was performed to avoid obstruction of the pulmonary veins when Glenn anastomosis was established.
Discussion: A four-year-old cyanotic boy diagnosed with unbalanced

AV septal defect, transposed great arteries, and severe pulmonary stenosis underwent modified superior cavopulmonary anastomosis after wide mobilisation of the branch pulmonary arteries up to the hilar level, using the translocated main pulmonary artery. The proximal SVC stump receiving the three pulmonary veins was left behind, draining into the atrium. The postoperative course was uneventful and the postoperative echo revealed unobstructed flow in the modified BDG anastomosis, and well flowing unobstructed pulmonary veins. The child recovered well and was discharged in seven days. This case report highlights the rarely described technique using the MPA stump to complete the Glenn anastomosis, while the lower stump of the SVC was left connected to the heart, which received the pulmonary veins. A more generous dissection and mobilisation of the branch pulmonary arteries up to hilar level is needed to avoid the kinking of the left pulmonary artery after translocation of the main pulmonary artery.

1286: ROLE OF TISSUE GLUE IN PAEDIATRIC CARDIAC SURGERY: OUR EXPERIENCE

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Introduction: Complex congenital cardiac surgery involving multiple suture lines at the arterial level increase morbidity due to bleeding and transfusion-related complications. Appropriate use of tissue glue in a suitable site and a convenient method is discussed in this article.
Methods: The study period was between March 2011 and the present. In 11 patients we used tissue glue, in five patients, tissel, in four, coseal and in two, flow seal. We used the rapid method for tissel and coseal in neonates and infant aortic surgery. We used tissel just before aortic cross-clamp removal, and coseal mainly in redo aortic surgery and coarctation of the aorta. For post-clamp removal bleeding we used flow seal along with surgical packing.
Results: Tissel worked in four patients but in the frst patient, due to inappropriate application, it was not effective. Coseal worked well in the drop technique but in one patient where we used the spray technique, it was not satisfactory due to low volume. There was no suture hole bleeding in 10 patients, which has reduced OT time, ventilation time, blood and blood products, and suture usage. This has helped fora speedy postoperstive recovery and prevented blood product-related complications.
Conclusion: The appropriate type of glue application in indicated patients with the correct amount and technique improves patient outcomes by reducing morbidity related to bleeding and it is also cost effective.

1292: CARDIAC ARREST AS A CONSEQUENCE OF CARDIAC FIBROMA IN A PREVIOUSLY HEALTHY INFANT: CASE REPORT

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Background: Cardiac tumours are very rare and mostly benign. Cardiac fibroma ranks second in frequency and clinical manifestations depend on the degree of ventricular filling and obstruction of the left ventricular outflow tract.
Case report: MMC, a previously healthy 14-month-old male infant, was admitted to the emergency room of a nearby hospital in cardiac arrest and was promptly ressuscitaded after cardiopulmonary manoeuvres. The patient developed heart failure and low cardiac output in the following days. The chest X-ray revealed cardiomegaly and pulmonary congestion. The ecocardiogram with Doppler showed an image suggestive of cardiac tumour with regular margins occupying the middle part of the interventricular septum and extension to the apex and anterior wall of the left ventricle, measuring 4.9 × 4.8 cm and displaying signs of dynamic obstruction of the left ventricular outflow tract. Computerised tomography and magnetic resonance imaging of the chest also showed an image suggestive of a large cardiac tumour. Cardiac surgery was undertaken via the transaortic approach and the presence of a tumour affecting most of the left ventricular cavity, compromising the left coronary bed was visualised, thereby restricting its total resection. So we opted for partial resection of the left ventricular outflow tract. Pathology of the material revealed a cardiac fibroma. The child had progressive clinical improvement in the postoperatory period and was discharged from hospital within three weeks with no signs of left ventricular obstruction or heart failure and is doing well as an out-patient with low-dosage beta-blocker.
Conclusions: Cardiac tumours can lead to cardiac arrest and heart failure, and must be included in the diagnostic hypothesis.

1297: BIVENTRICULAR OUTFLOW TRACT RECONSTRUCTION: THE AORTIC TRANSLOCATION (NIKAIDOH) PROCEDURE VS THE RASTELLI PROCEDURE

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Objective: To compare the Rastelli (R) and the Nikaidoh (N) procedures in terms of early and mid-term results.
Methods: Betweeen January 2000 and January 2012, 23 patients underwent a biventricular outflow tract reconstruction at our institution, with a mean age of 3.8 years old (SD ± 2.7) and a mean weight of 14 kg (SD ± 4.6). The anatomical variants were transposition of the great arteries (TGA) and double-outlet right ventricle (DORV) with non-committed ventricular septal defect (VSD) and pulmonary stenosis (PS). Patients were divided into group I = Nikaidoh (10) and group II = Rastelli (13).
Results: The mean CPB perfusion time was 227 min (SD ± 64) and aortic cross-clamping was 150 min (SD ± 46), which was more prolonged in group I ($p = 0.006$). The mean mechanical ventilation, inotropic requirement and hospital stay was five days (QI 25–75% = 2–19), six days (QI 25–75% = 3–28) and 10 days (QI 25–75% = 7.7–30). During the immediate postoperative period: nine patients experienced ventricular dysfunction and nine arrythmias, without significant differences between both groups. There was no mortality in group I, whereas four patients died in group II ($p = 0.05$). The mean follow up was 4.6 years (QI 25–75% = 2–7). Arrhythmias ($p = 0.04$) and right ventricular outflow tract obstruction (RVOTO) ($p = 0.03$) were more frequent in group II. All of the Nikaidoh group were free of LVOTO, whereas three patients in the other group developed subaortic stenosis. None developed aortic insufficiency larger than mild. In group II: five interventional procedures ($p = 0.01$) and nine re-operations ($p = 0.004$) were needed.
Conclusions: In the Nikaidoh procedure, even though cross-clamping time was prolonged, patients remained free of major events and mortality. In the Rastelli procedure with non-committed VSD group: mortality, arrhythmias, RVOTO, interventional procedures and re-operations were more frequent. Aortic translocation techniques such as the Nikaidoh procedure seem promising, especially in the presence of inadequate anatomy for a Rastelli repair. Further studies are needed to confirm this in the long term and with a larger number of patients.

1298: SOUTHAMPTON PRELIMINARY EXPERIENCE IN THE USE OF AN EXTRACELLULAR MATRIX TO REPAIR CONGENITAL AND ACQUIRED HEART DEFECTS

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Background: Surgical correction of congenital heart defects are performed early in life. An ideal material for these corrections would

encourage tissue regeneration with growth potential. The quest for an optimal material for vascular and intracardiac tissue repair is ongoing. Here we describe our experience with an extracellular matrix.
Methods: From April 2011 to June 2012, 109 CorMatrix patches were used in 89 operations on 82 patients aged one day to 81 years. In 82 cases, the ECM was used for cardiac or great vessel repair: pulmonary arterioplasty in 34, intracardiac tissue repair in 16, pulmonary monocusp valve creation in 10, ascending aortoplasty in five, aortic arch augmentation in five, right ventricular outflow tract patch in five, superior vena cava patch in four and valve leaflet augmentation in four patients. In 27 cases, the ECM patch was used for pericardial closure. Follow up was complete.
Results: There were no deaths, and at a mean follow up of 7.1 months (1.1–15.4), there was no evidence of ECM-related intracardiac or intravascular thrombosis. Two patients had pericardial effusions due to bleeding from the anastomosis. Six patients who underwent pulmonary arterioplasty had some element of re-stenosis but later had successful balloon dilatation in the catheterisation laboratory. Eight of the 10 monocusp valves were competent and none were stenotic.
Conclusions: Repair of congenital and acquired heart defects using CorMatrix ECM is feasible and safe. We particularly like this product due to the way it curves and conforms to the native tissue. It is also amenable to balloon angioplasty. These early results are encouraging but longer follow up is needed to evaluate the ability to grow and to determine the full potential of this material.

1299: CARDIAC OUTPUT MONITORING USING FEMORAL ARTERIAL THERMODILUTION DURING LEVOSIMENDAN INFUSION IN A NEWBORN WITH MYOCARDITIS

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Background: The pulse-induced continuous cardiac output (PiCCO) system is a method of continuous measurement of cardiac output by thermodilution in the femoral artery and analysis of the pulse contour curve. Levosimendan (Levo) is a calcium sensitiser that exerts inotropic action by binding to cardiac troponin C and increasing the sensitivity of the contractile apparatus to calcium. There are few reports on the use of Levo in children with myocarditis and no data exist on cardiac monitoring using PiCCO during Levo infusion. We analysed haemodynamic variations with PiCCO during Levo administration in a newborn with myocarditis.
Methods and Results: A 10-day-old term baby with acute myocarditis and severe cardiac dysfunction (FS 15%, FE 25%) received a Levo loading dose of 12 µg/kg over 10 minutes, followed by an infusion of 0.2 µg/kg/min for 24 hours. The femoral artery was catheterised using a 3-Fr PiCCO arterial thermodilution catheter, and a femoral central venous line was inserted. A total of 10 haemodynamic measurements before and during Levo infusion was performed using a 3-ml bolus of cold normal saline injected rapidly through the central venous catheter. Cardiac index (CI), cardiac function index (CFI), stroke volume index (SVI), systemic vascular resistance (SVR), and global ejection fraction (GEF) were recorded. Indicators of blood volume were also measured. Mixed venous saturation (SvO_2), NIRS and serial measurements of troponin I and pro-BNP were obtained.
Conclusions: To our knowledge, this is the first report of continuous haemodynamic monitoring with PiCCO during Levo administration. Levo significantly improved cardiac output. No side effects secondary to PiCCO were observed.

1300: DEFIBRILLATOR SURGERY IN PATIENTS WITH CONGENITAL HEART DISEASE

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Introduction: Congenital heart diseases (CHD) may develop significant arrhythmias resulting in implantation of a cardioverter-defibrillator device (ICD). In this study, we analysed the procedures and the indications for ICD surgery.
Methods: In a 12-month period (January to December 2010) ICD surgery was performed in 28 patients with CHD, 15 were male and 13 were female. The age ranged from three to 50 years.
Results: Twenty patients (71%) had undergone surgery for the underlying CHD. A new ICD device was implanted in 15 patients (54%); one-chamber device in seven patients, two-chamber device in five, and three-chamber device in three. A previously implanted device was changed in four patients (one-chamber device: $n = 3$, two-chamber device: $n = 1$). In three patients the complete (two-chamber) device including electrodes had been exchanged (in two cases with a staged approach). An upgrade of an implanted pacemaker was performed in four patients (one-chamber device: $n = 1$ or two-chamber device: $n = 3$). Finally, the previously implanted ICD was upgraded in two patients (from one- and two-chamber devices to three-chamber device, respectively).
Conclusion: Surgery for ICD in patients with CHD is challenging and increasingly complex. Therefore careful follow up is mandatory.

1304: HAEMODYNAMIC EFFECTS OF WEANING FROM POSITIVE-PRESSURE VENTILATION IN PRETERM NEWBORNS

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Background: To study haemodynamic and echocardiographic changes during weaning from synchronised conventional ventilation (sCV) to nasal continuous positive airway pressure (nCPAP) (T1) and from nCPAP to spontaneous breathing (T2) in uncomplicated preterm newborns.
Methods: We conducted a prospective study on preterm infants with gestational age (GE) ≤ 32 weeks. Each subject had an echocardiographic evaluation (echo) respectively one hour before, and after T1 and T2, for assessing left and right ventricular output (LVO and RVO), superior vena cava (SVC) diameter and flow, left ventricular end-diastolic diameter and shortening fraction (SF). Cardiorespiratory vital parameters and emogas analysis were also collected just before each echo was performed. Patients with haemodynamically relevant PDA and/or needing pharmacological closure were excluded.
Results: We identified 16 patients (mean GE 27.6 ± 1.9 week). We observed a significant increase of RVO, superior vena cava flow and SVC diameter, both at T1 and T2. There were no significant variations of ductal morphology and flow patterns or significant association between different ventilatory support and both LVO and SF. No significant variations were found in cardiorespiratory parameters and emogas analysis.
Conclusion: T2 was associated with significant increase in pulmonary output and superior vena cava flow, as previously reported. We found that weaning from sCV produced significant haemodynamic effects as well, influencing the same echo parameters. All studied variations were clinically well tolerated with no changes in cardiorespiratory parameters and systemic perfusion. Further studies are planned to show haemodynamic changes and any clinical deterioration, at both T1 and T2, in more 'complicated' preterm newborns (i.e. large PFO or PDA, reduced left ventricular function).

1316: DOSE INTERVENTIONAL CLOSURE OF ISOLATED ATRIAL SEPTAL DEFECT WITH SEVERE PULMONARY HYPERTENSION IMPROVES THE LONG-TERM CLINICAL OUTCOME IN ADULT PATIENTS

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Objective: To examine whether closure of isolated atrial septal defects (ASDs) (secundum and sinus venosus type) in adults with severe pulmonary hypertension improved their long-term clinical outcome.
Methods: In a retrospective study, there were 1 423 adult patients with ASD. We examined the major cardiovascular events and overall mortality of 68 adult patients with isolated ASDs and severe pulmonary hypertension without Eisenmenger change. Sixty-two of them underwent interventional closure either by means of percutaneous transcatheter occluder or by open surgical repair. The remaining six were on medical treatment. We compared the major cardiovascular events, including death, stroke or paradoxical embolism, new-onset atrial fibrillation or atrial flutter, infective endocarditis, progressed pulmonary hypertension, pneumonia requiring hospitalisation, and functional class deterioration in these two groups. Severe pulmonary hypertension was defined as right ventricular systolic pressure ≥ 60 mmHg, measured by transthorasic echocardiography. Follow-up period was between one and 214 months.
Results: The survival analysis did not reveal significant differences in overall mortality ($p = 0.805$) and major cardiovascular events ($p = 0.308$) between the interventional-closure group and the medical treatment group. In the interventional-closure group, the risk of having major cardiovascular events was significantly higher in patients aged between 18 and 30 years ($p = 0.019$) and over 40 years ($p = 0.022$) compared with those aged between 30 and 40 years, although there was no significant difference in overall mortality between these age groups ($p = 0.108$).
Conclusion: Interventional closure may not be superior to medical treatment in improving major cardiovascular events and overall mortality in adults with isolated ASD and severe pulmonary hypertension. In the interventional group, age was an important predictor of later cardiovascular events. This may have been due to the early development of pulmonary hypertension in the younger group (age 18–30 years old) and chronic change of pulmonary vascular hypertension in the older group (over 40 years old).

1317: REPAIR OF CONGENITAL MITRAL VALVE MALFORMATION IN INFANTS

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Objectives: Due to the wide spectra of morphological abnormalities, associated cardiac anomalies and body size, surgical treatment of congenital mitral valve malformations in paediatric patients remains a therapeutic challenge. We reviewed our experiences with mitral valve repair in infants.
Methods: All consecutive patients with congenital mitral valve disease who underwent surgery between 1998 and 2010 were studied retrospectively. Patients with atrio-ventricular septal defects, atrioventricular discordance, and functional single-ventricle anomalies were excluded.
Results: Between 1998 and 2010, 12 children (seven boys and five girls) with a median age of 5.2 months (interquartile range 2.1–8.1 months) underwent surgery. The median follow-up time was 3.9 years (interquartile range, 1.2–8.9 years). Twelve patients (52.2%) were under one year old. Ten patients (83.3%) were diagnosed with mitral valve incompetence, while two (16.7%) were diagnosed with stenosis. We attempted to repair the mitral valve in all patients, except one with Shone's complex and hammock valve. Associated cardiac lesions were presented in eight patients (66.7%). Five patients (41.7%) required re-operation, including three during the initial hospitalisation and two during the follow-up period. All patients who required re-operation needed mitral valve replacement with a mechanical prosthesis, and the mitral valves appeared extremely malformed in these patients during the first operation (hammock valve in two patients and parachute mitral valve in one). Overall, the success rate of mitral valve repair was 58.3% (seven patients). There were no early or late deaths, and all survivors were categorised according to the New York Heart Association classification system as class I or II, with mild or less mitral dysfunction.
Conclusion: Although mitral valve repair in infants with congenital mitral valve disease is a challenging procedure, low surgical mortality can be achieved. Despite the severity of mitral valve dysplasia, mitral valve repair should always be attempted. Patients with extremely malformed mitral valves have a significantly higher risk of requiring re-operation ($p = 0.03$, odds ratio = 9.33). Initial mitral valve repair can provide time for future mitral valve redo operations.

1318: HAND-MADE GORE-TEX VALVED CONDUIT FOR RIGHT VENTRICULAR OUTFLOW TRACT RECONSTRUCTION FOR PATIENTS WITH SEVERE PULMONARY REGURGITATION AFTER TOTAL CORRECTION OF TETRALOGY OF FALLOT

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Background: Surgical management of tetralogy of Fallot (TOF) results in anatomical and functional abnormalities in the majority of patients. Although right ventricular (RV) volume load due to severe pulmonary regurgitation (PR) can be tolerated for years, there is now evidence that the compensatory mechanisms of the right ventricular myocardium ultimately fail and that if the volume load is not eliminated or reduced, the dysfunction may be irreversible.
Methods: This was a retrospective chart review study. From 2008 to 2012, there were 32 patients receiving pulmonary valve replacement surgery due to severe pulmonary valve regurgitation and RV failure decades after total correction for TOF. Among them, 15 patients received hand-made valved Gore-Tex conduit reconstruction.
Results: Thirteen patients received our handmade tri-leaflet conduit reconstruction with tissue valves and two received monocuspid valved conduit reconstruction with the outflow tract diameter around 20–24 mm. Three patients received intra-operative radiofrequency ablation for pre-operative arrhythmia. There was no surgical morbidity or mortality in our series. During follow up, they all remained in New York Heart Association functional class I. The echocardiography revealed mild residual pulmonary regurgitation.
Conclusion: Pulmonary valve replacement with handmade valved tricuspid Gore-Tex conduits provides similar short-term outcomes when compared with traditional bioprosthesis and may avoid early graft calcification in growing adolescents. With this design, we can also provide a better landing area for later transcutaneous pulmonary valve implantation. However, we need further follow up to analyse our long-term surgical results.

1319: TRENDS IN EARLY RESULTS AFTER FONTAN SURGERY: THE ENTIRE AUSTRALIAN AND NEW ZEALAND EXPERIENCE OF 1 030 PATIENTS

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Objectives: To examine changes in Fontan surgerical practice in a population-based dataset and to analyse factors impacting on early outcomes.
Methods: The databases of all congenital cardiac centres in Australia and New Zealand were screened for patients who had undergone a Fontan procedure. A total of 1 087 patients were identified and 1 030 had sufficient available peri-operative data to be included in the audit. Peri-operative characteristics were analysed for their impact on mortality in hospital or within 30 days, Fontan failure (death, revision, takedown or mechanical support), prolonged or significant pleural effusion (lasting > 30 days or requiring re-operation) and prolonged length of stay (> 30 days).
Results: Since its introduction in 1975, the Fontan procedure has been increasingly performed in Australia and New Zealand with a peak of 2.5 Fontan procedures per one million inhabitants in 2006. The extracardiac technique (EC) has now been exclusively adopted: 234 atrio-pulmonary (AP) connections, 1975–1995; 288 lateral tunnel (LT), 1988–2006; and 508 EC, 1997–2011. The proportion of patients with hypoplastic heart syndrome (HLHS) rose throughout the study period (1% prior to 1991, 3% 1991–2000, 17% after 2000). After risk adjustment, early outcomes were similar between LT and EC and worse for AP. The only additional independent risk factors for mortality and Fontan failure were dextrocardia (OR 3.2, $p = 0.06$ and OR 2.7, $p = 0.03$) and pulmonary artery pressure (PAP) ≥ 15 mmHg (OR 3.2, $p = 0.02$ and OR 2.4, $p = 0.01$). HLHS morphology was an independent risk factor for prolonged hospital stay and significant effusions (OR 3.4, $p < 0.001$ and OR 3.2, $p = 0.01$, respectively).
Conclusion: The Fontan procedure is increasingly performed and is consuming a larger proportion of resources because its rise is driven by the larger proportion of patients surviving with HLHS. HLHS patients stay longer in hospital because of prolonged effusions. Early outcomes were similar after the LT and EC techniques.

1332: POSTOPERATIVE RESULTS OF SECONDERLY REPAIR OF RECURRENT RIGHT VENTRICULAR OUTFLOW TRACT LESION USING THE EPTFE MONOCUSPED OUTFLOW PATCHING FOR OLDER CHILDREN AND ADULTS

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Background: Chronic pulmonary insufficiency is a common problem after relieving right ventricular outflow tract obstruction. The importance of a competent pulmonary valve to preserve cardiac performance has been emphasised. However, controversy remains with regard to the best prosthesis to implant. We have expanded the use of ePTFE monocusp for re-operation for recurrent right outflow tract lesion in older children and adults.
Objective: The aim of this study was to evaluate the long-term results of ePTFE monocusped pulmonary valve, and to show the validity of the use of the ePTFE monocusp for older children and adults.
Methods: From 1997, 24 patients (mean 16.9 ± 6.8 years, range 11–36) underwent redo RVOT reconstruction using ePTFE monocusp at our institution. The primary diagnoses were TOF in 13, PA VSD in three, DORV in two, TGA in three, truncus in one, PA IVS in one, and LTGA in one. Seven had had valved conduit repair, and two *in situ* PVR. The patients underwent re-operation in the early postoperative period (within five years), and pre-adolescents (under 10 years old at re-operation) were excluded. The follow-up period was 11.2 ± 3.3 years.
Results: There was one late death due to non-cardiac disease. One patient had RVOT revision due to IE. Another patient had BCPS followed by RV aneurysmectomy and CRTD implantation due to RV failure. The ePTFE monocusp of both patients functioned well at re-intervention. Except for the patient who had BCPS, all patients showed good QOL at the last follow up. Actuarial freedom from re-intervention for RV lesion was 91.4± 5.7%, and freedom from ePTFE valve-related re-operation was 100% at 10 years.
Conclusions: Long-term results after pulmonary valve repair using the ePTFE monocusp were satisfactory. The ePTFE monocusp could be the first choice for even older children and adults.

1336: CHRONIC THORACIC PAIN IN CHILDREN AFTER CARDIAC SURGERY

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Background: Chronic pain after cardiac surgery in adults is common but has gained little attention in children. The aim was to investigate the prevalence and character of chronic pain after cardiac surgery via median sternotomy in children.
Methods: We carried out a prospective clinical examination with quantitative sensory testing three months after surgery, and a retrospective survey of children who had undergone cardiac surgery 10–60 months earlier. The questionnaire assessed pain descriptors, situations or activities that could worsen pain, and analgesic consumption. Faces pain scales (Bieri) were used to rate the pain intensity.
Results: Fourteen children were examined three months after surgery. One child reported pain. Brush allodynia and pin-prick hyperalgesia were present in five and nine children respectively, hypo-aesthesia to cold (20°C) was present in three children, and one child had cold allodynia in the scar area. The average pressure pain threshold was 80.4 kPa (range 34.3–127.7 kPa); 171 questionnaires were sent out, and 121 questionnaires (70.8%) were eligible for analysis. Worst pain intensity in the week after surgery was 5.6 (mean). Pain 'during the recent week' was rated positive by 26 children. Pain was evoked by pressure against the wound in 23 and by physical activity in 15 children. Itch and pressing were the most frequent pain descriptors chosen by 25 and 20 children, respectively. One child used paracetamol once a week. A history of two sternotomies increased the risk of complaints of pain; 15 complaints in 26 children compared with 28 complaints in 79 children who had undergone sternotomy only once. More than two sternotomies did not relatively increase the number of complaints.
Conclusions: The prevalence of chronic pain following cardiac surgery via median sternotomy in children was lower than in the adult population. The pain may have a neuropathic component, but appeared to be mild.

1348: ARTERIAL-SWITCH OPERATION FOR COMPLEX TRANSPOSITION OF THE GREAT ARTERIES: OUTCOMES IN ADULT PATIENTS

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Arterial switch operation (ASO) is the leading procedure for treatment of complex forms of transposition of the great arteries (TGA) associated with ventricular septal defect with/without aortic arch obstruction. This prospective study evaluates the status of survivors at adult age.
Methods: Among 688 patients who were operated on at our hospital for complex TGA between 1982 and 2011, 103 had reached adult age (> 18 years). All had haemodynamically significant ventricular septal defect, 23 had an aortic arch obstruction including coarctation (22 patients), and interrupted aortic arch (one). In 20 patients (19.4%), a two-stage management was performed after an initial palliative pulmonary artery banding and aortic arch repair when necessary.
Results: During a mean follow up of 19.2 ± 4 years, two late deaths occurred (1.94%, 95 CI: 1.92–2.02%), respectively, at five and 10 months postoperatively, both patients awaiting re-operation for severe aortic valve insufficiency. Actuarial survival was 97.1% at 10, 20 and

25 years. Permanent pacemaker implantation was required in four patients. Re-operations were performed in 22 patients (21.4%, 95 CI: 17–52%), mainly for pulmonary stenosis (seven), aortic valve insufficiency (four) and left ventricular outflow tract obstruction (three). Freedom from re-operation at 10, 20 and 25 years was 90.2, 76.9 and 65.7%. At last follow up, all patients were asymptomatic with normal left ventricular function. Aortic root diameters were collected in 52, of whom 29 (55.8%) had an aortic root dilatation (> 2 SD), which occurred more frequently in those with initial aortic arch obstruction compared to those without (respectively 86.7 vs 43.2% of patients, $p = 0.0043$).
Conclusion: Close to 20 years after ASO for complex TGA, late outcome was encouraging, with no deaths after the first year of life. However, the aortic root may dilate with time, more likely in patients with initial aortic arch obstruction, who justify close follow up.

1351: MODIFIED NIKAIDOH OPERATION FOR D-TRANSPOSITION OF THE GREAT ARTERIES WITH A VENTRICULAR SEPTAL DEFECT AND PULMONARY STENOSIS USING 'OVERSIZED' CONDUITS

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Objective: Our seven-minute film demonstrates the surgical technique of aortic root translocation with arterial-switch procedure and coronary artery re-implantation (modified Nikaidoh operation) for d-transposition of the great arteries with VSD and left ventricular outflow tract obstruction in a four- month-old patient (5.5 kg) previously palliated with systemic-to-pulmonary shunt.
Methods: The surgical approach consisted of harvesting the aortic root from the right ventricle, and the coronary arteries were excised as circular buttons. The pulmonary valve was excised and the conal septum was divided completely. The VSD was closed with a triangular-shaped patch. The aortic root autograft was then rotated 180° and was sewn to the LV outflow. Re-implantation of the coronaries and Lecompte manoeuvre were done, as in the arterial-switch procedure. The right ventricular-to-pulmonary artery continuity was re-established using a pulmonary homograft.
Results: The bypass time was 227 minutes, the cross-clamp time was 147 minutes. The patient was extubated in the operating room and had an uneventful postoperative period and excellent echo results.
Conclusion: Aortic translocation results in a more normal anatomical repair compared with the Rastelli operation. It is always possible to place an oversized conduit (homograft) with little risk of sternal compression. Individual coronary artery transfer during translocation may prevent coronary insufficiency.

1365: TRUNCUS ARTERIOSUS – EARLY VERSUS LATE INITIAL INTERVENTION: SINGLE CENTRE EXPERIENCE, OUTCOMES AND MANAGEMENT OVER MORE THAN 10 YEARS

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Background: Truncus arteriosus (TA) continues to be associated with significant morbidity and mortality. Neonatal primary complete repair has progressively become the treatment of choice for TA with encouraging survival rates. Our aim was to compare and contrast initial and late outcomes following surgery during the neonatal period (< 30 days) and infancy (> 30 days).
Methods: At a tertiary cardiac centre between 2001 and 2012, 51 patients with TA were reviewed in terms of staged versus primary complete repair, early versus late mortality and re-intervention rates. Associated cardiac anomalies were severe truncal regurgitation ($n = 4$), non-confluent pulmonary arteries ($n = 5$), interrupted aortic arch ($n = 5$), coarctation of the aorta ($n = 1$) and double aortic arch ($n = 1$).
Results: Fifty-one patients presented with TA between 2001 and 2012. Median weight was 3.14 (1.7–4.4) kg and median age was 22 (7–265) days; 49/51 were operated with 2/51 pre-operative deaths (seven, 13 days). Five patients had staged repair with subsequent surgeries within six months and none required further re-intervention. In the primary complete repair group, three patients had severe residual truncal regurgitation, while two had blocked conduits with infective endocarditis. Nine/33 (27%) required re-intervention: three conduit replacements, two truncal re-repairs and four trans-catheter pulmonary artery dilatation. The mean duration of PICU stay post-operatively was 10 (2–152) days, 43% were discharged within seven days and 32% within 15 days.
Conclusion: There was no significant difference ($p = 0.72$) in early or late mortality between the repair strategies. Alternative strategies such as staged repair will need to be explored for possible improved outcomes.

1383: SERUM CONCENTRATIONS OF PROCALCITONIN AFTER TETRALOGY OF FALLOT CORRECTION

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Background: Cardiopulmonary bypass in paediatric cardiac surgery may cause a systemic inflammatory response syndrome (SIRS). Clinical and laboratory signs of SIRS may mimics sepsis. An increase in procalcitonini (PCT) has been a known potential biomarker of postoperative infection, but may also increase in children after cardiac surgery.
Aim: To establish the baseline levels of PCT after open-heart surgery in order to analyse a possible induction of the inflammatory response that might interfere with the diagnosis of infection by PCT.
Methods*:* Serum samples were collected from nine out of a total of 32 tetralogy of Fallot patients who underwent total correction and showed signs of SIRS 24 hours postoperatively. Blood, sputum and urine specimens for culture were also studied. Patients were followed for the development of postoperative complications.
Results*:* The mean PCT value was 89.55 ng/ml (range 1.69–371) (reference value < 0.1 ng/ml). Only two patients showed positive culture from their sputum (patient with PCT 2.74 ng/ml showed *Acinetobacter pneumoni*, and patient with PCT 7.05 ng/ml showed *Streptococcus viridans* plus *Klebsiela pneumoni*).
Conclusions*:* An increase in PCT levels was observed in the first postoperative day after cardiac surgery in all patients with SIRS, however only 22% had proven infection.

1388: THE DISSOCIATION OF CEREBRAL TISSUE AND CENTRAL VENOUS OXYHAEMOGLOBIN SATURATION IN LOW-FLOW STATES DURING ECMO: EVIDENCE FOR CEREBRAL AUTOREGULATION

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Background: We report the relationship between central venous (S_vO_2) and cerebral tissue (S_cO_2) oxyhaemoglobin saturation during a case of veno-arterial ECMO. Problems with the arterial cannula resulted in periods of low systemic blood flow, conditions in which this relationship has not previously been studied. Transcranial near infra-red spectroscopy (NIRS) provides a non-invasive measurement of S_cO_2, which correlates positively with S_vO_2 and has been used in *lieu* to detect inadequate systemic oxygen delivery in neonates with hypoplastic left heart syndrome. NIRS is non-invasive, continuous and avoids the complications of indwelling catheters. However, its relationship with S_vO_2 is not well understood and may have wide limits of agreement, especially at lower values ($S_vO_2 < 50\%$). This observation may represent the influence of cerebral autoregulation.
Methods: A 6-kg, 14-month-old child underwent repair of mitral

stenosis. At the end of surgery, it was not possible to wean her from bypass due to severe pulmonary hypertension. Veno-arterial ECMO was instituted, which was initially complicated by excessive bleeding and cannula malposition. Data were collected retrospectively. Cerebral oximetry was compared to continuous monitoring of the ECMO venous drainage cannulae (S_vO_2) over the first 500 minutes.
Results: There were 20 epochs (median duration 150 seconds) of acute severe reduction in arterial ECMO flow (> 10% from baseline). The median drop was 58%. This was associated with a much more pronounced fall in S_vO_2 (median 55% from baseline) compared to S_cO_2 (median 14% from baseline).
Discussion: There was dissociation of S_vO_2 and S_cO_2 during low-flow states. This suggests oxygen delivery to the brain is preserved and supports the concept of cerebral autoregulation. S_cO_2 may be maintained when global oxygen delivery is critically low and should not be used in place of S_vO_2.

1390: ANESTHETIC TECHNIQUE FOR TRANSOESOPHAGEAL ELECTROPHYSIOLOGICAL STUDIES IN PAEDIATRIC PATIENTS WITH WOLFF-PARKINSON-WHITE SYNDROME

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Objective: All patients with Wolff-Parkinson-White syndrome require risk assesement to determine their potential for rapid conduction down the accessory pathway during atrial fibrillation resulting in sudden cardiac death. Transoesophageal electrophysiological studies (TEEPS) are an effective risk-stratification tool. The purpose of this study was to describe the least invasive and most effective anaesthetic technique for transoesophageal electrophysiological studies.
Methods: A retrospective review was done of anaesthetic techniques used during TEEPS. Inclusion criteria were: pre-excitation on ECG; age < 18 years; and no history of tachycardia, palpitations, or syncope. Patients had TEEPS under anaesthesia either as IV sedation, monitored anaesthesia care (MAC), or general anaesthesia (GA). Anesthetic technique (MAC vs GA) and airway management decisions were left to the anaesthesiologist. Midazolam, fentanyl and propofol were used in various combinations. Sevoflurane was used during the induction period in all GA cases and stopped 10 minutes prior to initiation of TEEPS.
Results: Inclusion criteria were met by 20 patients with an average age of 11.9 years, average weight of 48.9 kg and average height of 149.2 cm. IV sedation was performed on 15%, MAC on 10%, and GA on 75% of patients. Airway management techniques included 13.3% LMA, 20% endotrachael tube (ETT) and 66% mask. IV sedation as initial anaesthetic was found to be cumbersome and uncomfortable. Next was ETT and LMA but trouble pacing was encountered due to positional change of the oesophagus relative to the left atrium during positive-pressure ventilation. Mask induction was then performed in the remaining 10 patients with a TEEPS probe inserted through a nare while the anaesthesiologist continued mask ventilation. All were successful without complications.
Conclusions: Mask anaesthesia was the preferred method due to its overall simplicity and minimal interference with the TEEPs procedure.

1396: TRUNCUS ARTERIOSUS: REVIEW OF SURGICAL REPAIR OF CASES DONE AT THE WITWATERSRAND GROUP OF HOSPITALS FROM 1974–2012

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Background: Truncus arteriosus is an uncommon cardiovascular anomaly characterised by a single arterial trunk with a single semilunar valve and supplying blood flow to the aorta, pulmonary arteries and coronary arteries. Associated complex cardiac lesions and pulmonary hypertension lead to high morbidity and mortality rates.
Methods: This was a retrospective review of 53 patients who underwent surgical repair of the truncus arteriosus between 1974 and 2012. Data related to clinical features, diagnosis, operative procedures, preoperative and postoperative follow up were collected.
Results: Fifty-three patients (23 males and 30 females) underwent surgery to repair the truncus arteriosus. The most common presenting features were dyspnoea, tachypnoea, cardiac murmurs, congestive cardiac failure, shock and cyanosis. The median age at surgery was eight months (range: 3 weeks – 7 years). Median weight at surgery was 4.8 kg (range: 2.1–14.8 kg). Diagnosis was made with echocardiography in all patients and confirmed on angiography in 31 patients. Patients were classified according to Collert and Edwards as follows: 26 with type I; 24 with type II; two with type III, and one was unclassified. Additional surgery undertaken during primary repair included interrupted aortic arch repair in three patients and truncal valve repair in two patients. Seven patients (13.2%) had moderate to severe truncal valve regurgitation.The median cross-clamp time was 91 minutes and the median cardiopulmonary bypass time was 134 minutes. The right ventricular outflow tract was reconstituted using a bioprosthesis in 32 (60.4%) and a homograft in 21 (39.6%). Eight patients had subsequent surgery: seven required second pulmonary outflow replacements and one patient truncal valve replacement. Early mortality post primary repair was 24.5% including two patients who required interrupted aortic arch repairs. The major cause of early mortality was pulmonary hypertensive crisis.
Conclusion: Truncus arteriosus is associated with a high operative mortality if primary repair is delayed. A delay in the presentation associated with established pulmonary hypertension may be a contributing factor.

1403: CARE OF A VAD PATIENT ON A WARD HDU: A MULTIDISCIPLINARY PERSPECTIVE

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Background: Mechanical circulatory support is an important adjunct to the treatment of children with advanced heart failure. We are currently the second largest centre in the world for mechanical support and have been implanting the Berlin heart (our institution's preferred support device) since 2005. The research undertaken looks at the most suitable environment in which to nurse the stable paediatric VAD patient, comparing PICU ward care and HDU ward care.
Methods: A questionnaire was designed to look at four key issues: child development, emotional and psychological wellbeing, staffing/education, and cost implications. These questionnaires were completed by children, parents and carers. Members of the MDT all involved in the care of this patient group also completed questionnaires or underwent interviews.
Results: This study is ongoing we are currently gathering data.
Conclusions: As this study is ongoing, it is difficult to draw conclusions. However, so far, the research shows the most suitable environment in which to nurse this patient group is on a ward HDU. The availability and access to other service providers is better. The children nursed in this area also have more access to play and education facilities, therefore providing improved development. Emotionally the children and their families can adapt to a more 'normal' routine, parents become more involved in the care of their child, and the need for night time sedation is less frequent. The data we are currently gathering relates to funding and cost implications.

1418: POSTOPERATIVE FEEDING PRACTICE AND WEIGHT GAIN IN NEONATES POST ARTERIAL-SWITCH OPERATION

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Background: The progression of nutrition and its impact on feed tolerance and weight gain in postoperative transposition of the great arteries (TGA) babies is not widely studied.
Methods: A retrospective chart review was conducted to observe feeding practice in postoperative TGA infants between 2007 and 2012. A standardised feeding programme was not in place during the study period. Sixty-five infants aged 39 ± 2 (36–42) weeks' gestation underwent an arterial-switch procedure at a median age of nine (3–88) days. The progression of feeds in the postoperative period was documented.
Results: Total fluid intake (TFI) progressed to demand feeding (breast and/or bottle) by ward day 2 ± 2 (0–11), and by postoperative day 12 ± 11 (3–63). On transfer from intensive care to the ward, TFI was 131 ± 14 (100–150) ml/kg/day, of which 60% of infants were naso-gastric (NG) fed, 28% naso-jejunally (NJ) fed and 12% either parenteral or orally fed. There was no observed impact on incidence of feeding intolerance in those NJ versus NG fed, or those receiving hypercaloric feeds versus standard concentrations. Thirty-seven per cent of infants received one or more feeding-related referrals. It took 11 ± 6 (5–29) days until weight gain was achieved postoperatively. Age-appropriate weight gain (15–30 g/day) was achieved in 47% of infants by the time of discharge [11 ± 6 (5–29) days]; however 58% of patients were discharged below birth weight. Infants were discharged home on either oral (46%) or enterally supported (54%) feeds. Feeds consisted of mother's milk (49%), formula (14%), hypercaloric mother's milk (28%) or hypercaloric formula (6%).
Conclusions: More than half of infants were transferred or discharged home below birth weight. This suggests that more aggressive feeding may improve weight gain outcomes post TGA surgery. Standardised feeding progression to establish age-appropriate weight gain may be beneficial.

1420: SURGICAL INTERVENTION IN PATENT DUCTUS ARTERIOSUS IN NIGERIA

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Background: Congenital cardiac diseases are not uncommon in the Nigerian population and patent ductus arteriosus (PDA) accounts for a significant proportion of this burden. The management is still a challenge due to late presentation, financial constraints and availability of expertise and facilities. This surgically amenable anomaly can be dealt with in Nigeria with currently available expertise and facilities. This audit was done to review the division's experience and determine outcomes.
Methods: A retrospective review was done of patients with PDA operated on between July 2006 and June 2012. Data obtained included demographics, echocardiographic features, and intra- and postoperative data.
Results: There were nine cases done during this period, three males and six females. Median age at presentation and at surgery was 3.5 and nine months, respectively. Most (89%) had recurrent chest infections and failure to thrive. Three patients (33.3%) had associated anomalies. The median pre-operative weight was 5.6 kg. All the patients had left thoracotomy and PDA ligation under general anaesthesia. The median PDA size intra-operatively was 5.5 mm and 66% (six patients) had short, stout PDA morphology. All had chest tubes inserted intra-operatively. One patient had a lacerated PDA intra-operatively with primary haemorrhage. Five patients (55.5%) stayed in the intensive care unit (ICU). The median hospital stay was nine days and all were discharged home alive and well.
Conclusion: Patent ductus arteriosus can be safely managed with currently available expertise and facilities. We have excellent outcomes.

1423: LEFT THORACOSCOPIC SYMPATHECTOMY FOR CARDIAC DENERVATION IN CHILDREN WITH LIFE-THREATENING VENTRICULAR ARRHYTHMIAS

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Background: The sympathetic nervous system plays a prominent role in the development of many life-threatening ventricular arrhythmias. In recent times, a small number of limited case series have reported good long-term benefit using video-assisted thoracoscopic surgery (VATS) for left cardiac sympathetic denervation (LCSD) in patients with drug-refractory ventricular arrhythmias; predominantly those with long-QT syndromes and catecholaminergic polymorphic ventricular tachycardia (CPVT). It remains unclear how effective this minimally invasive surgical treatment is and whether it might be indicated in all paediatric patients with symptomatic ventricular arrhythmias.
Methods: We conducted a retrospective clinical review of all patients who underwent left cardiac sympathetic denervation by means of video-assisted thoracoscopic surgery at our institution. From August 2000 to December 2011, 24 paediatric patients (13 long-QT syndrome, nine catecholaminergic polymorphic ventricular tachycardia, and two idiopathic ventricular tachycardia) were identified from the cardiology database and surgical records.
Results: There were no intra-operative complications, and blood loss was minimal. Median post-operative length of stay was 2 ± 6 days. There were no major peri-operative complications. Longer-term follow up was available in 22 of 24 patients at a median follow-up time of 28 months (range 4–131 months). Sixteen (73%) of the 22 patients experienced a marked reduction in their arrhythmia burden, while 12 (55%) became completely arrhythmia-free after sympathectomy treatment.
Conclusions: Video-assisted thoracoscopic left cardiac sympathetic denervation can be safely and effectively performed in most children with life-threatening ventricular arrhythmias. This minimally invasive procedure is a promising adjunctive therapeutic option that achieves a beneficial response in the majority of symptomatic patients.

1434: HYBRID STRATEGIES FOR HIGH-RISK NON-HLHS PATIENTS: ROLE OF BILATERAL PULMONARY ARTERY BANDING WITH AND WITHOUT STENTING OF THE PDA

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Background: Hybrid procedures involving bilateral pulmonary artery banding (bPAB) and stenting of the PDA in HLHS are well described, but their use in other diagnostic categories is rarely reported.
Methods: Three neonates with IAA + truncus arteriosus, IAA + extreme prematurity and IAA + tricuspid atresia + DOLV, respectively, underwent a staged repair with initial palliative procedure.
Results: Patient 1 presented with a birth weight of 2.6 kg and diagnosis of persistent truncus arteriosus (Van Praagh type A4). Due to poor response to prostaglandin it was decided to proceed on day 3 of life with PDA stenting and bPAB, using sections of a 3-mm Gore-Tex™

tube. Five months later the patient underwent complete repair that included re-implant of an occluded left subclavian artery. At three months, normal biventricular function, mild AR and no pulmonary artery stenosis were demonstrated.

Patient 2, a 28-week twin baby with birth weight 1.2 kg, presented with IAA and underwent bPAB while on NICU using sections of a 3-mm Gore-Tex™ tube, to a mean systemic pressure of 30 mmHg and oxygen saturations in the 70s. Four months later, having reached the weight of 4.2 kg, the baby successfully underwent a full repair. At two years the baby is well and thriving although has required balloon dilatation of both pulmonary arteries.

Patient 3 presented with a background of DiGeorge syndrome, IAA, HRHS, TGA and severe systemic AV valve regurgitation. On day seven the baby underwent bPAB while on prostaglandin infusion. Five weeks later the baby underwent complete repair. Despite repeated attempts at correcting the persistent AV valve insufficiency the child died, aged three months, due to low cardiac output.

Conclusions: Manoeuvres aimed at limiting blood flow and maintaining patency of the duct can be used successfully in neonates with complex anomalies and prohibitive operative risks. A more accurate patient selection might improve survival rates.

1436: SINGLE-STAGE TWO-INCISION TECHNIQUE FOR THE MANAGEMENT OF COARCTATION WITH INTRACARDIAC DEFECT

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Background: Management of coarctation and intracardiac defect presents technical options that are not without complications. The optimal surgical strategy for the management of the neonate and infant with coarctation of the aorta and intracardiac defect is highly debated. The risks of the various surgical options need to be considered in the choice of technique.

Methods: A retrospective review was done of case notes, operation reports and intensive care unit (ICU) charts of neonates and infants admitted with the above diagnosis. A two-incision, single-procedure strategy was used: first a left thoracotomy to repair the coarctation and then, with the patient supine, a median sternotomy to correct the intracardiac defect. This is preferred to concomitant pulmonary artery banding or a median sternotomy with circulatory arrest.

Results: Eight patients were enlisted into the study with a male:female ratio of 1:0.6. Median age was 21.2 weeks (range 5.0–315.0) and median weight was 5.5 kg (range 3.3–21.0). Median cardiopulmonary bypass time was 51.0 minutes (range 44.0–77.0) and median aortic cross-clamp time was 33.0 minutes (range 27.0–55.0). Median length of stay in the ICU was 11 days (range 3.0–17.0). The intracardiac defects were ventricular septal defects (VSD) in four patients (50.0%), VSD, patent ductus arteriosus (PDA) and patent foramen ovale (PFO) in two patients (25.0%), while one patient each (25%) had VSD, PDA and double-outlet right ventricle (DORV), and VSD, PDA and atrial septal defect (ASD), respectively. There were no early or late deaths.

Conclusions: Single-stage, two-incision technique for the repair of coarctation and intracardiac defect achieves good results while avoiding the risks of pulmonary artery banding and circulatory arrest.

1442: THE EFFECT OF ACUTE-ANGLE CORRECTION ANGIOPLASTY (LEFT PULMONARY ARTERY BED EXTENSION) FOR LEFT PULMONARY ARTERY STENOSIS IN PATIENTS WITH TETRALOGY OF FALLOT

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Background: Left pulmonary artery (LPA) stenosis with acute angulation is the most common indication of re-operation following total correction of tetralogy of Fallot (TOF). We investigated the surgical outcome of acute-angle correction angioplasty in this study.

Methods: Among 183 patients who received total correction of TOF over the last 11 years, 53 patients underwent the surgical repair for LPA stenosis as a concurrent procedure (M:F = 28:25). The type of LPA stenosis was classified into two groups: LPA os acute-angle group (n = 29) (group I) [os focal (juxtaductal) (n = 20) and diffuse stenosis (n = 9)] and os obtuse-angle group (n = 24) (group II) [os focal (n = 14) and diffuse stenosis (n = 10)]. Acute-angle correction angioplasty (LPA bed-extension technique to turn the acute angle of the os into an obtuse angle) in group I and conventional patch angiography in group II, was performed for stenosis relief.

Results: There was no early or late mortality. Median follow-up duration was 1.8 years. There was no statistical difference [os focal stenosis (group I: 41.0%, n = 19), (group II: 41.2%, n = 11, p = 0.952), diffuse stenosis (group I: 25.4%, n = 7), (group II: 36.8%, n = 7, p = 0.113)] in the latest left lung perfusion scan. Z-score improvement of the os after surgery did not reach statistical significance between the two groups [os focal stenosis (group I: 2.0, n = 9), (group II: 2.9, n = 7, p = 0.615), diffuse stenosis (group I: 2.5, n = 8), (group II: 3.1, n = 7, p = 0.694)]. There was no statistical difference in re-operation or intervention [os focal stenosis (group I, n = 1, 5%), (group II, n = 1, 7.1%, p = 1.00), diffuse stenosis (group I, n = 4, 47.4%), (group II, n = 4, 40.0%, p =1.000)].

Conclusions: Acute-angle correction angioplasty carries prognostic implications for surgical outcome in patients with TOF undergoing surgical repair for LPA stenosis with acute angulation.

1448: EXPERIENCE WITH NUNN'S POLYTETARFLUOROETHYLENE (PTFE) BICUSPID VALVES IN RIGHT VENTRICULAR OUTFLOW RECONSTRUCTION FOR TETRALOGY OF FALLOT AND RELATED ANOMALIES

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Background: Much attention and many innovations have been focused on minimising postoperative pulmonary regurgitation in the repair of tertralogy of Fallot (TOF). Various valved outflow patches have been developed with variable beneficial but limited effects.

Methods: We employed a bicuspid polytetrafluoroethylene (PTFE) valve developed by Nunn, published in *JTCS* in 2008, in 20 cases with TOF or similar anomalies since 2007. Patients were aged eight month to 14 years (median 18 months) and weighed 6.3–36.8 kg (median 9.4). This valve, made intra-operatively on the operating table, consists of seagull-shaped, wide bicuspid leaflets of 0.1-mm thick PTFE with its middle free margin fixed to the posterior wall of the main pulmonary artery (MPA). Valve competency was assessed with ratio between velocity–time integrals of regurgitant and forward flows at the pulmonary annulus level.

Results: Reconstructed annulus size was 122% of expected normal (91–186). Echocardiographic evaluation in the early and mid-term results showed satisfactory valve function with regurgitant–forward flow ratio of 24% (5–85), subgrouped as trivial in one, mild in 17 and moderate in two patients. Median peak gradient across the valves was 20 mmHg (range 0–43). Cusp motion and trans-valvar flow characteristics were well demonstrated on 2DE imaging.

Discussion: Mid-term results so far are satisfactory in terms of valve competency. This valve has the benefit of simple reproducibility, easy handling, good function and is expected to offer better long-term outcomes than the conventional monocuspid patch. When the valve becomes stenotic from somatic overgrowth, the posterior fixation is amenable to be ablated with a balloon catheter, although substantial regurgitation could evolve.

Conclusion: This novel Nunn's bicuspid valve could be a good alternative, with at least satisfactory mid-term function, to other conventional valves for TOF, with similar anomalies requiring a trans-annular patch. Close observation and further evaluation are warranted.

1457: DOUBLE-ROOT SWITCH: TOTAL ANATOMICAL CORRECTION FOR TRANSPOSITION OF THE GREAT ARTERIES

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Introduction: Aortic and pulmonary root translocation are alternative surgical procedures used to correct several congenital defects. To correct transposition of the great arteries, ventricular septal defect and pulmonary stenosis (TGA/VSD/PS), we proposed the double-root translocation with preservation of the pulmonary valve (DRTPV). We are now proposing the double-root switch (DRS) to correct transposition of the great arteries (TGA).
Methods: From March 2006 to September 2011, 10 children with TGA/CIV/PS were subjected to some kind of translocation root. Group A: five patients underwent the Nikaidoh procedure, five were males, three newborn. The reconstruction of the right ventricular outflow tract was done with a valved conduit. Group B: five patients underwent the DRTPV procedure, three children were males, one newborn. Group C consisted of two neonates with simple TGA who submitted to DRS. The DRS procedure consists of aortic translocation with the aortic valve and the coronary arteries from the right to the left ventricle and pulmonary root translocation with the pulmonary valve to the right ventricle.
Results: In groups A and B there was one death (20%) each. The remaining children had excellent clinical outcomes in the ICU and were discharged from hospital in good clinical condition and with good echocardiographic control. In Group C there were no deaths.
Conclusion: DTPPV can theoretically be considered a better option to correct TGA/CIV/PS because it anatomically corrects the RVOT and LVOT. We also believe that DRS anatomically corrects all the structures of the heart with TGA, including the aortic valve and aortic root that are positioned to the LV, and the pulmonary root and the pulmonary valve that are positioned to the RV.

1465: RESOURCE UTILISATION AND ONE-YEAR OUTCOMES IN INFANTS WITH SINGLE-VENTRICLE LESIONS

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Background: Infants with single-ventricle (SV) circulations utilise a significant proportion of hospital resources. These children account for the largest group of long-stay patients in ICU at our institution (20% of patients staying ≥ 28 days). We reviewed our resource utilisation and one-year outcomes of infants with SV physiology.
Methods: From 2005 to 2011, 130 infants with SV lesions were admitted to ICU; 121 received surgical palliation. Data were collected in this group.
Results: One-year survival after stage 1 palliation was 76% (95% CI: 67–83). Median (IQR) ICU length of stay was 11 (7.3–21.6) days; 24 (20%) patients stayed in ICU for ≥ 28 days [40 (34–56)]. Mortality was higher in those who stayed ≥ 28 days than those who did not: 12/24 (50%) vs 17/97 (17.5%) ($p = 0.001$). In the first year of life, SV infants averaged three ICU admissions, utilised 8.7% of ICU bed days, accounted for 16% of ICU long-stay admissions and had a stay of 55 (38–83) days in hospital; 38 (31%) received mechanical support. Risk factors for long stay were lower birth weight (OR 0.29/kg, 95% CI: 0.11–0.76), early re-operation after stage 1 (OR 3.7, 95% CI: 1.2–11.4) and ECMO (OR 6.1, 95% CI: 2–18.3). Risk factors for one-year mortality were smaller ascending aorta diameter (OR 0.65/mm, 95% CI: 0.42–1), associated cardiac lesion (OR 3.8, 95% CI: 1.1–13.2), and ECLS requirement (one run: OR 22.1, 95% CI: 6.0–81.4 and two or more runs: OR 59.4, 95% CI: 5–709).
Conclusion: Mortality associated with SV operations remains high and these children utilise considerable hospital resources. Requirement for mechanical support after stage 1 operations is associated with a poor outcome: no ECMO: 92% survival; one run of ECMO: 48% survival; two runs of ECMO: < 15% survival.

1480: LONG-TERM FUNCTION OF HOMOGRAFTS USED IN INFANTS FOR RECONSTRUCTION OF THE RIGHT VENTRICULAR OUTFLOW TRACT

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Aim: Assessment of the outcome of homografts used to reconstruct the right ventricular outflow tract (RVOT) in infants in a single unit.
Methods: Between September 1995 and April 2011, 48 consecutive patients under the age of one year were identified from the departmental surgical database. Mean age at the time of operation was 111 ± 109 days, 51% were female. Diagnoses included tetralogy of Fallot with absent or severely stenosed pulmonary valve ($n = 16$), truncus arteriosus ($n = 26$) and complex pathology ($n = 9$). These patients received 52 homografts (20 pulmonary, 31 aortic; seven of the 52 homografts were bicuspidised) for reconstruction of the RVOT. Homografts were procured and stored locally at the New Zealand Heart Valve Bank from cadavers or organ donors. They were treated in antibiotic solution for up to 62 hours (mean 49.2 ± 7.7) and stored for a mean of 7.2 days (± 4.8) at 4°C (39.2°F) before they were placed in liquid nitrogen for 0.5 to 62 months (mean 16.7 ± 17.1). The primary endpoint was homograft failure, defined as operative replacement for stenosis (RV pressure > 60 mmHg on echocardiogram) or severe pulmonary regurgitation with right ventricular dysfunction. Follow up was complete. No early mortality but three late, non-homograft-related deaths occurred in the study period. Kaplan-Meier analysis demonstrated a homograft failure rate of 12% at one year, 32% at three years, 40% at five years, 60% at eight years and 80% at 10 years. Significant branch pulmonary artery stenosis (flow acceleration > 2 m/s on echocardiogram) was associated with early homograft failure ($p = 0.04$). Patient age, weight and homograft type and size were not associated with early failure.
Conclusion: In New Zealand aortic or pulmonary homografts are the preferred conduit for RVOT reconstruction in infants and give excellent durability.

1485: OUTCOMES OF SINGLE-VENTRICLE PALLIATION IN PATIENTS WITH ARCH OBSTRUCTION

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Background: Outcomes of patients undergoing single-ventricle palliation and arch repair are unknown.
Methods: The follow up of 70 consecutive patients undergoing single-ventricle palliation and arch repair, excluding HLHS between 1983 and 2008 were reviewed. Dominant diagnoses were DILV (28), tricuspid atresia (16), DORV (six), AVSD (seven), mitral stenosis/atresia (five), TGA hypoplastic RV (three) and other (six). Arch anomalies were coarctation (48), interrupted arch (10) and arch hypoplasia (12). A strategy of performing a neonatal stage 1 Damus procedure with arch repair and shunt became the dominant approach

being performed in 1/70 (1%) in the initial years of 1983–1989, 9/70 (13%) in 1990–1999 and 23/70 (33%) in the recent 2000–2008 period.
Results: All patients underwent an initial procedure at a median of six days (4–12) consisting of PA banding (four), PA band and arch repair (31), Damus and shunt (33) and other (two). A total of 26 patients died before Fontan completion. Three survivors were denied Fontan completion, and two are still awaiting discussion. Thirteen of the 35 patients who had initial banding later required a Damus and only three required outflow tract obstruction relief. Thirty-nine patients underwent Fontan completion at a median of five years (2–6). There was no hospital mortality, and after a mean follow up of 5 ± 6 years there was one death and no further adverse outcomes. Overall survival at 10 years was 60% (95% CI: 45–70).
Conclusion: Patients born with single-ventricle physiology and arch obstruction have a high risk of mortality in the first two years of life. Their outcomes seem excellent once they reach Fontan status. It is likely that in patients with single-ventricle and arch obstruction, strategies to avoid systemic outflow tract obstruction should be implemented early in life.

1493: DOES LIMITED RIGHT VENTRICULOTOMY PREVENT RIGHT VENTRICULAR DILATATION AND DYSFUNCTION IN PATIENTS WHO UNDERGO TRANSANNULAR REPAIR OF TETRALOGY OF FALLOT? ANALYSES OF MAGNETIC RESONANCE IMAGING DATA IN 113 PATIENTS

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Background: In the hope of decreasing long-term complications of conventional transannular repair with a large right ventriculotomy (RV-tomy) in patients with tetralogy of Fallot (TOF), a transatrial and transpulmonary approach with limited (< 1 cm) transannular RV-tomy has been adopted by many centres. However, long-term benefits of this technique have not been demonstrated.
Methods: Between June 2002 and April 2012, 113 patients (mean age of 18.5 ± 6.6 years) with transannular repair of TOF underwent magnetic resonance imaging (MRI) for evaluation of pulmonary regurgitation (PR). Patients were divided into limited RV-tomy group ($n = 39$) and conventional RV-tomy group ($n = 74$). We compared the MRI parameters of the two groups to test the hypothesis that limited RV-tomy in the setting of transannular TOF repair would result in less right ventricular (RV) dilatation and dysfunction compared with conventional RV-tomy.
Results: The interval between TOF repair and MRI examination was shorter in the limited RV-tomy group (limited: 12.7 ± 3.8 years vs conventional: 17.2 ± 4.7 years, $p < 0.001$). There was no difference in PR fraction between groups (43 ± 10 vs 45 ± 9%, $p = 0.270$). Indexed RV volumes were similar between groups (RV end-diastolic volume index: 149 ± 31 vs 152 ± 42 ml/m^2, $p = 0.704$, RV end-systolic volume index: 70 ± 24 vs 77 ± 38 ml/m^2, $p = 0.313$). There was no difference in RV ejection fraction between groups (54 ± 9 vs 51 ± 9%, $p = 0.160$).
Conclusions: We could not demonstrate long-term benefits of limited RV-tomy compared with conventional RV-tomy in patients who underwent transannular TOF repair, at least in terms of RV size and function. Further studies are necessary to define the role of limited RV-tomy in patients who undergo transannular TOF repair.

1525: DEEP SEDATION VERSUS GENERAL ANAESTHESIA IN CHILDREN UNDERGOING PERCUTANEOUS CARDIAC INTERVENTION IN THE CATHETER LABORATORY

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Background: Paediatric patients require sedation/anaesthesia more often than adults during both diagnostic and therapeutic procedures. Moreover the paediatric population represents the highest risk, lowest error-tolerance subgroup. The safety and efficacy of deep sedation versus general anaesthesia in children undergoing percutaneous cardiac intervention in the catheterisation laboratory is rarely discussed in the literature.
Methods: All patients under 18 years of age who were referred for elective percutaneous cardiac intervention during a period of six months were included in the study. The patients were divided into two subgroups; group 1, who were subjected to deep sedation using ketamine infusion and group 2 who completed the procedure under general anaesthesia. Different numerical and categorical data were collected using a custom-made sheet, and anaesthesia-related complications were analysed and compared between the two subgroups.
Results: Anaesthesia-related complications occurred in 14.1% of the study group, all were self limiting and none was life threatening. The commonest complications were delayed recovery ($n = 2$) and post-procedural vomiting ($n = 2$). There was no significant difference between the two subgroups regarding age, body weight, height, body surface area, procedure duration, anaesthesia duration and recovery time. There was also no significant difference between the two subgroups as regards the occurrence of anaesthesia-related complications ($p = 0.551$)
Conclusion: Both deep sedation and GA were as safe and effective as each other in establishing adequate sedation/anaesthesia in children with congenital cardiac malformations undergoing percutaneous cardiac intervention in the catheterisation laboratory.

1526: OUTCOMES OF INTRA-ATRIAL BAFFLING FOR PARTIAL ANOMALOUS PULMONARY VENOUS CONNECTION

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Background: Intra-atrial baffling for partial anomalous pulmonary venous connection (PAPVC) has been performed with excellent outcomes. However, there is the risk of superior vena cava (SVC) or pulmonary venous obstruction, as well as sinus node dysfunction. We studied the long-term results for this procedure.
Methods: From January 1992 to July 2012, 18 patients underwent intra-atrial baffling for anomalous right-side pulmonary vein draining to SVC. The median age was 10 years (45 days to 66 years). Two patients had bilateral PAPVC and the left-side anomalous pulmonary vein drain was repaired simultaneously. Two patients had intact atrial septum and the remaining 16 patients had atrial septal defect. The PAPVC was baffled to the left atrium through ASD using single-patch in 11 patients (61.1%) and two-patch in seven patients (38.9%). The duration of mean follow up was 47.4 months (1.0 months – 15.8 years) and two patients were lost to follow up. Medical records and echocardiographic data were retrospectively reviewed.
Results: There was no early or late mortality. Transient sinus node dysfunction occurred in three (16.7%) patients, but all patients were in normal sinus rhythm at the last follow up. SVC stenosis and thrombosis was noted in one patient who required stent insertion. Right superior pulmonary venous stenosis and thrombosis, and subsequent pulmonary infarction developed in a patient. In the other patients, there was no evidence of pulmonary vein or SVC stenosis.
Conclusions: Intra-atrial baffling of right-side PAPVC draining SVC does not affect the sinus node function. However, there is a risk of pulmonary vein or SVC stenosis. Thereore other surgical options should be considered for patients requiring long tunnel for intra-atrial baffling.

1527: OUTCOMES FOR SIMPLE TRANSPOSITION IN THE CURRENT ERA

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Background: We report outcomes for simple transposition of the great arteries (TGA) in a contemporary cohort.
Methods: Ninety-six patients were diagnosed between 2004 and 2011. Five did not undergo operations because of associated severe congenital anomalies ($n = 1$, antenatal diagnosis), hypoxic encephalopathy ($n = 3$, one antenatal diagnosis) or intracranial bleed ($n = 1$). Ninety-one neonates underwent surgery with preservation of the neo-aortic sino-tubular junction where possible.
Results: Mean age at operation was 8.1 days. There was one operative mortality in a premature low-birth weight neonate in circulatory shock prior to the operation. There were two late mortalities. One child had complicated coronary anatomy (operation involved pericardial patch augmentation of the left main coronary artery) and died suddenly at home 52 days after surgery. The other had unexplained severe pulmonary hypertension and died 169 days after operation. Coronary artery pattern was Leiden 1LCx2R (63%) and two intramural coronary arteries were encountered. The sino-tubular junction was preserved in 57 (63%) patients. Follow up ranged from 6–96 months (median 39 months), 81% complete. Neo-aortic regurgitation was mild to moderate (three), mild (10), and trivial (16). Two of the children with mild to moderate neo-aortic regurgitation had a single coronary origin implanted using a trapdoor. Otherwise, there was no specific coronary pattern or transfer technique identified as a risk factor for the development of neo-aortic regurgitation. Six required re-intervention for relief of supra-valvar pulmonary stenosis or right pulmonary artery narrowing at a median of 18 months. There were no aortic root or coronary arterial re-interventions.
Conclusions: In this series 5% of patients with TGA did not reach operation, highlighting the need for planning of delivery to optimise outcomes. Mortality in the operated group at one year was 3%. Re-operations for pulmonary artery stenosis are the most important late re-intervention (7%). Clinically apparent coronary problems are uncommon at mid-term follow-up.

1528: PRELIMINARY STUDY OF INTRA-OPERATIVE CHANGES IN RESPIRATORY AND HAEMODYNAMIC PARAMETERS IN THE MODIFIED BLALOCK-TAUSSIG SURGERY

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Objectives: The modified Blalock-Taussig surgery has a high mortality rate, especially in neonates with low birth weight. The objective of this study was to compare changes in haemodynamic and respiratory parameters in patients with cyanotic congenital heart defects undergoing modified Blalock-Taussig surgery.
Methods: Five consecutive patients submitted for modified Blalock-Taussig surgery were sampled for three arterial blood gas analyses (ABG): one at the beginning of the surgery as a control, followed by two more ABG taken five minutes after clamping the right pulmonary artery and five minutes after removal of the clamp to obtain acid–base, ventilation and systemic perfusion parameters. We used the Student's *t*-test, Wilcoxon test or the Mann-Whitney U-test to compare the same variable in two different times. A p-value < 0.05 was considered statistically significant.
Results: The mean age was 29.2 ± 35.8 days (range 2–86) and the mean weight was 3.11 ± 0.58 kg (range 2.66–4.05). The mean arterial blood gas parameters five minutes after pulmonary artery clamping were: pH: 7.20 ± 0.12, pCO_2: 53.9 ± 20.0 mmHg, HCO_3: 16.5 ± 1.7 mEq/l, pO_2: 30.3 ± 10.6 mmHg, and SaO_2: 48.1 ± 23.1%. The haemodynamic variables were: mean arterial pressure: 30.2 ± 7.4 mmHg and arterial lactate: 3.2 ± 2.6 mmol/l. The mean pulmonary artery clamping time was 8.4 ± 0.5 minutes.
Conclusions: In these patients submitted for modified Blalock-Taussig surgery, we found the following statistically significant changes in haemodynamic and ventilator parameters during the clamping of the pulmonary artery: decreased blood concentration of bicarbonate ions, decreased arterial oxygen partial pressure and decreased mean arterial pressure.

1534: DO IT YOURSELF: DEFINING INSTITUTIONAL PREDICTORS OF MAJOR ADVERSE EVENTS EARLY AFTER PAEDIATRIC CARDIAC SURGERY

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Background: Few reported predictors of major adverse events (MAE: cardiac arrest, cardiopulmonary resuscitation, emergency extracorporeal life support, or death) early after paediatric cardiac surgery have stood the test of time and shown universal applicability. Each institution needs to identify its own predictors and define local thresholds for intervention.
Methods: Over a three-year period, a range of reported clinical and laboratory predictors was assessed in patients following bypass surgery on admission to our tertiary-care paediatric intensive care unit (PICU). Peri-operative data were retrieved from our institutional Aristotle database, and automated extraction of clinical and laboratory data from our clinical information system was applied. Generalised estimating equations (GEE) were used to determine variables and their thresholds predictive of MAE.
Results: We analysed 505 procedures in 483 patients, median age was 40.8 (0–217) months, weight was 9.8 (2.2–110) kg. MAE occurred in 21 (4.2%) patients within 48 hours of surgery. The odds ratio (OR) for presence of pre-operative risk factors as defined by the Aristotle Institute was 3.4 ($p = 0.02$) for MAE. For continuous variables, GEE calculates the OR for any increase from the mean of the population by a predefined scale, e.g. the mean ± SD heart rate on admission to PICU was 133.7 ± 30/min, and for any increased heart rate by 20/min the OR for MAE was 2.11 ($p < 0.001$). Further identified predictors were: lactate, bypass time, vasoactive-inotropic score, pO_2–FiO_2 ratio, end-tidal CO_2–pCO_2 difference, and systolic arterial pressure.
Conclusions: Although not the most powerful outcome model, for units with low paediatric cardiac surgical patient volume, GEE is a validated and pragmatic tool for this purpose. However, while dealing with missing data, GEE is unable to examine interactions and combinations of predictors. In summary, we identified our institutional predictors of MAE and defined clinically useful local thresholds for early intervention. Remodelling is warranted at defined intervals, allowing for changes in our practice over time.

1541: PREDICTIVE VALUE OF ARISTOTLE COMPLEXITY SCORE FOR RISKS OF CONGENITAL HEART SURGERY IN KOREA

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Background: The Aristotle complexity score was developed to predict the risks of congenital heart surgery and evaluate surgical performance. However, every country has different healthcare environments, so we evaluated the predictive value of the Aristotle complexity scoring system in our country based on our institutional outcomes.
Methods: Aristotle basic complexity score and comprehensive scores were prospectively assigned to all consecutive surgical procedures for congenital heart surgery between January 2008 and August 2012. We defined the major morbidity based on the Society of Thoracic Surgeons' national database. The discriminator power of the scoring system for mortality and major morbidity was analysed by the goodness-of-fit test using a logistic regression model.
Results: We performed 821 surgical procedures for congenital heart defects. Aristotle basic score was 6.8 ± 2.2 and comprehensive score was 8.2 ± 3.0. There were 31 surgical mortalities and major morbidities developed in 223 cases. The *p*-value of the goodness-of-fit test for surgical mortality and major morbidities were 0.742 and 0.488, respectively. The C-index was 0.782 (95% CI: 0.69–0.874), and 0.711 (95 CI: 0.671–0.751).
Conclusions: Aristotle complexity score is an adequate model to predict the surgical mortality and morbidity rates in our country. The system can be used for stratification of the risks of congenital heart defects.

1564: VENTRICULAR-ARTERIAL COUPLING IN CHILDREN WITH STILL'S MURMUR

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Background: Still's murmur is the most common innocent heart murmur in children, but there is no reliable explanation of its origin. Ventricular-arterial coupling (VAC) is an index describing the cross talk between the left ventricle (left ventricular contractility E_{LV}) and the arterial system (effective arterial elastance E_A). It serves as an important measurement of cardiac performance and cardiac energetics.
Methods: In our exploratory study, data of 43 children with Still's murmur and 42 healthy children without murmur aged two to 10 years (mean age with murmur: 5.17 ± 2.0 years; controls: 5.8 ± 2.5 years) were analysed regarding blood pressure (BP) and heart rate (HR), echocardiographic measurements of aortic root (AoD) and left ventricular end-diastolic (LVIDd) and end-systolic (LVIDs) dimensions indexed to body surface area, and VAC, thereby including the arterial system into the investigation of Still's murmur and its possible origin.
Results: While there was no significant difference regarding BP, HR, AoD, LVIDd and LVIDs, or their relative proportions, significant differences could be found in VAC ($p = 0.005$). There was a tendency towards higher E_A ($p = 0.39$) and lower E_{LV} ($p = 0.14$) in children with Still's murmur. Also, there was a significantly higher ejection fraction (EF%) ($p < 0.005$) and a larger stroke volume (SV per kg body weight) ($p < 0.05$) in children with murmurs. All differences were more pronounced in children aged two to six years than in those aged six to 10 years.
Conclusion: The low-frequency vibratory murmur seems to be caused by a combination of lower E_A, higher E_{LV},and higher ejection fraction and stroke volume compared to children without murmur. The fact that all differences were more pronounced in young children is reaffirmed by the decreasing prevalence of Still's murmur in older children.

1568: 30 YEARS OF PAEDIATRIC CARDIAC SURGERY IN SERBIA AND 10 YEARS OF THE ARTERIAL-SWITCH OPERATION AT THE MOTHER AND CHILD INSTITUTE OF SERBIA

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Introduction: The Department for Paediatric Cardiac Surgery was officialy founded in Belgrade in 1982. Individual attempts to operate on patients with congenital heart disease prior to 1982 were performed at adult cardiac surgical centres in Belgrade. The Senning and Mustard modifications of the atrial-switch technique were the techniques of choice for treating D-TGA. Until 2001, there were 13 attempts at arterial switching, with three survivors. From 1998 to 2001, the majority of D-TGA patients were routinely sent abroad for treatment. The arterial-switch procedure has been the standard operative procedure for patients with D-TGA in our hospital since May 2003.
Methods: In the period from May 2003 to June 2012, a total number of 65 children with D-TGA or D-TGA + VSD patients were operated on by the arterial-switch technique. Other diagnoses were excluded from the study. There were two surgeons performing the operation: the senior surgeon performed 57 operations and the the second one, only five patients from 2010. Routine pre-operative treatment was applied to all children. The youngest patient was seven days old, the oldest 95 days, the body weight varied from 2 300 to 4 900 g. The coronary artery patterns were identified as normal in 40 patients, inverted in four, single coronary artery anatomy was diagnosed in three patients and other forms in 15 patients.
Results: The overall mortality has fallen from 42% in the first two years to 3% in the last 24 months. The identified risk factors were bleeding and coagulation disorders, clinical conditions of the children and surgical errors.
Conclusion: In a 10-year period, the arterial-switch procedure has become a sucessful technique in our hospital.

1597: VACUUM-ASSISTED CLOSURE IN THE TREATMENT OF DEEP POST-STERNOTOMY WOUNDS IN NEWBORNS

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Introduction: Deep sternal infections after paediatric open-heart surgery are rare. Mortality for post-sternotomy mediastinitis continues to be high. Treatment includes antibiotics, frequent change of wound dressing, and surgical debridement. The vacuum-assisted closure (VAC) therapy is a new treatment option. Nevertheless, especially in newborns, experience in deep wound treatment is limited.
Methods: In the five-year period from 2007 to 2011, two newborns developed deep sternal infection after cardiac surgery and were treated with a vacuum-assisted closure system.
Results: After primary cardiac surgery, both babies received circulatory support (ECMO) and delayed sternal closure. The duration of the VAC therapy was 21 and eight days. After good granulation was obtained, the patients underwent conventional wound dressing. Plastic surgery with muscle flap or skin graft was not required.
Conclusions: VAC therapy is an effective alternative treatment in post-sternotomy mediastinitis in neonates, reducing infection and providing good wound healing.

1618: NECESSITY OF BIOMEDICAL ENGINEERING SUPPORT FOR HUMANITARIAN MEDICAL MISSIONS IN DEVELOPING COUNTRIES

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Background: Due to less complex technology for medical equipment

in the past, there was little emphasis placed on the importance of biomedical engineering support for humanitarian medical missions. However, as technology for medical equipment has progressed at a rapid pace, and the complexity of procedures has increased, it is evident the role of biomedical engineering has expanded. An education in this field is not enough. A discipline in computer engineering, anatomy and physiology, and knowledge of equipment that is procedure specific must be incorporated. In underdeveloped countries it is quite common for these disciplines to simply not exist.
Methods: Data were analysed from the ICHF's (International Children's Heart Foundation) database over the past five years.
Results: The analysis provides a common link between the various types of equipment needed across all developing countries where humanitarian medical programmes have been started. During this five-year period, a total of 677 pieces of medical equipment were sent to 23 locations in 16 different countries. Some specific types of medical equipment included patient monitors, anaesthesia machines, cardiopulmonary bypass machines, ventilators, defibrillators, electro-surgical units, syringe pumps, hypo/hyperthermia units, and cardiac echo ultrasound units. ICHF biomedical engineering staff/volunteers have made 49 trips to provide biomedical engineering support and emergency repairs.
Conclusions: ICHF has created a paradigm shift and raised the bar of expectation on the level of education and expertise for biomedical engineering support. This support is defined as installing, servicing, repairing and providing staff training on biomedical equipment. The end result has allowed ICHF medical programmes in developing countries to grow at an accelerated pace, ensuring better patient safety and improving surgical results.

1632: USE OF HOMOGRAFT TISSUE FOR PAEDIATRIC AND ADULT CARDIAC SURGICAL DISEASE ON INTERNATIONAL COMPASSIONATE MISSIONS

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Background: International visiting cardiology and cardiac surgery teams that carry out paediatric and acquired cardiac surgery face serious limitations in the scope of surgery possible and restricted valve choices. These are complicated by late clinical presentations, accelerated anatomical distortion from growth, and rheumatic valve disease. The lack of effective primary care in many locations may force surgeons to change from a conceptually good operative decision to a temporary and unsatisfactory short-term fix.
Methods: In an attempt to address some of these issues and increase the operative choices available, CardioStart International teams have carried two homograft valves (donated by Cryolife Inc, Ga, USA) on many two-week missions since 1996. In three countries receiving cardiothoracic surgical assistance, 18 patients between four and 56 years underwent conduit reconstructions to the aortic (five) or pulmonary (13) position; two patients underwent the Ross procedure. Six conduits were later implanted by the local surgical team based on the intense learning experience they had gained during the mission.
Results: Recent follow-up information is available in 15 patients (indirectly, in a further three). One adult patient (Ross) died at 15 days from a chest infection. All remaining patients are known to be alive, extending this small series follow-up experience to 16 years. Six patients have now reached adulthood and are in gainful employment. None currently need revision/re-operation of their primary operation. Ten patients are being tracked by a full international database evaluation already in operation (UMN and OHSU, USA).
Conclusions: Despite limitations of supply, homograft technology can logistically be provided to the diverse patients groups seen; and conduit implantation can readily be taught to local teams with excellent initial outcome. In two of these countries, local surgical teams have since developed local competence and limited availability, but proper follow up remains a priority in assisting new programmes to grow successfully.

1646: A TECHNIQUE TO CORRECT SEVERE STENOSIS OF LEFT CORONARY ARTERY AS A LATE COMPLICATION OF DIRECT RE-IMPLANTATION FOR TREATMENT OF ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

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A 11-year-old was admitted with unstable angina. At eight months old the baby underwent a correction of ALCAPA using the technique of re-implantation of the left coronary artery to the aorta with a button of the pulmonary artery. The early and medium-term outcomes of the operation were good. In the last six months the patient developed angina and deterioration of the ejection fraction. Cardiac catheterisation revealed severe stenosis of the coronary artery from the anastomosis with the ascending aorta to the bifurcation, and poor distal opacification of both anterior descending and circumflex arteries. At re-operation, the left coronary artery was stenotic with less than 1 mm diameter, and was solidly adhered to the posterior wall of the pulmonary artery. The technique used to correct the stenotic coronary artery was transection of the aorta distal to the origin of the left coronary artery. A longitudinal incision of 3–4 mm was made in the aortic wall, down to the anastomic orifice and the left coronary artery, all the way to the bifurcation. Using a segment of the left subclavian artery, opened longitudinally as a flap, the aortic wall, the new ostia and the left coronary artery were enlarged. Both the ascending aorta and the left subclavian artery were reconstructed, the first with end-to-end anastomosis, and the second with a polyester graft. The patient had an uneventful postoperative course. A cardiac catheterisation performed one month after the re-operation revealed an adequate flow through the left coronary artery and branches, and a better segmental contractility of the left ventricle. The enlargement of the stenotic coronary arteries with autologous arterial grafts is feasible and shows good results. With the technique of direct implantation of the left coronary artery to the aorta, excessive tension may occur with the compression of the bovine pericardial graft. These were possible causes of the coronary obstruction.

1652: STRICTLY POSTERIOR THORACOTOMY FOR MODIFIED BLALOCK-TAUSSIG SHUNT

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Background: In resource-poor settings, the modified Blalock-Taussig shunt (MBTS) is often performed for symptomatic relief in patients unable to afford the out-of-pocket expense for primary complete correction. Posterolateral thoracotomy and sternotomy are valid approaches for the creation of MBTS. However, in populations predisposed to keloids and hypertrophic scars, the aesthetic insult from either approach forms the basis of complaints from many

patients. The strictly posterior thoracotomy (SPOT) may yield a cosmetically superior result with a short posterior scar not evident to the patient.
Methods: From August 2011, we adopted the SPOT approach for creation of the MBTS, which is preferably performed on the right side. The patient is placed in a left lateral decubitus position with a slight counter-clockwise chest rotation. The incision extends 6–8 cm between the scapular spine and posterior axillary line, 2 cm parallel to the vertebral border of the scapula. Limited division of the trapezius and lattisimus dorsi may be required beneath the skin incision to facilitate exposure. The chest is entered through the bed of the fourth rib. We used this approach in 23 patients.
Results: There were 15 males and eight females with symptomatic Fallot's tetralogy. Ages ranged from 16 months to 17 years (median four years). Shunt sizes used ranged from 4–6 mm. Mean postoperative SPO_2 was 84%. No shunt failures were encountered. Hospital stay was 7–10 days. There was one re-operation for clotted haemothorax; this 17-year-old patient was the only mortality (4.3%) from sudden cardiac death on the seventh postoperative day. The cosmetic result has been very appealing.
Conclusion: The SPOT approach is a safe alternative for creation of the MBTS; it yields a cosmetically superior result to both posterolateral thoracotomy and sternotomy.

1662: DOUBLE-SWITCH PROCEDURE FOR C-TGA: IS IT WORTH DOING?

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Aim: To present our limited experience with the double-switch procedure in SRMC over a period of 15 months (April 2010 – June 2012).
Methods: This group includes five patients with a diagnosis of corrected TGA undergoing the double-switch procedure. The surgical options for C-TGA are anatomical and physiological repair. Anatomical repair includes the double-switch procedure in the form of either ASO + Senning or Rastelli + Senning. All our patients underwent Rastelli + Senning as they were not suitable candidates anatomically for ASO.
Results: Four patients performed well in the immediate postoperative period and were discharged from hospital. One patient died on the fourth postoperative day.
Conclusion: We believe that although the double-switch procedure is complex, the immediate postoperative results were satisfactory. Long-term follow up is needed in these patients. Hence we conclude that the double-switch procedure should be considered in patients with C-TGA.

1698: CONTEMPORARY SURGICAL APPROACH TO SCIMITAR SYNDROME USING DIRECT RIGHT PULMONARY VEIN-TO-LEFT ATRIAL ANASTOMOSIS VIA STERNOTOMY

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Background: There is no consensus on the optimal surgical technique for the repair of scimitar syndrome, an anomalous right pulmonary venous connection to the inferior vena cava. We report our experience with an anatomical approach involving direct anastomosis of the right pulmonary vein to the left atrium, performed under conventional cardiopulmonary bypass without the need for circulatory arrest through a sternotomy.
Methods: This was a retrospective review of nine patients operated from 2009 to 2011 for anomalous pulmonary venous connections from the right lung to the IVC. The age of patients at presentation varied between three months and 27 years (mean 16 months). Through a median sternotomy, the anomalous right pulmonary vein was mobilised and disconnected close to the diaphragm and the caval end was secured. The pulmonary end of the vein was brought superiorly through a generous pericardial opening behind the right phrenic nerve without tension, and via a natural or surgically created atrial septal defect, a wide opening was created on the right side of the posterior left atrium, wherein the scimitar vein was directly anastomosed.
Results: Eight out of nine patients were primary venous repair and one patient had a previous failed repair using an intra-atrial baffle. There were no operative deaths. The median cardiopulmonary bypass time was 109 min and median cross-clamp time was 47 min. The median ventilator time was 16.8 hours and the median stay at the intensive care unit was 2.4 days. There were no re-explorations in the postoperative period. There were no instances of pulmonary venous obstruction. There were no late deaths within follow up of one year.
Conclusions: Anatomical repair of the scimitar vein based on re-implantation onto the left atrium via sternotomy results in safe and reliable repair in patients with a wide age spectrum, including re-operation.

1701: HAEMODYNAMIC CHARACTERISTICS OF PORCINE DECELLURISED HENOGRAFTS FOR AORTIC VALVE REPLACEMENT IN CHILDREN

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Objectives: Aortic valve repair and the Ross procedure are not always applicable in aortic valve disease in children. We wanted to demonstrate an alternative in a preliminary study of the performance of decellularised porcine xenografts in children implanted into the systemic circulation.
Methods: Between January 2010 and March 2012, a total of nine aortic valve replacements with Matrix decellurised porcine xenografts was performed. Operative results and postoperative haemodynamics were evaluated.
Results: The mean patient age was 6.4 years (range 3–15 years). The mean weight was 15.2 kg (range 11–45 kg). The underling diagnoses were isolated aortic valve disease in six, common arterial trunk in two and single-ventricle type CHD with obstructed systemic outflow tract in one patient. Among the children, three underwent additional procedures: lateral tunnel Fontan operation, Damus-Kaye-Stansel procedure and pulmonary homograft replacement. All children with isolated aortic valve disease underwent previous balloon valvuloplasty or surgical valvulotomy. The diameter of aortic grafts was 1 mm in two, 14 mm in two, 17 mm in three, and 19 mm in two children. Postoperative echocardiography demonstrated mean transvalvular gradient 19 mmHg (range 10–27 mmHg) that did not tend to increase over time. No significant valve insufficiency was observed. Follow-up time varied between three and 24 months.
Conclusion: Our preliminary results suggest that porcine decellurised tissue-engineered xenografts may be the alternative option for children who require aortic valve implantation or replacement.

1709: MANAGEMENT OF POST-OPERATIVE CHYLOTHORAX IN PAEDIATRIC PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Chylothorax is a well-known postoperative complication affecting patients with congenital heart disease (CHD). No clear consensus has been reached regarding the management strategy to date.
Methods: Data were collected retrospectively regarding the diag-

nosis and management of chylothorax. The cardiac database and dietetic records were used to identify the paediatric population with CHD that underwent cardiac surgery at the Yorkshire Heart Centre.
Results: Between July 2005 and June 2012, 2 290 operations were performed in paediatric patients with CHD. Chylothorax was diagnosed in 117 of them, an incidence of 5.1% (95% CI: 4.2–6%), of which 69 (59%) were under one year of age. The average time to diagnosis was six days postoperatively. All patients were commenced on medium-chain triglyceride diets (MCT) and 33 (28.2%) were treated with octreotide, when drainage was refractory. Surgical intervention was performed in seven patients. Chest drains remained for an average of 11 days, nine (range 3–22) for those on only MCT and 15 days (range 6–32) for those on octreotide. Fourteen patients died. Among the survivors, 33 (32%) were on a six-week MCT diet (our policy until the beginning of 2008), one was treated for five weeks, 59 (57.3%) had a four-week diet, and the rest (9.7%) had less than four weeks (range 2 days to 3 weeks) mainly for nutritional reasons. There were two recurrences (1.9%, six-week regime).
Conclusions: Chylothorax is a rare complication following paediatric cardiac surgery. The MCT diet remains the gold standard in the management of chylothorax. A maximum of four weeks has proven to be sufficient for our patients. A shorter duration of the MCT diet could be considered. Octreotide can be reserved for the more persistent cases. Surgical intervention is required in a limited number of patients.

1710: TRANSITION OF SURGICAL REPAIR FROM 'CONVENTIONAL' TO 'PRIMARY SUTURELESS' TECHNIQUE FOR TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

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Background: The 'sutureless' repair has been described for post-repair pulmonary vein stenosis. It has since been reported as a modified technique for the primary repair of infracardiac total anomalous pulmonary venous connection and recently for the primary repair of other types as well.
Methods: One hundred and two consecutive patients (median age 60 days; median weight 3.5 kg) underwent conventional or sutureless total anomalous connection repair between July 2009 and July 2012.
Results: Types of total anomalous pulmonary venous connection included supracardiac in 54 patients (53%), cardiac in 24 (23%), infracardiac in 17 (16%), and mixed type in seven patients (6%). Median follow-up time was six months. Vertical vein obstruction was found pre-operatively in 43 patients (40%). A primary sutureless repair was carried out in 19 patients (18.6%; supracardiac, $n = 4$; cardiac, $n = 1$; and infracardiac, $n = 14$). A proportionately greater number of patients with high-risk infracardiac total anomalous pulmnary venous connection underwent the sutureless technique (73 vs 3.6%). There were seven early operative deaths, of which four were from non-cardiac causes. Early outcomes for death and re-operation for pulmonary venous stenosis were not significantly different between these two disparate groups.
Conclusion: The sutureless group had more infracardiac total anomalous pulmonary venous connection and a higher rate of decline in the postoperative right ventricular systolic pressure with a shorter ICU stay and earlier hospital discharge. Early results of sutureless repair has led to a change in our practice from conventional to primary sutureless repair for all cases of infracardiac total anomalous pulmonary venous connection and for selected cases of supracardiac, cardiac and mixed types with pre-operative pulmonary vein stenosis or confluence obstruction.

1712: DOBUTAMINE DOES NOT OFFER SUSTAINED CONTRACTILITY IN A PIGLET MODEL OF RIGHT VENTRICULAR FAILURE

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Background: The immature myocardium has significantly different receptor kinetics, metabolism and enzyme activity. Previous haemodynamic studies have primarily focused on the inotropic effect on left ventricular function. A piglet study was planned to investigate the effect of three different inotropic strategies on right ventricular failure.
Methods: Twenty-one piglets aged four days old were prepared to measure right ventricular pressures. Stunning of the right ventricle (RV) was induced by 10 cycles of ischaemia–reperfusion injury. We randomised the animals to placebo or one of three inotropic protocols: AM (adrenaline): 0.09 $\mu g.kg^{-1}.min^{-1}$ and milrinone: 50 $\mu g.kg^{-1}$ bolus and 0.4 $\mu g.kg^{-1}.min^{-1}$; DM (dopamine): 6 $\mu g.kg^{-1}.min^{-1}$, milrinone: 50 $\mu g.kg^{-1}$ bolus and 0.4 $\mu g.kg^{-1}.min^{-1}$; and Dob (dobutamine): (8 $\mu g.kg^{-1}.min^{-1}$) for 240 minutes. We used maximum pressure development over time (dP/dt_{max}) as a marker of contractility, and minimum pressure development over time (dP/dt_{min}) to evaluate diastolic function. Arterial elastance and end-systolic pressure–volume relationship were calculated to evaluate arterio-ventricular coupling (Ea/ESPVR). One-way ANOVA was used to analyse differences between area under the curve (AUC) and mean values from specified time points.
Results: Maximum response was fastest in the Dob group: 30 min, followed by DM: 40 min, and AM: 80 min. After 180 min, dP/dT_{max} decreased to a level indistinguishable from baseline in the Dob group (non significant), whereas dP/dt_{max} in the AM and DM groups continued to increase throughout the study period ($p < 0.01$ and $p = 0.03$, respectively). AUC did not differ between groups ($p = 0.48$). dP/dt_{mi}n improved in all groups, and remained stable in all groups. Only DM improved Ea/ESPVR compared to controls ($p < 0.05$).
Conclusions: In the stunned RV, Dob effectively increased contractility, however the effect was not sustained over the infusion period in comparison with the AM and DM groups. Only DM improved the arterio-ventricular coupling after right ventricular failure. The three inotropic interventions improved diastolic function to a similar degree.

1715: TRANSITIONING PAEDIATRIC HEART SURGERY IN MONGOLIA, FROM HYPOTHERMIC SURGERY TO CARDIOPULMONARY BYPASS: LESSONS LEARNED

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Introduction: Mongolian paediatric cardiac and cardiac surgery care faced a growing need to develop a new cardiac programme after its 1990 independence from the USSR. This was due in part to budgeting constraints and a lack of advanced cardiac surgery care and infrastructure. Therefore, hypothermic cardiac surgery without cardiopulmonary bypass was instituted as the standard approach to congenital defect repair.
Methods: In 2005 a visiting paediatric team composed of a cardiothoracic surgeon, cardiologists, cardiac anaesthesiologists, a perfusionist, cardiac intensivists, and cardiac intensive care nurses started systematically training the Mongolian cardiac team in all aspects of paediatric cardiac surgery using cardiopulmonary bypass.
Results: From 2005 to 2011 the visiting cardiac team visited eight times and performed 30 closed- and 50 open-heart surgeries with the Mongolian team. During that time period the Mongolian team successfully transitioned from hypothermic surgery to cardiopulmonary bypass surgery. In 2011 the total number of bypass cases was 176, with a 30-day operative mortality rate of less than 5%.
Conclusions: Our institution has safely and cost-effectively transitioned to the surgical repair of congenital cardiac defects using cardiopulmonary bypass. We will discuss the procedures and lessons learned from this transition to more advanced cardiac surgery techniques which can be used as a model for other developing countries such as Mongolia.

1728: PAEDIATRIC CARDIAC TUMOURS: A 43-YEAR REVIEW

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Background: Cardiac tumours in children are rare. Of the 800 cases reported in the literature, 97% were benign. The most common paediatric cardiac tumour is rhabdomyoma; most are asymptomatic, regress over time, and often are only observed. When benign tumours become obstructive and produce symptoms, they should be resected. We retrospectively reviewed our experience over the last 40 years to evaluate presentation, management and outcomes in this patient population.
Methods: Since 1969, 67 children were diagnosed with paediatric cardiac tumours. Thirty-three (49%) were surgically managed and further analysed. Twelve (39%) presented with symptoms of CHF. Mean age at the time of diagnosis and surgery was 8.9 years (range: 1 day – 27 years).
Results: Rhabdomyoma was the most common tumour. Although there were only eight (24%) malignant cardiac tumours, five of the six deaths in our series were from malignant tumours. Two malignancies, however, a teratocarcinoma and a rhabdomyosarcoma, were successfully excised and treated with chemotherapy. These two patients have survived tumour-free for 15 and 25 years, respectively. At a mean follow up of 4.6 (range: 0–26) years, 21 (91%) survivors of all tumours remained free of lasting postoperative cardiac symptoms. Four (12%) had minimal follow up.
Conclusions: Cardiac tumours are rare in children. Surgical excision or de-bulking should be carried out for rhabdomyomas and fibromas causing significant obstruction. Myxomas and teratomas require complete excision because they can embolise or recur locally. Malignant cardiac tumours should undergo radical excision, if possible, followed by adjuvant chemoradiation therapy. An aggressive surgical approach can yield long-term survival in some patients.

1738: ULTRASONOGRAPHY FOR VOCAL CORD MOBILITY AFTER PAEDIATRIC CARDIAC SURGERY

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Objectives: Upper airway obstructions after paediatric cardiac surgery are not uncommon and have many reasons. Vocal cord paresis or paralysis is not an unusual cause of airway obstruction or failure of extubation after cardiac surgery. In this study we aimed to evaluate the feasibility and accuracy of ultrasonography (US) assessment of vocal cord mobility and compare it to fibre-optic laryngoscopy (FL).
Methods: A prospective pilot study was conducted in the paediatric cardiac ICU (PCICU) from 1 June 2009 to 31 July 2010. Patients who had cardiac surgery and manifested with significant signs of upper airway obstruction were subjected to US screening of their vocal cord mobility, followed by FL assessment. All operators were blinded to each other's reports. Results of invasive (FL) and non-invasive (US) investigations were compared.
Results: Ten patients developed persistent significant upper airway obstruction after cardiac surgery and were included in the study. Their mean weight and age were (4.6 ± 2.5 kg) (2.7–9.4) and (126.4 ± 162.4 days) (8–480), respectively. All patients were referred to bedside US screening for vocal cord mobility. The results of US were subsequently compared with FL findings. Results were identical in nine (90%) patients and partially different in one (10%). Six patients showed abnormal glottal movement while the other four patients demonstrated normal vocal cord mobility by FL. Sensitivity of US was 100% and specificity was 75%.
Conclusion: US assessment of vocal cords is a simple, non-invasive and reliable tool to assess vocal cord mobility in the critical care setting. This screening tool requires skills that can be easily obtained.

1742: DEFINING NEURODEVELOPMENTAL OUTCOMES FOLLOWING MECHANICAL CIRCULATORY SUPPORT USING STANDARDISED CARE ASSESSMENT AND MANAGEMENT PLAN (SCAMP) METHODOLOGY

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Background: ECMO support for cardiorespiratory failure increases survival but neurodevelopmental outcomes remain ill-defined. Currently no national/international-level consensus on neurological follow up exists.
Aims: To (1) establish a collaborative standardised clinical assessment and management pathway (SCAMP) for neurodevelopmental outcome between ECMO centres in England, (2) identify surveillance, screening and early interventions to improve the level of functional neurodevelopment, quality of life and family satisfaction in children post mechanical support.
Methods: Published literature, international recommendations and local guidelines were reviewed. Expertise from neurology, neuroradiology and neuropsychology was sought. Consensus was evolved through a series of local and national specialist group meetings. A background position paper outlining prevalence, uncertainties in follow up, and appraisal of available neuropsychological assessment was developed. Evidence-based management algorithms were agreed on, incorporating plausible hypotheses pertaining to 'knowledge gaps'. A framework for cyclical analysis allowing variability in outcomes to emerge alongside refinements in care and resource utilisation was defined.
Results: The incidence of neurodevelopmental sequelae is 20–35% in neonatal populations and up to 50% in children supported post-cardiac surgery, with limited literature on paediatric respiratory ECMO patients. Risk factors for neurological injury have been identified, however few are modifiable. Neuro-imaging can help categorise risk but cannot predict degree of neurodisability. An algorithm was developed after consensus meetings between ECMO centres in England standardising baseline assessment, follow up, neuro-imaging and sequential, age-appropriate, neuropsychological testing. Multiple plausible outcomes for future evaluations based on prospectively collected data were identified.
Conclusion: There is a clear need for uniformity in early identification, risk evaluation and structured follow up of these children. Timely, relevant and nationally resourced tests may minimise neurological morbidity and maximise potential by the introduction of early interventions. Using SCAMP methodology, it is possible to seek national consensus, formulate evidence-based practice and achieve a national framework to provide the longitudinal screening of neurological deficits.

1754: CRITICAL ULTRASOUND, THE NEW ESSENTIAL SKILL IN PAEDIATRIC CARDIAC ICU (PCICU)

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With the recent introduction of high-quality, reasonably priced and completely portable neonatal ultrasound machines, a new paradigm shift has also emerged in the philosophy of ultrasound imaging in PCICU, which is the philosophy of 'critical ultrasound'. This is a shift from being an organ-based, systematic, comprehensive examination done by radiologists to a new concept of problem-based, goal-directed, focused multi-organ, time-dependant examination done by the treating neonatologist. The new paradigm is not trying to describe an organ pathology but rather involves 'focused, simple yes/no' examinations for the airway, lung, heart, abdomen or limbs that are directed to answer specific clinical problems (hypoxia, hypotension,

etc) and are performed not only for diagnosis but also for monitoring management.

The presentation will give the results of four years of experience in our centre with regard to this new essential skill in PCICU in obtaining hour-by-hour information to help the management of critically ill paediatric cardiac patients. In addition, it shows the multiple procedures that were guided by ultrasound, such as vascular access, lumbar puncture or pleural tap. It will also shed light on its important application in the telemedicine field.

1765: IDENTIFYING THE RISK FACTORS FOR CHYLOTHORAX FOLLOWING CARDIAC SURGERY IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Chylothorax is a rare but well-recognised complication following cardiac surgery in paediatric patients with congenital heart disease (CHD).
Methods: We retrospectively studied the paediatric patients with CHD who developed chylothorax following cardiothoracic surgery at the Yorkshire Heart Centre. Data regarding age, diagnosis, type of surgery, management and outcome were collected from the cardiac database and the dietetic records of the department for a seven-year period (July 2005 to June 2012).
Results: A total of 2 290 cardiothoracic operations were performed in our centre in paediatric patients with CHD during the above period. Chylothorax complicated 117 of these procedures, incidence of 5.1% (95% CI: 4.2–6%). The age ranged from one day to 14 years: 59% were under the age of one year, 31.6% between one and five years, and the rest were older. The incidence of chylothorax was higher following repair of tetralogy of Fallot (n = 22, 18.8%), atrioventricular septal defect repair (n = 16, 13.7%), Glenn (n = 14, 12%), ventricular septal defect closure (n = 11, 9.4%), arch repair (n = 11, 9.4%), arterial switch (n = 8, 6.8%), Fontan (n = 6, 5.1%), totally anomalous pulmonary venous drainage (n = 5, 4.3%). Previous cardiac surgery was performed in 40 (34.2%) patients. Median sternotomy was the approach in 103 (88%) patients, whereas a left thoracotomy was performed in 14 (12%). There was no difference in the percentage of patients requiring further treatment with octreotide (n = 29, 28.2% for the median sternotomy group, n = 4, 28.6% for the left thoracotomy group). Death occurred in 14 patients, 12 of whom were under the age of one year.
Conclusions: Younger age, median sternotomy and certain types of operation (TOF, AVSD repair, Glenn) seem to constitute risk factors for the development of chylothorax. Increased mortality rate was observed in infants. The type of sternotomy did not correlate with the need for the use of octreotide.

1776: ULTRASOUND-GUIDED VASCULAR ACCESS IN CRITICALLY ILL CARDIAC CHILDREN

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Vascular access in the paediatric age group is a challenging procedure, especially in compromised children and those who require multiple vascular cannulation. Critically ill paediatric cardiac patients particularly are in frequent need of vascular access. We report on our experience of vascular access under ultrasound guidance in children with congenital heart disease and we discuss the technique of line insertion and its difficulties in small infants.
Methods: In the paediatric cardiac intensive care unit we prospectively enrolled all the trials of vascular access (central or arterial lines) guided by ultrasound from January to September 2010. Age, weight, the time from first needle puncture to wire insertion, site of insertion, number of attempts, type of the line and complications were documented.
Results: Seventy-seven vascular access trials were performed in 43 patients. They included 15 arterial and 62 venous cannulations (32 femoral veins and 30 internal jugular veins). Mean age and weight of patients were 15 months (6 days – 11 years, median 2.5 months) and 7.2 kg (2 – 46 kg, median 3.8), respectively. Success rates were 93 and 95% for arterial and venous cannulation, respectively. Mean time from first needle puncture to wire insertion was 3.9 min (0.5–15 min, median 2 min). Fifty-five central-line cannulations (75%) were successful from the first puncture, 17 (23%) were successful from the second puncture and one case (2%) required three punctures. Lower body weight did not affect success rate, as 30 patients (45%) were less than 3.5 kg with 96.6% successful cannulation rates. There were no associated complications.
Conclusion: Ultrasound-guided vascular cannulation in critically ill paediatric patients is very useful. It is associated with high success rates and minimal complications.

1802: COMBINED ECMO AND CRRT IN CHILDREN

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Background: Venous–arterial/venous–venous extracorporeal circulatory membrane oxygenation (VA/VV-ECMO) is needed in cases of severe respiratory and/or circulatory failure. Reported mortality rates for ECMO-treated paediatric patients are high and even higher when combined with renal failure (> 80%). These children are often grossly fluid overloaded and oliguric. Successful CRRT in infants depends on good vascular access, which can be impossible to achieve. Therefore we have incorporated connections for CRRT apparatus as a standard in our paediatric ECMO circle.
Methods: Seven PICU patients were enrolled [median age 10 days (3–665); median weight 3.4 kg (2.9–10)]. Six of the patients were on VA-ECMO after surgery for congenital heart disease and one oncological patient with *Aspergillum pneumonia* was on VV-ECMO. We used the Prismaflex device (Gambro, Lund, Sweden). Dialysis filters were HF-20 filters, except for one patient where M60 was used (Gambro Industries, Lyon, France). The dialysis filters were primed with blood, apart from the M60 filter, which was primed with fresh frozen plasma. Blood flow targeting 7.5–10 ml/kg/min; modi CVVHDF/CVVHF using dialysis/effluent fluid doses targeting 2 000 ml/1.72 m^2/h. All patients received heparin infusion aiming at APTT range 60–80 s. Both the inlet and outlet connections for the blood flow to the Prisma apparatus were placed after the ECMO pump head and before the oxygenator to minimise the risk of air embolism.
Results: CRRT treatment time was a median of 89 hours; range 0.8–328 hours. A precise filter lifetime was not recorded but no filter was reported to have stopped due to clotting. The CRRT treatment was without impact on the haemodynamic parameters. We obtained intended negative fluid balance, and were able to administer TPN and reduce azotemia levels. Unfortunately all seven seven patients died in hospital.
Conclusion: In infants, CRRT using Prismaflex with HF20 and M60 filters inserted into the ECMO circle is technically feasible. Incorporation of preformed connections for CRRT in ECMO circles had no impact on ECMO treatment and provided steady blood flow for the CCRT.

1807: VALIDATION STUDY OF A CLINICAL HEART FAILURE SEVERITY SCORING SCALE IN CHILDREN, USING BRAIN NATRIURETIC PEPTIDE AND ECHOCARDIOGRAPHIC PROFILES

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Background: The New York Heart Association (NYHA) heart failure severity grading scale has stood the test of time in serving as a useful clinical grading scale for the severity of heart failure, particularly in adults but has its limitations as a useful scale in children. The Ross grading scale on the other hand has its usefulness in infants for whom it was designed but it has no use in older children. The Ibadan heart failure grading scale (IHFGS), applicable in children (aged 1 day – 12 years) derived from bedside-elicitable parameters without the use of sophisticated tools has been designed. The severity grading scale needs to undergo validation studies involving the use of known 'gold standards'.
Methods: A clinical team using the IHFGS, an echocardiographic team using 2D and M-mode derived standard parameters, and a clinical chemistry team measuring plasma BNP levels evaluated 100 consecutively recruited children aged one to 120 months with a clinical diagnosis of congestive heart failure from a variety of causes, along with 100 age- and gender-matched apparently healthy controls. Each team, blinded to the findings of other teams and separate from the primary care team, evaluated each child within half an hour of the others at presentation. Findings were compared between study subjects and those of the controls. Specificity and sensitivity of the IHFGS were determined. Correlation coefficients were determined between the IHFGS scores and the echocardiographic parameters and plasma BNP levels, correcting for age and body surface area and other confounding variables.
Results: Causes of heart failure included acute respiratory infections (35%), severe anaemia (30%), congenital heart disease (25%) and dilated cardiomyopathy (10%). High coefficient of correlation (+0.85) between IHFGS and plasma BNP levels and echocardiographic parameters were recorded. High specificity and sensitivity were recorded in respect of the IHFGS definition of no HF, and the three grades of HF.
Conclusion: The IHFGS is proposed for use in grading severity of heart failure in children, especially where sophisticated facilities are lacking.

1837: CO-ORDINATING CARE FOR LONG-TERM POSTOPERATIVE PICU CARDIAC PATIENTS

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Introduction: Cardiac patients in the PICU at the Royal Children's Hospital (RCH), Melbourne, account for approximately 50% of bed occupancy. Of these patients, more than 50% have a length of stay (LOS) longer than seven days. The PICU care manager's role is a nursing position introduced in 2008 to provide co-ordination and consistency in communication and care for all long-term and complex PICU patients. The following case study of a complex postoperative cardiac patient provides an example of how the role supports the complex care needs of a patient during the PICU admission.
Methods: A 13-year-old with a past history of tetralogy of Fallot (TOF) and Alagille syndrome presented to the PICU post right and left pulmonary artery plasty and redo of RV-to-PA conduit, during which a left main bronchial injury was sustained intra-operatively. Surgical complications and projected prolonged ICU stay led to inclusion as a care managed patient. Support to enable consistent and co-ordinated care is tailored to specific patient needs according to age of the patient, surgical pathway, illness acuity, social situation and postoperative complications. A close working relationship with all members of the multidisciplinary team, including medical, nursing and allied health, along with pre-established professional relationships and processes, enable the PICU care manager to be a link between patient and family and the hospital team.
Results: Consistent communication was maintained with weekly minuted multidisciplinary team meetings; detailed daily and weekly bedside care plans, and daily booked interpreter sessions (as English was a second language). Complex care needs were met with regular bedside support, including care plans for allied health sessions, ventilation weaning, analgesia and sedation weaning, mental health management and inotrope rotation management. Bedsides education, designated nursing care teams and the provision of clinical support were all instigated for ongoing nursing consistency of care.
Conclusion: Care co-ordination instigated and supported by a singular service for complex and long-term PICU patients at RCH provides a consistent and focal service for holistic and thorough patient management. The role enables a singular service provider overview of the patient to improve patient-centred care and to provide the ongoing perspective for all teams (which are often varied due to shift structure) to utilise.

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CATHETER INTERVENTIONS FROM FOETUS TO ADULT

1054: RHYTHM AND CONDUCTION ABNORMALITIES AFTER TRANSCATHETER CLOSURE OF VSDS: A SINGLE-CENTRE EXPERIENCE

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Background: We describe our experience focusing on the acute and mid-term disturbances of conduction and arrhythmias after transcatheter ventricular septal defect (VSD) closure and review their therapy and follow-up.
Material and methods: The medical records of consecutive 45 patients undergoing transcatheter closure of VSD were retrospectively assessed from August 2007 to July 2012. All electrocardiogram (ECG) and 24-hour ECG-Holter monitoring records were analysed for disturbances of conduction and arrhythmias.
Results: Patients' ages at implantation range between 15 months and 58 years (mean 10.8 years, median 8 years). Mean body weight was 30.2 kg (range 8.7–78 kg, median 24.5 kg). The diameters of VSDs were between 3.5 and 8.5 mm measured by echocardiography (mean 5.6 mm). All defects were occluded with Amplatzer devices. Amplatzer perimembranous VSD occluders were used for 14 patients (mean size of device 6.4 mm, range 4–9 mm), muscular VSD occluder for 26 patients (mean size of device 6.9 mm, range 4–10 mm) and duct occluder for 5 patients (3 of them 6/4, and 2 of them 8/6). Mean device diameter to defect diameter ratio was 1.2 + 0.13 (range 1–1.4). The mean duration of follow-up was 25.7 + 14.2 months (range 1–49 months). Two of the 29 patients with perimembranous VSDs developed cAVB within 2 and 6 days of the closure. Insignificant rhythm disturbances developed in 26.7% (12/45) of the patients after transcatheter closure of VSDs.
Conclusion: With early and late occurrence of cAVB, careful monitoring of rhythm and atrioventricular conduction is essential during the follow-up period. However, Holter monitoring should be performed before the procedure to understand the insignificant rhythm disturbances which were related to the procedure.

23: DEVICE CLOSURE OF LARGE TUBULAR PATENT DUCTUS ARTERIOSUS WITH SEVERE PULMONARY HYPERTENSION: ANALYSIS OF FORTY-SEVEN CASES

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Objective: Large patent ductus arteriosus (PDA) can cause severe pulmonary hypertension (PHT) as a result of increased blood flow or increased pulmonary vascular resistance (PVR). Surgical ligations of these large tortuous hypertensive ducts are very difficult. Device closure is an alternative option and here we report 47 such cases of device closure in our centre.
Patients and methods: A total of 47 patients (29 male) had large tubular PDA with severe PHT from January 2009 to December 2010. A retrospective review of data was done from the records kept in the department.
Results: The median age of the patients was 4.5 years (2 months to 45 years); median weight was 9.7 kg (3.5–60 kg). Narrowest PDA diameter varied from 3.4 to 14 mm (median 4.8 mm). Pulmonary to systemic blood flow ranged from 1.1:1 to 6.7:1. PVR ranged from 4.2 to 8.1 Wood units. Pulmonary arterial pressure ranged from 60/45 (50) to 120/84 (97) mmHg. Systemic blood pressure ranged from 65/40 (48) to 120/88 (99) mmHg. With 100% oxygen, pulmonary arterial pressure was reduced in 26 cases and remained the same in 9. In 12 cases the device was implanted without this test. Minimum PDA device size was 5/4 mm ADO 11 and maximum device size was 18 mm Amplatzer septal occluder. Forty-three patients had complete occlusion and 4 had residual shunt. Three patients had complete occlusion within the first month. One patient developed haemolysis and was managed with coil implantation. Embolisation was experienced in one case. On echocardiography, right ventricle pressure was reduced to normal by 3 months in all cases except two.
Conclusion: Tubular PDA associated with severe PHT could be successfully closed by ductal occluders. After occlusion, pulmonary pressure had definite reduction and results in our centre are encouraging.

39: IMPACT OF FENESTRATION CREATION ON MANAGING PATIENTS WITH PROTEIN-LOSING ENTEROPATHY COMPLICATING FONTAN PROCEDURE

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Introduction: Protein-losing enteropathy (PLE) is a well-known complication following the Fontan procedure and one way of managing such complication is to create fenestration if it is not present, or enlarging it if it is small, to reduce the Fontan pressure and reduce venous congestion which results in intestinal protein loss.
Aim: To evaluate the effect of such a procedure in our population.
Method: From February 2006 through October 2011, 9 patients who underwent fenestration creation due to development of PLE were assessed with regard to clinical, laboratory result and haemodynamic effect prior to and post procedure.
Result: Median age was 7 years (range 4–21), median weight 23 kg (15–52), and male: female ratio was 3.5:1 (7 male and 2 female). Median saturation pre and post procedure was 93% and 82% respectively, median albumin pre and post procedure was 18 g/dl and 31 g/dl during the first 2 wks and 36 g/dl thereafter, and median pulmonary artery pressure before and after was 25 mmHg (17–32 mmHg) and 16 mmHg (14–19 mmHg). Transpulmonary pressure gradient reduced from a median of 11 mmHg to 5 mmHg. There were no immediate deaths; 2 patients needed redilatation, and there were 2 (22%) late deaths (1 had stent thrombosis 1 month after the procedure, followed by fulminant pulmonary embolism, though all patients were on anticoagulant; 1 with sudden arrest could not be resuscitated at the emergency unit). Three (33%) patients have persistent low albumin though the fenestration is patent.
Conclusion: Transcatheter fenestration creation as a management of PLE following Fontan procedure is a feasible procedure, can be done in the catheter laboratory with little morbidity and mortality and with beneficial effect; however, late complications and complete resolving of PLE is of concern, especially if procedure is done late.

40: RIGHT VENTRICULAR OUTFLOW TRACT STENTING IN THE SYMPTOMATIC INFANT WITH TETRALOGY OF FALLOT

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Background: Debate continues regarding the initial management of cyanotic or duct-dependent infants with tetralogy of Fallot (TOF) especially those patients with pulmonary artery hypoplasia. While repair can and has been performed in these patients, it is associated with increased morbidity.
Objective: We review the effectiveness of right ventricular outflow tract (RVOT) stenting in the symptomatic young infant with TOF.
Methods: Clinical, echocardiographic, angiographic and haemodynamic data were reviewed for 13 patients who underwent 17 RVOT stenting procedures from March 2008 to Januay 2012.
Results: There were 8 girls and 5 boys; median age was 3 months and weight 3.5 kg . The pulmonary valve was hypoplastic in all patients. Median pulmonary valve diameter was 3.1 mm (range 2.7–5.2), Z-score -5.5 (range -8.9 to -4.4) RVOT stenting improved arterial oxygen saturation from a median of 60% (55–66%) to 91%

(82–94%). Median Z-score for the left pulmonary artery increased from -4.2 (-7.2 to -2.9) before stent implantation to -1.8 (-4.6 to -0.8) at time of surgery. Median Z-score for the right pulmonary artery increased from -3.1 (-6.2 to -2.1) to -0.5 (-2.1 to 0.2). There were no complications. Nine patients have undergone successful repair. There were no immediate or early deaths.
Conclusions: In the symptomatic young infant with TOF in whom surgery has been abandoned for any reason or patients at high risk, stenting of the RVOT provides a safe and effective management strategy, improving arterial oxygen saturation and encouraging pulmonary artery growth.

113: TRANSCATHETER VSD CLOSURE: COMPARISON OF DIFFERENT DEVICES AND METHODS IN SUCCESS RATE

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Background: Ventricular septal defects (VSD) were previously treated surgically. With the introduction of percutaneous VSD closure with different devices, the management of PM VSD, muscular VSD and residual VSDs has evolved. In our centre, Amplatzer and coil devices have been implanted for selected PM VSD, muscular VSD and residual post surgical VSDs since 2006.
Methods: The charts of all VSD closures since 2006 were reviewed retrospectively. Clinical, electrocardiographic, and echocardiographic data were analysed. The pre closure, immediate post closure, and 1-month, 6-month, and 12-month post closure results were assessed.
Results: One hundred and seven patients (50 male 46.7%, 57 female 53.3%) with mean age 9.40 ± 4.22 years (0–18 years) were evaluated .The mean diameter was 4.69 ±1.79 mm (minimum 2 up to maximum 12 mm) and mean pulmonary artery pressure (PAP) patients was 17.98 ± 6.86 mmHg (13–70 mmHg). The mean ratio QP:Qs 1.4 ± 0.13 was calculated respectively (1.3–1.8). Nineteen patients (17.9%) had LVH before the procedure. Twelve patients (11.2%), had trace, 5 patients (4.7%) mild and 11 patients (10.3%) had moderate aortic regurgitation. Sixteen patients (15%), had trace, 22 patients (20.6%) had mild and 7 patients (6.5%) had moderate tricuspid regurgitation. The results showed successful closure in 104 cases; 3 cases failed. Completely closed shunts were detected in 91.6% of patients (98 patients), while shunt remained in 5 patients (4.7%) and there was a trace tricuspid regurgitation in 4 patients (3.7%). Onset for all arrhythmias was within the first 24 hours of the procedure, and right bundle branch block was the most common.
Conclusion: Congenital VSD closure using transcathether devices resulted in stable improvement in clinical status and decreased interventricular shunting. Transcatheter device closure is an effective management option for patients with complex muscular VSDs that are difficult to approach surgically and for postoperative residual VSDs. The selection of patients with VSD still remains a challenge to avoid intervention complications such as complete heart block and aortic insufficiency. The selection of suitable device size and shape is most important for success rate.

115: OUTCOME AFTER PRENATAL DIAGNOSIS OF CARDIAC MALFORMATIONS PREDICTED TO REQUIRE A UNIVENTRICULAR PATH AFTER BIRTH

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Background: Data on the prognosis for children with univentricular (UV) hearts is mainly based on surgical follow-up. During prenatal counselling however, knowledge of the outcome after foetal diagnosis is necessary.
Materials and methods: This two-centre study analyses the outcome in 105 consecutive foetuses predicted to certainly (n = 79) or probably (n = 26) require a UV path. Termination was an option at < 22 weeks.
Results: The most common diagnoses were hypoplastic left heart syndrome (HLHS) + variants (37+7), unbalanced AVSD (17), coarctation of the aorta (CoA) + left ventricular (LV) hypoplasia (9), pulmonary atresia with intact ventricular septum (PA-IVS) (8), TA (8), AS (6), double inlet left ventricle (DILV) (5). Eight had aneuploidy, and another 9 extracardiac malformations. Eighty-eight had isolated cardiac defects. Forty of 67 (60%) diagnosed at < 22 weeks chose termination. Five foetuses died: AVSD + heart-block, CoA + diaphragmatic hernia, Ebstein + large TR, HLHS + trisomy 18, and PA-IVS + large TR. Of 60 live births, 12 died unoperated: 5 were inoperable, 4 preterm, 1 hydrops, 1 trisomy 18, 1 with extracardiac malformations. Forty-eight were operated on; 40 stage I, 32 stage I + II, 20 stage I–III, 1 stage I–III + transplant and 8 with biventricular (BV) correction (1 certain and 7 probable prenatal prediction of UV). Thirteen of the 48 operated on died (27%), 7 between stages I and II, 1 after an HLHS-hybrid procedure, 2 after stage II, 1 from malignancy after transplant and 2 after BV correction. Median age at postoperative death was 3 months (0.2–40). Median follow-up in survivors was 5.6 years (0.3–10). Diagnoses in survivors were HLHS + variants (15), TA (6), DILV (3), CoA (3), AVSD (2), PA-IVS (2), VSD + hypoplastic RV (1), complex UV (2) and complex transposition of great arteries (TGA) (1). Survival rate was 33% (35/105) after foetal diagnosis, 54% (35/65) in continuing pregnancies, 58% (35/60) after live birth and 73% (35/48) after surgery. Survival to surgery in continuing pregnancies was 74% (48/65).
Conclusions: Seventy-four per cent of the foetuses in continuing pregnancies survived to have surgery. Medium-term surgical survival was 73%. If diagnosed before 22 weeks, 60% chose termination. This is useful information in prenatal counselling.

144: CAN PRENATAL INTRACARDIAC ECHOGENIC FOCI AFFECT POSTNATAL CARDIAC FUNCTION?

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Objective: To determine whether prenatally diagnosed intracardiac echogenic foci (ICEF) are associated with neonatal cardiac dysfunction and persistence.
Methods: Foetuses with ICEF shown on prenatal sonography (between January 2010 and December 2011) at 1 perinatal centre underwent postnatal echocardiography at ages 1 month to 1 year. A single perinatal cardiologist assessed cardiac function by measuring the left ventricular shortening fraction (LVSF) and myocardial performance index (MPI). The presence of tricuspid valve regurgitation (TR) was sought.
Results: Prenatally, 24 foetuses had ICEF, mean age at diagnosis was 25 ± 3.1 weeks. Eighteen (75%) foetuses had left ventricular intracardiac echogenic foci (LVIEF) and 6 (25%) had right ventricular intracardiac echogenic foci (RVIEF). Postnatally, those infants, 14 (58%) males and 10 (42%) females were examined. at a mean age of 8.2 ± 4.1 months. Prenatally, all infants had normal LVSF. The overall mean left ventricular MPI (reference value, 0.36 ± 0.06), was normal for both infants with LVIEF (0.34 ± 0.06) and RVIEF (0.33 ± 0.04). Trace TR was noted in 12 (50%) of the infants. LVIEF persisted in 14 infants (77%), whereas RVIF persisted in 2 infants (33%).
Conclusions: Prenatally diagnosed ICEF can be persistent but is not associated with myocardial dysfunction in the first year of life.

150: OVAL FOSSA DEFECTS: MORPHOLOGIC VARIATIONS AND IMPACT ON TRANS-CATHETER CLOSURE

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Background: Incomplete formation of the partition between the two atrial chambers in the region of the oval fossa results in a range of defects, which extend from patent foramen ovale (PFO) to large secundum atrial septal defects (ASDs). There is wide variation in the morphology of the latter lesions. The spatial orientation of the margins of ASDs relative to the persisting flap valve is not easily definable with standard echocardiographic imaging. Careful evaluation of the morphology is essential in optimising successful trans-catheter closure to minimise complications. The advent of three-dimensional trans-oesophageal echocardiography (3DTOE) has changed our understanding of the morphology of these defects, and facilitates successful percutaneous closure.
Methods and results: Since 2007, over a 4-year period, we performed transcatheter closure of ASDs in 104 patients. During this period there were two instances of embolisation of the device. We evaluated carefully the morphology of the defects in the patients suffering embolisation, and noted an unusual spiral configuration of the flap valve relative to the rims of the oval fossa. These findings were then found in 4 additional patients, and serve as the focus of this report. To facilitate our understanding of the unusual morphology, we compared the clinical findings with images showing the mechanism of development of the atrial septum in the mouse, revealing a striking similarity.
Conclusions: Though uncommon, spiral spatial orientation of the margins of ASDs predisposes to embolisation of devices used for percutaneous closure. Standard cross-sectional techniques have limited use in identifying this variation. Understanding of development of the atrial septum in the mouse heart may help explain morphogenesis of the defect, and the mechanism predisposing to embolisation.

190: PREMATURE CLOSURE/RESTRICTION OF THE FORAMEN OVALE WITH SEVERE PRENATAL CARDIAC ASYMMETRY: WILL THE LEFT HEART BE SUFFICIENT POSTNATALLY?

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Background: Premature closure of the foramen ovale (FO) is a rare but serious condition that can be associated with severe right heart failure and hydrops, supraventricular tachycardia and left heart obstructive lesions.
Methods: We report 2 cases of early prenatal restriction/closure of the FO.
Results: Both patients were referred for foetal echocardiography, at 22 + 4 and 26 + 1 weeks respectively, for severe cardiac asymmetry with suspicion of 'hypoplastic left heart' (HLH) at ultrasoundUS. In the first case, a 30-year-old G1P0 woman, cardiac asymmetry was initially attributed to an aortic coarctation (CoA). Flow in the FO appeared normal up to 29 + 3 weeks. At 34 + 3 weeks, cardiac asymmetry worsened with a bulging septum towards the left atrium. At 37 + 1 weeks, the left heart appeared tiny with no visible interatrial flow. A 2.3 kg baby boy was born at 37 + 3 weeks. Postnatal echocardiogram showed a tiny restrictive FO, a dilated right heart with transient pulmonary hypertension and a smallish left heart. The aortic arch was small and disharmonious but no CoA developed. In the second case, a 32-year-old G2P0 patient, cardiac asymmetry was immediately attributed to a restrictive FO. A CoA was suspected at 31 + 1 weeks and the FO was closed at 34 + 4 weeks with increasing cardiac asymmetry. A 2.5 kg baby girl was born at 38 + 1 weeks. Postnatal echocardiography confirmed an intact atrial septum with suspicion of HLH, a bicuspid aortic valve and CoA. She was treated with prostaglandins. The follow-up echocardiograms showed a progressively better-filled left heart and reduction of pulmonary pressures. She underwent a CoA repair on day 13. Both children are currently well at 6-year and 2-month follow-up respectively.
Conclusions: Premature closure of the FO may mimic HLH pre- and post-natally. In some cases, despite severe cardiac asymmetry, postnatal evolution may be very good. Further research is necessary to better predict outcome.

226: ANDRASTENT IN THE MANAGEMENT OF COARCTATION OF THE AORTA

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Background: Stenting in coarctation of the aorta (CoA) has emerged as an alternative to surgery with good intermediate result. Recently a new bare metal stent made of a cobalt-chromium alloy (Andrastent XL, XXL , Andramed, Germany) was introduced to clinical practice. Its strong radial force, flexibility and good radio-opacity should be advantageous in implantation in CoA.
Objective: To evaluate the use of Andrastent XL and XXL in the management of CoA at a single tertriary care centre with immediate result and midterm follow-up.
Methods: Andrastents were implanted over a 30-month period in 29 patients: 25 with native CoA and 4 with recurrencet after previous surgery (ReCoA). The Andrastents were manually mounted on Maxi LD and BIB balloons and delivered through 10 to 14 Fr Mullins sheaths using a conventional femoral approach.
Results: Mean patient age was 28.3 ± 15.6 (range 9–65) years. The systolic gradient across the native CoA decreased from a mean 48.3 ± 20.2 before to 11.9 ± 10.2 mmHg after the procedure and in case of ReCoA from 37.8 ± 20.7 before to a mean 9.7 ± 12.4 mmHg after the procedure. No aneurysm formation, stent migration or rupture of the aorta were observed in any patient during the procedure. The mean fluoroscopy time was 6.1 ± 2.3 min. Procedural outcome remained favourable during mean follow-up 1.1 ± 0.8 without stent fracture. Planned redilatation of implanted stent was performed between 4 and 14 months in 6 patients. In one man with secondary LV failure ejection fracture (EF) 15% (49 years old), the procedure was performed urgently during cardiogenic shock with good clinical result.
Conclusions: Implantation of Andrastents XL and XXL is a very good therapeutical option for the treatment of native and recurrent CoA.

235: PRENATAL DIAGNOSIS OF HETEROTAXY SYNDROME: IMPACT OF SUBTYPE ON PERINATAL AND SHORT-TERM OUTCOME

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Objective: After prenatal diagnosis (Dx), to assess the influence of heterotaxy syndrome (HS) subtype on perinatal and short-term survival.
Methods: We included foetuses with HS from 1995 to 2011. We collected data on gestational age (GA) at Dx, cardiac anomaly, congenital heart block (CHB), extracardiac anomalies, pregnancy outcome, surgery, current survival and circulation.
Results: Of the 154 foetuses, 61(40%) were categorised as asplenia syndrome patients (ASP) and 93 (60%) polysplenia syndrome patients (PSP). Median GA at Dx was 21 (range 14–39 weeks). Complex cardiac anomalies were more frequent in ASP than PSP (98% vs 58%). Bradycardia-CHB was exclusive to PSP (18/93). Extracardiac anomalies were present in 45%. In ASP, 24/36 (66%) Dx at < 24 w elected for termination of pregnancy (TOP), 3 foetuses died from non-cardiac lesions, and 34 (56%) were liveborn. In PSP, 11/61 (18%) Dx < 24 w elected for TOP, 5 foetuses died (4 with CHB) and 77 (83%) were liveborn. Mean follow-up was 6.1 years (± 4.6). In the liveborn ASP, 15/34 (44%) died, 6 with pulmonary vein stenosis, 5 postoperatively and 4 from non-cardiac lesions. In the liveborn PSP, 10/77 (13%) died, 6 postoperatively, 2 with CHB, and

2 from extracardiac lesions. Of the current survivors, 100% of ASP have undergone cardiac surgery compared to 53% of PSP. Starting with foetal Dx, 64/93 (69%) of PSP are still alive, 57% with biventricular circulation. Of the ASP, 17/61 (28%) are still alive, only 5% biventricular.
Conclusion: After foetal Dx, there were significant perinatal survival differences between ASP and PSP. ASP had higher rates of TOP and postnatal mortality. The majority of PSP were biventricular compared to few ASP. CHB was a risk factor for foetal demise. Neonatal deaths were cardiac related in both groups whereas late deaths were due to extracardiac causes in PSP and to pulmonary vein stenosis in ASP.

249: VENTRICULAR SEPTAL DEFECT CLOSURE WITH A NEW PLATINUM-COATED NITINOL DEVICE

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Background: Three types of new platinum-coated nitinol VSD (COCOON™) devices were designed: perimembranous, aneurysmal perimembranous and muscular VSD devices, according to the morphology of ventricular septal defect (VSD). This study reports the animal trial and early clinical results.
Materials and methods: In the animal trial, VSD was created in 12 pigs by retrograde aortic approach for ventricular septal puncture with Brokenborough needle and followed by balloon dilation of the septum. The clinical trial included 16 VSD patients; age ranged from 2 to 56 years, weight from 10 to 72 kg. There were 8 perimembranous, 5 aneurysmal, 2 muscular and 1 post myocardial infarction VSDs.
Results: In the animal study, the device was successfully deployed to close the created VSD in the 12 pigs. Imaging studies demonstrated complete VSD closure in 11 animals. The autopsy findings demonstrated minimal and complete neo-endothelialisation over the device at 1 week and 8 weeks after implantation, respectively. In the clinical trial, the device was successfully deployed in 14 patients. Two unsuccessful cases underwent surgical closure. Two of the 14 successfully deployed cases developed haemolysis after the procedure; one due to residual VSD leakage and another due to undiagnosed aortic regurgitation with the regurgitant flow hitting the device. Both of them had surgical device removal and VSD closure. During the echocardiographic follow-up period (1 to 6 months), 3 cases still had slight leakage. There was no device-related complication.
Conclusion: The new nanoplatinum-coated nitinol VSD devices had a good outcome in animal trials. They also had attractive results in the early clinical trial.

287: OUTCOME OF TRANSCATHETER DEVICE CLOSURE OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT

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Purpose: To evaluate the safety and efficacy of transcatheter closure for perimembranous ventricular septal defect (PmVSD).
Methods: From September 2002 to June 2011, 155 PmVSD patients were admitted for percutaneous device closure (age 26 months to 21 years, weight 11–67 kg. Among these 155 patients, 12 patients were post surgery residual VSD, 1 patient was associated with Down syndrome. All patients had transthoracic echocardiography(TTE), ECG and 24h Holter pre- and post occlusion, and were asked to attend follow-up at 1 m, 3 m, 6 m, 12 m, and then once every year for 5 years, and then every 3 years. ECG and TTE were performed at every follow-up, and 24h Holter was performed only when necessary.
Results: Devices were deployed successfully in 151 patients (97.5%). The VSD size measured by left ventricle angiogram was 2.0–8.46mm on right ventricle side with multiple shunt in most patients with aneurism, and Qp/Qs was 1.4–2.18. The device used included 4 types with the difference in the shape of the left disc and the height of device, and the size of the device was 4–12mm. All patients were followed up until June 2012, with the follow-up duration 12–82m. During follow-up, 15 immediate adverse events (9.9%) were reported, including 2 complete atrioventricular block (AVB), 5 CRBBB, 3 CLBBB, 1 premature ventricular contraction, and 4 moderate TR. Almost all recovered, except 3 CRBB and 2 moderate TR were still present with normal heart chamber and function. There was also 1 late complete AVB at 3-month follow-up. To date no patient has required permanent pacemaker implantation or surgery.
Conclusions: Transcatheter PmVSD closure can be performed safely and successfully with low morbidity and mortality. Medium- to long-term prognostic results are favourable; however, late complications especially high-degree AVB should be additionally considered by all interventional cardiologists.

301: OUTCOME AND FEASIBILITY OF TRANSCATHETER ATRIAL SEPTAL DEFECT OCCLUSION UNDER TRANSTHORACIC ECHOCARDIOGRAPHY GUIDANCE IN LIMITED RESOURCES CENTRE: INITIAL REPORT

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Background: Transcatheter atrial septal defect (ASD) occlusion has become the established treatment of secundum ASD under transoesophagal echocardiography (TOE) guidance in many centres; nevertheless TOE may not be available in several areas.
Aim: To evaluate the outcome and feasibility of transcatheter ASD occlusion under transthoracic echocardiography (TTE) guidance in our institution
Methods: TTE was performed to assess the suitability of transcatheter occlusion. The occlusion procedures were performed under sedation and local anaesthesia. Routine right heart catheterisation was performed in all cases to assess the pulmonary vascular resistance. Balloon sizing was done in case of inadequacy of the rims. Amplatzer septal occluder device was used for occlusion. TTE and angiogram were done to assess the position of the device and the result of occlusion. All patients were followed up periodically clinically, with ECG and TTE at 24 hours and 1, 6 and 12 months after closure.
Results: Of a total of 45 patients who underwent non-surgical cardiac interventions from April 2011 to December 2011, there were 9 cases of secundum ASD which were suitable for occlusion. The median age, weight, and diameter of defect were 15 years (range 10–42 years), 32 kg (27–54 kg), and 24 mm (16–34 mm), respectively. Seven patients achieved complete immediate closure, and two cases failed as a result of deficit of inferior and floppy of posterior rim. There were no complications during procedures and follow-up. All patients were discharged the next day. Complete closure was seen on further follow-up.
Conclusions: Transcatheter ASD occlusion may be performed under TTE guidance which showed effectiveness and safe therapy, but it has limitations in assessing the inferior and floppy posterior rim. Further evaluation of this method is mandatory.

317: HEART SOUNDS AT HOME: PROSPECTIVE MATERNAL SURVEILLANCE OF SSA POSITIVE PREGNANCIES USING A HAND-HELD FOETAL HEART RATE MONITOR

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Background: The purpose of this study was to test a simple hand-held Doppler foetal heart rate (FHR) monitor in the ambulatory setting to prospectively identify the onset of atrioventricular (AV) block. This

may be useful in monitoring in maternal SSA/SSB disease.
Methods: We recruited pregnant women with SSA and/or SSB antibodies at gestational age (GA) 16–17 weeks from a large perinatal cardiology practice. Mothers were instructed and then given handheld Doppler FHR monitors. Between 17 and 27 weeks, mothers were instructed to listen for 1 minute twice daily and seek attention immediately if there were 'skipped beats' or a 'slow rate (< 120 bpm). The mothers returned to clinic every other week until GA 27 weeks for a foetal echocardiogram to assess the mechanical PR interval, endocardial fibroelastosis (EFE) and AV valve insufficiency. At the conclusion of the monitoring period the mothers filled out a questionnaire rating their experience. Newborns were evaluated by 12-lead ECG to confirm and verify conduction.
Results: Between 2009 and 2012, 11 mothers with SSA (9) or SSA/SSB (2) antibodies were given monitors at GA 16.5–18 weeks. Three mothers had previously had a child with AV block. Foetal AV block recurred in 2 of 3 foetal siblings. Each foetus had 'skipped beats' at GA 17.5 and 18 weeks, 12 hours after auscultation of normal foetal heart tones. The mother of foetus #1 sought medical attention 24 hours later; the foetus had EFE and 3° AV block. The mother of foetus #2 was seen within 4 hours; her foetus had intermittent 2° AV block. Both mothers were given high-dose vitamin D, IVIG and dexamethasone. Sinus rhythm was restored in foetus #2 and AV block unchanged in foetus #1. No foetus with normal FHR Doppler monitoring (n = 9) had echocardiographic abnormalities; echo did alter later management in foetuses #1 and #2. Ninety per cent of mothers were reassured by the FHR monitor, would recommend it and use it during subsequent pregnancies.
Conclusions: This pilot study suggests that, using a simple FHR monitor, mothers can prospectively detect the onset of SSA-mediated conduction system abnormalities; this capability may make early successful treatment feasible.

322: PERCUTANEOUS BALLOON ANGIOPLASTY FOR SEVERE NATIVE AORTIC COARCTATION IN YOUNG INFANTS UNDER 6 MONTHS

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Purpose: To assess the effects and the potential role of percutaneous transcatheter balloon angioplasty as an alternative therapy to surgical operation.
Methods: Thirty patients aged 6 d to 6 m (body weight 2.5–6.1 kg) with severe coarctation of the aorta (CoA) were admitted for balloon angioplasty. All 30 patients had cardiac dysfunction, and 8 patients had cardiac shock with severe metabolic acidosis. Ten patients had isolated CoA, while the others had other associated cardiac malformations. Cardiac catheterisation and aortic angiogram were performed under general anaesthesia with intubation. Balloon size was from 3 × 20 mm to 8 × 20 mm, and PTCA balloon was preferred for its low profile and small sheath.
Results: The femoral artery was successfully punctured in all patients except for 1 with further carotid artery puncture. The pressure gradient (PG) across the coarctation was 13–76 (40 ± 17.0) mmHg. The narrowest diameter was 0.5–2.8 (1.7 ± 0.6) mm. All patients had successful dilatation with PG significantly decreased to 0–40 (14.7 ± 11.7) mmHg, and the diameter significantly improved to 2.5–4.8 (3.7 + 0.9) mm. No intraoperative complications occurred. However, in the 1 case with carotid arteries puncture, a giant aneurysm at the puncture site needed surgical repair. During 6-month to 5-year follow-up, it was shown that: (1) PG crossing coarctation measured by echocardiography further decreased or remained stable in 28 cases (the remaining 2 patients, whose PG gradually increased, needed a second dilatation; no case needed further surgery because of CoA); (2) aortic aneurysm formation occurred in 1 case, which disappeared within 18 m; (3) tricuspid regurgitation and pulmonary hypertension improved in all patients; and (4) all patients were asymptomatic.
Conclusions: Percutaneous balloon angioplasty is relatively safe and effective and should be considered a valid alternative to surgical operation in view of its good effect, less trauma and low incidence of complications.

356: PRENATAL FOETAL ECHOCARDIOGRAPHIC FINDINGS IN TOTAL ANOMALOUS PULMONARY VENOUS RETURN IN A SERIES OF 25 PATIENTS

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Background/hypothesis: Prenatal diagnosis of total anomalous pulmonary venous return (TAPVR) is crucial given the possibility of clinically important obstruction to venous return which manifests postnatally. Optimal perinatal management in these cases involves timely identification and delivery at an appropriate centre. Antenatal diagnosis has been considered a challenge. We aim to show that consistent prenatal echocardiographic features exist in this condition.
Methods: This was a retrospective cohort review of foetal echocardiographic studies performed at the University of California, San Francisco from 2001 to 2012. We systematically reviewed the sonographic features that led to the diagnosis of TAPVR in each case.
Results: Over the study period, 25 patients were diagnosed prenatally with TAPVR, at a mean gestational age of 24 weeks. Four were isolated cases of abnormal pulmonary venous return, 21 had heterotaxy syndrome with additional cardiac abnormalities. Abnormal connections were supracardiac in 18 cases, cardiac in 1 and infracardiac in 6. Identification of a venous confluence posterior to the left atrium on axial images and presence of additional vertical venous channels apart from the vena cavae on the 3-vessel view or sagittal views were consistent diagnostic markers on 2D imaging. Cardiac asymmetry was not consistently noted. The most consistent Doppler finding was one of low-velocity mildly pulsatile mono- or bi-phasic flow in individual pulmonary veins, seen in 24/25 (96%) of the patients studied, irrespective of presence or absence of downstream obstruction.
Conclusions: The diagnosis of TAPVR can be suspected by a targeted approach with attention to important B-mode and spectral Doppler clues. Diagnosis can be suspected from standard cardiac views used in routine anomaly screening. The spectral Doppler waveform pattern is extremely helpful in identifying patients with abnormal connections and in patients with isolated TAPVR may be the most useful finding upon screening when gross 2D and colour Doppler appearance is apparently normal.

358: LEFT HEART STRUCTURES IN FOETUSES WITH CONGENITAL DIAPHRAGMATIC HERNIA AND THE EFFECT OF FOETAL ENDOSCOPIC TRACHEAL OCCLUSION

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Background/hypothesis: Small left heart structures have been observed in foetuses with left-sided congenital diaphragmatic hernia (CDH). Compression from herniated abdominal organs, and reduced filling of the adjacent left ventricle, seem to be the main pathophysiologic mechanisms. Fetoscopic tracheal occlusion (FETO) is a novel procedure performed in mid-gestation which seems to promote lung growth in foetuses with CDH, however, the cardiac effects of FETO are poorly described. Our aim was to study the effects of FETO on the heart and analyse the left cardiac structures size at birth.
Methods: This is a retrospective case-control study. We performed measurements of mitral, tricuspid, aortic and pulmonary valve and pulmonary artery diameters, ventricular lengths, left ventricular end-diastolic volume indexed (LVEDVi) to body surface area.

Comparisons were made between foetuses who underwent FETO and matched controls with similar CDH disease severity and foetal cardiac dimensions who did not undergo foetal intervention.
Results: A total of 35 foetuses were studied, 9 with FETO and 26 controls. All had liver herniation and lung-head ratio < 1 at foetal evaluation (average gestational age (GA) 24 wks). At birth, prior to CDH repair, the intervention group had: larger LVEDVi (16.8 ml/m^2 vs 12.76 ml/m^2, $p = 0.04$), increased LV length z-score (-2.05 vs -4, $p = 0.006$), larger LV:RV length ratio (1.43 vs 1.04, $p = 0.03$), increased LPA diameter z-score (1.71 vs -1.04, $p = 0.021$), better growth of aortic valve (-1.66 foetal to -2.18 neonatal in FETO, -1.09 foetal to -3.3 neonatal in controls, $p < 0.005$). A trend towards better growth of mitral and tricuspid valves was observed.
Conclusions: Left heart structures and LPA were larger postnatally in foetuses with CDH who underwent FETO than in those who did not. These data suggest that haemodynamic alterations are introduced with foetal tracheal occlusion which are associated with alterations in ventricular loading and may influence growth; further study of these observations is necessary to determine overall significance.

365: RELATIONSHIP BETWEEN LEFT HEART HYPOPLASIA AND CONGENITAL DIAPHRAGMATIC HERNIA IN HUMAN FOETUSES

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Background: Congenital diaphragmatic hernia (CDH) is associated with small left ventricle (LV), mitral valve (MV) and aortic valve (AV) *in utero*, but the pathophysiologic processes behind this observation and implications for prognosis are not well known.
Hypotheses: We hypothesised that both left and right-sided CDH severity would correlate with worsening left heart hypoplasia and lower left cardiac output. We analysed factors leading to abnormal foetal blood flow patterns that potentially contribute to left heart hypoplasia: compression, altered ductus venosus and reduced pulmonary blood flow.
Materials and methods: Retrospective cohort of foetuses with CDH, 2000-2010. Ultrasound-derived lung:head ratio (LHR), liver position, and hernia side were recorded. CDH severity was categorised as severe (LHR ≤ 1.0 and liver herniated into thorax) or mild (LHR > 1.0 or no liver herniation). Cardiac dimensions and combined ventricular output (CVO) were measured on echocardiogram at presentation. Foetuses with anomalies other than CDH were excluded.
Results: A total of 171 left CDH and 17 right CDH were included. Foetuses with severe left CDH had smaller MV (z -2.24 ± 1.3 vs -1.33 ± 1.08), AV(z -1.39 ± 1.21 vs -0.51 ± 1.05), and LV length (z -1.66 ± 1.28 vs -1.11 ± 1.45) z-scores and had a lower mean LV output as percentage of CVO (26% ± 10% vs 32% ± 10%) versus those with mild CDH (all $p < 0.01$). Although LV outputs were similar in severe right vs severe left CDH, right CDH average dimensions were larger (MV z -0.9 ± 1.25, AV z -0.8 ± 1.1, LV z -1.0 ± 1.34). Severe dextroposition and abnormal liver/ductus venosus position were independently associated with smaller left heart structures, while LHR and branch pulmonary artery measures were not.
Conclusions: Severity of left heart hypoplasia correlates with severity of CDH when the left diaphragm is affected, but not when the lesion is right-sided. Results suggest that both compression by left CDH and preload alterations from changes in ductus venosus flow are present and may be responsible for poor ventricular growth in LCDH.

389: USE OF LOCAL ANAESTHETIC (0.25% BUPIVACAINE) FOR PAIN CONTROL IN PAEDIATRIC CARDIAC CATHETERISATION: A RANDOMISED CONTROLLED TRIAL

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Background: In paediatric cardiac catheterisation procedures performed under general anaesthesia, local anaesthetic is used for some patients prior to femoral sheath removal. We found no published reports of the impact of local anaesthetic on pain after paediatric cardiac catheterisation, and mixed reports on its effectiveness in adults. Our purpose was to investigate the effects of local infiltration of 0.25% bupivacaine on self-report measures of pain and analgesic use up to 6 hours post-procedure in paediatric patients undergoing cardiac catheterisation procedures.
Hypothesis: We hypothesised that administering 0.25% bupivacaine around the femoral catheter insertion site in children under general anaesthetic at the end of cardiac catheterisation procedures would have no effect on pain scores and use of analgesics up to 6 hours post-procedure.
Materials and methods: A randomised controlled trial with 140 participants aged 7 to 18 years undergoing cardiac catheterisation under general anaesthesia via femoral vein/artery is being conducted. Participants were randomised to the intervention group receiving usual care plus subcutaneous infiltration of 0.25% at the femoral site just prior to sheath removal or to the control group who had femoral sheaths removed under general anaesthetic but without subcutaneous bupivacaine.
Results: To date 133 of 140 participants have been recruited and followed; the remaining 7 will be recruited by September 2012. Blinded midpoint data analysis was conducted for data safety monitoring purposes and found that recruitment should continue. Full results will be available by October 2012. Primary outcomes are patient-rated pain scores and use of analgesics post-operatively. Procedural information, such as size and number of catheters used and post-operative bleeding, will also be compared between groups.
Conclusion: This study's results will be the first to contribute to an evidence base regarding the effectiveness of 0.25% bupivacaine given just prior to femoral sheath removal for pain control after paediatric cardiac catheterisation.

391: CORONARY INTERVENTIONS IN CHILDREN WITH CONGENITAL HEART DISEASES

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Background: The small vascular anatomy of infants and children makes interventional treatment of impaired coronary perfusion, such as stenoses, complete occlusions and fistulae, demanding. Materials and techniques, appropriate for this young age group, have to demonstrate their ability to effectively treat these lesions and avoid problems, such as disruption, myocardial infarction or malfunction.
Methods and results: Between 2004 and 2011, 14 patients with an age of 9 days to 25 years (median 4.6 years) and a bodyweight of 1.7 to 65 kg (median 14 kg) underwent coronary intervention. In three cases emergency revascularisation of the left coronary artery was performed successfully, followed by stent implantation in one patient. Embolisation of coronary arterial fistulae with coils and vascular plugs was effective in 10 patients. An antegrade, retrograde or combined approach to achieve the most distal device placement preserved all side branches. One infant with pulmonary atresia and an intact ventricular septum was prepared for biventricular repair by step-by-step closure of the right ventricular to the coronary artery connections. No procedure-related deaths occurred.
Conclusion: Congenital and post-procedural coronary obstructive lesions can be treated effectively and safely with balloon dilation at any age. In coronaries impaired by external compression stent implantation can restore perfusion, but long-term results are still awaited. Interventional closure of coronary fistulae improves peripheral coronary arterial perfusion. A combination of these strategies allows the preparation of patients with pulmonary atresia and intact ventricular septum for biventricular repair.

392: INTERVENTIONAL CLOSURE OF MUSCULAR VSDS AT A YOUNG AGE

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Purpose: The presence of muscular ventricular septal defects (mVSD) may pose a high circulatory burden in young patients. Surgical closure is difficult because of the multilayer structure of these malformations. We present the course of 13 patients (age 10 days – 7.3 years, median 9 months; bodyweight 2.2 and 18 kg, median 8.7 kg) undergoing transcatheter closure of such defects.
Method: All closures were assisted by transoesophageal echo guidance. For transvascular closure of 5 singular and 8 multiple mVSDs 5 Amplatzer PDA II occluder, 6 Amplatzer VSD occluder and 4 Amplatzer vascular plugs IV were used. Follow-up time ranged from 23 days to 2.8 years (mean 1.2 years).
Results: Three defects were closed immediately; 9 mVSD showed a residual shunt immediately after the implantation procedure, which further reduced during follow-up in all patients. Two intra-procedural complications occurred. A sudden complete atrioventricular (AV) block led to transcatheter explantation of the device in one patient. This patient with a restrictive cardiomyopathy died during follow-up. In one patient on ECMO a left ventricular perforation with the device already in place had to be oversewn in an emergency operation.
Conclusion: With some of the newer devices available, which pass through smaller delivery sheaths of 4–6 F, interventional closure of mVSD has become a feasible option in the treatment of patents of all ages and a bodyweight from 2.2 kg on, who present with elevated right ventricular pressures and high shunt volume. The procedure is challenging in newborns and infants and severe complications may occur.

422: AMPLATZER DUCT OCCLUDER II IN ALIEN POSITIONS

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Background: Amplatzer Duct Occluder II (ADO II) is specially designed for closing long ducts in infants. There are few reports of 'off-label' use of ADO II in alien positions.
Aim: To evaluate the advantages and disadvantages of ADO II in alien positions.
Material and results: In this prospective study 51 cases of ventricular septal defect (VSD) and one case of coronary artery venous fistula (CAVF), as well as one aorto-right ventricular tunnel (ARVT) were closed with ADO II. Seventy-two cases of VSDs were closed with regular devices. Age ranged from 8 months to 21 years (mean 9.7 years). M:F sex ratio: 1.1:1. In 2 patients associated atrial septal defects (ASDs) were simultaneously closed with ASD devices. One patient had dextrocardia, 11 cases had mid or high muscular VSDs, 38 had perimembranous VSDs, and 2 had Gerbode defect. In one patient, 2 ADO II were used. The shortest fluoroscopic time was 4.2 min (mean 8.4 ± 4.1 min). Complete closure was achieved in all. One case of Gerbode defect closed with ADO II developed complete heart block and recovered with temporary pacing and steroids. None of the patients developed tricuspid regurgitation or aortic regurgitations. Fluoroscopic time was greater (5.6 ± 3.4 min) in the control group; 3 cases had residual shunt, and transient complete heart block and haemolysis occurred in one patient each.
Discussions: The advantages of ADO II are a very low profile and easy delivery through a 5F guiding catheter with short fluoroscopic time, at one-third of the cost of a ventricular septal occluder. A disadvantage is that it is not useful in VSDs of more than 6 mm.
Conclusions: ADO II is an excellent device in an alien position like VSD, Gerbode defects, CAVF, ARVT. The procedure time and the cost are significantly less than regular devices. The success rate is very high and complication rate is very low.

425: TRANSCATEHETER CLOSURE OF ATRIAL SEPTAL DEFECTS : SINGLE CENTRE EXPERIENCE IN A LIMITED RESOURCES PAEDIATRIC HEART CENTRE

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Background: Transcatheter closure of atrial septal defects (ASD) has been accepted worldwide as an alternative to surgical closure with good clinical results. This procedure plays an important role in most developing countries with limited resources: lack of cardiac surgeons and paediatric cardiac intensivists, limited intensive care unit (ICU) beds and limited funding.
Objective: To report our clinical experience in transcatheter closure of ASD in a paediatric heart centre where very limited funding and few human resources are available.
Materials and methods: We have performed successful transcatheter closure of 118 ASD, out of a total of 556 various interventional procedures of congenital and structural heart disease (21.2 %) from September 2002 to June 2012. The procedures were performed under general anaesthesia guided by fluoroscopy and transoesophageal echocardiography.
Results: The median age was 7.6 yrs (range 1.3–69.3yrs) with 93 (78.8%) female and 25 (21.2 %) male. Most patients' body weight was between 10 and 30 kg. The defect size was 17.8 mm (range 4–36 mm) and the implanted device size was 22 mm (range 10–38 mm). All devices were Amplatzer septal occluders. The mean procedure time was 109 minutes (range 50–74 minutes) with fluoroscopic time 30 minutes (range 12–83 minutes). One patient had an embolised right ventricle requiring surgical intervention; the success rate was 99.1%.
Conclusion: Transcatheter closure of ASD is an effective and safe procedure. In a country with limited resources this procedure can play an important role as an alternative to surgery in treating ASDs.

432: TAILORED MANAGEMENT APPROACH FOR CRITICALLY SICK CHILDREN AND LATE PRESENTERS WITH CONGENITAL HEART DISEASE

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Background and objectives: Re-conditioning before cardiac surgery in critically sick children is often needed. We report our experience using a tailored management approach in these patients.
Methods and patients: The charts of patients with congenital heart disease (CHD) who were judged to have high operative risk were reviewed. *Included* were patients with: large left to right shunt and ventilation for longer than 2 months, significant left to right shunts at multiple levels combined with malnutrition or recent infection, severely impaired cardiac function needing inotropic support and antifailure medications, recent infection, severe malnutrition (body weight < 5th centile), and critically sick patients during early postoperative course. *Excluded* were patients with: significant left to right shunts, who presented early, with minor growth retardation, and without recent active infection.
Results: Six patients were included. The median age was 13 months (2–48 months) and median weight was 4.6 kg (2.3–12.6 kg). Two patients had multiple left to right shunts and ventilator dependency. One with huge ventricular septal defect (VSD) presented at 4 years of age. Another had low body weight, large VSD and impaired left ventricular (LV) function, while the other two had early postoperative complications and ventilator dependency.

In the first category, a staged approach was essential. This was done by transcatheter closure of the patent ductus arteriosus (PDA), followed by pulmonary artery banding. This approach led to extubation. After proper nutrition, total correction was done. The patient

who presented at 4 years of age underwent banding of PA, later surgical repair of VSD. Because of long postoperative ICU stay, he needed percutaneous closure of his residual VSD and was then discharged. The patient with large muscular VSD and impaired LV function underwent percutaneous VSD closure with good result despite low weight (4.2 kg). One patient needed balloon dilation of residual coarctation; the other because of severe obstruction to his bilateral Glenn anastomosis, underwent stenting of these stenoses. Both had good results.
Conclusions: In severely sick children and late presenters with multiple shunts, a tailored management including therapeutic catheterisation and supportive measures are essential before cardiac surgery. Catheterisation and possible intervention should be considered early during the postoperative phase in severely sick patients.

433: PERCUTANEOUS CLOSURE OF CORONARY ARTERY FISTULAS: VARIOUS EMBOLISATION TECHNIQUES AND DEVICES

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Background/hypothesis: Percutaneous closure of coronary artery fistulas (CAF) has emerged as an alternative to surgery. Closure of CAF with coils has been well described. We aimed to review our experience of the closure of CAF with various techniques and devices.
Materials and methods: We retrospectively reviewed 6 patients (4 male, 2 female) with a median age of 10.6 years (range: 4.1–24.1 years) who had undergone percutaneous closure of CAF from March 2010 to March 2012. The closure results and clinical follow-up were analysed.
Results: The origin sites of the fistulas were left (3) and right (3) coronary arteries. The fistulas drained to the right atrium (3) and right ventricle (3). One patient had multiple drainage sites to the right ventricle. A single device was used in 4 patients: vascular plug (2), vascular plug II (1) and duct occluder II (1). Two patients required the use of more than one device (duct occluder and duct occluder II, 2 duct occluders with multiple coils). All devices were deployed retrogradely except in one patient. One patient had significant residual shunt at 6-month follow-up and was occluded by percutaneous technique. Follow-up studies 1.6–22.0 months (mean 13.9 months) showed complete occlusion in all patients.
Conclusions: Percutaneous closure of CAF is effective and safe with good results. With the availability of newer devices, most CAF can be closed percutaneously. Transcatheter closure should be considered as the treatment of choice.

442: RECOGNITION OF SURGICAL OUTCOME BIAS IN MULTI-CENTRE STUDIES: A METHODOLOGICAL APPROACH USING 109 CASES OF FOETAL AORTIC STENOSIS

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Background/hypothesis: Foetal Working Group of Association for European Paediatric and Congenital Cardiology studied influence of foetal valvuloplasty on natural course of aortic stenosis (AoS) from foetal diagnosis to determination of surgical pathway. To recognise bias, we tested a committee approach for the following hypothesis: institutional preference for neonatal AoS treatment overrides a committee's decisions based on morphological and clinical characteristics.
Material and methods: The committee re-assessed postnatal, pre-procedure imaging studies (including measurements and z-scores) of 109 liveborn infants with prenatally diagnosed AoS (2005-2011), treated in 24 institutions, blinded to foetal intervention, location and outcome. **Outcome measures for decision concordance:** Individual and consensus decision on univentricular (UV) or biventricular (BV) pathway; concordance for first (BV) and second (UV) decision for those converting BV-UV; image quality; specialism (foetal/paediatric cardiologist, interventionalist or surgeon) and institutional ethos.
Results: Of 109 infants 64 had initial BV outcome, with later conversion to UV circulation in 6. Consensus concordance for first pathway was 85/109 (78%) and for second: 81/109 (74%). There was concordance with initial BV surgical pathway in 4 of 6 requiring later conversion to UV. Poor imaging led to consensus 'undecided' in 3 of 4. Committee decision was BV in 16/45 (36%) final UV decisions and undecided in 1 (poor imaging). The interventionalist disagreed with first ($p = 0.015$) and final outcome ($p = 0.009$), grading more UV than other specialists. Those with greatest foetal intervention experience (GT, RM) agreed more with first and final outcomes: odds ratio (OR) 1.86 (1.03, 3.33; $p = 0.039$) and OR 1.87 (1.04, 3.38; $p = 0.037$). Committee's discordance was less for cases treated in surgical centres performing neonatal Ross-Konno (11% vs 27%).
Conclusions: The committee's consensus was more optimistic than eventual outcome, perhaps reflecting study members' experience in foetal intervention and neonatal Ross-Konno. This approach recognised greater discordance with institutions not performing Ross-Konno, leading to the potential to underestimate the influence of foetal valvuloplasty in achieving BV outcomes.

444: DOPPLER ECHOCARDIOGRAPIC DIAGNOSIS OF FOETAL LONG QT SYNDROME WITH FUNCTIONAL SECOND-DEGREE HEART BLOCK

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Background: A rare presentation of foetal long QT syndrome (LQTS) is a functional second-degree atrioventricular block (2°AVB), sometimes in association with ventricular tachycardia leading to congestive heart failure. Junctional and/or ventricular tachycardia has also been suggested as being characteristic of an acute stage of antibody-mediated heart block, requiring a completely different strategy of treatment. Recently observing a foetus with 2°AVB and abnormal diastolic relaxation, later confirmed to have LQT1, we speculated that this probably was an effect of a long refractory period in the ventricles, and a possible marker to differentiate LQTS from other causes of 2°AVB.
Material and methods: Isovolumetric relaxation (IRT) and contraction (ICT) time intervals were retrospectively determined from left ventricular inflow/outflow Doppler records obtained from 21 cases of foetal bradycardia. Five had 2°AVB (one LQTS), 7 3°AVB, and 9 blocked atrial bigeminy.
Results: A markedly prolonged IRT (105 ms) and a short ICT (7 ms) clearly distinguished our LQTS case from all other cases, where IRT values ranged between 29 and 67 ms and short ICT values only were seen in those with blocked atrial bigeminy. Long ICT values were seen in 75% of cases with antibody-mediated 2°AVB.
Conclusions: Even if our observation of IRT prolongation in LQTS with 2°AVB is based on only one case, it is not unexpected and could well be explained by a QT-interval exceeding the duration of mechanical systole. Moreover, IRT prolongation was not seen in any other case with other mechanisms causing the foetal bradycardia, suggesting that measurements of IRT and ICT could be a valuable complement to get a correct diagnosis.

449: STENT IMPLANTATION IN THE AORTIC ISTHMUS: AN ANIMAL MODEL OF HAEMODYNAMIC, HORMONAL, PRESSURE AND GENE EXPRESSION EFFECTS

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Background: Stenting of the isthmus has been increasingly adopted for treatment of the aortic coarctation in children and adolescents. However it is not ascertained whether this technique can contribute to the development of late systemic hypertension.
Hypothesis: Stent implantation in the isthmus in growing subjects can affect haemodynamic, hormone incretion, arterial pressure and gene expression in an animal model.
Methods: Sheep 3–5 months old underwent 2D-echocardiography, cardiac catheterisation, haemodynamic study and stent implantation in the isthmic region. Pressure was measured in the LV, ascending and descending aorta before and after stent implantation; sham animals underwent the same procedure without stent implantation. All were followed for 1 year, to adulthood. Quarterly pressure measurements and echocardiography were performed; blood samples were drawn for determination of renin-angiotensin-aldosterone levels. A second catheterisation, with haemodynamic study and a dobutamine test, was carried out 1 year after the enrolment, before the sacrifice. Samples were taken from myocardium, ascending and descending aorta for molecular biology examination.
Results: Four sheep received stent implantation, 4 sham animals were enrolled. The stent was oversized, considering the future growth. No differences in echocardiographic parameters or pressure measurements have been detected between the two groups in the interim evaluations. No pressure gradient across the isthmus was present at the follow-up; angiography did not show any narrowing. The dobutamine challenge test did not reveal differences, or differences in renin-angiotensin-aldosterone levels .The oxidative stress genes expression (MMP-9 and Caspase-3), showed a trend in significance in the expression of the MMP-9 that resulted higher in the ascending aorta of the animals with a stent.
Conclusion: Stent implantation in growing sheep does not affect haemodynamic, hormonal, or pressure parameters. The increased expression of MMP-9 in ascending aorta of stented animals could indicate a subliminal endothelial dysfunction, promoted by the presence of a rigid metal segment.

492: IMPACT OF FOETAL ECHOCARDIOGRAPHY ON PATIENTS' DECISIONS IN CHINA: A PRELIMINARY REPORT FROM A SINGLE CENTRE IN SHANGHAI

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Background: Foetal echocardiography allows for prenatal diagnosis of congenital heart disease or severe arrhythmia, and now serves as a routine screening tool for foetal cardiac anomalies in Shanghai. The aim of this retrospective study was to evaluate the impact of prenatal echocardiography diagnosis on parents' decisions.
Materials and methods: A total of 112 patients were referred to the prenatal counselling clinic in Shanghai First Maternity and Infant Hospital from July 2010 to Dec 2011, for detailed foetal echocardiography and prenatal counselling. The serial echocardiographic assessment was performed by an experienced foetal sonographer and paediatric cardiologist. Based on the diagnosis of foetal echocardiography, the paediatric cardiologist, obstetrician and neonatologist provide prenatal counselling for the couples, to outline the treatment options and to provide a clear picture of prognosis.
Results: Among these 112 patients, 41 foetuses (36.6%) were diagnosed with congenital heart disease, 5 (4.5%) with severe cardiac arrhythmia leading to haemodynamic compromise and 1 with a huge cardiac tumour. There was 1 stillbirth. A total of 23 patients (56.1%) with prenatally diagnosed congenital heart disease, 4 (80%) with severe arrhythmia and 1 with a cardiac tumour chose abortion. After birth two patients abandoned treatment, including 1 diagnosed with transposition of great arteries with abnormal coronary arteries and 1 with congenital complete atrioventricular block with extremely low heart rate.
Conclusions: In China, most parents are not willing to accept multiple staged surgical repair and possible long-term complications. Medical cost, neurodevelopmental outcome and ultimate quality of life play important roles in influencing parents' prenatal decisions.

533: NON-CONVENTIONAL USE OF OCCLUDER DEVICES

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Device closure is now an accepted modality of treatment for cardiac septal defects .We report the efficacy of closure of non-septal defects with devices conventionally used for septal cardiac defects.
Study design: Retrospective study.
Material and methods: We studies 46 patients, age group 2–67 yrs. These were divided into two groups: group 1: with no available customised device, group 2 : For which customised devices are available but alternative devices have been used. These included 37 in group 1: ruptured sinus of Valsalva (duct occluder 11), coronary arteriovenous (CAV) fistula (duct occluder 5), closure of paravalvular leak (5), mitral (4; duct occluder devices 3,ventricular septal defect (VSD) device 1) and aortic (duct occluder 1), closure of AP window (duct occluder 3), Fontan fenestration closure (3; atrial septal defect (ASD) occluder, patent foramen ovale device, vascular plug,1 each), pulmonary atrioventricular (AV) fistula (duct occlude 2), systemic AV fistula (vascular plug 1), closure of ascending aorta perforation (septal occluder 1), occlusion of subclavian artery (vascular plug1), splenic artery (duct occluder1), Blalock Taussig shunt (duct occluder 1). In group 2 there were 9 patients, VSD closure by ADO II device (6), PDA closure by muscular VSD device (2) and ASD device (1).
Results: Residual shunt was detected in 2 patients each of coronary AV fistula and mitral paravalvular leak. No shunt detected in ruptured sinus of Valsalva, fenestrated Fontan and ascending aorta perforation. Complications: Local site haematoma was observed in 4 patients. Haematuria was observed in 4 patients. There was one mortality in a large RSOV with gross congestive heart failure. On follow-up (2 months–6 years), all patients are asymptomatic with no late complications.
Conclusion: It is feasible to treat selected lesions successfully with the use of non-prototype occluder devices without significant complications.

535: SUCCESSFUL TREATMENT OF MULTIPLE MYCOTIC ANEURYSMS OF AORTIC COARCTATION WITH COVERED STENTS

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Introduction: Coarctation of the aorta constitutes 7% of all congenital heart diseases and is 2–5 times more prevalent in males. Infective endarteritis has been reported in18% of aortic coarctation and the resulting mycotic aneurysm has a high mortality. The treatment reported in different case reports for these aneurysms has been surgical. We report a case of a long segment coarctation treated with multiple covered stents.
Case report: A 22-year-old female was investigated for hypertension and fever for 3 months. She was known to have autoimmune disease with anaemia and moderate splenomegaly. Transthoracic echocardiogram revealed coarctation of aorta and transoesophageal echocardiogram showed multiple vegetations and aneurysms in the post stenotic segment. She was treated with ceftrioxone (for 4 weeks) and genta-

mycin (for 2 weeks) for *Streptococcus viridans*. MRA revealed a long segment coarctation distal to the left subclavian with multiple aneurysms in the post stenotic area. Conventional aortogram confirmed the above findings. The pressure gradient across the coarctation was 100 mmHg. Two overlapping covered CP stents (Nu-Med, New York, USA) were placed to dilate the coarcted segment as well as completely cover the aneurysms, reducing the pressure gradient to 5 mmHg. The recovery was unremarkable.

Discussion and conclusion: Osler introduced the term mycotic aneurysm in 1885 to describe any aneurysm that develops in context with bacterial endocarditis irrespective of the pathogen. Mycotic aneurysm has a high risk of rupture in coarctation of aorta with up to 50–100% mortality. Treatment of adult coarctation with covered stent is a standard practice in several centres. The PTFE covering is useful in treating aneurysms complicating bare metal stent implantation or coexisting patent ductus arteriosus. This report highlights the safety of endovascular covered stents in managing mycotic aneurysms of coarctation of the aorta not reported to date.

538: UNUSUAL SYSTEMIC VENOUS COLLATERALS CHANNEL TO LEFT ATRIUM CAUSING DESATURATION AFTER FONTAN OPERATION CLOSED PERCUTANEOUSLY

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Abstract: We present an unusual cause of progressive cyanosis in a child 2 years after successful Fontan surgery. One large venous channel and two small venous channels were draining into the left atrium (LA) directly; the larger channel was closed by Amplatzer vascular plug and small venous collaterals were closed by coils, resulting in improvement of oxygen saturation.

Case report: A 7-year-old girl with tetralogy of Fallot (TOF), with hypoplastic right ventricle and adequate branch pulmonary arteries, had undergone modified right BT shunt in infancy followed by BD Glenn, ligation of pulmonary artery and takedown of BT shunt at 16 months, followed by an extracardiac Fontan at 5 ½ years. She had a smooth postoperative course with saturation of 95 %. She had gradual return of cyanosis, exertional dyspnoea and easy fatigability over 6 months. Her saturation was 85% at rest and 78% after 10 minutes walking. Transoesophageal echocardiogram (TEE) revealed normally functioning Fontan circuit, normal ventricular function, and no atrioventricular (AV) valve incompetence, pericardial or pleural effusions. One venous channel was seen arising from the brachiocephalic vein. On cardiac catheterisation, intracardiac and Fontan pressures were normal. Inferior vena cava (IVC) angiogram revealed a large tortuous venous channel arising at T8-T9 level and draining into the LA of 12 mm diameter, an additional small vein was seen connecting the larger venous channel to the LA. Innominate vein angiogram revealed a small venous channel communicating with the LA. These channels were occluded with 8 mm Amplatzer plug and two coils. Post-deployment angiograms revealed complete occlusion of all the venous channels, with 94% saturation. The patient remains well at 6-month follow-up. We could not find a similar case report in the English literature.

Conclusion: Large venous channels draining directly into LA can be closed percutaneously. Long-term follow-up is necessary.

545: PRENATALLY BORDERLINE LEFT HEART A PROBLEM AT ALL?

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Background: Borderline left heart (Border-LH) raises the question of two- versus single-ventricle circulation. Aim of the study was detailed analysis of prenatal Border-LH and further postnatal outcome.

Patients and methods: We analysed 14 patients (9 female) with Border-LH identified prenatally between 2008 and 2012. 1. Prenatal echocardiographical measurements included: mitral/tricuspid valve (MV/TV) annulus, aortic/pulmonary valve (AOV/APV), left/right ventricular (LV/RV) width and length; these were compared to 35 healthy matched controls (NORMAL) and to 17 patients with hypoplastic left heart syndrome (HLH). 2. Postnatal anatomy and outcome were evaluated, with median follow-up 15 months.

Results: 1. Prenatally, in Border-LH significantly smaller structures were found compared to NORMAL but bigger compared to HLH. Border-LH/NORMAL/HLH: median MV/TV 0.60/0.92/0.42 ($p < 0.0001$), AO/AP 0.54/0.91/0.32 ($p < 0.0001$), LV/RV width 0.59/0.93/0.42 ($p < 0.0001$), LV/RV length 0.92/0.99/0.41 ($p < 0.0001$). 2. Postnatal z-scores: median MV -1.2, AOV -1.05; smaller (z-score ≤ 2) MV and/or AOV in 3 patients (21.4%). Additional findings were: ventricular septal defect (VSD) in 7 (50%), bicuspid aortic valve in 6 (42.9%), and persistent left superior vena cava to coronary sinus in 3 (21.4%) patients. 3. All patients were able to maintain a biventricular circulation postnatally, 1 (7.1%) without (w/o) intervention. Thirteen (92.9%) developed pathology, 10 (71.4%) underwent coarctation of aorta (CoA) repair, and 1 VSD closure. Three (21.4%) early deaths occurred: 2 prior to surgery, 1 after CoA repair. 4. No late deaths were registered. By 2 months MV and AOV were all within normal range. In 2 (14.3%) balloon angioplasty for re-CoA was performed, in 2 (14.3%) mild mitral stenosis developed, w/o intervention needed.

Summary: Border-LH means a thinner LV with almost normal length and smaller MV and/or AOV prenatally; but with normal postnatal growth potential. Despite the positive consequence of biventricular circulation, these patients are not free of interventions. CoA can be expected with arterial duct closure. Frequently associated pathology indicates that this is not just prenatally 'lower-flow' circulation but probably represents a more complex morphological problem.

563: FOETAL PULMONARY VALVULOPLASTY BY PERCUTANEOUS TRANSHEPATIC ACCESS IN A LAMB MODEL

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Background/hypothesis: Foetal pulmonary valvuloplasty may ameliorate hypoplastic right heart syndrome and mitigate postnatal disease. Foetal heart access by direct foetal heart puncture is well-described. We have recently developed a percutaneous transhepatic foetal cardiac catheterisation technique, which may be safer and offer technical advantages. We hypothesised that foetal pulmonary valvuloplasty could be performed by a percutaneous transhepatic approach at mid-gestation.

Materials and methods: Nine foetal lambs at 97-100 (term 147) days' gestation (average weight: 1 215 g) were studied under maternal general anaesthesia. Under ultrasound guidance, the foetal hepatic vein was percutaneously punctured using a 16 GA IV-cannula with needle *in situ*. A 2.4/1.9Fr coronary catheter was inserted over a 0.014 inch floppy-guidewire, and the inferior vena cava (IVC), right atrium (RA), right ventricle (RV), pulmonary artery, ductus arteriosus and descending aorta were catheterised. After removing the guiding catheter, but with the guidewire in place, a coronary PTCA dilatation catheter was positioned across the pulmonary valve, and several inflations of the balloon were performed simulating a valvuloplasty. Seven foetuses were euthanised post-procedure, and 2 were euthanised after term-delivery, for postmortem examination.

Results: Percutaneous cannulation of the foetal hepatic vein followed by RA and RV catheterisation was successful in all cases. One foetus

died during catheterisation following RV perforation. In the remaining 8 cases the coronary catheter was advanced to the descending aorta. Pulmonary valvuloplasty was successful in 5 cases using catheters with a 6 mm long balloon; postmortem showed minimal haemorrhage without cardiac trauma. The procedure was unsuccessful in 2 cases (both survived) using a 12 mm long balloon which could not be turned into the pulmonary artery; postmortem showed small RV perforations. In 1 case the PTCA catheter could not be inserted as the cannula became dislodged.
Conclusions: Foetal pulmonary valvuloplasty by percutaneous transhepatic cardiac catheterisation is feasible, providing an alternative route for human foetal cardiac intervention.

572: SPECTRUM AND OUTCOME OF CARDIOMYOPATHIES DIAGNOSED DURING FOETAL LIFE

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Background: Primary cardiomyopathies (CM) are potentially irreversible, life-threatening heart muscle disorders that typically affect the cardiac filling and/or contractility in the absence of any anatomical abnormality. These are rare *in utero* diagnoses and, in consequence, there is a paucity of knowledge on the entity. We sought to clarify the foetal disease spectrum and to identify early echocardiographic predictors of outcome.
Methods: We reviewed 53 consecutive cases with prenatally detected CM at our tertiary care centre since 2000. Hypertrophic CM (HCM) was defined by the presence of ventricular wall thickness > 2 z-scores. Non-hypertrophic CM (NHCM) was defined by cardiac dysfunction in the absence of myocardial hypertrophy. Excluded were cases with cardiac hypertrophy secondary to maternal diabetes and twin-twin transfusion.
Results: NHCM was diagnosed in 34 and HCM in 19 cases at 24.5 + 5.6 weeks' gestation. Aetiology included familial (9%), inflammatory (17%), and genetic-metabolic disorders (36%), while 38% were idiopathic. The pregnancy was terminated in 9/53 (17%). Of 44 actively managed cases, 20 (64%) survived to infancy and 3/17 (18%) with HCM versus 14/27 (52%) with NHCM remain alive (Hazard ratio (HR) 2.9; 95% confidence interval (CI) 0.98–8.7; p = 0.03). Baseline echocardiographic variables associated with mortality of actively managed cases included ventricular septal wall thickness (HR 1.15 per z-score increase; 95% CI 1.03–1.29; $p < 0.02$); foetal hydrops (HR 5.22; 95% CI 1.13–24.16; $p < 0.03$); umbilical vein pulsations (HR 9.5; 95% CI 2–45.3; p =0.005); > 2 diastolic Doppler flow abnormalities (HR 3.54; 95% CI 0.98–12.77; $p < 0.02$); left ventricular myocardial performance index (HR 2.6 per 10% rise in index; 95% CI 1–9; $p < 0.05$) and cardiovascular profile score (HR 1.45 per deducted point; 95% CI 1.10–1.90; $p < 0.01$).
Conclusions: Foetal CM is caused by a broad spectrum of causes and associated with significant perinatal mortality, especially those with HCM. Early echocardiographic detection of diastolic dysfunction is most useful in predicting adverse outcomes.

574: PERCUTANEOUS RECONSTRUCTION OF FUNCTIONAL AORTIC INTERRUPTION USING PERFORATION AND COVERED STENT

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Introduction: Coarctation of aorta is usually surgically treated in infancy and childhood, if untreated may lead to mortality. Rarely may result in acquired interruption.
Objective: Reconstruction of functional aortic interruption using perforation technique and covered Stent.
Methods: Three cases were suspected clinically and diagnosis confirmed by echocardiogram, cardiac CT and/or MRI. Cardiac catheterisation was done under general anaesthesia. Pre and post interruption areas were accessed by radial artery/transseptal puncture and femoral artery respectively. Pre and post interruption areas were assessed by angiograms. Transseptal needle 6 Fr with dilator and sheath was advanced from femoral access and with aid of angiogram through other access. The needle was used to perforate the atretic part into the ascending aorta, position confirmed by pressure and angiogram. Then with help of a PTCA (0.014) wire, dilator and sheath were advanced and the sheath was replaced to Mullens 12 Fr. After appropriate measurements, a covered stent was selected and mounted in BIB balloon taking into consideration the diameter of the transfer aortic arch and length to cover the whole atretic part of aortic arch and extended to descending aorta about 10–15 mm avoiding left subclavian artery origin. Post stent deployment pressure and angiograms were recorded.
Results: There were no early or late complications. All patients were discharged home within 3 days. Mean follow-up was at 2 years and the gradients obtained were unchanged.
Conclusion: Percutaneous treatment of functional aortic atresia is a safe, feasible and effective alternative

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593: FOETAL ECHOCARDIOGRAPHY IN A DEVELOPING COUNTRY: REFERRAL PATTERNS AND IMPACT ON PREGNANCY OUTCOMES

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Background: Limited data exist about the practice of foetal echocardiography in developing countries with logistic limitations to treat complex congenital heart disease (CHD).
Objective: To report referral patterns for foetal echocardiography and its impact on outcomes of affected pregnancies in a newly established foetal cardiology unit in India.
Patients and methods: A foetal cardiology service was started in January 2008. Prospective data (2008–2010) was collected in a dedicated database; retrospective data (2006–2007) was obtained from hospital records. Outcomes were tracked by direct evaluation of newborns, questionnaire or telephonic interviews.
Results: A total of 1 084 women were referred for foetal echocardiography; mean gestational age at referral was 24.9 + 5.9 weeks. The most common indication for referral was abnormal screening ultrasound (38.4%). Referrals for indications other than suspected CHD were more in the current period (2008–2010) (63.8% vs 50.5%; p =0.0006). CHD was diagnosed in 297 foetuses (27.5%), 114 (38.4%) were simple and 183 (61.6%) were complex. The most common outcome was termination of pregnancy (36 %) or intrauterine/neonatal death (18.9%). Termination rates were higher for complex CHD (47% vs 18.4%; $p < 0.0001$). Continuation of pregnancy with planned perinatal care was more common in the current period (25.5% vs 10 %; $p = 0.02$) and for simple CHDs (36.4% vs 14.2%; $p < 0.001$).
Conclusions: Referral patterns for foetal echocardiography were characterised by late referrals, especially when CHD was suspected. Although half of the pregnancies affected with CHD resulted in termination or foetal/neonatal deaths, with improved awareness, there was a trend towards planned deliveries and postnatal care, especially for repairable CHDs.

619: THE EDWARDS VALEO LIFESTENT® FOR TREATMENT OF CARDIOVASCULAR LESIONS IN CHILDREN

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Background: The Edwards Valeo Lifestent® is a stainless steel, premounted, open cell stent. Easy dilation to larger diameters and low profile are advantages in growing children. Radial force is however poor.
Results: Between April 2011 and July 2012, 26 VALEO® stents were placed in 22 children during 23 procedures. Median age was 1 year (5 d–14.3 yrs), median weight 8 kg (2–53). Most insertions (87%) were percutaneous (group 1) while 3 were peri-operative (group 2). Indications and locations were: right pulmonary artery (RPA) or left pulmonary artery (LPA) (rehabilitation strategy) in pulmonary atresia with intact ventricular septum (PA-IVS) in 6; LPA stenosis in tetralogy of Fallot, PA-IVS or ventricular septal defect (VSD) in 4 (3 peri-operative); PA stenosis in single ventricle in 2, PA stenosis after arterial switch in 2, sub-hepatic vein thrombosis in 1, patent ductus arteriosus (PDA) stenting (hybrid approach) for hypoplastic left heart syndrome (HLHS) in 6 and for complicated IAA in 2 patients. In group 1, stent placement was straightforward, through femoral venous access in all except 3 (2 jugular, 1 transhepatic). Predilation was performed in 5, all with PA stenosis; subsequent postdilation was performed in 2. Immediate angiographic and/or haemodynamic results were considered satisfactory in all. Acute complications occurred in 3 (haemoptysis in 2, reperfusion oedema in 1). Six patients have so far been recatheterised, a median of 1.7 mo after initial procedure. The stent remained fully patent in all but 1. In group 2, the stents were always secured with a single proximal stitch and flared at their proximal end. All 3 patients had early post-op recatheterisation (median 6 mo). All stents were redilated to achieve better wall apposition. In 1 a possible stent fracture without obstruction was seen.
Conclusion: The VALEO® stent is an easy and effective stent for use in growing children. Radial force was sufficient in the lesions we encountered. When used peri-operatively, early recatheterisation is warranted to improve wall apposition. Longer-term follow-up is needed.

628: HEMOPTYSIS AFTER FENESTRATED EXTRACARDIAC FONTAN - A MULTIDISCIPLINARY APPROACH TO MANAGEMENT

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Background: Haemoptysis after fenestrated Fontan repair for univentricular physiology is a well-known complication of chronic cyanosis. We present our management of a 10-year-old who presented with massive haemoptysis 5 years after completion of extracardiac Fontan.
Summary: TD underwent a superior cavopulmonary anastomosis at age 2 for his transposition of great arteries with a small left ventricle (LV) and PS. He had a fenestrated extracardiac Fontan done at age 5. At age 10 he presented with sudden-onset haemoptysis. After stabilisation he underwent a right heart catheterisation to delineate the conduit-anatomy; an aortic digital subtraction angiogram (DSA) was also performed to delineate possible bronchial collaterals. Right heart catheterisation revealed a 7 mm conduit-right atrium (RA) fenestration while the aortic DSA revealed multiple and large bronchial collaterals. Our interventional radiologist used 300–500 PVA (polyvinyl alcohol) particles to successfully embolise the bleeding collaterals. We planned to close the large conduit-fenestration at a staged setting with an ASD occlusion device. Angiography and trans-oesophageal echocardiogram (TEE) revealed an anatomy which favoured using a Amplatzer Duct Occluder -2 (ADO-2, St Jude Medical) device for closing the fenestration. This was successfully achieved using an ADO-2 6-6 device under fluoroscopy and TEE guidance. Post procedure angiogram revealed no right-to-left shunting. Pressure in the venous-PA conduit prior to device closure was 11/5 mmHg and post-procedure was 10/5 mmHg. Arterial blood gas oxygen saturations improved to 96%. He remains well and fully saturated 3 months later.
Conclusion: We would like to report this case as using a multidisciplinary approach helped in reducing procedure time and radiation time in a haemodynamically unstable patient. It was also a novel use of PVA particles for successful embolisation of the bronchial collaterals (with possible advantages over coil occlusion). There have been no reports of using of ADO-2 device for percutaneous closure of the fenestration.

638: BALLOON VALVULOPLASTY IN PREMATURE INFANTS WITH SEVERE AORTIC AND PULMONARY VALVE STENOSIS OR PULMONARY ATRESIA

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Background: The rationale for foetal balloon valvuloplasty is to prevent the development of left or right ventricular hypoplasia in the setting of severe aortic, pulmonary stenosis or pulmonary atresia intact septum.
Material and methods: A retrospective review of premature infants, weight < 2 kg, gestation < 36 weeks, undergoing balloon valvuloplasty was performed
Results: There were 6 with severe aortic stenosis, gestational age 32–36 weeks, birth weight 1.4–1.9 kg. treated in the first 10 days of life. Nine with severe/critical pulmonary valve stenosis, gestational age 28–36 weeks, birth weight 1.2–1.9 kg were treated during the first 9 days. Three with pulmonary atresia, weights 850, 1700 and 1 900 g, had radiofrequency assisted balloon pulmonary valvuloplasty. Three with aortic stenosis developed transient loss of a femoral artery pulse. The systolic gradient or left ventricular systolic pressure fell in every case. One developed severe aortic regurgitation and required a Ross operation at two months. One patient required repeat aortic valvuloplasty at 6 months. In 2 cases a Ross operation was undertaken at 5 and 7 years respectively. One patient has reached 9 years without a second intervention. Relief of pulmonary stenosis occurred in every case. In one case repeat pulmonary valvuloplasty was required at the age of 12 months but no other patient has required re-intervention 6 months to 12 years following the initial valvuloplasty.

Of 3 patients with pulmonary atresia one born at 27 weeks died 10 days later from non-cardiac causes. The 2 others did not require an additional source of pulmonary blood supply and have required no further intervention 8 and 1 year later.
Conclusions: Balloon valvuloplasty is a safe procedure in premature infants. Elective premature delivery before 36 weeks' gestation with postnatal balloon valvuloplasty could be an alternative to foetal intervention in selected cases.

689: PRENATAL ECHOCARDIOGRAPHIC FINDINGS IN HYPOPLASTIC LEFT HEART SYNDROME WITH CORONARY FISTULA

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Background: Prenatal diagnosis of hypoplastic left heart is commonly performed during foetal life. Identifying risk factors to better predict outcome is still challenging.
Case report: We describe here the prenatal echocardiographic features of a rare association of hypoplastic left ventricle with coronary fistula. Foetal echocardiography at 33 weeks' gestation showed a small left ventricle (LV) of 6 mm with mitral hypoplasia. The LV had a globular appearance with filled apex and a poor systolic function. A small 1.5 mm aorta arising from the LV and a biphasic Doppler flow recorded in the aortic root suggested aortic regurgitation. The ductus arteriosus perfused the horisontal and ascending aorta. The

pulmonary artery diameter was 8 mm. The delivery was planned in a tertiary centre. In the delivery room, Apgar was 5/8 and evidence for low cardiac output was already noted. Prostaglandin perfusion was started. Echocardiography showed a large left coronary fistula filling the LV. This was confirmed by computed tomography (CT) angiography. Subsequently, compassionate care was planned and the child died. In prenatal echo, the LV cavity size was overestimated because the LV was filled by the coronary fistula. In addition, this fistula was responsible for a steal phenomenon that further compromised LV function while no endocardial fibrosis could be evidenced.
Conclusion: Coronary fistula is a rare associated lesion in hypoplastic left ventricle. Awareness on this association might be useful to better predict postnatal outcome and to improve prenatal counselling.

695: INTRACARDIAC ECHOCARDIOGRAPHIC GUIDANCE FOR TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT IN AN UNSELECTED ADULT PATIENT POPULATION

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Background: Intracardiac echocardiography (ICE) is an alternative to transoesophageal echocardiography (TEE) for transcatheter closure of ostium secundum atrial septal defect (ASD). However, studies on ICE guidance generally include a majority of patent foramen ovale and only few ASDs. Our aim was to assess transcatheter closure of ASDs under ICE guidance in an unselected patient population, paying particular attention to complex cases with large (> 38 mm) ASDs and/or deficient rims other than the anterior-superior.
Materials and method: From January 2006 to January 2012 in our centre, all ASDs in the adult population were closed percutaneously under ICE guidance, except for 3 patients with Down syndrome who underwent closure under general anaesthesia and TEE guidance. During this period, 93 patients (59 females) underwent transcatheter ASD closure at a median age of 48 (21–90) years with Amplatzer devices under local anaesthesia and ICE guidance. All patients had routine TEE before catheterisation.
Results: Complex cases comprised 17 patients (18%) including 13 with one or more deficient rims other than the anterior-superior and 8 with an ASD size > 38 mm. The median ASD diameter by TEE and ICE was 20 (6–40) mm and 25(10–40) mm, respectively. Ninety cases (97%) were successfully closed. Three cases failed because of insufficient rims and/ or defect size superior to 40 mm. Minor and transient complications (atrial arrhythmia, groin haematoma) occurred in 10 patients (9.6%). Three patients experienced a major complication with favourable outcome: one arterial tear (laceration of a branch of hypogastric artery) during venous puncture attempt successfully treated by percutaneous embolisation, one blood transfusion for a groin haematoma and one retroperitoneal haematoma. The only risk factor for failure to close the ASD was deficient rims ($p = 0.05$).
Conclusion: In an unselected adult population, ICE provides safe and efficient guidance for device closure of ASD, even for large defects with deficient rims.

706: TRANSCATHETER CLOSURE OF SECUNDUM ATRIAL SEPTAL DEFECT ASSOCIATED WITH DEFICIENT RIMS OTHER THAN THE ANTERO SUPERIOR

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Background: We aimed to assess the feasibility of transcatheter closure for secundum atrial septal defects (ASDs) with deficient rims (< 5 mm), other than the antero-superior because it is well demonstrated that deficiency in the anterior rim toward the aorta does not influence the success rate for transcatheter ASD closure.
Materials and method: Between 1 January 2008 and 31 December 2011, 192 patients underwent percutaneous closure of ASDs in our institution, under transoesophagal echocardiography (TEE) guidance in children and intracardiac echocardiography guidance in adults. Amplatzer devices (Amplatzer Septal Occluder or Cribriform) were used in 191 whereas 1 case without deficient rim was closed with intrasept ASD occluder. We retrospectively analysed the outcomes of the 43 patients (22.4%, 26 children) with one or more deficient rim.
Results: The median age and weight was 16 (1.1–85) years and 56 (8.8– 99) kg, respectively. Deficiency of the inferior (11), inferior-posterior (15), or of the superior-posterior rim (17) was confirmed by TEE in all the cases. Transcatheter closure was successfully accomplished in 37 (86%) of the cases with a median Amplatzer septal occluder size of 28 (10–40) mm. A modified method of implantation (sizing balloon technique) was used in 30 patients (70%).In 6 patients (5 children) the ASD could not be closed. Four other children experienced device embolisation a few hours after successful transcatheter closure and underwent surgical ASD closure without further complication. With use of Fisher's exact test, deficiency of the inferior-posterior rim was associated with failure or embolization ($p = 0.05$) and there was a trend for adult age to be associated with a low risk of embolisation or failure ($p = 0.06$).
Conclusion: Transcatheter closure of secundum ASD is feasible in patients with deficient rims. However, embolisation may occur, especially in the paediatric population with deficient inferior-posterior rim.

712: EARLY RESULTS FROM THE UNITED STATES MELODY^{s} TRANSCATHETER PULMONARY VALVE POST-APPROVAL STUDY

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Background: This study was undertaken to evaluate short-term outcome after percutaneous transcatheter pulmonary valve (TPV) implantation in the 'real-world' clinical setting. The primary outcome is TPV function and freedom from reintervention at 6 months. We present procedural and early results.
Methods: Implantation procedures were based on IFU guidelines. Data were collected prospectively on all consecutive subjects enrolled. Procedural success was defined as TPV appropriately fixed in position; catheter gradient < 35 mmHg; and < trivial pulmonary regurgitation by angiography. Successful TPV function at endpoint was mean echocardiographic gradient < 30 mmHg and mild or less regurgitation.
Results: Between 29 July 2010 and 23 May 2012, 127 patients were enrolled at 10 USA centres for possible TPV implantation at a mean age of 20.0 ± 9.7 y. The primary indication was conduit stenosis in 19.8%, regurgitation in 42.9%, both in 30.2%. Most (78%) patients were NYHA I or II. Most conduits were homografts (65.9%); 12.7% were bioprosthetic. TPV implant was attempted in 98 patients, 97 (99%) were successful. Procedural success rate was 93% (90/97) with no mortality. Serious procedure-related events included conduit rupture (6 – all treated with covered stents; 5 underwent TPV, 1 surgery), pulmonary artery perforation (1), and coronary compression requiring emergent surgery (1). At discharge, the average mean

gradient was 16.9 ± 7.6 mmHg; 97% had no or trace pulmonary regurgitation. At mean follow-up of 7.7 ± 6 mo 2 TPV were explanted, 1 for endocarditis and 1 during aortic root replacement. Another patient was treated for presumed endocarditis.
Conclusion: These early data suggest TPV in standard clinical practice yields procedural results equivalent to those in the IDE study. The procedure appears an acceptable alternative to surgical conduit replacement. Endocarditis risk warrants further study.

735: ATRIAL LEVEL RESTRICTION INFLUENCES CEREBROVASCULAR IMPEDANCE IN THE FOETUS WITH TRANSPOSITION OF THE GREAT ARTERIES

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Background: In transposition of the great arteries (TGA) the aorta arises from the right ventricle, which leads to alteration in cerebral blood flow and oxygen delivery *in utero*. Restriction at the atrial septum requires balloon atrial septostomy (BAS) after birth, but may also influence patterns of cerebral blood flow before birth.
Objective: To characterise cerebrovascular impedance in the foetus with TGA and investigate the relationship to atrial septal restriction.
Methods: Foetuses with TGA, intact ventricular septum, 2 good-size ventricles and no outflow tract obstruction had middle cerebral artery pulsatility indices (MCA-PI) measured and compared to normal gestational age-matched controls. TGA group was divided into BAS and no-BAS after birth, reflecting degree of prenatal atrial restriction.
Results: Thirty-six foetuses with TGA (17 no-BAS; 19 BAS) were compared to 126 controls. Overall there was no difference in MCA-PI between TGA and normal. Comparing cohorts of similar gestational age, at the end of gestation (36–39 w) BAS group had MCA-PI similar to normal, which was significantly lower than no BAS group (BAS 1.52 + 0.07 vs no BAS 2.02 + 0.15, $p < 0.01$).
Conclusions: MCA-PI normally decreases at the end of gestation (36–39 w). MCA-PI is normal in the foetus with TGA and a restrictive atrial septum, but is abnormally elevated near the end of gestation when there is an open atrial septum. We speculate that a restrictive atrial septum leads to increased mixing in the right atrium, with cerebral oxygen delivery that is closer to normal, while under conditions of open atrial septum, streaming leads to severely deoxygenated blood delivery to the brain. Chronic cerebral hypoxia in TGA with open atrial septum leads to cerebrovascular dysregulation, with markedly elevated MCA-PI. Childhood neurocognitive deficits in TGA may in part have their origins in prenatal life as a consequence of variability in cerebral blood flow and oxygen delivery.

753: ARE COVERED STENTS PROTECTIVE AGAINST ACUTE LOCAL COMPLICATIONS IN HIGH-RISK PATIENTS WITH AORTIC COARCTATION?

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Background: Percutaneous dilatation and stenting of aortic coarctation (CoA) has proved to be an effective alternative to surgery. Because of complications associated with bare metal stent (BMS) such as local dissection, covered stent (CVS) were introduced.
Aim and hypothesis: To compare the results and complication rate between BMS and CVS in the treatment of CoA. We tested the hypothesis that a 50% or more stenosis in relation to the *aorta* at the *diaphragm* level is a significant risk factor associated with BMS for acute local complications including *aneurysm* formation and bleb tears.
Materials and methods: We did a retrospective chart review of patients treated at the collaborating centres, comparing pre- and post-procedural data and procedural details. Patients who received BMS with stenosis ≥ 50% were considered high-risk (HR-BMS). The 2 BMS groups we compared to a group of patients who received a CVS, all of whom had stenosis ≥ 50%. Acute complications were classified as '*in situ*' (*stent migration, intimal tear, and aortic aneurysm*) or major (death, *stroke, wall rupture, cardiac arrest, transfusion*, and necessity of urgent reintervention). Early and mid-term MRI studies were reviewed for potential *in situ* complications.
Results: There were 25 BMS recipients (0.74–71.47 yo; 25.7 ± 18.49), 9 were HR-BMS (0.74–49.46 yo; 20.63 ± 15.97, and 18 CVS recipients (12.4–58.0 yo; 26.74 ± 14.63). Early total complication rate was 9/25 (36%) in the BMS group, and 6/9 (66.6%) in the HR-BMS compared to 4/18 (22.2%) in the CVS group ($p = 0.503$ & 0.108 respectively). Major complications occurred in 3/25 (12%) BMS patients, and in 3/9 (33.3%) HR-BMS, compared to 1/18 (5.56%) CVS, ($p = 0.628$ & 0.093 respectively). *In situ* complications were encountered in 3/25 (12%) BMS patients, or 1/9 (11.1%) HR-BMS, compared to 0/18 (0%) CVS patients; ($p = 0.25$ & 0.33 respectively). Finally, the recatheterisation rate was 6/25 (24%) for BMS, and 4/9 (44.4%) in the HR-BMS compared to 3/18 (16.67%) in CVS; ($p = 0.71$ & 0.175 respectively).
Conclusion: Based on our limited series, the use of covered stent did not significantly reduce the risk of early *in situ* complications. Long-term follow-up imaging is needed for appropriate assessment of the hypothetical risk of long-term aortic wall integrity following CoA stent procedures.

774: PROFILE AND PREGNANCY OUTCOMES OF FETUSES WITH CONOTRUNCAL ANOMALIES IN A NEWLY ESTABLISHED FOETAL CARDIOLOGY UNIT IN SOUTH INDIA

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Objective: To describe the profile and immediate pregnancy outcomes of foetuses with conotruncal anomalies (CTAs) in a newly established foetal cardiology unit in South India.
Methods: Records of 68 women identified with diagnosis of CTA on foetal echo (mean gestational age 26.75 ± 5.93 weeks; range 17–38 weeks) during the period 2008–2011 were reviewed.
Results: The most common indication for referral was suspected congenital heart disease (CHD) during routine antenatal scan (89.7%). The various CTAs diagnosed included tetralogy of Fallot (TOF, 44.1%), double outlet right ventricle (DORV, 27.9%), transposition of great vessels (TGA, 8.8%), TOF with pulmonary atresia (TOF-PA, 8.8%), TOF with absent pulmonary valve (TOF-APV, 7.4%) and truncus arteriosus (2.9%). Extracardiac anomalies were reported in 4 foetuses (7.1%). Pregnancy outcomes were reported for 65 foetuses (95.6%), with 4 lost to follow-up. The most common outcomes included termination of pregnancy (41%), planned delivery and post-natal care (41%), intra-uterine death (13%) and neonatal death (5%). Termination of pregnancy rates, based on individual diagnosis, were as follows: TOF (43.3%), DORV (42.1%), TGA (50%), TOF-PA (16.7%), TOF-APV (40%) and truncus (50%). Overall post-natal survival among patients with intention to treat was 86.9% with 5 patients undergoing surgery in the neonatal period. The accuracy of foetal echo in identifying the primary diagnosis was 96.4%.
Conclusion: Pre-natal diagnosis of CTA prompted post-natal care in less than half of the diagnosed patients with a very high proportion of even repairable CTAs undergoing termination of pregnancy. Post-natal survival of those with intention to treat was excellent.

803: TRANSCATHETER CLOSURE OF PERIMEMBRANOUS VENTRICULAR SEPTAL DEFECT USING AMPLATZER DUCTAL OCCLUDER

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Background: Transcatheter closure of perimembranous ventricular septal defect (PMVSD) is a challenging procedure. Recently, the Amplatzer PMVSD occluder (AGA Medical Corp, Plymouth, Minnesota) has been shown to be effective in closing haemodynamically significant PMVSDs. However, the high incidence of complete atrioventricular block (CAVB) after device occlusion of a PMVSD using it has been a controversial issue, too. Among several devices tried to close PMVSD percutaneously, we prefer to use the Amplatzer ductal occluder (ADO, AGA Medical Corp, Plymouth, Minnesota) because of the anatomic resemblance between PMVSD with aneurysm and patent ductus arteriosus.
Materials and methods: Between August 2009 and June 2012, 21 patients underwent percutaneous PMVSD closure using ADO. There were 5 males and 16 females; median age was 6 (6–42) years, and median weight was 23 (18–60) kg.
Results: All patients showed echocardiographic signs of left ventricular volume overload and trivial to small mitral regurgitation (Qp/Qs = 1.7 ± 0.4). The mean defect size of the right ventricular side was 4.7 ± 0.8 mm. Devices 2 mm larger than the measured narrowest VSD diameter were selected in most patients. The ADOs were successfully implanted in all patients without any significant complications except one transient CAVB and one case of delivery wire fracture. Small residual shunts were observed immediately after device implantation, but have been disappeared during follow-up in 20 of 21 patients. Mean follow-up period was 18 ± 8 months, and CAVB or aortic regurgitation was not observed in any patients.
Conclusions: Transcatheter closure of PM VSD with the ADO is a safe and promising treatment option, but long-term follow-up in a large number of patients would be warranted.

808: UTILITY OF FOETAL ECHOCARDIOGRAPHY IN THE EVALUATION AND TREATMENT OF FOETAL ANAEMIA

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Background: Significant foetal anaemia can cause foetal compromise as a result of high foetal cardiac output (CO), with intrauterine transfusion (IUT) being the treatment of choice. IUT carries risks including preterm delivery and foetal demise. Peak middle cerebral artery (MCA) Doppler velocity of > 1.5 multiples of the mean (MoM) is sensitive for moderate-severe foetal anaemia with false-positive rates of ≥ 12%. We sought to explore the additive value of foetal echocardiography (FE) and calculated foetal combined CO (CCO) in assessing foetal anaemia.
Methods: We identified pregnancies that underwent FE prior to IUT for suspected foetal anaemia from June 2009 to June 2012 in our programme. FE was used to calculate foetal CO. Prenatal records were reviewed for peak MCA velocity at IUT, pre-transfusion haemoglobin (Hb) and hydrops. Anaemia was graded as moderate if foetal Hb was < 0.65 MoM and severe if < 0.55 MoM.
Results: Eleven pregnancies complicated by foetal anaemia had 20 IUTs. Nine of the 11 pregnancies had FE within the preceding 30 days. One case was excluded because of no pre-transfusion Hb measurement. Median gestation at FE was 24 wks (range 19–29wks). Peak MCA Doppler was > 1.5 MoM (range 1.5–2.5) in all. Of the nine foetuses with measured Hb, 44% had severe ($n = 4$, 3 of whom with hydrops), 22% moderate ($n = 2$) and 33% ($n = 3$) mild anaemia. Foetal CO was above the 90th centile in all foetuses with moderate or severe anaemia. In 3 with mild anaemia, 2 had normal CO and 1 had CO at the 90th centile.
Conclusions: FE-derived CO may be a useful additional tool in the evaluation of foetal anaemia severity. A normal foetal CO, even with elevated peak MCA Doppler, may allow the clinician to predict mild anaemia at foetal blood sampling. Surveillance using FE may avoid or delay the need for invasive management and the associated foetal morbidity and mortality.

811: EASE OF RETRIEVABILITY OF ADO II - AN ADDED ADVANTAGE

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Background: Amplatzer Duct Occluder II (ADO II - AGA Medical Corporation, Plymouth, USA) has found wide favour in transcatheter closure of large ductal shunts as well as in retrograde transcatheter closure of perimembranous and muscular vertricular septal defects (VSDs). This is attributable to the fabric-free design and thinner nitinol wire mesh rendering device delivery through smaller catheters feasible. However, this design also enhances the flexibility of the device, making it prone for deformation and migration when deployed across large (> 5.5 mm) high pressure shunt lesions, despite optimum placement of the device.
Materials and method: We report our institutional experience of ADO II from August 2011 to date; 27 devices were implanted (perimembranous VSD 19, patent ductus arteriosus (PDA) 6, muscular VSD 1, aortopulmonary window 1) in 27 patients aged 4 months to 12 years (mean age 4.7 years) with 3 devices migrating to the pulmonary artery. One was in an infant with a 5.5 mm Type A PDA closed with 6-4 ADO II and the other two were children with 5 mm perimembranous VSDs where a 5-4 ADO II was deployed. The retrieval was done by first positioning a 7 Fr Mullins sheath in the distal main pulmonary artery through which a 5 Fr snare catheter and 5 mm gooseneck snare (Cook incorporated, Bloomington, USA) is advanced to the embolised device. The proximal screw of ADO II was snared and the device withdrawn into the 7 Fr Mullins sheath.
Results: In all three cases, the devices were retrieved successfully and subsequent transcatheter closure performed with a nitinol duct occluder.
Conclusion: ADO II is not ideal for use in high pressure shunt lesions measuring > 5.5 mm. However, the fabric-free design and the thinner nitinol wire mesh renders it easily retrievable even after being completely deployed.

832: TEN YEARS' EXPERIENCE OF TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECT IN CIPTO MANGUNKUSUMO HOSPITAL, JAKARTA, INDONESIA

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Background: Recently transcatheter closure of ventricular septal defect (VSD) is more popular compared to surgery because of shorter hospitalisation, eliminated need for thoracotomy, and better cosmetic results without scar.
Objective: To evaluate the efficacy and safety of transcatheter closure of VSD.
Methods: A case series of patients who underwent transcatheter closure of VSD in Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia.
Results: In our hospital since February 2002, transcatheter closure of VSD has been done in 43 patients. For perimembranous VSD, 38 were closed using membranous VSD occluders and 3 were closed using ADO II; the 2 muscular VSDs were closed using Amplatzer muscular VSD occluder and Amplatzer septal occluder, respectively. There were 24 males. Age ranged from 5 to 46 years (median 6 years) and the median body weight was 19 (range 8–68) kg. The size of VSD was 3.5 (2.0–11.0) mm. Systolic pulmonary artery (PA) pressure was

108 (87–164) mmHg and flow ratio was 1.3 (0.6–5.0). The size of device used was 5 (4–12) mm. Fluoroscopy time was 42 (10–159) minutes and procedure time was 147 (38–305) minutes. Small residual shunt was observed in 1 patient following VSD closure using ADO II. All patients were discharged on day after procedure. Patients were followed-up periodically clinically and by echocardiography to monitor the presence of residual shunt or complications. During procedure and follow-up no significant complications occurred and no patient had complete atrioventricular block.
Conclusion: Based on our experience we conclude that percutaneous transcatheter closure of VSD is effective and save.

887: IMPROVED OUTCOME AFTER PERCUTANEOUS ULTRASOUND-GUIDED STENTING OF THE FOETAL ATRIAL SEPTUM IN LEFT ATRIAL HYPERTENSIVE CARDIAC LESIONS

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Background: Hypoplastic left heart syndrome with a highly restrictive or intact atrial septum (HLHS-RAS) has a very high mortality. Foetal left atrial (LA) hypertension results in abnormal lung development with lymphangiectasia and severe pulmonary vascular disease. We report our experience of *in utero* percutaneous ultrasound-guided stenting of the foetal atrial septum to decompress the LA.
Methods: Retrospective review of foetuses with HLHS-RAS or a variant that underwent active perinatal management since 2000 at our tertiary care centers.
Results: Ten foetuses were identified. Of 6 cases without prenatal intervention, 2 died *in utero* (33, 29 weeks) from progressive hydrops. The 4 foetal survivors required the urgent creation of an atrial communication immediately after birth but died subsequently (5-54 days). Percutaneous ultrasound-guided catheter interventions were performed in 4 more recent foetuses at 28-36 weeks. A 20 cm non-bevelled 18 G needle was advanced via the maternal uterus, amniotic fluid, the foetal chest and right atrium across the muscularised thickened inter-atrial septum to stent the atrial septum with a coronary stent expandable to about 3.5 mm. Elevated LA pressure, dilatation and Doppler flow pattern of the pulmonary veins, and magnetic resonance imaging (MRI)-estimated lung perfusion all immediately improved after the procedure. Three of the 4 stented foetuses were born by vaginal delivery. Atrial septectomy and additional surgical procedures were performed within 72 hours of birth. Intraoperative lung biopsy demonstrated muscularised pulmonary veins and lymphangiectasia in all 4. Two of the 4 foetuses developed severe or moderate stent stenosis *in utero* and both died after birth from pulmonary hypertension and sepsis, respectively. The other 2 remain alive >12 months of life, a significant improvement in survival compared with non-stented cases ($p = 0.03$).
Conclusion: Foetal LA decompression by atrial septal stenting may decrease lung injury and has the potential to improve survival in HLHS-RAS.

925: SUCCESSFUL PALLIATIVE BALLOON DILATION OF NATIVE COARCTATION OF THE AORTA IN A PRETERM VERY LOW BIRTH WEIGHT INFANT

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Balloon dilation of coarctation of aorta may provide adequate palliation in preterm infants, by relieving symptoms and allowing growth until definitive surgical repair can be performed. We report successful balloon angioplasty in a preterm very low birth weight (VLBW) infant with coarctation of the aorta.

Case report: The patient was a 30-week gestation preterm female infant with birth weight of 1 050 g. She had respiratory distress syndrome and required mechanical ventilation. The echocardiogram revealed moderate size of ventricular septal defect and patent ductus arteriosus at 13 days. The aortic arch could not be identified because of poor echo window. She was treatmed with a diuretic. At 1.5 months, she developed hypertension with 50 mmHg blood pressure differential in upper and lower extremities. The echocardiogram revealed coarctation diameter of 1.6 mm. with maximum pressure gradient (PG) 60 mmHg and mean PG 17 mmHg. The balloon dilation was done at 2 months of age (body weight 1 000 g) via 4 Fr sheath at right femoral artery. The aortogram revealed hypoplastic transverse arch 3.2 mm, aortic isthmus 2.2 mm, coarctation 1.8 mm. and descending aorta 4.3 mm. Balloon dilation was done with coronary balloon (Hirye) 4 × 20 mm, requiring 2 attempts. The peak to peak PG across coarctation after balloon insertion was reduced from 70 to 12 mmHg. She was successfully extubated after 2 days of intervention. The echocardiogram revealed residual coarctation 2.5 mm. with mean PG 9 mmHg after 12 days of intervention. Two weeks later, she was weaned off oxygen. She was discharged from hospital 3 months after intervention with body weight 2 090 g.
Conclusion: We were successful in palliative balloon dilation of coarctation of the aorta in a preterm VLBW infant.

926: ROUTINE ULTRASOUND-GUIDED VENOUS ACCESS FOR ELECTRO-PHYSIOLOGY PROCEDURES REDUCES INADVERTENT ARTERIAL PUNCTURE AND THE RISK OF ARTERIOVENOUS FISTULA FORMATION

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Background: Electro-physiology (EP) studies are commonly performed in children and require the insertion of multiple electrodes into the heart. This requires multiple points of venous access, including femoral, subclavian or brachial venous access. There is evidence to suggest that percutaneous femoral venous access carries a risk of inadvertent arterial puncture (IAP) with development of arteriovenous (AV) fistulae. In the United Kingdom, National Institute of Clinical Excellence (NICE) guidance suggests that ultrasound guidance should be used for central venous access.
Objective: To assess whether the routine use of a vascular ultrasound machine (SONOSITE M-TURBO) reduces the risk of arterial puncture and thereby the causation of AV fistulae.
Methods: We utilised a departmental database (Oscar 4D) to perform a retrospective review of the procedural notes of patients who underwent EP studies in our institution from June 2008 to June 2011. Our database contains information on access methods and complications.
Results: A total of 122 EP studies (87 radio frequency ablations) were performed in 108 children under the age of 16 in the time period reviewed, with between 3 and 5 sites of venous access per study. Successful femoral venous puncture was performed in 100% of patients, successful brachial venous puncture was performed in 97.5% (119/122) of patients under echo guidance, with IAP reported in none. There were no reported cases of AV fistulae in over 500 episodes of access, with up to 3 years of post-procedural clinical outpatient review.
Conclusion: Our experience shows that ultrasound-guided venous access is safe and achievable for children undergoing EP studies, with a reduction of the expected risk of IAP from published data and no episodes of post-procedural AV fistulae. We recommend the routine use of ultrasound to reduce these potentially significant complications.

964: RESULTS OF TREATMENT OF FOETAL TACHYCARDIA IN 80 SUBJECTS IN SWEDEN

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Background: Foetal tachycardia represents a risk for morbidity and mortality and there is no consensus concerning medical treatment.
Aim. To evaluate the treatment of foetal tachycardia with emphasis on choice of first-line agent.
Materials and methods: Eighty of 128 referrals with foetal tachycardia received transplacental antiarrhythmic treatment and were reviewed.
Results: Fifty-one foetuses were diagnosed with atrioventricular re-entry tachycardia (AVRT), 23 had atrial flutter (AF), 5 permanent junctional reciprocating tachycardia (PJRT), one junctional ectopic tachycardia (JET) and one atrial ectopic tachycardia (AET). Drugs used included sotalol, digoxin, flekainide and amiodarone.
Conclusions: In this small series sotalol was more effective than digoxin in terminating tachycardia without hydrops. In hydropic foetuses neither sotalol nor digoxin was effective in terminating the tachycardia as first-line agent.

993: TRANSCATHETER CLOSURE OF RIGHT DUCT IN INFANT WITH ISOLATED RIGHT SUBCLAVIAN ARTERY FROM MAIN PULMONARY ARTERY WITH PULMONARY AND SUBCLAVIAN STEAL SYNDROME: AN EXTREMELY RARE CASE REPORT

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Background: Isolated subclavian artery without associated congenital cardiac defect is an extremely rare. This anomaly can cause pulmonary and subclavian steal with perfusion of the arm and lung via the vertebrobasilar system. Reimplantation of subclavian artery into the ascending aorta is considered a conventional management. However, transcatheter embolisation may be a proper alternative for small infants with this condition.
Methods and results: We report an infant girl diagnosed with 22q11.2 deletion syndrome who presented with continuous murmur, heart failure and poor perfusion of right arm. The MDCT angiography demonstrated isolated right subclavian artery from main pulmonary artery via right duct with retrograde flow of right vertebral artery. Moreover, collateral branches of prominent right intercostal arteries supplied the right subclavian artery. The right duct was completely occluded with vascular plug II for stopping pulmonary overcirculation. The angiogram showed that subclavian steal phenomenon was diminished as a result of closure of the duct. Right subclavian artery was supplied by right vertebral artery and right intercostal arteries. Consequently, her perfusion of right arm and heart failure were clinically improved.
Conclusions: Transcatheter occlusion of isolated right subclavian artery from main pulmonary artery with steal phenomenon is an effective and alternative treatment for small infants.

995: HYBRID MANAGEMENT OF A PRENATALLY DIAGNOSED PULMONARY ARTERY TO LEFT ATRIUM COMMUNICATION

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Background/hypothesis: Symptoms of a pulmonary artery (PA) to left atrium (LA) fistula appear at different stages of childhood depending on the severity of the shunt. In newborns, central cyanosis and severe heart failure are signs that an emergency management must be done. Open heart surgery is the gold standard for the management of PA to LA fistula but with the arrival of new technology, transcatheter interventions are becoming the elective choice in some cases.
Materials and methods: We report a case of a female newborn with a prenatal diagnosis of right pulmonary artery (RPA) to LA communication managed with a hybrid technique. Pulmonary aneurysm was diagnosed in the last trimester.
Results: After caesarean section, because of severe heart failure, central cyanosis and hypoxaemia, urgent catheterisation was performed and a RPA to LA fistula with a large pulmonary cavity is found. She was taken to emergency surgery. Intraoperative findings included type III PA to LA fistula and 4 mm patent ductus arteriosus (PDA). On extracorporeal circulation without aortic clamping the fistula and the PDA were ligated. Because of a persistent image of an upper right pulmonary lobe cavity, catheterisation was done and a 6 mm plug was implanted in the efferent branch of the fistula, creating a successful embolisation. The patient was discharged a few days later with normal development seen on follow-up. Hybrid management of this rare entity was successful.
Conclusions: Prenatal diagnosis is important for planning and performing emergency operations, and in the presence of anomalies in the PA, a fistula between the PA to LA must be suspected. Team work on the hybrid procedure is a useful tool to manage this rare entity.

1012: FOETAL AND POSTNATAL OUTCOME OF CONGENITAL HEART DISEASE DIAGNOSED IN UTERO - A SINGLE CENTRE EXPERIENCE

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Background: Foetal echocardiogram (FE) is an established procedure to diagnose foetal heart disease; however foetal and postnatal outcomes are variable depending on social, financial and cultural factors. We report our experience of FE and their outcome.
Materials and methods: A total of 1 390 FE for suspected abnormalities were performed from January 2004 to June 2012 at our centre; 430 (31%) foetuses had a significant finding in FE. The maternal risk factors, indications, diagnosis and outcome of the foetal heart disease were analysed.
Results: The mean maternal and foetal age was 25.9 ± 4.1 years and 25.6 ±4.6 weeks respectively. The indications for FE were high-risk pregnancy (45%), suspected congenital heart disease (CHD) (22%), previous sibling with CHD (18%), arrhythmia (7%) and intracardiac echogenic focus (8%). Among the abnormal FE of 374**,** CHD accounted for 83.4%, arrhythmias 13.6% and cardiac tumours 2.9%. Heterotaxy syndrome was the commonest diagnosis contributing 20%, followed by hypoplastic left heart syndrome (HLHS) 11.6%, isolated ventricular septal defect (VSD) 9%, double outlet right ventricle (DORV), dextro-transposition of the great arteries (d-TGA), single ventricle and tetralogy of Fallot (TOF) contributed 6–7% each. Other defects contributed 21.4%.

One foetus with severe aortic stenosis underwent balloon valvotomy at 24 weeks. Two hundred and eight women opted for medical termination of pregnancy, a few beyond the legal gestational age limit. Foetal SVT was seen in 11, of which 10 were successfully controlled by transplacental pharmacotherapy. One foetus with sinus bradycardia was diagnosed as LQTS after birth. Twenty babies underwent emergency intervention after birth, 5 babies underwent definitive cardiac surgery as a single stage. The remaining babies were followed up medically.
Conclusions: Late referral FE is commonly seen in our geographical location. Good outcome is predicted in a subset of CHD with definitive treatment, arrhythmias and tumours. Late terminations beyond the legal limit are noticed in complex foetal CHD.

1013: CHANGES IN HEALTH-RELATED QUALITY OF LIFE OVER 3 YEARS FOLLOWING TRANSCATHETER PULMONIC VALVE IMPLANTATION: RESULTS OF A CONTROLLED INTERNATIONAL MULTICENTRE TRIAL

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Introduction: Transcatheter pulmonic valve (TPV) implantation has been shown to be an effective treatment for patients with right ventricular conduit failure. In addition to improvements in ventricular function, overall health-related quality of life (QOL) is an important measure of patient satisfaction. The aim of this study is to evaluate long-term clinical performance and whether TPV leads to QOL improvement.
Methods: TPV was implanted in 63 patients between October 2007 and April 2009 at 7 centres throughout Europe and Canada. Procedural outcomes, haemodynamic effects, and QOL (EQ-5D) measures were evaluated during pre-implant, discharge, 6 month and annual visits.
Results: Forty-two patients (age ≥15 years) who completed the EQ-5D assessments at pre and post-implant were included for analysis: 30 were male; mean age 26 ± 11 years (range 15–59). The underlying congenital heart diseases included tetralogy of Fallot (45%) and truncus arteriosus (17%). Eighty-three per cent of conduits were homografts. Compared to pre-implant (81%), patients in NYHA class I/II increased at 1 year (93%) and sustained through 3 years. Mean pressure gradient pre-implant was 37±12 and 1 year post-implant 17 ± 6 mmHg which remained similar through 3 years. None of the patients experienced moderate/severe pulmonary regurgitation at discharge and throughout 3 year follow-up. EQ-5D index and health state significantly improved at 6 months and remained improved through 3 annual visits ($p < 0.05$). Pain/discomfort and anxiety/depression components of the EQ-5D index improved significantly at 6 months and 1 year (anxiety/depression through 3 years, $p < 0.05$).
Conclusion: In addition to the positive haemodynamic effects, TPV leads to QOL improvement. Whether these results support indication of TPV at an earlier stage of valve dysfunction needs to be discussed.

1059: TRANSCATHETER CLOSURE OF PDA USING AMPLATZER DUCT OCCLUDERS: A SINGLE CENTRE EXPERIENCE

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In this report we review initial and 4-year results following transcatheter occlusion of patent ductus arteriosus (PDA) using Amplatzer duct occluders (ADO). Between November 2004 and May 2012, 91 patients, ages 3.5 months to 66 years, underwent transcatheter closure using the ADO and ADO II. Mean pulmonary artery pressure was 25.4 ± 8.6 mmHg (median 23 mmHg, 12–60 mmHg); 51 were greater than 25 mmHg. The mean PDA diameter (at the pulmonary end) was 3.2 ± 1.2 mm (range 1.6–7.5 mm), the PDA length was 6.9 ± 3 mm (median 6, range 3–16 mm) and the mean ampulla diameter (at the aortic end) was 10 ± 3.9 mm (median 9, range 4–23 mm). For closure of PDA, 130 ADO and 41 ADO II and 14 ADO AS were used. Occlusion of femoral artery developed in only one patient and there were no other complications in early period. Complete closure was observed in 56/57 (98.2%) in ADO patients, 19/20 (95%) in ADO II patients and 14/14 (100%) in ADO II AS patients at the end of the 6th month after closure.
Conclusion: All ADO types can be used safely and effectively in treatment of PDA. ADO II and ADO II AS have many advantages in small infants and short ductus. These devices can be deployed completely in the ductal body without any obstruction because of their special configuration. They have smaller sheath sizes and a flexible, user-friendly delivery system.

1066: EXTRACARDIAC FINDINGS IN FOETAL LATERALITY DISTURBANCE - THE UNUSUAL SUSPECTS

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Background: In prenatal congenital heart disease, the group with laterality disturbance are most likely to have an associated extracardiac abnormality (ECA). ECAs are multi-system, vary in severity and may be difficult to detect prenatally. We report a large series of prenatal cases with laterality disturbance and describe their associated extracardiac malformations.
Materials and methods: We performed a retrospective search of our foetal cardiology database from 1985 to 2010 for cases of laterality disturbance. Post-mortem (PM) and postnatal notes were reviewed to document ECA.
Results: A total of 255 cases of laterality disturbance were identified. Three cases were excluded as outcome data was not available. Of the 252 cases, 120 resulted in termination of pregnancy (TOP), 16 intrauterine deaths occurred and 54 patients were still alive. PM reports were available in 86 (72%) cases following TOP. Common findings were bowel malrotation (84 cases, 33%), polysplenia (62 cases, 25%) and asplenia (60 cases, 24%). An additional 43 patients had other ECAs. Of those live born, 23 (20%) patients underwent 29 non-cardiac surgical procedures including 17 Ladd procedures, 2 Kasai procedures for biliary atresia and 2 congenital diaphragmatic hernia (CDH) repairs. Four patients died as a direct result of ECA. Three had significant respiratory compromise as a result of CDH or major airway abnormality. One patient died following surgery for volvulus.
Conclusions: Intra-cardiac pathology is the predominant cause of death in the foetus diagnosed with laterality disturbance. The ECAs influence survival in a minority of cases, though they may have a significant impact on the quality of life of survivors. Particular attention should be made to discussing potential ECA when laterality disturbance is diagnosed.

1077: USEFULNESS OF LONG-TERM ADMINISTRATION OF HEPARIN IN THE RECANALISATION OF FEMORAL ARTERIES IN YOUNG CHILDREN AFTER PERCUTANEOUS PROCEDURES

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Introduction: As a result of interventional and diagnostic procedures performed in young children because of congenital heart defects some vascular complications can appear. The obstruction of the femoral artery is the most common problem.
Material and methods: Between March 2010 and July 2012 we performed 176 heart catheterisations. Among these were 95 children under 3 years of age who underwent catheterisation through the femoral artery access. The presence of arterial pulse on lower extremities was examined by palpation in all patients. In cases of doubt, or lack of pulse, ultrasound vascular flow was performed. In 6 patients, aged 20.5 ± 9.7 months (0.2–30) weighing 9.2 ± 4.0 kg (33–15), there was no flow through the punctured artery. The consulting vascular surgeon found no need for surgical treatment in all cases.
Results: All 6 patients received continuous infusion of heparin. When the absence of flow was still observed we started the treatment with low molecular weight heparin. As a result, after 19.7 ± 10.5 days (6–40) of intake of heparin, in 5 patients complete vessel patency returned. We have no data about the last patient because he was transferred to another department.
Conclusions: Performing heart catheterisation through the femoral

artery access in young children is associated with an increased probability of its closure or obstruction. Long-term administration of heparin is associated, in most cases, with a total arterial recanalisation.

1086: PERCUTANEOUS CLOSURE OF SHUNT FISTULAS WITH THE AMPLATZER VASCULAR PLUG IV IN PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Vascular plugs allow the interventional closure of large to medium-sized abnormal vessels, but their application is limited by the need for long sheaths or large guiding catheters. We report our experiences with the new Amplatzer Vascular Plug IV (AVP IV). This self- expanding spindle-shaped occluder is made of a nitinol wire mash and it is possible to place it through a 4 to 5 French catheter with a lumen of 0.038'.
Method and results: From October 2009 until June 2012, 14 AVP IV devices were deployed in 12 patients (age range 3 months to 48.8 years, weight 6.3–60.6 kg). Nine patients had venovenous or arteriovenous collaterals in functional univentricular hearts. One patient had pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries, one patient had a pulmonary arteriovenous fistula and one child had a large coronary artery fistula. We used AVP IV devices with diameters of 4–6 mm and 8 mm. In all 12 patients the AVP IV was implanted successfully, in 2 patients 2 different vessels were occluded with AVP IV devices and there was no occluder dislocation. In 2/14 vessels it was necessary to place additional devices in the presence of a residual shunt. Complete vessel occlusion was achieved in 7 cases; in a further 7 patients a residual shunt was present at the end of the procedure while patients were still fully heparinised. There were no complications related to the procedure.
Conclusion: Based on our experience, the AVP IV allows safe and effective occlusion of medium-sized and large abnormal vessels. It is also well suited for tortuous high-flow vessels such as coronary or pulmonary arteriovenous fistulas. In case of suboptimal positions it is possible to reposition the occluder with ease. The AVP IV represents a valuable new device in the interventional treatment of complex congenital vessel malformations.

1092: PRENATAL DIAGNOSIS OF MAJOR CONGENITAL HEART DISEASE: COMBINING UK CENTRAL CARDIAC AUDIT DATABASE AND MATERNITY AUDIT TO OBTAIN CENTRE-SPECIFIC DETECTION RATES IN 120 198 SCREENED WOMEN

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Background/hypothesis: Foetal cardiac screening standards introduced by NHS Foetal Anomaly Screening Programme for English maternity units, 2010, have no current baseline data. In contrast, prenatal diagnosis for infants undergoing transcatheter or surgical treatment is routinely collected and validated by Central Cardiac Audit Database (CCAD) (www.ccad.org.uk). A national system is vital to measure prenatal detection of major congenital heart disease (CHD).
Material and methods: A total of 120 198 unselected pregnant women were screened in three maternity units referring to one surgical centre:
A: co-located with foetal medicine unit; ready access to second opinion; training in major cardiac malformations
B: received on-site training and telemedicine support by perinatal cardiologist;
C: supported by local obstetricians with scanning expertise
We cross-referenced maternity records and CCAD to include cases unreported by obstetric screening.
Results: Of 120 198 screened women, 388 cases of CHD were documented; detection by unit (A,B,C): simple transposition of great arteries (TGA) (78%, 50%, 0%) and coarctation (82%, 20%, 13%).
Conclusions: Combining prenatal diagnosis (PD) with CCAD produced maternity hospital and lesion-specific detection rates and confirms that continued improvement in CHD detection depends on integration of health information systems to enable tracking between maternal and infant records.

1095: FOETAL LATERALITY DISTURBANCE: DO ATRIAL APPENDAGES MATTER?

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Background: Laterality disturbance in the foetus is associated with a wide spectrum of cardiac abnormalities. Many present prenatally with complex morphology where single ventricle palliation is the only management option after birth. The cumulative effect of associated lesions means many do not achieve Fontan completion long-term. We report a large prenatal series describing their associated malformations and review factors that influence their outcome.
Materials and methods: We performed a retrospective search of our foetal cardiology database from 1985 to 2010 for cases of laterality disturbance. Pre- and postnatal management and long-term outcomes were documented.
Results: A total of 255 cases of laterality disturbance were identified. Three cases were excluded as outcome data was not available. Of the 252 cases, 120 resulted in termination of pregnancy (TOP) and 16 intra-uterine death (IUD). Fifty-four patients remain alive, 20 with a Fontan circulation; 2 patients have undergone cardiac transplantation. In those receiving active treatment after birth, univentricular physiology, complete heart block, total anomalous pulmonary venous drainage and right heart outlet obstruction were independent risk factors for death or transplantation ($p < 0.05$).
Conclusions: Prognosis remains poor for those with univentricular physiology with an ongoing high risk of mortality throughout childhood. Survival in this group has not significantly changed across the study period. Detailed prenatal assessment to identify specific morphological features associated with poor prognosis may help guide foetal counselling. In addition reduced quality of life remains a serious concern for the few survivors.

1124: OUTCOMES OF AMPLATZER DUCT OCCLUDER II (ADO II) DEVICE OCCLUSION OF PATENT DUCTUS ARTERIOSUS (PDA) – THE NORTH WEST EXPERIENCE

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Background: The Amplatzer Duct Occluder II (ADOII) is a self-expanding nitinol device for closure of patent ductus arteriosus (PDA). This device is designed to produce high rates of occlusion but there has been recent concern as to device deformability and embolisation.
Objective: To evaluate our experience of using the ADOII device and document the closure rate and incidence of complications.
Method: A retrospective analysis of data of patients on whom we attempted closure of PDA using the new device from July 2008 to June 2012 were included ($n = 67$).
Results: Median (range) age was 20 (5–130) months and weight 10 (5.2–39) kg. The narrowest ductal diameter was 2.6 (1.5–4.9) mm, length of duct 7 (3.3–17) mm, and diameter of aortic ampulla 7.7 (3–13) mm. Procedure time was 48 (22–114) min and fluoroscopy time 7 (2.8–23.3) min. Krichenko classification of PDA morphology: A = 28 (42%), B = 12 (18%), C = 18 (27%), D = 2 (03%), E =

7 (10%). Devices used were 3/4 (12), 4/4 (30), 5/4 (15), 6/4 (3), 4/6 (3), 5/6 (2) and 6/6 (2). The venous approach was employed in all. Echocardiography was done on D1 and 4-6 weeks post implantation. One device was not deployed successfully. Complete angiographic closure occurred in 55, trivial contrast flow in 10 and mild flow in one patient. In 65 patients, echocardiography the next day and on follow-up scan at 6 weeks showed no residual shunts. In one patient mild flow was noted which resolved at 4 weeks. Three devices embolised, 1 occurring within 6 hours and 2 the next day. There were no procedure-related complications.
Conclusion: ADOII is a safe and effective device for PDA occlusion and has a high early closure rate and low embolisation rate.

1133: IMPROVED LONG TERM OUTCOME IN PATIENTS UNDERGOING PERCUTANEOUS PULMONARY VALVE IMPLANTATION AT A YOUNGER AGE

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Background/hypothesis: Percutaneous pulmonary valve implantation (PPVI) has become increasingly accepted as a safe, less invasive alternative to surgical pulmonary valve replacement for congenital heart disease patients with right ventricular outflow tract dysfunction. Several series have demonstrated favourable short-term outcome after PPVI. The aim of this study was to determine if reverse right ventricle (RV) remodelling following PPVI is persistent in the long term and whether earlier timing of PPVI may be favourable.
Methods: PPVI patients from Hospital for Sick Children (HSC) and Toronto General Hospital (TGH) were studied. Cardiac magnetic resonance imaging (MRI), echocardiography, metabolic exercise testing, and haemodynamics prior to intervention were compared to repeat measures on follow-up, using paired *t* tests and linear regression models assessing changes over time.
Results: Fifty-one patients were followed for up to 6.6 (4.2 ± 1.9) years after PPVI, including 35 HSC patients, averaging 14.8 ± 8.9 years of age at intervention and 16 TGH patients, 32.0 ± 2.1 years old at implantation. Freedom from re-intervention was 87% and 68% at 3 and 5 years, respectively, and 92% from re-operation at 5 years. Younger age at implantation was associated with an increase of 4.17 ± 1.29%/10 years of age in echo left ventricle ejection fraction (LVEF) ($p = 0.001$), 0.23 ± 0.10 points (on a 3-point scale) per 10 years in qualitative RV function ($p = 0.03$), 2.68 ± 0.90 ml/kg/min per 10 years in max VO2 ($p = 0.003$) and a decrease of 0.81 ± 0.19 cm per decade in RVED ($p < 0.001$). Preparing the conduit for implantation (prestenting) yielded an increase of 5.32 ± 1.95% in LVEF ($p = 0.006$) and 0.63 ± 0.18 points in RV function ($p < 0.001$) as well as a decrease of 0.80 ± 0.30 cm in RVED ($p = 0.007$) and 8.77 ± 3.39 mmHg in RV systolic pressure ($p = 0.01$).
Conclusions: This is the largest series to show that PPVI at a younger age has improved long-term outcomes. Strategies to preserve right ventricular function should be considered in management planning for this population.

1154: TRANSCATHETER CLOSURE OF PERSISTENT DUCTUS ARTERIOSUS AT CIPTO MANGUNKUSUMO HOSPITAL JAKARTA: CLINICAL CHARACTERISTIC AND OUTCOME

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Background: Transcatheter closure of persistent ductus arteriosus (PDA), as alternative to PDA ligation, has shorter hospital stays, fewer major complications, and comparable success rates. Studies suggest a success rate of 90% to 99% for transcatheter closure of PDA, with the higher success rates being associated with newer devices.
Methods: A descriptive analysis was done in 285 consecutive patients with diagnosis of small to moderate PDA (279 patients) and residual PDA after PDA ligation (6 patients) from January 2002 to July 2012 who underwent transcatheter closure of PDA. Data were collected from medical records and analysed by descriptive statistics.
Results: From January 2002 to July 2012, 285 patients had transcatheter closure of PDA. The median of age was 3.5 years (40 days to 37 years). One hundred and ninety-seven of the patients were female (69.1%) and median weight was 12 (3.6–59) kg. Median size of PDA was 3.6 (1–15.6) mm. Amplatzer Ductal Occluder I (ADO I) was used in 259 (90.9%) patients; median size of ADO I device was 6/8 (4/6–14/16) mm. Other devices were Gianturco coil (5.6%), ADO II (1.8%), pfm nit coil (1.1%), and Amplatzer septal occluder (0.4%). The median of flow ratio was 1.8 (0.6–14.5), fluoroscopy time was 15.4 (2.3–80) minutes, and procedure time was 76 (30–198) minutes. Median length of hospital stay was 3 (3–4) days. Overall success rate was 99.3% and 98.2% of patients had no major complications. Transient bradycardia occurred in 3 patients and migration of device in 2 patients. Evaluation of echocardiography showed most patients had complete closure of PDA. Almost all of them were discharged one day after procedure.
Conclusions: Transcatheter closure of PDA is a safe and effective alternative to surgery. This procedure has a high success rate, short length of stay in hospital, and minimal complications.

1181: FOETAL CARDIAC DISEASE AND FOETAL LUNG VOLUME: AN IN UTERO MRI INVESTIGATION

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Background: Foetal magnetic resonance imaging (MRI) has become a good non-invasive tool to study foetal lung volumes after 18 gestational weeks *in vivo*. In foetuses with congenital heart disease (CHD) proper lung function is essential for postnatal survival. Pulmonary hypoplasia is associated with high morbidity and mortality. Antenatal detection of abnormal pulmonary development may help to optimise pre- and perinatal management of the foetuses at risk. The aim of our study was to investigate foetal lung volume in foetuses with prenatally diagnosed heart disease.
Material and methods: A cross-sectional retrospective study of 105 consecutive singleton pregnancies that underwent MRI was performed. The heart defects were divided into 4 groups and compared within the groups, and also with a normal collective. To compare the lung volumes we calculated z-scores (normal range: z-score -2 to +2). Our focus of interest was the lung volume of foetuses with heart disease, compared to healthy foetuses and to calculate z-scores for these foetuses.
Results: Foetuses with CHD have significantly smaller lung volumes compared to healthy foetuses corrected by gestational age ($p = 0.049$). No difference was found within the specific groups. Z-scores: 18/105 foetuses had lung volumes with a z-score < -2.
Conclusion: Foetuses with CHD show similar lung volume within different types. Our data might indicate that postpartum pulmonary symptoms in neonates with CHD may have contributed more to the lower lung volume than to the cardiac disease itself.

1184: PRIMARY STENT THERAPY FOR SHUNT MALFUNCTION IN INFANTS

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Background: Stent therapy (ST) for BT shunt malfunction (SM) is a viable alternative to surgical shunt revision. In our institution, ST is the preferred method of treatment for SM.
Methods: A retrospective analysis of all infants with suspected SM was performed including demographics, catheterisation findings, intervention type, survival to second operation and evidence of bacteraemia.
Results: Seventeen patients (pts) presented with SM (s/p Norwood (5), Damus-Kaye-Stansel/shunt (12)) with a median age and weight of 31 days (7d - 2 yrs) and 3.2 kgs (2.6 - 14.2) kg. Five pts presented to the lab requiring mechanical cardiopulmonary support. Primary ST was attempted in 15/17 pts with technical success in all 15 pts. One pt underwent primary angioplasty and one patient arrested during the diagnostic part of the case and underwent emergent surgical revision. Both of these patients survived to second operation. Of 15 stented pts, 4 (26%) died prior to discharge; 2 arrived at the lab requiring mechanical support, and 2 had pre-existing bacteraemia. All remaining ST patients (11/15 (73%)) survived to second operation, resulting in overall survival to second operation of 77% (13/17). Aetiology of SM included discrete stenosis (12), thrombus (3), or a diffusely small shunt (2). Stenosis at the distal shunt anastomosis occurred in 10/12 patients, 3 of whom also had evidence of thrombus within the shunt. Two patients with diffusely small shunts underwent successful primary stenting with enlargement of overall shunt diameter, both surviving to second operation.
Conclusions: A programmatic approach of primary stent therapy for SM in infants has a high technical success rate and acceptable survival to second planned operation. Discrete stenosis most often occurs at the distal anastomosis and may be associated with thrombosis. Pre-existing bacteraemia and/or requirement for mechanical circulatory support prior to ST were associated with high mortality in this small series.

1187: FOETAL SUPRAVENTRICULAR TACHYCARDIA (SVT): COMPARISON OF TWO DRUG TREATMENT PROTOCOLS

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Background: The best treatment for sustained foetal SVT with 1:1 atrioventricular relationship is not known. We compared 2 treatment protocols.
Methods: A total of 155 consecutive foetuses with supraventricular tachyarrhythmia presented from 2000 to 2012; 127 had SVT with 1:1 atrioventricular conduction, and 86 received drug treatment according to one of two protocols: first-line maternal intravenous digoxin (n = 52, centre 2), or maternal oral flecainide (n = 34, centre 1). Treatment success was defined as live birth after conversion to sinus rhythm, or rate reduction by >15%.
Results: Short ventriculo-atrial (VA) interval occurred in 69 and long VA in 17. Median age at start of treatment start was 31 weeks' gestation in each centre. Hydrops was present in 30/86 (35%). Digoxin was successful in 23/28 (82%) and flecainide in 26/27 (96%, p = 0.19) of non-hydropic fetuses, compared to 8/21 (38%) and 6/7 (86%, p = 0.07) respectively when hydrops was present. For short VA SVT, conversion to sinus rhythm and rate control was 31/44 (70%) and 0/44 for digoxin, and 23/25 (92%) and 1/25 (cumulative 96%, p = 0.01) for flecainide. For long VA SVT, conversion to sinus rhythm and rate control was 4/8 (50%) and 0/8 for digoxin, and 5/9 (55%) and 2/9 (cumulative 78%, p = 0.3) for flecainide. Second-line drug treatment was added to digoxin in 19/52 (37%), and to flecainide in 2/34 (6%, p = 0.002).

Intrauterine or neonatal death occurred in 9/21 (43%, including 1 termination) hydropic foetuses treated with digoxin compared to 0/9 (p = 0.03) of those treated with flecainide.
Conclusions: Flecainide was more effective than digoxin in short VA SVT, especially when hydrops was present. Additional treatment was used more often in the digoxin protocol. No adverse foetal outcomes were attributed to flecainide.

1188: FOETAL ATRIAL FLUTTER: COMPARISON OF DRUG TREATMENT PROTOCOLS

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Background: The best treatment for sustained foetal AFl, defined as regular atrial rate >200 bpm and faster than the simultaneous ventricular rate, is not known. We compared drug treatment protocols.
Methods: Of 155 consecutive foetuses with supraventricular arrhythmia, presenting from 2000 to 2012, 28 had AFl; 25 received drug treatment according to one of two drug protocols: digoxin ± sotalol/other drug (n = 16, centre 2), or digoxin ± flecainide (n = 9, centre 1). Treatment success was defined as conversion to sinus rhythm.
Results: Hydrops was present in 7/16 (44%) in centre 2, and in 1/9 (11%, p 0.18) in centre 1. Median age at treatment was 32 weeks' gestation in centre 2 and 31.4 weeks' gestation in centre 1 (p = 0.96). In non-hydropic fetuses, sinus rhythm occurred with digoxin monotherapy in 7/17 (41%), with digoxin + flecainide in 1/17 (cumulative 47%), and with digoxin + sotalol in 1/17 (cumulative 53%). In hydropic foetuses, sinus rhythm occurred with digoxin monotherapy in 3/8 (38%), with digoxin + sotalol in 2/8 (cumulative 63%), digoxin + sotalol + amiodarone in 1/8 (cumulative 75%), and with digoxin + propranolol in 1/8 (cumulative 88%). Hydrops was present in 1/9 (11%) of foetuses with sustained AFl compared to 7/16 (44) of those converting to sinus rhythm (p = 0.18). Intrauterine death did not occur (0/8 hydropic foetuses, 95% confidence interval (CI) 0–0.37). Gestation at delivery was median 38 (range 31–39) weeks' gestation.
Conclusions: Surprisingly, hydrops did not reduce the likelihood of conversion to sinus rhythm, raising the possibility that the natural history of foetal AFl may be more important than a drug effect. Hydrops was also well tolerated *in utero* in comparison to foetal supraventricular tachycardia with 1:1 atrioventricular relationship, possibly because of later onset of AFl and more favourable atrioventricular coupling.

1196: TRANSCATHETER CLOSURE WITH USE OF THE CARDIO-O-FIX OCCLUDERS IN 153 PATIENTS WITH CONGENITAL AND STRUCTURAL HEART DEFECTS.

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Introduction: Transcatheter closure of different congenital heart disease (CHD) with the use of nitinol wire mesh occluders has been widely accepted as preferred treatment; however, the high cost of these devices limits their clinical application in many countries. Few clinical data are available regarding lower-cost products.
Aim. We evaluated the efficacy and safety of CARDIO-O-FIX occluders (COF) type: patent ductus arteriosus (PDA), atrial septal defect (ASD),and patent foramen ovale (PFO).
Materials and method*:* From September 2009 to August 2012 a total of 153 patients (pts) with congenital and structural heart defects (ages 0.5–72 years) underwent transcatheter closure with use of different COF occluders: 63 pts - PFO, 36 - ASD, 47 - PDA ,6 - ruptured sinus of Valsalva aneurysm (SOVA) and one coronary artery fistula to right ventricle (CAF). Patients with PFO had cryptogenic

stoke in anamnesis and confirmed right-to-left shunt through PFO. Mean diameter of ASD was 21 mm, PDA 4.1 mm, SOVA 10.0 mm and CAF 6 mm.
Results*:* All procedures were successful. In one PFO case COF was withdrawn because of the unusual position of the device caused by long tunnel (27 mm) of PFO; application of ASD COF was efficient in this case. In all cases complete closure of undesirable connections was achieved without any complications at follow-up (1–35 months). In ASD mean diameter of applied devices was 20 (10–28 mm), for PFO mostly 25/18 mm devices were used, for PDA devices 4/6 to 10/12, for SOVA PDA COF 10/12 to 16/18 and for CAF PDA COF 8/10 mm
Conclusions*:* These results suggest that the COF are safe and effective devices for the transcatheter closure of CHD. For confirmation a randomised controlled trial with more patients and longer follow-up is warranted.

1205: MID-TERM FOLLOW-UP RESULTS OF TRANSCATHETER TREATMENT IN PATIENTS WITH UNROOFED CORONARY SINUS

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Objectives: We present the mid-term results of transcatheter treatment of unroofed coronary sinus (CS) using the Amplatzer septal occluder.
Background: The unroofed CS is a rare atrial septal defect connecting the left atrium and CS. Surgery has been the mainstay of treatment.
Methods: In a 4.5-year period, 9 patients (5 males) with ages ranging from 26 to 69 years (median 39 years) diagnosed with an unroofed CS but without a persistent left superior vena cava (PLSVC) underwent transcatheter treatment. Computed tomography (CT) was performed in 8 patients. Transoesophageal echocardiography (TEE) was used to monitor the procedure.
Results: The mean Qp/Qs ratio was 2.4 ± 1 and mean systolic pulmonary artery pressure was 35 ± 19 mmHg. An Amplatzer septal occluder was deployed in all 9 patients. The device was implanted in the defect in 1 patient and at the CS ostium in the other 8 patients. The median device size used was 22 mm (16–28 mm). The left disc herniated into the CS in the single patient in whom the device was implanted within the defect. All patients were available for the 3-month follow-up. None had a residual shunt on the 3-month follow-up echocardiography. One patient died of a stroke 4.5 months after the procedure. At mean follow-up 42.6 ± 18.3 months, symptomatic improvement was documented in the remaining 8 patients. All 8 patients had an O_2 saturation above 96 %.
Conclusions: Transcatheter treatment for unroofed CS without PLSVC using the Amplatzer septal occluder is safe and feasible.

1213: ASSESSMENT OF THE DUCTUS ARTERIOSUS IN FOETUSES WITH RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION AND VENTRICULAR SEPTAL DEFECT

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Objectives: To describe the ductus arteriosus (DA) in foetuses with right ventricular outflow tract obstruction (RVOTO) and anteriorly malaligned ventricular septal defect (VSD).
Background: Foetuses with RVOTO may develop significant cyanosis after birth. Prenatal diagnosis may improve the early postnatal management approach. Although prenatal detection of RVOTO has already been reported, there is lack of information on ductal morphology and ductal blood flow pattern, during pregnancy and after birth.
Patients and methods: The department registry was retrospectively searched for all patients born between January 2006 and June 2012 with a prenatal diagnosis of RVOTO and VSD. RVOTO was considered severe if pulmonary artery/aortic diameter ratio was ≤ 0.5 and pulmonary circulation was ductus dependent. Foetuses with pulmonary atresia or absent pulmonary valve syndrome were excluded.
Results: Out of 50 foetuses, 33 had tetralogy of Fallot (TOF) (66%) and 17 (34%) had double outlet right ventricle with subaortic VSD. Five (10%) foetuses had severe RVOTO with reverse flow across the DA; parents opted for termination of pregnancy (TOP) in 2 cases and the other 3 liveborn infants underwent modified Blalock Taussing (mBT) shunt. Thirty-three (66%) foetuses had mild RVOTO with normal ductal morphology and flow pattern; there were 9 TOP and 2 neonatal deaths for extracardiac reasons. Twelve (24%) more foetuses had mild to moderate RVOTO but very small ductus with changeable flow pattern, spontaneously closed immediately after birth. Two of them developed severe hypoxic spells respectively at birth and during the first month of life and both had mBT shunt.
Conclusion: In less severe degrees of RVOTO, flow across the DA would be reduced and ductus diameter would be decreased even up to prenatal closure. Ductal morphology assessment may be useful for improving management of neonates with prenatal mild to moderate RVOTO and small DA who may become cyanotic at birth.

1221: THE AMPLATZER DUCT OCCLUDER II ADDITIONAL SIZES DEVICE FOR TRANSCATHETER PATENT DUCTUS ARTERIOSUS CLOSURE: INITIAL EXPERIENCE

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Background: Effective and safe patent ductus arteriosus (PDA) transcatheter closure requires a device suited to the specific ductal anatomy which is appropriate for the patient size. The new Amplatzer Duct Occluder II Additional Sizes (ADOIIAS) device is designed with a range of diameters (3–5mm) and lengths (2–6mm) and small disks to avoid flow disturbance in the aorta and pulmonary artery.
Materials and methods: Population: All patients who underwent attempted closure with an ADOIIAS device. Following aortography the PDA was closed using a 4-5 Fr delivery system from the pulmonary or aortic approach with an ADOIIAS of waist ≥ 1.5 times the narrowest part of the PDA. Aortography to confirm position and leak was performed before and after device release. Echocardiography was performed the following day.
Results: Between June 2011 and August 2012, 45 consecutive patients (25 female), median age 3.3 years (0.6–15.8) and weight 14.5kg (4–79), underwent PDA closure with an ADOIIAS device. In 42/45 (93.3%) a device was successfully deployed (38 aortic). At echocardiography the next day 41/42 (97.6%) of PDA were closed; in 1 case there was a tiny residual leak. In 3 patients the ADOIIAS was unstable and an alternative device was implanted. The mean ratio of ADOIIAS:PDA diameter was 2.4 (± 0.52):1. There were no complications.
Conclusions: We present a large initial clinical experience with the new ADOIIAS device which is easily implanted from the aortic side and is safe and effective for PDA closure. The variety of sizes affords intuitive suiting of the device to the specific PDA anatomy.

1224: IDENTIFYING PREDICTORS OF MITRAL VALVE (MV) TEARS RESULTING FROM PERCUTANEOUS BALLOON MITRAL VALVULOPLASTY (PBMV): A DESCRIPTIVE STUDY BASED AT TYGERBERG ACADEMIC HOSPITAL

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Background/hypothesis: Despite selection and procedural refinement for PBMV, the rate of catastrophic mitral regurgitation (MR) from mitral valve (MV) tears remains fixed at ±5%, reflecting our

inability to predict this complication. Valve tears not related to catastrophic MR are incompletely studied. The mechanisms underlying MV tears are presumably common to both these scenarios, creating an opportunity to study predictors of MV tears in this cohort.
Materials and methods: Forty-five consecutive patients undergoing PBMV (Inoue-technique) with pre- and post-PBMV digital echocardiographic images available were assessed. Patients with a clearly identifiable tear of the mitral leaflets or apparatus (MVA) were identified. The mechanism and severity of MR pre- and post PBMV were evaluated. We recorded detailed morphological descriptions of each case; including Massachusetts General Hospital score (MGHS), parameters quantifying subvalvular involvement, detailed leaflet edge and commissural assessment and tear location.
Results: Twelve patients (26.7%) developed clearly identifiable tears of the MVA; 3 patients (6.7%) had catastrophic tears necessitating early surgery. All patients developed moderate or greater MR from their tears. The mean MGHS was 9.5 with subvalvular thickening (3.5) and calcification (2.33) contributing most. Tear location: anterior leaflet 6; posterior leaflet 5; subvalvular 1. Nine of 11 leaflet tears were located in a thin, calcium-free area of the leaflet adjacent to an area of significant calcification. Six of 7 tears that occurred laterally (not centrally) were located on the side of the most severe subvalvular involvement.
Conclusions: Non-catastrophic MVA tears related to PBMV are more common than previously thought. Longitudinal follow-up of this cohort is required to evaluate outcomes in patients who develop non-catastrophic tears post-PBMV. An interesting interplay appears to exist between the morphology at the edge of the MV leaflets, where tears presumably start, and factors that restrict free movement of leaflet segments. This is being assessed in an ongoing study.

1229: FOETAL BRADYCARDIA AND SINUS NODE DYSFUNCTION

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Introduction: Sinus node dysfunction is a common condition in elderly patients or those with postoperative congenital heart disease. It is characterised by an abnormality of cardiac impulse formation. In this report I present a rare case of foetal bradycardia and sinus node dysfunction.
Case description: A 26-year-old pregnant patient was referred because of persistent foetal bradycardia after obstetric ultrasonography. The 23 weeks' gestation echocardiogram showed sinus bradycardia (heart rate (HR) 80–87 bpm) with normal conduction time from the atria to the ventricles (1:1) and normal anatomy. Follow-up echocardiograms performed at 29, 32, 35 and 38 weeks' gestation showed profound bradycardia (HR 53–62 bpm) with no hydropsy or blocked atrial extrasystoles. Maternal antibody titres for SS-A/SS-B and the screen for QT long syndrome were negative. A female infant was born by caesarean section at 39 weeks' gestation and had Apgar scores of 8/8. After birth, the baby developed profound bradycardia (HR 50 bpm), poor perfusion, and signs of shock. Atropine and isoproterenol were administered intravenously to the patient with no significant improvement in HR (55–60 bpm). The initial ECG showed junctional rhythm at 55 bpm, normal QTc (0.42 s), atrial extrasystoles, and variable second-degree atrioventricular (AV) block. A 24-hour ECG and oesophageal ECG allowed the diagnosis of sinus node dysfunction to be confirmed. The echocardiogram demonstrated two small ostium secundum atrial septal defects. Possible sinus bradycardia causes were dismissed, thus confirming the above diagnosis. The baby developed oliguria and heart failure at which time the implantation of a pacemaker was indicated. The surgery was successful, and the patient is currently growing and thriving at 24 months of age.
Conclusion: The author reports the importance of foetal echocardiography for the diagnosis and appropriate therapy approach to avoid complications.

1233: SUCCESSFUL CONSERVATIVE TREATMENT FOR HAEMOLYSIS AFTER TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECT (VSD) USING NIT-OCCLUD® LÊ VSD OCCLUDE

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Background: We report a haemolysis-associated transcatheter VSD closure using Nit-Occlud® Lê VSD occluder (LVO) that led to transient renal failure and recovery by conservative treatment.
Case: A 1-year-old (8 kg) boy with an underlying disease of cri du chat syndrome underwent transcatheter VSD closure. An LV angiogram revealed a large perimembranous VSD with a tricuspid pouch (11 mm). Initially, a Cocoon Duct Occluder (CDO) (12/14) was placed, resulting in mild aortic regurgitation. A CDO (6/8) was placed instead but resulted in a residual moderate shunt. When an LVO coil (12/6) was used instead of the CDO it showed another RV exit (5 mm). We placed a second LVO coil (8/6) into that exit. A repeat LV angiogram revealed a trivial residual shunt. Unfortunately the LVO coil (12/6) became stuck during deployment, but it was retrieved and another LVO coil (10/6) was placed instead. After the 4-hour procedure, the result was a mild residual shunt. Unfortunately, the patient did not gain full consciousness when weaned off anaesthesia. An emergency craniotomy and clot removal was done after a competd tomography (CT) brain scan showed a large subdural haematoma. In the intensive care unit (ICU) his dark brown urine suggested haemolysis, which was confirmed by urinalysis. Because of the complicated procedure, we avoided unnecessary intervention. Our strategy for the haemolysis was aspirin cessation, IV hydration, alkalinisation of urine and to maintain the haemoglobin >12 g%. His baseline blood, urea, nitrogen (BUN)/creatinine was 14.1/0.32 mg%, which increased slowly in the first 10 days then rose rapidly to 31.2/0.57 mg% within 1 day after the IV fluid was unintentionally withdrawn. We re-administered our strategy plus dexamethasone. The next day his BUN/Cr dramatically declined to 17/0.35 mg%. Gradually his renal functions resolved within 4 days.
Conclusion: In some circumstances, haemolysis can be overcome by conservative treatment to avoid redoing percutaneous or surgical closure.

1255: NORMAL GROWTH OF FOETAL CARDIOTHORACIC STRUCTURES DURING THE LATE FIRST/ EARLY SECOND TRIMESTERS

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Background/hypothesis: Foetal echocardiography is used increasingly early in pregnancy to screen for congenital heart disease (CHD). Data are lacking for normal growth of the foetal heart in the late first/early second trimester. Foetal weight increases in a non-linear fashion and we hypothesise that growth of cardiothoracic structures is also non-linear.
Materials and methods: A total of 197 healthy pregnant women were prospectively recruited to have detailed echocardiography between 8 and 14 + 6 weeks' gestation. Atrial, ventricular, heart, chest, pulmonary artery and ascending aorta dimensions were measured. Statistical analysis included ANOVA.
Results: There was non-linear growth of all measured structures, with faster growth after 12 weeks' gestation. From 8 to 10 + 6 weeks' gestation, 95% (20/21) had a pericardial effusion. Heart and atrial size relative to chest size decreases significantly over time. There is

no difference between right versus left ventricle and main pulmonary artery versus ascending aorta dimensions at any stage (p >0.05).
Conclusions: We have documented growth of cardiothoracic structures during the late first/early second trimesters and established normal values. Pericardial effusion is almost universal in early pregnancy. The relatively large heart in the late-first trimester may relate to atrial size, possibly reflecting the importance of atrial function to filling the non-compliant early foetal ventricles. These data provide insight into cardiac development and should assist in early diagnosis of CHD.

1278: ANTENATAL FOETAL CARDIAC SCREENING: USE OF OUTFLOW TRACT VIEWS IMPROVES DETECTION OF TRANSPOSITION OF GREAT ARTERIES (TGA) AND TRUNCUS ARTERIOSUS (TA): EXPERIENCE FROM TWO REGIONAL FOETAL CARDIAC UNITS AND A SINGLE PAEDIATRIC TERTIARY CARDIAC CENTRE.

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Introduction: Outflow tract assessment of foetal hearts was introduced in the UK in 2008. We compared detection of transposition of the great arteries (TGA) and truncus arteriosus (TA) in our region before and after the introduction of the new recommendation.
Methods: We undertook a retrospective review of foetal medicine and cardiac database at two regional foetal cardiac units and a single paediatric tertiary cardiac centre between January 2001 and June 2012. The detection rates for TGA and TA in babies scanned between 2001 and 2008 (before the introduction of the mandatory national outflow tract guidelines) and after 2008 were compared. A survey of practice at the referring obstetric units was also performed.
Results: The antenatal detection percentage before mandatory outflow tract guidelines (Jan 2001 – Dec 2008) was 12.5% for TGA and 17.9% for TA, whereas after mandatory outflow tract guidelines (Jan 2009 – June 2012), percentages were 32.9% and 50% respectively.Prior to the mandatory national guidelines, only 15/27 obstetric units within the region performed outflow tract views for foetal cardiac screening. Now all obstetric units routinely perform 4 chamber as well as outflow tract views.
Conclusions: There is significant improvement of antenatal cardiac detection of outflow tract abnormalities with introduction of the mandatory outflow tract views. It is hoped that as the screening sonographers get more used to the outflow tract views, and with more dedicated training of sonographers, the detection rate will increase further.

1289: ASYMMETRIC RIGHT CARDIAC CHAMBERS ENLARGEMENT IN FETUSES: A RESPONSE TO FOETAL HYPOXIA AFTER THE 30TH WEEK?

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Introduction: Foetal hypoxia (FH), with or without intrauterine growth restriction (IUGR) evaluated by Doppler ultrasound (DU), was defined by measuring the ratios between the index of the middle cerebral (MCA) and umbilical arteries (UA) (Rc/u) < 1. The lack of symmetry between the cardiac chambers in favour of the right chambers (relation right atrium(RA)/left atrium(LA) and right ventricle(RV)/left ventricle (LV) >1.5:1) not detected before the 30th week of gestational age (GA) without cardiac malformation (CM) could indicate the presence of a functional anomaly.
Objectives: The asymmetric right predominant cardiac chambers (ARPCC) manifested after week 30 could be an indicator of FH. This does not occur before 30 weeks. We discuss the possible physiopathology.
Materials and methods: Fifteen foetuses (f) were referred for ARPCC between week 30 and 37(mean 32.5) and 10 f between week 26 and 29 (mean 27.1) with an Rc/u < 1. A complete echocardiogram and DU in the UA, MCA, aortic isthmus and A wave of the venous duct were performed.
Results: Of the 15 f, 4 (26.6%) had a weight above percentile 50 (P50), 4 f had P30 and 7 f (46.6%) had less than P5. All of them presented with ARPCC >1.5:1 and Rc/u < 1. The flow at the aortic isthmus was reversed in all. The A wave was reversed in 8 f (100%) without IUGR and 4/7 (57.1%) less than P5. Of 7f with IUGR, 6 (85,7%) presented as a T21. In 1 foetus coronary flow was increased. Of the 10 f of < 30 weeks with a Rc/u index < 1, none presented with ACCD.
Conclusion: After week 30, ARPCC without CM could indicate severe FH for which foetal DU should be performed. ARPCC associated with RCIU < P5 can indicate risk of associated T21. ARPCC apparently does not present in foetuses less 30th week with Rc/u< 1.

1307: THE ABSENCE OF PHYSIOLOGICAL SHUNTS DURING THE FOETAL PERIOD CAN HELP PREDICT SEVERE POST-NATAL HYPOXIA IN FETUSES WITH TRANSPOSITION OF THE GREAT ARTERIES WITH INTACT VENTRICULAR SEPTUM

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Background: Transposition of the great arteries with intact ventricular septum (TGA-IVS) is amenable to complete repair with low mortality rate. However, some neonates may experience profound cyanosis leading to rapid haemodynamic compromise.
Objective: We evaluated whether the assessment of physiological shunts during the foetal period could help predict postnatal profound hypoxia in neonates with TGA-IVS.
Methods: Echocardiographic data of 35 foetuses with TGA-IVS were retrospectively reviewed. The size of the foramen ovale (FO), septum primum (SP), main pulmonary artery (MPA) and aorta were measured. Doppler characteristics and output in the MPA, aorta and ductus arteriosus (DA) were assessed. The net pulmonary output was calculated as [output in the MPA – output in the DA]. Patients were divided into 2 groups based on postnatal saturation: group 1 had peripheral saturation < 50% and group 2 had saturation ≥50%.
Results: Eleven of the 35 foetuses (31.4%) were in group 1. Foetuses in group 1, in comparison with group 2, had smaller FO (2.93 vs 4.07 mm, p = 0.02). In addition, holo-diastolic intermittent or persistent retrograde flow in the DA was observed in 10 of the 11 patients (91%) in group 1 in contrast to only 6 of the 24 patients (25%) in group 2 (p < 0.01). Patients with decreased mobility of the SP or a thick septum experienced higher output in the DA (0.71 vs 0.32 ml/min, p = 0.01). There was a positive linear correlation between the size of the fossa ovalis and the net pulmonary output (r = 0.54, p = 0.004).
Conclusion: A decrease in the size of the FO with retrograde holodiastolic flow in the DA is predictive of severe postnatal hypoxia in foetuses with TGA-IVS. Patients with smaller FO experience a lower net pulmonary flow with higher output through the DA, possibly related to greater pulmonary hypertension, and should be flagged for a Rashkind immediately after birth.

1310: UHL'S ANOMALY: A DIFFICULT PRENATAL DIAGNOSIS

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Introduction: Uhl's anomaly is a rare form of congenital heart disease with partial or complete absence of right ventricular (RV) myocardium, parchment-like appearance of the RV wall, often associated with tricuspid anomalies. Only 3 prenatal cases have been

reported so far. We describe a foetus with Uhl's anomaly that initially mimicked Ebstein's disease.

Case report: A 22-year-old pregnant woman was referred for suspicion of congenital heart disease at 21 weeks' gestation (WG). Initial echocardiographic evaluation concluded apical displacement of the tricuspid septal leaflet compatible with an Ebstein's anomaly, with mild RV and right atrial (RA) dilatation. The RV free wall was surprisingly thin and hypokinetic. Two additional muscular ventricular septal defects (VSDs) were also reported. There was no tricuspid regurgitation and pulmonary flow, and flow in the ductus arteriosus (DA) and ductus venosus (DV) were normal. At 27 WG, significant cardiomegaly and profound A waves in the DV were reported, with moderate tricuspid regurgitation and RA dilatation. The DA showed bidirectional flow reflecting decreased RV output. At 30 WG foetal supraventricular tachycardia at 230 bpm occurred leading to foetal hydrops. There was no antegrade pulmonary blood flow. The RV free wall was even more abnormally thin and Uhl's anomaly was suspected. Foetal demise occurred at 31 WG.

Discussion: Uhl's anomaly is a rare cause of foetal RV dilatation and can be associated with tricuspid dysplasia or pulmonary atresia. A cautious analysis of the RV free wall showing an abnormally thin myocardium and decreased myocardial thickening should help making the differential diagnosis between tricuspid disease and Uhl's anomaly. The evolution is poor with low RV output, progressive dilatation of the right heart chambers and rhythm disturbances leading to foetal demise.

1333: INTERMEDIATE AND LONG-TERM OUTCOME OF DEVICE CLOSURE OF PATENT ARTERIAL DUCT WITH SEVERE PULMONARY HYPERTENSION

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Background: A subset of patients with large patent ductus arteriosus (PDA) and severe pulmonary arterial hypertension (PAH) progresses to adulthood without established pulmonary vascular disease. Whether to close or not to close and how to close these PDAs is often a difficult decision.

Objectives: To evaluate the results of device closure of large PDA with severe PAH after 2 years of age and report their intermediate and long-term outcome.

Methods: Fifty-seven patients with large PDA and severe PAH (pulmonary artery pressure (PAP) more than 2/3rd systemic) with no desaturation were considered for closure. The reversibility of PAH was assessed with 100% O_2 and trial balloon/device occlusion. Mean age was 12.5 + 1.4 (2–55) years and mean weight 27.4 + 2.1 (9–70) kg. The follow-up was primarily based on echocardiography while 4 patients with persistent PAH underwent repeat catheterisation.

Results: Fifty patients demonstrated a decrease in PAP and device closure was successful in 47/49 (96%). Three patients underwent surgery, two after unsuccessful device closure. Seven (12.3%) patients did not demonstrate a decrease after trial occlusion. Amplatzer ductal occluder (ADO) was used in 30/49 (61.2%), MVSD device in 14 (28.6%) and Amplatzer septal occluder (ASO) in 5 (10.2%) patients. There was a significant decrease in peak systolic PAP (81 ± 4 vs 61 ± 3 mmHg, $p < 0.001$) and mean PAP (63 ± 2 vs 46 ± 2 mmHg, $p < 0.001$) following device occlusion. The device embolised to the left pulmonary artery (LPA) in one patient needing surgery. At a mean follow-up of 49.2 ± 3.7(6–121) months, there were no deaths, late embolisation or aortic protrusion. Mild LPA stenosis in 2 patients did not progress. Significant PAH has persisted in 4/50 (8%) patients.

Conclusions: Device closure of large PDA causing severe PAH with Amplatzer devices is safe and effective. Despite symptomatic improvement, PAH may not regress in 8% of the patients. In addition to more diligent non-invasive assessment, an accurate measurement of pulmonary vascular resistance, together with direct measurement of PAP after trial occlusion may help further exclusion of borderline patients.

1352: MEDIUM-TERM FOLLOW UP OF ENDOVASCULAR STENTING FOR NATIVE AND RECURRENT COARCTATION OF THE AORTA: A SINGLE CENTRE EXPERIENCE

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Aim: To describe the complete single-centre experience of transcatheter stenting of native and recurrent coarctation of the aorta (CoA).

Patients and methods: From December 2000 to October 2011 160 patients (92 male) underwent transcatheter stenting for coarctation of the aorta; 107 had native coarctation (67%) and 53 re-coarctation, 11 of whom had previously undergone balloon aortoplasty. Median age and weight at procedure were 20.5 yrs (6–65) and 63 kg (24–116) respectively. Bare metal stents were used in 131 patients and covered stents were used in 29 (14%). Four patients had more than one stent inserted during the procedure. Follow-up data were available for 129 at a median interval of 56 months (range 4 months – 10 years).

Results: Invasive peak systolic gradient decreased from mean 28 mmHg (± 13) to 5 mmHg (± 6). Coarctation site increased from mean 6 mm (± 3) to 13 mm (± 2.5). There were no deaths, 8 (5%) patients suffered procedure-related complications (1 acute pulmonary oedema, 1 para-aortic aneurysm, 1 aortic rupture, 5 embolised stents). Right arm systolic blood pressure decreased from mean of 150 ± 20 mmHg to 128 ± 16 mmHg ($p < 0.0001$). Seventy-five (47%) remain on antihypertensive medication. Echocardiographic Doppler-derived gradient decreased from mean of 58.7 mmHg (± 15.8) to 25.1 mmHg (± 14.3) ($p < 0.0001$). Eighty patients (50%) had post-procedural computed tomography (CT) angiograms at approximately 3 months. Only one aneurysm was identified which was unchanged from the post-procedure angiogram. There were no late complications.

Conclusions: Transcatheter stenting of native and recurrent CoA is a safe and effective therapeutic option in this age group with benefits maintained in the medium term. Complications tend to occur at the time of the procedure. Routine post-procedural CT angiography does not appear to be necessary and the place of covered stents has still to be established.

1356: SUCCESSFUL TRANSCATHETER CLOSURE OF A PATENT DUCTUS VENOSUS WITH AN AMPLATZER VASCULAR PLUG II IN AN INFANT

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The ductus venosus is an embryological vascular structure that connects the umbilical vein with the inferior vena cava. The ductus venosus rarely persists beyond 2 weeks after birth. If however it remains patent beyond this period and the resulting portosystemic shunt is significant, it can be an extremely rare cause of liver failure. We present a rare case of a large patent ductus venosus causing cholestatic jaundice and severe liver dysfunction in a 6-day-old neonate. Abdominal sonography demonstrated a large vascular channel adjacent to the ligamentum teres which was identified to be patent ductus venosus.

Computed tomography (CT) angiography was unsuccessful because of technical difficulties, and because of the deteriorating liver function a combined diagnostic and interventional catheterisation was planned.

At catheterisation a 6F sheath was inserted into the right internal jugular vein. Angiography demonstrated a large patent ductus venosus measuring 6 mm at its widest diameter and 26 mm in length. Balloon occlusion was performed with a wedge pressure catheter in order to measure the portal pressures once the ductus was occluded, which showed no significant increase.

The decision was then made to occlude the ductus venosus using a 12 mm Amplatzer vascular plug type II. The device was deployed with some initial occlusion to the normal hepatic venous drainage. It

was then retrieved into the sheath and redeployed with a good result. The child did well post catheterisation with the liver functions recovering to within normal limits shortly after closure.

1360: LONG-TERM OUTCOME OF BALLOON DILATATION OF VALVULAR AORTIC STENOSIS AS PRIMARY TREATMENT: THE NEED FOR REINTERVENTION

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Background: Although balloon dilation of congenital aortic stenosis has become a primary therapeutic strategy, few data are available regarding long-term outcomes.
Objectives: We evaluated the long-term (median 6.0, range 1.6–11.6 years) results of balloon dilation of severe or critical aortic valve stenosis in children in our institution where all invasive paediatric cardiology in our country is centralised.
Methods: Retrospective long-term follow-up study comprised 47 children who underwent balloon aortic valvuloplasty (BAV) between 1/2000 and 12/2010. There were 20 neonates, 16 children aged 1–12 months, and eleven aged >12 months. We assessed clinical, catheterisation and echocardiographic outcome and need for reintervention.
Results: The median (range) age at dilatation was 0.11 (0–16.8) years and weight 4.4 (2.2–84) kg. Fourteen children received prostaglandins, 66% had bicuspid aortic valve and 47% had other cardiac abnormalities. At catheterisation, median peak gradient before dilatation measured 60 (35–113) mmHg and after dilatation, 17 (3–48) mmHg ($p < 0.0001$). Valve regurgitation before dilatation was graded as none or mild in all children, and after dilatation as moderate in 7 (15%) and as none or mild in others. Mean balloon to annulus ratio measured 0.91. Complications in 15% of patients included transient pulse loss in 4, transient haematuria in 1, transient hypotension in 1, and pericardial tamponade and resuscitation in 1, leading to neurological damage and death. One child died 2.5 months after BAV, after operation for CoA. Acute and long-term mortality rates were 2% and 4%, respectively. During median follow-up of 6 years, survival was 96% and freedom from reintervention 57%. Twenty children had undergone reintervention: repeat balloon valvuloplasty in 6 (13%), and surgery in 14 (30%) including valvotomy (5), Ross operation (6), and aortic valve replacement (3).
Conclusions: In our centre, balloon dilatation of aortic stenosis has been effective with low complication and mortality rates. Reintervention is frequently needed.

1368: ACUTE DISSECTION AND PSEUDOANEURYSM WITH TRANSCATHETER PATENT ARTERIAL DUCT DEVICE OCCLUSION

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Background: Transcatheter device occlusion of patent ductus arteriosus (PDA) is a well-established and safe procedure with a high success rate. Previous reports describing pseudoaneurysms as complications with PDA devices have alluded to femoral artery pseudoaneurysms at the vascular access site. A literature review did not identify reports of acute dissection and pseudoaneurysm formation during transcatheter PDA occlusion.
Case report: A 3.1 kg 74-day-old infant with a moderate atrial septal defect (ASD), PDA and pulmonary valve stenosis was planned for transcatheter balloon pulmonary valvotomy (BPV) and PDA device closure. Pulmonary valve annulus measured 8 mm. An aortogram with a 4F vessel sizing pigtail with the end cut off showed a long and tubular PDA with a slight constriction at the pulmonary end. BPV was performed with a 9 mm X 3 cm Tyshak II balloon. A 4-4 ADO II AS device was deployed from the aortic side. The device was however freely mobile on stability testing and pushed through the duct into the main pulmonary artery (MPA) with easy retrieval of the fully deployed device back into the aorta. The delivery system and unreleased device were removed. Repeat angiography into the PDA with a cut pigtail catheter demonstrated dissection and pseudoaneurysm of the duct with 2 exit points into the MPA. Transthoracic echocardiography confirmed a tissue flap at the proximal pulmonary end of the duct and a pseudoaneurysm which prolapsed into the MPA. The infant was referred for surgical PDA ligation. Post ligation, there was no residual PDA with resolution of the pseudoaneurysm.
Conclusions: The dissection could have occurred during BPV, during retrieval of the occlusion device or with positioning the cut vessel sizing pigtail during the second aortogram. We feel the injury most likely occurred with the cut pigtail catheter. This highlights the risks associated with the sharp edges of a cut catheter.

1372: FEASIBILITY AND SAFETY OF TRANSCATHETER CLOSURE OF ATRIAL SEPTAL DEFECT IN SMALL CHILDREN WEIGHING 10 KG OR LESS

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Background: Transcatheter closure of atrial septal defects (ASD) has been accepted as a standard treatment for patients with haemodynamic significance in children and adults. Little is known about very small children and infants with poor weight gain and symptoms with congestive heart failure.
Materials and methods: From April 2004 to March 2008, 316 patients underwent transcatheter closure of ASD using Amplatzer septal occluder (ASO, Golden Valley, MN) in our institute. Among them 94 patients weighed 10 kg or less. The indication of early treatment in each group was symptoms of congestive heart failure with volume overload of the right side of the heart. We analysed the demographic data, clinical characteristics and outcome of the patients.
Results: There were 28 males and 66 females. Median age was 15 months (7–15months) and average weight was 9.1 kg (5–10 kg). Median ASD size was 15 mm (10–24 mm). Four patients were sent to surgery because of the encroaching mitral valve by left atrium (LA) disk after device placement. The procedure was successful in the rest of the patients. There was no mortality. Complete closure rates at discharge were 81.8%. Only one minor complication was noted during the procedure (transient arrhythmia). The mean hospital stay was 4.7 days.
Conclusions: Transcatheter closure of secundum ASD with the ASO is technically feasible, safe and effective even in very small children and infants of less than 10 kg. Meticulous patient selection is of critical importance to avoid undue invasive procedures in this unique group of patients.

1376: DUCTAL CLOSURE USING AMPLATZER DUCT OCCLUDER TYPE II ADDITIONAL SIZES: EARLY EXPERIENCE IN PORT ELIZABETH HOSPITAL COMPLEX, SOUTH AFRICA

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Background: Various devices have been developed for percutaneous closure of patent ductus arteriosus (PDAs) in all patients except the small infant. Such devices lead to either pulmonary artery or descending aorta obstruction.
Methods: Records of patients undergoing ductal closure were reviewed. Demographics; haemodynamic and angiographic characteristics including size, shape, length and aortic diameter, device to close the duct and closure approach, screening time, complications

and outcomes were recorded.
Results: From June 2011 to July 2012; 22 patients (11 females and 11 males) were assigned to percutaneous closure using Amplatzer Duct Occluder II Additional Sizes (ADO II AS). Mean patients' age was 9 months (range 1–60 months), and weight was 5.4 kg (1.1–12.9). The QP:Qs ratio mean was 2.2 (1.1–4.89) and pulmonary vascular resistance mean was 1.88 WU (0.12–7.12 WU). The ductal size mean was 1.9 mm (0.6–3.2). Ten patients had Krichenko Type A duct, 2 type B, 4 type C, 1 type D and 5 type E. The screening duration mean was 21.1 minutes (9–45.3). Nine patients were occluded with ADO II AS 4 x 6 mm device; 8 with 5 x 6 mm; 2 with 3 x 6 mm; 1 with 3 x 4 mm; 1 with 4 x 2 mm and 1 with 5 x 2 mm. In 19 patients the device was deployed via the pulmonary side and in 3 patients via the aortic side. In one patient, the device dislodged to the pulmonary arteries immediately following deployment, with successful retrieval. Complete ductal occlusion was achieved in all (100%) other patients (n = 21) before discharge (day one).
Conclusion: The ADO II AS is a safe and effective device for closure of small ducts even in smaller infants. There is minimal risk for aortic and pulmonary artery obstruction with the device.

1386: TRANSCUTANEOUS AORTIC VALVE REPLACEMENT: INITIAL EXPERIENCE IN A DEVELOPING COUNTRY

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Aim: Critical aortic stenosis is a common structural heart disease in the elderly and symptomatic stenosis has a mortality of 50% over 24 months. Surgery for this group carries 3–15% mortality. Transcutaneous aortic valve insertion (TAVI) is a non-invasive alternative to surgery. The aim of this study was to assess the initial experience of TAVI in a high risk aortic stenosis population of central South Africa.
Methods: Prospective study of all patients undergoing a TAVI procedure over a 12-month period. Seventeen patients with a median age of 79.6 yr (range 62.8–87.9) were included. Median aortic valve area and gradient were 0.6 cm^2 (range: 0.5–1.5) and 70 mmHg (46–120), respectively. Co-morbidities were present in all with a median Euroscore of 15 and predicted Society of Thoracic Surgeons (STS) mortality of 21.4%.
Results: Vascular access was percutaneously gained in 16 and surgically in1 patient. All valves were successfully implanted: 26 mm (6), 29 mm (7) and 31 mm (4). Procedure time ranged from 45 to 160 min. No procedural or peri-procedural deaths occurred. Aortic valve peak gradient decreased significantly to a median of 0 mmHg (range: 0–31) ($p < 0.0001$). Mean hospital stay was 3.9 ± 1.3days. Four patients (23%) required permanent pacemaker insertion. NYHA class symptomatology improved significantly ($p < 0.001$) during follow-up. There were 2 late deaths, not related to the procedure (78 & 93 days, respectively).
Conclusion: Our results show that TAVI is feasible in our population. The procedure is successful and follow-up shows that stable device position is maintained in all patients. Clinical and haemodynamic improvement could be demonstrated in all patients. Short-term mortality and morbidity show that TAVI is an acceptable alternative to conventional surgical valve replacement in this high-risk group of patients.

1407: RADIOFREQUENCY ABLATION OF MULTIFOCAL RIGHT ATRIAL TACHYCARDIA FACILITATED BY 3D MAPPING AND ROBOTIC ASSISTANCE IN A PATIENT WITH REPAIRED TETRALOGY OF FALLOT

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Background: Patients with congenital heart disease often have complex cardiac arrhythmias with extensive underlying substrate. Significant structural variations in the underlying anatomy provide further challenges to the success of ablation therapy. Cumulative radiation exposure of the operator is an added concern.
Materials and methods: A 50-year-old male patient with repaired tetralogy of Fallot presented with incessant tachycardia and heart failure. The cardiac ejection fraction (EF) was reduced at 43%. A cardiac electrophysiological study was performed and a multifocal right atrial tachycardia was identified. A detailed 3D map of an enlarged right atrium was generated using an integrated robotic sheath and RF Ablation catheter (LYNX, Hansen Medical) and Ensite Velocity (St Jude Medical). Arrhythmogenic foci were identified along the crista terminalis and around the sinus node. The sinus node was carefully tagged on the 3D map. Radiofrequency (RF) ablation was remotely performed with precision guidance of the LYNX RF ablation catheter.
Results: The arrhythmogenic foci were successfully ablated and tachycardia was rendered non-inducible. Sinus node function was unchanged post ablation. At 2-months follow-up the patient remains in stable sinus rhythm and left ventricle (LV) function has recovered completely with the EF now 60%.
Conclusion: Ablation of complex and challenging cardiac arrhythmias is greatly facilitated by 3D mapping and robotic assistance. These technologies also improve safety and reduce operator fatigue and radaition exposure.

1408: RADIOFREQENCY ABLATION OF SYMPTOMATIC PREMATURE VENTRICULAR COMPLEXES (PVCS) IN THE YOUNG PATIENT

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Background: Although often benign, premature ventricular complexes (PVCs) can cause severe symptoms in some patients and may induce ventricular dysfunction. Drugs are frequently not efficacious in this condition and have major side-effects and toxicity which is particularly relevant in the young patient.
Materials and methods: Two young patients presented with symptomatic high-grade PVCs. The first, a 21-year-old male patient was almost permanently in a bigeminal rhythm with bizarre atypical left bundle branch block (LBBB) morphology ectopics. The ensuing compensatory pauses produced sinus bradycardia. The second, a 10-year-old girl had 1233 PVCs per hour on Holter monitoring. These had positive inferior axis and morphology consistent with a right ventricular outflow tract (RVOT) source. Aside from mildly dilated RVOTs on the echocardiogram, both had otherwise structurally and histologically normal hearts as determined by right ventricle (RV) angiography, cardiac magnetic resonance imaging (MRI) and RV biopsy. Both patients underwent cardiac electro-physiology (EP) studies. In the first patient, a 3D map was constructed and an activation map identified an arrhythmogenic focus on the free wall of the RV adjacent to the tricuspid annulus. This was ablated with robotic assistance. An arrhythmogenic focus was identified in the second patient in the RVOT and manually ablated.
Results: Both patients were rendered completely asymptomatic off all drug therapy. Follow-up ECG and Holter studies confirmed elimination of the PVCs.
Conclusion: Radiofrequency ablation is an effective and safe treatment of symptomatic PVCs in the young patient.

1428: ANGIOPLASTY PROCEDURE IN AORTIC COARCTATION

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Background: Follow up of patients (p) with aortic coarctation (AoCo) post angioplasty (BCA) procedure.
Methods: We performed retrospective analysis of 47 p post angio-

plasty procedure, grouped by severity as mild < 20 mmHg, moderate 20–40 mmHg, and severe > 40 mmHg. Topics reviewed: age at diagnosis, clinical manifestations (cardiac insufficiency (CI), arterial hypertension (AH)), defect extension (localised, extensive, isthmus hypoplasia), associations, initial treatment (catheterisation or surgery), and long-term evolution. Multivariable statistical analysis t test $p <$:0.05.

Results: Forty-seven p with AoCo post BCA were followed up. Age at diagnosis was < 1 month (11 p), 1-2 years (17 p) and >2 years (13 p); the median age at BCAwas 8 ± 8 months. On clinical examination 19 p had CI and 8 p AH; 1 p had AH without CI. Electrocardiography revealed right ventricular hypertrophy in 13 p, and left in 11 p. Associations included ventricular septal defect in 13 p (5 perimembranous, 7 muscular, 1 subaortic), ductus in 8, mitral stenosis in 1, mitral insufficiency in 3, aortic insufficiency in 3, and right aberrant subclavian artery in 1. Anatomic types: 41 p localised: 6 p membranous, 35 p fibromuscular; 1 p extensive; 7/47 p had hypoplasia of the transverse aortic arch. Severity was judged as moderate in 18 p and severe in 29. BCA involved 52 procedures on 47 p; 44 p had one BCA, 4 p had two BCA (2 p had native AoCo), 1 p had four BCA, and 1 p needed a stent. The procedure was efficient in 38/47 p (80.8%), but not efficient in 8 p; 1 p died of cardiogenic shock during the procedure. Pressure gradient post procedure: dropped from 50 ± 18 mmHg to 14 ± 10 mmHg ($p = 0.00$). Recoarctation (RC): Seventeen of 30 p with native AoCo, all < 1 year old with CI, had early RC X: 12 ± 10 month; 2 p had small saccular aneurysms. Four of 17 p had postoperative RC (23%); 1 p required a stent and 3 p another BCA.

Conclusion: Angioplasty is an efficient recourse to solve complicated types of AoCo in infants (localised, without isthmus hypoplasia). This procedure in infants has a high risk of recoarctation (57%) and saccular aneurysms (4.2%). Therefore, except in those special situations, it is not recommended as the first therapeutic choice.

1437: A META-ANALYSIS ON THE EFFICACY AND SAFETY OF TRANSCATHETER DEVICE CLOSURE OF VENTRICULAR SEPTAL DEFECTS (VSD)

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Background: Advances in interventional techniques now allow for transcatheter treatment of some VSDs. Widespread use of this option is limited, unlike atrial septal defects (ASDs), to several factors, one being concerns about adverse events. We performed a systematic analysis to look at outcomes and complications associated with use.

Method: A PubMed search for series in English on device closure of VSD from 2003 to June 2012 was performed. We excluded small series which were included in multicentre studies so as not to double count, and also patients who had acquired VSD following myocardial infarction. The random effect model was used to obtain pooled estimates of success and complications.

Results: A total of 37 publications comprising 4 406 patients with VSDs (perimembranous 3 758 , muscular 419, intracristal 47, doubly commited subarterial 36, multiple 16, post-surgical 123, unclassified 7) were included in this analysis. The age of patients ranged from 3 days to 84 years with the mean age ranging from 0.4 to 37.9 years at the time of surgery. The pooled estimate of success of device implantation was 96.6% (95% confidence interval (CI):95.7–97.5; heterogeneity test $p < 0.001$). Complications included residual shunt, rhythm abnormalities, valvular defects and others.

Discussion: Our analysis suggests that transcatheter device closure of VSD is safe and yields good results. However, this study does not analyse different devices individually, and it does not segregate age range and different VSD types.

Conclusion: Transcatheter closure of VSD appears a feasible alternative to surgery (with very high success and low complication rates). However, there are several complications which require attention. Further data stratifying type of VSD, age range of patients and prevention of complications would be useful before this can be recommended for routine treatment of haemodynamically significant VSD.

1441: FOETAL AORTIC VALVULOPLASTY TECHNICAL ASPECTS AND FACTORS INFLUENCING POSTNATAL OUTCOME

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Objectives: There are still many queries concerning foetal aortic balloon valvuloplasty. The aim of this study was to evaluate early experience and find the main factors influencing the postnatal outcome.

Material and methods: Between June 2011 and July 2012 foetal aortic valvuloplasty was performed in 10 fetuses. Evolving hypoplastic left heart syndrome (HLHS) was diagnosed in 8, severe heart failure with foetal hydrops in 2. The mean age of treatment was 24 weeks. Maverick2 catheters were used, with balloon size slightly bigger than the aortic valve. Six procedures were performed under general anaesthesia, 4 under mothers' intravenous and foetal analgesia with fentanyl.

Results: All aortic valves were successfully dilated, without major complications during and just after the procedure. Pericardial puncture was necessary in 2, and adrenaline injection in 2. One emergency caesarean section was performed because of placental abruption on the 2nd day after the procedure, 1 foetus died at the end of pregnancy from obstetric causes. Two fetuses are still *in utero*. Aortic diameter was within lower limits of normal in all children. Three neonates with big left ventricles (LVs) and poor function had balloon valvuloplasty just after birth. Conversion to univentricular circulation was attempted in two of them, the third is still a neonate. All three of them had severe heart failure. Three Norwood operations were performed without complications.

Conclusions. Foetal aortic valvuloplasty is technically possible and can be performed by an experienced team without major complications. Good aortic development was observed in all children, whichmade the Norwood procedure easier in neonates with HLHS ventricles. Neonates with borderline left ventricles with high gradients across the aortic valve were the most difficult group of patients. After our early experience, we think that early aortic valve replacement, i.e. neonatal Ross-Konno operation, should be considered to improve long-term outcome after prenatal aortic valvuloplasty.

1467: FERP (FOETAL ECHOCARDIOGRAPHY REFERRAL PROJECT): THE DISSEMINATION OF GUIDELINES AIMED TO IMPROVE PROVIDER KNOWLEDGE

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Although clinical practice guidelines do exist regarding foetal echocardiography referrals, these guidelines are often not utilised in practice, partly because of lack of provider knowledge. The primary purpose of this 6-week Quality Improvement (QI) Research Project was to increase the knowledge of obstetrical providers on the guidelines for foetal echocardiography referrals. A 3-phase intervention project using a non-experimental descriptive design methodology was implemented, employing both pre- and post-project surveys to a convenience sample of obstetrical providers within a 50-mile radius of a specified Southern California zip code.

A paired-samples t test was used to evaluate the statistical signifi-

cance of pre- and post-project obstetrical provider scores. Study findings indicated a positive correlation between the use of an education intervention, implemented by an advanced practice nurse and her team, utilising a multi-modal approach and increased provider knowledge on foetal echocardiography guidelines.

1482: STENTING THE PATENT DUCTUS ARTERIOSUS IN NEONATES WITH CRITICAL CONGENITAL CYANOTIC HEART DISEASE IN A DEVELOPING COUNTRY

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Background: Critical cyanotic heart disease in newborns is life-threatening and difficult to manage. Systemic to pulmonary artery shunts are used as temporary palliative interventions. This is associated with increased morbidity and mortality. Patent ductus arteriosus (PDA) stenting has been introduced as a non-surgical alternative. The aim of this study was to determine the short- and medium-term outcomes of PDA stenting in neonates with duct-dependent pulmonary circulations in central South Africa.
Materials and methods: Sixteen neonates with duct-dependent pulmonary circulation requiring PDA stenting were included in a retrospective review from September 2005 to March 2012. In 3 cases stenting was not performed because of inability to obtain vascular access. Stenting was performed via the femoral artery and/or vein. Standard balloon expandable coronary bare metal stents were used off-label. Follow-up included clinical assessment and echocardiography.
Results: Thirteen cases were successfully stented at first attempt. Five underwent concomitant Rashkind septostomy. Diagnoses consisted of pulmonary atresia (n = 9) and tricuspid atresia (n = 4). Median age at procedure was 2 days (range 0–23) and median weight 3.0 kg (1.5–4.3). Number of stents used were 1 (n = 3), 2 (n = 9) and 3(n = 1). Implanted stent diameters were 3.5 mm (n = 4), 4 mm (n = 7), 4.5 mm (n = 1) and 5 mm (n = 1). Median saturations were 80% (52–92) pre-procedure and 90% (64–97) post-procedure (p = 0.07). Periprocedural complications included stent migration into pulmonary artery (n = 1), femoral artery compromise (n = 1) and temporary dysrhythmia (n = 3). There was no periprocedural mortality. All patients received low-dose acetylsalicylic acid following the procedure. At last follow-up (median 136 days; range 7–334), balloon angioplasty (n = 1) and restenting (n = 1) of stenotic stents had been performed. Three cases had proceeded to palliative surgery. Four cases had demised at a median of 9.5 days (7–14) post-procedure, secondary to septicaemia (n = 1), airway obstruction(n = 1), pneumonia(n = 1) and severe hypertrophic cardiomyopathy (HOCM) (n = 1). All deaths were unrelated to the procedure.
Conclusions: PDA stenting is a safe and effective alternative to surgical shunts as a palliative procedure for neonates with duct-dependent pulmonary circulation.

1489: GIANT INTRACARDIAC HAEMANGIOMA DIAGNOSED IN UTERO

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We describe a case of a foetus diagnosed at gestational age (GA) 31 weeks with a large right atrial cavernous capillary haemangioma who had elective delivery by caesarean section and successful elective resection of tumour at 1 week of life. A 36-year-old G3P2001 mother was referred for a foetal echocardiogram when a mass was visualised on routine anatomy scan at 31 weeks' gestation. Foetal echocardiogram was significant for a large loculated visualised mass occupying the majority of the right atrial cavity with sparing of the posterior third of the right atrium. There was no obstruction to the tricuspid valve inflow, systemic venous return or coronary sinus. There was a small pericardial effusion of no haemodynamic significance. The foetus was in sinus rhythm. The patient was followed weekly with echocardiograms. The night prior to delivery, the foetus was in atrial flutter in 2:1 at a ventricular rate of 300/minute without hydrops fetalis. Rhythm had spontaneously converted to sinus before delivery. Delivery was uneventful with Apgar scores 8 and 9. Echocardiogram post-delivery revealed a wide-based large mass (19.9 x 21.3 mm) in the lateral wall of the right atrium, attached to the right atrial appendage extending anteriorly to the aortic root. The mass had irregular borders with cystic formation inside in addition to echogenicity with blood flow with connection to the right coronary artery. Elective resection of the tumour was done at 1 week of age. Pathologic evaluation also confirmed the diagnosis. Patient was discharged home a few days after surgery.

Compared to the most frequently seen rhabdomyomas and less common teratomas and fibromas, cardiac haemangioma is one of the rarest types of benign cardiac tumours found in neonates. Foetal echocardiogram is an essential diagnostic tool for early detection, close monitoring and appropriate and early treatment.

1506: COVERED STENTS IN THE MANAGEMENT OF NATIVE COARCTATION OF THE AORTA: INTERMEDIATE AND LONG-TERM FOLLOW-UP

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Objectives: To evaluate the results of the use of covered Cheatham-Platinum (CP) stents in the management of native coarctation of the aorta (CoA) and report intermediate and long-term follow-up.
Background: Covered stents are being used increasingly in CoA to reduce risk of aortic wall complications; however, there are limited data on the intermediate and long-term outcome.
Patients and methods: Fifty-nine covered CP stents were implanted in 56 patients. In 54 patients these were as primary treatment for severe CoA with near atresia and complex CoA, and in 2 as rescue for complications related to bare stent implantation. The follow-up was based on echocardiography and computed tomography (CT).
Results: Mean patient age was 22.25 ± 1.2 (11–56) years and weight was 58.6 ± 2.1 (32–99) kg. The systolic gradient across the CoA decreased from mean of 51.4 ± 3.4 to 4.6 ± 0.7 mmHg ($p < 0.0001$). The diameter of the CoA segment increased from 4.69 ± 0.20 to 15.1 ± 3.2 mm ($p < 0.0001$). There was one death 3 days post procedure due to cerebral anoxia. There was one dissection diagnosed 24 hours post procedure. At a mean follow-up of 45.9 ± 3.9 (3–120) months, all stents were patent and in good position on CT. Four (7.1%) patients underwent successful redilation. Antihypertensive medication was decreased or stopped in 37 (66%) patients.
Conclusions: Covered CP stents may be used effectively as the therapy of choice in selected patients with severe CoA. Aortic wall complications can occur even with covered stents. The stents can be redilated safely to keep pace with somatic growth. Covered stents provide a safe alternative to conventional stenting in the intermediate and long term.

1512: THE DEVELOPMENT OF THE LEFT VENTRICLE IN PRENATALLY DIAGNOSED LEFT SIDED OBSTRUCTION

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Background: The asymmetry in four chamber view with small left ventricle (LV) in the foetal heart could be caused by the obstruction in different levels of the left heart, by the presence of left superior vena cava draining coronary sinus (LSVC-CS), aortic stenosis (AS), coarctation of aorta (COA) or hypoplastic left heart syndrome (HLHS). The aim of the study was to follow up the development of

the LV in foetuses with left sided obstruction.
Materials and methods: The length of the LV was measured at the time of diagnosis prenatally and after birth in echocardiographic evaluation in 131 foetuses with left-sided obstruction and small LV.
Results: Of these 131 foetuses detected at 11–37 weeks gestation, 42 were diagnosed with LSVC-CS, 30 with AS, 24 with COA, 21 with HLHS and 14 without final diagnosis (non-specified disproportion) as a result of early stage of gestation. There were extracardiac abnormalities in 24 foetuses and chromosomal abnormalities in 20. The pregnancy was terminated in 51 (39%) cases: for HLHS 100%, for non-specified disproportion 93%, for AS 43%, for COA 8% and for LSVC-CS 4%. Eighty (61%) children were born; 6 of them died. The size of LV was normal postnatally in 100% of foetuses with LSVC-CS, in 95% of foetuses with COA and in 68% of foetuses with AS.
Conclusion: There was normal development of the LV in foetuses with prenatally diagnosed LSVC-CS and COA (small number of terminations of pregnancy and low mortality after birth). In AS there was good development of LV after birth in the majority, but there was a high number of terminations of pregnancy (43%). There were associated abnormalities in the majority of foetuses with non-specified disproportion and none of them survived.

1515: TRANSCATHETER CLOSURE OF AN AORTO-CAMERAL FISTULA WITH A VASCULAR PLUG USING A RETROGRADE APPROACH: A CASE REPORT

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Background: Aorto-cameral fistula is a rare cardiac anomaly. Few reports in the medical literature discuss the use of a vascular plug using a retrograde approach to close these fistulae.
Materials and method: We describe the case of an 8-year-old girl who presented with complaints of frequent respiratory infections since early infancy, shortness of breath (NYHA class II) and palpitations for the last 2 years. She had a 3/6 continuous murmur at the left lower parasternal area. Echocardiography revealed a huge aorto-cameral fistula originating from the right sinus of Valsalva emptying into the right ventricular (RV) inlet just below the anterior leaflet of the tricuspid valve (TV).The right coronary artery (RCA) was faintly visualised. As the child was symptomatic, a cardiac catheterisation was planned with a goal of possible transcatheter closure of the defect. Aortogram confirmed the echo findings. The RCA was faintly visualised originating 8 mm away from the aortic sinus with decreased flow. The narrowest segment of the fistula measured 6.1 mm.There was retrograde filling of the RCA from the left coronary artery (LCA) branches. On temporary balloon occlusion beyond the origin of the RCA with a 6F SwanGanz catheter, no electrocardiograph (ECG) changes were seen. A 10 mm vascular plug (166% of the narrowest diameter of the fistula) was placed into the narrowest part of the fistula through a retrograde approach, through a patent ductus arteriosus (PDA) device delivery system, resulting in complete occlusion of the fistula. Post procedure RCA angiogram showed normal flow through the RCA with visualisation of the marginal branches which were hitherto not seen as a result of steal through the fistula. The retrograde approach obviated the need for an arteriovenous loop. No procedural complications occurred with the patient showing better exercise tolerance on follow-up.
Result: Complete occlusion of fistula with improvement in symptoms was achieved.
Conclusion: Coronary artery fistula closure by vascular plug through a retrograde approach is safe and should be considered as the standard approach**.**

1559: VARIABLE APPROACH AND OUTCOME OF STENTING OF ARTERIAL DUCT IN DUCT-DEPENDENT PULMONARY CIRCULATION

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Background: Stenting of arterial duct has emerged as an alternative for systemic to pulmonary shunt in young infants with duct-dependent pulmonary circulation (DDPC). However, the approach of stenting is determined by the anatomy and orientation of the arterial duct. We report our experience of stenting of arterial duct by various approaches and their outcomes.
Materials and methods: Data of 45 (31 male) young children with DDPC were included for the procedure. Seven children were excluded after angiogram because of unfavourable anatomy in 4 and inability to negotiate wire in 3. Various parameters and outcomes were analysed.
Results: The anatomical diagnosis includes 24 cases of ventricular septal defect (VSD) pulmonary atresia, 12 of pulmonary atresia with intact septum and 9 with single ventricle physiology and pulmonary atresia. The median weight was 3 kg (range 1.7–6.5) and age 20 days (range 2–370 days). Successful deployment of stent was done in 37 (82%) children. The approach for stent deployment was determined by the anatomy of the arterial duct and physician experience; femoral artery approach in 19, axillary 8, carotid (hybrid) 15 and femoral vein in 3 cases. There were two deaths; rupture of the ductus in one and the other was due to sepsis. There were three major procedure-related complications: spasm, dissection and acute thrombosis, which were managed. Six children died at various hospitals during follow-up – as a result of pneumonia (1) , gastroenteritis with dehydration (1), and awaiting surgery (2); the cause could not be detected in 2. Successful second-stage procedures (systemic to pulmonary shunt in 4, bidirectional Glenn in 4 and biventricular repair in 4) were done.
Conclusion: Stenting of the arterial duct in DDPC is an alternative palliation using various approaches as a result of variable anatomy of tortuous vertical arterial ductus. The outcome is determined by the presentation, associated co-morbid condition and underlying complex anatomy.

1599: MANAGEMENT OF DEXTROCARDIA, SITUS INVERSUS TOTALIS, MIXED TYPE TAPVC, COMPLETE AV CANAL DEFECT, COMMON ATRIUM, BILATERAL SVC, PDA, SEVERE PAH'

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Objective: To present the management of dextrocardia, situs inversus totalis, TAPVC – mixed, complete atrioventricular (AV) canal defect, common atrium, bilateral SVC, patent ductus arteriosus (PDA), severe pulmonary arterial hypertension (PAH).
Diagnosis: This 9-month-old girl baby girl was admitted with failure to thrive and respiratory distress. Echo evaluation revealed the above-mentioned diagnosis. After preoperative evaluation, surgery was performed. Operative finding included mixed type TAPVC – right superior and middle pulmonary veins and left inferior pulmonary vein were draining into common venous chamber. A vertical vein from the common chamber was joining RSVC, and a left superior vein to LSVC. The right upper lobe vein joined the RSVC. Morphologic right atrium (RA) was on right side receiving RSVC and hepatic vein. Morphologic left atrium (LA) was on the left side receiving LSVC and IVC. Other findings were: unroofed coronary sinus; complete AV canal defect – Rastelli Type A with large inlet ventricular septal defect (VSD); severe mitral regurgitation (MR) – anterior mitral leaflet (AML) cleft; common atrium.
Results: The procedure included TAPVC repair & AV canal repair. Rerouting of pulmonary veins was done with wide anastomosis

between the common venous chamber and mLA. Vertical vessel was ligated. Since innominate vein was present, LSVC was interrupted, hence diverting left superior vein to mLA. Regarding AV canal repair, a double patch technique was used. VSD closure, MV repair with AML cleft repair, tricuspid valve repair, atrial septal defect (ASD) closure with Pericardia patch created an atrial baffle, diverting IVC, RSVC and hepatic vein to mRA. Core cooling was done to 28C. Total CPB time was 325 min and aortic cross clamp time 235 min. Total ventilatory support was given for 86 hours and inotropes for 6 days. Intensive care unit (ICU) stay was until the 8th post-operative day (POD).
Conclusion: Baby was discharged on 16th POD with stable haemodynamics and on normal oral feeds.

1570: USEFULNESS OF JOINT FOETAL ECHOCARDIOGRAPHY AND COUNSELLING SERVICE - EXPERIENCE IN HONG KONG

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Aim: We collaborated with our obstetricians to run a Joint Foetal Echo and Counselling service. This study aims to review the usefulness of this service.
Methodology: We reviewed our results from 1993 to 2012. Medical records were retrieved and analysed retrospectively.
Results: A total of 150 pregnant women underwent foetal echocardiography in our department; 56 foetuses were found to have cardiac abnormalities and prenatal counselling was offered to their families. Detailed anatomy, expected management, prognosis and our life experience of similar cases were explained to the expectant parents. Psychological support was offered as well. Out of the 56 foetuses, 45 had major congenital cardiac abnormalities; only 13 (29%) of the pregnancies were electively terminated, 1 aborted spontaneously and the rest were born at or near full term.
Follow-up: After birth, these babies were reassessed, stabilised and referred to our neonatal cardiac centre for surgery. The prenatal diagnoses were accurate in 98.7% of cases. The transfer and preoperative course was smooth as appropriate stabilisation was instituted soon after birth. The survival rate after surgery is best in the right ventricular outflow tract obstruction and transposition of great artery groups, amounting to 100%. The oldest child is 13 years of age. However, all died after surgery for truncus arteriosus, pulmonary artery sling and hypoplastic left heart syndrome, though the number was very small. Of those with complex heart lesions including heterotaxy syndrome, 60% survived with or without surgery. The only foetus diagnosed with premature closure of ductus survived after prompt delivery and neonatal intensive care.
Conclusions: In our experience, the Joint Foetal Echocardiography and Counselling service is invaluable for the management of foetuses with cardiac abnormalities. It does result in a low elective abortion rate but also improves babies' survival and parents' acceptance after birth.

1591: EXPERIENCE WITH THE COOK FORMULA STENT IN PAEDIATRIC CARDIAC INTERVENTIONS

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Introduction: Balloon-expandable stents are an integral part in the catheter treatment of congenital heart disease (CHD). In the growing child, stents have to be dilatable to greater diameters over time. All current stent designs have limitations. The pre-mounted Cook Formula stent is a recent 316 stainless steel open-cell design licensed for peripheral vascular work.
Methods: Extensive *ex vivo* studies were carried out to better understand the stent behaviour regarding shortening and ability to overdilate the stent. Subsequently 30 stents were implanted in 29 children (median age 0.96 (0.03–9.8) years; median weight 7.7 (3.8–43)kg).
Results: Stents were implanted in the right ventricle (RV) outflow tract in 11 patients with Fallot-type lesions, in 9 for branch pulmonary artery stenosis (3 post Fontan), 4 conduit stenosis, 2 Fontan fenestrations, and 1 each for SVC, coarctation of the aorta (CoA) and patent ductus arteriosus (PDA). Stent delivery up to 7 mm was over a 0.014' wire via a 4F sheath or 6F guide catheter. 8 or 10 mm stents (from 3/2012) were placed over a 0.035' wire using a 7F Mullins sheath. Stent tracking and delivery was excellent. There was no stent shortening for dilatation to nominal diameter and beyond. This allowed for precise placement, avoiding protrusion into adjacent vessels. Sixteen stents were primarily or subsequently overdilated without any shortening. The 5 mm stent could be dilated to 10 mm, and the 10 mm stent could be dilated to 17 mm without shortening. There was one circumferential balloon fracture requiring retrieval, and one stent slipped and was removed.
Conclusion: The Cook Formula stent is a versatile pre-mounted balloon-expandable stent that can be significantly overdilated with virtually no shortening. It is a great addition to the range of stents for use in the catheter treatment of complex CHD in children.

1601: STENTING OF BILATERAL ARTERIAL DUCTS IN COMPLEX CONGENITAL HEART DISEASE

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Background: Maintaining ductal patency in duct-dependent congenital heart lesions by implantation of coronary stents is an alternative to systemic pulmonary shunt in selected cases and lesions with suitable anatomy. Bilateral arterial ducts are a very rare pattern of pulmonary blood flow in congenital heart disease with pulmonary atresia with non-confluent pulmonary arteries. Ductal closure leads to severe systemic hypoxia indicating emergent surgical palliation or repair; percutaneous arterial duct stenting might be an alternative to surgery, in such high-risk patients.
Methods: We describe two critically ill neonates (both male) with complex heart disease and discontinuous pulmonary arteries surviving on bilateral arterial ducts who successfully underwent transcatheter ductal stenting as first-step palliation toward lower-risk surgery.
Results: Patient #1 (weight 3.8 kg) was referred at 4 weeks with single double inlet left ventricle, atretic right atrioventricular valve, restrictive atrial septal defect (ASD), pulmonary atresia, disconnected BPAs supplied by bilateral ducts. The procedure entailed atrial septostomy/bilateral coronary stent placement in each ductus. Patient #2 was diagnosed antenatally then delayed to complete fungal sepsis treatment. At 8 weeks (weight 4.3 kg) bilateral coronary stent placement in each ductus was performed for single ventricle, atretic left atrioventricular valve, pulmonary atresia, right arch disconnected BPAs supplied by bilateral ducts. Ductal stabilisation was achieved with coronary stents, patient #1 is awaiting first palliative surgical procedure, patient #2 underwent a successful first palliative surgical procedure, and he continues to do well at clinical follow-up at 14 months of age.
Conclusions: Stent implantation is a technically feasible, safe and effective palliative option in high-risk surgical patients with bilateral arterial ducts with pulmonary atresia with non-confluent pulmonary arteries.

1630: TEMPORARY USE OF SMALL STENTS IN CRITICAL CONGENITAL HEART DISEASE

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Background: Surgery in children with critical congenital heart disease is associated with a high morbidity and mortality. The aim of this study was to look at the short and medium-term outcomes of stent implantation in these patients and to determine whether surgery could be delayed in a developing country.

Materials and methods: Eighteen children were included in a retrospective review from October 2005 to May 2012. Vascular access was obtained via the femoral artery and/or vein. Balloon-expandable coronary bare metal stents were used. Follow-up included clinical assessment and echocardiography.
Results: Eighteen cases were successfully stented. Indications were aortic arch obstruction (9), right ventricular outflow tract obstruction (4), systemic-to-pulmonary artery shunt stenosis (3), pulmonary venous chamber stenosis (1) and patent ductus arteriosus (PDA) stenting to preserve a discontinous left pulmonary artery (1). Eight patients had residual problems following prior surgery. Median age was 4.8 months (range 0.1–65.1) and median weight 4.6 kg (1.7–17). Number of stents used: 1 ($n = 12$), 2 ($n = 5$) and 4 ($n = 1$). Implanted stent diameters: 2.5 mm ($n = 2$); 3.5 mm ($n = 2$); 4.0 mm ($n = 4$); 4.5 mm ($n = 3$); 5.0 mm ($n = 9$); 5.5 mm ($n = 1$); 6.0 mm ($n = 3$) and 8.0 mm ($n = 2$). Paired pressures were not obtainable in all cases as most patients were critically ill. Saturations improved from a median 83.5% to 90% in patients with obstructive pulmonary blood flow ($n = 4$). Periprocerdural complications included temporary rhythm disturbances ($n = 3$). There was no periprocedural mortality. Last follow-up was at a median 200 days (range: 10–1406). A second stent procedure was performed in 6 patients ranging between 3.8 and 17.8 months post initial stent placement. Five cases proceeded to surgery. Five cases demised at a median of 73 days (range: 20–422), all at home secondary to unknown causes. Three patients have been lost to follow-up. Five patients remain in follow-up.
Conclusions: Mortality in this high-risk group remains high. Stenting is feasible and may delay surgery in some; however, problems in developing countries present unique challenges to patient follow-up.

1695: ANEURYSM OF THE FORAMEN OVALE FLAP AND VENTRICULAR SIZE DISCREPANCY IN PRENATAL ECHOCARDIOGRAPHY

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Background: Ventricular size discrepancy (VD) is easily detected in routine general foetal echography; echocardiography is therefore performed, the aim being to detect prenatally left ventricle inflow or outflow obstructive lesions and aortic arch anomalies. However VD can be a physiological aspect late in pregnancy or associated to anomalies such as left vena cava or aneurysm of the foramen ovalis flap (AFOF).
Materials: Among 3 500 prenatal echocardiographies performed in our unit from January 2008 to July 2012, 45 AFOF were detected with variable degree of VD late in pregnancy: mean term was 33.5 ± 0.7 weeks of gestation (w). In all patients ventricular and arterial ratio, right/left ventricular diameter (VR), aorta/pulmonary artery diameter (AR) and aortic isthmus diameter (AID) were measured. Colour Doppler images and velocities were recorded. In most cases analysis of the aortic arch showed a complete aortic arch with normal flow velocity and direction; AID was considered as normal when > 3 mm after the 30th w or 0.1 mm x w when earlier in gestation.
Results: In group 1 (G1), 33 patients with VR < 1.5 had 31 normal AIDs and 2 small AIDs. In group 2 (G2), 12 patients with VR ≥1.5 had 8 normal AIDs and 4 small AIDs. Only 15% of 45 patients with AFOF and VD had AID suspicious of postnatal coarctation. In these few cases birth was planned in our hospital in conjunction with the intensive care and paediatric cardiology units.
Conclusion: AFOF is frequently associated with VD in prenatal echocardiography. If the AID is normal with forward flow and normal velocity, AFOF can be considered as the main factor responsible for VD and there is low risk of postnatal coarctation. This screening is useful to plan *in utero* transfers from peripheral hospitals and avoid unnecessary ones.

1697: SCREENING OF FOETAL CARDIAC DISEASE IN SAUDI ARABIA: A TERTIARY CARDIAC CARE CENTRE EXPERIENCE

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Introduction: Antenatal detection of cardiac malformations remain suboptimal in most areas of the world with a gap in diagnosing serious cases of congenital heart disease (CHD).
Method: To describe the current status and factors affecting detection of CHD antenatally in Saudi Arabia.
Results: Foetal echocardiography/cardiology started in the 1980s at our institution; however rates of detection of foetal CHD remain low with almost half the cases referred with serious CHD not picked up by antenatal screening. Contributing factors include high incidence of CHD in low-risk pregnancies, suboptimal referral system, lack of regional/national foetal cardiac screening programmes and the need for more countrywide training for cardiac screening at obstetric units.
Conclusions: The overall rate of detection of major congenital heart disease before birth remains low. Cases will only be detected by screening of the low-risk population at the time of routine obstetric scanning. A common effective practice standard for antenatal detection of CHD needs to be established nationally/regionally and globally, with enhanced training and education in the screening of the foetal heart with appropriate referral/networking.

1770: PRENATAL DIAGNOSIS OF EBSTEIN'S ANOMALY: REPORT OF FOUR CASES

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Ebstein's anomaly (EA) is a rare malformation of tricuspid valve leaflets and can be detected in foetal life by important cardiomegaly during obstetric ultrasonographic examination, the most common indication of foetal echocardiogram. We describe the evolution of four cases of foetal diagnosis of EA, from 3 units of the Fetal and Pediatric Cardiology Centre of Rio de Janeiro are reported. Patients were referred for foetal echocardiogram because of cardiomegaly observed during obstetric assessment, between 25 and 37 weeks' gestation age. All foetuses had significant tricuspid regurgitation, and significantly enlarged right atrium. Ventricular septal tricuspid leaflet was attached to ventricular septum, obstructing right ventricular outflow tract with functional pulmonary atresia (PA) in two cases, and true PA, associated with pulmonary branches hypoplasia, in the other two foetuses. One foetus had heart failure with periods of atrial tachycardia treated with oral maternal digitalis. Two patients died, one in the 38th gestational week due to corioaminionitis confirmed by anatomo-pathologic study after birth, and another due to pulmonary hypoplasia. Another case of PA needed a systemic-pulmonary shunt during the neonatal period and had a tricuspid valve repair one month after birth with good postoperative evolution, although he developed tachyarrhythmia that was controlled with amiodarone. The last one had functional PA but pulmonary blood flow became normal, after measures to lower pulmonary vascular resistance.

Foetal echocardiogram is an important tool of diagnosis of congenital heart disease with severe manifestations and complications during the perinatal period. It permits the medical team to create strategies to prevent the dangers of metabolic acidosis and hypoxaemia in the first hours after birth. The presence of intrauterine pulmonary hypoplasia, severe tricuspid regurgitation and pericardial effusion are signs of poor prognosis.

1775: PERCUTANEOUS TRANSMITRAL COMMISSUROTOMY IN CHILDREN: INTERMEDIATE TERM RESULTS WITH SPECIAL REFERENCE TO PULMONARY HYPERTENSION

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Objective: To know the long-term results of percutaneous transmitral commissurotomy (PTMC) and fate of pulmonary hypertension in children who have undergone PTMC.
Material: During the last 11 years 138 children, aged 10.48 ± 2.24 years (range 3.5 -16) had PTMC. Indications were echocardiographic evidence of moderate to severe mitral stenosis (MS) plus pulmonary arterial hypertension (PAH) more than 50 mmHg and/or FC III-IV
Result: Pre PTMC, mitral pressure gradient (MPG) across the mitral valve (MV) was 18.6 mmHg ± 3.4 which decreased to 7.6 mmHg ± 2.21 .MVA was 0.62 cm^2 ± 0.12 which increased to 1.6 cm^2 ± 0.3. Pulmonary artery pressure (PAP) was 83 mmHg ± 13 decreased to 50 mmHg ± 14.5. Three procedures were unsuccessful: there were 2 deaths, one within half an hour in patient (pt) with PAP of 100 mmHg and left ventricle (LV) dysfunction, had no mitral regurgitation (MR) or tamponade. Another died after 24 hours, after thromboembolism of LAD, he revived and had successful PTMC but died after 24 hours. One had tamponade drained and had a successful PTMC after 1 week. There was no change in MR in 50 pts.

After a mean follow-up period of 44.3 yrs ± 30.6 of 110 pts (10 months to 10 years), 8 needed repeat PTMC after 5.37 ± 2.3yrs (0.5–8 yrs), and 2 had mitral valve regurgitation (MVR) after 1 and 7 years.

Systolic PAP was 83 mmHg ± 13 pre procedure which decreased to 50 mm Hg ± 14.5 immediately after and to 42 mmHg ± 13 after 6 months . Immediately after PTMC 54 pts had PAP more than 50 mmHg, after 6 months 24pts .Out of these 2 died: one had severe PAH despite adequate relief of MV, died after 3 yrs; another had mild MS and +2 MR with persistent severe PAH, had MVR after 1 year. PAP did not decline after surgery and he died two years after PTMC. Both had initial PAP of more than 100 mmHg.
Conclusion: PTMC is effective in reliving stenosis, however initial high PAP is predictor of persistent pulmonary hypertension.

1783: CONGENITAL LEFT VENTRICULAR ANEURYSM: FOETAL DIAGNOSIS

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Background: Congenital left ventricular aneurysm (CLVA) is a rare cardiac malformation occurring in 0.5:100 000 births, regardless of gender and presents as a single lesion in 75% of cases. Most patients are asymptomatic, although some complications such as congestive heart failure, arrhythmias, thromboembolism and ventricular wall rupture may occur.
Case report: A 29-year-old primigravida, without risk factors was referred for foetal echocardiography at 33 weeks gestational age with diagnosis of cardiomegaly during obstetric ultrasound. Foetal echocardiogram showed a 1.9 cm expansive rounded area in left ventricle apex, consistent with a CLVA with slow velocity flow inside. Doppler analysis detected supraventricular extrasystoles and bigeminism periods. No other structural changes, thrombi or signs of heart failure were observed during subsequent evaluations. Delivery occurred at term without complications and postnatal echocardiogram confirmed the diagnosis. During the first weeks of life, the newborn developed supraventricular tachycardia, successfully controlled with betablockers; since then she has remained asymptomatic
Discussion: Aetiology of CLVA is unknown and may occur by localised weakening of the ventricular wall during embryogenesis, infection or ischaemia of the ventricular myocardium. It can be produced by stenosis, hypoplasia or abnormal development of coronary arteries. Foetal echocardiogram is essential for early diagnosis, as well as allowing monitoring during pregnancy and birth to detect complications such as rupture of the wall, thromboembolism, arrhythmias and heart failure, should intrauterine treatment be necessary. The natural history of CLVA is not well studied so the approach must be individualised for each case.

1789: TRENDS IN PRENATAL DIAGNOSIS OF TRANSPOSITION OF THE GREAT ARTERIES WITH INTACT VENTRICULAR SEPTUM AND IMPACT ON OUTCOMES

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Background: In transposition of the great arteries with intact ventricular septum (TGA/IVS), changes occur after birth that, if untreated, lead to haemodynamic compromise and death. Prenatal diagnosis (Dx) allows for delivery site planning and timely neonatal management. Despite TGA being a major heart defect, detection is challenging for obstetricians.
Objective: To evaluate temporal trends in prenatal Dx and its impact on neonatal morbidity and mortality.
Methods: Patients with TGA/IVS referred for surgical management between 1995 and 2012 were included. The study time was divided into three 6-year periods. We compared variables pre-op and post-op between those diagnosed pre or postnatally. Variables included gestational age and weight at birth, age at admission, mechanical ventilation, ECMO, metabolic acidosis, timing of septostomy and surgery, intensive care unit (ICU) and hospital stay, and mortality.
Results: Of the 295 patients, 78 (26%) had prenatal Dx. There was an increase in the prenatal diagnosis over the years, from 10%, during the first period, to 42% during the last period. Gestational age and birth weight was similar between groups. Age at admission (1.4 vs 0), at septostomy (1.2 vs0.3) and surgery (4.7 vs 3.9) were greater in the postnatal Dx group. The postnatal Dx group had more metabolic acidosis (19% vs 7%), mechanical ventilation (71% vs 40%) and need for ECMO (3.7% vs 1.3%) prior to surgery. The overall mortality was 2.7%. In the postnatal Dx group 7/217 patients died, 3 pre-op and 4 post-op. There was 1 death (pre-op) in the prenatal Dx group. Hospital and ICU stay was not significantly different between groups.
Conclusions: Prenatal detection rate of TGA/IVS increased significantly over the study period, but is still disappointingly low at < 50%. Although the mortality rate was not different between pre- and postnatal Dx groups, patients with prenatal Dx had significantly less metabolic acidosis, earlier admission and septostomy compared to those diagnosed postnatally.

1798: FOETAL CARDIAC MASSES WITH LARGE PERICARDIAL EFFUSION: A CASE REPORT

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Background: Primary tumours of the heart are rare with a prevalence rate that ranges from 1 per 1.000 to 1 per 100 000 of the general population autopsy studies. Most of all primary cardiac tumours are benign and rhabdomyoma is the most common in the paediatric population.

Case report: A 16-year-old pregnant woman (37 weeks' gestation) was referred for foetal echocardiography evaluation which demonstrated a large pericardial effusion with partial right atrium collapse and a great number of myocardial well-delimitated masses distributed in the right and left ventricle walls and left atrium posterior wall without outflow obstruction. Magnetic resonance images were performed to study the heart tumours distribution and screening to search for extracardiac anomalies confirming the tuberous sclerosis complex diagnosis. Restrictive pericardial effusion was postnatally observed and pericardiocentesis was performed. A new effusion was observed; the newborn evolved a severe sepsis and died on the 7th day of life. Unfortunately neither an autopsy nor a cytological study of the pericardium effusion was performed. The teenage mother was referred for paediatric ambulatory attendance because she also had tuberous sclerosis diagnosis.
Discussion: Supportive evidence was obtained for the theoretical rhabdomyomas diagnosis in this case. Foetal echocardiography can provide an accurate diagnosis and the necessary follow-up considering the disorder morbidity. Serious complications can occur such as cardiac arrhythmia, flow obstruction with low cardiac output, hydrops foetalis, and sudden death. Knowledge of the outcome of affected foetuses and the association of tuberous sclerosis complex with cardiac rhabdomyoma is necessary for adequate prenatal counselling and planning of prenatal or postnatal treatment.

1804: REFERRAL PATTERNS FOR FOETAL ECHOCARDIOGRAPHY AND IMPACT OF PRENATAL DIAGNOSIS OF CONGENITAL HEART DISEASE AT A TERTIARY CARE CENTRE IN GREECE

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Objective: The aim of this study is to follow the patterns of referral for foetal echocardiograms (FE) and the impact of the prenatal diagnosis of congenital heart disease (CHD) at a tertiary paediatric cardiology/cardiac surgery care centre in Greece.
Methods: This is a retrospective study; all FE performed from 2005 to 2011 at our centre were reviewed. The main indication for the study, and the positive diagnoses of CHD were documented and analysed.
Results: A total of 4 694 FE studies were performed between 2005 and 2011 in 4 120 patients at our centre. The gestational age (GA) at which the examination was performed ranged from 16 to 37 weeks. There was gradual rise of the annual number of studies with the rate of detection of significant CHD being almost constant. There was a gradual increase in the number of patients being referred because of findings during the first trimester foetal scan – increased nuchal translucency, tricuspid regurgitation, flow reversal at the ductus venosus. Referral rate for suspicion of CHD at the anomaly scan, positive family history of CHD, arrhythmias, maternal diabetes and treatment with medications, as well as suspicion of chromosomal anomalies, remained important factors for cardiac evaluation of the foetus by a specialist. A total of 392 cases of significant CHD were diagnosed and 145 terminations of pregnancy occurred.
Conclusions: This is a large series of FE in a small country's rather homogeneous population in regard to racial, cultural and religious aspects. Our study suggests that first trimester findings in the foetus have influenced the referral patterns for FE over the last few years compared to known pre-existing indications, and that the GA for initial cardiac evaluation by a specialist is gradually decreasing. Termination of pregnancy is also the preferred option for significant CHD in our country, despite availability of appropriate postnatal interventional or surgical treatment.

1805: MATERNAL DRUG LEVELS AND RESPONSE TIME TO MATERNAL ANTIARRHYTHMIC TREATMENT IN FOETAL TACHYARRHYTHMIA

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Background: Foetal tachycardia is an emergency and requires rapid evaluation of foetal rhythm and prompt initiation of antiarrhythmic medication. Foetal mortality is unacceptably high even in treated cases. Foetal response time and maternal antiarrhythmic drug levels may play an important role in unfavourable outcomes.
Methods: We undertook a 10-year review of foetuses presenting with tachyarrhythmia to a tertiary institution. Flecainide and digoxin combination was preferred treatment. Maternal antiarrhythmic levels and foetal response time to tachycardia resolution have been evaluated.
Results: There were 44 patients, and 37 foetuses were given flecainide and digoxin combination treatment. The sinus rhythm was restored in a mean of 4.85 days (range 1–14 days) in foetuses with supraventricular tachycardia (96%). The foetal response time in atrial flutter was slightly longer with a mean of 10 days (range 1–18 days) but all hydropic foetuses had complete resolution of ascites, pleural, and pericardial effusions with rate control. Resolution of hydrops took as long as 2 weeks after normalisation or reduction of foetal heart rate below 160 bpm. There was no correlation between maternal drug levels and maternal electrocardiographic parameters or foetal response time to tachycardia.
Conclusion: Combination treatment is well tolerated by pregnant women, and there was no major side-effect requiring withdrawal of antiarrhythmic medication. The 2-drug combination was effective in improving foetal haemodynamics in most cases but foetal response time showed no significant correlation with maternal drug levels.

1824: TRANSCATHETER OCCLUSION OF LARGE CONGENITAL CORONARY CAMERAL FISTULAE USING THE AMPLATZER VASCULAR PLUG II

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Background: A coronary cameral fistula is a rare connection between a coronary artery and a cardiac chamber or vein. Most of these fistulae have a congenital origin; though they can be acquired. Congenital coronary cameral fistulae (CCCF) are rare abnormalities. Their management has ranged from cardiac surgery to transcatheter occlusion using multiple coils, bags, glue, and more recently the Amplatzer devices. The most recently developed Amplatzer Vascular Plug II (AVPII), for peripheral vascular occlusions has only been used for the occlusion of coronary cameral fistulae in few published case reports.
Case 1: A 28-year-old woman presented with a history of decreased effort tolerance and episodes of chest pain on exertion. On examination she was in NYHA (cardiac failure) class II. Her pulse was 75 beats/min and was collapsing in nature. All pulses were palpable. Her blood pressure (BP) was 120/50 mmHg. A 2/6 continuous murmur was audible in the left lower sternal border. Echocardiography demonstrated a large right CCCF draining into the right ventricle. At cardiac catheterisation under general anaesthesia, a 14 mm AVPII was selected and deployed into the distal portion of the fistula with successful occlusion of the CCCF. She remains well 2 years after the procedure.
Case 2: An 8-year-old boy presented with an incidental murmur found during evaluation for an upper respiratory tract infection (URTI). He had no cardiac symptoms. On examination he had a pulse of 80/min and bounding pulses. His BP was 110/45 mmHg with a wide pulse pressure. Echocardiography demonstrated a large left CCCF draining into the left ventricle (LV). During cardiac catheterisation the fistula was successfully embolised using the AVPII.
Results: Both patients had successful transcatheter occlusion of the CCCF. They have remained well more than 18 months since the procedures.No complications were encountered.

Conclusion: We report successful occlusion of large CCCF using the AVPII. The advent of the Amplatzer vascular plug for peripheral vascular applications has added another tool in the interventionist's armamentarium for closing large coronary artery fistulae.

1835: SUBPULMONARY STENOSIS ASSESSED IN MID-TRIMESTER FOETUSES WITH TETRALOGY OF FALLOT: A NOVEL METHOD FOR PREDICTING POSTNATAL PULMONARY VALVE Z-SCORE AND SURGICAL MANAGEMENT

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Background/hypothesis: The severity of right ventricular outflow tract obstruction impacts postnatal outcomes in tetralogy of Fallot (TOF). There are no existing data relating sub pulmonary stenosis (SPS) severity in the second trimester to postnatal TOF course. We hypothesised that quantification of prenatal SPS in the second trimester would identify infants with smaller pulmonary valves who would require earlier surgery and/or undergo trans annular patch (TAP) repairs.
Materials and methods: We retrospectively identified foetuses with TOF from 1998 to 2010 diagnosed at ≤26 weeks gestation. Data evaluated included prenatal and postnatal pulmonary valve z-scores (PVZ). To quantify foetal SPS, we created a novel index, the SPS/DAO ratio, measured as the ratio of the minimum infundibular diameter to descending aorta diameter (DAO) at the level of the diaphragm. Multiple linear regression was used to predict postnatal PVZ from prenatally determined parameters, including SPS/DAO. Foetal parameters were analysed by logistic regression for association with postnatal outcomes: timing of surgery (< 1 month) and type of surgery (TAP)vs 'valve-sparing').
Results: Twenty-three foetuses met inclusion criteria. Mean gestational age was 21.8 ± 1.9 (16.6–25.4 weeks). There was excellent correlation between predicted and measured PVZ-, r = 0.82, $p <$ 0.0001, using the derived equation:

$$\frac{-3.68 + 0.91*\text{prenatal PVZ} - 4.44*\text{SPS}}{\text{DAO} - 3.19\ (\text{prenatal PVZ}*\text{SPS/DAO})}.$$

An SPS:DAO value of < 0.5 had 100% sensitivity and 56 % specificity for repair < 1 month and < 0.47 had 100% sensitivity and 75% specificity for TAP repair.
Conclusions: Postnatal PVZ can be predicted using prenatal PVZ with the SPS/DAO ratio in < 26-week foetuses with TOF. Quantification of SPS with the SPS/DAO ratio identifies patients who may require early intervention and/or TAP repair, thereby impacting prenatal counselling.

1252: RADIOFREQUENCY PERFORATION IN THE TREATMENT OF PULMONARY ATRESIA - INTACT VENTRICULAR SEPTUM: CHALLENGES FACED IN THE CATH LAB OF DEVELOPING COUNTRIES

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Background: Pulmonary atresia with intact ventricular septum (PA-IVS) is an infrequent disorder with significant morphological heterogeneity. The use of percutaneous radiofrequency (RF)-assisted perforation of the atretic valve and subsequent balloon dilation provides an easy but expensive overall procedure that is expected to establish ante-grade flow successfully through the pulmonary valve in most of patients.
Methods: Twenty patients presenting with PA-IVS presenting to Cairo University Children Hospital (CUCH) were taken for RF perforation using Baylis RFP-100 generator with trials for cost limitations. Cost limitation methods to minimise expenses included the following: we stopped using the Protrack microcatheter and replaced this in most cases with the 'wire tracks a wire' technique; we also replaced the use of the micro-snare from the arterial end to mark the pulmonary valve by looping a coronary wire; telescopic Luma catheter was replaced by using Mullin long sheath 5 or 6F through which the 4F catheter is passed; fixing the wire and trying to limit the number of balloons used, replacing the Tyshak Mini balloons which were not always available with regular coronary balloons which are readily available; if 4F sheath with special curve was unavailable we mostly used 4F VER and reshaped it.
Results: Successful opening of the atretic valve with cost limitation was successful in 70% of cases. We resorted to a hybrid procedure with the surgeon opening the chest and fixing the sheath in RV directed towards the pulmonary valve in case of failed peripheral vascular access.
Conclusion: Cost limitation is essential in our developing countries and innovative ideas to reduce cost are essential, especially if they give the same rate of success.

HEALTH SYSTEMS AND HEART DISEASE

2: CHANGING POLICY IN SCREENING LOW-RISK PREGNANCIES FOR FOETAL HEART DISEASE CAN IMPROVE EARLY DETECTION OF CONGENITAL HEART DISEASE: LESSONS LEARNT FROM AN INTEGRATED HEALTH-CARE SYSTEM

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Background: A large percentage of significant congenital heart disease (sCHD) is not detected in utero or immediately after birth. Prenatal detection rates of sCHD of around 30% have been reported, even in the current era. Utilising the electronic medical record (EMR) in an integrated healthcare system, incidence, outcome and prenatal detection rate of sCHD within a defined low-risk population was documented. Influence of policy to document foetal cardiac outflow tracts on screening prenatal ultrasound was explored.
Methods: During a six-year period, patients/foetuses under one year of age with sCHD were identified. Mother and child EMR were used to gather data. Incidence was determined using the number of live births within the defined population and time period. The outcomes, prenatal detection rate and postnatal timing of detection were documented. The prenatal detection rate of patients undergoing cardiac surgery under one year of age was compared to a similar group within the same healthcare system where there was no policy to document foetal cardiac outflow tracts.
Results: sCHD was identified in 101 patients/foetuses. The number of births within the same period was 25 666, giving an incidence of 4.0 per 1 000 live births. The overall prenatal detection rate of these infants was 74%. Detection rate prior to discharge was 95%. A significant difference in prenatal detection rates was found when there was a policy in place to obtain foetal cardiac outflow tract views in prenatal screening (58 vs 28%).
Conclusions: Within an integrated healthcare system and use of an EMR, a detection rate of sCHD of 95% can be demonstrated before hospital discharge and 74% can be detected *in utero*. A concerted programme that includes documentation of foetal cardiac outflow tracts in pregnancy screening can result in improved prenatal detection of sCHD.

60: A NATIONAL VIDEO CONFERENCING SYSTEM FOR PAEDIATRIC CARDIOLOGY

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Sweden is a sparsely populated Nordic country with a population of 10 million, with 100 000 births annually. With only 32 paediatric cardiologists and two operating centres, all located in the major cities and university departments, rural small hospitals must rely on local paediatricians for primary diagnosis and early treatment of heart disease. In order to help ensure correct diagnosis and optimal early treatment of congenital heart disease, a national video conferencing system was installed in all 36 hospitals with delivery units and paediatricians or paediatric cardiologists on call. A commercially available codec was customised for high-resolution (1 080 p) and high frame-rate (60 fps) video conferencing. Dual streaming over a secure national broadband network allowed for part-to-part conferences, or up to all 36 hospitals simultaneously participating. The Swedish Heart–Lung Foundation financed the system that was installed in 2011.

The system is frequently used by small hospitals for acute consultations with university departments and operating centres. Echocardiographic examinations are presented and discussed. Decisions are made on acute treatment, time and mode of transportation and information given to referring paediatricians and parents. Life-saving decisions have been made using the system. Follow-up out-patient visits have been made with the patient, parents and paediatrician in the remote local hospital and the paediatric cardiologist or cardiac surgeon in the university centre. The system is also used for training, clinical rounds, research and business meetings.

This telemedicine video conference system and network facilitates improvement of diagnostic skills and enhances knowledge about heart disease in infants and children, especially in the small hospitals. It gives patients and local paediatricians access to instant specialist knowledge and operating centres improved information about patients to be referred for treatment. Infants with critical heart disease arrive in a better condition and results are likely to be improved.

68: PATTERN OF CARDIAC DISEASE AMONG PATIENTS IN A PAEDIATRIC CARDIAC CLINIC, JUSH, 2012

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Background: JUSH is the only specialised referral hospital in south-western Ethiopia. Paediatric care is given by specialist and subspecialist doctors. Cardiac diseases are one of the leading causes of morbidity and mortality in children. Both congenital and acquired heart diseases are common in children. The paediatric cardiac clinic gives regular follow up for these groups of patients every Friday afternoon.
Methods: A retrospective chart revision was made in those paediatric patients who came to JUSH and were diagnosed to have cardiac illnesses and were on follow up. Data collected from a structured questionnaire was filled in by the physician at the time of chart review. Analysis using SPSS version 16 was made after clearing the data.
Result: Mean age of patients was 3.8 years with 51% females, and mean duration of follow up was 1.3 years. Echocardiography was done for 89% of patients who were on follow up in the paediatric cardiac clinic. Acquired heart disease scored the highest value, with a predominance of CRVHD. Among congenital heart disease patients, ventricular septal defect (VSD) was the leader, followed by patent ductus arteriosus (PDA) with 50 and 21% prevalence, respectively.
Conclusion: The majority of patients were receiving echocardiography at the time of follow up. The leading cause of cardiac illness in our children is still CRVHD from acquired, and VSD from congenital heart diseases. A more detailed study needs to be done to find further management options.

189: THE DEVELOPMENT OF PAEDIATRIC CARDIAC SURGICAL NURSING IN MONGOLIA

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Introduction: The mission of WSPCHS is to promote the highest quality comprehensive care to all patients with paediatric and/or congenital heart disease. As mission teams help developing countries establish paediatric cardiac surgery programmes, the mission of the WSPCHS must be extended to include nursing; realising a perfect surgical repair is not enough to assure a successful recovery.
Methods: Since 2005 the Mongolian Minister of Health has invited a team from the United States to Mongolia to perform cardiac surgery and catheterisation procedures for children with congenital and

acquired heart disease. Each year a nursing conference is conducted in collaboration with the nursing department at Shastin Medical Centre in Mongolia. Mongolian ICU nurses then partner with a visiting nurse to care for the children after surgery. The goal for the Mongolian nurses is to be fully responsible for the care provided by the end of the week.
Results: Since 2005, over 350 nurses have attended the conference and 840 hours of clinical mentoring have been provided. In addition, nursing rounds have been conducted each year to encourage the Mongolian nurses in their roles. The team has provided over 200 stethoscopes and provided assessment training and practice. The nurses, who had never used stethoscopes before our team's arrival, have used their new assessment skills with the postoperative patients.
Discussion: This abstract describes one mission team's efforts to improve the postoperative nursing care of children in Mongolia. The nurses in Mongolia have been eager to learn and have taken seriously their role in the success of a paediatric cardiac surgery programme for their country. In order to meet the mission of the WSPCHS, education and training of nurses must be included whenever cardiac surgery teams are working in developing countries.

191: MEDICAL MISSION OR OUTREACH CLINIC? OPERATIONALISING PAEDIATRIC CARDIOLOGY SERVICES WITH A PORTABLE ELECTRONIC MEDICAL RECORD (EMR) AND MEDICAL SCRIBE IN THE FEDERATED STATES OF MICRONESIA

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Background: We operationalised our paediatric cardiology services in the Federated States of Micronesia (FSM). Located in the Western Pacific Ocean just north of equator, FSM comprises 600 islands spread over an area of 1 000 000 square miles and is a medically underserved area. Previous outreach trips took five weeks and consisted of a visit with a brief handwritten entry in the patient's medical record. Our team consisted of a paediatric cardiologist, an echocardiography technician with a portable Philips CX-50 echocardiography machine, and a medical scribe for data entry. Using a portable EMR we hypothesised that we could improve the efficiency of our outreach. In addition, we sought to characterise the spectrum of congenital and rheumatic heart disease (CHD and RHD) in the region.
Methods: We created a customised mini-EMR using a Microsoft Excel© spread sheet. Drop-down lists with common diagnoses, surgeries, physical examinations, echo findings, and medications were used to fill in the spread sheet. Additional data were free-texted as needed. Data entry for each patient took under two minutes by the medical scribe and a consultation note was generated using the Mail Merge feature in Microsoft Excel©.
Results: We evaluated 328 patients on four different islands over a period of three weeks (compared to five weeks previously). The portable EMR generated comprehensive individualised notes with complete treatment plans for local providers. Perimembranous and/or supracristal VSDs are the most common forms of CHD in this population. The clinical spectrum of RHD is similar to previous studies of Asian/Pacific islanders.
Conclusions: We improved the efficiency of our paediatric cardiology outreach services to FSM using a customised portable EMR along with a medical scribe for data entry. In addition, we documented the clinical spectrum of CHD and RHD in this region.

229: INNOVATIVE ORGANISATIONAL STRATEGY FOR CRITICAL CONGENITAL HEART DISEASE

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Objective: We described an innovative organisational strategy for neonates who have prenatal diagnoses of critical congenital heart disease (cCHD) with a focus on the earliest surgical repair, using autologous umbilical cord blood (AUCB). The programme, First Hours of Life Cardiac Surgery, was innovated and applied at the Ukrainian Children's Cardiac Centre.
Methods: From September 2009 to May 2012, 350 neonates with cCHD were operated on at our institution. For patients with prenatal diagnoses, new management was proposed. This included: (1) re-examination by Echo at the 36th week of gestation to confirm the diagnosis; (2) checking the pregnant woman for blood infection and referring to the maternity department; (3) planning labour for an early morning on a working day by induction or caesarian section due to the obstetric indications; (4) harvesting of AUCB in utero and testing it at the blood transfusion service in accordance with standards for blood products; (5) immediate transfer of the newborn to the cardiac department; (6) complete primary surgical repair of cCHD in the first hours of life after specification of the anatomy; (7) using AUCB for peri-operative blood management.
Results: During this period, 47 neonates underwent the new strategy. In 27 cases (57%) labour was induced, and 20 patients (43%) were delivered through caesarian section. Mean volume of harvested cord blood was 85 ± 24 ml (50–140). Neonates were admitted to the cardiac department within an hour of birth. Mean age at operation was 3.9 ± 1.1 hours (2–6). No patients required ICU admission, interventional procedures, mechanical ventilation or medications before surgery, which resulted in significant positive economic effects, compared with the conventional approach. Thirty-seven neonates (78%) underwent open cardiac surgery without homologous blood transfusion.
Conclusion: The proposed innovative organisational strategy for cCHD allows avoidance of pre-operative ICU stay, balloon atrioseptostomy and reduction in homologous blood transfusions, and shows significant positive economic effect.

255: ATTENDING TO THE UNATTENDED: THE ESTABLISHMENT OF THE FIRST PAEDIATRIC CARDIOLOGY OUTREACH CLINIC IN UGANDA

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Background: The burden of paediatric cardiac disease in the developing world is substantial and under-resourced. Several million children in sub-Saharan Africa suffer with acquired conditions such as rheumatic heart disease (RHD) and endomyocardial fibrosis (EMF). More than 90% of these children live in areas of inadequate or absent care. Our aim was to create an outreach clinic in an under-served community in Uganda to address this care deficit.
Methods: A paediatric cardiology outreach clinic was started in 2011 in the Luwero region of Uganda. This clinic was a coordinated effort between Kiwoko Hospital (clinic site) and the Uganda Heart Institute. Funding for the clinic was established by the Uganda Heart Institute and the ISIS Foundation (a non-governmental organisation). Kiwoko Hospital supplied a portable echocardiogram machine (Acuson Cypress Portable Ultrasound) and laboratory support. Prescriptions were supported by the National Medical Stores. The Uganda Heart Institute provided skilled clinic personnel.
Results: A total of four outreach clinics took place in 2011. The mean number of patients seen was 11 (range 7–15). The cost of a single clinic day was 650 000 Uganda shillings ($263 US dollars), covering allowance for two outreach doctors, two outreach nurses, a single driver, and 40 l of fuel. Several conditions were identified, including EMF, RHD, non-pathologic murmurs, and patent ductus arteriosus. All of the patients cared for in the clinic did not have the

necessary means to pursue care outside of their local community. Follow up was successfully managed for several patients.
Conclusion: The outreach clinic represents the first-ever successful and sustained paediatric cardiology outreach clinic in Uganda. The financial cost of the clinic is not over-burdensome, and the clinic allows children to obtain much-needed subspecialty care. Efforts are on-going to expand the number of patients served and to improve the available diagnostic and therapeutic tools.

312: IMPACT OF AN INTEGRATED ELECTRONIC HEALTH RECORD SYSTEM ON PAEDIATRIC CARDIOLOGY CLINIC DOCUMENTATION AT A TERTIARY HEALTHCARE FACILITY

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Background: We hypothesised that the introduction of an electronic health record system (EHR) in an ambulatory clinic setting would significantly improve time-lines and uniform availability of paediatric cardiology consultation reports.
Methods and Results: Clinic report records of three full-time paediatric cardiologists were reviewed from four consecutive typical weeks (fully staffed for five days, not preceding prolonged physician absence of over one week) immediately before rollout of an integrated EHR and from four consecutive typical weeks one year later. Pre-EHR reports (164 in total) were prepared from transcribed dictation with subsequent editing by a physician. Documentation methods of post-EHR reports (159 in total) varied by physician. Total report preparation time (RPT) was defined as the number of days from patient visit to final signature and report release.
Conclusions: Institution of an integrated EHR at this tertiary healthcare facility significantly reduced out-patient consultation report preparation times on average over 65% (> 1 week), and markedly improved uniformity of practice among physicians, despite variable documentation methods. Prior performance did not predict results post-EHR. EHR benefits included improvements in patient care communication with positive implications for referring provider satisfaction and healthcare system fiscal performance.

402: PARENT EDUCATION AND DISCHARGE INSTRUCTION: THEMES FROM INDIA

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Background: Discharge teaching has been associated with improved patient and family satisfaction and earlier recognition and treatment of complications. Traditionally, in developing countries physicians give orders and nurses carry out those orders. The role of the patient and family discharge teaching is not typically within nurses' scope of practice. We hypothesised that Indian nurses can effectively expand their role to include discharge teaching for parents after a child's cardiac surgery.
Methods: Study participants were recruited from a tertiary paediatric heart hospital in India. Paediatric cardiac nurses and parents of children undergoing cardiac surgery were eligible to participate in the qualitative aspect of this mixed-methods study. Study participant interviews were conducted by the PI with assistance from a trained Indian interpreter and translated into English. Methodological and conceptual memos were developed and a multi-phase analysis occurred. The research team read the text and open-coded data according to themes. The thematic analysis included a group discussion for salient themes among several researchers and coding by two separate researchers. Salient themes were integrated into higher-order categories and given conceptual labels. The meanings of these categories were interpreted and validated by data illustration.
Results: Three themes were generated from the study and included role expansion, agency, and easing recovery for nurses and parents. Nurses and parents reported taking an active role in contrast to their previously passive role during a child's transition to discharge. This active role was described as satisfying to nurses and parents alike.
Conclusions: Parent teaching offers an opportunity for nurses to help parents care for their sick children.

490: IMPROVEMENT IN RESULTS AND PROGRESS OF INDEPENDENT SURGERY WITH INTERNATIONAL CO-OPERATION IN A SINGLE UKRAINIAN CENTRE

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Background: Surgery for congenital heart disease has been slow to develop in parts of the Ukraine. We describe the impact of our four-year surgical collaborative assistance programme between the International Children's Heart Foundation (ICHF) and Institute of General and Urgent Surgery, AMS of Ukraine, Kharkov.
Methods: Data were analysed from ICHF and Kharkov databases prior to and since commencement of assistance (era A: January 2000 to May 2008, era B: May 2008 to May 2012). We evaluated differences between era A and era B for: case volume per year, (± SD), 30-day/hospital discharge mortality, case complexity (RACHS-1 model), and RACHS adjusted standardised mortality ratio (SMR: observed/expected mortality). For era B, we evaluated year by year the number of collaborative operations where a Ukrainian surgeon was the primary operator.
Results: In era A, 154 surgeries were performed, mean annual case volume was 17.3 ± 4.8, with an overall mortality of 4.55% and an SMR of 3.6. RACHS category 1 comprised 75% of the total. In era B, 361 surgeries were performed. Mean annual case volume increased to 90.2 ± 30.9 ($p < 0.001$) with higher case complexity, and an overall mortality of 5.82% and SMR of 1.6. In era B, 237 surgeries were performed during 16 trips, 124 between trips; 140/237 (59.1%) cases were led by a Ukrainian surgeon with either the visiting surgeon (87/140) or local surgeon assisting (53/140).
Conclusions: An assistance partnership in the model applied significantly reduced mortality, increased case volume and complexity, and developed independent operating skills in an economically disadvantaged centre in a relatively short time period. This model of assistance to developing countries is not 'surgical tourism', and should always be open to scrutiny and evaluation by proven clinical and educational outcomes.

494: REAL-TIME MOBILE TELEMEDICINE USING SCALABLE VIDEO CODING FOR NEONATAL HEART DISEASE

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Background: An efficient system of real-time telemedicine requires being applicable to a variety of devices and networks that consulting medical providers utilise. Scalable video coding (SVC) enables a video stream to be broken into multiple resolutions, quality levels and frame rates, and to be flexibly conformed to device capabilities and network conditions. We evaluated the applicability of H.264/SVC to neonatal tele-echocardiography and devised a system of real-time mobile telemedicine using SVC for underserved and disaster regions in east Japan.

Methods: Echocardiograms of newborns with critical congenital heart disease with resolutions of 640 × 448 and frame rates of 30 fps were encoded into two layers for scalability with resolutions of 640 × 448 and 320 × 224. Four sets of bit-rate were tested; no compression, 2 Mbps, 1 Mbps and 0.5 Mbps. Images were coded by VPN system and randomly transmitted to decoders in three device/network conditions; PC/local network, PC/wide area network (WAN) and tablet/WAN. Fifteen blinded board-certified paediatric cardiologists subjectively assessed images and scored between 0, i.e. unsuitable for diagnosis and 1.0, i.e. compatible with normal studies. Subjective assessments were compared with objective quality metrics peak signal-to-noise ratio (PSNR).
Results: In PC/WAN condition, SVC images with resolutions of 640 × 448 required bandwidths of more than 1 Mbps to get average scores of 0.5 or more. At bandwidths less than 1 Mbps, scores for images with resolutions of 320 × 224 were significantly higher than values for 640 × 448 images (0.47 vs 0.27). Images in tablet/WAN condition at 1 Mbps scored 0.42. Subjective assessments were significantly correlated with PSNR. An application (Vidyo Inc) using SVC showed successful transmission of images with 950 × 540 and 30 fps at 768 kbps on mobile devices in a pilot LTE environment, allowing interaction between participating medical staff.
Conclusion: The real-time mobile system using SVC may be useful for neonatal tele-cardiology in the unreliable wireless network.

539: SAVE A CHILD'S HEART: SPECIFIC ETHICAL QUESTIONS IN A CHARITY PROGRAMME

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Save a Child's Heart is a charity programme that has treated over 2 900 paediatric cardiac patients from 43 underdeveloped countries during the past 17 years. Throughout the years, many ethical questions have arisen. The purpose of this presentation is to bring up these questions, describe our approach to solving them and open the subject for further discussion.

A relative paucity of available openings for treatment due to both money restraints and bed limitations imposes ethical problems that are not faced by most programmes. Patients' acceptance to the programme induces a conflict between the referring and accepting systems due to the medical severity of cases, chances of success and, sometimes, due to secondary benefits. The feasibility of further medical follow up needs to be considered before acceptance to a charity programme, especially if there is a potential need for a prosthetic valve or pacemaker and the possible continuation of expensive medications. A very important issue is whether patients should be admitted for palliative treatment, considering the availability of future surgeries and catheterisation as a part of the planned repair of the specific patient. As a paediatric charity, what is our commitment to adults who were treated by us as children?

Air travel is a risk for cyanotic children. Is there a benefit in a medical escort on the fight and if one is not available should we avoid transferring cyanotic children for treatment? In charity programmes where the parent and physician have a language and cultural barrier, the meaning of consent forms is questionable. There is a relative absence of parental supervision and when parents have no choice, quality control may be jeopardised. So far we have dealt with all these questions in accordance with our personal and cultural morals and attitude. These dilemmas are open for different approaches and merit further discussion.

585: HOSPITAL VARIATION IN POST-OPERATIVE INFECTION AND ASSOCIATED OUTCOMES FOLLOWING CONGENITAL HEART SURGERY

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Background: While previous studies have demonstrated the association of post-operative infection with morbidity and mortality following congenital heart surgery, variation across hospitals has not been well described. This study evaluated hospital infection rates across a large multi-centre cohort, and association with other hospital-level outcome measures.
Methods: The Society of Thoracic Surgeons Congenital Heart Surgery database was linked to resource utilisation data from the Pediatric Health Information Systems Database for hospitals participating in both databases (2006–2010). Hospital infection rates (sepsis, wound infection, mediastinitis, endocarditis, pneumonia) adjusted for patient risk factors, case mix and delayed sternal closure were calculated using Bayesian methodology. Association with hospital mortality rate, post-operative length of stay (LOS), and total costs were evaluated.
Results: The cohort included 32 856 patients (28 centres). Across hospitals, the adjusted infection rate varied from 0.9 to 9.8% (median 4.1%). The most common types of infection were sepsis (51%) and wound infection (35%). On a patient level, infection was associated with increased mortality (OR 2.8, 95% CI: 2.2–3.6, $p < 0.001$), prolonged LOS (25.5 vs 11.2 days, $p < 0.001$) and increased hospital costs ($115 800 vs $63 300, $p < 0.001$). Similar results were observed when hospitals at the extremes of infection rates were excluded. Hospitals were divided into tertiles according to adjusted infection rate. Hospitals with the highest infection rates (vs lowest) had longer average LOS (13.2 vs 12.0 days, $p < 0.001$) and hospital costs ($70 900 vs $58 200, $p < 0.001$), but no significant difference in mortality (OR 0.9, 95% CI: 0.7–1.1, $p = 0.2$).
Conclusions: Post-operative infection following congenital heart surgery contributes to prolonged LOS and increased costs on a hospital level. Initiatives aimed at reducing post-operative infection may reduce variation and improve outcomes across centres.

702: TRACKING CARDIOVASCULAR MORBIDITY: UTILITY OF A HAND-HELD DEVICE TO MONITOR CARDIOVASCULAR COMPLICATIONS

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Background: In this era of declining cardiovascular mortality, other measures of quality assurance (QA) become important. We have previously described the utility of a hand-held device to track complication rates (chosen from a pre-defined pick list including all body systems) related to cardiac procedures [cardiac catheterisation, closed-heart surgery or open-heart surgery (OHS)].
Objective: To record the complication rate for OHS and document variation related to changes in clinical practice.
Methods: Five cardiologists used Palm OS-based software (Smart-

list to Go) to enter all procedures and related complications. Data were analysed over five years, the last two years encompassing changes in surgical and peri-operative practice aimed at facilitating early extubation and decreasing ICU and hospital length of stay (LOS).
Results: Complication rates were expressed as total number per 10 OHS, and rates were summarised quarterly with 95% confidence limits (CI). In the past two years, complications have decreased significantly from 5.2 (CI 4.6–5.8) per 10 procedures to 3.0 (CI 2.4–3.6) per 10 procedures ($p < 0.0001$). The rate in each era has remained stable and, despite an increase in case complexity in the recent era, the ICU (6.3 vs 2.6 days, $p < 0.0001$) and hospital LOS (12.6 vs 6.2 days, $p < 0.0001$) have decreased.
Conclusions: Implementation of new strategies for reducing patient morbidity, ICU and hospital LOS has been successful. Control charts with quarterly rates for all complications were readily derived, and these allow estimation of current morbidity trends. The effect of QA processes can be assessed in a timely manner.

772: BEATING THE ODDS: AN ORIGINAL ADVENTURE IN FAVOUR OF MEXICAN CHILDREN WITH CONGENITAL HEART DISEASE

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Mexico has a total population of 112 336 538, a growing population index of 1.40, and every year 21 151 babies are born with congenital heart disease, our second cause of child mortality before the age of five. We have 10 public hospitals that can perform paediatric heart surgery but with more than 50 million people living in poverty, all the public hospitals are overcrowded, with not enough infrastructure or human resources to give appropriate attention to our heart patients. An estimation of the number of annual surgeries that are performed on paediatric patients per year in public and private hospitals hardly reaches 25% of the children that are at risk. Some children are born into a family that can pay for medical attention in a private hospital, but the majority of Mexicans have no medical cover or social security. These families need our help the most.

We face a serious health problem that is difficult to change. On one hand we have public hospitals that have many heart patients who cannot pay for private medical attention, or, because of the large number of cases treated, cannot be cared for by best heart specialists. On the other hand, private hospitals have money to spend but few cases to work on, and do not support the huge investments required for medical equipment, human resources and training. We decided to transform this situation, joining three groups together: a private hospital, a government-funded paediatric hospital and a foundation dedicated to the medical care of children with congenital heart disease. We share our strategy, programme, and how we have started to make it work.

888: CAN A COMPREHENSIVE CONGENITAL HEART PROGRAMME BE DEVELOPED IN RUSSIA WITH LIMITED RESOURCES AND MAINTAIN QUALITY OUTCOMES?

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Background: Worldwide, less than 15% of children born with congenital heart disease have access to appropriate medical care. The purpose of this study was to evaluate the effectiveness of the Heart-to-Heart International Children's Medical Alliance's multidisciplinary training programme to improve the care of children with congenital heart disease in Russia by creating sustainable centres of excellence.
Methods: The process begins with site assessment encompassing evaluation of medical, administrative, governmental support and desire for the programme. It is followed by a multi-year commitment to the US–Russian training programme consisting of: (1) strategic guidance and team building: year-round guidance on programme development, and specific recommendations for the 'next steps'. (2) Data collection and analysis: to provide detailed feedback to improve results. (3) Annual surgical educational missions: the Heart-to-Heart team works side by side with Russian colleagues to diagnose, perform cardiac surgery and catheterisation, and provide post-operative care. (4) Scholar exchange: Russian physicians travel to centres of excellence in the USA or Russia, and attend international conferences. (5) Continuing education: professional educational materials, participation in US journal clubs, and attending medical conferences.
Results: Composite outcome data from two centres (Samara and Tomsk) are presented. Over a five-year time frame, the total number of cardiac operations increased from the baseline 186 in year one to 514 in year five; the complexity of cases RACHS 3–6 increased from 11.5 to 26.5%; children less than 12 months of age at the time of surgery increased from 21 to 46%; mortality decreased from 14 to 4%. The median cash expenditure per site per year was 99 612 USD (range 73 977–182 030 USD).
Conclusions: The goal of creating a comprehensive, sustainable programme to care for children with congenital heart disease can be accomplished in a fairly short period of time with modest financial investment. The individualised, multidisciplinary training and educational strategy developed by Heart-to-Heart International Children's Medical Alliance has proven successful in increasing patient complexity over time, with excellent surgical results.

946: SAVE A CHILD'S HEART: 17 YEARS OF ACTIVITY

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Save a Child's Heart (SACH) is a hospital-based, non-governmental organisation founded by Dr Ami Cohen in 1994 at the Wolfson Medical Centre in Israel. Its goal is to improve the cardiac care of children with heart diseases from developing countries. This is achieved through two main channels: (1) treating children with heart disease through surgery and/or cardiac catheterisation in Israel, and (2) training medical personnel from partner countries. Over 2 900 children have been treated so far and over 70 physicians and nurses have been trained. The main mode of action is by direct cooperation with a medical facility trying to help their patients.

Fifty per cent of our patients are Palestinians, referred by their local physicians to our free clinic for further treatment. With regard to children from our overseas partner countries, we travel to the country to screen patients, discuss their problems with the local team and the child is wait-listed to be brought to the SACH centre in Israel. In addition, we offer training positions in an attempt to build a local team who will treat their own patients. As a part of this endeavour, we also go on surgical missions to the partner country and operate together with the local team. Individual patients who contact us via different channels are also accepted according to feasibility of treatment. The presentation will describe our activity during the past 17 years, the structure of the organisation, mode of action, problems and achievements.

1014: STATE OF THE ART HUMANITARIAN PAEDIATRIC CARDIAC MISSIONS: A DESIGN FOR SUSTAINABLE DELIVERY OF ASSISTANCE

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Background: Paediatric cardiac development assistance is widely but sporadically practiced. The delivery of more than one assistance mission per year is beyond the ability of many centres. We describe an organisational structure enabling the sustained delivery of assistance through an NGO-based multi-national, multi-institutional team-member approach. Our results over the last 20 years are presented.
Methods: The database of the NGO was reviewed since inception in 1993. Totals were harvested for trips made, sites of assistance, number of team members, country and institution of origin of team members, and number of operations. Data were analysed by five-year periods (eras) for differences. A p-value of < 0.05 was considered significant
Results: Total number of years of assistance was 20. Total team members were 3 578 on 289 trips. Team members came from 106 institutions in 43 countries. The average number of trips and team members/year increased between all eras ($p < 0.001$) from 3.6/48 in era 1 to 29.0/401 in era 4. The maximum number of trips in one year was 36, requiring 509 team members. The number of institutions and countries of origin of the team members increased over time and the average was significantly different between eras ($p < 0.01$). The single largest number of institutions and countries represented on a trip was 13 and 11. Previous recipients of assistance became team members in 2000. A total of 26 countries have received assistance, with the greatest number of trips and operations provided in Central America and Asia, at 61/1 132 and 58/1 128, respectively.
Conclusions: Paediatric cardiac education and service can be provided to multiple sites simultaneously utilising this model. We are not aware of a single hospital, institution or charitable entity that can provide a similar level of assistance

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1024: NECESSITY OF BIO-MEDICAL ENGINEERING SUPPORT ON HUMANITARIAN MEDICAL MISSIONS

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Background: Due to less complex technology for medical equipment in the past, there was little emphasis placed on the importance of biomedical engineering support for humanitarian medical missions. However, as the evolution of technology for medical equipment has progressed at a rapid pace, and the complexity of procedures has increased, it is evident the role of biomedical engineering has expanded. An education in this field is not enough. A discipline in computer engineering, anatomy and physiology, and knowledge of equipment that is procedure specific must be incorporated. In under-developed countries, it is quite common for these disciplines to simply not exist.
Methods: Data were analysed from ICHF's (International Children's Heart Foundation) database over the past five years. During this period, a total of 677 pieces of medical equipment was sent to 23 locations in 16 different countries. Some specific types of medical equipment included patient monitors, anaesthesia machines, cardiopulmonary bypass machines, ventilators, defibrillators, electro-surgical units, syringe pumps, hypo/hyperthermia units, and cardiac echo ultrasound units. Additionally, the ICHF biomedical engineering staff/volunteers have made 49 trips to provide biomedical engineering support and emergency repairs.
Results: The analysis provides a common link between the various types of equipment needed across all developing countries where humanitarian medical programmes have been started.
Conclusions: The ICHF has created a paradigm shift and raised the bar of expectation on the level of education and expertise for biomedical engineering support. This support is defined as installing, servicing, repairing and providing staff training on biomedical equipment. The end result has allowed ICHF medical programmes in developing countries to grow at an accelerated pace, by ensuring better patient safety and improving surgical results

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1116: RHEUMATIC HEART DISEASE HEALTH WORKER TRAINING AND SYSTEM STRENGTHENING IN FOUR PACIFIC ISLAND NATIONS

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Background: The Pacific region has the highest documented rheumatic heart disease (RHD) prevalence globally. Since 2005 the World Heart Federation and Menzies School of Health Research have been working to strengthen rheumatic heart disease prevention and control with Pacific Island nations. Additional funding was secured in 2011 to assist four additional Pacific countries, Nauru, Tuvalu, Kiribati and the Solomon Islands.
Methods: The programme is based on WHO international recommendations for register-based prevention and control. An Australian-based team assists local staff from the Ministries of Health to expand capacity and strengthen existing activities.
Results: Health professionals from the four Pacific countries have received RHD training. Each country has been supported to develop clinical information materials, patient injection cards and national protocols for the diagnosis and prevention of ARF. A regional RHD trainer's manual has been developed to support local staff to extend RHD training to health workers on the outer islands and provinces. National registers have been developed for each country. In Nauru, 2011 baseline data showed that 46% of patients were receiving 50% or more of their injections. Echocardiography screening to define baseline burden of disease has been conducted, showing a prevalence of 15.1/1 000 in Nauru and 35.1/1 000 in Tuvalu. Screening will be conducted in the Solomon Islands and Kiribati in 2012–13. A community and patient-education campaign plan has been developed, which includes: patient–peer support groups and multi-media campaigns to raise awareness of ARF and RHD.
Conclusion: The Pacific RHD programme has seen an increase in disease notification and awareness among health staff and the community. It is anticipated that system and capacity strengthening will contribute to a more sustainable programme, including a more efficient and user-friendly service for patients and an increase in the delivery of secondary prophylaxis in patients, and improve primary healthcare for RHD patients.

1162: DONOR EXPECTATIONS IN PAEDIATRIC CARDIAC SURGERY: ARE THE SELECTION CRITERIA JUSTIFIED?

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Background The purpose of this research was to determine the best practice in case selection to ensure that the use of donor funds is effective for sustainable and maximum impact. Bearing in mind that the objective of donors is to save as many children per donated rand as possible, it is therefore incumbent on the Foundation's selection

committee to select cardiac cases that are correctable with a good prognosis and outcome. The researcher created a strategy to guide the members of the selection committee in their selection of cardiac beneficiaries. The cases selected should have predictable, good outcomes that meet the donors' expectations and achieve sustainability for the Foundation.
Methods: The research question is: How can donor funding be allocated in a fair, evidence-based and sustainable way to achieve maximum beneficial impact on indigent children requiring cardiac surgery in Africa? I chose to do a focus group and held a question-and-answer group interview and discussion with 15 people selected from all of the stakeholder parties in the Foundation.
Outcomes: The key outcomes of the focus group included the need for surgical hubs to be developed in Africa and skills transfer to occur, the correct diagnosis being key to the outcome of the cases selected, the selection of complex cardiac cases directly impacting on the costs involved, and the appropriate use of donor money being in keeping with the avoidance of selecting cases that may have difficult outcomes and extended length of stay in hospital.
Conclusion We want WSPCF to make a real difference in the lives of children with cardiac disease on the African continent. The result of this study shows that the current selection criteria are sound and meet the donor expectations.

1168: CHALLENGES IN IMPLEMENTING A PAEDIATRIC CARDIOVASCULAR HOME TELEHEALTH PROJECT

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Background: Infants with single-ventricle physiology are at high risk of sudden death following discharge (8–12%) despite palliative management, presumably due to intravascular volume depletion, shunt occlusion, and/or arrhythmia. Therefore, we developed a pilot home telemedicine project to evaluate the feasibility of using web-based daily reporting of clinical parameters to reduce unscheduled events and death.
Methods: Subject enrollment began following IRB approval. We enrolled six single-ventricle subjects following palliation over a 12-month interval. Parents electronically recorded and transmitted daily weight and peripheral oxygen saturation data, then completed an automated 10-point phone questionnaire related to nutrition, activity and distress. Subject enrollment in the project continued until a second palliative surgical procedure ($n = 4$) or sudden death ($n = 2$). All transmitted data were managed on a customised website with hardcopy backup. We collected comprehensive clinical data in all enrolled subjects and 12 historical controls. We analysed subject clinical management including success in out-patient telephone surveillance, scheduled and unscheduled office and emergency room (ER) visits, hospitalisations, procedures and adverse events, including death.
Results: Subject recruitment was more difficult than expected. We found a high success rate in transmitting subject weights but poor correlation between oxygen saturation values measured by the study saturation monitor and monitor provided to discharged patients (Massimo). Success rate for out-patient telephone surveillance for historical controls, independent of this telemedicine project to date was approximately 30%. After technical adjustments, all enrolled subjects (100%) were able to transmit questionnaire data. There were 14 unscheduled ER visits for controls versus two ER visits for study subjects. Sudden death occurred in one of 12 controls and two of six enrolled subjects.
Conclusion: Home telemedicine monitoring for high-risk patients with congenital heart defects is feasible, may reduce unscheduled visits, but may not impact on the primary endpoint of preventing sudden death.

1587: PREPARATION OF THE VISITING TEAM FOR TECHNOLOGY TRANSFER TO THIRD-WORLD COUNTRIES

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Background: The visit of a medical team for the purpose of technology transfer from the developed world is a useful tool in capacity building of cardiologists and cardiac surgeons in Third-world countries. Such visits, when taking place between two countries with diverse socio-economic conditions, involve a number of difficult issues. The visitors are often not aware of these facts, which reduce the effectiveness of their dedication and effort. Between 1996 and 2012, we had the experience of hosting 15 foreign teams or individuals in Bangladesh for technology transfer. Various arrangements had to be made for them. Sharing those experiences through this article may be of interest for the future visitors.
Methods: Hospital records served as the major source of information. The involved doctors, nurses and other staff were interviewed. We also contacted the visitors for their comments. After collection of all data, we discussed our own experiences and compiled it in an organised manner.
Results: The important issues for the visitors, identified by our research are addressed under the following headings:

- Selecting a suitable site for the visit
- Need assessment of the recipient institute
- Travel and immigration
- Government permission and registration issues
- Safety and security
- Arranging logistic support
- Patient selection and management
- Local hospitality and team healthcare
- Organising technology transfer
- Budget and finance
- Press and media
- Conclusion and reporting.

Conclusions: Visiting a Third-world country for the purpose of technology transfer is often not an easy job. There are a number of issues making it a highly specialised subject. Analysing our experience, we have identified some key features for making such trips successful. This may be useful for future visitors preparing for such missions.

1613: COST-EFFECTIVENESS ANALYSIS OF CONGENITAL HEART SURGERY IN DEVELOPING COUNTRIES

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Background: Of the estimated more than one million children born worldwide every year with congenital heart defects (CHD), more than 95% are born in developing countries with inadequate access to surgical repair. Since surgery for CHD seems expensive, it may be given a lower priority in the allocation of scarce economic resources. The World Bank uses $100 per year of life saved as the marker for highly cost-effective interventions in poor countries. Cost-effectiveness analysis is warranted.
Methods: Total cost data were gathered for the first five years of a congenital heart surgery programme in a developing country. Cost per case included local costs plus the value of donated medical supplies sent to support the programme. Local costs were obtained from the business plan of the local foundation and included all costs, including paying the hospital, the physicians, and all administrative

costs of the programme, assuming an annual case volume of 100 operations, and including pre-operative evaluation and postoperative care. Cost per year of life saved was calculated based on known average life expectancy of less than two years for unoperated CHD, versus an estimated postoperative life expectancy of 40 years, minus our reported mortality of 7%.
Results: The value of the donated medical supplies was $190 329 over five years. Total local programme costs averaged $216 600 per year, making overall costs $2 800 per surgery or $3 011 per survivor. Cost per year of life saved is $75.28.
Conclusion: The cost of surgery for CHD falls well within the World Bank's definition of highly cost-effective interventions. Cost of surgery for CHD compares favourably with other interventions such as BCG vaccination for children, condom distribution to attempt to prevent HIV, and general/trauma surgery. Future work should attempt to incorporate disability data and cost of medical care for unoperated children.

1614: IS IT DANGEROUS TO LIVE ON AN ISOLATED ISLAND IF YOU ARE A CHILD WITH CONGENITAL HEART DISEASE WHO NEEDS OPERATION?

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Background: Sweden has very good results for paediatric cardiac surgery with a 30-day mortality of < 2%. These results depend on a functioning chain of care from the first suspicion of a congenital heart defect (CHD) to primary examination, transportation, operation, perioperative care and follow up. In order to achieve safe treatment and to optimise long-term prognosis, all these links have to be efficient and of high quality all the time and for inhabitants in all geographic locations. Sweden has nine million inhabitants and 21 counties. The island of Gotland, with its isolated location in the Baltic Sea is the smallest county with approximately 57 000 inhabitants and 500 deliveries/year.

The paediatric cardiac service is provided by a paediatrician with additional training in paediatric cardiology, and a radiologist performing echocardiography. The hospital collaborates with the Children's Heart Centre in Lund for tertiary-level care, mainly paediatric cardiac surgery and interventions. In addition, a senior consultant from Lund visits Visby twice a year. The aim of this study was to validate the results of paediatric cardiac care in Gotland.
Methods: Retrospective studies were done of hospital files in Visby and Lund for children (< 18 years) born between 1 January 2000 and 31 December 2009, who had undergone treatment for cardiac conditions.
Results: Thirty-four children underwent surgery (31) or catheter treatment (16) (one to five per patient). The median age at operation was 4.6 months (range four days to 16.9 years). There has been no surgical acute or late mortality during follow up (three to 18 years). Forty children visited the centre on a total of 63 occasions: 1.5 visits/patient (range 1–6); 25% of the visits were cardiac investigations such as MRI or diagnostic catheterisation.
Conclusion: Care of children with congenital heart disease can yield excellent survival rates, even in an isolated area such as the Island of Gotland. Close collaboration between all health professionals on the local level and between the local hospital and the tertiary-level centre are cornerstones for high quality.

1623: MULTINATIONAL, MULTILINGUAL VOLUNTEER TEAMS OPTIMISE DELIVERY IN PAEDIATRIC CARDIAC SURGICAL ASSISTANCE MISSIONS

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Background: The International Children's Heart Foundation (ICHF), a US-based non-governmental organisation (NGO), has operated on over 6 000 children in collaborative educational programmes in 45 cities, in 35 developing countries since 1993. The majority of programmes receive three or more trips per year in a partnership of five or more years, which has been shown to be effective in increasing surgical numbers and complexity. W e described the origin and numbers of our volunteer pool and drew conclusions about the sustainability of our assistance model.
Methods: We analysed the ICHF volunteer database from 2012 back to 1996. Of interest was city of origin, country of origin and volunteer participation from previously assisted programmes.
Results: Over 16 years there were 260 trips, 1 124 volunteers, originating from 185 departure cities. Analysis of contributing institution was not possible with accuracy but we can confidently infer that this volunteer pool came from more than 185 contributing cardiac centres, since many cities have more than one cardiac programme. Volunteers were of 42 separate country origins, with 350/1 124 (31%) volunteers as non-US volunteers. Therefore a significant pool of volunteers from close-proximity Spanish- and Russian-speaking countries enabled cost efficiency and a working practice in the local language. Of the 45 assisted sites, 11 eventually became volunteer contributors to teams, with 66 volunteers from such programmes contributing the knowledge and skills developed during our mutual partnership to newer programmes.
Conclusions: A mixed, multicentre, multinational volunteer pool, as presented above, enables the delivery of multiple annual assistance missions without excessive strain on any single centre. Furthermore, it enables targeted optimisation of volunteers for location and language skills, and sustains an ongoing partnership with 'graduated centres' by utilising their staff as volunteers.

1624: DATA MINING WITH NATURAL LANGUAGE PROCESSING FOR A PEDIATRIC CARDIOVASCULAR PROBLEM LIST

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Background: A problem list is a basic building block to ensure effective medical management of children with cardiac diseases by facilitating electronic health record (EHR) use. We hypothesised that problem lists can be generated from discharge summaries using natural language processing (NLP).
Methods: We identified concept-unique identifiers (CUIs) for each diagnostic term using Metamap software with standard settings. After analysing Metamap output in the paediatric cardiovascular domain, we extracted noun, verb, prepositional and adjectival phrases using the Stanford parser, and weighted each phrase by occurrence frequency and overlap with the Paediatric Cardiac Care Consortium (PCCC) diagnosis list augmented with acronyms and lexical variants. The weighted-phrases method was applied to discharge summaries from 884 patients using the Biomedical Information Collection and Understanding System (BiomedICUS). A random sample of 100 records was manually analysed to compare output of the weighted-phrases method to primary cardiac diagnosis.
Results: The Metamap analysis found 982 terms, of which 345 had cardiovascular meaning; of these 205 did not map to any CUI. From this we concluded Metamap was neither sensitive nor specific for identification of paediatric cardiovascular diagnosis. The weighted-phrases method found the primary cardiac diagnosis in the highest-weighted phrase (report line 1) in 87/100 subjects (87% sensitivity). Although 10 to 20 phrases per patient were available, these added little to sensitivity beyond the first five. Sensitivity with the five highest-weighted phrases was 97% for primary diagnosis.
Conclusion: The low rate of CUI mappings to the PCCC diagnostic list using Metamap indicates inadequate inclusion of paediatric cardiac diagnoses in standard biomedical terminologies. Our NLP method using phrases weighted by frequency and overlap with a paediatric

cardiac diagnostic terminology allows BiomedICUS to accurately generate diagnostic phrases from discharge summaries. This early use of NLP in the paediatric cardiovascular domain offers promise to facilitate EHR implementation for these patients.

1625: A NEW CARDIAC SURGERY PROGRAMME IN ASWAN: WORKING TOWARDS CREATING A SUSTAINABLE HUMANITARIAN ACADEMIC CENTRE OF EXCELLENCE

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Background: Until recently, no paediatric cardiac services were available for 20 million people in upper Egypt and adult services were difficult to access. The Aswan Heart Centre started in April 2009. It provides care free of charge and aims to become an academic and clinical centre of excellence.
Methods: Surgical activity between the start-up of the Centre and the end of July 2012 were retrospectively reviewed.
Results: The total number of procedures performed was 761. Initial operations took place during missions by international teams who created a culture of training and knowledge transfer. With their on-going support, the local team increasingly performed operations, resulting in more continuous activity. A large proportion of patients underwent surgery for rheumatic valve disease, aiming at reconstructive procedures. Surgery for congenital heart disease dealt with advanced cases in children and adults, but increasingly also in small children, including neonates. A HOCM service was established, and over the last 18 months coronary revascularisation commenced. Intra-operative TEE was used routinely with a low threshold to revise repair if the technical result was suboptimal. Post-operative patients were initially managed by early extubation and ICU fast-track. Sicker patients and neonatal surgery required further development of ICU. In spite of increasing complexity and number of small children, mortality has remained below 5%. Outpatient and imaging services were established. Research and teaching are an integral part of the service.
Conclusions: A shared vision and on-going collaboration between visiting experts and the host team facilitate the development towards a sustainable centre of excellence.

1650: A CARDIOSTART (CS) PERFORMANCE AND COMPLIANCE SCORING SYSTEM FOR EMERGING PAEDIATRIC AND ADULT CARDIOTHORACIC AND VASCULAR HEALTHCARE PROGRAMMMES

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Background: Paediatric cardiac surgery is especially challenging in emerging healthcare programmes that are personnel and/or equipment deprived; priorities include improving outcome, reducing waiting time and teaching new techniques. No uniformity in structure or technique exists to ensure that best standards of cardiological surgical management are implanted; outcomes may not reflect individual or team competence or programme worthiness. Local teams may be dazzled but afterwards, overwhelmed and financially depleted. We have devised a dual-language performance-score (part of a full international database evaluation already in operation, UMN & OHSU, USA) to monitor and assist various systems improvements.
Methods: Visiting and local team performance during the peri-operative period was evaluated discreetly, without direction or encouragement, using a scoring system addressing admission, transfer from intensive care and discharge. Admission scoring included: use of medications, dental checks, antiseptic/antibiotic use, laboratory tests, consent, allergic status, 'time out', and completion of CS's anaesthetic and perfusion checklists. Transfer from intensive care scoring evaluated airway management, respiratory rate, oxygen requirement, chest X-ray, ECG/monitor normality, bleeding, serum electrolytes, neurology and central line status. Discharge from hospital scores evaluated exercise limit checks, incision and electrolyte status, completeness of charts, communication with family members and readiness for discharge.
Results: Total possible scoring for admission (19), transfer (38) and discharge (23) (total = 80) were similar and above 80% among most 128 patients evaluated. Minor differences between countries did not impact on peri-operative management or outcome. Local doctors responded well to the structured check list, which helped self-organisation.
Conclusions: CS scoring encourages better discipline in patient care and transfer, helps identify shortcomings, assists teams to evolve components of peri-operative management that are vital to success and a platform to help programme building. This will be refined in the future by statistical analyses to develop the scores that are predictive of survival and positive outcomes.

1667: PEDIATRIC CARDIOLOGY IN PUBLIC HEATH: A TELE-NETWORK COVERING OVER 55 000 KM² OF UNDERSERVED REGIONS IN BRAZIL

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Objective: To describe the initial results from a paediatric cardiology network set up to reverse a picture of high mortality and morbidity from CHD in a population of around 2.5 million poor people.
Methods and Results: This prospective, multicentre project was divided into stages, including problem definition, network structure, and training and initiation of medical services. Thirteen institutions have been linked and equipped with iPads, pulse oximeters and portable echo machines. Physicians from the referral centre travelled once a week to perform key activities such as cardiac surgery, and local teams were supervised by telemedicine. Between January and July 2012, 10 858 pulse-oximetry tests were performed, 554 (5.1%) were considered abnormal and 217 babies underwent a screening echo; 138 anomalies were detected. The relationship between collected/abnormal oximetries decreased from 17.2% in January to 3.1% in July, suggesting a learning curve in the use of the test. The number of screening echoes increased over the time of the study. The network has clinically evaluated over 2 500 patients and 74 surgeries were performed, with seven deaths. Previously, most of these children had lengthy stays on waiting lists to be referred outside the State for diagnosis and treatment.
Conclusions: Diagnosing and managing children with CHD is a major challenge in developing countries. The establishment of partnership programmes between reference and primary and secondary centres with the aid of telemedicine is one way to ameliorate this problem. Careful planning, training, supervision and performance of key activities by trained personnel are requirements to achieve successful results.

1676: STATUS OF PAEDIATRIC CARDIAC SURGERY IN THE LEAST-DEVELOPED COUNTRIES

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Background: The least-developed countries (LDCs) are a group of countries that have been identified by the United Nations in terms of their low gross national income (GNI), weak human assets and high degree of economic vulnerability. As per 2011 statistics, the average GNI in these countries is US$ 567, far below the world average of US$ 9 488. Currently there are 36 countries in this category, mostly in Africa and Asia. Paediatric cardiac surgery in these nations is poorly developed or totally absent. The developed world has a responsibility to recognise the deficiency and to help the growth of cardiac surgical facilities in these countries.
Methods: Cardiac surgeons and cardiologists of these 36 countries were contacted through e-mail, phone calls and postal questionnaires to gather information about the status of paediatric cardiac surgery in the respective countries, with an emphasis on quality, cost and number, if any. The cardiac societies and national heart foundations were also approached for information.
Results: The estimated total population of these countries is 816 810 477, which is 11.7% of the world population. Their combined GDP is only 0.6% of the world GDP. This unfair distribution of wealth makes these countries vulnerable in terms of healthcare facilities. Facilities for paediatric cardiac surgery are either absent or primitive but where present, the cost of surgery may be surprisingly low, sometimes as low as US$ 1 000.
Conclusions: LDCs are part of the same world. This unfavourable economic situation deprives 11.7% of the world's population of this expensive modality of cardiac treatment. Cardiac surgeons from the developed world should help grow the facilities in the LDCs. They can take advantage of relatively inexpensive surgery and boost growth, thereby saving thousands of lives in these parts of the global village.

1677: A BOTTOM-UP MULTIDISCIPLINARY APPROACH TO IMPROVE PAEDIATRIC CARDIOLOGICAL CARE IN SWEDEN

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Background: Paediatric cardiac surgery in Sweden is concentrated at two centres (paediatric cardiology is practised in about 30 hospitals) and the results of surgery regarding survival are excellent, with a mortality rate of below 2%. In order to promote survival and good quality of life, collaboration between all levels of care is mandatory, however distances, diagnostic ability and accuracy, follow up and resource use varies greatly within the country. Every third year, a multi-professional (including representatives from the parents' association) meeting is held, with the aim to improve communications and cooperation in different levels of care. At the fourth meeting in 2011 a new approach was tried in order to promote national collaboration, inter-disciplinary work and evidence-based care.
Methods: Four areas of concern were defined: follow up, communication, education and incoming referrals. Locally used guidelines, healthcare programmes and 'tips and tricks' were presented on a public webpage. About 150 persons (of approximately 300 possible), mainly physicians and nurses, but also medical secretaries, biomedical laboratory scientists, physiotherapists, dieticians, occupational therapists, psychologists and social workers attended this three-day meeting. The congress' main part consisted of multi-professional workshops, with the objective to share and develop material and improvement ideas. The workshops were led by professional facilitators and the results were further discussed in a plenary session, and plans of actions were created.
Results: Thirty specific topics were defined, called 'heroic deeds' and for every topic a leader with responsibility for the continuing progress was defined. Seven new professional networks were also created.
Conclusion: With a bottom-up approach, involving different professions nationwide, it is possible to create a foundation for national guidelines and standardised care, which has the potential to make better use of resources and increase the quality of care for children with heart disease.

1688: FIVE CENTRES, ONE HEARTBEAT: A NETWORK APPROACH TO HEALTHCARE DELIVERY

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Background: For more than 10 years, the Western Canadian Children's Heart Network (WCCHN) has coordinated and integrated cardiac and surgical care for patients across the four western Canadian provinces. The WCCHN clinicians work collaboratively across all jurisdictions to ensure the highest standard of care is accessible to all patients and families. Surgical care is regionalised in two surgical centres ensuring adequate volumes to maintain a high standard of technical skill and optimal outcomes.
Objectives: to develop an understanding of the factors to consider when looking at a regionalised approach to providing specialised healthcare services; to identify the benefits/challenges to regionalising specialised services; impacts of regionalisation of specialised services on families – the good and the bad; sustainability of this model.
Discussion: History of paediatric cardiac surgery in western Canada

- Outcome data that necessitated looking at a different approach to providing highly specialised services to paediatric cardiac patients
- Stakeholders – how they came together and reached the decision to regionalise services
- Building a network of paediatric cardiac and surgical care
- Successes/challenges encountered along the way
- Reflection after 10 years of practicing this way
- Future directions for this model of delivering healthcare.

1720: THE 'VIRTUAL ICU': ADDING EXPERIENCE IN PAEDIATRIC CARDIAC SURGERY

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Introduction: Similar to other developing countries, some areas in Brazil experience a shortage of resources to establish paediatric cardiology (PC) programmes. Within this context, a telemedicine PC network was established between two states to add competences and train local teams.
Objective: To report on the experience with a new model of telemedicine, the 'virtual ICU', within a PC network for poor children in north-east Brazil.
Methods: Besides medical equipment and a TV screen, an iPAD and wireless Internet connection were added to the ICU. Local teams underwent face-to-face and online training sessions. Surgical and post-operative checklists were developed. Cases are discussed online. Specialised teams travel once a week to perform more complex surgical cases and initial post-operative follow up (24-hour). Local teams, under online supervision, perform the remaining post-operative and surgical cases of lesser complexity.
Results: From October 2011 to July 2012, 74 surgeries were performed in children aged 20 days to 17 years. Cardiac bypass was used in 59.6%. Jenkins' complexity scores were one to three in 50%, three to six in 44.5% and over seven in 5.4%. Total mortality was

9.9%. Infection and bleeding were the most frequent post-operative complications (14.8%). 'Virtual ward rounds' took place daily and there was an average three to five extra consultations per week, most related to clinical instability, drug management or echo revision. On a qualitative review, ICU teams reported a feeling of security from the new system, although some still regard telephone calls as simpler and more efficient than Internet connections.
Conclusion: Our results do not focus on surgical numbers, as this experience, despite being aligned with current literature, is still incipient. The emphasis is on the model provided by the 'virtual ICU', which adds experience and diffuses knowledge between centres, thus providing a safer environment to initiate such complex programmes in less-developed areas.

1727: PULLING THE PATIENT THROUGH THE SYSTEM: PROACTIVE CARE AT GREAT ORMOND STREET CHILDREN'S HOSPITAL, LONDON, ENGLAND

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A streamlined patient journey provides a process where clinical staff, families and children can manage their expectations of the system. With the present high financial pressures on the National Health Service, a current review of national children's surgical heart services and the opening of a new cardiac unit in 2012, our aim was 'to reduce over a six-month period the hospital stay by 5% in children with congenital heart disease, by removing inefficiencies, and improving quality of service provision, meanwhile ensuring that each child is discharged when medically ready'. Quality improvement tools used were affinity diagrams, interrelationship diagrams and rating final sub-groups. The final drill down highlighted areas of 'plan, process, journey and structure'. The group of champions looked at four keys areas, patient and family preparation, medication and discharge, investigations and pre-discharge ECHO. Data were obtained using Tally sheets and the Great Ormond Street Hospital cardiorespiratory database: length of stay in hours, discharge times, with balancing measures around patient experience questionnaires and repeat prescriptions. The improvement project is currently on-going using a range of interventions, with an aim to review data monthly and monitor outcomes over the next six months as to whether we achieved our aim. Learning points have however been evident around timing and current work pressures, culture and structure, and engaging clinically based individuals to make it a real and tangible experience and outcome.

1744: QUALITY OF CARE THAT CHILDREN WITH HEART PROBLEMS RECEIVE IN THE SOUTH AFRICAN HEALTH SYSTEM

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[3]Child Healthcare Problem Identification Programme, South Africa

Background: The quality of care children with heart problems receive in the broader South African health system is not well described. The Child Healthcare Problem Identification Programme (Child PIP) uses mortality audits to assess quality of care. Over 150 South African hospitals have used Child PIP since 2005, recording over 700 000 admissions and over 29 000 deaths. This paper provides information on the children dying with heart problems.
Methods: The study population included children admitted to and dying in participating South African hospitals. Data were gathered from 2005 to 2012. The mortality review process established cause of death and modifiable factors.
Results: Of the 29 000 deaths, 800 died with a heart problem as the main cause of death. Most cardiac deaths occurred in infants, 12% were severely malnourished. HIV status was unknown in one-third. Over one-third died within 24 hours of admission, 16% of deaths were considered avoidable. There were 1.7 modifiable factors per death, 52% of modifiable factors occurred within the health system, of which 55% were attributed to health workers. For rheumatic heart disease there were 2.2 per death. A substantial number were attributed to caregivers' health-seeking behaviour. A lack of high-care facilities was most significant for administrators, and inadequate history, assessment and investigations were the most common for health workers.
Conclusion: These findings for children dying with heart problems suggest areas of concern. Co-morbidities of HIV and malnutrition may influence decision making about definitive cardiac care. The high proportion dying within 24 hours of admission and the higher rate of modifiable factors occurring in children with rheumatic heart disease call for further investigation and may present opportunities for improving cardiac care systems. Resource allocation, in particular critical care, is a problem that needs attention, and improved awareness of heart-related danger signs in the general population is necessary.

1791: HOME-BASED PALLIATIVE CARE IN CHILDREN WITH CARDIAC PROBLEMS

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[2]Warsaw Hospice for Children, Poland

Introduction: Warsaw Hospice for Children (WHC) is the first non-governmental organisation in Poland that provides home-based palliative care for children with life-limiting conditions and their families. Children in whom curative treatment was completed could be admitted to the palliative home-care programme. The aim of this study was to evaluate patients with cardiac problems who were under hospice care between 1994 and 2012.
Methods: During 18 years, 96 children with different cardiac problems were under WHC care. There were 17 teenagers with Duchenne muscular dystrophy, 13 children with unoperable heart defects, mainly due to pulmonary hypertension or severely hypoplastic pulmonary arteries, eight children with post-operative complications, four with Down syndrome and pulmonary hypertension, 54 with lethal chromosomal aberrations, mainly trisomy 13, and 18 with heart defects.
Results: Until 2000, seven teenagers with heart defects complicated by pulmonary hypertension, heart failure or hypoplastic pulmonary arteries were under palliative care. All of them died after 31 days to seven years in hospice care. Since 1999, perinatal palliative care has been established, mainly for children with lethal chromosomal aberrations complicated by congenital heart defects, 37 patients were admitted to the hospice programme after prenatal diagnosis and consultation. None of those children was operated on. Period of palliative home care lasted from three to 1 269 days, mean 161 days. Just one newborn with hypoplastic left heart syndrome diagnosed prenatally was in the hospice care for 35 days.
Conclusions: Palliative care should be considered in all children with life-limiting conditions. In patients with complicated heart defects in whom surgical treatment failed, such options should be discussed with the parents. Palliative care should be the method of choice for foetuses and neonates with lethal chromosomal disorders whose parents are against termination of pregnancy.

1826: CARDIAC CARE IN AFRICA: THE NAMIBIAN CHILDREN'S HEART PROJECT

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[2]Christiaan Barnard Memorial Hospital, Cape Town, South Africa

Background: There is limited access to cardiac care in Africa, and prior to 2010, there were no cardiac services in Namibia. The Ministry of Health and Social Services commissioned a heart centre

at Windhoek Central Hospital (WCH) on 18 October 2010. We report early results from a new single-surgeon programme over the first two years.

Methods: This was a case series of 231 patients between October 2010 and 31 August 2012. In year one, surgery was limited to children above 20 kg and in year two, those above 10 kg body mass. Data were entered prospectively into a hospital-based registry, File maker pro data.

Results: Two hundred and thirty-one patients had surgery, 208 on cardio-pulmonary bypass. Age ranged between four months and 83 years, of whom 56% were under 18 years; 105 cases (45%) had rheumatic heart disease (RHD) with aortic valve replacement (20), mitral valve replacement (41), and a mitral valve-sparing annuloplasty and repair in 33; 86 patients had congenital heart disease (25 adults). The common lesions were VSD (31), secundum ASD (20), primum ASD (16), PDA (16) tetralogy (17), aortic coarctation (four), DORV (three), Ebstein's anomaly (two). There have been nine post-operative (< 28 days) deaths with a mortality rate of 3.6%.

Conclusion: This new service is a novel model of integrated cardiac care that dispensed with age-related barriers to delivery and concentrated on 'uncomplicated' and common diseases. Adults with CHD represent survivors of native heart disease. Low numbers of small babies reflect strategic choices in a resource-limited environment but also the parlous state of diagnostic services for children with CHD. High numbers of patients needing surgery for RHD reflect the absence of a national programme for prevention and control of RHD. Without further development and support, the future of this new service remains precarious.

Major Advances in Cardiology

2010
SHIFT

2005
PREAMI
Perindopril and Remodelling in Elderly with Acute Myocardial Infarction

2008
BEAUTIFUL

2005
Anglo Scandinavian
ascot
Cardiac Outcomes Trial

2008
HYVET
HYPERTENSION IN THE VERY ELDERLY TRIAL

2003
EUROPA

2007
ADVANCE
ACTION IN DIABETES VASCULAR DISEASE: PRETERAX AND DIAMICRON MR CONTROLLED EVALUATION

2001
PROGRESS
PERINDOPRIL PROTECTION AGAINST RECURRENT STROKE STUDY

ADULTS WITH CONGENTIAL HEART DISEASE AND THE PREVENTION OF AQUIRED HEART DISEASE STARTING IN CHILDHOOD

76: ELEVATED LIPOPROTEIN A IN A NEWBORN WITH THROMBOEMBOLIC DISEASE AND A FAMILY HISTORY OF ATHEROSCLEROSIS

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[2]Pediatrics, Taubate University, Brazil

Background: Thromboembolic disease is uncommon in young children. Catheters, sepsis and inherited diseases are important risk factors for thrombosis. Familial hyperlipoprotein A is an inherited condition that leads to higher levels of lipoprotein A (Lp (a)). Lp (a) is a subclass of low-density lipoproteins, plasminogen-like, with thrombogenic properties.
Case description: The male infant was born by caesarean section at 37 weeks' gestation. The baby was cyanotic with little improvement in the delivery room. The first echocardiogram (ECHO) demonstrated pulmonary arterial hypertension (PASP = 80 mmHg). During the first days of life, the newborn developed intracranial haemorrhage and worsening of cyanosis. The follow-up ECHO showed 2 small clots within right ventricle and a larger one (13.4 × 9.2 mm) near the left pulmonary artery with flow obstruction. Radionuclide scanning showed signs of pulmonary thromboembolism. An umbilical venous catheter was removed. Enoxaparin was administered to the patient (1.5 mg/kg SQ q12h). The results of the collected blood tests were normal (thyroid hormones, liver enzymes, glucose, white blood cell and platelet count, haemoglobin electrophoresis, factors IX, V and VIII). Owing to haemodynamic instability, the newborn did not undergo cardiac surgery. The conservative treatment showed clinical improvement and gradual reduction of clots. He was discharged on oral anticoagulation (warfarin). On further investigation, the prothrombin gene mutation, the anticardiolipin antibodies and the proteins C and S were collected and were negative. The family history was positive for premature heart disease. Serum levels of Lp (a) were obtained from the patient and his parents. The child (55 mg/dl) and his mother presented with higher levels of Lp (a). Antiagregant (aspirin) was prescribed. After the neonatal period, the patient presented no new thrombus.
Conclusions: Congenital thrombophilia needs to be strongly considered in neonates with a clinically significant thrombosis. The increased Lp (a) is a risk factor for thrombosis, coronary artery disease and cerebrovascular accident.

108: SELF-REPORTED HEALTH-RELATED QUALITY OF LIFE IN CHILDREN AND ADOLESCENTS WITH HEART DISEASE: A SWEDISH REGISTRY STUDY

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[2]Malmo University and Skåne University Hospital, Sweden

Background: During the last decades survival of children with congenital heart disease has improved significantly. It is important to assess health-related quality of life (HRQoL) in these patients. Some of the variables in the Swedish national registry of congenital heart disease (SWEDCON) include HRQoL measurements.
Aim: To describe HRQoL in children and adolescents with heart disease.
Materials and methods: A descriptive study based on data from SWEDCON, collected from one paediatric cardiac outpatient clinic. The sample consisted of 104 girls and 154 boys, 9–18 years old with registrations of HRQoL variables and the questionnaire DISABKIDS short version. The patients were divided into three groups depending on the number of cardiac operations or catheter treatments they had undergone.
Results: DISABKIDS mean total score (max 100) in patient group 1 (no cardiac procedures) was 92.9, in group 2 (one cardiac procedure) 94.4, and in group 3 (2 or more cardiac procedures) 86.0. NYHA classes II and III were more frequent in patient group 2 and particularly in group 3. Patients in groups 2 and 3 had significantly more cognitive difficulties than those with no cardiac procedures. However, more than 50 % of all patients had more than 3 hours' physical activity/week.
Conclusions: In this unselected group of patients with congenital heart disease it is shown that HRQoL is better for children with fewer cardiac procedures and that HRQoL generally is better than for other chronically ill children, which confirms previous results on selected materials. In this single-centre study a web-based registry was used for evaluation of HRQoL in children and adolescents with heart disease. In the future it may be used for larger groups, such as the national population of children with heart disease.

114: COMPLIANCE WITH LIFESTYLE RECOMMENDATIONS IN CHILDREN AND YOUNG ADULTS WITH HYPERTROPHIC CARDIOMYOPATHY

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Background: Hypertrophic cardiomyopathy (HCM) is the most common medical cause of sudden death during exercise. Previous studies suggest that restriction in competitive sports participation result in lower mortality rates. This study evaluates the effects of lifestyle recommendations on physical and leisure-time activities in HCM-patients.
Materials and methods: Twenty-seven consecutively recruited asymptomatic patients with HCM diagnosed through family screening were asked to participate in the study. All received lifestyle recommendations according to international guidelines. Twenty (median age 14.5 years, range 5–25) filled out a questionnaire regarding the frequency of strenuous physical activities (defined as becoming exhausted), sports participation and leisure time activities (such as spending time with friends, using a computer, watching TV, visiting the cinema, theatre or playing or listening to music), before diagnosis and after 1 year. Patients were classified in 5–6 groups according to the frequency of activities. Results were analysed using Wilcoxon signed rank test.
Results: There was a significant decrease in the frequency of strenuous physical exercise from before diagnosis compared to 1 year later ($p = 0.002$). Forty per cent performed strenuous exercise > 7 hours/week before diagnosis compared to 5% 1 year later. The number of patients who never participated in sports activities increased from 10% to 20% ($p = 0.007$). No change was detected regarding leisure-time activities. Time spent watching TV and using a computer did not increase significantly.
Conclusions: Lifestyle recommendations significantly affect the physical activity habits in HCM patients. Our results indicate a high level of compliance with the recommendations. Leisure-time activities do not seem to be negatively affected and time spent on sedentary activities did not increase.

128: VENTRICULAR HYPERTROPHY IN ADULTS OPERATED FOR VENTRICULAR SEPTAL DEFECT AS TODDLERS

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Background: Cardiac morphology is expected to restore to normal after surgical closure of ventricular septal defects (VSD) in early childhood. However, long-term abnormalities in cardiac morphology may exist and are the subject of this study.

Methods: Patients (n = 15), median age at surgery of 2.6 years (1.5–4.1 years) and 21.1 years (19.8–23.2 years) at the time of examination, and age-matched control subjects (n = 10) underwent cardiac magnetic resonance (MR) scanning using a Philips Intera, 1.5T MR scanner. Stacks of 3 contiguous long-axis and 12 short-axis slices encompassing both ventricles were acquired. Quantitative flow measurements were made using phase-contrast gradient imaging. For data analyses OsiriX and Medviso Segment software was used.
Preliminary results: Compared to controls, left ventricular ejection fraction was unaffected, median 63.2% (56.6–68.7%) vs 65.9% (53.5–69.9%), p = 0.99, and so was cardiac index, median 3.6 l (min m^2)$^{-1}$ (3.2-4.1 l (min m^2)$^{-1}$) vs 4.0 l (min m^2)$^{-1}$ (3.6–4.1 l (min m^2)$^{-1}$), p = 0.81. Ventricular mass indexes were larger in VSD patients, $p <$ 0.05 in both ventricles. Left ventricular peak ejection/filling rate was higher in patients; median 87.3 ml sec^{-1} (72.5–111.3 ml sec^{-1})/145.5 ml sec^{-1} (82.7–174.5 ml sec^{-1}) compared to controls 69.3 ml sec^{-1} (55.0–87.1 ml sec^{-1})/87.4 ml sec^{-1} (76.3–126.2 ml sec^{-1}), $p < 0.05$ and p = 0.08, respectively. In contrast, right ventricular peak ejection/filling rate was lower in patients; median 57.7 ml sec^{-1} (50.4–73.0 ml sec^{-1})/50.4 ml sec^{-1} (37.0–60.2 ml sec^{-1}) as compared to controls 89.9 ml sec^{-1} (48.5–131.0 ml sec^{-1})/94.5 ml sec^{-1} (51.1–137.2 ml sec^{-1}), $p <$ 0.01 for both parameters.
Conclusion: Twenty years after surgically closed VSDs, larger ventricular masses combined with superior peak rates of ejection and filling in the left ventricle are noticed. In contrast, right ventricular peak ejection and filling velocities were inferior compared to control subjects. The consequence for long-term outcome is unknown and needs further studies.

134: MENSTRUAL BLEEDING AFTER OPEN HEART SURGERY

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[2]Department of Cardiology, Aarhus University Hospital, Denmark

Background: We investigated whether open heart surgery with the use of extracorporeal circulation has impact on menstrual bleeding (MB).
Material and methods: MB pattern was registered retrospectively and MB during admission was registered prospectively in fertile women undergoing heart surgery for congenital heart disease. Hematocrit and 24-hour postoperative bleeding were compared with men also undergoing congenital heart surgery in the period 2010-2011.
Results: Women (n = 22), mean age 35 years (range: 17–60) were operated on and hospitalised for 4–5 postoperative days. Mean postoperative bleeding in the first 24 hours was 312 ml (range 50–1442 ml). Three to four (16%) women were expected to have MB during their hospital stay. Unplanned MB (lasting 2–5 days) was detected in 13 patients (60%). Six had expected MB and 3 had, as expected, no MB during hospital stay. Of the 13 unexpected MB, 4 were 1–7 days early, 4 were 8–14 days early, 3 were 1–7 days late and 2 had MB despite having had MB within the last 2 weeks. None had unusually severe or long-lasting MB. Ten women took oral contraceptives, 7 of whom had unexpected MB. Men (n = 22), mean age 35 years (17–54) had a mean 24-hour postoperative bleeding of 331 ml (range 160–796 ml) which was not significantly different from that of the women. The mean preoperative haematocrit was 40% (29-53%) among men, not significantly different from that of women (mean 40% [32–60%]).
Conclusion: MB patterns are disturbed by open heart surgery in the majority of fertile women. Nevertheless, the unexpected MB is neither particularly long lasting nor of excess quantity, and postoperative surgical bleeding is unaffected. We recommend giving information about irregular MB, but no special precautions when operating on women of fertile age.

138: LIMITED KNOWLEDGE AMONG LOCAL CARDIOLOGISTS OF THE MANAGEMENT OF PREGNANT WOMEN WITH MECHANICAL HEART VALVES AND MINIMAL IMPACT OF FOCUSED EDUCATION

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Background/hypothesis: There are significant risks to the mother and foetus in pregnant women with mechanical heart valves. While these patients are frequently managed by experienced multidisciplinary teams, general cardiologists should be aware of the relevant issues. We assessed the knowledge of local cardiologists on these issues and evaluated the impact of focused education on the subject.
Methods: A questionnaire on the principles of management of pregnant women with mechanical heart valves was distributed to cardiology consultants and trainees from the UK's Yorkshire and Humber region at an educational meeting. A lecture was then given and a leaflet summarising the issues circulated. At a further meeting 11 months later the participants were re-questioned.
Results: The questionnaire was completed by 35 doctors on the first occasion and 26 on the second. Questions included: What is the risk of maternal death? [*Accepted answer*: 1–5%; *% correct, 1st questioned*, 48%; *% correct, re-questioned*, 69%.] What is the risk of foetal wastage? [*Accepted answer*: 20–40%; *% correct, 1st questioned*, 18%; *% correct, re-questioned*, 31%.] What is the risk of major bleeding? [*Accepted answer:* 1–4%; *% correct, 1st questioned*, 23%; *% correct, re-questioned*, 23%.] What is the risk of warfarin embryopathy? [*Accepted answer:* 5–6%; *% correct, 1st questioned*, 20%; *% correct, re-questioned 31%.]* By what gestation should warfarin be stopped to avoid embryopathy? [*Accepted answer:* 6 weeks; *% correct, 1st questioned*, 3%; *% correct, re-questioned*, 15%.] The risk of warfarin embryopathy may be lower if the daily dose is < ? mg. [*Accepted answer:* 5 mg; *% correct, 1st questioned*, 49%; *% correct, re-questioned*, 31%.] How does pregnancy alter the pharmacokinetics of low molecular weight heparins? [*Accepted answer:* Various; *% correct, 1st questioned*, 40%; *% correct, re-questioned*, 38%.] How would you determine the dose of low molecular weight heparin? [*Accepted answer:* Anti-factor Xa levels; *% correct, 1st questioned*, 40%; *% correct, re-questioned*, 35%.] Rank in order thromboembolic risk during pregnancy: A) heparin throughout, B) warfarin throughout, C) heparin first trimester, warfarin thereafter. [*Accepted answer:* A>C>B; *% correct, 1st questioned*, 49%; *% correct, re-questioned*, 35%.]
Conclusions: Knowledge among local cardiologists regarding the management of pregnant women with mechanical valves was limited. There was also a limited effect of improving knowledge with targeted education.

139: VASCULAR FUNCTION IN ADULTS WITH REPAIRED COARCTATION OF THE AORTA – ASSESSMENT BY MULTIMODAL MAGNETIC RESONANCE IMAGING

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[2]University of Leeds, Leeds, UK

Background/hypothesis: Patients with surgically repaired coarctation of the aorta remain at increased risk of premature cardiovascular events including myocardial infarction and stroke. Magnetic resonance imaging (MRI) can be used to assess a number of measures of vascular function, including endothelial function (as flow-mediated dilatation (FMD) of the brachial artery) and large artery stiffness (as aortic pulse wave velocity (PWV) and distensibility).
Aim: Our aim was to gather preliminary comparative data on vascular function by MRI in adults with repaired coarctation and healthy controls.
Methods: Seven patients with previous coarctation repair and 14 age-matched healthy controls underwent multimodal MRI assessment of vascular function. Blood pressure was measured in the

right arm. Distensibility of the ascending aorta was measured at the level of the right pulmonary artery. Phase contrast MRI was used to measure aortic arch PWV. The transit time between the arrival of the flow wave at ascending aorta and the proximal descending aorta was calculated by a foot-to-foot method. Cine imaging of the right brachial artery was performed to measure brachial artery reactivity to reactive hyperaemia (FMD) and glyceryl trinitrate. Comparisons were made using Student's *t*-tests. Data are expressed as mean (SD).
Results: In 2 patients PWV data was of poor quality and was therefore excluded, along with the corresponding data from controls. Ascending aortic distensibility was lower in patients than in controls (4.3(2.2) vs 7.2(2.1) $mmHg^{-1}.10^{-3}$, $p < 0.01$), aortic arch PWV was higher (6.3(1.5) vs 4.3(1.1) m/s, $p < 0.05$). There was no difference in brachial artery FMD (13(10)% vs 16(6)%, $p = 0.3$) or response to glyceryl trintrate (22(12)% vs 27(7)%, $p = 0.08$).
Conclusions: In small numbers MRI appears to have identified differences in vascular function between patients with repaired coarctation and controls. Incorporation of these measures into surveillance MRI assessment of repaired coarctation may determine whether they have any prognostic value in the long term.

153: WOMEN WITH CONGENITAL HEART DISEASE: THEIR IMPERATIVE TOWARDS CONCEIVING

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[2]Monash University, Melbourne, Australia

Background: Advances in medical care have increased the number of women with congenital heart disease (CHD) reaching child-bearing age and wishing to conceive. Pregnancy adds an additional stress on their compromised cardiovascular system, with risks to both themselves and their unborn child. Despite these risks, they still proceed to conception. A qualitative study was undertaken to determine their motivations for conception.
Methods: A two-group design of women with ($n = 20$) or without ($n = 20$) CHD, over 18 years of age who had completed one or more successful pregnancies were recruited from a tertiary centre and private clinics. The women completed a questionnaire and a semi-structured interview. Their medical records were reviewed. Thematic analysis was conducted on the interview data.
Results: CHD in the women ranged from minor lesions (e.g. atrial septal defect) to moderately severe lesions (e.g. Fontan circulation). The motivations for conceiving in women with CHD were similar to those of healthy women. These motivations included: the influences of relationships with their partners, family and friends; concern for the reproductive changes associated with increasing age; their innate desires for motherhood; the women's personal goals; and cultural and social expectations. Women with CHD had a tendency to underestimate the severity and associated risks of their CHD. They exhibited a strong reliance on their treating clinicians and assumed advances in medical care would carry them through their pregnancies.
Conclusions: Motivations to conceive are similar for healthy women and women with CHD. Those with CHD had a tendency to harbour an unrealistic understanding of the severity of their CHD and its implications in pregnancy. This perception emphasises the importance for clinicians caring for these patients to be knowledgeable about the impact of CHD on the woman's pregnancy and of pregnancy on their CHD, as they carry great responsibility in caring for these patients.

201: SURGERY OF TETRALOGY OF FALLOT IN ADULTS IN AHMED GASIM CARDIAC CENTER

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Background: Corrective surgery of tetralogy of Fallot (TOF) beyond childhood is very rare as a result of improved methods of early detection and early repair. In our centre 20% of TOF patients who underwent repair were adults, with good early results.
Method: This is a descriptive retrospective study. The data were collected by the authors from the records of the patients who underwent surgical repair of TOF from 2003 to 2011 in our centre. We included all patients above 18 years. The data were collected manually and analysed by a computer program SPSS.
Results: Adult TOF surgical repair represents 20% of TOF surgery. Fourteen patients were male (75%). The mean age was 20.47 years. Eleven patients were classified as NYHA III, and 60% of patients had pulmonary gradients of more than 50 mmHg. There was no previous palliative surgery or associated major cardiac anomaly. All patients underwent total surgical repair through right atriotomy and the ventricular septal defects (VSDs) were closed by synthetic patch. In one patient the pulmonary artery was opened and enlarged with a pericardial patch. The mean follow-up time was 4.5 years. Two patients died during their hospital admission and another 3 died during the period of follow-up.
Conclusion: Although operating on adult patients with TOF is a challenge and rarely seen, we still had a reasonable number of patients with good early surgical outcome but poor in the mid-term follow-up period.

233: DEMONSTRATION OF STRONG FEASIBILITY OF THE IHEARTCHANGE WEBSITE FOR TRANSITIONING PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Published guidelines recommend initiatives to improve the rate of transfer and quality of transition of patients with congenital heart disease (CHD) from paediatric to adult cardiac care. Unfortunately, there are few published interventions for transitioning patients with CHD, proposed initiatives are often resource intensive, and strategies for engaging adolescents are unknown. We created and evaluated the feasibility of the iHeartChange website targeting transitioning CHD patients.
Methods: Patients transferring from a paediatric to adult CHD programme in a 1-year period were invited to participate in the study. Feasibility outcomes, specifically eligibility, participation rate, and patient feedback about the website, were investigated.
Results: A total of 207 CHD patients aged 16–19 were transferred, of whom 187 (mean age 17.7 years, 58% male, 75% with moderate/great defect complexity) met full inclusion criteria and were invited to participate. Thirty-four per cent (63/187) of eligible patients logged on and completed baseline surveys; there were no significant differences in age, sex, or defect complexity between patients who did and did not complete baseline surveys. Forty-eight patients completed follow-up surveys at least 1 month after initial website access. The majority of patients mostly or strongly agreed that the website was easy to use (95%), fun to use (82%), gave information that was useful (91%) and they could trust (95%), gave a better understanding of CHD medical issues (86%), should be offered to all patients moving to adult care (91%), and is something they would use again if available (87%).
Conclusion: Strong feasibility of the iHeartChange website was demonstrated. One-third of patients with CHD chose to access a website as part of a research study; higher use is anticipated if offered in a clinical setting. Adolescent feedback in terms of design and content was extremely positive. In conclusion, a website targeting transitioning CHD patients holds significant clinical promise.

258: CHANGING RISK FACTORS FOR ARRHYTHMIAS AND SUDDEN DEATH AMONG ADULTS WITH REPAIRED TETRALOGY OF FALLOT IN THE CURRENT ERA

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Background: Among adults patients (pts) with repaired congenital heart disease, those with tetralogy of Fallot (TOF) historically have exhibited increased risk for tachyarrhythmias (Ar), AV block (AVB) and sudden cardiac death (SCD) associated with right ventricular (RV) systolic pressures (RVSP) > 60 mmHg, outflow tract (OT) gradients > 20 mmHg and QRS duration > 180 ms. However, surgical practices have changed over the years. The purpose of this study was to evaluate Ar/SCD risk with symptoms, echocardiographic (ECHO), electrophysiologic (EP), haemodynamic and RV volume on cardiac MRI (CMRI) findings among the current generation of older repaired TOF pts.
Methods: TOF pts seen in our institution's Adult and Adolescent Congenital Heart Programs were divided into 2 groups: group I: +Ar/SCD; group II -Ar/SCD, and correlated with current pt age, gender, age at surgical repair, repair types (transannular patch; no patch; conduit; shunt), ECHO, CMRI, ECG/Holter, haemodynamic and EP results.
Results: Of 136 pts (66 male/70 female) ages 11–58 y (mean 26), 61 (45%) had Ar (22% atrial, 60% ventricular, 10% both) and 8% AVB. Of these, SCD occurred in 3 (5%) (group I). These pts were chronologically older (mean 32 vs 22 y) with repair performed at an older age (mean 50 vs 22 mo) than those without Ar/SCD (group II) ($p < 0.05$). QRS duration (mean 158 ms) and RVSP (mean 44 mmHg) were persistent risk factors. However, there was no correlation with type of surgical repair, gender, RV pressure > 60 mmHg, RVOT gradient > 20 mmHg, or RV volume. Ar were induced in 91% of group I pts studied, requiring ablation or device implant.
Conclusions: In the current era, repaired TOF pts still remain at risk for Ar/SCD depending on their chronological age and age at surgical repair. However, QRS duration is shorter (158 ms) and RVSP less (44 mmHg) than previously reported. Type of repair, RV outflow gradients or CMRI volumes did not correlate with Ar/SCD.

362: SURGERY FOR ADULTS WITH CONGENITAL HEART DISEASE: AN 11-YEAR SINGLE-CENTRE EXPERIENCE

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Background/hypothesis: Adults with congenital heart disease (ACHD) are increasing in number as a result of asymptomatic lesions during childhood or improved paediatric cardiac care. Some of these patients are not discovered in childhood, while others require subsequent cardiac interventions. We retrospectively reviewed the early and late outcomes of ACHD operations/re-operations over the last decade.
Materials and methods: Between January 2001 and December 2011, 1 044 congenital lesions were repaired during 659 surgical operations in 538 patients (287 males). Mean age at operation was 32.5 ± 13.1 (range: 18–84) years. Excluding rhythm-related problems from postoperative analysis, 200/449 (45%) were first-time operations, and 98/449 (21.9%) had NYHA Class symptoms 3 or 4 pre-operatively.
Results: Early and late mortality (EM) (LM) were 1.3% ($n = 6$) and 2.9% ($n = 13$) respectively. Actuarial survival at 1, 3, 5, and 10 years was 98%, 96%, 94%, and 86% respectively. Major postoperative complications occurred in 54/449 (12.0%). Re-operations were required in 7.1% ($n = 32$). Actuarial freedom from re-operation at 1, 3, 5, and 10 years was 98%, 92%, 87%, and 70% respectively. At mean follow-up of 33.1 ± 35.7 (range: 0–137) months, 3.1% ($n = 14$) had residual Class 3 or 4 symptoms, although 4.5% ($n = 20$) had minimal follow-up.
Conclusions: A wide variety of CHD pathology is seen in adults. Surgical repair can be accomplished with low mortality and morbidity. Pulmonary valve, aortic valve, aortic pathologies and rhythm abnormalities dominate the spectrum of ACHD pathology.

379: MODE OF DELIVERY AND PREGNANCY OUTCOME IN A TERTIARY CENTRE FOR ADULT CONGENITAL HEART DISEASE

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Background: As a result of the continuing improvement of congenital heart disease (CHD) therapy, a growing number of women are reaching child-bearing age. Contemporary maternal risks of pregnancy and preferred modes of delivery are not well known.
Aim: To review all deliveries of CHD patients in our centre since 2000, regarding mode of delivery and maternal outcome.
Results: There were 391 deliveries. Mean age at delivery was 30.3 (± 5.5) years, age range 17.4–48.5 years; 208 women had 1 delivery (4 twins), 58 had 2, 17 had 3 and 4 had 4.

Spontaneous deliveries occurred in 231 (59%), 94 (24%) were by caesarean section (CS) and 66 (17%) after medical induction of labour; 120/231 (52%) spontaneous deliveries were entirely uneventful. The rest had premature rupture of membranes (26), fetal distress (23), bleeding requiring transfusion (10), maternal arrhythmia (3), vacuum delivery (30), forceps (6) and perineal laceration grade II or more and/or uterine exploration.

Of 66 medical inductions of labour, 56 were for cardiac indications; 18 had shunt lesions, 17 valve disease, 10 complex transposition of the great arteries (TGA), 7 tetralogy of Fallot (TOF), 2 Fontans and 2 PFO post stroke.

Of 94 CS, 58 (62%) were elective (both term and preterm); 7 were for very high risk CHD – 4 Eisenmenger syndrome, 2 PHT, 1 TGA-Rastelli, 1 Fontan. Thirty-three CS were urgent. Two of four Eisenmenger patients died. Two more patients died, both with repaired DSS: one after induction and one died at home 3 days after an uneventful delivery. There were 7 more serious complications but with good outcome.
Conclusions: This large single-centre series of deliveries in CHD shows that except for Eisenmenger patients, maternal outcome is very satisfactory with a very low cardiac complication rate in all modes of delivery. A higher than expected rate of CS suggests that the threshold for this intervention was lower in CHD patients.

448: SUDDEN CARDIAC DEATH IN CHILDREN AND ADOLESCENTS: IT IS PREVENTABLE WITH A MULTI-DISCIPLINARY, MULTI-INSTITUTIONAL APPROACH

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Background/hypothesis: SCD in children and adolescents occurs with unclear frequency and is devastating. There are many aetiologies of SCD in children/adolescents. We hypothesise that survival is achievable with a multi-disciplinary/multi-centre primary and secondary prevention approach emphasising awareness/education as well as advocacy for secondary prevention approaches (lay-public cardiopulmonary resuscitation (CPR), education, automated external defibrillation (AED) use and placement of school CPR-AED programmes.
Material and methods: Project ADAM (PA) was initiated in the state of Wisconsin at Children's Hospital of Wisconsin in November 1999. PA is a primary and secondary prevention programme. Warning sign education, teaching importance of comprehensive pre-participation exam, advocacy for individual school emergency-preparedness plans and school CPR-AED programmes are all part of PA. Since inception, affiliate PA programmes have been established in other states: Georgia, Pennsylvania, Florida, Illinois, Alabama, Washington, Tennessee, Texas and Michigan.
Results: PA has saved 60 lives since November of 1999. Of the total, 24 lives of children/adolescents and 36 lives of adults have been saved: Wisconsin (total 23, children/adolescents 10, adults 13); Georgia (total 29, children/adolescents 11, adults 18); Pennsylvania (total 4, children/adolescents 2, adults 2); Texas (total 2, children/adolescents 1, adults 1).
Conclusions: PA has saved the lives of both children/adolescents and adults. Education/awareness and implementation of CPR-AED programmes across the country aimed at children and adolescents will have the potential to save many mores lives, adults included.

505: METABOLIC SYNDROME-RELATED CHARACTERISTICS ARE ASSOCIATED WITH AN INCREASED CAROTID-FEMORAL PULSE WAVE VELOCITY IN CHILDREN

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Background/hypothesis: The origins of cardiovascular disease are in childhood, with changes to vessel structure and function often apparent from early life. Carotid-femoral pulse wave velocity (PWV) is a validated non-invasive measure of arterial stiffness – a determinant of vascular function. We hypothesised that features of metabolic syndrome – obesity, insulin resistance and hypertension – would be associated with higher PWV in children.
Materials and methods: Carotid-femoral artery PWV was performed on a population cohort of 147 twin pairs aged 7–11 years. Fasting bloods, blood pressure, anthropomorphic and other measures were collected concurrently. The participants were analysed as individuals by multiple linear regression, after adjusting for age and sex. Further between-twin pair analysis was performed to investigate the effect of twin-pair clustering.
Results: Of the 147 twin pairs, 47 were monozygotic and 100 were dizygotic. Mean age was 9.03 years, and half were male. There were no significant differences between the characteristics of monozygotic and dizygotic twins. Mean PWV was 5.95 m/s, SD 0.655.

In individual-based analysis, age (β = 0.262, 95% confidence interval [CI] 0.170–0.348) and height (β = 0.015, 95% CI 0.004–0.027) were positively associated with PWV, as were markers of adiposity; weight (β = 0.012, 95% CI 0.002–0.023), truncal skin-fold thickness (β = 0.006 95% CI 0.001–0.010), waist circumference (0.011 95% CI 0.000–0.021) and hip circumference (β = 0.010 95% CI 0.000–0.020). Strong associations were found between markers of insulin resistance – HOMA (β = 0.116 95% CI 0.051–0.181) and c-peptide (β = 0.588 95% CI 0.248–0.927) – and increased PWV. Dyslipidaemia was also associated with increased PWV, most strongly with increased triglycerides (β = 0.278 95% CI 0.082–0.476). There was a strong association with increased systolic (β = 0.015 95% CI 0.008–0.021) and diastolic blood pressure (β = 0.034 95% CI 0.023–0.044). Relationships weakened within twin pairs, suggesting that the associations are at a familial level.
Conclusions: Accepted adult characteristics of metabolic syndrome are associated with increased PWV in pre-pubertal children.

581: NEW PREDICTORS OF SUSTAINED VENTRICULAR TACHYARRHYTHMIA IN REPAIRED TETRALOGY OF FALLOT

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Objectives: Investigation of whether cardiac anatomy and function predict arrhythmia.
Background: Repaired tetralogy of Fallot (rtoF) patients are at risk of ventricular tachyarrhythmia and sudden cardiac death. Risk stratification for arrhythmia remains difficult.
Methods: One-hundred-and-fifty-four adults with rtoF, median age 30.8 (21.9–40.2) years, were studied with a standardised protocol including cardiovascular magnetic resonance (CMR) and prospectively followed-up over median 5.6 (4.6–7.0) years for the prespecified endpoints of new-onset ventricular tachyarrhythmia (sustained ventricular tachycardia/ventricular fibrillation).
Results: Nine patients had ventricular tachyarrhythmia (6%) during follow-up. Patients who developed ventricular tachyarrhythmia were older (42.5 [34.9–50.2] vs 29 [21–40] years; $p = 0.01$), had a later repair (12.8 [6.2–13.9] vs 4.4 [2–8] years; $p = 0.02$), larger akinetic right ventricular outflow track (RVOT) region (length 55 [34–60] vs 30 [20–40] mm; $p = 0.002$) and a lower RV ejection fraction (42 [40–52] vs 53 [51–55] %; $p = 0.01$), compared to the other patients. On univariate Cox analysis, RVOT akinetic region length and RV ejection fraction were predictive of ventricular tachyarrhythmia. On stepwise Cox regression analysis, the RVOT akinetic region length was the only remaining predictor (hazard ratio 1.05, 95% confidence interval 1.01–1.08 per mm; $p = 0.004$). The survival ROC curve analysis indicated a cut-off value of 30 mm as a predictor of ventricular arrhythmia during 6-year follow-up with an AUC of 0.77, sensitivity of 83% and specificity of 61%. RVOT akinetic area length >30 mm predicted reduced ventricular arrhythmia-free survival (logrank $p = 0.002$)
Conclusions: RVOT akinetic region length predicts ventricular arrhythmia in late follow-up of rtoF. This is a simple, feasible measurement for inclusion in serial surveillance and risk stratification of rtoF patients.

583: IS GROWN UP CONGENITAL HEART DISEASE (GUCH) DIFFERENT IN A DEVELOPING COUNTRY? AN EXPERIENCE FROM A TERTIARY CARE HOSPITAL IN PAKISTAN

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Background: In the current era grown up congenital heart disease (GUCH) patients undergoing surgical intervention are increasing. There is considerable literature to suggest that most of the interventions in developed countries are in the form of re-operation in patients who had previously undergone repair, palliation or correction. However in a developing country like ours, most of the interventions are primary and corrective.

Methods: This is a descriptive study of 176 GUCH patients (over the age of 16 years) who underwent surgical intervention. Data were acquired from the cardiac surgery database over 6 years from July 2006 to June 2012. The majority of surgical procedures were performed by paediatric cardiac surgeons as first-time surgery.
Results: Of 176 patients, 54.3% were male, ranging in age from 16 to 76 years (mean 37.3 years). The majority of patients underwent surgical interventions for closure of atrial ($n = 77$) and ventricular septal defect ($n = 36$); 16 patients were operated for tetralogy of Fallot, and 5 for coarctation of aorta. The average length of hospital stay was 7.4 days. Overall mortality was 4%, and morbidity was 30.7% including re-operations, prolonged ventilator use, arrhythmias, heart block and others. Re-operations were performed in 1.7% patients as compared to almost 50% in developed countries. A similar trend was observed in literature from other developed countries along with a decline in primary procedures.
Conclusion: GUCH surgery in our practice is often a primary procedure for simple diagnoses. At present we do not have dedicated facilities for GUCH patients. This report highlights the fact that most of these simple lesions should have been treated before patients reached adulthood. There is a need to focus on developing GUCH services in developing countries to decrease the morbidity of untreated congenital heart disease.

591: AORTOPATHY IN ADULT PATIENTS WITH REPAIRED TETRALOGY OF FALLOT: HOW MUCH SHOULD WE WORRY?

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Background: We investigated the prevalence and progression of aortic dilatation (AD) late after repair of tetralogy of Fallot (rtoF) and sought to determine its predictors.
Materials and methods: Retrospective study of 110 rtoF adults with native aorta studied with cardiovascular magnetic resonance (CMR) at baseline and at ≥ 1 year follow-up. Aortic measurements were performed in diastole at sinus and ascending aorta (AA) level. AD was defined as diameter 2 standard deviations larger than published normal values.
Results: Age at baseline CMR was median [interquartile range]; 30.9 [22.9–39.4] years and at repair was 4.5 [2.1–9.1] years. Time interval between CMR scans was 6.3 [5.1–7.6] years.

Sixty-eight patients (61%) had AD, 65 had sinus dilatation (SD) (39 [35–41] mm, range 31–55) and 24 had associated AA dilatation (AAD) (40 [37–45] mm, range 36–51). Predictors of SD were age at baseline ($p = 0.009$), male gender ($p = 0.039$) and previous palliation ($p = 0.0004$); previous palliation remained an independent predictor ($p = 0.0004$). Predictors of AAD were age at repair ($p = 0.01$), systemic hypertension ($p = 0.04$), pulmonary atresia variant (PA) ($p < 0.0001$), male gender ($p = 0.008$), and sinus diameter ($p < 0.0001$); PA ($p < 0.0001$) and male gender ($p = 0.004$) were independent predictors for AAD. On ROC curve analysis, sinus diameter > 39 mm predicted AAD (AUC 0.90, $p < 0.0001$).

Mean rate of sinus and ascending aortic diameter progression was respectively 0.05 ± 0.1 and 0.12 ± 0.26 mm/year. No risk factor for increasing sinus diameter was identified. Predictors of AA diameter increase were age at repair ($p = 0.008$) and previous Blalock-Taussing shunt ($p = 0.01$). The latter was the only independent predictor ($p = 0.01$). No patient underwent aortic surgery.
Conclusion: AD is common in patients with rtoF but the rate of progression of aortic size is low in this large contemporary adult cohort. This should be taken into account when planning for scans in follow-up and/or prophylactic aortic replacement. Patients with sinus diameter ≥ 39 mm need cross-sectional imaging to assess for ascending aortopathy.

648: CLINICAL FEATURES OF ADULT CONGENITAL HEART DISEASE: A CHINESE INSTITUTIONAL EXPERIENCE

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Background: A substantial number of children with congenital heart disease (CHD) now reach adolescence and adulthood as a result of advances in paediatric cardiology, surgery and other subspecialties over the past few decades. Consequently, there has been fast development in the management of this special group of patients in many countries. However, it remains a new phenomenon in China. We obtained clinical information about adults with CHD at Beijing Anzhen Hospital, China.
Methods: A total of 431 consecutive patients aged 18 years or older diagnosed with CHD from January 2005 to March 2012 were enrolled. Demographics and disease characteristics including diagnosis, haemodynamics, treatments and outcomes were recorded.
Results: Patients' age was 30.3 ± 11.0 years; 71% were female. Four hundred and one (92%) patients were diagnosed with CHD for the first time. The majority had simple left to right shunt defects, including atrial septal defect (ASD) in 182 patients (42%), ventricular septal defect (VSD) in 90 (21%) and patent arterial duct (PDA) in 78 (18%). Other defects in the remaining patients included tetralogy of Fallot and total abnormal drainage of pulmonary veins. Cardiac catheterisation was performed in 82 patients with severe pulmonary hypertension. Systolic pulmonary arterial pressure was 79.6 ± 14.7 mmHg, and pulmonary vascular resistance 24.4 ± 14.0 Wood*m^2. Thirty-five patients were diagnosed with Eisenmenger syndrome and received pulmonary vasodilator treatment. Surgical repair was performed in 129 patients, and cardiac interventions in 180 patients including VSD, ASD and PDA device closures. There was no in-hospital death.
Conclusions: In our centre, the majority of patients were first diagnosed with CHD at adulthood, with severe pulmonary arterial hypertension and increased pulmonary vascular resistance. Patients who underwent surgical or cardiac interventional repairs had good early outcomes. A formal organisation to define treatment strategies and long-term follow-up in adults with CHD is urgently needed in China.

709: THE VALUE OF A SPECIALISED PSYCHOLOGICAL SERVICE FOR ADULTS WITH CONGENITAL HEART DISEASE

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Background: Adult congenital heart disease (ACHD) patients are at increased risk of developing mood and anxiety disorders; there are few dedicated psychological programmes for this patient population. A specialised ACHD psychological service has not previously been described, but the information would be valuable to programmes considering expanding their scope to include psychological care.
Methods: A retrospective review of 100 ACHD patient psychology files was performed and the following data were abstracted: sociodemographic and medical variables, presenting psychological concerns, and course of psychological treatment.
Results: Of 100 patients, the mean age was 33 ± 11 years, 51% were female, and the majority were single ($n = 63$), educated beyond high school ($n = 70$), and had defects of moderate or great complexity ($n = 90$). The most common psychological concerns were general anxiety ($n = 82$), health/heart-related anxiety ($n = 71$), depressed mood ($n = 60$), and/or coping with a medical condition ($n = 49$); 65 patients met diagnostic criteria for a psychiatric disorder. Following assessment, individual psychotherapy with the ACHD psychological service was recommended to 87 patients, of whom 75 agreed to proceed with treatment. The median number of psychotherapy sessions was 8.

Therapy most commonly included cognitive restructuring (94%), supportive listening (90%), and/or training in skills for relaxation (56%) or communication (46%). Of 36 patients who have thus far completed the recommended course of psychotherapy, 35 had reduced or absent psychological concerns at the end of treatment.
Conclusions: The majority of patients referred to a specialised ACHD psychology service have significant mood and/or anxiety problems; over 70% present with health/heart-focused anxiety. Therefore, ACHD programmes are encouraged to integrate specialised ACHD psychological services in order to most effectively address patient needs. Reduced psychological distress can be achieved among ACHD patients who receive appropriate psychological intervention.

720: BODY MASS INDEX, PREVALENCE AND PREDICTORS OF OBESITY IN URBAN AND RURAL COMMUNITIES IN ABIA STATE, SOUTH EASTERN NIGERIA

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Background/hypothesis: Obesity is assuming an epidemic dimension globally. Currently more than 1 billion adults are overweight, and at least 300 million of them are clinically obese. There is no previous study on the prevalence and predictors of obesity in Abia State, South Eastern Nigeria, necessitating this study.
Methods: The study was a cross-sectional study aimed at ascertaining the prevalence and predictors of obesity in the state. Participations in the study were recruited from the three senatorial zones in the state. In each of the zones an urban and rural community were randomly selected. Screening for obesity was carried out in these patients using the body mass index (BMI).
Results: Fifty-three (2.1%) of the participants were underweight while 1 456 (57.4% had normal weight. Furthermore 706 (28.2%) of the participants were overweight, while 313 (12.3%) were obese. Two hundred and seventeen (8.6%) of the participants fell into class 1 obesity, 66 (2.6%) into class 2 obesity, while 30 (1.2%) had class 3 obesity.

Of the obese patients, 180 (57.5%) were urban dwellers, while 133 (42.4%) were rural dwellers. These male to female and urban to rural differences in prevalence of obesity were statistically significant.

In a multiple logistic regression analysis in which variables that were significant in the univariate analysis were entered into a model, two variables appear to independently predict obesity in our population. These were gender (odds ratio (OR) = 2.83, 95% confidence interval (CI) 2.150-3.717), $p \leq 0.001$, and income (OR = 1.51, 95% CI, 1.154-1.979, $p = 0.003$).
Conclusion: The prevalence of obesity and overweight in the state is significantly high, and there is a need for interventions to halt this trend.

722: PREVALENCE AND DETERMINANTS OF HYPERTENSION IN ABIA STATE NIGERIA: RESULTS FROM THE ABIA STATE NON-COMMUNICABLE DISEASES AND CARDIOVASCULAR RISK FACTORS SURVEY

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Background/hypothesis: Hypertension is the commonest non-communicable disease in sub-Saharan Africa. In Abia State, Nigeria, no previous study has been carried out on the prevalence and correlates of hypertension among the populace. The purpose of this study is therefore to determine the prevalence and determinants of high blood pressure in Abia State, South Eastern Nigeria.
Methods: The study was a community based cross-sectional house-to-house survey conducted in rural and urban communities in the state. Participants in the study were men and women aged 15 years and above and were recruited from the three senatorial zones in the state.
Results: A total of 2 983 consented to be interviewed. There were 1 430 men (47.9%); the mean age of the population was 41.7 ± 18.5 years (range 18–96 years). Women had significantly higher BMI than the men. Similarly waist circumference was also larger in women but waist-to-hip ratio was only significantly higher in women in the urban area. Thirty-one per cent of all the subjects had systolic hypertension (33.5% of men and 30.5% of women). This gender difference was statistically different in the urban area. Diastolic hypertension was 22.5% in all the population (23.4% in men and 25.4% in women. Age and indices of obesity were the strongest predictors of blood pressure.
Conclusion: The prevalence of hypertension was high in our study, in both rural and urban settings. The major determinants of blood pressure in our subjects included age, gender, indices of obesity and pulse rate.

824: RIGHT VENTRICLE IN CONGENITAL HEART DEFECTS WITH SEVERE PULMONARY HYPERTENSION: ANATOMIC AND HAEMODYNAMIC CONSIDERATIONS

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Background: Right ventricular (RV) adaptation mechanisms to chronic pressure and/or volume overload are hypertrophy, dilatation and decreased function. Compared to other causes, in patients with severe pulmonary arterial hypertension associated with congenital heart defects (PAH-CHD) long well-preserved RV function is typical. The aim of the study was to evaluate specific RV features in these patients and in different subtypes of PAH-CHD.
Patients and methods: A total of 56 patients (40F/16M, median age 32 years) with PAH-CHD were analysed: 41 (73.2%) post-tricuspid defects (Post-TD), 9 (16.1%) pre-tricuspid defects (Pre-TD) and 6 (10.7%) after surgery without residual shunt (NO-SHUNT). Anatomic/haemodynamic parameters were established by echocardiography and invasively; and compared to age- and sex-matched healthy controls (NORMAL).

Results: Echocardiography in PAH-CHD/NORMAL showed significant RV dilatation (RV diameter (RVD) $p < 0.0001$), RV hypertrophy (RV anterior wall (RVAW) $p < 0.0001$) and lower RV function (fractional area change (FAC) $p = 0.024$). Comparing Post-TD/Pre-TD/NO-SHUNT showed difference in median RVD 25/47/35 mm ($p = 0.0001$), with RVD > 22 mm present in 24/100/60% patients ($p = 0.0001$); median RVAW 11/7.5/8.2 mm ($p = 0.009$), with RVAW > 10 mm in 63.2/12.5/16.7% patients ($p = 0.01$), median FAC 60/43/41% ($p = 0.003$), with decreased RV function FAC < 40% present in 5.3/37.5/50% patients ($p = 0.02$). Comparing Post-TD/Pre-TD/NO-SHUNT showed significant differences in the following invasive parameters: median mean pulmonary arterial pressure (mPAP) 78/41/49 mmHg ($p = 0.0014$); median systolic pulmonary-to-systemic pressure ratio (sPAP/sAoP) 1.06/0.61/0.6 ($p = 0.0011$), with suprasystemic sPAP present in 72.2/0/16.7% patients ($p = 0.02$); and median pulmonary vascular resistance (PVR) 18/7.4/8.2WU ($p = 0.08$).
Conclusions: Compared to NORMAL, in PAH-CHD the RV shows compensatory hypertrophy and dilatation. Patients with various cardiac shunt locations demonstrate different haemodynamics. Post-TD patients are able to tolerate extreme PAP/ PVR, they show most dominant RV hypertrophy but less severe RV dilatation and good RV function. On the contrary, Pre-TD/NO-SHUNT present with a less favourable picture. Defect location may therefore play an important role, enabling better RV off-load and so preserving RV function - which is crucial for patients' long-term outcome.

893: NEUROPSYCHOLOGICAL ASSESSMENTS OF 40 ADOLESCENTS AFTER CONGENITAL HEART DEFECT REPAIR

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Congenital heart defects (CHD) are a risk factor in cognitive development. A growing number of post-CHD adolescents increases incidence of neurodevelopmental abnormalities.
Objective: Neuropsychological assessment of adolescent's post-surgical and/or interventional procedures as a result of CHD in childhood.
Material and methods: We assessed 40 patients (pts) aged 17.6 ± 19.1 years (28 with simple, 12 with complex CHD), who because of age had been transferred to adult centres by means of Wechsler Intelligence Scales and clinical trials assessing memory, attention, praxis, abstract thinking and visuospatial functions.
Results: Only 8 pts (1 post-dextro-transposition of the great arteries (dTGA), 3 post-coarctation of the aorta (CoAo), 2 post-atrial septal defect $(ASD)_2$, 2 post-ventricular septal defect (VSD) repair) scored normally, while the others demonstrated cognitive difficulties typical of organic central nervous system (CNS) dysfunction/damage. Three pts (1 post-PDA, 1 post-VSD, 1 post-CoAo repair) showed only visual memory impairment. Thirty-two pts revealed frontal region dysfunctions, while 20 pts additionally showed visual memory impairment (characteristic of right temporal and frontal dysfunction). In 1 pt with complex CHD (post-tetralogy of Fallot repair) and in 8 simple CHD pts (2 post-PVS, 2 post-CoAo, 2 post-ASD_2, 1 post-VSD, 1 post-AVS), visuospatial impairment was noted, typical for temporoparieto-occipital lesions. In 12 complex CHD pts, the number and severity of cognitive dysfunctions were greater: 11/12 pts had executive and learning dysfunctions, 10/12 disorders with three error types (perseverative, confabulation, meaning changes), 8/12 visuospatial and visual memory disorders. Two pts had only frontal dysfunction, one temporal, while others had both types of dysfunction. In the post-simple CHD repair group, 71.4% revealed disorders typical for frontal and 10.9% temporal, while 28.5% had temporoparieto-occipital dysfunction. Most of these pts (85.7%) committed only one type of error (confabulation).
Conclusions: Of most commonly demonstrated neurodevelopmental abnormalities, grown up congenital heart disease (GUCH) patients show executive function impairment.

In post-simple CHD pts, frontal region dysfunctions are less severe than in pts with post-complex CHD and isolated types of cognitive dysfunctions are more common.

910: THE RELATIONSHIP BETWEEN BIRTH WEIGHT AND OTHER ANTHROPOMETRIC PARAMETERS AND AORTIC INTIMA-MEDIA THICKNESS AT ONE MONTH, IN UNSELECTED BIRTH COHORT

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Background/hypothesis: Evidence suggests that early-life fetal programming can have long-term effects on the adult health. A number of small studies report an association between intrauterine growth restriction and a recently validated and reproducible marker of early atherosclerosis - aortic intima-media thickness (aIMT) in early life. However there are currently no aIMT data from an unselected population of infants, nor on the relationship between aIMT and body composition or growth parameters other than birth weight.
Materials and methods: Infants are assessed as part of The Barwon Infant Study, an unselected birth cohort of Australian mothers and babies, recruited before 28 weeks' gestation ($n = 1\,250$). To date 651 babies have been recruited, with > 80% participant retention to 12 months. Aortic IMT measurements have been obtained on > 95% of those babies attempted at 4 weeks of age. Birth weight, length, head circumference and placental weight are recorded and adiposity is assessed by standardised skinfold thickness measurements in the first week of life. Maternal weight and weight change in pregnancy is recorded, together with co-morbities such as gestational diabetes and pre-eclampsia. At 4 weeks of age aIMT of the posterior wall of the abdominal aorta is measured by trans-abdominal ultrasound using standardised and reproducible methods.
Results: Data on the first 500 babies will be available by early 2013. We will present data on the relationship between placental weight, birth weight, head circumference, length, skinfold thickness and the 4-week aIMT.
Conclusions: This will be the largest study of the association between birth weight and other growth parameters, determined by the *in utero* environment, and a validated early marker of atherosclerosis.

917: REACTIVE HYPERAEMIA INDEX AND DETECTION OF ENDOTHELIAL DYSFUNCTION IN HIGH-RISK CHILDREN

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Background: Reactive hyperemic index (RHI) is considered as a new indicator of endothelial function (EF). This plethysmographic method is based on non-invasive assessment of endothelium-dependent changes in vascular tone (PAT) in patient fingertips. Type 1 diabetes mellitus (T1DM) is a well-known a risk factor for development of premature cardiovascular disease. As a result of highly efficient treatment protocols of acute lymphoblastic leukaemia (ALL) in children, the number of children leukaemia survivors has dramatically risen. Data on the premature manifestation of atherosclerosis (PMA) in ALL children are insufficient in the literature.
Materials and methods: Following approval by the institutional review board 73 eligible participants were enrolled in the study (33

T1DM (16 ± 2.2 yrs) and 28 ALL patients (14 ± 3.4 yrs) matched with 12 healthy controls (HC) (16 ± 1.7 yrs). Endo-PAT recorder was used for the determination of RHI and specific biochemical markers of EF were assessed (hsCRP, ADMA, E-selectin, VCAM). RHI was evaluated in T1DM and ALL children and compared with HC. In addition, RHI was correlated with anthropometric and biochemical parameters.

Results: Significantly lower RHI were revealed in T1DM and ALL patients in comparison with HC (1.44 ± 0.41, 1,57 ± 0.50 and 1.99 ± 0.68; $p \leq 0.05$ respectively) implying impaired endothelial-dependent dilation. No association was revealed between RHI and anthropometric parameters, arterial blood pressure or glycated haemoglobin in both T1DM and ALL groups.

Conclusions: Our results support the hypothesis of impaired EF in T1DM children which is in agreement with previously published data in adults. Our study also showed impaired EF in ALL patients. We believe that a non-invasive method such as RHI is a promising future prospect for EF assessment in children with high risk of PMA and may aid tailoring their treatment strategies.

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961: KEY POINTS OF CHEST PAIN IN CHILDREN AND ADOLESCENTS

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Objective: Paediatric cardiologists frequently encounter chest pain in children and adolescents. This study is performed to characterise the common complaint and to emphasise the significance of the symptom.

Methods: The medical records of children and adolescents under 19 years old who presented to Seoul National University Bundang Hospital, Gyeonggi-do, Korea between 10 May 2003 and 30 June 2012 with a complaint of chest pain were reviewed retrospectively.

Results: Data of 479 patients were reviewed. The proportion of males to females was 1.52 (289 males and 190 females) and the median age was 9.16 years old (range: 2.44–18.79 years). Cardiac causes of chest pain were decided in only 4 cases (0.8%). The remaining 475 patients had chest pain of unknown (264), musculoskeletal (155), psychiatric-related (28), pulmonary (14) or gastrointestinal (12) origin. During the follow-up period, only 34 patients (7.1%) had medication or interventional procedures. Interestingly, 89.4% (428) had no more anxiety about chest pain after reassurance from a doctor; however 49 patients (10.2%) still suffered from sustained chest pain, headache, abdominal pain and so on. There were no deaths during follow-up and 2 patients were diagnosed with breast mass.

Conclusion: Chest pain in children and adolescents is a very common problem but it rarely has a cardiac cause. However, in 10% of patients the problem remained unsolved. Although it is not a life-threatening condition as in adults, more meticulous history taking and a careful approach is needed.

1072: TRANSITION 'READINESS' AMONG YOUNG ADULTS WITH CONGENITAL HEART DISEASE

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Background: Given the lifelong nature of congenital heart disease (CHD), adolescents and young adults with CHD are expected to assume increasing responsibility for their health care management independent of their parents/guardians. The Transition Readiness Assessment Questionnaire (TRAQ) is a 29-item questionnaire with two domains (self-management, self-advocacy) designed to measure transition 'readiness' among adolescents and young adults with chronic health conditions. The TRAQ has not previously been evaluated in the CHD population. We investigated whether TRAQ scores differed by age, defect complexity (moderate vs great), and/or clinic in which patients are followed (paediatric vs adult).

Methods: Cross-sectional study of 18–25-year-olds with moderate or complex CHD managed in Edmonton or Toronto, Canada. Subjects completed the TRAQ following a CHD clinic visit. TRAQ scores have a range of 1–5, with higher scores reflecting greater independence.

Results: Of 128 patients, 80 (63%) had defects of moderate complexity and 48 (37%) had defects of great complexity; 55 (43%) were female. Self-management scores were significantly higher among 23–25-year-olds than 18–19-year-olds (4.4 ± 0.6 vs 3.6 ± 0.9, $p < 0.0001$). Self-advocacy scores were also higher among 23–25-year-olds compared to 18–19-year-olds (4.6 ± 0.5 vs 4.0 ± 0.7, $p < 0.0001$). Neither self-management scores nor self-advocacy scores differed significantly between patients with defects of moderate vs great complexity or between patients followed in paediatric vs adult CHD clinics.

Conclusions: Among young adults with CHD, transition readiness does not appear to be influenced by the severity of the CHD lesion or whether patients are followed in a paediatric vs adult CHD clinic. What is apparent, however, is that self-management and self-advocacy both improve with increasing age. This is consistent with the notion of a 'successful' transition process in which young people with CHD gradually assume increasing responsibility for their health care.

1078: COMMITTED TO LIFE: ADOLESCENTS AND YOUNG ADULTS' EXPERIENCES OF LIVING WITH FONTAN CIRCULATION

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Background: Single-ventricle defects are among the most complex congenital heart defects and the development of advanced surgical procedures in the last decade has created the first generation of adolescents and young adults living with this condition. Yet little is known about how these individuals experience life and what impact the heart defect has on their life in general.

Aim: The aim was to illuminate and gain a deeper understanding of adolescents' and young adults' experiences of living with a surgically palliated univentricular heart.

Method: All adolescents and young adults operated before 1995 according to the Fontan or the total cavo-pulmonary connection procedure, at one paediatric cardiology unit were included in the study. They were 17–32 years of age (mean age 22 years). Seven open-ended in-depth interviews were conducted, transcribed and analysed according to the phenomenological hermeneutical method.

Results: The interpretation of the interview texts showed that the participants experienced living with a surgically palliated univentricular heart in terms of feeling exceptional, strong and healthy. This was supported by two structural analyses, where three themes emerged: happiness about being me, focusing on possibilities and being committed to life.

Conclusion: Living with a Fontan circulation included negative experiences but the analyses clearly demonstrated a feeling of being strong and healthy. An appreciation of having survived and being committed to life was found to be part in the development of the interviewees' existential growth. This probably strengthens them further in their ability to balance expectations and hurdles in life.

1110: TECHNOLOGY AND TEENAGERS - DEVELOPING A MOBILE PHONE APP FOR TRANSITIONAL CARE

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Background: Children are now surviving into adult life with conditions which were previously fatal in childhood. These young people often require complex interventions from numerous members of the multidisciplinary team and there is an obligation to ensure that their health care needs are met as they move into adulthood. The National Service Framework for Children, Young People and Maternity services have highlighted transitional care as an integral component for all young people including those with chronic illness and/or disability. Transition is further supported by the intercollegiate report 'Bridging the Gaps: Health Care for Adolescents', guidelines from the Royal College of Nursing, 'You're Welcome' standards for adolescent-friendly services. There is now an evolving evidence base for development in this area and the unmet transitional care needs of young people are well documented. There is an emphasis on increasing the knowledge of young adolescents about their condition, treatment and the personnel concerned with their care. Previously paper records had been given to young people; however the clinic nurses identified that young people did not return to clinic with these.
Method: In discussion with young people in transition and those in the adult clinic who had recently transitioned, it was identified that a phone app for use by patients as a health passport would be beneficial to the young people during a difficult time.
Results/conclusion: Phase 1 was the development of the app and initial trial involving young people at all stages. Phase 2 will happen later in the year as we build on the initial design, modify the app and roll out to all. We believe that technology is the way to engage young people in their own care and that this design can be shared with colleagues in other centres and across other specialties.

1135: PSYCHOSOCIAL MATURITY, QUALITY OF LIFE, AND PARENTAL FOSTERING OF AUTONOMY AMONG YOUNG ADULTS WITH CONGENITAL HEART DISEASE

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Background: Congenital heart disease (CHD) may impose psychosocial challenges for youth and their parents, especially during the transition to adult health services. We compared psychosocial maturity, quality of life, perceived health status, and parental fostering of autonomy among young adults with CHD of moderate vs great complexity, and between those followed in a paediatric vs adult clinic.
Methods: Cross-sectional study of 18–25-year-olds with CHD of moderate or great complexity managed in Edmonton or Toronto, Canada. Subjects completed the Satisfaction with Life Scale (SWLS), Short Form 12 health status survey (SF-12), Erikson Psychosocial Stage Inventory (EPSI), and Kenny's Parental Attachment Questionnaire (PAQ, a measure of parental fostering of child autonomy). Student t tests and Pearson's product-moment correlations were performed.
Results: A total of 164 subjects participated, 106 (65%) with moderate and 58 (35%) with complex CHD; 71 subjects (43%) were female. Mean age was 21.4 ± 2.4 vs 21.9 ± 2.3 years among those followed in a paediatric vs adult setting, respectively (p = 0.29). Mean SWLS score was 27.2 ± 5.4 among patients (pts) with moderate CHD, compared with 25.8 ± 6.7 among pts with complex CHD (p = 0.17). SF-12, EPSI, and PAQ scores also did not differ by defect complexity. Pts followed by a paediatric cardiologist had more favourable PAQ scores compared to those followed by an adult cardiologist (55.7 ± 10.9 vs 50.9 ± 12.3, p = 0.02). Within the total sample, SF-12 mental component summary scores correlated moderately with SWLS scores (r = 0.52, p < 0.0001).
Conclusions: Somewhat unexpectedly, young adults followed in the paediatric setting reported higher parental fostering of their autonomy than those followed in the adult setting. Survey scores did not significantly differ between patients with defects of moderate vs great complexity. These results suggest that defect complexity should not be a focus when considering the transition needs of young adults with CHD.

1136: PREPARING YOUTH FOR TRANSITION FROM PAEDIATRIC TO ADULT CONGENITAL HEART PROGRAMMES: AN INTERVENTION PROTOCOL FROM EDMONTON, CANADA

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Background: Many adolescents living with CHD have limited knowledge of their cardiac lesion or prior interventions, impairing their potential to transition successfully from child-centred to adult-oriented care. Although transition programmes have been introduced by some centres, there are no outcome data with respect to CHD transition.
Methods: A 1-hour nurse-led one-on-one clinic-based intervention preparing 15–17-year-olds with moderate or complex CHD was developed in Edmonton, Canada and tested on 24 subjects. The aim of the intervention was to improve the knowledge level of the adolescent regarding their CHD. The intervention protocol included a) review of a diagram depicting the subject's cardiac anatomy and prior surgical or catheter interventions; b) review of potential late cardiac complications; c) name, dose and rationale for the subject's cardiac medications; and d) creation of a portable health summary (MyHealth passport). The intervention was conducted the same day as the teen's cardiology appointment in most cases. Email or texting (teen choice) was employed by the nurse as a follow-up in the week after the intervention. Intervention effectiveness was evaluated through a CHD knowledge questionnaire (MyHeart score) administered pre intervention and 1 month post intervention.
Results: The intervention lasted 67.8 ± 17.9 minutes. The nurse was able to complete the intervention with every subject. The nurse reached 17 of the 24 teens for a follow-up contact; 13 by texting, 4 by email and 0 by phone. Mean MyHeart score improved from 70.5 ± 16.5% correct responses pre-intervention to 79.3 ± 13.3% correct responses 1 month post intervention (p = 0.003).
Conclusion: This transition intervention was feasible in the outpatient setting. A modest improvement in CHD knowledge was demonstrated 1 month post intervention. Further study is required to demonstrate the impact of transition interventions on self-management skills among adolescents with CHD. A two-stage, multicentre intervention is being planned by our group.

1186: SELF-REPORTED PHYSICAL FUNCTIONING MAY BE MISLEADING IN PREDICTING ACTUAL EXERCISE CAPACITY IN ADULTS WITH CONGENITAL HEART DISEASE

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Objectives: To compare self-reported physical functioning with the results of cardiopulmonary exercise test (CPX) in adults with congenital heart disease (ACHD).
Patients and methods: Between November 2010 and October 2011, 76 ACHD (≥18 years) received a questionnaire survey regarding quality of life (QoL) and self-estimated exercise performance, as well as a symptom-limited CPX on the same day. The QoL was investigated using the Taiwanese version of the QoL questionnaire designed by the World Health Organization, and the subscores of physical domain were extracted. Self-estimated exercise performance was defined as the subjective prediction of exercise capacity compared to the general population and expressed as a percentage. Maximal oxygen consumption (VO_2max) in CPX was expressed as a percentage of predicted VO_2max for age and sex to represent the actual exercise capacity. Overestimation of exercise capacity was defined as the percentage of predicted VO_2max lower than self-estimated exercise performance.
Results: After excluding 10 patients (6 had premature termination of CPX and 4 had missing questionnaire data), a total of 66 patients (age 31.8 ± 11.7 years; 67% women) were studied; 70% of patients were classified as having moderate to severe CHD. Overall, ACHD had significantly decreased exercise capacity (VO_2max: 65.4 ± 13.2% of the predicted value), which differed among different CHD severities ($p = 0.043$). Although self-estimated exercise performance correlated with actual exercise capacity ($r = 0.345$, $p = 0.005$), overestimation of actual exercise capacity is common (59%), regardless of age, sex and disease severity. In our ACHD cohort, physical QoL score was not related to actual exercise test results. Furthermore, a higher physical QoL score may paradoxically increase the probability of overestimation of the actual exercise capacity in multivariate logistic regression (odds ratio: 1.34, $p = 0.039$).
Conclusions: Overestimation of actual exercise capacity is common in ACHD. A higher self-reported physical functioning did not necessarily predict a better exercise test result, and may be even more frequently observed in patients who overestimated their own actual exercise capacity.

1201: CAROTID ELASTOGRAPHIC ELASTIC-PRESSURE-MODULUS: A NEW PREDICITIVE FACTOR OF VASCULAR FUNCTION IN LONG-TERM CARDIOVASCULAR HEALTH STATUS EVALUATION OF PATIENTS BORN WITH IUGR

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Introduction: Several studies have suggested that intrauterine growth restriction (IUGR) increases the risk of cardiovascular disease and early atherosclerosis. Early detection of vascular modulation is essential for implementing early intervention.
Objective: To assess cardiovascular health status of patients born with IUGR at an earlier stage than adulthood using known and novel non-invasive techniques.
Material and methods: We evaluated 18 adolescents born with IUGR and formerly evaluated *in utero* in our fetal echocardiographic laboratory. Data were compared to those obtained in 17 controls with normal fetal cardiovascular profile. Cases and controls were assessed at 13.96 ± 0.51 versus 14.72 ± 1.02 years old. Work-up included ascending aortic diameters and peak flows, pulse wave transit time around the aortic arch with simultaneous blood pressure (BP) recording. Aortic pulse wave velocity (PWV), input and characteristic impedances (Zi, Zc), stiffness index and elastic modulus of the ascending aorta (AA-Ep) were calculated. Common carotid intima media thickness (CC-IMT) and elastographic elastic modulus (CC-eEp) (a novel non-invasive method studying the carotid artery wall response to stroke volume) were calculated.
Results: IUGR subjects are shorter and smaller than controls, but comparable in BMI. IUGR subjects yielded higher systolic BP. In contrast to a previous report, classic biophysical properties of the aorta were comparable to those in controls; aortic impedance indices showed a supernormal adaptive response with a preserved stiffness index. From the carotid artery perspective and despite a normal CC-IMT, IUGR subjects had a significantly higher CC-eEp.
Conclusions: IUGR subjects present higher systolic BP and CC-eEp at adolescence, which is a probable precise predictive factor of vascular dysfunction. Our findings in the carotid arteries seem to detect intrinsic anomaly of the homeostasis of its arterial wall.

1211: SURGICAL OUTCOMES IN ADULTS WITH CONGENITAL HEART DISEASE: SINGLE CENTRE EXPERIENCE FROM THE DEVELOPING WORLD

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Background: Lack of data exists on the spectrum and surgical outcomes of congenital heart surgery (CHS) in adults in the developing world. With the number of adults requiring CHS being projected to rise, we undertook this study to improve our understanding of this patient population.
Materials and methods: Data were collected of all patients 18 years and older with congenital heart disease, undergoing a surgical procedure between January 1998 and May 2012 at our institution. Categorisation of collected data was done according to the Society of Thoracic Surgeons Congenital Heart Surgery database data collection form. Outcomes data were analysed and compared with available European and developing world data.
Results: A total of 161 procedures were performed in 157 patients. Mean age at surgery was 30.1 ± 11.3 years (47.3% male). Septal defects and right heart lesions constituted two-thirds of the diagnoses. Diagnostic preoperative cardiac catheterisation was performed in 86.3% of patients. Incidence of preoperative risk factors was 18.7%, with endocarditis and severe pulmonary hypertension being the most frequent. Operative mortality was 1.2% ($n = 2$). Postoperative complications occurred in 24.2% of patients. Re-operative procedures constituted 26.3% of procedures performed, one-third of which were repeat re-operative procedures. Right ventricle to pulmonary artery conduit placement constituted 57% of the re-operations.
Conclusions: Although the incidence of complications is high, operative mortality is low in the setting of a tertiary referral centre with dedicated congenital cardiac surgeons. The clinical profile of our patient population was similar to that reported in a multicentre European series. The spectrum of patients and incidence of re-operations did however differ from recently reported series from developing countries. Our utilisation of cardiac catheterisation as diagnostic modality was excessive.

1217: COMPARING THE SPECTRUM OF HEART DISEASE IN TWO CITIES IN SUB-SAHARAN AFRICA UNDERGOING SOCIO-ECONOMIC TRANSITION

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Background: With rapid westernisation in sub-Saharan Africa, cardiovascular disease is gradually becoming the major cause of morbidity and mortality in this part of the world. Unfortunately, there is still a dearth of data on the pattern of heart disease in sub-Saharan Africa given the size and diversity of the region and population. We therefore studied the pattern of heart disease in Abuja, Nigeria and compared our findings with similar data derived from the Heart of Soweto Study.
Methods: We prospectively studied 1 515 subjects with confirmed cardiac diseases referred to the cardiology clinic of the University of Abuja Teaching Hospital during a 4-year period. We then developed a prospectively designed registry and gathered detailed clinical data.
Results: There were slightly more women (50.7%) than men, and the mean age of the study cohort was 49.0 ± 13.7 years. Hypertension was the primary diagnosis in around two-thirds of the study cohort, comprising more women than men (odds ratio (OR) 1.96 95% confidence interval (CI) 1.26–2.65). Hypertension was also the commonest cause of heart failure (HF) accounting for HF in 60.6% of cases. The Abuja cohort were more likely to present with a primary diagnosis of hypertension (adjusted OR 2.10, 95% CI 1.85–2.42), hypertensive heart disease/failure (OR 2.48, 95% CI 2.18–2.83); $p < 0.001$ for both comparisons and representing more than two-thirds of presentations in Abuja. Alternatively, they were far less likely to present with coronary artery disease (OR 0.04, 95% CI 0.02–0.11), DCMO (OR 0.35, 95% CI 0.26–0.46), right HF (0.09, 95% CI 0.05–0.17) and valve disease (predominantly RHD in both sites, OR 0.21, 95% CI 0.16–0.28); $p < 0.001$ for all comparisons.
Conclusions: Hypertension and its complications is the commonest cardiovascular disease in the Nigerian population in Abuja, and unlike in Soweto, coronary artery disease is not common.

1253: EXERCISE TESTING IN COARCTATION OF AORTA: AN UPDATE

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Background: Long-term follow up of coarctation of aorta (COA) poses issues of optimal evaluation non-invasively. Routine clinical and ambulatory/home blood pressure (BP) measurements and echo may not provide enough answers.
Aim: To assess usefulness of exercise testing (ET) in COA follow-up (fu).
Patients and methods: Forty patients (pts) (7–32 yrs, median 21.06) (29 male) were followed up to 20 years post COA repair (surgical 29, native 11).
Methods: Modified Bruce treadmill protocol was done with VO2 uptake measured where possible. Tests were undertaken to evaluate BP, endurance and VO2max responses to exercise where there was suspicion of hypertension, aortic arch hypoplasia and questionable response to medication. Pts had no residual pressure gradients. Seventeen pts had two or more studies during individual fu. Response to stenting and growth of patients during fu were important factors needing repeat studies. Twenty-three pts had one study (4 pre stents and 19 post stent/surgery).
Results: Nine pts had hypertensive peak BP response, 5 resolving post stenting, 1 unchanged post stenting (with weight gain), 3 waiting for stent and 1 resolving with medication. Endurance time and VO2 max tended to improve post stenting. Seventeen pts had normal pre and peak BP response and 13 pts had borderline pre BP and normal peak BP response not requiring active management. One pt had marked improvement with endurance time post stent but still had hypertensive peak BP response (super athlete).
Conclusion: ET allows peak BP response to be assessed and appropriate interventions/surgery/medication undertaken. ET is a useful tool to evaluate optimal long-term management for COA.

1254: COARCTATION OF AORTA: 31-YEAR FOLLOW-UP

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Background: Coarctation of aorta (COA), although eminently treatable, continues to present with ongoing morbidity/mortality even after good treatment. This study explores potential areas of improvement in follow-up (fu) by highlighting detection of hypoplasia (hao)/distortion of aortic arch (ao)
Aim: We analysed the fu of 70 patients (pts) with special emphasis on ongoing persisting hypertension (hbp).
Patients and methods: Fu to 31 years was analysed for 70 pts (6–36 yrs, median 19.96 yrs; 22 female); surgery infancy/early childhood: 30 subclavian flap, 12 end to end; 27 native ballooned (ba) – 7 infants. Records were analysed for initial and subsequent adequacy of treatment, upper and lower limb blood pressure (BP), periodic echo ambulatory/home blood pressure (amb bp/home bp), and especially during periods of rapid growth (adolescence), exercise testing, and imaging of ao by computed tomography/magnetic resonance imaging (CT/MRI) with hbp and suspicion of hao/ao distortion. No pressure gradient at catheterisation does not mean good outcome in presence of hbp. Where hao (indexed) was present, stenting of ao was undertaken to normalised (indexed) ao to produce BP improvement. Stented pts fu was 1–4 yrs.
Results: Three infants/toddlers had failed ba with resultant surgery; 2 still had hbp 2–4 yrs post surgery. Many pts tend to develop hbp at puberty. Of 22 pts followed up post ba, 10 remained stable, and 12 had stents. Post surgery 42 pts, 22 pts had stents, 8 remain normotensive, 5 await stents. Post stenting, there is improvement in exercise endurance and BP control. One pt needed surgery, with adequate BP response. Stented pts tend to maintain BP response thus far.
Conclusion: Persistent hbp warrants imaging of the ao. Many COA pts develop hbp with proven hao (indexed) and/or distortion and are improved with stenting and/or surgery. Further long term fu is necessary.

1288: LATE PULMONARY VALVE REPLACEMENT IN PATIENTS WITH TETRALOGY OF FALLOT REPAIRED IN INFANCY AND CHILDHOOD

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Background: Total repair of tetralogy of Fallot (TOF) has a good long-term prognosis, but the frequent chronic pulmonary regurgitation (PR) established after the correction can lead to exercise intolerance, right ventricular (RV) dilatation and failure, deleterious arrhythmias and even sudden death. Pulmonary valve replacement (PVR) may improve and/or avoid these situations.
Material and methods: As the ideal substitute for PVR remains a source of dispute, since January 2004, in our Unit, we have been implanting the bovine pericardium biological prosthesis. This study was retrospective at its inception and prospective during the later years. Eighty-two patients were operated on between January 2004 and June 2012. Median age was 22.15 years. The average interval between TOF repair and PVR was 17.2 years.
Results: Two patients died in the early postoperative period. Mean follow-up has been 3.5 years (0.2–8.5 yrs). There was one late death after heart transplant for persisting biventricular failure. Of surviving patients 95% are classified as NYHA class I. The postoperative (> 1 yr postop) assessment of RV volumes and function using magnetic resonance imaging (MRI) improved significantly compared to MRI preoperative data. Mean postoperative peak transprosthetic echo-Doppler gradient at follow-up is 19.38 ± 14.47 mmHg.

Conclusions: PVR can be performed with low operative mortality. The bovine pericardium biological prosthesis presents good short- and mid-term results, although a longer follow-up is necessary to confirm the initial results regarding its haemodynamics and durability. As with the ideal substitute for PVR, the optimal timing of PVR remains controversial.

1323: ADULT CONGENITAL HEART DISEASE SUPERVISION IN REGIONAL NEW ZEALAND

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Background: Patients born with congenital heart disease now commonly live into adult life. Over the last 10 years the care model in New Zealand for adult congenital heart disease (ACHD) has changed from follow-up centrally in Auckland to regional follow-up in conjunction with Starship ACHD service. Tauranga Hospital (TH) drains a population of just over 210 000 and is approximately a 3-hour drive from Auckland.
Patients and methods: Our research describes the cohort of 103 patients with ACHD under active follow-up at TH at the end of August 2012 for the purposes of audit as well as service planning. Median age is 27 y (range 15–79 y); females 50.5%. The primary congenital cardiac lesion was classified as severe in 46%, simple shunt in 23%; the other 31% in accordance with the schema used by Mackie *et al*. In addition, 3 patients have Eisenmenger physiology and 1 patient has uncorrected cyanotic congenital heart disease. In the 2 years until August 2012 196 clinic appointments were attended, and 140 echocardiograms and 27 cardiac magnetic resonance imaging (MRI) scans performed. While under local follow-up 14 patients have undergone cardiac surgery (including a single cardiac transplant) and there have been 16 pregnancies resulting in 13 live births. Five patients have died (age range 23–55 y) and are not included in the main analysis. Two of the five patients suffered from Eisenmenger syndrome.
Conclusions: A significant proportion of ACHD patients under local follow-up at TH have severe or complex congenital heart lesions. Although ACHD patients represent a small proportion of patients seen in the outpatient setting, they utilise a significant amount of health resource, particularly during pregnancy. Furthermore, given that most ACHD patients require lifelong follow-up, clinical leaders should account for this group when projecting for growth in service demands.

1366: THE LATE OUTCOMES OF SURGICAL TREATMENT OF SCIMITAR SYNDROME: NO SIMPLE SOLUTION TO A COMPLEX FORM OF ANOMALOUS PULMONARY VENOUS DRAINAGE

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Background: Scimitar syndrome is a rare congenital cardiac defect where right-sided pulmonary veins drain to the inferior vena cava. Complications arise in adulthood in both patients who have had surgical correction by intra-atrial baffle, and those who have had no intervention.
Methods: We contrast the presentation, treatment plans and outcomes of 18 patients (11 female) with mean age 41 yrs followed between 1978 and 2012 at an adult congenital heart disease unit in Brisbane, Australia. Age of surgical repair in 13 cases was a mean of 28.1 yrs (range 3–72). Five patients were never operated on. Clinical and imaging follow-up was obtained.
Results: A broad spectrum of presenting complaints was seen. Associated lesions included right lung hypoplasia and abnormal right lung arterial supply in 12 patients (66%) and atrial septal defect (ASD) in 3 (17%). The treatment plan varied between conservative management, medical therapy and surgical intervention. Complications in adulthood in the operated group included occlusion of intra-atrial baffles, right heart dilatation, haemoptysis, pulmonary hypertension and atrial arrhythmia.
Conclusions: Management of scimitar syndrome in adulthood remains complex as patients treated both conservatively and surgically develop complications. Care should be individualised.

1367: ADVANCED THERAPIES TO TREAT PULMONARY HYPERTENSION IN COMPLEX CYANOTIC CONGENITAL HEART DISEASE WITHOUT EISENMENGER PHYSIOLOGY ARE AN EFFECTIVE AND SAFE LONG-TERM TREATMENT OPTION

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Background: Significant improvement in exercise capacity has been reported in patients with Eisenmenger syndrome with advanced pulmonary hypertension (PAH) therapies. There is limited evidence of safety and efficacy in complex congenital heart disease (CHD) patients who do not have Eisenmenger physiology with shunt reversal.
Aim: To report the long-term efficacy and safety data with use of advanced PAH therapies in patients with PAH associated with complex CHD.
Methods: A prospective, single-centre study of 16 patients (11 female) with mean age 31.3 years (range 15–52 years) was done; Fontan circulation (2), pulmonary atresia with ventricular septal defect (VSD)/MAPCAs (11), truncus arteriosus (1), single ventricle with VSD/transposition of great arteries (TGA) (1), and TAPVD/atrial septal defect (ASD) (1). Thirteen patients were prescribed mono PAH therapy and 3 combination, between 2004 and 2012. Outcome measurements of oxygen saturation (SaO2), NYHA-FC, 6-minute walk test distance (6MWD) and adverse events were analysed.
Results: Mean duration of therapy was 33.7 months. Significant improvement was noted: NYHA-FC (baseline mean 3.19 to 2.4) and 6MWD (pre therapy 343.7 m to 420.3 m). There was no significant change in SaO2 or echocardiographic parameters. Two deaths occurred because of failure of Fontan circulation; 2 patients switched to sildenafil because of worsening heart failure.
Conclusion: This single-centre study suggests a significant improvement in functional class and exercise capacity after treatment of PAH in complex CHD with advanced PAH therapies.

1393: RUPTURED SINUS OF VALSALVA ANEURYSMS IN ADULT CONGENITAL HEART DISEASE - CLINICAL PRESENTATION, ECHOCARDIOGRAPHIC DIAGNOSIS AND HISTOPATHOLOGICAL FINDINGS IN 8 CASES

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Background: Sinus of Valsalva aneurysms (SVAs) are rare anomalies, often associated with congenital cardiac malformations and connective tissue disorders. Aneurysm rupture usually triggers clinical deterioration.
Materials and methods: We searched our echocardiography, histopathology and electronic patient databases between 2004 and 2012. Clinical details and procedures were obtained from case notes.
Results: A total of 8 patients (5 females; mean age 35.7 years) were studied. Aneurysm of right coronary sinus was found in 5 and non-coronary sinus in 3 cases. Rupture into the right ventricle (RV) occurred in 5 cases (3 into RV body; 2 into outflow tract) and the right atrium in 2. Five patients had congenital defects: bicuspid aortic valve (BAV) 2, ventricular septal defect (VSD) 1, resected subaortic stenosis 1. Another patient had Klippel-Feil syndrome.

Acute onset of breathlessness or heart failure was the commonest manifestation (5/8). Clinical examination revealed a continuous murmur in 7/8 patients. Two patients were asymptomatic at time of diagnosis and one with Klippel-Feil syndrome died suddenly.

Echocardiography demonstrated a continuous shunt across the rupture site in 7/8 cases. Seven patients had a successful procedure (6 surgical, 1 device closure) and remain well to date. Histopathological findings in the patients with concomitant BAV were focal disorganisation of the aortic wall elastic layers with increase in smooth-muscle cells and medial and subintimal fibrosis. In the Klippel-Feil fatal case, a linear rupture was found in the non-coronary SVA with thinning of the aneurysmal wall and medial elastin replaced by fibromyxoid connective tissue.

Conclusions: SVAs arise mainly from congenital defects of the aortic media. Sudden onset of dyspnoea with continuous murmur suggests aneurysm rupture. Rarely it can result in sudden death. Commonly coexisting cardiac lesions are BAV and VSD. Echocardiography is the key to diagnosis. Timely surgical or device closure has a good long-term outcome.

1440: PULMONARY CONDUIT FUNCTION AFTER THE ROSS OPERATION: LONGITUDINAL ANALYSIS OF THE GERMAN-DUTCH ROSS REGISTRY EXPERIENCE

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Background: The Ross operation presents with excellent mid-term results. However, the prevalence and predictors of late pulmonary conduit failure after the Ross procedure have been addressed only in few reports with small patient numbers. The objective of this study was to determine the natural dynamics of pulmonary conduit stenosis and regurgitation.

Materials and methods: Among 1 775 consecutive patients (mean age ± standard deviation (SD), 43.7 ± 12.0, range 16.1–70.5 years) that underwent the Ross procedure, 1 645 (93%) received an allograft (pulmonary 1 612, aortic 12, unknown 21), 120 (6%) received a bioprosthesis, and 5 (0.3%) received a bovine jugular vein for the reconstruction of the right ventricular outflow tract. Using non-linear longitudinal models, serial echocardiographic records (n = 6 950) from these patients were studied with a maximum echocardiographic follow-up of 22.4 years (mean ± SD, 5.5 ± 4.3 years).

Results: A slight increase in pulmonary conduit regurgitation grade was observed during follow-up. Freedom from regurgitation grade ≥ 2+ was 95% after 14 years of follow-up. Female *patient* gender, the use of an allograft (compared to bioprosthesis), male *donor* gender, antibiotic treatment of the allograft, and specific surgical adjustments were associated with a significantly higher regurgitation grade. Mean conduit gradient increased from about 4.7 mmHg at 1 month to about 10 mmHg by 14 years after the procedure. Peak gradient increased from about 8.4 mmHg at 1 month to about 18.5 mmHg by 14 years after the procedure. Smaller conduit diameter, male *patient* gender, younger *patient* age, younger *donor* age, and use of bioprosthesis were associated with significantly higher mean and peak gradient. The changes in echocardiographic measurements were mainly observed in the first 2 years after surgery.

Conclusion: Echocardiographic follow-up of pulmonary conduits shows outstanding conduit durability. Clinically important conduit regurgitation and stenosis is rare in patients after the Ross operation. Consideration of risk-associated predictors may improve pulmonary conduit outcome.

1454: A VERY RARE CASE OF UNOPERATED ADULT WITH TRICUSPID ATRESIA AND SUCCESSFUL PREGNANCY

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A 46-year-old female with tricuspid atresia, ventricular septal defect (VSD) and d-transposition of great arteries (d-TGA) was diagnosed during infancy and her family deferred surgical palliation becaue of lack of symptoms (1966). The patient transferred her care to us in 2009 for routine yearly follow-up. Her review of symptoms included cyanosis, occasional palpitations, and dyspnoea on exertion (NYHA class II) but no chest pain, dizziness or easy fatigability. She works at an office. Current medications include digoxin 0.25 mg once daily. Her obstetric history was G1P1L1A0. She had an uneventful pregnancy and delivered a healthy baby girl at 36 weeks' gestation.

The imaging studies including transthoracic echocardiogram and magnetic resonance imaging (MRI) studies revealed tricuspid atresia type IIb, d-TGA and bilateral branch pulmonary artery stenosis, and adequate sized VSD and atrial septal defects (ASDs). The electrocardiograph (ECG) showed sinus rhythm and normal QRS axis, which is usually seen in about 50% of cases with type II tricuspid atresia.

The reason for her long-term survival was very well balanced pulmonary and systemic circulation. This is facilitated mainly by adequate size of ASD which allowed blood flow from right atrium to left atrium and adequate size of VSD with free flow of blood into aorta. She also had bilateral branch pulmonary artery, controlling amount of pulmonary blood flow.

Long-term survival is very rare in unoperated tricuspid atresia patients. Only three patients including our patient have survived into the 5th decade. Our patient had long-term survival without surgery because of her favourable anatomy for this condition and well-balanced circulation. To our knowledge, this is the first report of successful pregnancy in an unrepaired tricuspid atresia with TGA.

1471: ASSOCIATION OF EMERGENCY ADMISSION WITH HOSPITAL LENGTH OF STAY IN ADULTS WITH CONGENITAL HEART DISEASE

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Background: In the field of emergency care, the increasing needs of adults with congenital heart disease (CHD) are going to add to the burden on medical resources. The aim of this study was to investigate the reasons for emergency admission and its associations with increased hospital length of stay (LOS).

Methods: We evaluated 2 720 adults with CHD in a single tertiary cardiac centre database. Patients who required emergency admission between September 1994 and December 2011 were reviewed from medical records.

Results: A total of 669 adult CHD patients (24%) required emergency admission, culminating in a total of 1 726 admissions. Mean age was 48.4 ± 18.5 years, with a female predominance (57%). Most were either studying or working (62%) and were married (64%). Atrial septal defect (43.8%), ventricular septal defect (21.5%) and tetralogy of Fallot (7.2%) formed the majority of diagnoses. One-third of the admissions were for cardiac reasons including heart failure, endocarditis, arrhythmias, haemoptysis and thromboembolism. The remaining two-thirds were admitted for non-cardiac reasons. Median hospital LOS was 14.5 ± 5.6 days. Those who were older (p = 0.02), and neither employed nor studying (p = 0.021) had longer LOS. Endocarditis accounted for longer LOS (p < 0.001); 41% of the admissions that required interdisciplinary referrals had

increased LOS ($p < 0.001$), with utilisation of diagnostic evaluation of non-cardiac causes ($p < 0.001$). Increased LOS was not associated with adverse clinical outcome ($p = 0.72$).
Conclusions: Adult CHD patients require admissions for both cardiac and non-cardiac reasons. Older age group and endocarditis were associated with increased LOS. Non-cardiac conditions required interdisciplinary resources and involved increased LOS. Understanding their diverse acute needs may be possible to improve care and outcome for these patients.

1491: MICROALBUMINURIA IN CHILDREN WITH CONGENITAL HEART DISEASE: SENSITIVE MARKER OF CYANOTIC NEPHROPATHY

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Background: As a result of advances in cardiovascular surgery in congenital heart disease (CHD), a large number of patients with CHD are now entering adulthood, and are facing many problems. Renal dysfunction is one such problem. Microalbuminuria is a sensitive marker of early-stage renal impairment, and microalbumin-creatinine ratio (μALB/Cre) is reported to be a useful predictor of future cardiovascular events. We investigated whether early-stage renal impairment assessed by μALB/Cre already exists in children with CHD, and examined the determinants inducing μALB/Cre elevation.
Methods and results: We examined urinary concentrations of Cre and microalbumin, serum levels of Cre and cystatin C, arterial oxygen saturation (SaO_2) and haemodynamic variables in 150 patients with CHD (90 male, median age 5.5) who underwent cardiac catheterisation between 2011 and 2012 in our hospital. Among these patients 32% showed abnormal levels of μALB/Cre (> 30). There was no significant correlation between μALB/Cre and Cre or cystatin C. Multivariate analysis with age, sex, systemic cardiac output, blood pressure, pulmonary artery pressure, central venous pressure, and SaO_2 including independent variables demonstrated that only SaO_2 significantly correlated with μALB/Cre ($p = 0.019$). In a separate group of cyanotic CHD, there was also a statistically significant correlation between μALB/Cre and SaO_2 ($r = 0.70$ $p < 0.05$), independent of the levels of Cre and systatin C.
Conclusions: Low SaO_2 is an important determinant of renal impairment even in children. μALB/Cre appears to be more useful to detect the early phase of cyanotic nephropathy than serum Cre or cystatin C.

1518: SUCCESSFUL PERINATAL OUTCOME IN A PATIENT WITH SINGLE VENTRICLE PHYSIOLOGY WITH EISENMENGER SYNDROME

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Background: Pregnancy in patients with Eisenmenger's syndrome (ES) is associated with high maternal and foetal mortality as pulmonary hypertension (PAH) is aggravated, leading to poor outcomes. Here we report a rare case of successful outcome of pregnancy in a patient with double inlet left ventricle (DILV) with ES. Although pregnancy is discouraged in such patients, a coordinated multidisciplinary team effort for a patient presenting in late gestation is imperative.
Materials and method: A 26-year-old primipara with short stature and torticollis presented at 32 weeks' gestation; ultrasound showed a single live fetus. She was haemodynamically stable, in NYHA class II and saturating at 85% in room air. A loud second heart sound along with an ejection systolic murmur could be appreciated. Echocardiography diagnosed DILV with a large inlet ventricular septal defect (VSD) amounting to a single ventricle, unrestricted pulmonary blood flow and severe PAH. Foetal echocardiography revealed no major cardiac abnormalities. Chest X-ray alluded to established ES and she had a sinus rhythm on electrocardiograph (ECG). Inputs from the obstetrician, anaesthesiologist and neonatologist were taken. The patient was admitted for monitoring and oxygen inhalation. Heparin infusion was started and diuretics were minimised to prevent volume depletion. Intravenous betamethasone was administered for foetal lung maturity. A caesarean section was undertaken at 35 weeks' gestation along with tubal ligation. Spinal anaesthesia had to be converted to general anaesthesia for adequate analgesia. A healthy, appropriate-for- gestational-age, baby girl was delivered. The postpartum period was uneventful. The newborn's echocardiogram was normal. The mother was prescribed oral sildenafil and bosentan in the postpartum period along with advice to refrain from excessive physical exertion.
Conclusion: A multidisciplinary approach, judicious use of medications and anaesthesia and delivery at a centre with expertise in grown up congenital heart diseases is mandatory for favourable outcome in an otherwise extremely high-risk pregnancy in cases of ES.

1566: FONTAN OPERATION IN ADULTS: IS IT NECESSARY TO STAGE THE PROCEDURE?

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Background: Data on immediate postoperative results of Fontan operation in adult patients are limited. We present a single institution's experience with single-stage and staged Fontan procedures in adults.
Materials and methods: Case records of adults (> 18 years of age) who underwent Fontan operation from February 2004 to July 2012 were reviewed.
Results: Among 28 patients, 15 underwent single-stage Fontan whereas 13 patients underwent completion of staged Fontan. Mean age at surgery was 21.96 ± 1.37 years. The ventricular morphology was left in 16, right in 6 and biventricular in 6 patients. Mean pulmonary artery (PA) pressure was 13.14 ± 1.13 mmHg and mean ventricular end diastolic pressure was 11.7 ± 1.15 mmHg. Extracardiac Fontan was done in 14 and lateral tunnel was done in 14 patients. All of them had fenestration. Atrioventricular (AV) valve repair was performed in 4 patients. Two patients had pacemaker implantation during the procedure for pre-existing complete heart block. The mean cardiopulmonary bypass time was 185 ± 26.26 min and cross clamp time 56 ± 12.34 min. Among survivors, the mean duration of mechanical ventilation was 17 ± 4.39 hours and that of inotropic support was 51 ± 9.4 hours. Average intensive care unit (ICU) stay was 3.25 ± 0.42 days and chest drain duration was 7.16 ± 1.7 days. There was no statistical significance (unpaired t test) between those who had single-stage Fontan and staged Fontan completion in terms of survival and abovementioned outcome measures. Four patients (14%) died in the postoperative period; 2 patients died due to postoperative arrhythmias, 1 due to failing Fontan and 1 due to severe anaphylaxis.
Conclusion: Single-stage Fontan operation can be performed safely in the adult age group, if PA pressure is normal. There seems to be no significant difference in the early postoperative results between single-stage and staged Fontan in adults.

1582: ROLE OF ROUTINE USE OF CONTINUOUS POSITIVE AIRWAY PRESSURE THERAPY (CPAP) IN FAST TRACKING AFTER OPEN HEART SURGERY IN ADULTS WITH CONGENITAL HEART DISEASES

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Introduction: Prolonged elective ventilation after open-heart surgery in adult congenital heart disease patients for respiratory complica-

tions, high pulmonary artery pressures, poor respiratory reserve and on high inotropic support is a routine practice. Mandatory elective ventilation also has its risks and complications such as sudden tube displacement, mucus plug blockages, atelectasis, infection and bleeding due to tracheal trauma. Sudden deaths in the intensive care unit (ICU) mostly occur in patients on ventilators as a result of one of the complications mentioned above, but are under-reported. Fast tracking emerged as one of the major advances in the recent era but has limitations, requiring an extremely vigilant and expert intensive care team which most cardiac surgical ICUs lack in India. We have evolved a strategy employing routine use of continuous positive airway pressure (CPAP) therapy in all elective early weaning from mechanical ventilation and instituting CPAP mask ventilation with secured airway preserving normal swallowing, speech and feeding. Exclusion criteria include renal failure, low cardiac output state or state of confusion or non-co-operation.

Material and methods: Between January 2010 and April 2012, CPAP mask ventilation was prospectively used in 125 patients who had high respiratory or cardiac risk factors. All patients were counselled in the pre-operative period and were given a short trial of CPAP ventilation a day prior to their surgery. Analysis included retrospectively: operated age-, sex-, disease- and NYHA-matched population of 135 patients operated earlier. Patient's co-operation and absence of chest infection were mandatory requirements.

Results: Incidence of lung atelectasis, retained secretions, poor arterial blood gases (ABGs) were significantly less in the CPAP ventilation group. Cardiac arrhythmia (AF) did not differ between groups. Only 1 patient required re-intubation in the CPAP group except1, who had cardiac arrest as a result of acute severe mitral insufficiency after a failed mitral repair. Two patients in the non-CPAP group required re-intubation. Total ICU and hospital stay was significantly less in the CPAP group, who also complained of less postoperative pain and less need for physiotherapy and coughing. More importantly there was significant cost reduction in the CPAP group. Detailed data and group analysis will be presented.

1631: FREQUENCY OF MISCARRIAGE, STILLBIRTH AND PREGNANCY TERMINATION IN WOMEN WITH CONGENITAL HEART DEFECTS IN GERMANY, HUNGARY AND JAPAN

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Background: The 2011 'ESC Guidelines on the management of cardiovascular diseases during pregnancy' define maternal predictors for neonatal events (preterm birth, small for gestational age, respiratory distress syndrome, cerebral haemorrhage, foetal/neonatal death) in pregnancies in women with heart disease. It is unknown whether these predictors also lead to an increased number of miscarriages, stillbirths and terminations of pregnancy (TOP), particularly regarding patients with congenital heart defects (CHD). In the general population, miscarriages and stillbirths occur in 15–20%. In the participating countries, stillbirth occurs in below 0.5% of all pregnancies.

Material and methods: In a multicentre cross-sectional questionnaire-based study, 634 women with CHD (Germany 61%, Hungary 24%, Japan 15%) were surveyed over a period of 12 months concerning courses of pregnancy. Of 634 women, 309 had been pregnant at least once (578 pregnancies). Patients were grouped into those with and those without existing maternal predictors for neonatal events. The predictors were NYHA classification greater than class II or cyanosis, maternal left heart obstruction, smoking during pregnancy, multiple gestation, use of oral anticoagulants during pregnancy, and mechanical valve prosthesis. The outcomes were miscarriage/stillbirth and TOP (miscarriages and stillbirths were grouped together).

Results: In 75 women with predictors, a total of 141 pregnancies occurred (group 1, 24%). In 234 women without predictors, a total of 437 pregnancies occurred (group 2, 76%). There were no significant differences between the participating countries. There were 27 (19.1%) miscarriages/stillbirths in group 1 and 70 (16.0%) in group 2 ($p = 0.532$), and 22 (15.6%) and 24 (5.5%) TOP respectively ($p = 0.002$). The combined figures were 49 (34.8%) and 94 (21.5%) respectively ($p = 0.042$).

Conclusion: Underlying maternal predictors for neonatal complications do not lead to a significantly higher number of miscarriages or stillbirths. However, TOP occurred significantly more frequently in this group. In the presence of maternal predictors for neonatal events pregnancies in women with CHD are less likely to be successful.

1641: EVALUATION OF CONTRACEPTIVE METHODS IN WOMEN WITH CONGENITAL HEART DEFECTS IN GERMANY, HUNGARY AND JAPAN

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Background: The 2011 'ESC Guidelines on the management of cardiovascular diseases during pregnancy' recommend that maternal risk assessment for women with heart diseases is carried out according to the modified World Health Organization (WHO) risk classification. Particularly for women with congenital heart defects (CHD) who are at a higher pregnancy-related risk for cardiovascular complications or in whom pregnancy is contraindicated, early and effective contraception is necessary.

Material and methods: In a multicentre cross-sectional questionnaire-based study, 634 women with CHD (Germany 61%, Hungary 24%, Japan 15%) were surveyed over a period of 12 months concerning contraception. Median age was 30 years. According to the modified WHO classification of maternal cardiovascular risk, patients were grouped into three risk groups (low, medium and high/pregnancy contraindicated). The contraceptive methods (CM) used by each group were determined. In this study CM with a Pearl index ≤ 2 (at ideal use) was classified as 'safe'.

Results: In all three risk groups almost one-third of the women was using a CM classified as unsafe (low-risk 85 (27.9%); medium-risk 64 (30.5%); high-risk 20 (29.9%)). In 29% of all cases an unsafe CM was used. There was no significant difference between the participating countries.

Conclusion: Alarmingly, almost one-third of the women with CHD and increased pregnancy-related risk of cardiovascular complications or contraindication for pregnancy were using a CM deemed as unsafe. More efficient education regarding contraception in women with CHD is necessary.

1713: THE FATE OF YOUNG PEOPLE WITH FAMILIAL HYPERCHOLESTEROLAEMIA II/A TREATED BY HELP

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Familial hypercholesterolaemias represent an important group of diseases responsible for significant cardiovascular morbidity and mortality. Sustained elevation of serum cholesterol levels lead to early onset of acquired atherosclerotic heart disease.

Type II/A familial hyperlipoproteinaemias are inherited autosomal disorders with incomplete penetrance. The manifestation of the homozygous form is 1:1 000 000. Serum low-density lipoprotein (LDL) plasmapheresis techniques such as heparin-induced extracorporeal LDL-cholesterol (Ch) pheresis (HELP) are useful for controlling serum cholesterol and LDL levels. The HELP system completely removes LDL-Ch, fibrinogen and lipoprotein A (Lp(a)) from the blood. The treatment is tolerable, safe and effective.

Two cases of the homozygous form of familial hypercholesterolaemia were confirmed in childhood in Hungary. The authors present the case of a 31-year-old man whose valvular aortic stenosis was discovered at age 2. In 1989 he was examined because of xanthomatous skin alterations and familial hypercholesterolaemia was diagnosed. The extended examination of lipid metabolism revealed normal level of receptors but decreased function. His aortic stenosis was operated on at Munich in 1994 and was followed by regular plasmapheresis and oral statin treatment. He has been on this combined treatment from 9 years of age and recently he had no sign of cardiovascular disease!

An 18-year-old girl was examined first at 7 years of age because of granulomatous and xanthomatous tendon lesions. The extended examination of lipid metabolism confirmed the homozygous form of familial hypercholesterolaemia. Her treatment was started with cholesterol plasmapheresis using the HELP system combined with esetimide and rasorvastatin therapy. She is on this combined treatment without any sign of cardiovascular disease. The authors consider it worthwhile to present the two cases because of the young age of the patients, the unusual presentations of the disease and the long-term successful therapy by HELP.

1721: LONG-TERM ADVANCED THERAPY IN EISENMENGER SYNDROME: SERIAL RIGHT HEART CATHETERISATION AND 6 MINUTES WALK DISTANCE

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Background: Despite similarities in pulmonary vascular changes, the survival is markedly different between patients with idiopathic pulmonary arterial hypertension (IPAH) and Eisenmenger syndrome (ES). Furthermore, while patients with IPAH slowly deteriorate in spite of advanced therapy (AT) with pulmonary vasodilatators, few or no data exist regarding long-term effect of AT on haemodynamics and 6-minute walk distance (6 MWD) in ES. The aim of this study was to examine this by serial right heart catheterisation (RHC) and 6 MWD.
Methods: Nineteen adult patients with ES (15 with ventricular septal defect and 4 with atrial septal defect) were followed for 5 years with RHC before and after 3 months of AT, and then yearly. 6 MWD was performed at baseline and then every 6 months.
Results: None of the patients died or were transplanted during the study period. RHC revealed a continuous, significant improvement in pulmonary vascular resistance (PVR) (before AT 29 ± 12 vs 5 years 14 ± 5.2 Wood units, $p < 0.0001$) and pulmonary blood flow (PBF) (before AT 2.6 ± 1 vs 5 years 4.7 ± 1.8 l/min; $p < 0.0001$) over time after initiation of AT. However, the 6 MWD improved over the first 2 years, and then showed a more fluctuating pattern.
Conclusions: This study showed that AT is a beneficial but also long-lasting treatment, which improves both haemodynamics as well as 6 MWD in ES. Furthermore, these data suggest that despite inconsistent performance as measured by 6 MWD, a long-lasting improvement of haemodynamics is obtained by AT. The variation in 6 MWD may be due to conditions such as iron deficiency, which is known to influence 6 MWD. Finally, RHC is the golden standard in evaluating the effect of AT, but since ES on AT seems to be stable, yearly RCH is not necessary, but may instead be performed when clinical deterioration is present.

1729: LIFE EXPECTANCY AND CAUSES OF DEATH IN A COHORT OF FONTAN CONVERSION PATIENTS

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Background/hypothesis: The development of late atrial arrhythmias following Fontan palliations is associated with 3-year mortality approaching 40% in a recent multi-centre series. Fontan conversion and arrhythmia surgery (FCAS) is performed with the expectation that life expectancy would be improved, but the durability of this circulation is not known. We sought to determine mid-term survival and causes of death in a large series of patients (pts) following FCAS.
Materials/methods: Current status of 137 consecutive pts undergoing 138 FCAS at a single institution between 1994 and 2012 was reviewed. Current status was ascertained by phone interview with patients or review of physicians' records. Age at last follow-up, time to heart transplantation (OHT), and causes of death were assessed.
Results: Median age at original Fontan surgery was 5.7 yrs (range 1.1–34.9 yrs), and at FCAS was 23.6 yrs (range, 2.6–47.3 yrs). Ventricular morphology was single left in 100, single right in 13, and complex/indeterminate in 24 pts. Early operative mortality was 1.5%. Of 135 early survivors, 16% (22/135) died (13) or underwent OHT (9). OHT occurred at a mean post-FCAS interval of 31 months; 4/9 died in the peri-transplant period. Of the remaining 13 deaths, 2 died of non-cardiac causes. Circulatory causes of death included: sudden (3), congestive heart failure (3), sepsis following urological procedure or defibrillator change (2), renal failure (1), liver failure (1), and hepatocarcinoma (1). At mean follow-up of 8.6 ± 4.2 yrs, total post-FCAS transplant-free survival is 83%. The mean current age of 113 transplant-free survivors is 32.4 ± 6.6 yrs.
Conclusions: Survival into the 4th-6th decades of life was achieved in this population of Fontan pts with predominantly single left ventricular morphology, posing challenges for ongoing medical surveillance and management of co-morbidities.

1731: CONGENITALLY CORRECTED TRANSPOSITION OF GREAT VESSELS IN A YOUNG PREGNANT PATIENT

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Background: Transposition of great vessels is a rare form of congenital heart defect and occurs in less 1% of patients with congenital heart disease. Normally the development of ventricular situs occurs during the 5th week of gestation, when twisting of the primordial heart tube to right (d-looping) places morphologic right ventricle on right side of heart and morphologic left ventricle on left side of heart. Normal development of the great arteries occurs during the 5th to 6th week of gestation and this is genetically influenced by neural crest cells. Abnormal development (congenitally corrected transposition of the great arteries (ccTGA) or l-TGA) results from looping of the primordial heart tube to the left instead of the right.
Objective/aim: To show a rare case of a congenitally corrected transposition of great vessels in a 27-year-old pregnant patient.

History and progress: The patient was first seen at our combined obstetrics and cardiology clinic, at 33 weeks' gestation. On her initial presentation she reported symptoms of grade 2 dyspnoea and intermittent episodes of palpitations. The examination revealed a dextrocardia with a 3/6 pansystolic murmur along her right parasternal area. In addition she had a palpable and loud pulmonary component of the second heart sound. Her echocardiography revealed a dextrocardia, L-type TGA and a small restrictive ventricular septal defect. The patient subsequently had an elective caesarian section and her perioperative period was uneventful. She and her baby remained haemodynamically stable. The patient declined any form of invasive stratification or intervention for her heart. She was discharged and then followed up in the cardiology outpatient department.
Discussion and conclusion: There is limited literature on transposition of great vessels in pregnancy; however it can be associated with disastrous outcomes. The main recommendations in these patients include: a) consultation with a cardiologist who has an experience with adult congenital heart disease before pregnancy; b) scheduled cardiology evaluation and follow-up during pregnancy; and c) a multidisciplinary co-ordination for labour, delivery, and postpartum periods. Long-term effect of pregnancy on right ventricular function is unclear; however there is an increased risk of heart failure and arrhythmias with tricuspid valve regurgitation. Long-term outcomes/problems depend on presence/severity of associated lesions and right ventricular (systemic ventricle) failure.

1737: COMPARATIVE EVALUATION OF NORMAL WEIGHT AND OBESE CHILDREN WITH ESSENTIAL ARTERIAL HYPERTENSION

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Aim: To compare normal weight and obese children with essential hypertension.
Materials and methods: A total of 114 patients diagnosed with primary hypertension (87 boys and 27 girls, aged 10–18 years, mean 15.2 ± 1.8 years, were analysed. Patients were divided into two groups according to body mass index (BMI): group I normal BMI (< 85 percentile), group II overweight and obese patients (BMI > 85 percentile). Analysed parameters included systolic and diastolic blood pressure (SBP, DBP), fasting glucose, total cholesterol, high-density lipoprotein (HDL) cholesterol, triglycerides levels, and heart rate at rest (HR). Mean values of the parameters in both groups were compared using Student's t test. The correlations between studied parameters and blood pressure in the selected groups were assessed by Pearson's correlation coefficient (r).
Results: Statistically significantly higher SBP, triglycerides levels, and HR were observed in overweight and obese children, and positive correlations between SBP-triglycerides levels ($r = 0.34$), HR-DBP ($r = 0.4$), DBP-BMI ($r = 0.2$), BMI-triglycerides levels ($r = 0.25$) were found. In children with normal BMI, no relationships between the studied parameters were observed.
Conclusions: The overweight and obese children with hypertension are characterised by higher blood pressure values, serum triglyceride levels and heart rate compared to children with hypertension and normal BMI. The moderate apparent correlations between blood pressure values and BMI, serum triglycerides levels and heart rate are observed in children with overweight and obesity.

1782: CAN ARRHYTHMIA SURGERY AT THE TIME OF PULMONARY VALVE REPLACEMENT PROTECT FROM FUTURE ARRHYTHMIA?

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Background/hypothesis: Longstanding pulmonary regurgitation after surgery for tetralogy of Fallot (TOF) or pulmonary stenosis leads to progressive right ventricle (RV) dilatation and arrhythmia. Pulmonary valve replacement (PVR) is often required in adult life. Arrhythmogenesis is complex, relating both to scars and ongoing haemodynamic problems. Our unit has performed surgical cryoablation in selected patients at the time of PVR since 2007.
Materials/methods: Retrospective analysis of case notes for all patients undergoing PVR by a single surgeon at our institution between 2007 and 2010.
Results: Fifty operations were performed in 47 patients; 42 had underlying TOF. The main indication for PVR was significant pulmonary regurgitation; 33 patients with arrhythmia were also referred for surgical cryoablation to the right atrium (RA) and RV outflow tract (RVOT). Mean age at operation was 27.8 y (range 14–61). Twelve patients had atrial fibrillation/flutter pre-op; all had cryoablation performed. Three (25%) had a recurrence of AF post-op. In 22 cases, RVOT cryoablation was performed; in 11 there had been pre-op ventricular tachycardia. Four patients developed VT post-op after a mean time of 12 months (range 1–30); only 2 required long-term medication. One already had an implantable cardioverter defibrillator (ICD); 1 required a new device insertion years later. Three further patients had ICDs inserted post-op: this had been planned electively pre-op and was not due to new arrhythmia. Three patients who underwent cryoablation required insertion of a pacemaker post-op for new bradyarrhythmias at a mean time of 5 months (range 0–12).

Three deaths occurred; 2 in patients post-ablation. None appear related to arrhythmia.
Conclusions: Surgical cryoablation at time of PVR appears to reduce arrhythmia recurrence in the short-term although long-term outcome data are required. Patients selected for this have already demonstrated a clinical arrhythmia burden and although initial results appear promising, whether this provides effective future protection remains to be seen.

CARDIOLOGY AND THE IMAGING REVOLUTION

1: REAL-TIME THREE-DIMENSIONAL ECHOCARDIOGRAPHIC CHARACTERISTICS OF LEFT VENTRICLE AND LEFT ATRIUM IN NORMAL CHILDREN

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Background: The accurate assessment of left atrial (LA) and/or left ventricular (LV) volume and contractility is crucial for the management of patients with congenital heart disease. The real-time three-dimensional echocardiography (RT3DE) is reported to show better correlation with magnetic resonance imaging (MRI) in estimating LV and LA volume than conventional two-dimensional echocardiography (2DE). On the other hand, the volume measurement in RT3DE is also reported to be significantly smaller than those in MRI, necessitating the establishment of normal values of RT3DE itself. The aim of this study was to identify the normal values of LV and LA volume measured by RT3DE in Japanese children.
Methods: Sixty-four normal school children [age: median 9.6 years; range (5.5–14.5); male 26, female 38] were enrolled in this study. End-diastolic and end-systolic LV and LA volumes were analysed using M-mode in short-axis view, 2D biplane method, and RT3DE. We used IE-33 (PHILIPS) with matrix probe X7 and X4 off-line assessment to calculate LA, and LV volume was done using QLAB 8.1 (Philips).
Results: Forty-nine children [age: median 9.1 years, range (6–14); male 21, female 28] had adequate RT3DE data sets and were analysed.

- RT3DE: LV end-diastolic volume index (LVEDVI) = 51.4 ± 5.7 ml/m^2, LV end-systolic volume index (LVESVI) = 21.0 ± 4.2 ml/m^2, max LA volume index (LAVI) = 21.4 ± 5.0 ml/m^2, min LAVI = 7.7 ± 2.7 ml/m^2, LV ejection fraction (LVEF) = 59.1 ± 6.9%, and LA volume change [(max LAV–min LAV)/max LAV × 100%] = 63.7 ± 9.3%.
- M-mode: LVEDVI = 70.9 ± 10.8 ml/m^2, LVESVI = 23.5 ± 5.5 ml/m^2, LVEF = 66.8 ± 6.1%.
- 2DE biplane: max LAVI = 22.8 ± 5.6ml/m^2.

LV end-diastolic volume on RT3DE showed good linear correlation with body surface area (BSA): LVEDV = –4.52 + 55.75 × BSA, R^2 = 0.746.
Conclusion: Approximately 77% of normal children had adequate RT3DE images. The discrepancy of LVEDV between RT3DE and M-mode was significant and the measurements of RT3DE were constantly smaller than those of M-mode.

4: REDUCED AORTIC ELASTICITY AND VENTRICULAR DYSFUNCTION LATE AFTER PAEDIATRIC MENINGOCOCCAL SEPTIC SHOCK: A PRECURSOR OF ATHEROSCLEROSIS?

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Background: Given the strong similarities in inflammatory pathways between septic shock and atherosclerosis, aortic wall abnormalities and associated ventricular sequelae may be expected after MSS.
Objectives: To prospectively assess aortic elasticity and biventricular systolic and diastolic function in paediatric patients after meningococcal septic shock (MSS) using magnetic resonance imaging (MRI).
Methods: Eighteen paediatric MSS survivors (eight male; age 14.5 ± .9 years; MRI 8.2 ± 2.4 years after MSS) treated with at least two inotropic and vasoconstrictive agents for > 48 hours, and 18 matched controls were studied. Routine MRI was used to assess aortic pulse-wave velocity (PWV) and systolic and diastolic biventricular function.
Results: MSS patients showed reduced aortic elasticity vs controls (PWV aortic arch: 4.1 ± 0.3 vs 3.3 ± 0.5 m/s, $p < 0.01$; PWV descending aorta: 3.9 ± 0.9 vs 3.2 ± 0.4 m/s, $p < 0.01$). Systolic biventricular function was preserved (LV ejection fraction 57 ± 8 vs 56 ± 6%, $p = 0.74$; RV ejection fraction 56 ± 8 vs 52 ± 6%, $p < 0.01$), whereas biventricular mass was increased (LV 52.1 ± 8.4 vs 36.0 ± 9.9 g/m^2, $p < 0.01$; RV 26.8 ± 6.5 vs 10.4 ± 5.0 g/m^2, $p < 0.01$). Also, delayed biventricular relaxation was found after MSS: E-wave deceleration time was significantly prolonged across the mitral valve (MV) (184 ± 61 vs 116 ± 28 ms, $p < 0.01$) and tricuspid valve (TV) (192 ± 67 vs 126 ± 40 ms, $p < 0.01$) with loss of diastasis time (MV: 22 ± 35 vs 159 ± 92 ms, $p < 0.01$; TV: 13 ± 24 vs 113 ± 70 ms, $p < 0.01$). Also, peak filling rates corrected for end-diastolic volume (PFR_{EDV}) across the MV and TV were significantly reduced (MV: PFR_{EDV} of E wave 2.54 ± 0.56 vs 3.08 ± 0.63, $p = 0.01$; PFR_{EDV} of A wave 1.10 ± 0.26 vs 1.31 ± 0.30, $p = 0.03$; TV: PFR_{EDV} of E wave 1.81 ± 0.44 vs 2.09 ± 0.29, $p = 0.04$; PFR_{EDV} of A wave 1.11 ± 0.22 vs 1.42 ± 0.39, $p < 0.01$). Increased PWV in the aortic arch and descending aorta were associated with increased LV mass ($r = 0.62$, $p < 0.01$, and $r = 0.51$, $p < 0.01$, respectively) and delayed LV relaxation parameters (MV diastasis: $r = 0.50$, $p < 0.01$, and MV E deceleration time: $r = 0.38$, $p = 0.03$, MV diastasis: $r = 0.34$, $p = 0.04$, respectively).
Conclusions: Despite adequately preserved systolic biventricular function, reduced aortic elasticity in paediatric patients after MSS may indicate aortic wall pathology, being associated with ventricular hypertrophy and concomitant delayed ventricular relaxation. Long-term prognosis in MSS survivors may therefore be negatively affected considering the cumulative effects of cardiovascular disease and aging during a lifetime.

6: PREDICTING SUBENDOCARDIAL ISCHAEMIA IN HUMANS

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In 1972 we demonstrated that a ratio DPTI/SPTI < 0.8 predicted relative subendocardial ischaemia in normal dogs. (DPTI – area between aortic and left atrial diastolic pressures; SPTI – area below systolic LV pressure curve.) To correct for anaemia, multiply DPTI by arterial oxygen content (ml/100 ml blood); the critical ratio is –10. This ratio probably applies to normal human hearts, but not to hearts with hypertrophy or dilatation in which SPTI underestimates myocardial oxygen demand (MVO_2) in proportion to excess wall tension or wall thickness. In most abnormal hearts, wall tension remains normal, so that $MVO_2 \propto$ mass or wall thickness, and the critical ratio must be multiplied by relative wall thickness. If wall tension rises because of ventricular dilatation, then the ratio must be multiplied also by the relative wall tension. These variables can be quantitated easily by echocardiography and applied to patients.

7: ABNORMAL VENTRICULAR TORSION: KEY TO DIASTOLIC DYSFUNCTION

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Helical muscles in the LV wall are essential for effective systolic emptying and subsequent refilling. During systole, the base of the heart rotates clockwise and the apex counterclockwise; the angular difference in these rotations (torsion) is measured by magnetic resonance imaging or speckle tracking echocardiography.

After systole, rapid untwisting is needed for ventricular suction and optimal LV filling. Torsion loses efficiency if the LV dilates and

helical fibre angles become less steep, muscle contraction weakens with disease, or torsion is excessively prolonged.When the aortic valve closes, the right side of the ventricular septum (ascending segment of left helix) contracts for 60 to 90 ms after relaxation begins in the muscle on the LV side of the septum and free wall (descending segment of right helix). Ascending segment recoil facilitates ventricular suction and early LV filling. The 60 to 90-ms hiatus is essential for normal function. In disease, excessive prolongation of torsion by persistence of contraction of the LV free-wall spiral and circumferential fibres decreases the hiatus, impairs untwisting, leaving less time for rapid early LV filling, a cardinal sign of diastolic heart failure. For example, during forceful contraction in aortic stenosis, systolic torsion is exaggerated but also prolonged so that it encroaches on the early filling period after aortic valve closure. Suction cannot happen if torsion persists.

Therefore the hallmarks of diastolic dysfunction – elevated LV diastolic pressure despite a normal ejection fraction, and a delayed fall of LV pressure after aortic valve closure – are both manifestations of abnormal systolic muscle function leading to defective untwisting and filling of the LV.

17: LOW RECURRENCE RATE IN TREATING ATRIOVENTRICULAR NODAL RE-ENTRANT TACHYCARDIA WITH TRIPLE FREEZE–THAW CYCLES

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Background: Cryoablation is an alternative to radiofrequency ablation in treating atrioventricular nodal re-entrant tachycardia (AVNRT). However, its long-term effectiveness is in question when compared to radiofrequency ablation. We reviewed the results of cryoablation in children with AVNRT at our institute.
Methods: We performed retrospective single-centre chart review of consecutive patients ≤ 18 years of age with AVNRT who underwent cryoablation between January 2007 and August 2009. During cryoablation, a 6-mm tip cryocatheter was used with temperature set to –80°C. Test lesions were performed at the presumed slow pathway location based on combined anatomical and electrophysiological approaches. If successful, ablation was then continued with triple freeze–thaw cycles (FTC) of 4 min each.
Results: A total of 53 patients (age range: 6.1–18.4 years, mean: 13.6, median: 13.2) underwent slow pathway modification with cryoablation. Acute success was achieved in 51 (96.2%) cases. Transient atrio-ventricular block was seen in 19 (35.9%) cases, but no patient had permanent heart block. Number of FTC was three in 47 (92.2%) patients. Less than three FTC were given in two patients due to transient heart block and four FTC were given in two patients with suspected catheter movement. Procedure duration was 177 ± 56 min; fluoroscopic time was 14 ± 11 min. Mean follow up was 30.7 ± 10 (range 12–52, median 31) months. Recurrence of supraventricular tachycardia was seen in only one (1.96%) patient.
Conclusion: Triple FTC cryoablation lesions resulted in a comparable low recurrence rate to RF ablation in treating AVNRT without increased complications.

25: THE OUTCOME OF THE PATIENTS WITH RIGHT ATRIAL ISOMERISM IS POOR

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Background: Right atrial isomerism is a heterotaxy syndrome with disturbances in the left–right axis development, resulting in complex heart malformations. Previously we have shown that in some families with autosomal recessive inheritance, right atrial isomerism is associated with mutations in the growth/differentiation factor 1 (GDF1).
Methods and Results: The outcome of the patients diagnosed with this syndrome in Helsinki University Central Hospital between January 1976 and December 2010 were reviewed. Among 32 patients (girls 32%) the survival was 22% with a follow-up time of 13.8 years (median, range 0.1–33 years). Extra-cardiac malformations occurred in 91%, mostly aspenia. Cardiac defects included dextrocardia 44%, single ventricle 66% and common atrio-ventricular valve in 100% of cases. Transposition or double-outlet ventricle was seen in 56% and 44%, respectively. Total anomalous pulmonary venous drainage appeared in 75%. Pulmonary outflow obstruction was identified in 91%. Arrhythmias were evident in 28%. Two (6%) had atrio-ventricularblock and pacemaker treatment. Surgery was performed on 78% of patients, seven (22%) were inoperable. Biventricular repair was not possible in any of the patients. In long-term follow up there was no significant difference between the patients with total anomalous pulmonary venous return or normal or partially anomalous venous drainage ($p = 0.5$). Infants requiring their first surgical intervention before four weeks of age had a mortality rate of 60% at five years and those requiring surgery at four weeks or later had a mortality rate of 80% at 15 years.
Conclusions: Right isomerism is one of the most severe forms of cardiac disease. The prognosis remains poor in spite of modern surgical techniques. Therefore, prenatal diagnosis and termination are recommended or prompt treatment after delivery.

33: CARDIOVASCULAR DISORDERS IN ADOLESCENTS WITH CHEST PAIN

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Background: Chest pain is one of the chief complaints that cause parents to bring their children to the paediatrician, paediatric cardiologist, or to the emergency room. Chest pain in children and adolescents can be caused by abnormalities of the heart, but more often are due to musculoskeletal problems, gastrointestinal, lung and idiopathic causes, or are psychogenic.
Aim: To acknowledge the involvement of cardiovascular abnormalities in adolescents with chest pain.
Methods: The subjects of this study were 25 adolescents with chest pain who came to the Cardiac Centre, Hasan Sadikin General Hospital, Bandung during the period January 2008 to January 2011. The presence of established cardiovascular disorders was based on history, physical examination, electrocardiography and echocardiography.
Results: We found 13/25 adolescents with chest pain had cardiovascular abnormalities. Of the 25 teens who came in with chest pain, most showed normal electrocardiographic results, and only nine had dysrhythmias and sinus tachycardia and eight a first-degree AV block. Echocardiography examination showed only four patients with abnormal cardiac anatomy. There was no correlation between nutritional status and adolescents with cardiovascular abnormalities or adolescents with chest pain ($p = 0.206$ and $p = 0.632$, respectively). There was a positive correlation of gender and cardiovascular abnormalities in adolescents with chest pain ($p = 0.007$).
Conclusion: There were cardiovascular abnormalities in adolescents, with symptoms of chest pain in some cases. Female adolescents with chest pain had no correlation with cardiovascular abnormalities.

36: THERAPEUTIC ROLE OF MOBILISED BONE MARROW CELLS IN CHILDREN WITH NON ISCHAEMIC DILATED CARDIOMYOPATHY

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Dilated cardiomyopathy is an important cause of congestive cardiac failure in infants and children. Mobilising haematopoietic progenitor cells is a promising intervention to this deadly disease.
Aim: to evaluate the granulocyte colony-stimulating factor as a therapeutic modality in children with idiopathic dilated cardiomyopathy.
Methods: This case–control prospective study was conducted on 20 children with idiopathic dilated cardiomyopathy following up at the cardiology clinic of the Children's Hospital, Ain Shams University, Cairo, who were compared to 10 age- and gender-matched children as a control group. They were subjected to history taking, clinical examination, echocardiographic study of the left ventricle and cluster of differentiation 34 T-cell assessments in the peripheral blood before and one week after granulocyte colony-stimulating factor intake, for five consecutive days.
Results: A significant improvement in echocardiographic data and increase in the cluster differentiation of 34 T cells was found in patients post granulocyte colony-stimulating factor intake. The percentage change in the cluster of differentiation 34 T cells showed no significant correlation with the percentage change of left ventricular dimension and systolic function.
Conclusion: Administration of granulocyte colony-stimulating factor to children with dilated cardiomyopathy resulted in clinical and echocardiographic improvement that was not correlated to the mobilised cluster of differentiation 34 T cells, implying the involvement of additional mechanisms than simple stem cell mobilisation.

42: THE ROLE OF ECHOCARDIOGRAPHY IN THE ASSESSMENT OF RIGHT VENTRICULAR SYSTOLIC FUNCTION IN PATIENTS WITH TRANSPOSITION OF THE GREAT ARTERIES AND ATRIAL REDIRECTION

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Background: Dysfunction of the systemic right ventricle (RV) in patients with complete transposition of the great arteries (TGA) after atrial redirection by Mustard or Senning procedures is well recognised. However, the complex crescentic shape and the trabeculation of the RV, exaggerated by chronic systemic pressure overload, hampers echocardiographic assessment of the systemic RV. The purpose of this study was to examine feasibility, and variability of echocardiographic parameters for the assessment of systemic RV, and to discuss the role of echocardiography in the assessment of RV systolic function in patients with a systemic RV.
Methods: Multiparametric transthoracic echocardiographic analysis, including global function parameters for RV ejection fraction (RVEF), fractional shortening (RVFS), Tei index and dP/dt_{max}; longitudinal function parameters (tricuspid annular plane systolic excursion (TAPSE), lateral tricuspid annulus TDI peak systolic velocity (S'), tricuspid regurgitation (TR) evaluation and asynchrony assesssement, was performed in 35 patients with TGA after atrial redirection. Functional parameters were compared with MRI. Inter- and intra-observer variability on echographic assessment were analysed from 10 randomly selected cases.
Results: RVEF, RVFS, dP/dt_{max}, TAPSE and 2D strain of the RV were not correlated with RVEF calculated by MRI. Peak systolic velocity (S') was weakly correlated with MRI-RVEF ($r = 0.37$, $p = 0.02$). Inter- and intra-observer variability was high (> 10%) for RVEF, RVFS, and $dP/_{dtmax}$, and low (5%) for TAPSE and S'. Assessment of asynchrony and TR was feasible in all patients.
Conclusion: Conventional echocardiographic parameters for RV function assessment are neither very reliable nor reproducible. However, asynchrony and TR assessment are feasible in routine practice and highly reproducible. Echocardiography does not permit complete assessment of systemic RV but is complementary to MRI and should not be abandoned.

43: MULTIPARAMETRIC ASSESSMENT OF RIGHT VENTRICLE BY ECHOCARDIOGRAPHY IN ADULT PATIENTS WITH REPAIRED TETRALOGY OF FALLOT UNDERGOING PULMONARY VALVE REPLACEMENT: A COMPARATIVE STUDY WITH MRI

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Aim: Evaluation of the right ventricle (RV) using transthoracic echocardiography is challenging in patients with congenital heart diseases affecting the right ventricular outflow tract such as tetralogy of Fallot (TOF). MRI is commonly used to determine the best timing for PV replacement but accessibility remains limited. The objective of this study was to evaluate the feasibility and the accuracy of a multiparametric echographic approach including 2D strain and 3D for RV volumes and function assessment, in comparison with MRI.
Methods and Results: We performed a complete echocardiographic study including 2D parameters [TAPSE, S' TDI, Tei indice, Fractional area change (FAC)], 2D strain and 3D and an unsedated cardiac MRI in 26 consecutive patients with repaired TOF before pulmonary valve replacement and one year after surgery. TAPSE, S' TDI and 2D strain parameters were poorly correlated with MRI regarding RV function assessment. FAC was well correlated with REVF before and after PVR ($r = 0.70$, $p < 0.01$ and $r = 0.68$, $p < 0.01$, respectively). Despite RV volume underestimation, 3D analysis using dedicated software was well correlated with MRI values in both pre- and post-operative assessment ($r = 0.88$, $p < 0.01$ and $r = 0.91$, $p < 0.01$, respectively for RV end-diastolic volume; $r = 0.92$, $p < 0.01$ and $r = 0.95$, $p < 0.01$, respectively for RV end-systolic volume).
Conclusion: A global approach of RV function using 2D (FAC) or (3D) parameters seemed reliable in patients with repaired TOF. The commonly used TAPSE and S' TDI focused on segmental analysis of RV inflow were less sensitive, probably because RV inflow is less affected by RV remodelling related to initial surgical repair.

45: CONGENITAL ANOMALIES OF THE MITRAL VALVE: A CLINICAL AND ECHOCARDIOGRAPHIC STUDY

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Background: Congenital anomalies of the mitral valve (CAMV) comprise a wide range of leaflet and subvalvular apparatus anomalies. Their presentation in adults is not frequent.
Objective: In this work, we assessed the clinical and echocardiographic aspects as well as the treatment of five adult patients with CAMV.
Methods: A complete clinical history was taken for each patient and also an electrocardiogram, chest X-ray and transthoracic echocardiogram.
Results: Two patients were in functional class (FC) I, one in FC II, and two in FC III of the New York Heart Association. Diagnoses were: prolapsed mitral valve with severe mitral failure; parachute mitral valve associated with a subvalvular aortic fibrous ring and patent ductus arteriosus; trileaflet mitral valve with subaortic obstruction; double mitral orifice associated with bicuspid aorta and aortic coarctation; and tunnel-forming mitral valve associated with ostium primum interatrial communication and pulmonary arterial hypertension and pulmonary artery hypertension. One patient was subjected to mitral valve change, one to dilation of the aortic coarctation, and another was put on sitaxentan treatment, with improvement in the FC. The two remaining patients are awaiting surgery.
Conclusions: Review of these cases with CAMV reveals the relevance of the echocardiogram in evaluation of the mitral valve and subvalvular apparatus, because it allows for identification of different

types of malformations and their haemodynamic repercussion, so as to be able to propose precise and timely treatment for these patients.

46: ANATOMICAL–ECHOCARDIOGRAPHIC CORRELATION IN COMPLEX CONGENITAL HEART DISEASE

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Background: Echocardiography is a valuable non-invasive technique to identify the anantomy and function of complex congenital heart disease. Knowledge of the morphological details of each type of complex congenital heart disease is the basis for the correct interpretation of diagnostic images and clinical decisions.
Objective: The objective of this study was to establish the anatomical–echocardiographic correlation in complex congenital heart disease from heart samples with equivalent findings to those of echocardiographic images.
Methods: Thirty hearts and 50 patients with Ebstein's anomaly, 60 hearts and 24 patients with atrio-ventricular septal defect, 15 hearts and 24 patients with absence of right atrio-ventricular connection were studied. The samples correspond to the collection of the Embryology Department.
Results and Conclusions: The anantomical–echocardiographic correlation clearly showed that the anatomical findings of the hearts corresponded with the echocardiographic images of complex congenital heart disease and provided an adequate understanding of the echocardiographic images in terms of an accurate diagnosis, treatment, therapeutic decisions and prognosis.

50: CONTRIBUTION OF MRI AND CT SCAN IN DIAGNOSIS AND MANAGEMENT OF CONGENITAL HEART DISEASE: ABOUT 18 CASES IN THE DEPARTMENT OF CARDIOLOGY, FANN UNVERSITY HOSPITAL, DAKAR, SENEGAL

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Introduction: For diagnosis of congenital heart disease (CHD), cardiovascular magnetic resonance imaging (MRI) and cardiac angioscan (CT) can provide additional useful information for therapeutic decisions. The objective of this study was to identify the indications for MRI and CT and to determine their contribution to diagnosis and management of CHD.
Methods: This was a descriptive study from January 2008 to March 2011 including patients with CHD who underwent trans-thoracic echocardiography (TTE) and in whom MRI or CT was performed.
Results: We identified 10 patients who underwent MRI and eight cases CT scan. Patients' average age was 10 years.The indications for MRI were always pre-operative assessment of: tetralogy of Fallot; coarctation of the aorta; patent ductus arteriosus with a right aortic arch; congenital aneurysm of the left ventricle (LV); and migration of pulmonary banding.

The MRI allowed precise anatomical diagnosis of: hypoplastic left pulmonary artery (LPA); tight coarctation of the aorta; a bi-carotid trunk; rupture of a LV aneurysm within the pericardium; distal migration of a pulmonary banding. CT was indicated for: stenosis at the origin of the pulmonary branches after ligation of the ductus arteriosus; tetralogy of Fallot with pulmonary hypoplasia. A CT scan provided a better description of the aorta, but also the pulmonary arteries and their abnormalities.
Conclusion: MRI and CT scans can be very useful in pre-operative assessment of congenital heart defects. They can be indicated for more accurate anatomical diagnosis prior to surgery and may avoid the use of invasive cardiac catheterisation.

54: INTRA-ATRIAL RHABDOMYOMA IN A NEWBORN PRODUCING PSEUDO PRE-EXCITATION

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Pre-excitation is a common phenomenon in paediatric patients with an accessory pathway. We present a term newborn, male patient with one giant atrial rhabdomyoma and multiple ventricular rhabdomyomas that showed pseudo pre-excitation in the 12-lead surface ECG due to tumour caused atrial depolarisation and repolarisation disturbances. First presentation to our clinic was for further diagnosis and therapy. Except for the cardiac tumours, other physical findings were unremarkable for his age and gender.

Initial ECG showed atrial fibrillation with ventricular rate of 230/min, which was terminated by a single direct current shock of 6 Joule. After cardioversion, ECG showed slow atrial rhythm with frequent premature atrial contractions (PAC) and deformation of the PR interval posing as pseudo-pre-excitation, followed by a normal QRS complex with seemingly abnormal ventricular repolarisation.

We suspected the origin of this pseudo pre-excitation was isolated atrial depolarisation disturbances due to the tumour, which caused heterogenous endocardial activation. Also the seemingly abnormal ventricular repolarisation represented excessive delay in atrial repolarisation superimposed on the ventricular repolarisation. Due to the developement of inflow congestion, the atrial tumour was resected with consequent vast atrial reconstruction using patch plastic, whereas the ventricular tumours were left without manipulation. After surgery pre-excitation and repolarisation abnormalities vanished entirely, with development of an alternans between sinus rhythm and ectopic atrial rhythm.

The QRS complex before and after surgical resection of the rhabdomyoma were identical, underlining the atrial origin of the repolarisation abnormalities before surgery.

57: CONGENITAL JUNCTIONAL ECTOPIC TACHYCARDIA: CLINICAL PRESENTATION, RESPONSE TO TREATMENT AND LONG-TERM EVOLUTION

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Objective: To examine the clinical presentation, response to pharmacological treatment and evolution of congenital JET treated in our hospital.
Methods: Twenty patients were seen in our practice in arrhythmias with congenital JET during the period 2001–2010, with a mean follow up of 5.9 years (± 5.3). All were evaluated with physical evaluation, ECG and Holter periodically. The diagnosis was based on ECG: tachycardia with normal QRS morphology, atrioventricular dissociation and ventricular rate higher than sinus rhythm. Patients were subdivided according to the age of presentation into group I: younger than one year old: 16 patients, with a mean age at diagnosis of 38 days old (1–90 days) and group II: older than one year: four patients with a mean age of 8.7 years (7–11 years).
Results: Group I: median HR at diagnosis was of 182 ± 32 beats/min, and it was constant in all our patients ($p = 0.002$); 87.5% ($p = 0.006$) showed signs of heart failure and ventricular dysfunction, and 81% ($p = 0.02$) needed to be admitted. In all cases HR (mean 122 beats/min, range 90–157) was controlled by 45 days (15–120). Only one patient had a positive family history.

Group II: median HR of the JET patients was 127 ± 38 beats/min and in two patients, it was paroxismic ($p = 0.09$). All of them referred because of palpitations ($p = 0.0002$) and one patient required admission. In all cases the HR (mean 81 beats/min, range 69–98) was controlled by 35 days (5–120). In all cases there was partial control of

the arrhythmia (control of the HR, with intermittent periods of sinus rhythm) and amiodarone was the most effective anti-arrhythmic drug, either alone or in combination with other drugs (p = 0.02). During follow up, all patients persisted with intermittent JET, with HR under control and within normal ranges for age.
Conclusions: Children younger than one year old presented generally with incessant JET, ventricular dysfunction and heart failure requiring admission and a combination of drugs.

In children older than one year, the JET was better tolerated, with less HR and occasionally paroxistic. The treatment with amiodarone either alone or in combination with other drugs was effective in controlling HR, but the arrhythmia remained in all patients during the follow up, without spontaneous cure in serial Holter studies.

59: SCALING CARDIAC STRUCTURES IN CHILDREN

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Background: Body composition influences cardiac dimensions. Body surface area (BSA) or body mass are the most commonly used denominator in paediatric echocardiography to normalise cardiac dimensions for differences in body size. Lean body mass (LBM) has been suggested by many investigators to be the best scaling factor. Data on LBM are, however, not available in clinical practice. We assessed whether cardiac dimensions normalised by LBM compared with cardiac dimensions normalised by various anthropometric methods in younger children.
Methods: This was a cross-sectional study of 139 healthy children (boys $n = 82$ and girls $n = 57$) aged 9.7 ± 0.6 years (range 7.9–10.7). Dual-energy X-ray absorptiometry measured total lean body mass (LBM). Echocardiography was performed with two-dimensional guided M-mode according to current guidelines and left atrium diameter (LA), left ventricular diastolic diameter (LVDD), left ventricular systolic diameter (LVSD), septal wall thickness in diastole (Sep), posterior wall thickness in diastole (Post) were measured and left ventricular mass (LVM) was calculated.
Results: There were significant ($p < 0.05$) univariate correlations between various cardiac structures normalised by LBM and with various anthropometric methods.
Conclusions: This investigation showed close correlations between cardiac dimensions normalised by LBM and by BSA or height$^{2.7}$. These results support the use of them as appropriate denominators in paediatric echocardiography to normalise cardiac dimensions for differences in body size.

63: EVIDENCE FOR AORTOPATHY OF THE NATIVE DESCENDING AORTA IN CHILDREN WITH HYPOPLASTIC LEFT HEART SYNDROME

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Background: Patients with hypoplastic left heart syndrome (HLHS) after Norwood operation show dilatation and reduced distensibility of the reconstructed proximal aorta. Cardiovascular magnetic resonance imaging and angiographic examinations indicate that the native descending aorta is also dilated, but this has not been intensively studied.
Methods: Seventy-nine children with HLHS in Fontan circulation (6.4 ± 3.3 years) and 18 controls (6.8 ± 2.4 years) underwent 3.0-Tesla cardiovascular magnetic resonance imaging. Gradient-echo ciné and phase-contrast imaging were applied to measure cross-sectional areas, distensibility and pulse wave velocity (PWV) of the entire thoracic aorta. Cross-sectional areas were compared with normal values for healthy children.
Results: Patients had significantly elevated cross-sectional areas of the descending aorta at different levels ($p < 0.05$). In 41 (51%) patients they exceeded the 95th percentile. These HLHS patients also showed a higher PWV of the descending thoracic aorta compared to those with normal cross-sectional areas (4.0 ± 1.1 vs 3.4 ± 1.3 m/s, $p < 0.05$). Distensibility of the descending aorta was not significantly different between patients and controls (12.4 ± 6.5 vs 9.9 ± 3.0 10^{-3} $mmHg^{-1}$, $p > 0.05$).
Conclusions: About half of our HLHS patients showed aortic dilatation and increased PWV as a marker of aortopathy of the descending aorta. These findings require further scientific evaluations to investigate potential clinical implications in the Fontan circulation.

64: IMPAIRED AORTIC BIOELASTICITY IS ASSOCIATED WITH DIASTOLIC DYSFUNCTION IN PATIENTS AFTER SUCCESSFUL COARCTATION REPAIR

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Background: Arterial hypertension and accelerated arteriosclerosis are late complications even after successful coarctation (CoA) repair, which may clinically affect left ventricular (LV) function in the long-term follow-up. However, only few data exist on the occurrence of abnormal bioelastic properties of the aortic root and their effect on diastolic LV function in asymptomatic CoA patients at midterm follow up. Therefore, we evaluated this question in children and young adults after successful CoA repair using magnetic resonance imaging (MRI).
Materials and methods: Fifty-two patients (18.9 ± 10.7 years) were examined 14.6 ± 9.2 years after CoA repair using 3.0-Tesla MRI and compared with 39 controls (17.1 ± 7.9 years). The data were used to calculate aortic distensibility and pulse wave velocity (PWV) at different levels. LV ejection fraction (EF), volumes (LVEDV, LVESV) and mass were assessed from short axis views. Axial cine images were used to measure left atrial (LA) volumes and functional parameter ($LAEF_{Passive}$, $LAEF_{Contractile}$, $LAEF_{Reservoir}$) as markers for diastolic function.
Results: In patients aortic distensibility was reduced at all levels of the thoracic aorta (aortic root: 5.5 ± 3.8 vs 7.6 ± 2.9 $10^{-3}mmHg^{-1}$, ascending aorta: 5.8 ± 3.1 vs 9.1 ± 3.6 $10^{-3}mmHg^{-1}$, descending aorta at the isthmus: 5.6 ± 3.0 vs 6.9 ± 2.1 10^{-3} $mmHg^{-1}$, descending aorta at the diaphragm: 6.7 ± 2.8 vs 8.3 ± 3.1 $10^{-3}mmHg^{-1}$; all $p < 0.05$) and PWV in the aortic arch was significantly elevated (4.7 ± 1.8 vs 3.3 ± 0.6 m/s, $p < 0.01$). The minimal LA volume ($LA\text{-}Vol_{min}$) and the LA volume before atrial contraction ($LA\text{-}Vol_{bac}$) were higher in patients ($LA\text{-}Vol_{min}$: 25.3 ± 7.6 vs 20.8 ± 5.4 ml/m^2, $LA\text{-}Vol_{bac}$: 33.2 ± 9.8 vs 26.8 ± 6.2 ml/m^2; all $p < 0.01$). $LAEF_{Passive}$ and $LAEF_{Reservoir}$ were reduced ($LAEF_{Passive}$: 31.7 ± 8.4 vs 38.3 ± 5.7 %; $LAEF_{Reservoir}$: 48.0 ± 7.2 vs 52.0 ± 7.1 %; all $p < 0.01$). $LAEF_{Reservoir}$ and $LAEF_{Passive}$ correlated negatively with aortic arch PWV ($p < 0.05$). LVEF, LVEDV, LVESV, LV mass and blood pressures were not different compared to controls.
Conclusions: Patients after CoA repair show reduced aortic bioelasticity of the entire thoracic aorta and this likely contributes to LV diastolic dysfunction. Therefore this aspect should be focused on during long-term follow-up.

72: ECHOCARDIOGRAPHIC FEATURES OF CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

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Objective: To elucidate the pathological anatomy of congenitally corrected transposition of the great arteries (ccTGA) by echocardiography.
Methods: Twenty-seven consecutive patients (20 males and seven females), aged from one month to 15 years, diagnosed as ccTGA by echocardiography between June 2006 and June 2010 in our paediatric heart centre were included. According to van Praagh sequential segmental analysis, transthoracic echocardiography including M-mode, two-dimensional mode, colour Doppler, pulse-wave Doppler and continous-wave Doppler was performed.
Results: (1) Combination of atrio-ventricular discordance and ventriculo-arterial discordance was diagnosed in all of the 27 cases. Levocardia was detected in 23 cases, including 21 cases of {S, L, L} and 2 cases of {I, D, D}, while dextrocardia with {I, D, D} was seen in the other four cases. (2) Co-existence of atrioventricular valvular abnormalities was seen in three cases, including complete atrio-ventricular septal defect (two cases) and straddling tricuspid valve (one case). In addition, functional tricuspid regurgitation was detected in 19 cases (severe in four, moderate in eight and mild in seven cases), and functional mitral regurgitation in 10 cases (severe in one, moderate in one and mild in eight cases). Compared with functional mitral regurgitation, functional tricuspid regurgitation was more frequent (41.7 vs 79.2%) and worse (moderate to severe regurgitation 20.0 vs 63.2%). (3) Other co-existing abnormalities were seen in 24 cases (88.9%), among which, ventricular septal defect (18 cases, 66.7%) and left ventricular outflow obstruction (18 cases, 66.7%) were the most common structural lesions.
Conclusion: Based on the findings of echocardiography, {S, L, L} was the most frequent pathological anatomy of ccTGA, and ventricular septal defect and left ventricular outflow obstruction were the most common co-existing abnormalities. Additionally, attention should be paid to the functional tricuspid regurgitation in patients with ccTGA.

73: LUNG BIOPSY DIAGNOSIS OF OPERABILITY ASSOCIATED WITH CONGENITAL HEART DISEASE AND PULMONARY HYPERTENSION IN 358 PATIENTS

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Background: I performed a lung biopsy diagnosis in order to determine operability in a total of 358 patients over five years, as requested by the departments of Paediatric Cardiology and Cardiac Surgery from all over Japan, and introduced the prospective data collection.
Method: Decision regarding surgery in simple cardiac anomalies (SCA) or atrioventricular septal defect (AVSD) was based on the index of pulmonary vascular disease. In total anomalous pulmonary venous connection (TAPVC), the operative indication was determined by the degree of hypoplasia of the small pulmonary arteries. Operability of the Fontan procedure was based on the degree of residual medial hypertrophy after pulmonary artery banding.
Results: In SCA, pathological examination revealed radical surgery was indicated in 145 patients but was not indicated in 19. In simple atrial septal defect (ASD), ASD closure was indicated in 21 patients but not in six. Radical surgery was indicated in 50 patients with AVSD but not in 12 –these 12 patients were all associated with Down syndrome. In 26 patients with TAPVC, radical surgery was indicated in all patients. In 68 Fontan candidates, surgery was not indicated in 49. Among seven patients with tetralogy of Fallot, one was not indicated for radical surgery because of occlusive longitudinal smooth muscle cells. Four patients with IPPHN were treated with nitric oxide or bosentan.
Conclusion: Although this was a prospective study in patients with congenital heart disease and pulmonary hypertension, the results have the confidence of cardiac surgeons, paediatric cardiologists and cardiologists who asked for lung biopsy diagnosis of pulmonary vascular disease.

77: CLINICAL IMPACT OF HUMAN RHINOVIRUS IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Community-acquired bronchiolitis is common in infants, and presents with varying clinical severity. Limited information is available regarding the impact of human rhinovirus (HRV) on the management of children with congenital heart disease (CHD). The purpose of this review was to evaluate and describe the clinical impact and management strategies of HRV in children with CHD.
Methods: This was a retrospective review of children with CHD hospitalised and diagnosed with HRV from 1 January to 1 May 2012. Outpatient, pre-operative, operative and postoperative variables were reviewed with an interest in management strategies and clinical impact.
Results: Nine patients tested positive for HRV. Median age was five months (range 1–9); single-ventricle defects (n = 3); trisomy 21 (n = 2); community-acquired HRV (n = 5); nosocomial HRV (n = 4); postoperative patients (n = 3). Surgical management was altered with elective palliative procedure instead of complete repair in two patients. Median length of mechanical ventilation associated with HRV was 11 days (range 0–70); median length of non-invasive ventilation was nine days (range 0–45). Postoperative hospital stay increased significantly in patients with single-ventricle physiology and in patients with trisomy 21 compared to a similar cohort without HRV. Comprehensive stage II (BDG and Norwood type aortic arch reconstruction) median length of stay 14 days without HRV vs 62 days with HRV; atrio-ventricular septal defect repair median length of stay six days without HRV vs 72 days with HRV. There was one death in this patient population that was not attributed to HRV.
Conclusions: The clinical impact of HRV was significant in patients with CHD undergoing cardiac surgery, especially in patients with single-ventricle physiology and patients with trisomy 21. This study emphasises the importance of pre-operative evaluation of respiratory viral bronchiolitis in high-risk children prior to cardiac surgery.

80: CARDIAC ENTRAPMENT AND INJURY DUE TO EPICARDIAL PACEMAKER WIRES

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Placement of epicardial pacemaker wires in neonates and young infants is a common procedure for congenital or surgically related complete heart block. It is estimated that > 40 000 epicardial pacing systems were implanted in patients in North America over the past 20 years. Historically, the excess loops of the wires were placed in the pericardial space to provide sufficient length for anticipated cardiac and thoracic growth. Reports of cardiac compression by the pacing leads are available in seven patients (incidence of < 0.02%).
Case reports: At our centre, we have now seen 3/93 patients (incidence of 3.2%) with severe entrapment of the heart by fibrous fixation of these wires to the atrio-ventricular groove. In one seven-year-old patient, unidentified pacemaker lead compression was felt to have caused sudden unexpected death. Post mortem angiography confirmed compression of the circumflex coronary artery. Two other

patients, aged two and five years, respectively, have shown compression of cardiac structures identified by chest radiography and echocardiography. One underwent cardiac catheterisation without angiographic evidence of coronary compromise. The other underwent lead revision without antecedent angiography. Both of these children had successful lead adjustments with the compressing portions of the leads re-positioned and the redundant portions of the leads anchored to the diaphragm with sutures.
Conclusions: An ongoing review of all 93 patients who have undergone epicardial pacemaker lead placement at our centre is underway. Estimating the degree of cardiac compression by conventional means is limited and requires a high index of suspicion along with investigations including echocardiography, radiography, angiocardiography and CT angiography. No single modality is definitive. We caution all physicians involved with patients who have epicardial pacing systems about this rare but important and potentially lethal complication associated with pacemaker lead placement.

88: TOWARD BETTER VENTRICULAR PACING IN PATIENTS WITH A SYSTEMIC RIGHT VENTRICLE

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Background: Patients treated by atrial redirection surgery (Senning or Mustard procedure) for transposition of the great arteries (TGA), have an important risk for heart failure caused by dysfunction of the systemic RV. Conventional non-systemic ventricular pacing (non-systVP) may even further increase this risk. We investigated whether these patients may benefit from biventricular pacing (BiVP) and/or single-site systemic ventricular pacing (systVP).
Methods and Results: During clinically indicated catheterisation in nine patients with TGA and status post-atrial redirection surgery (SenningMustardTGA), endocardial ventricular stimulation (overdrive DDD-mode, 80–90 beats/min) was applied with temporary pacing leads at the non-systemic and the systemic ventricle. Acute changes in dP/dt_{max} and systolic pressure of the systemic ventricle, as induced by non-systVP, systVP and BiVP compared to reference, were assessed with a pressure wire within the systemic ventricle. Reference was AAI pacing with similar heart rate ($n = 7$), or non-systVP at a lower heart rate than during stimulation at experimental sites (85 vs 90 beats/min; $n = 2$). Systemic dP/dt_{max} and systolic ventricular pressure were significantly higher during systVP (+15.6 and +5.1%, respectively) and BiVP (+14.3 and +4.9%, respectively, compared with non-systVP). In six out of seven patients, systemic dP/dt_{max} was even higher during BiVP and systVP than during AAI pacing.
Conclusions: In a population of patients with SenningMustardTGA, acute haemodynamic effects of endocardial systVP and BiVP were significantly and equally better than those of non-systVP. Single-site systVP and BiVP might also be beneficial in patients with a systemic RV and intrinsic ventricular dyssynchrony.

90: EVALUATION OF LEFT VENTRICULAR SYSTOLIC FUNCTION WITH THE USE OF TISSUE DOPPLER ECHOCARDIOGRAPHY IN CHILDREN WITH PRIMARY ARTERIAL HYPERTENSION

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Background: Arterial hypertension (HA) has become an increasing problem in recent years. The aim of the study was to assess the left ventricular systolic function in children with primary arterial hypertension with the use of tissue Doppler method.
Methods: The analysis included 30 children, 10–18 years old (mean 15.4 ± 2.06) with diagnosed primary arterial hypertension, without overweight or obesity. The control group included 30 children, 10–18 years old (mean 15.43 ± 2.08) with normal values of arterial pressure. All patients underwent: physical examination, manual measurements of arterial pressure, ambulatory blood pressure monitoring, echocardiographic examination with cardiac function evaluation with the use of standard parameters (ejection fraction, shortening fraction, myocardial performance index) and tissue Doppler examination (systolic mitral annular velocity profile and regional function parameters: velocity, strain, strain rate).
Results: Mean values of ejection fraction (EF) as well as shortening fraction (SF) were correct in both groups of patients. Mean values of left ventricular myocardial performance index were significantly higher in children with arterial hypertension (0.46 ± 0.08 vs 0.36 ± 0.03). Significantly lower mean values of systolic mitral annular velocity profile at the intraventricular septum (Sm) and at the lateral level (Sml) were found in children with HA (respectively: 8.7 ± 1.27 and 11.66 ± 2.84 cm/s vs 10.9 ± 2.19 and 16.16 ± 3.30 cm/s). Mean values of regional function parameters (velocity, strain, strain rate) were significantly lower in the hypertensive children group.
Conclusions: In children with primary arterial hypertension, on the basis of evaluation the parameters with the use of tissue Doppler method, subclinical systolic dysfunction of the left ventricle was observed. Left ventricular systolic function, estimated with the use of standard echocardiographic indices was normal, except for myocardial performance index, the value of which was significantly higher compared to the control group.

91: SYNCOPE UNIT IN PAEDIATRIC POPULATION: A SINGLE-CENTRE EXPERIENCE

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Background: Syncopes are frequent in the paediatric population. The majority are benign but, for a minority of children, a cardiac disease is the underlying cause and has to be recognised promptly, as it can be fatal. Syncope units developped in adult populations have demonstrated major improvement in diagnostic processes, reduction in hospitalisation time, with favourable long-term outcomes. We report our experience of syncope management in chidren and adolescents through a dedicated syncope unit.
Methods: In this ongoing study, we prospectively enrolled 45 consecutive patients (13 ± 3 years, 65% male) between January 2011 and June 2012, referred for loss of consciousness (LOC), in a dedicated paediatric syncope unit involving a paediatric cardiologist, nurse, physiotherapist and psychologist. All patients underwent initial evaluation including medical history assessment, physical examination, 12-lead ECG and echocardiography to exclude non-cardiogenic syncopes. If initial assessement was abnormal, they underwent targeted tests that differed according to suspected aetiology. Patients with neurocardiogenic syncope underwent specific physiotherapy training and a consultation with a psychologist.
Results: The most common causes of LOC were neurocardiogenic syncope: 32 patients (71%) and psychogenic LOC: 11 patients (23%). One patient (3%) had a long QT syndrome and received beta-blocker therapy. One patient had typical epileptic seizures and was transferred to a neurology department. Mean hospitalisation time was 0.9 ± 0.5 days. Head-up tilt testing was positive in 62% with neurocardiogenic syncope. Echocardiograms and exercise tests were not contributive for diagnosis. After a mean follow up of 9 ± 4 months, including physiotherapy and/or phychological care, syncope reccurred in five patients (12%).

Conclusion: A syncope unit for the paediatric population with a dedicated team improved diagnostic processes, reduced hospitalisation and decreased syncope recurrence when adapted follow up was proposed.

94: THE IMPACT OF HUMAN DEVELOPMENT INDEX ON SURVIVAL OF CHILDREN AND ADOLESCENTS WITH RHEUMATIC HEART DISEASE HOSPITALISED FOR HEART FAILURE IN BRAZIL

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Background: Rheumatic heart disease (RHD) is still the main aetiology of acquired heart disease in children and adolescents worldwide. The major outcomes are heart failure (HF) and death. Their survival rate after the onset of HF is unknown. In addition, the impact on prognosis of social conditions in an individual community's human development index (HDI) remains unknown.
Purpose: To assess children and adolescents' survival rate with RHD and HF, using probabilistic database linkage methods and to study the influence of geographic regions and HDI on the prognosis.
Methods: We performed probabilistic database linkage from Brazilian hospital admission and death certificates (2001–2007). We used Chi-square, analysis of variance, and Kaplan-Meier method for survival curves, and compared groups by log-rank tests. We estimated hazard ratios (HR) with 95% confidence intervals, followed by Cox proportional hazards model. The significance was achieved at $p < 0.05$.
Results: Seven hundred and eighty patients were hospitalized for the first time for RHD and HF, with 421 (54.0%) deaths in seven7 years of follow up. The median age was 12.8 ± 4.36 (4–19) years old, and 53.8% were boys. The overall survival rates were 61.4% at one, 54.9% at two and 37.2% at six years. There was no difference in survival between genders ($p = 0.107$), but by geographic regions ($p = 0.0001$), with the lowest in north and north-east regions. HDI was lower in the north (0.727 ± 0.727) and north-east (0.682 ± 0.078) compared with the mid-west (0.796 ± 0.049), south-east (0.784 ± 0.051), and south (0.800 ± 0.038) regions ($p < 0.0001$). With Cox analysis, the increase in the overall HDI of 0.01 points reduced the HR for death (0.959; 0.949–0.970; $p < 0.001$), as for income (0.966; 0.958–0.975; $p < 0.001$), longevity (0.961; 0.947–0.974; $p < 0.001$), and educational (0.968; 0.958–0.977; $p < 0.001$) criteria.
Conclusion: Patients with RHD hospitalised for HF are at increased risk of death. An increase of 0.01 point in HDI reduces that risk.

97: ATRIAL SEPTAL DEFECT IN INFANCY: SINGLE-CENTRE EXPERIENCE

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Background: Isolated ASD usually does not present in infancy, and such presentation should prompt careful search for additional lesions. It is also known that the development of pulmonary vascular disease is not uniformly related to age or degree of shunting across the ASD. We present our experience in the management of five patients who were operated in infancy.
Methods: Between January 2009 and June 2012, five patients were operated on with an isolated diagnosis of ASD, mean age was six months, weight was 4.5 kg. Three were operated for failure to thrive, one for repeated respiratory tract infections and one had an emergency operation as the child was ventilator dependent with no other associated anomalies or infection. Two children had cathetersation before surgery to ensure that (1) there were no associated lesions, (2) the LV end-diastolic pressure was not elevated, and (3) the pulmonary vascular resistance was reversible. All had autologous pericardial closure. Two patients had a prolonged postoperative course (15 and 28 days) due to signs and symptoms of RV failure. There was no mortality.
Results: The mean follow-up duration was 18 months (six months to three years). Three children have normalised pulmonary pressures with good catch-up growth. Two children still have elevated PAP with mild FTT, and they are being followed up closely. They are on sildenafil and bosentan.
Conclusion: ASD can present in infancy, and would benefit by surgical closure. A proportion of them will have pulmonary hypertensive issues. Whether a concomitant lung biopsy or a fenestrated closure would prognosticate and hasten their postoperative course needs to be evaluated.

102: NEW INSIGHTS INTO ASPECTS OF PULMONARY DIFFUSING CAPACITY IN FONTAN PATIENTS

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Background: Functional univentricular heart patients were palliated in the morning. Fontan patients lack a sub-pulmonary ventricle and consequently have non-pulsatile pulmonary blood flow. They are known to have reduced pulmonary diffusing capacity, however, the cause of this reduction remains unclear. We aimed to assess possible determinants in the aetiology of reduced diffusing capacity and to assess if it could be increased. Furthermore, we aimed to search for predictors of reduced diffusing capacity.
Methods: Eighty-seven Fontan patients (mean age 16.3 ± 7.6 years) performed sitting pulmonary function tests using the single-breath method and two different mixtures of gas. Furthermore, 72 of the 87 patients performed a maximal cycle ergometer test using the Innocor™ rebreathing technique, thereby estimating cardiac output and stroke volume. Ten Fontan patients and nine matched controls performed a supine pulmonary function test after a supine rest.
Results: In the sitting pulmonary function test, mean *z*-scores were: diffusing capacity corrected for haemaglobin and alveolar volume (DLCOc/VA): –2.38 ± 1.20, alveolar capillary membrane diffusing capacity: –0.14 ± 0.84, and pulmonary capillary blood volume (Vc): –2.04 ± 0.80. In the supine compared to sitting pulmonary function test, DLCOc/VA increased 51.7 ± 11.9% in the Fontan group and 23.3 ± 17.7% in the control group ($p < 0.001$) and Vc increased 48.3 ± 17.4% in the Fontan group and 20.2 ± 13.9% in the control group ($p = 0.001$). In a univariate and multiple linear regression analysis including explanatory variables of surgical data and exercise data at rest and peak exercise, resting cardiac index was an independent predictor of sitting DLCOc/VA (regression coefficient; 0.18, $p < 0.001$).
Conclusions: Pulmonary diffusing capacity was reduced in Fontan patients due to reduced pulmonary capillary blood volume while the function of the alveolar capillary membrane was preserved. The diffusing capacity was highly increasable in Fontan patients compared to controls and resting cardiac index was an independent predictor of diffusing capacity.

103: ARRHYTHMIA AND EXERCISE INTOLERANCE IN FONTAN PATIENTS: CURRENT STATUS AND FUTURE BURDEN

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Background: Long-term survival after the Fontan procedure shows excellent results but is associated with a persistent risk of arrhythmias, exercise intolerance and other severe complications. We aimed to analyse the current burden of clinically relevant arrhythmia and severe exercise intolerance in Danish Fontan patients and, furthermore, to estimate the future burden from analysis of mortality and from the current burden related to age.
Methods: All Danish citizens with Fontan completion from 1981 to 2009 were identified (n = 235). Surviving patients performed exercise test, Holter monitoring, echocardiography, pulmonary function test, and blood sampling and medical history were retrieved from medical records.
Results: Twenty-six (11%) patients died or had heart transplantation (HTx) after a mean (± SD) post-Fontan follow up of 8.3 ± 5.7 years. Excluding peri-operative deaths (n = 8), a linear probability of HTx-free survival was observed and estimated to 99.1% per year. Prevalence of clinically relevant arrhythmia and severe exercise intolerance increased significantly with age and was found in 32 and 85% of patients ≥ 20 years, respectively. Furthermore, resting and maximum cardiac index, resting stroke volume index and pulmonary diffusing capacity decreased significantly with age while diastolic and systolic ventricular function were unchanged. From survival data and logistic regression models the future prevalence of patients, clinically relevant arrhythmia and severe exercise intolerance were estimated, revealing a considerable augmentation. For example, assuming Fontan completion at four years of age and survival of the peri-operative period, the probability of being alive at 40 years old is 72%, the probability of clinically relevant arrhythmia is 45% and the probability of severe exercise intolerance is 88%.
Conclusions: The prevalence of clinically relevant arrhythmia and severe exercise intolerance increased significantly with age in Danish Fontan patients. The future Fontan burden was estimated showing a considerable increase in the prevalence of older patients, clinically relevant arrhythmia, and severe exercise intolerance.

104: PREDICTIVE VALUE OF RESPONSE TO ACUTE VASOREACTIVITY TESTING IN CHILDREN WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION

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Background: Acute testing of vasoreactivity during catheterisation is an important factor in the evaluation of idiopathic pulmonary arterial hypertension (IPAH). However, although the presence of an acute response has important clinical consequences, its definition remains controversial, especially in childhood, where a response is generally defined as decrease of mean pulmonary artery pressure (mPAP) of ≥ 20%. The purpose of our study was to assess the predictive value of vasoreactivity testing and to compare different response criteria.
Methods: Forty-two children with IPAH (mean age 10.1 ± 5.4 years) were included in the study. The assessment of pulmonary vasoreactivity was performed according to the guidelines of the German Association for Paediatric Cardiology. Receiver-operating characteristic curve (ROC) and Kaplan-Meier analysis were used to define the predictive value of three different response criteria: reduction of mPAP ≥ 20%, mPAP ≥ 30%, mPAP ≤ 40 mmHg.
Results: Baseline mPAP was 65.2 ± 18.3 mmHg, mean change in mPAP during vasoreactivity testing was 25.8 ± 19.1%; 24 of the 42 patients showed a more than 20% reduction of the mPAP, 18 of them to an mPAP below 40 mmHg. Mean follow up after catheterisation was 55.3 ± 40.5 months. Freedom from serious cardiovascular events (lung transplantation or death) was 86% after two years, 76% after three years, and 57% after five years. ROC curve revealed a reduction of mPAP ≥ 30% as best cut-off value [area under the curve 0.753 (95% confidence interval 0.603–0.904); p = 0.006]. A predictive value of mPAP ≥ 30% and ≤ 40 mmHg was superior to a reduction of mPAP ≥ 20% [log rank (Mantel Cox) Chi-square 9.98; p = 0.002].
Conclusions: Acute vasoreactivity testing has an impact on outcome in paediatric IPAH patients, however, a stricter definition of the response criteria seems to be more reliable to predict serious cardiovascular events.

109: RHEUMATIC FEVER IN ESTONIAN CHILDREN DURING THE LAST TWO DECADES (1992–2011) BASED ON DATA FROM THE CHILDREN'S CLINIC OF TARTU UNIVERSITY HOSPITAL

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Backround: There are almost no data published on RF among Estonian children. We report our data from the south Estonian region with a population of approximately 100 000.
Methods: We carried out a retrospecive analysis of hospital and outpatient cases.
Results: There were 37 cases of RF (24 girls, 13 boys). The diagnosis of RF was based on modified Jones criteria. Echocardiography was performed on al patients.The first attack of RF occurred at the age of thee to 16 years. Most cases occurred from November to April. Cardiac involvement was present in 26 cases (70%): endocarditis in 24 cases with mitral valve involvement, in eight cases with aortic valve involvement, in five cases with tricuspid and in 11 cases with multiple valve involvement, pericarditis in three cases, and myocarditis in one case. Huntington's chorea was diagnosed in 15 cases (41%); and only chorea in seven (19%). Joint involvement was present in 22 cases (59%). All children were treated with conventional antirheumatic and symptomatic therapy. They received prophylaxis with benzathine benzylpenicillin. After the first attack in children with cardiac involvement, six recovered without seqalae. Others had only mild mitral or aortic valve regurgitation and in two cases, mild dilatation of LV persisted, requiring administration of enalapriI. In five cases there were no futher data after the first attack. A second attack occurred in seven cases: five with cardiac involvement and two with chorea. Even after the second attack only two mild mitral insufficiencies persisted. None of our children needed to be operated during childhood.
Conclusions: RF is a rare disease in Estonia nowadays. In 1995 a small outbreak of 8.5:100 000 was evident. Since 2000 there have been up to two cases diagnosed per year in our hospital. However there may still be some new cases in years to come and it is important to take the possibility of RF into consideration, and implement timeous, appropriate treatment to prevent subsequent complications.

110: NO DIFFERENCE IN CARDIAC PERFORMANCE BETWEEN CRITICAL NEONATAL AND NON-NEONATAL PATIENTS ONE YEAR POST COARCTECTOMY

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Background: Nowadays, repair of a critical neonatal coarctation has low surgical mortality. It is unknown if subsequent postoperative cardiac performance in this subgroup of coarctation patients is different from patients who have undergone correction at an older age. Accordingly, we aimed to characterise in both neonatal and non-neonatal coarctation patients the changes in right (RV) and left ventricular (LV) systolic and diastolic performance within the first year of coarctectomy.
Methods: Children (0–17 years) undergoing an aortic coarctectomy were included and subsequently classified as critical neonatal (prostin dependent < 1 month old) or non-neonatal coarctation patients. To evaluate RV and LV systolic and diastolic performance peak systolic (S') and early diastolic (E') tissue Doppler imaging velocities and E/E' were assessed in the basal LV lateral wall and

RV free wall. Echocardiographic studies were performed pre-operatively, one day postoperatively and one year postoperatively (11.4 ± 8.3 months postoperatively). Additionally, controls age-matched to patients were included for echocardiographic evaluation.
Results: In both neonatal (n = 20) and non-neonatal (n = 19) coarctation patients LV systolic and diastolic performance significantly improved within the first year following repair. One year postoperatively LV systolic performance had normalised, while LV diastolic performance was still impaired compared to controls in both neonatal (LV E' 8.7 ± 3.1 vs 13.3 ± 3.8 cm/s, p = 0.005; LV E/E' 20.0 ± 13.8 vs 9.1 ± 3.4, p < 0.001) and non-neonatal patients (LV E' 12.1 ± 3.5 vs 15.1 ± 2.4 cm/s, p = 0.008; LV E/E' 11.4 ± 4.2 vs 7.4 ± 1.6, p = 0.001). In neonatal coarctation patients, RV systolic and diastolic performance significantly increased within the first year following repair. Subsequently, one year postoperatively no differences were observed in RV systolic or diastolic performance between neonatal or non-neonatal coarctation patients and controls.
Conclusions: One year postoperatively LV diastolic performance was still impaired in both neonatal and non-neonatal coarctation patients, while RV systolic and diastolic performance were normal. Hence, current results reveal that a similar pattern of cardiac dysfunction is present in neonatal compared to non-neonatal coarctation patients one year after coarctectomy.

112: IMPLANTABLE CARDIOVERTER DEFIBRILLATOR THERAPY IN PAEDIATRIC PATIENTS IN THE NETHERLANDS

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Introduction: In paediatric patients, implantable cardioverter defibrillators (ICDs) are increasingly used but unfortunately, the high incidence of shocks limits its use in this particular population. This multicentre retrospective study was undertaken to evaluate the clinical outcome of ICD therapy, the incidence of complications, and the frequency of (in)appropriate shocks.
Methods and Results: We retrospectively reviewed the medical records of all children [n = 95; female: 38; male: 57; median age 13.0 years (0.23–19)] who underwent ICD implantation between 1990 and 2012 in The Netherlands. Median follow-up period was 3.8 years (0.1–16). Six patients died during follow up. ICD was implanted as a primary prevention in 56, and after aborted cardiac arrest in 39 patients. Underlying cardiac disorders were primary electrical heart disease (n = 41), cardiomyopathy (n = 37), congenital heart disease (n = 7) and several other diagnoses in 10 patients. Re-interventions in 56 patients included ICD change because of end-of-life (N = 28) and lead-related problems (n = 17); 31 (33%) patients received an appropriate shock after a median time of 6.6 months and 14 (15%) children experienced an inappropriate shock after a median period of 5.8 months. Patients with primary electrical heart disease were significantly more likely to receive an appropriate shock (p = 0.036). With regard to the occurrence of appropriate shocks, no differences could be found between the primary and secondary prevention group. Patients younger than six years were more at risk of an appropriate shock than older children. Methods to prevent further shocks need to be investigated, as increasing beta-blocker dosage or changing ICD settings were rather unsuccessful, since 80% of the patients received further shocks.
Conclusion: ICD therapy is effective and safe in paediatric patients. Regardless of the type of treatment change, patients who received a shock once remain at risk for further shocks. Lead-related problems are an important reason for re-interventions.

119: VARIANTS OF SCIMITAR SYNDROME

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Background: Scimitar syndrome is a rare condition with anomalous drainage of the right inferior pulmonary vein into the inferior vena cava. This can present either in infancy as a sick child or as a relatively asymptomatic older child. There can be other associated conditions that determine the timing of presentation.
Case report 1: A 45-day-old neonate presented with respiratory distress and echo and X-ray evidence of scimitar syndrome with large patent ductus arteriosus and hypoplastic right lung. The PDA was ligated through a left thoracotomy with partial improvement in the clinical condition. Catheterisation showed multiple collaterals from the descending aorta to the right lung which was embolised. The condition again transiently improved but the child continues to have severe PAH.The child is four months old and is now scheduled for pneumonectomy.
Case report 2: A seven-year-old relatively asymptomatic child presented with symptoms of mild failure to thrive. X-ray and echo raised a suspicion of scimitar syndrome which was confirmed by CT angiography. A dilated inferior pulmonary vein was draining into the inferior vena cava and there were no systemic collaterals. This child underwent successful re-implantation of the inferior pulmonary vein into the left atrium without cardiopulmonary bypass through a right thoracotomy, with an uneventful postoperative course.
Conclusion: Scimitar syndrome can present as extremes, with one child having persistent severe pulmonary hypertension and respiratory issues, presenting as a neonate, and ending up requiring a pneumonectomy, and the other as a relatively asymptomatic child where off-pump re-implantation of the pulmonary vein to the left atrium was possible. The degree of associated pulmonary artery and lung hypoplasia and the presence of systemic collaterals to the lung determined the timing of presentation.

121: ARRHYTHMIA IN PATIENTS WITH CONGENITAL CORRECTED TRANSPOSITION OF THE GREAT ARTERIES AFTER DOUBLE SWITCH OPERATION AND CONVENTIONAL RASTELLI

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Background: Patients with congenitally corrected transposition of the great arteries (ccTGA) often develop arrhythmia.
Methods: We analysed the incidence of arrhythmia in 64 and 29 patients after they underwent double-switch operation (DSO) and conventional Rastelli (CR), respectively.
Results: Fourteen of 64 patients (21%) after DSO and seven of the 29 (24%) after CR showed significant tachycardia. Of these, 12 after DSO and five after CR had atrial tachycardia (AT). The mean duration from the repair to the first AT was 7 ± 5 and 21 ± 7 years after DSO and CR, respectively (p < 0.02). Two cases (3%) after DSO and two cases (7%) after CR developed ventricular tachycardia, one with DSO was treated medically and the other was given an implantable cardioverter defibrillator. Thirteen of the 64 patients (20%) after

DSO and five of the 29 (17%) after CR had bradycardia. Of the 13 cases with bradycardia after DSO [complete atrio-ventricular block (CAVB) in seven, advanced AVB in two, and sick sinus syndrome (SSS) in four], 10 underwent permanent pacemaker implantation (PMI). Among the four cases with SSS, two underwent PMI four to 10 years after DSO. Among the nine cases with AV block, four underwent PMI soon after DSO. CAVB progressed in these four patients and PMI was performed one to 13 years after DSO. One case already had congenital CAVB, and for this case, PMI was performed at the DSO. Five cases had CAVB soon after CR, and all five underwent PMI.

Conclusion: There was no significant difference in the incidence of tachycardia and bradycardia between the DSO and CR groups. Patients with ccTGA were found to have a high incidence of arrhythmia after both DSO and CR. Therefore, close observation of patients with ccTGA is essential.

123: JUST A HEADACHE? BRAIN ABSCESS IN RIGHT-TO-LEFT SHUNT LESIONS

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Background: Although congenital heart disease is a major risk factor to develop brain abscess in the paediatric population, the overall incidence is low, becoming even lower nowadays with early correction of right-to-left shunt lesions.

Case reports: We encountered two adolescents with brain abscess in recent years. The first one was a 14-year-old girl from Morocco with tetralogy of Fallot. The child underwent corrective surgery within a charity programme without any adverse event. Her major complaints were headaches, which, according to her mother, was a known, chronic problem. Just before her return to Morocco she had a seizure that led to the diagnosis of a large temporal abscess that was drained surgically. After four weeks of antibiotics she could return home without any residuals.

The second patient was a 21-year-old man with functional univentricular heart and Eisenmenger syndrome. Due to new onset of headaches he presented at our clinic on the weekend, was admitted and was to be discharged on the following day. As in the young girl, he did not have any fever, neurological deficits or elevated inflammatory markers. Having the previous case in mind, he had an MRI, which showed a brain abscess in the basal ganglia. Due to the location and the reduced general condition, surgery seemed to be too risky. Initial treatment included meropenem and vancomycin. Since repeated MRI after 10 days showed progression, clindamycin was added for a total of six weeks, leading to regression of the abscess.

Conclusion: In immunocompetent patients congenital heart defects with right-to-left shunt remain a major risk factor for brain abscesses. There should be a high index of suspicion in this population. Conservative treatment is an option in high-risk cases or if abscess location is unfavourable.

125: REFERENCE VALUES FOR QT AND QTC MEASUREMENTS AFTER BRISK STANDING IN HEALTHY (7–13 YEARS) PREPUBERAL SCHOOLCHILDREN

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Background*:* Long QT syndrome (LQTS) is an inheritable cardiac disorder that can lead to ventricular arrhythmias associated with sudden death, especially in young and apparently healthy individuals. LQTS is caused by mutations in cardiac ion channels, which decrease cardiac repolarisation reserve and cause QTc prolongation. In borderline cases risk assessment is difficult but important to prevent sudden death. In adults the brisk standing test (BST) has been shown to discriminate between normal individuals and genetically proven LQTS patients. This research was undertaken to establish reference values for prepuberal children (7–13 years of age) for QT duration and QTc prolongation after brisk standing.

Methods: Fifty-seven prepuberal (Tanner score ≤ 3) healthy schoolchildren underwent a BST during continuous ECG registration. The subjects rested in the supine position for two minutes, stood up briskly and stayed in a vertical position for one minute upon which they lay down quickly and rested for two minutes in a supine position. QTc maximal, minimal and mean values were measured during the pre- and post-standing supine resting position, as was QTc at maximal sinus tachycardia and shortest TP interval after brisk standing.

Results: All 57 subjects were included in the measurements. Of these, 29 were boys (age 10.2 ± 1.1) and 28 girls (age 9.9 ± 1.1). Baseline characteristics and response to standing did not differ between boys and girls. Whole group QTc prolongation at maximal tachycardia at BST was significantly longer in children compared to adults (79 ± 26 vs 50 ± 30 ms).

Conclusions: QTc prolongation after brisk standing in children was more pronounced than in adults. Using adult values for children would therefore yield false positive results with the a risk of over-diagnosis and over-treatment. Comparison of these BST reference values with DNA-positive children seven to 13 years old is therefore warranted.

129: MANAGEMENT AND OUTCOME OF ISOLATED PARTIAL ANOMALOUS PULMONARY VENOUS CONNECTION FROM THE LEFT UPPER LOBE

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Background: An anatomical subset of partial anomalous pulmonary venous connection (PAPVC) is venous drainage of the left upper lobe (LUL) of the lung to the innominate vein via a vertical vein. The operative approach, outcomes, and follow-up strategy for this lesion are not well reported. Our objective was to describe the operative approach and outcome for repair of LUL-PAPVC.

Methods: All patients who underwent surgical repair of PAPVC at our institution were retrospectively reviewed. The incidence of LUL-PAPVC, postoperative imaging and complications, need for re-intervention, and survival were analysed.

Results: The incidence of LUL-PAPVC was 12% (n = 12) of all PAPVC patients (n = 102). Median patient age was 14 (6.37) and six patients were female (50%). The operative procedure involved median sternotomy, cardiopulmonary bypass, ligation of the vertical vein at its insertion to the innominate vein, and anastomosis of the vertical vein to the left atrium. There were no peri-operative or late deaths. Postoperative echocardiography was performed in 92% (n = 11) of patients, and the vertical vein to left atrium anastomosis was visualised in 64% (7 of 11). Three patients (25%) had postoperative MRIs performed, all with excellent visualisation of the anastomosis. There were no cases of pulmonary venous obstruction, no imaging evidence of pulmonary hypertension, and no patients required re-intervention. Four patients (33%) had postoperative complications, including two with atrial fibrillation, one with post-pericardiotomy syndrome, and one with left phrenic nerve dysfunction. Median length of hospital stay was 4 (3.5) days.

Conclusions: PAPVC from the LUL to the innominate vein via a vertical vein can be repaired using this technique, with low morbidity and mortality, and without postoperative pulmonary venous obstruction. Pulmonary hypertension was not observed during follow up. Postoperative imaging of LUL-PAPVC repair can be difficult, and MRI should be considered as an alternative to echocardiography in imaging these patients.

131: THE TARANAKI REGIONAL AREA PLAN TO SAVE CHILDRENS' LIVES (THE TARANAKI PROJECT)

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Background: Sudden cardiac death can occur in adults (usually as a result of coronary artery disease) and in younger people under 30 (usually as a result of structural heart disease or channelopathies), ventricular fibrillation or tachycardia being a common pre-mortem arrhythmia that can be treated with direct current (DC) cardioversion. Time to DC shock is proven to be critical to outcome.
Methods: We report on a project in the Taranaki province of New Zealand to place ambulatory automatic defibrillators (AEDSs) throughout the community. Taranaki is on the west coast of North Island of New Zealand, population 109 000, area 7 257 km^2. The population is 84% of European ancestry and 15% Maori. Since sudden unexpected deaths in the young (SUDY) from presumed long QT syndrome, we have initiated a project to place AEDs in every single senior school in the province. The majority of school teachers also have received basic CPR training. All pupils at the schools are aware of the sites of the devices. We have also placed the devices in police vehicles, some volunteer fire brigades, certain shops, several gyms, golf courses, and some Maraes. The aim is to get as good a geographical coverage of the region as possible. An iPhone app shows the site of every device. Devices sourced by us and also those from other sources are on the site. Emergency services are aware of all AED placements and will either tell the emergency caller or ask a nearby business that has one to deliver it.
Results: Several devices have been used appropriately since being placed, but the programme has not been successful yet.
Conclusion: It is feasible to get wide geographical coverage of AEDs and use modern technology to improve useage.

132: TRIALS, TRIBULATIONS, TERRORS, FEARS, FRUSTRATIONS OF SETTING UP A SUDDEN UNEXPECTED DEATH IN THE YOUNG (SUDY) PROJECT IN A SECONDARY HOSPITAL AND ASSESSMENT OF SYNCOPE IN YOUNG PERSONS

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Methods: Taranaki is a province on the west coast of New Zealand's North Island, population 109 000, 85% white, 14% Maori. As part of the National Congenital Inherited Diseases group (CIDG), we have set out to collect and investigate all SUDY/syncope cases, and screen relatives where possible. All sudden, unexpected/unexplained deaths in persons two to 40 years of age are reported to us. Referrals for syncope are also investigated once/if referred. Investigations include autopsy, DNA studies in index cases: clinical examination, ECG, ETT, echocardiography, and where appropriate, cardiac MRI, DNA testing.
Results: Pathologies found to date include long QT syndrome, hypertrophic cardiomyopathy, catecholaminergic polymorphic ventricular tachycardia, arrhythmogenic right ventricular cardiomyopathy, Brugada syndrome, and aortic stenosis. Challenges have included reluctance of family members to be investigated, reluctance to accept therapy where appropriate, inaccurate diagnoses, cultural issues, missed referrals, and lack of notification. Several asymptomatic relatives carrying potentially lethal genetic diseases have been identified; some have accepted therapies as appropriate.
Conclusion: Investigation of relatives of victims of sudden deaths/syncope can prevent further deaths but it is not an easy service to implement, as it is very time and resource consuming.

142: BORN WITH A HUGE MEDIASTINAL MASS, THE STORY OF AN AMAZING SURVIVAL

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Introduction: Cardiac tumours are benign or malignant neoplasms arising primarily in the inner lining, muscle layer, or the surrounding pericardium of the heart. They can be primary or metastatic. Primary cardiac tumours are rare in paediatric practice. Intrapericardial teratomas are significantly rare and generally seen in infants and young children. Interestingly, malignant teratomas are extremely rare with a prevalence of four to six cases in 10 000. It presents mainly in female newborns weighing less than 3 kg. Both forms of teratomas may lead to respiratory distress, pericardial effusion and cardiac compression.
Methods: We present a 16-day-old female infant with a massive intrapericardial mass that caused fatal cardiac compression and respiratory distress. CXR, echocardiography and magnetic resonance imaging (MRI) of the heart are the main non-invasive diagnostic tools. Surgery was lifesaving. The removed tissues were examined pathologically.
Results: Lifesaving surgery was performed without complete resection due to the involvement of the great arteries. Pathological samples revealed a malignant immature teratoma. Chemotherapy was started on day four post extubation. Thr child has remained in stable condition to date.
Conclusion: Immediate post birth detection and surgical management were essentially life saving. Yet the prognosis of this child is considered to be poor in light of the incomplete resection of the tumour and the course of chemotherapy required.

146: PULMONARY ARTERY BANDING FOR LEFT VENTRICULAR DILATED CARDIOMYOPATHY: A NOVEL THERAPEUTIC STRATEGY INSTEAD OF HEART TRANSPLANTATION

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Background: Dilated cardiomyopathy (DCM) in childhood has a considerable morbidity and mortality rate and high incidence of heart transplantation (HTX). Pulmonary artery banding (PAB) has been proposed in patients with ccTGA to retrain the sub-pulmonary left ventricle (LV) and to improve a failing sub-aortic right ventricle. We used PAB in young patients with LV-DCM.
Methods: A retrospective single-centre observational study was performed to evaluate dilatable surgical PAB in infants and young children with LV-DCM.
Results: Since April 2006, 10 infants and two toddlers with LV-DCM referred for HTX received a PAB. Additional, four patients underwent repair of the left-sided total TAPVR, re-implantation of an ALCAPA, mitral valve repair or replacement. There was no hospital mortality, and clinical improvement in all patients. Median age at operation was 126 (1–756) days in the 12 patients without additional operation. The pressure gradient across the PAB increased significantly. The LV ejection fraction increased from a median 15% pre-PAB to 43% at discharge home, and 47%, three to six months later. The median LVEDD and *z*-score decreased from 45 to 30 mm ($p > 0.001$) and +6.1 to +3.2, respectively. Plasma B-type natriuretic peptide levels decreased in comparison to the functional class improvement ($p > 0.001$). Six children were subsequently de-banded by trans-catheter technique and are currently in functional class 1. Two patients, both with non-compaction DCM, deteriorated five and six months after PAB de-banding and finally died, one listed for HTX.
Conclusion: In young children with LV-DCM and preserved right ventricular function, PAB led to an improvement of LV and mitral valve function by ventricular interaction and as yet unknown factors.

147: GLOBAL AND REGIONAL CIRCUMFERENTIAL AND RADIAL MYOCARDIAL DEFORMATION AND TORSION IN ELITE HIGH SCHOOL ATHLETES: PHYSIOLOGICAL IMPACT OF HIGH-INTENSITY ATHLETIC TRAINING ON VENTRICULAR ADAPTATION AND PERFORMANCE

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Background: Cardiac adaptation to high-intensity athletic training is characterised by increases in LV chamber dimension, wall thickness and mass. Previously, we identified regional differences in longitudinal LV myocardial strain in elite high school athletes compared to sedentary controls, most notably with increases in apical relative to basal myocardial deformation. The purpose of this study was to assess the physiological impact of long-term training and the type of sports participation on parameters of LV radial and circumferential deformation as well as LV torsion in this same cohort.
Methods: Standard two-dimensional (2D), spectral Doppler and tissue Doppler were prospectively performed using a GE Vivid 7 system to evaluate LV systolic function in 107 elite high school athletes (68 males) compared to controls. Radial and circumferential strain was performed to evaluate 18 regional (apical, mid and basal short-axis segments) and global LV strain.
Results: Traditional measures of LV systolic and diastolic function as well as tissue Doppler, global circumferential 2D strain, and averaged radial 2D strain were not different between groups. Similar to previously identified changes in longitudinal strain, regional differences in both radial and circumferential strain were identified, with apical deformation increased relative to basal function in athletes compared to controls ($p < 0.001$). LV torsion was decreased in athletes versus controls ($p < 0.01$). Similar changes in regional deformation and torsion were seen in all athletes.
Conclusions: Regionally increased apical deformation compared to basal function was a consistent finding demonstrated in all athletic groups for all components of myocardial deformation. These findings, in addition to an overall decrease in LV myocardial torsion in elite athletes, add novel insights into the physiological basis of augmented regional contractile reserve seen with high-intensity training and may also assist in distinguishing athletic from myopathic increases in LV mass.

149: ASSOCIATED SYMPTOMS OF KAWASAKI DISEASE

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Background: In addition to the diagnostic criteria, a broad range of non-specific clinical features are observed in patients with Kawasaki disease (KD). This can cause it to be confused with other febrile illnesses and delay diagnosis. The purpose of this study was to identify common associated symptoms and their clinical significances in children with KD.
Methods: We retrospectively reviewed the medical records of 121 children who were treated for KD at Presbyterian Medical Centre from January 2005 to June 2010. We collected and analysed all clinical symptoms, laboratory data, and echocardiographic findings.
Results: We identified nine associated symptoms: cough, rhinorrhea, sputum, abdominal pain, vomiting, diarrhoea, arthralgia, headache and seizure. There were only 32 (26.4%) children with no associated symptoms. Patients with abdominal pain and headache were older than those without such symptoms. Compared with complete KD, the incidence of seizure was significantly higher in patients with incomplete KD. Vomiting was strongly associated with the IVIG non-response group.
Conclusions: To decrease the incidence of serious coronary complications due to delayed diagnosis, physicians need to be aware of the manifestation of KD and the possibility of associated symptoms.

151: REDUCED EXERCISE CAPACITY IN PATIENTS OPERATED FOR VENTRICULAR SEPTAL DEFECT

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Background: Ventricular septal defects (VSDs) are generally closed very simply, and postsurgery, the patients are considered healthy with normal physical capacity. However, if this is true, is has not been verified, and this therefore was the aim of this study.
Methods: We tested cardiopulmonary exercise capacity in 21 patients and 11 healthy control subjects on an ergometer cycle. Pulmonary ventilation and gas exchange were simultaneously measured breath by breath with Jaeger MasterScreen CPX®. Each test was performed as a maximal incremental test. The graded cycling test protocol was chosen individually to ensure test time to be approximately the same for all participants. During the test session, respiratory gas exchange was measured along with heart rate, blood pressure, and ECG. Endpoints were: maximal oxygen uptake, maximal workload, and anaerobic ventilatory threshold. For the last-mentioned, both absolute and relative thresholds were measured using V slope. Before each test a spirometry was made to measure FVC, FEV_1 and PEF.
Preliminary results: VSD patients had a median age at surgery of 2.6 years (1.5–4.1 years) and 21.1 years (19.8–23.2 years) at the time of examination. Compared to controls they had a markedly impaired maximal oxygen uptake, median 38.0 ml O_2 $kg^{-1}min^{-1}$ (31.6–40.8 ml O_2 $kg^{-1}min^{-1}$) vs 45.8 ml O_2 $kg^{-1}min^{-1}$ (41.1–49.9 ml O_2 $kg^{-1}min^{-1}$) in control subjects, $p < 0.01$. Furthermore, absolute and relative anaerobic thresholds were reduced in VSD patients, median 22.1 ml O_2 $kg^{-1}min^{-1}$ (17.5–25.9 ml O_2 $kg^{-1}min^{-1}$) and 60.0% (54.0–72.7%), respectively, vs 33.5 ml O_2 $kg^{-1}min^{-1}$ (25.1–41.6 ml O_2 $kg^{-1}min^{-1}$) and 76.1% (64.0–86.4%), respectively, $p < 0.05$ for both parameters. Lastly, maximal workload were significantly reduced, median 3.2 W kg^{-1} (2.7–3.6 W kg^{-1}) vs 4.1 W kg^{-1} (3.2–4.3 W kg^{-1}) in control subjects, $p < 0.01$.
Conclusion: Patients with a surgically closed VSD had a markedly reduced cardiopulmonary exercise capacity compared to healthy controls; findings include effort-independent measurements.

157: SHADES OF BRADYCARDIA: AN EVALUATION OF THE FOETAL HEART RATE ACROSS GESTATIONAL AGE IN LONG QT SYNDROME

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Introduction: Long QT syndrome (LQTS) is rarely recognized *in utero* although sinus bradycardia is a common manifestation. Normally, foetal heart rate (FHR) decreases as gestation progresses. The effects of gestational age on FHR in foetal LQTS have not been examined. Little is known of gene-specific associations with FHR in LQTS. The purpose of this study was to evaluate FHR of LQTS subjects across gestation to determine a FHR predictor of LQTS and determine if foetal bradycardia is gene-specific.
Methods: We reviewed FHR throughout gestation from obstetrical records of 42 paediatric patients with LQTS at three paediatric cardiac centres from 2002 to 2011. For comparison, we determined 3rd to 95th percentile range of FHR at 10 to 40 weeks from 547 normal foetuses. Bradycardia was defined as FHR ≤ 3rd percentile for gestational age or ≤ 110 bpm (perinatal definition). We assessed best FHR predictor of LQTS. LQTS was confirmed by postnatal ECG changes (QTc, 2nd-degree AVB, torsades de pointes) and in most confirmed by genetic testing.
Results: We ascertained 309 FHR from 42 LQTS patients: 96% had a mutation confirmed in a known LQTS gene: KCNQ1 (23), KCNH2 (four), SCN5A (six), KCNE1 (two) and multiple (one). Three had

uncharacterised (*de novo*) mutations and three were not tested. Bradycardia incidence depended on the definition: 15% of FHR readings were ≤ 110 bpm and 66% ≤ 3rd percentile. Confirmation of foetal LQTS resulted in diagnosing unsuspected LQTS in three families.
Conclusions: FHR varies widely in LQTS. A FHR ≤ 3rd percentile for gestational age increases the sensitivity to ascertain foetal LQTS compared to the FHR ≤ 110 bpm definition. LQTS should be suspected if FHR ≤ 3rd percentile for gestational age even in the absence of arrhythmias. *De novo* mutations result in the most severe foetal bradycardia. These findings may improve detection of foetal and neonatal LQTS.

160: PREDICTORS OF DISEASE PROGRESSION IN PAEDIATRIC DILATED CARDIOMYOPATHY

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Background: Dilated cardiomyopathy (DCM) is the leading indication for heart transplantation in children. Identifying patients at risk for disease progression/transplantation remains elusive.
Methods: The Paediatric Heart Network Ventricular Volume Variability Study evaluated chronic DCM patients with serial prospective echocardiographic and clinical data collection over an 18-month follow up. Inclusion criteria were age < 22 years and DCM disease duration > 2 months with exclusion of those needing IV inotropic or mechanical support, and those listed as status 1A/1B for transplant. Disease progression was defined as an increase in transplant listing status, hospitalisation for heart failure, IV inotropes, mechanical support, or death during follow up. Predictors of disease progression were identified using logistic regression and classification and regression tree (CART) analysis.
Results: Of the 127 patients, 28 (22%) met the criteria for disease progression during the 18-month follow-up period. Multivariable analysis (*c*-statistic = 0.90) identified older age at diagnosis (OR = 1.16 per year, $p = 0.003$), larger left ventricular (LV) end-diastolic M-mode dimension *z*-score (LVEDDz) (OR 1.77, $p < 0.001$) and lower septal peak systolic tissue Doppler velocity *z*-score (OR = 0.68, $p = 0.04$) as independent predictors of disease progression. CART analysis risk-stratified patients for significant disease progression with 89% sensitivity and 94% specificity based on LVEDD z ≥ 7.7, LV ejection fraction < 38.2%, LV inflow propagation velocity (color M-mode) *z*-score < –0.28, and age at diagnosis ≥ 8.5 months.
Conclusion: In paediatric patients with DCM, diagnosis after late infancy and echocardiographic parameters of LV size, systolic and diastolic function were independently associated with disease progression, and may be used to reliably risk stratify DCM patients.

161: PHYSICAL ACTIVITY RECOMMENDATIONS IN CONGENITAL AND ELECTROPHYSIOLOGICAL HEART DISEASE: A SURVEY OF CANADIAN HEALTHCARE PROVIDERS

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Background: Determining safe levels of physical activity for children and adolescents with congenital and arrhythmic heart disease is a challenging clinical problem. The body of evidence for making these recommendations is limited and based on the perceived risks of sudden cardiac death (SCD) with activity. The Bethesda guidelines were designed to establish consensus guidelines for eligibility and disqualification from competitive sports in athletes with cardiovascular abnormalities. However, literature on non-competitive physical activity is not available.
Methods: A survey was designed to determine practice patterns for patients with structural and arrhythmic heart disease**.** Between July 2011 and December 2011, approximately 350 healthcare providers working with this group of patients were recruited by e-mail or while attending professional meetings. We received 81 responses, primarily from paediatric cardiologists (70%).
Results: Our findings indicate that the majority of Canadian cardiac care providers surveyed are only partially implementing current recommendations. Areas of disagreement included physical activity recommendations for hypertrophic cardiomyopathy, long QT syndrome, catecholaminergic polymorphic ventricular tachycardia and heart transplantation, among others. The development of consensus guidelines for activity recommendations was supported by 96% of respondents.
Conclusions: The heterogeneity of our responses may be attributed to conflicting information in the literature, an entrenched tendency towards bed rest in the cardiology community and a lack of awareness by cardiac care providers regarding the role of physical activity in structural and arrhythmic heart disease. Balancing the risk of SCD with the long-term morbidity and mortality associated with cardiovascular disease needs to be strongly considered.

170: LONG-TERM RESULTS OF PERCUTANEOUS BALLOON VALVULOPLASTY FOR CRITICAL NEONATAL AORTIC STENOSIS

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Background: Since the late eighties, balloon valvuloplasty has been the first-choice treatment of neonatal aortic stenosis. The aim of the study was to offer long-term results of the balloon valvuloplasty to compare with new surgical methods such as valve shaving and plasty that provide promising short-term results in selected newborns.
Methods: This was a retrospective follow-up study of all the 126 patients initially treated with the balloon valvuloplasty and regularly followed up in a single high-volume tertiary referral centre. Only 94 of them (74.6%) fulfilled the revised criteria for biventricular repair published by Colan *et al*. The age at valvuloplasty was 0 to 28 days (median 2 days) and the follow-up period was up to 22.4 years (median 4.8, in 111 early survivors 6.9 years).
Results: Thirty patients (23.8%) died, 28 (22.2%) developed re-stenosis, 16 (12.7%) severe aortic regurgitation and 20 (15.9%) both re-stenosis and regurgitation. Surgery was needed in 41 (32.5%) patients. Mean (SEM) actuarial probabilities 20 years after the procedure were as follows: freedom from re-stenosis 0.43 (0.11), freedom from severe aortic regurgitation 0.48 (0.07), survival 0.70 (0.07), and surgery-free survival 0.24 (0.06). Twenty-year survival probability in patients who fulfilled the Colan criteria was 0.89 (0.03) and in those who did not 0.32 (0.10), $p < 0.001$. Risk factors for the death identified by Cox analysis were small body surface area, small aortic annulus, left ventricular failure/duct-dependent systemic circulation, severe endocardial fibroelastosis, mitral stenosis, and severe pulmonary hypertension.
Conclusions: Balloon aortic valvuloplasty is a good life-saving palliation for critical neonatal aortic stenosis. Less than a quarter of patients treated will survive childhood without surgery. Small newborns and those with small aortic annuli and concomitant left heart diseases are at greater risk of death.

181: EFFECT OF E-HEALTH INDIVIDUALLY TAILORED ENCOURAGEMENTS TO PHYSICAL EXERCISE ON AEROBIC FITNESS AMONG ADOLESCENTS WITH CONGENITAL HEART DISEASE: A RANDOMISED CLINICAL TRIAL. DESIGN AND RATIONALE FOR THE PREVAIL STUDY

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Background: Intensive exercise may be an important part of rehabilitation in patients with congenital heart disease (CHD). However, performing regular physical exercise is challenging for many adolescent patients. Consequently, effective exercise encouragement may be needed. Little is known on the effect of e-Health encouragements on physical fitness, physical activity and health-related quality of life (HRQoL) in adolescents. This trial is a nationwide interactive e-Health rehabilitation study lasting one year, centred on interactive use of mobile phone and Internet technology.
Hypothesis: We hypothesised that e-Health encouragements and interactive monitoring of intensive exercise over one year could improve physical fitness, physical activity and HRQoL.
Methods: Two hundred and sixteen adolescents (13–16 years) with surgically corrected complex CHD, but without significant haemodynamic residual defects and no restrictions to participate in physical activity are in the process of being enrolled by invitation after informed consent. Physical fitness is measured as the maximal oxygen uptake (VO_2 peak) at baseline and after 12 months by an assessor blinded to the randomisation group. After baseline testing, the patients are 1:1 randomised to an intervention group or a control group. Individually fully automated tailored e-Health encouragements: SMS, Internet and mobile applications aimed at increasing physical activity are delivered to the participants in the intervention group once a week. Bandura's social cognitive theory inspires the behavioural theoretical background.
Results: The e-Health intervention and the Godfrey cycle ergometer protocol has been feasibility tested and seems applicable to adolescents with CHD. The trial is expected to contribute to new knowledge regarding how physical activity in adolescents with CHD can be increased and possibly co-morbidity reduced.

182: PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM: SINGLE INSTITUTION EXPERIENCE

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Objective: This study analyses outcomes and predictive factors in cases of percutaneous and surgical treatment of pulmonary atresia with intact ventricular septum (PA-IVS).
Methods and Results: Between September 2003 and March 2012, 53 consecutive newborns with PA and IVS were reviewed. Ten patients had a concomitant pathology, Ebstein's anomaly (n = 6), severe dysplasia of TV and RV (n = 4) including Uhl's syndrome (n = 2). All patients were divided into three groups: no to mild RH hypoplasia (n = 35), moderate hypoplasia (n = 10), and severe in eight patients. Thirty-two neonates with PA-IVS underwent attempts with percutaneous balloon pulmonary valvoplasty as primary procedure, which was successful in 28 patients. Mortality rate was 10.7%, there was no procedure-related death or major complications. Freedom from re-intervention was 32%. Nineteen patients required 23 additional surgical procedures after BPVP, 10 of them in the 10 days after BPVP. Primary surgical procedure was performed in 21 patients (including three patients after unsuccessful BPVP). There were 11 one-stage repair with mortality rate 55% (n = 6) and BTSh in 10 with mortality rate 20% (n = 2). Mortality rate after surgical procedures was associated with presence of Ebstein's anomaly (four of six), severe dysplasia of TV (two of two) ($p < 0.001$). Five patient was refused treatment because of RVDCC (n = 3) and severe dysplasia of the right ventricle (n = 2). Of 44 patients who survived, 23 already had biventricular circulation and four are planned for biventricular repair, four are planned for 1.5-ventricle circulation, and eight are awaiting staged univentricular correction.
Conclusions: Percutaneous balloon valvotomy is an effective treatment strategy for cases of PA-IVS with well-formed right ventricle and absence of right ventricular dependent coronary circulation. Severe dysplasia of RH structures and Ebstein's anomaly were associated with high mortality.

188: QUANTIFICATION OF QUALITY IN CONGENITAL HEART SURGERY

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Background: Estimation of quality of congenital heart surgery should reflect procedural complexity, achieved survival and observed postoperative unfavourable events. It should be quantified to facilitate bench-marking.
Methods: Procedural complexity was determined by Aristotle's basic complexity score. Hospital and 30-days survival was applied. Surgical performance was estimated as the product of complexity score and achieved survival. Observed morbidity was calculated as score according to the methodology of Sata and co-authors. The following formula was used: Quality in congenital heart surgery = surgical performance – morbidity score. Means are given with ± standard deviation.Year 2011 results were analysed.
Results: Primary procedures (542), including 46 (8.5%) without cardiopulmonary bypass, were evaluated. Total cavopulmonary connection with external fenestrated conduit constituted the most frequent operation (n = 34). Mean Aristotle basic score amounted to 7.78 ± 2.65 points. Survival reached 98.15% (532/542). Surgical basic performance attained was 7.64 ± 2.60 points. No adverse event occurred following 183 (33.8%) procedures. Calculated morbidity score averaged 2.26 ± 1.80 points for the whole cohort. Consequently, quality of congenital heart surgery for year 2011 was quantified at 7.64 – 2.26 = 5.38 points.
Conclusions: Such quality quantification adequately reflects complexity of performed procedures and related observed mortality and morbidity. Once accepted, it could serve as a reliable tool for monitoring and comparing the achievement of various programmes of congenital heart surgery.

200: FACTORS AFFECTING GROWTH FROM BIRTH TO NORWOOD DISCHARGE: RESULTS FROM THE SINGLE VENTRICLE RECONSTRUCTION TRIAL

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Background: Growth failure after the Norwood procedure is a potentially modifiable risk factor for medical morbidity and neurodevelopmental outcome. We sought to characterise growth patterns and to determine risk factors for poor growth between birth and Norwood discharge.
Methods: We performed a secondary analysis of growth using the Single Ventricle Reconstruction Trial (SVR) database, in which subjects undergoing a Norwood procedure were randomised to modified Blalock-Taussig vs right ventricular-to-pulmonary artery shunt.

Infants < 37 weeks' gestation and those who underwent stage II without being discharged after the Norwood procedure were excluded. The primary outcome was change in WHO weight-for-age *z*-score (WAZ) between birth and discharge. Growth faltering was defined as a drop of ≥ 0.5 in WAZ and failure to thrive (FTT) as a WAZ < –2. The effect of predictor variables on change in WAZ was analysed using multivariable linear regression with bootstrapping.
Results: The change in WAZ from birth to discharge (at 1.1 ± 1.0 months) for 424 infants in the analytic cohort was –1.40 ± 0.80 and was similar in the two shunt groups ($p = 0.32$). Growth faltering was present in 91%, with FTT in 37% at discharge. Independent risk factors associated with a decline in WAZ ($R^2 = 0.33$) were male gender ($P < 0.001$), higher birth weight ($p < 0.001$), pre-Norwood enteral feeds ($p = 0.01$), longer cardiopulmonary support time ($p = 0.03$), and more ventilator ($p = 0.04$), and hospitalisation days ($p < 0.001$).
Conclusions: Nearly all infants discharged after the Norwood procedure in the SVR trial had growth faltering and over a third had FTT, irrespective of shunt type. Males were at higher risk for a decline in WAZ and pre-Norwood enteral feeds did not prevent growth failure. Suboptimal growth was associated with longer intra-operative support and medical morbidity. Strategies to improve growth during the Norwood hospitalisation warrant further attention.

203: OUTCOME OF CONGENITAL HEART DEFECTS ASSOCIATED WITH 22Q11 DELETION

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Background: 22q11.2 deletion is the most common chromosomal microdeletion syndrome. It has a strong association with conotruncal heart defects and may exert negative influence on the clinical outcomes. We aimed to define the prevalence of congenital heart defects in patients with 22q11.2 deletion, timing of detection and the outcomes.
Methods: We reviewed the patients with 22q11.2 deletion who either presented to the University Hospital of Wales with congenital heart disease or were reported to the Congenital Anomaly Register and Information Service for Wales (CARIS) between 1990 and 2011.
Results: One hundred and two cases of 22q11.2 deletion were reported during the study period, suggesting a prevalence of 1.2 per 10 000 total births (one in 8 335 total births); 95 cases had both 22q11.2 deletion and at least one associated congenital heart defect (CHD) (93%). Interrupted aortic arch, ventricular septal defect, tetralogy of Fallot and truncus arteriosus, and pulmonary atresia were the most common defects. Seven cases exhibited a normal heart. In 18 cases, 22q11.2 deletion was detected in foetal life; 13 babies were delivered alive (72%), pregnancies were terminated in four, and still birth occurred in one. In 84 patients 22q11.2 deletions were confirmed postnatally; 88 of 102 patients remained alive during the mean follow up of 16.2 ± 11.4 years (range 1–52 years), but 16 patients died (19%) of whom eight died after birth, seven in the first year of life, and one at age 3.8 years.
Conclusions: Ninety-three per cent of patients with 22q11.2 deletion exhibited CHD. In spite of the antenatal detection rate being low, survival rates were similar for all cases irrespective of the presence of CHD. Conotruncal abnormalities were most prevalent heart defects. Most deaths occurred within the first year of life but beyond infancy survival was favourable.

204: MISDIAGNOSIS OF BLAND–WHITE–GARLAND SYNDROME: REPORT OF TWO CASES WITH DIFFERENT PRESENTATIONS

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Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or Bland–White–Garland syndrome is usually an isolated cardiac anomaly but, in rare incidences, has been described with patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, and aorta. This syndrome may cause sudden death in infants and young people but in this case report we present two different types of presentation.
Case report: The first case was a three-year-old girl diagnosed with dilated cardiomyopathy since infancy. Her electrocardiography showed prominent Q waves in the lateral leads. A dilated right coronary artery was shown by echocardiography. The second case was a girl with prolapsed mitral valve and chest pain but similar to the first case as she had prominent Q waves in the lateral leads on electrocardiography and a dilated right coronary artery but without heart failure.
Conclusion: ALCAPA in children may present with ambiguous presentations from dilated cardiomyopathy and full-blown heart failure to an atypical chest pain attributed to prolapsed mitral valve.

205: LIVER STIFFNESS: A NEW, RAPID AND NON-INVASIVE METHOD OF CENTRAL VENOUS PRESSURE EVALUATION IN PATIENTS WITH CONGENITAL HEART DISEASE

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Background: Transient elastography is a rapid, non-invasive and reproducible approach to assess liver fibrosis by measuring liver stiffness (LS). However, because the liver is enveloped by a capsule, any variation in parenchymal fluid content could theoretically affect LS. LS has been correlated to central venous pressure (CVP) in an animal model. We aimed to determine the correlation between LS and CVP in children and adults with congenital heart disease.
Methods: In this ongoing prospective study, all patients referred for right heart catheterisation were included. Measurements of mean right atrial pressure were obtained under general anesthesia (Fi02 = 21%) using an Optitorque 5 French catheter. The patients underwent 10 LS measurements (median value taken as representive) by transient elastography (Fibroscan, Echosens, France) within the 24 hours before catheterisation. The results of LS are expressed in kilopascals (kPa).
Results: Fourteen children (mean age = 9 ± 6 years, 64% male) and 14 adults (mean age = 34 ± 17 years, 66% male) have been included so far. Catheterisation indications were pulmonary angioplasty ($n = 5$), Melody valve implantation ($n = 2$), fenestration occlusion after a Fontan procedure ($n = 1$), aortic coarctation stenting ($n = 1$), atrial septal defect closure ($n = 4$) and pre-operative assessement of a complex congenital heart defect ($n = 16$). Mean right atrial pressure was 8.2 ± 3.3 mmHg and mean LS was 8.1 ± 4.4 kPa. Correlation between LS and mean right atrial pressure was excellent for these first 28 patients ($r = 0.86$, $p < 0.001$).
Conclusion: Liver stiffness is a new, rapid and reliable method to evaluate CVP in patients with congenital heart disease. This non-invasive parameter could potentially be useful for patients in whom CVP play a key role, especially in patients with a Fontan circulation.

206: IS QRS AXIS PATTERN ASSOCIATED WITH THE TYPE OF SURGICAL REPAIR IN ADULTS WITH OPERATED TETRALOGY OF FALLOT?

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Background: Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital heart disease. Until the development of the transatrial–transpulmonary approach, surgical repair was achieved through a right ventriculotomy causing right bundle branch block (RBBB). We aimed to study the QRS axis pattern in adults with repaired ToF and to correlate it to the type of surgery.
Methods: Adults with repaired ToF referred at our institution for ECG, echocardiography and cardiovascular magnetic resonance were included except paced patients. Surgical history was obtained from hospital records. Electrocardiographic measures (maximum PR, QRS and QT duration, QRS axis) were analysed manually from standard 12-lead electrocardiograms.
Results: Twenty-nine patients were included (72% male, 33 ± 13 years). All patients but one had a transventricular repair (37% had a transannular patch, 60% had a transverse or longitudinal ventriculotomy without transannular patch). Mean CMR right and left ventricle indexed end-diastolic volumes were, respectively, 150 ± 29 and 71 ± 17 ml/m^2. Mean QRS duration was 156 ± 16 ms with a RBBB pattern for all patients. There was no significant difference concerning BMI, CMR measures or QRS duration between different sub-groups. All patients from the ventriculotomy group had a normal or right ECG axis pattern (axis between 34 and 160°). Nine patients (90%) from the transannular group had a left ECG axis pattern (axis between –10 and –76°).
Conclusion: QRS axis pattern in adults with repaired ToF was correlated with the type of surgery.

208: THE INCIDENCE AND NATURAL HISTORY OF INNOCENT HEART MURMUR IN NEWBORN BABIES

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Introduction: This study aimed to determine the incidence, and define the origin and natural history of the innocent heart murmur in newborns.
Methods: In a 21-months prospective study, 2 849 newborn babies underwent routine examination by paediatricians. Infants with a murmur were re-examined by paediatric cardiologists. Term babies with a clinical diagnosis of an innocent heart murmur were studied. Each baby had a complete echocardiographic study and was followed up at two and six months until the murmur had disappeared or the heart was totally normal.
Results: Clinically suspected innocent heart murmur was found in 30 cases. The incidence of innocent murmur of term babies was 9:1 000 live births. Normal echocardiogram was found in 10 cases (33%), peripheral pulmonary branch stenosis (PPS) in six (20%), small patent ductus arteriosus (PDA) in five (17%), small ventricular septal defect (VSD) in three (10%), mild pulmonary valve stenosis in one (3%) and isolated mild tricuspid regurgitation in five cases (17%). Only VSD and pulmonary valve stenosis were considered to be pathologic. Follow up was performed at the age of two months: the heart murmur had disappeared in 23 cases (82%), PDA and mild pulmonary stenosis had resolved in all patients but PPS was still present in one of six cases. At six months, the murmur had disappeared in 26 cases (93%), two of the three with asymptomatic small VSD still had murmur and the defects were still patent.
Conclusion: An innocent heart murmur in a term baby is often related to non-clinical significant conditions. The suspected innocent heart murmur diagnosed at birth had resolved in most of the babies at six months.

212: SEVERITY AND PROGNOSTIC INDICES IN CHILDHOOD CARDIAC FAILURE

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Background: Childhood cardiac failure remains a major cause of morbidity and mortality in the developing world. The diagnosis and management outlook of these children in resource-poor countries remains poor when compared to their developed counterparts. There is still a paucity of data on the severity and determinants of outcome in childhood cardiac failure in Nigeria.
Objectives: The aim of the study was to evaluate the severity of congestive cardiac failure (CCF) and to highlight the factors that affect outcome in children presenting at the Lagos University Teaching Hospital (LUTH), Lagos, Nigeria.
Methods: Two hundred and twenty consecutive cases of CCF admitted into paediatric wards and the children's emergency rooms of LUTH with a diagnosis of heart failure over a one-year period were studied prospectively. Diagnosis of heart failure was based on the presence of at least three or four cardiac signs of heart failure. The severity of CCF was determined using heart failure severity index scores for children, proposed by Omokhodion *et al*., which was validated using the Ross heart failure scores for infant and children. All cases were followed up daily until a definite outcome was determined.
Results: Predominant underlying causes of CCF were severe anaemia (39.4%), respiratory tract infections (26.1%), intrinsic heart disease (22.5%), and others (12.0%); 23.8, 30.3 and 45.9% of the patients presented in mild, moderate and severe heart failure, respectively, and the difference in distribution was statistically significant ($p < 0.01$). The factors that affected the severity of heart failure were low packed-cell volume on admission ($p = 0.04$), low socio-economic class ($p = 0.03$) and the cause of the heart failure ($p = 0.02$). The case fatality rate was 15.1% among the study population. Poor prognostic indices identified were renal disease ($p = 0.03$) and acquired heart disease ($p = 0.004$) as the cause of the heart failure, presence of severe heart failure on admission ($p < 0.001$), lower socio-economic status ($p = 0.003$) and re-admission for heart failure within the study period ($p < 0.01$).
Conclusion: Heart failure in Nigerian children is associated with an unacceptably high mortality rate. Identification of high risk factors in children with heart failure and routine use of heart failure severity index to grade heart failure severity may aid in early and effective interventions.

222: FAMILIAL RECURRENCE OF CONGENITAL HEART DISEASE CAUSED BY MUTATION IN NKX2-5

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Background: Familial recurrence presents in 3 to 5% of non-syndromic congenital heart disease (CHD). The transcription factor *NKX2-5* is known to cause a variety of CHDs, but most previous studies have focused on a single specific diagnosis. The aim of this project was to identify a large number of families with recurrent CHD and to screen them for mutations in *NKX2-5* regardless of the CHD diagnosis.
Methods: We included 46 families with 136 affected individuals. Blood samples were collected from family members after informed consent. A medical interview was conducted by phone and the diagnoses confirmed by examination of hospital files. One affected member of each family was screened for mutations in *NKX2-5*. Malformations were listed.
Results/Discussion: One family had a single nucleotide deletion in exon 1 of *NKX2-5* causing a frameshift. The family had five living (three with ASD2, one with ASD2 and first-degree AV-block, one with muscular VSD and ASD) and one deceased individual with DORV-TOF, CoA, VSD and ASD. The mutation segregated with the affected individuals, and we also found one healthy carrier of the

mutation. It is known that mutations in *NKX2-5* can cause ASD and AV-block. In mice the AV-block is progressive and sudden death has been reported.
Conclusion: Screening for known mutations in 46 CHD families revealed a mutation in *NKX2-5* in one family. Further investigations are needed to determine if the combination of ASD and AV-block should lead to genetic investigations.

223: GLOBAL STRAIN RATE IS AN INDEX OF RIGHT VENTRICULAR CONTRACTILITY IN HYPOPLASTIC LEFT HEART SYNDROME (HLHS)

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Background: Systemic right ventricular (RV) function in patients with HLHS is an important aspect during long term-follow up after Fontan repair. Echocardiographic evaluation of RV function is affected by loading conditions. The only load-independent parameter of ventricular function, the end-systolic elastance (Ees), can only be generated with invasive catheterisation. Therefore we sought to determine if parameters obtained by two-dimensional speckle tracking (2DST) are affected by acute changes in preload and correlate with intrinsic RV contractility measured by Ees in HLHS patients after Fontan palliation.
Methods: In 34 patients [median age 5.0 (range 2.9–12.7) years] 2DST and conductance catheter studies were perfomed simultaneously. A balloon catheter in the intra-atrial lateral tunnel was used to modify preload. Measurements were repeated with dobutamine infusion.
Results: RV Ees correlated with global strain rate (SR) (r_s = –0.4, p < 0.01), but not with global strain (S) (r_s = 0.01). S and SR did not change with preload reduction (S: –17.6 ± 3.6 vs –17.3 ± 4.1%, p = 0.6; SR: –0.92 ± 0.20 vs –0.95 ± 0.26 1/s, p = 0.4). S did not change with dobutamine infusion (–17.6 ± 3.6 vs –18.6 ± 4.0%, p = 0.09) whereas SR increased significantly (–0.92 ± 0.20 vs –1.675 ± 0.49 1/s, p < 0.001).
Conclusion: SR was not affected by preload and correlated with Ees of the systemic right ventricle. It may therefore be a useful non-invasive parameter of RV contractility suitable for routine follow up in patients with HLHS after Fontan palliation.

231: CURRENT OUTCOMES OF POSTOPERATIVE EXTRACORPOREAL MEMBRANE OXYGENATION SUPPORT IN CHILDREN WITH FUNCTIONAL SINGLE VENTRICLE PATHOLOGIES

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Background: Improved survival with postoperative extracorporeal membrane oxygenation (ECMO) support has expanded its application to children with complex single-ventricle (SV) pathologies. We sought to examine current era outcomes of postoperative ECMO and compare results between children with SV versus biventricle (BV) pathologies.
Methods: Demographics, anatomical, surgical and support details of children who received postoperative ECMO (2007–2012) were entered into multi-variable regression analysis to determine factors affecting survival.
Results: There were 95 patients (3 days to 16 years) who were divided into SV (n = 28) and BV (n = 67) groups. ECMO was initiated in the OR for failure to wean off bypass (n = 30) or ICU for haemodynamic compromise (n = 65). Thirty-four patients (36%) received rescue ECMO (ECPR) during chest compression and 13 (14%) required re-operation while on ECMO. Forty patients (42%) survived > 24 hours after ECMO discontinuation and 37 (39%) were discharged alive. Mean ECMO duration was 4.9 ± 3.1 days (3.8 and 5.5 days in survivors vs non-survivors, p = 0.003). Survival for ECMO initiated in the OR vs ICU was 43 and 40% (p = 0.66), and 38 and 39% for ECPR and non-ECPR patients (p = 1.0). Survival of SV and BV patients was 32 and 42% (p = 0.18). In the SV group, outcomes diverged with best survival after BT shunt (60%) or Norwood procedure (50%). On the other hand, survival was dismal following PA band, Glenn, Fontan, TAPVC in heterotaxy patients (0%). On multi-variable analysis, cardiac re-operation and leaving cannulation snares were predictors of survival while longer CPR duration, higher pre-arrest and post-ECMO lactate and longer time to lactate normalisation were predictors of mortality. In addition, markers of end-organ injury such as higher creatinine and bilirubin levels, in addition to pulmonary haemorrhage, dialysis requirement and ischaemic brain injury were associated with death.
Conclusions: ECMO plays a valuable role in children requiring postoperative support, including SV patients. Results in SV vary with Norwood procedure and BT shunt having a better prognosis. Timely ECMO prior to emergence of complications and surgical correction of residual lesions may improve survival.

240: INCIDENCE OF ISOLATED AORTIC DILATION IN PATIENTS WITH TURNER SYNDROME

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Introduction: Dilatation of the ascending aorta (AoD) is described in Turner's syndrome (TS) with variable prevalence (6.8–32%). Reported series include patients with associated cardiac anomalies, e.g. aortic coarctation, left outflow tract obstruction and bicuspid aortic valve.
Methods: A retrospective study was carried out with data collected from medical records and echocardiography studies. Patients with TS seen at our centre from 1992 to 2010, free of structural cardiac malformations were eligible when they had ≥ two echocardiographies. Patients with previous cardiac surgery were excluded. Age ranged from infancy to adulthood. From 120 patients, 33 (27.5%) had one or more cardiac anomaly; 18 (15%) with bicuspid aortic valve, 14 (11.6%) with coarctation of the aorta, seven (5.9%) with aortic valve stenosis and three (2.7%) with left superior vena cava. Ascending aorta diameter measurements were collected for all patients and adjusted for body surface area based on our institutional regression equation derived from 1 300 healthy children. AoD was defined as a z-score > 2.
Results: Of the 87 subjects, 28 (31%) were further excluded due to missing data or no follow-up echo. Age was (13.6 ± 8.6 years) at first echo and (20.0 ± 8.1 years) at last follow up. Follow-up duration was (6.4 ± 3.8 years). At initial echo, 10 (16.9%) patients had AoD. A total of 18/59 (30.5%) patients had AoD throughout follow up, with actuarial survival analysis showing freedom from AoD in 90, 77 and 50% at 10, 20 and 30 years old, respectively.
Conclusion: The prevalence of AoD increases with age in TS even in the absence of bicuspid or obstructive left-sided lesions. Specific attention should be brought to healthcare providers attending to TS patients. The impact of confounding factors such as genetic variants (mosaicism), growth hormone therapy and vasoactive medication is yet to be determined.

242: USEFULNESS OF PRO-BNP AS A MARKER FOR MYOPERICARDITIS IN THE EARLY DIAGNOSIS OF KAWASAKI DISEASE

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Background: Early diagnosis of Kawasaki disease (KD) is sometimes difficult and critical to achieve the optimal treatment result, especially in incomplete or atypical KD. We investigated the correlation of serum level of pro-B-type natriuretic peptide (pro-BNP) and the incidence of echocardiographic abnormalities (especially mitral valve regurgitation or pericardial effusion) in patients with KD to discover whether pro-BNP level might be helpful in the early diagnosis of KD.
Methods: Pro-BNP concentrations were measured and echocardiography was performed in the acute stage of 96 patients with KD. Abnormal pro-BNP level and echocardiographic findings were classified into two categories and defined as follows: (1) significant mitral valve regurgitations, (2) significant pericardial effusion.
Results: In the KD patients, significant mitral valve regurgitations were present in 20 patients (20.8%), significant pericardial effusion in eight patients (8.3%). Pro-BNP level was correlated with echocardiographic findings of myopericarditis (mitral valve regurgitation or pericardial effusion). Receiver operating characteristic analysis showed a high value of the area under the curve (0.78) for the detection of myopericarditis with a sensitivity of 69.6% and a specificity of 75.5% for a cut-off value of 927.3 pg/ml.
Conclusions Highly elevated baseline levels of pro-BNP in the acute phase of KD are associated with the presence and extent of myopericarditis and may be helpful in the diagnosis.

243: SUPRA-VALVULAR PULMONARY STENOSIS AFTER ARTERIAL SWITCH OPERATION: EARLY IDENTIFICATION OF PATIENTS AT HIGH RISK FOR RE-INTERVENTION

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Background: Supra-valvular pulmonary stenosis (SVPS) is the most common complication after the arterial switch operation (ASO) for D-transposition of the great arteries (D-TGA) in neonates. While the majority improve over time, some require re-intervention to relieve symptomatic lesions. We hypothesised that early postoperative echocardiography will predict which patients are at higher risk of re-intervention for significant SVPS following ASO.
Methods: A single institution retrospective review of Manitoba newborns ($n = 63$) who had ASO for D-TGA from 1991 to 2010 was undertaken (independent of when, where and who performed their surgery). First postoperative and most recent transthoracic echocardiograms (TTE) of all surviving patients ($n = 59$) were reviewed for SVPS. Patients were categorised as needing re-intervention for SVPS (re-intervention group) versus those who did not (no re-intervention group). Univariate analysis using Fisher's exact test was used to analyse parameters. Significant TTE parameters (gradients > 40 mmHg plus two or more levels of stenosis) were analysed using the Kaplan-Meier method to calculate probability of freedom from re-intervention.
Results: Mean follow-up period was 9.3 years. First postoperative TTE demonstrating two or more levels of stenosis, stenosis gradients > 40 mmHg, both last-mentioned parameters combined, as well as D-TGA plus VSD were all significantly more prevalent in the re-intervention group. Patients who had gradients > 40 mmHg and stenosis at two or more levels, the five-year probability of freedom from re-intervention for SVPS was 40% compared to 100% for those without the two aforementioned parameters (log rank $p = 0.0001$).
Conclusion: SVPS with multiple levels of stenosis causing a gradient > 40 mmHg at initial TTE post-ASO allowed paediatric cardiologists to identify patients at higher risk of future re-intervention for supra-valvular pulmonary stenosis. These findings need to be validated in a larger cohort.

245: RANDOMISED CONTROLLED TRIAL OF INTRAVENOUS IMMUNE GLOBULIN IN ACUTE MYOCARDITIS IN PAEDIATRIC AGE GROUP

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Background: Acute myocarditis is a life-threatening disease which may progress to dilated cardiomyopathy. An autoimmune mechanism has been postulated and immune-modulatory therapies tried with little evidence to support it. The paediatric literature is scant, and there are no randomised controlled studies of immunosuppressive therapy in childhood myocarditis. Hence, this study was designed to assess whether intravenous immunoglobulin (IVIG) improves left ventricular function and survival in children with acute myocarditis.
Methods: A prospective, randomised controlled trial was designed in children with acute (< 3 months) onset of congestive heart failure and echocardiographic documentation of diminished left ventricular function. Children were randomised to receive either IVIG (2 g/kg) plus prednisolone (2 mg/kg/day) or prednisolone (2 mg/kg/day) alone for six weeks' duration. Left ventricular function was assessed at seven days, one month, three months, six months and 12 months after presentation. Primary outcomes included survival and recovery of left ventricular function.
Results: The baseline characteristics were comparable in both groups. Of the 21 children with acute myocarditis, 12 (57.1%) were treated with IVIG plus steroids and nine (42.8%) were treated with steroids. All the patients received anti-congestive therapy and inotropic support as required. Compared with the non-IVIG group, those treated with IVIG had a smaller mean adjusted left ventricular end-diastolic dimension (LVED) at six and 12 months ($p = 0.01$ and $p = 0.009$, respectively). Left ventricular ejection fraction (LVEF) was also higher in IVIG group at six months but did not reach statistical significance. However, at 12 months it was statistically significant ($p = 0.03$). Patients treated with IVIG were more likely to achieve normal left ventricular function ($p = 0.02$). Survival was similar in both groups.
Conclusion: Compared to steroids, IVIG was associated with significant improvement of left ventricular function in acute myocarditis at the end of 12 months but without significant difference in survival rate.

246: REPAIR OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION IN INFANCY-A SINGLE CENTRE EXPERIENCE FROM WESTERN INDIA

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Background: Repair of total anomalous pulmonary venous connections in infancy still carries a significant morbidity and mortality in India. Here we report our experience from a single centre in western India.
Methods: Seventy-two patients were operated in our institute from July 2006 to March 2012. There were 48 males and 21 females. Median age was 89 days and median weight was 3.77 kg; 37% had prior admissions in other hospitals, 84% were prepared with PDE inhibitors and 38% needed emergency surgery.
Results: Sixty-nine patients were included in the analysis, 75% of patients had a delay in diagnosis and 42% deaths were due to preoperative infections. The mean age of surgery in the delayed group was 115 ± 84 days and in the timely diagnosed group was 54 ± 49 days ($p = 0.006$). Preparation for more than two days significantly reduced the mortality ($p = 0.49$), 40% had delayed sternal closure. Mean ventilation was 109 hours; 98.5% of patients received anti-PH medications and 38% received inhaled nitric oxide. Mean inotrope score was 3618; 13% needed additional cardiac surgeries, 5.7% needed additional non-cardiac surgeries, 37% had pre-operative

infections and 58% had postoperative infections; 42% ofdeaths were due to delay in diagnosis and pre-operative infections and 14% to pulmonary hypertensive crisis. There was one late death. In a follow up of three months to six years (n = 53), no patient had residual pulmonary hypertension.
Conclusion: Delay in diagnosis and surgery and rampant use of broad-spectrum antibiotics still contribute to the high mortality rate of this treatable CHD with good long-term prognosis.

250: CHANGES IN PLASMA HYDROGEN SULPHIDE AND THE SIGNIFICANCE IN THE DIAGNOSIS OF KAWASAKI DISEASE

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Objective: Kawasaki disease (KD) is an acute systemic inflammatory disease in childhood. Our aim was to find out a possible biomarker which might represent the development of KD and the subsequent coronary artery lesions.
Method: A prospective unmatched case–control study was designed. Forty-eight KD patients, 57 non-KD fever patients and 27 non-fever children were recruited for the research. KD patients were further divided into subgroups according to coronary artery abnormalities. Plasma hydrogen sulphide (H_2S) was examined by a sulphide-sensitive electrode method.
Results: Plasma H_2S levels in KD patients during the acute period were significantly lower than those during the convalescent period (34.37 ± 8.11 vs 38.17 ± 8.63, $p < 0.05$). Plasma H_2S levels in KD patients during the acute period were significantly lower than those in non-KD fever patients (34.37 ± 8.11 vs 55.77 ± 17.88, $p < 0.05$). There was a negative correlation between the plasma level of H_2S and C-reactive protein, and between the plasma level of H_2S and erythrocyte sedimentation rate in all participants, respectively ($r = –0.511$ and –0.481, respectively; $p < 0.05$). Receiver operating characteristic (ROC) curve analysis revealed a diagnosis of KD (ROC area: 0.905 ± 0.028, $p < 0.001$, 95% confidence interval: 0.849–0.960, optimal cut-off value: 44.705 μmol/l) and a prediction of coronary artery injury (ROC area: 0.834 ± 0.045, $p < 0.001$, 95% confidence interval: 0.745–0.922, optimal cut-off value: 43.78 μmol/l).
Conclusions: Plasma H_2S level in the acute period may be a potentially useful biomarker for assisting the diagnosis of KD and predicting coronary lesions.

252: CHILDREN SUFFERING FROM POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME WITH A MARKED INCREASE IN ERYTHROCYTIC HYDROGEN SULPHIDE HAVE A BETTER THERAPEUTIC RESPONSE TO MIDODRINE HYDROCHLORIDE

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Background: Midodrine hydrochloride is an important therapeutic option for children with postural orthostatic tachycardia syndrome (POTS). However, there are few methods to predict response to the drug. Endogenous hydrogen sulphide plays an important role in the pathogenesis of POTS. The present study was to explore the predictive value of erythrocytic hydrogen sulphide in predicting the therapeutic efficacy of midodrine hydrochloride for children with POTS.
Methods: Sixty-eight children were included in the study, of whom 28 suffered from POTS (POTS group) and 40 healthy children served as control group. Children in the POTS group received midodrine hydrochloride treatment. The erythrocyte hydrogen sulphide production was measured by sensitive sulphur electrode and a receiver operating characteristic (ROC) curve was used to test if erythrocyte hydrogen sulphide could predict the therapeutic response to midodrine hydrochloride for children with POTS.
Results: Hydrogen sulphide production from erythrocytes was significantly higher in children with POTS than in control subjects ($p < 0.001$). Erythrocyte hydrogen sulphide production in responders to midodrine hydrochloride was significantly higher than that in non-responders ($p < 0.05$). ROC curve analysis revealed that the area under curve was 0.857 with a 95% confidence interval (CI) of 0.715–0.999. Erythrocytic hydrogen sulphide production yielded both high sensitivity (81.0%) and specificity (85.7%) in predicting the efficacy of midodrine hydrochloride therapy for POTS in children.
Conclusion: Erythrocytichydrogen sulphide could serve as a useful predictor of therapeutic response to midodrine hydrochloride in POTS of children.

253: SINGLE-VENTRICLE FUNCTION: PREDICTORS OF CARDIAC INDEX AND RELATION TO CAVOPULMONARY HAEMODYNAMICS

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Background: Single-ventricle (SV) lesions are associated with gradual attrition following surgical palliation with the total cavopulmonary connection (TCPC). Abnormalities in ventricular function (VF) are frequently noted. The role of TCPC haemodynamics, which are potentially unfavourable, is also still unclear. In this study, we assessed the ventricular dynamic characteristics of 40 SV patients, and compared those results to cardiac index (CI), systemic venous flow (Q_s), and simulated TCPC power loss (TCPC-EDI) to elucidate biases and functional relationships.
Methods: Cardiac magnetic resonance (CMR) data were retrospectively analysed for 40 patients. Cine ventricular short-axis scans were semi-automatically segmented for all cardiac phases. In addition to traditional VF volume measures, the maximum time rate of volume change (dV/dt_{max}) was calculated and normalised by end-diastolic volume (EDV) for systole and diastole. TCPC geometry was acquired from an axial CMR image stack; relevant flow rates were taken from phase velocity CMR data. TCPC-EDI was calculated from computational fluid dynamics simulations. Statistical significance was assumed for $p < 0.05$.
Results: The natural logs of EDV and stroke volume (SV), and the systolic and diastolic dV/dt_{max} were significantly related to CI. A multivariate regression model was constructed using natural log of EDV (preload), systolic dV/dt_{max} and normalised heart rate, and was strongly predictive of CI ($R^2 = 0.877$). Ventricular volumes were all negatively correlated with TCPC-EDI; EDV had the strongest relationship. Q_s did not significantly correlate with any investigated variables.
Conclusions: EDV (preload), BSA-normalised HR, and systolic dV/dt_{max} were independent predictors of CI in single ventricles. TCPC power loss was inversely related to ventricular volumes, particularly preload.

256: CARDIOVASCULAR PROFILE IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Objective: To study the cardiovascular profile in children with chronic kidney diseases (CKD).
Methods: Clinical data, biochemical profile and staging of 43 children with CKD were correlated with echocardiographic/colour Doppler profile and large-vessel disease [carotid intimal–medial thickness (CIMT)].
Results: Male:female ratio was 1:4 with mean age eight years (4.5 months to 16 years) (n = 43). The mean GFR was 23.22 ml/1.73m^2/min. Malnutrition was universal in all stages, with reflux nephropathy as the commonest aetiology. Nine/41 patients with LVH (LV mass index > 38 g/m$^{2.7}$) and eight/39 patients with high CIMT were symptomatic; 68% had LV dilatation with hypertrophy. The mean EF in early CKD (stage II and III) was 54.3 ± 4.46% vs 63.7 ± 1.9% seen in advanced CKD (stage IV and V) (p = 0.03). Patients with ESRD had higher mean CIMT values (0.14 cm, p = 0.03) than other stages; 57% of patients with uncontrolled systolic hypertension and 43% with uncontrolled diastolic hypertension had LV dysfunction (p = 0.04). Significantly higher CIMT was noted in those with uncontrolled systolic and diastolic hypertension (0.15 cm), which persisted even after control. Children with LVH had significantly higher mean CIMT (0.124 ± 0.006 cm) than those without (p < 0.0001). Vitamin D intake was associated with better LV function (p = 0.03).
Conclusion: Cardiovascular disease was mainly subclinical and noted in all stages of CKD, with large-vessel disease worsening exponentially with ESRD. Eccentric LV hypertrophy with hypertension was an important risk factor contributing to LV dysfunction and persistent large-vessel disease. Vitamin D therapy had a positive impact on global LV function.

261: SENNING OPERATION FOR CORRECTION OF TRANSPOSITION OF THE GREAT ARTERIES: IS THE SURGICAL TECHNIQUE ADOPTED RESPONSIBLE FOR A LONG PATIENT SURVIVAL?

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Objective: Long-term results after the Senning operation for transposition of the great arteries are not known. Sinus node dysfunction and systemic ventricular dysfunction are crucial in patient survival. We evaluated the results, long-term outcome and quality of life in a group of 39 patients.
Methods: The study was a retrospective analysis of 39 (39/40 = 97.5%) surviving surgical patients submitted to Senning operation, with a mean follow-up time of 14.7 ± 3.1 years. Electrocardiogram, echocardiogram and chest radiograph series were performed every six months. Thirty-six patients of this series underwent Holter study and ergometric tests to evaluate the physical capacity. Three patients living overseas were excluded.
Results: There was only one late death (1/39) (late mortality = 2.5%): a 16-year-old patient had an accidental death. The actuarial survival was 95.0% (38/40) (simple or with little VSD, TGA). The probability of staying in sinus rhythm in the 39 surviving patients was 77.1% (30) or normal right ventricular function was 76.5% (29), 10 to 20 years after operation. The incidence of sinus node and right ventricular dysfunction increased gradually over time. No re-operation and pacemaker implantations were performed. Functional class I = 30 (85.7%) cases and functional class II = five (14.3%) cases.
Conclusions: Patients with simple TGA submitted for the Senning procedure, in our experience, presented during late follow up with: (1) low incidence of right ventricular dysfunction and active arrhythmias, (2) low mortality and no sudden death recorded, (3) good quality of life, and (4) satisfactory surgical results (free of re-operation or definitive pacemaker implantation).

263: REGRESSION OF AORTIC DILATION IN PATIENTS WITH HEART BLOCK AFTER PACEMAKER IMPLANTATION

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Introduction: The appearance of ascending aortic dilation in patients with structurally normal hearts and congenital heart block (CHB) has been recently described, but the aetiology and management have yet to be determined. We hypothesised that aortic dilation occurs in children with CHB and that it will regress following the implantation of a permanent pacemaker. We also attempted to identify an association between the presence of maternal antibodies and the degree of aortic dilation.
Methods: We carried out a retrospective review of charts and echocardiograms of patients with CHB, followed at our institution over the past 27 years. Comparison of the diameter of the target structures was based on z-scores derived from normal populations and dilation was defined as z-score ≥ 2.
Results: Seventeen patients met the inclusion criteria in our preliminary review. Mean age at diagnosis was 5.2 months and 53% were males. Positive maternal antibodies were found in five cases, two were negative and 10 were unknown. All the patients underwent pacemaker implantation (mean age of 17 months). Aortic dilatation was found in seven (41%) patients. In those patients, there was a trend towards reduction of the z-score when comparing ascending aortic diameters before (z-score = 5.10) and after implantation (z-score = 2.89, p = 0.01). Also, four patients among the seven had positive maternal antibodies (the other three had unknown serological status).
Conclusion: Although these are preliminary data, ascending aortic dilatation seems to be present in a large proportion of patients with congenital heart block. This associated aortic dilatation seemed to regress after implantation of a pacemaker. Mechanisms involved in this phenomenon might be related to the regularisation of stroke volume. Positive serological status in a large proportion of affected patients might indicate susceptibility secondary to possible *in utero* inflammation of aortic wall tissues. Further studies are needed.

265: DECREASED MORTALITY AND PRESERVATION OF LEFT VENTRICULAR FUNCTION IN DUCHENNE MUSCULAR DYSTROPHY TREATED WITH STEROIDS

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Background: Duchenne muscular dystrophy is a debilitating X-linked recessive disease that afflicts one in 3 500 boys. Cardiomyopathy is a major source of morbidity and mortality. While steroid therapy slows musculoskeletal impairment, the effects on cardiac function and mortality remain to be determined.
Methods and Results: We conducted a retrospective cohort study on 86 patients with Duchenne muscular dystrophy, age 9.1 ± 3.5 years, all of whom received antagonists of the renin–angiotensin–aldosterone system. Over a follow up of 11.3 ± 4.1 years, seven of 63 (11%) patients on steroids died, in comparison to 10 of 23 (43%) without steroids (p = 0.0010). Survival rates at five, 10, and 15 years of follow up were 100, 98 and 79% for patients with steroids versus 100, 72, and 28% for patients without steroids (log rank p = 0.0005). In multivariate analyses, steroid use was associated with an 85% lower mortality rate (hazard ratio 0.15, 95% confidence interval 0.04–0.56,

p = 0.0046). The mortality reduction was driven by significantly fewer heart failure-related deaths (0 vs 22%, p = 0.0010). In multivariate analyses, steroids were associated with an 89% lower rate of new-onset cardiomyopathy (hazard ratio 0.11, 95% confidence interval 0.04–0.29, $p < 0.0001$). The annual rate of decline in left ventricular ejection fraction (–0.43 vs –1.09%, p = 0.0101) and shortening fraction (–0.32 vs –0.65%, p = 0.0025) was less steep in steroid-treated patients. Consistently, the increase in left ventricular end-diastolic dimension was of lesser magnitude (+0.47 vs +0.92 mm per year, p = 0.0105).
Conclusion: In patients with Duchenne muscular dystrophy, steroid therapy was associated with a substantial reduction in all-cause mortality and new-onset and progressive cardiomyopathy.

266: ASSESSMENT OF ATRIAL SEPTAL DEFECTS WITH REAL-TIME THREE-DIMENSIONSAL TRANSOESOPHAGEAL ECHOCARDIOGRAPHY: A NEW INSIGHT INTO DYNAMIC CHANGES WITH CARDIAC CYCLE

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Background: Accurate assessment of ASD anatomy and size are paramount in selecting treatment option and guiding transcatheter device closure. ASDs are well known to have complex geometry and real-time three-dimensional transoesophageal echocardiography (RT3D TEE) may provide superior imaging assessment of ASD compared to conventional two-dimensional TEE. The objectives of this study were to compare measurements of ASD size obtained by RT3D and 2D TEE and to study the dynamic changes of ASD during the cardiac cycle.
Methods: RT3D and 2D TEE imaging were acquired in 34 patients with ASD (age 40.1 ± 16.4 years). *En face* views were used to assess the shape and type of defects. Following full volume acquisition, the ASD diameters in both major and minor axes were measured offline using Xcelera QLab software and compared with values obtained by 2D imaging. Defect areas by 3D planimetry were evaluated for changes during cardiac cycles.
Results: Out of 34 ASDs, 28 were oval, four were multiple and two were complex. RT3D TEE *en face* views enabled better appreciation of ASD shape and orientation in those with multiple and complex ASDs. There was high agreement between ASD diameters measured by RT3D and 2D TEE along both major axis (19.6 ± 5.5 vs 19.0 ± 5.3 mm, r = 0.92, $p < 0.001$) and minor axis (14.8 ± 4.4 vs 14.8 ± 4.1 mm; r = 0.93, $p < 0.001$). There was significant change in ASD size during the cardiac cycle; being smallest during atrial systole and largest during ventricular end-systole. The major axis, minor axis and defect area varied as much as 4.5 ± 3.1 mm, 4.1 ± 2.5 mm and 0.69 ± 1.8 cm^2 ($p < 0.05$), respectively. The defects became more elliptical during atrial systole (eccentricity index increased from 0.61 ± 0.19 to 0.67 ± 0.17, p = 0.029).
Conclusions: RT3D TEE was highly accurate in assessing the anatomy and size of the ASD. It also provided new insight on the dynamic changes of ASD size and shape during the cardiac cycle.

271: LONG-TERM OUTCOMES OF LYMPHOCYTIC MYOCARDITIS IN INFANTS AND CHILDREN: A SINGLE-CENTRE EXPERIENCE

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Background: Lymphocytic myocarditis is a cause of dilated cardiomyopathy (DCM) in children. Survival in the era of immunosuppressive therapy approaches 80 to 100% in registry populations. We assessed long-term outcomes in patients with recovery of ventricular function during early follow up.
Methods: Institutional databases identified patients diagnosed by endomyocardial biopsy with DCM due to lymphocytic myocarditis (Dallas criteria) at the Royal Children's Hospital, Melbourne between 1989 and 2007. Patients younger than six years at latest follow up and post-partum women were excluded. Baseline characteristics and most recent investigations were obtained. Consenting patients were prospectively reviewed with standardised cardiac ultrasound, including assessment of left ventricular diastolic function and tissue Doppler imaging.
Results: Nineteen local patients met eligibility criteria. Fifteen patients consented for study, with no baseline difference between eligible and study populations. Median presenting age was 1.3 years (IQR 0.9, 2.8); four of 15 patients were male. All patients were treated with corticosteroids and cyclosporine, two of 15 patients also received intravenous immunoglobulin post diagnosis. Long-term follow up occurred at a median of 10.3 years (IQR 6.2, 14.3) post illness. No eligible patients had died. All patients were symptom and medication free. Echocardiographic parameters demonstrated median LVEDD z-score 2.1 (IQR 1.4, 2.6) and median LVEF 62% (IQR 58, 66). Seven of 15 patients (47%) had mild left ventricular dilatation, LVEDD z-score median 2.6 (IQR 2.4, 3.0); 1/15 (7%) also had impaired systolic function, LVEF 53%. Diastolic function assessed by mitral inflow Doppler, pulmonary venous flow Doppler, and tissue Doppler imaging was within normal limits in all patients.
Conclusions: Long-term outcomes of lymphocytic myocarditis in children are good with rare systolic dysfunction, but almost 50% had mild left ventricular dilatation 10 years post illness. There was no evidence of diastolic dysfunction by routine echocardiographic assessment in our paediatric population.

273: THE DIAGNOSTIC VALUE OF PLASMA BRAIN NATRIURETIC PEPTIDE IN SYNCOPE OF CHILDREN AND ADOLESCENTS

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Background: Syncope is a common problem in childhood and caused by a variety of underlying diseases. The study was designed to evaluate the diagnostic value of B-type natriuretic peptide (BNP) in syncope in children and adolescents.
Methods: Fifty-seven consecutive children and adolescents hospitalised for syncope were collected from Peking University First Hospital from January 2008 until December 2011. Routine evaluation included patient's history, physical examination, measurement of supine and orthostatic blood pressure and heart rate, standard ECG and basic laboratory examinations. Plasma BNP concentration was measured by radioimmunoassay method. Logistic regression analysis was used to find the independent predictors for cardiac syncope. The diagnostic value of plasma BNP was assessed by ROC curve analysis.
Results: Out of 57 children and adolescents hospitalised for syncope, 34 patients (59.6%) had autonomic-mediated reflex syncope (22 vasovagal syncope, 10 postural othrostatic tachycardia syndrome and two micturition syncope), 10 (17.5%) had cardiac arrhythmias, 11 (19.3%) had structural cardiac/cardiopulmonary disease, and two (3.6%) patients with unknown causes. Patients with structural cardiac syncope had significantly higher plasma BNP values than those with non-cardiac and arrhythmic cardiac syncope (1 955.95 ± 3 322.24 vs 31.01 ± 23.59 and 36.83 ± 25.63 pg/ml, p = 0.000). However, there was no significant difference in plasma BNP level between patients with arrhythmic cardiac syncope and with non-cardiac syncope (31.01 ± 23.59 vs 36.83 ± 25.63 pg/ml, p = 0.991). At cut off of 71.24 pg/ml, plasma BNP was associated with a significant risk for

structural cardiac cause of syncope, yielding high sensitivity (90.9%) and specificity (94.4%) in distinguishing structural cardiac syncope from non-cardiac syncope.
Conclusion: Plasma BNP was helpful in differentiating structural cardiac syncope from non-cardiac syncope, but it had a limited value in the differential diagnosis of arrhythmic cardiac syncope and non-cardiac syncope in children and adolescents.

274: WORLDWIDE EXPERIENCE WITH PORCINE SMALL INTESTINE SUB-MUCOSAL EXTRACELLULAR MATRIX GRAFT (CORMATRIX) IN CONGENITAL HEART SURGERY: A SINGLE-INSTITUTIONAL EXPERIENCE

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Methods: All patients receiving SIS-ECM implants were entered into the ICHF database. This report is a retrospective review of this database. The patients were from Iraq, India, Egypt, Ukraine, Honduras and Ecuador.
Results: There were 174 patients who received 176 implants, including 33 pulmonary valved conduit procedures (14 Ross, six tetralogy with absent pulmonary valve, four truncus arteriosus, four Rastelli, three DORV with RV to PA conduit, two TOF redo RV to PA conduit), 39 primary tetralogy repairs, 34 valve leaflet repairs (included with primary repair), 65 septal patches, seven arterial switch repairs, five AVSD repairs and five Glenn procedures with pericardial closure. Overall mortality was 19/174 or 10.9%. There was no mortality, which could be directly attributed to the ECM. Two patients required early replacement of an ECM pulmonary valved conduit for obstruction distal to the conduit. Two patients receiving aortic leaflet augmentation required aortic valve replacement. All patients having the Ross procedure have had excellent early and midterm results with no mortality or re-operations.
Conclusions: The SIS-ECM (Cormatrix) is an easy-to-use haemostatic alternative to traditional materials. It can be used to fashion valve conduits for right-sided reconstructions and establish competent pulmonary valves in tetralogy repairs needing trans-annular patch. The valve constructs demonstrate excellent results in short- and mid-term follow up. Complete freedom from calcification is an added benefit.

282: MODIFIED CALGARY SCORE IN DIFFERENTIAL DIAGNOSIS BETWEEN CARDIAC SYNCOPE AND POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME-ASSOCIATED SYNCOPE IN CHILDREN

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Objectives: The present study was designed to analyse the usefulness of a modified Calgary scoring system during differential diagnosis between cardiac syncope and postural orthostatic tachycardia syndrome-associated syncope through a large-sample sized clinical investigation.
Methods: The study included 213 children with cardiac syncope or postural orthostatic tachycardia syndrome-associated syncope (101 male and 112 female children), aged between two and 19 years (mean 11.8 ± 2.9). A modified Calgary score was created, which was analysed to predict differential diagnoses between cardiac syncope and postural orthostatic tachycardia syndrome-associated syncope using a receiver operating characteristic curve.
Results: The median of modified Calgary scores for cardiac syncope was –5.0, which significantly differed from that of postural orthostatic tachycardia syndrome (0.0) ($p < 0.01$). The sensitivity and specificity of a differentiation score of less than –2.5 was 96.3 and 72.7%, respectively. Since the modified Calgary score was an integer, when less than –3.0, the diagnosis could be considered as cardiac syncope.
Conclusion: The modified Calgary score could be used to make an initial differential diagnosis between cardiac syncope and postural orthostatic tachycardia syndrome-associated syncope in the clinic.

290: DOES QUALITY OF CONGENITAL HEART SURGERY VARY WITH MOON PHASES?

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Background: Popular belief bestows to some moon phases adverse effects on human activities. Accordingly, worried parents refuse operations for their children during these 'critical' periods. Is quality of congenital heart surgery affected by lunar phases?
Methods: Quality was measured by observed postoperative mortality and morbidity. Lengths of stay at the intensive care unit (ICU) and in hospital were used as morbidity surrogates. Morbidity scores were attributed to each hospital stay according to complications and adverse events occurring after surgery, in conformity with Sata's methodology. Two groups of patients with similar age who underwent similar types of procedures, with the same Aristotle complexity score (8 points) from January 2006 to June 2012 were compared, using the Mann-Whitney test, unpaired *t*-test and Fischer's exact test. Group 1 ($n = 75$) consisted of patients operated upon during so-called unfavourable periods (full moon and moon in Leo). Patients in group 2 ($n = 79$) underwent surgery during presumed favourable moon phases (last week before new moon). The difference was considered not significant at a *p*-value above 0.05.
Results: No patient died: a mortality of 0% for both groups. ICU and in-hospital lengths of stay were 3.85 ± 3.91 and 13.19 ± 7.31 days for group 1, versus 4.21 ± 4.96 and 14.53 ± 8.51 days for group 2: $p = 0.62$ and 0.30, respectively; 57.3% (43/75) of patients in group 1 and 57.0% (45/79) in group 2 had an uneventful postoperative course; $p = 1$. The severity of complications that occurred in the other patients was similar for both groups; $p = 1$. The morbidity score reached 1.55 ± 1.72 points for group 1 and 1.50 ± 1.58 points for group 2; $p = 0.85$.
Conclusions: Quality of results did not vary with moon phases. One should not worry at all about lunar phases when planning and performing congenital heart surgery.

295: PERMANENT PACEMAKER IN PREGNANCY: ANALYSIS OF SIX CASES

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Background: Outcome of pregnancy in patients with a pacemaker is not fully understood because of limited numbers of cases. We present

six cases and discuss their management issues.
Methods: We conducted a retrospective analysis of seven pregnancies in six patients with pacemaker implantation from 2008 to 2012 in our institution. The course of pregnancy, perinatal outcome, maternal baseline disease, and pacing mode were studied.
Results: All six patients had atrio-ventricular block (AVB). Of these, three patients had structural cardiac lesions (single ventricle in two and atrio-ventricular septal defect in one), and the other patients had normal structure heart (cardiomyopathy in one and congenital AVB in two). In five patients, implantation of the pacemaker was performed before pregnancy. The mode of pacemaker was DDD in four (transvenous lead), and VVI in one patient (epicardiac lead). One patient who had congenital AVB required pacemaker implantation during pregnancy due to a large difference between the atrial and ventricular rate. There was no maternal or foetal death. Three patients had four uneventful vaginal deliveries. One patient delivered a baby by cesarean section for intra-uterine growth retardation and non-reassuring foetal status. One patient who had a single right ventricle with a pacing due to surgical AVB developed cardiac decompensation in the third trimester, and was induced at 36 weeks' gestation. One patient with VVI pacing system had to have the ventricular pacing rate changed due to failure of cardio-acceleration during the intrapartum period.
Conclusion: There were no serious maternal or neonatal complications. The outcomes of pregnancy in patients with pacemakers were uneventful by multidisciplinary medical care.

309: LONGITUDINAL COMPARISON OF OUTCOMES BETWEEN PRE VERSUS POSTNATALLY DIAGNOSED INFANTS WITH TRANSPOSITION OF THE GREAT ARTERIES

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Prenatal diagnosis of transposition of the great arteries (TGA) may decrease peri-operative morbidity but few data exist regarding its long-term impact or costs. Linking the Utah Birth Defects Network (UBDN, a statewide surveillance system) and the Utah Population Database (UPDB, statewide in-patient discharge abstracts and charges) captures all hospitalisations regardless of whether ICD-9-DM codes include the primary heart defect. We sought to determine differences in longitudinal medical encounters and in-patient costs for prenatally (predx) and postnatally (postdx) diagnosed TGA.

TGA cases identified at the UBDN (born 1997–2009) were reviewed and preterm gestation excluded. Linkages to the UPDB identified in-patient encounters and charges for cases (up to 12/2011) and their mothers (10 months prior to birth). Charges were adjusted to 2011 using the consumer price index and cost/charge ratio. We compared cost, using generaliszed linear regression with gamma distribution and log link function, and in-patient encounters, using multivariate modeling, adjusting for demographic and clinical risk factors.

Of 108 cases identified, 12 (11%) were prenatally diagnosed. Predx were similar to postdx in gestational age (38.6 vs 38.8 weeks), birth weight < 2.5 kg (0 vs 4%, $p = 0.62$) and additional congenital defects (8 vs 10%, $p = 0.39$). There were seven deaths (6%) with no difference in survival between groups (mean follow up 7.4 years). Predx cases had similar hospitalisations (median two in both groups), but higher total LOS (23 vs 19 days, $p < 0.004$) as well as higher costs for both mothers ($8 200 vs $5 200) and index cases ($290 000 vs $151 000) after adjusting for gestational age, birth weight and additional congenital defects ($p = 0.001$).

Predx TGA cases had longer hospitalisations after adjusting for available risk factors. Both mothers and infants in the predx group had higher longitudinal in-patient medical costs. Ongoing investigation should explore specific cost components responsible, including additional testing, surgical delays and other risk factors.

311: MYOCARDIAL PERFORMANCE INDEX (TEI INDEX) IN CHILDREN AND ADOLECENTS IN A REGION OF SOUTHERN BRAZIL

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Introduction: Myocardial performance index (MPI) is a relatively simple and non-invasive echocardiographic approach to a quantitative assessment of the global ventricular cardiac function.
Objectives: The main purpose of this study was to define values for the right and left ventricular MPI in healthy children and adolescents and to compare the values between healthy subjects and a small sample of patients with dilated cardiomyopathy (DCM).
Methods: Observational and cross-sectional study was done at the Hospital Infantil Joana de Gusmão, in Florianopolis, Santa Catarina, Brazil. The left ventricular MPI (LVMPI) and right ventricular MPI (RVMPI) were measured in 121 healthy children and adolescents and in seven children with DCM. Also measured were the mitral valve peak E/A ratio (MVE/A) and tricuspid valve peak E/A ratio (TVE/A). The index was obtained from conventional inflow and outflow Doppler velocity waveforms of each ventricle. For evaluation of the intra- and inter-observer reproducibility, the author and another independent observer analysed 28 children and adolescents, randomly selected from the healthy group, at a minimum of 30-day intervals between the measurements.
Results: Among the healthy individuals, the age ranged from three to 173 months. The LVMPI was 0.37 ± 0.08, RVMPI was 0.32 ± 0.06, peak MVE/A ratio was 2.13 ± 0.57 and the peak TVE/A ratio was 1.01 ± 0.08. Among the DCM children, the LVMPI was 1.16 ± 0.16, RVMPI was 1.05 ± 0.42, peak MVE/A ratio was 1.14 ± 0.38 and the peak TV/EA ratio was 1.01 ± 0.08.
Conclusions: Age did not affect MPI in healthy children and adolescents. The LVMPI and RVMPI to DCM individuals were significantly prolonged compared to the values in healthy children and adolescents. The intra-observer reproducibility was high for LVMPI and moderate for RVMPI and inter-observer reproducibility was considered high for LVMPI and low for RVMPI.

316: ARRHYTHMIA PHENOTYPE DURING FOETAL LIFE PREDICTS LQTS MUTATION: RISK STRATIFICATION OF PERINATAL LONG QT SYNDROME

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Background: Torsades de pointes (TdP) and/or second-degree atrio-ventricular block (2° AVB) are signature rhythms for perinatal LQTS, known for their high morbidity and mortality rates. We hypothesised that the clinical profile of patients with these complex foetal arrhythmias might be genotype specific.
Methods: Perinatal records of LQTS subjects exhibiting complex foetal arrhythmias were reviewed. Foetal echocardiograms, neonatal ECG and genetic testing were evaluated.

Results: We studied 11 LQTS subjects exhibiting complex foetal arrhythmias. Mutations in one of the three major LQTS susceptibility genes were identified in nine: SCN5A (five), KCNH2 (two), and KCNQ1 (two). Most mutations were *de novo* including four with SCN5A-R1623Q. TdP occurred in seven foetuses (mean gestational age = 30.5 weeks) and eight neonates; 2° AVB occurred in four foetuses/neonates. TdP exhibited two patterns: incessant and fast (> 270 bpm) or intermittent and slow (< 250 bpm). All cases with SCN5A mutation had fast-incessant TdP while cases with KCNH2 mutations had slow-intermittent TdP. Cases with KCNQ1 mutations had 2° AVB. Foetuses with TdP were delivered earlier (33.6 weeks) than those with 2° AVB (38.5 weeks). Neonatal QTc of subjects with TdP were longer (652 ± 42 ms) than subjects with 2° AV block (507 ± 43, $p = 0.01$). Prenatal treatment was administered in six cases without maternal complications; four foetuses improved and TdP ceased in two. Despite medical and pacemaker therapy, cardiac arrest ($n = 6$) resulting in sudden death ($n = 1$) was common.
Conclusion: Complex rhythm phenotypes of foetal LQTS have genotype-suggestive features that, along with QTc duration, may risk stratify the perinatal management of LQTS.

321: LEFT-AXIS DEVIATION IN THE PAEDIATRIC POPULATION: A MODERN STUDY

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Background: Few recent large studies have investigated left-axis deviation on electrocardiogram with associated medical conditions in children. Using electronic medical records and the computer database of electrocardiograms within an integrated healthcare system, we documented these associations.
Methods: During a five-year period (2007–2012), the computer database of all electrocardiograms performed (3 267 893 in 1 360 489 individuals) were screened. Patients 0–18 years of age with left-axis deviation (–30° to –90°) were identified. Each patient's electronic medical record was used to identify known associations with left-axis deviation and these were recorded.
Results: There were 122 484 electrocardiograms performed on 79 938 individuals (0–18 years of age). Of these individuals, 886 demonstrated left-axis deviation on ECG (1.1%). Of these, 491 were considered normal as they had no record of heart disease, thoracic or genetic abnormalities. There were 279 individuals with structural heart abnormalities: ventricular septal defect (65), complete atrio-ventricular septal defect (59), partial atrio-ventricular septal defect (22), tricuspid atresia (10), single-ventricle arrangement (24), atrial septal defect (19), pulmonary stenosis (18), coarctation of the aorta (10), left ventrticular hypertrophy/cardiomyopathy, aortic stenosis (12), tetralogy of Fallot (11) (majority postoperative), patent ductus arteriosus (five), transposition of the great arteries (five) and isolated anomalous pulmonary venous return (four). There were 57 with conduction abnormalities: Wolff–Parkinson–White syndrome (37), paced rhythm (17), long QT syndrome (two) and complete heart block without a pacemaker (one). There were 10 patients with Noonan syndrome, and 32 with short stature or other genetic abnormalities. Eighteen had scoliosis, pectus excavatum or dextrocardia.
Conclusions: This study documents the associations of left-axis deviation in children and demonstrates the utility of using electronic medical records within an integrated medical system to gather such data.

326: SURGICAL AND INTERVENTIONAL MANAGEMENT OF PATENT DUCTUS ARTERIOSUS IN PREMATURE INFANTS IN WESTERN AUSTRALIA: A NINE-YEAR EXPERIENCE

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Background: There is significant controversy regarding optimum PDA management in preterm infants. KEMH is the sole tertiary perinatal hospital caring for all infants born at < 30 weeks in WA. KEMH has adopted a conservative approach to neonatal duct ligation for many years. After discharge it was apparent some preterm infants required intervention for PDA at the only tertiary children's hospital (PMH) in WA.
Methods: The aim of this study was to review a geographic cohort of all surviving preterm infants born at < 30 weeks in WA from January 2003 to December 2011 requiring surgical or interventional closure of a PDA. The medical records of these infants were reviewed to determine complications and outcomes. The neonatal survival and major duct-related morbidity data for the whole cohort were compared to data from the Australian and New Zealand Neonatal Network (ANZNN).
Results: A total of 1 652 live births (GA < 30 weeks) had neonatal management at KEMH, of which 1 475 (89.5%) infants survived to discharge. Only 2.0% of survivors (median GA 25 weeks and median BW 735 g) received surgical or interventional closure at PMH during the study period. Eleven infants had surgical closure during their initial neonatal hospital admission and 20 after discharge; 17 interventional and three surgical. All surgical and interventional procedures produced successful closure. Post surgery, three had a pneumothorax and one vocal cord palsy. One death occurred late and unrelated to surgical closure. No significant complications occurred with interventional closures and 14 were day cases. Overall, the incidence of CLD and NEC in this cohort was similar to the ANZNN mortality and morbidity data.
Conclusions: Most PDAs close with conservative medical treatment. Continued surveillance is needed to manage PDAs that remain open. There is no evidence of any increase in morbidity or mortality using this approach.

332: NURSE-LED ECHOCARDIOGRAPHIC SCREENING FOR RHEUMATIC HEART DISEASE IN FIJI: DESIGN OF A TRAINING SYLLABUS

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Background: Rheumatic heart disease causes significant morbidity and mortality in Fiji. Antibiotic prophylaxis is most effective for mild cases, many of whom are asymptomatic. Screening with echocardiography is the most sensitive screening approach, but is limited by lack of resources, particularly experienced cardiologists and technicians. Training nurses in basic echocardiography for screening is a potential solution. A previous pilot programme showed that training nurses is feasible, but found that improvements were needed in screening protocols, referral criteria and the extent of practical experience. We hypothesised that a structured eight-week syllabus, including extensive supervised field experience, would provide appropriate and adequate training for echocardiographic screening for RHD.

Methods: We designed an eight-week training syllabus for primary health workers. The syllabus included tutorials in basic cardiac anatomy and physiology, pathophysiology of RHD, and practical sessions in basic echocardiographic screening. We developed a 14-step screening protocol, and devised referral criteria based on identification of significant mitral or aortic regurgitation. The syllabus included seven weeks of supervised screening practice in schools.
Results: Seven nurses from across Fiji participated in the training. Despite minimal relevant experience, nurses were quick to learn basic anatomy and pathophysiology for understanding rheumatic valvular pathology. Nurses demonstrated rapid acquisition of basic echocardiographic skills. Use of a simplified screening protocol was highly valuable.
Conclusions: Training of primary health workers with limited prior knowledge or experience in basic screening echocardiography for RHD is feasible. The results of the current fieldwork phase, involving screening of 2 000 children, will provide further information about the sensitivity and specificity of this approach to screening. A structured syllabus, including screening and referral protocols, has been developed, which may be useful for training other workers in Fiji and other resource-poor settings.

336: FIRST EXPERIENCES WITH ANGIOTENSIN II RECEPTOR BLOCKER IN PAEDIATRIC PATIENTS WITH MARFAN'S SYNDROME AND ENLARGED AORTIC ROOT

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Background: Progressive enlargement of the aortic root leading to dissection or rupture is the main cause of premature death in patients with Marfan's syndrome (MFS). Standard prophylaxis is beta-blockers (BB). A new concept in adults with MFS is the treatment with angiotensin II receptor blockers (AT). So far, there is a lack of experience regarding the treatment of children with AT. We present a comparison of our first patients treated with AT or BB.
Methods: We identified a cohort of 41 paediatric MFS patients with aortic root enlargement (mean age 7.8 years). In 28 MFS patients a prophylaxis with BB (n = 15) or AT (n = 13) was subscribed. A control group of 15 children has not been under any medication during evaluation period while seven of these patients later had to switch to AT treatment due to progressive enlargement of the aortic root. Retrospective analysis of the impact of medical treatment was performed by comparing the rates of change in diastolic aortic root diameter (DDAR) over a mean period of 8.21 ± 5.1 (1.3–30.6) months.
Results: Mean DDAR increase over 8.7 months was 0.69 ± 1.37 (–0.7–4.0) mm in patients with BB prophylaxis while mean DDAR increase over a 5.9-month period was –0.08 ± 1.11 (–2.0–1.0) mm in patients with AT prophylaxis respectively ($p > 0.05$). Mean DDAR in the control group over 6.6 months was 1.59 ± 1.22 (–0.5–4.0) mm. DDAR was significantly lower in patients on medication with BB ($p = 0.056$) and AT ($p = 0.001$) compared to the control group.
Conclusion: In this small, non-randomised cohort study, the use of AT or BB therapy in children with Marfan's syndrome slowed the rate of progressive aortic root dilation while therapy with AT seemed to be more effective. These findings require confirmation in further studies with a prospective and randomised study design.

342: THE SIX-MINUTE WALKING TEST AS A PROGNOSTIC MARKER IN CHILDREN WITH DILATED CARDIOMYOPATHY: PRELIMINARY DATA FROM THE CARDIOMYOPATHY STUDY (CARS) IN CHILDREN

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Background: The maximal oxygen uptake (VO_{2max}) has been used as a prognostic marker to stratify adults with (severe) heart failure. In children, VO_{2max} may also have prognostic value, but it may be challenging to measure, especially at a young age. Therefore, we determined whether the six-minute walking test (6MWT) could be used as a prognostic marker in children with heart failure and dilated cardiomyopathy (DCM). The 6MWT, which measures the distance that is voluntarily covered in six minutes, is easy to perform and a good reflection of daily activity.
Methods: In a prospective longitudinal multicentre study the 6MWT was performed in children with DCM. Using reference data from Geiger *et al.* (2007), the 6MWT results were transformed to *z*-scores. Death, heart transplantation and institution on mechanical support were defined as primary endpoints. The 6MWT performance of children with and without an endpoint was compared.
Results: Twenty-four children with DCM (mean age 13 ± 3 years) were included and performed 61 6MWTs without adverse events, during a mean follow-up period of 11 ± 5 months. The mean (SD) 6MWT distance *z*-score was –3 ± 3.2 compared to reference data ($p < 0.01$). In children who reached one of the pre-defined primary endpoints, the distance covered during the 6MWT was significantly lower (*z*-score –5.7 ± 2.9) in the three months before the endpoint was reached, compared to those not reaching a primary endpoint (–2.2 ± 2.1) ($p < 0.05$).
Conclusion: The six-minute walking test is easy to perform and safe in children with DCM. The 6MWT performances of children with DCM were significantly reduced compared to normative data. Children reaching a primary endpoint performed significantly worse than children with a favourable course of the disease. Longitudinal assessment of the six-minute walking test may have prognostic value in children with DCM.

344: REDUCED HEALTH-RELATED QUALITY OF LIFE IN CHILDEREN WITH DILATED CARDIOMYOPATHY

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Background: Dilated cardiomyopathy (DCM) is an important cause of heart failure in children and carries a high risk of mortality and morbidity. In adults with heart failure moderate to low health-related quality of life (HRQoL) has been reported. Information on HRQoL in children is lacking. Therefore, in this multicentre study we evaluated the HRQoL in children with DCM.
Methods: HRQoL was measured with international standardised QoL questionnaires: the Infant–Toddler Parental QoL questionnaire (age 0–3 years); Child Health questionnaire (CHQ) Parental Form 50 (age 4–17 years) and the CHQ Child Form 87 (children aged 10–17 years). Results were compared with Dutch normative data. A subgroup analysis was done to evaluate the parent–child agreement.
Results: Parent reports of children aged 0–3 years (n = 19) showed significantly poorer scores on 'physical functioning', 'general health' and 'growth and development' compared to normative data ($p < 0.05$). In contrast, on subscales 'general behaviour', 'getting along' and family cohesion', significantly better scores were found. Parents of children aged 4–17 years (n = 33) reported impairments (significantly reduced scale scores) on nearly all subscales, except for 'general behaviour' and 'family cohesion'. On self reports, patients with DCM (aged 10–17 years, n = 24) also scored significantly lower on half of the subscales. Remarkably, they reported better 'general behaviour' ($p < 0.05$) and their scores for 'bodily pain' and 'mental health' were comparable to normative data. Strong parent–child agreement was found for most physical subscales (r^2 = 0.5–0.8; $p < 0.01$). To a lesser degree we observed agreement between parents' and children's reports on the psychosocial scales as 'mental health' and 'self esteem' ($r^2 = 0.2$, $p < 0.05$).
Conclusion: In children with DCM, HRQoL is considerably impaired. Low scores were specifically found on physical subscales, in which strong parent–child agreement was found. Interestingly, our results showed better psychosocial and family functioning in families with young children with DCM.

346: RESOURCE UTILISATION AND OUTCOMES OF INFECTIVE ENDOCARDITIS IN CHILDREN: A MULTICENTRE STUDY

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Background: The objective of this study was to evaluate the resource utilisation and outcomes of IE in children with and without pre-existing heart disease (HD vs NHD).
Methods: Children < 19 years old without chronic non-cardiac conditions hospitalised from 2004 to 2010 with IE at one of 37 US children's hospitals in the Paediatric Health Information Systems database were included. Comparisons were made between HD and NHD cohorts. Regression analysis was used to evaluate factors associated with poor outcomes (mortality, mechanical cardiac support, renal failure or stroke).
Results: Of 1 033 IE cases, 663 had HD and 370 had NHD. Factors associated with poor outcome in the HD cohort: higher risk of mortality score (OR = 7.9), mechanical ventilation (OR = 3.1), anti-arrhythmic use (OR = 2.7) and vasoactive medication use (OR = 3.8) and in the NHD cohort: renal failure (OR = 19.3), higher risk of mortality score (OR = 4.2), anti-arrhythmic use (OR = 3.8) and mechanical ventilation (OR = 3.2).
Conclusions: Compared to IE in NHD, IE associated with HD was more likely to occur in younger patients, less often related to *Staphylococcus* and more often related to *Streptococcus*, and was associated with higher mortality, greater likelihood of needing cardiac surgery, but a lower risk of stroke. Factors associated with poor outcome in the entire group included mechanical ventilation, higher risk of mortality score at admission and anti-arrhythmic use. In addition, renal failure in the NHD and vasoactive medication use in the HD cohort were associated with poor outcomes.

363: EPIDEMIOLOGY OF RHEUMATIC HEART DISEASE IN TUVALU

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Background: The World Heart Federation, Menzies School of Health Research and Tuvalu Ministry of Health conducted the first school-based echocardiographic screening in Tuvalu in 2012. The aim of the screening was to detect cases of rheumatic heart disease among school-aged children and to determine the burden of rheumatic heart disease in this Pacific Island nation.
Methods: A technical team from Australia and the Fiji Islands travelled to Tuvalu for 10 days to conduct the school-based screening. Screening was undertaken in three schools. A local radiographer assisted the team. The visiting paediatric cardiologist conducted 1:1 training in echo screening with the local radiographer and doctors.
Results: A total of 571 children between the ages of six and 15 years were screened on the main island of Funafuti. This represents approximately 50% of the age-eligible Funafuti population and 25% of the national age-eligible population. Of the 571 children screened, a total of 26 definite and probable cases of rheumatic heart disease (RHD) were detected using the recently published World Heart Federation criteria for echocardiographic screening, 65% of the cases detected were female. RHD prevalence in Tuvalu is estimated at 31.5 per 1 000. The majority of those affected had mild disease. Four cases of congenital heart disease were detected. All children who were found to have either definite or probable RHD were counselled with their parents/guardians, registered with the programme and secondary prophylaxis was commenced. Probable cases of RHD will be reviewed after one year when the technical team returns to Tuvalu.
Conclusion: The RHD prevalence in Tuvalu is consistent with rates found in nearby Pacific nations of Tonga 33 per 1 000 and Fiji 32 per 1 000. Funafuti is densely populated, with poor living conditions. Further work is required to determine risk factors and to estimate the burden in outlying islands of Tuvalu.

364: SCHOOL-BASED SCREENING FOR RHEUMATIC HEART DISEASE IN NAURU

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Background: Nauru, a Pacific nation with an estimated population of just 10 000, commenced a rheumatic heart disease (RHD) prevention programme in 2006. In 2012, the Ministry of Health with the support of an international team conducted their first school-based screening for RHD. The aims of the screening were to estimate disease burden to enable for the prioritisation of RHD-related activities and to identify children with RHD early, to allow for timely enrolment into secondary prophylaxis programmes.
Methods: School-based screening of grades 3, 6 and 9 students at all four schools was undertaken using portable echocardiography.

Echocardiography images were reviewed on-site by a paediatric cardiologist and were reported on using the World Heart Federation echocardiographic criteria. The local team was assisted by an RHD coordinator from Fiji to build capacity, support and expand the local RHD programme.
Results: Of the total of 2 374 school-aged children of Nauru, 462 pupils aged six to 15 years underwent echocardiographic screening for RHD; 53% were female. Seven cases of definite RHD were identified including two previously known cases; a prevalence of 15.1 per 1 000. Five had mild disease, one had moderate and one had severe. Nine additional borderline RHD cases were detected. All pupils with RHD had a clinical review, received counselling and were registered with the RHD programme. Those with definite RHD were commenced on secondary prophylaxis. Pupils with borderline RHD and a suspected history of rheumatic fever based on clinical review were also commenced on benzathine penicillin. All cases were offered follow up by the visiting cardiologist in 12 months.
Conclusion: A prevalence of definite RHD of 15.1 per 1 000 is consistent with similar screening studies conducted in neighbouring Pacific Island countries. A contributing factor to the prevalence of RHD may include the overcrowded living conditions in Nauru.

367: CURRENT OUTCOME VARIABLES IN NEONATAL EBSTEIN'S ANOMALY

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Background: Neonates with Ebstein's anomaly have a high mortality rate. Currently, cases of Ebstein's anomaly have frequently been detected before birth, and they make up the main group of the population of neonatal Ebstein's anomaly. We performed this study to find the outcome variables in current populations of neonatal Ebstein's anomaly.
Methods: The records of 59 patients with neonatal Ebstein's anomaly who were managed in Asan Medical Centre between January 2001 and June 2012, were investigated retrospectively. In 46 of them, a prenatal diagnosis was done.
Results: Operative procedures: pulmonary valvotomy, palliative shunt operation, tricuspid valvuloplasty or right ventricular exclusion, were performed in 27 patients. Median follow-up period was 1.96 (range 0.0–10.4) years. The overall mortality rate was 23.7% (14/59). Ten of 14 died during the neonatal period. One-year and five-year survival rates were 78.6 and 76.3%, respectively. Variables found to be related to the time to death on univariate analysis were foetal distress ($p = 0.002$), prematurity ($p = 0.036$), low birth weight ($p = 0.003$), diameter of atrial septal defect ($p = 0.022$), pulmonary stenosis/atresia ($p = 0.001$). Carpentier-Edwards classification ($p = 0.175$) and Celermajer index ($p = 0.958$) were not significant variables. Multivariate analysis showed that foetal distress ($p = 0.004$) and pulmonary stenosis/atresia ($p < 0.001$) remained significant.
Conclusion: Foetuses in whom Ebstein's anomaly are diagnosed should be closely monitored throughout pregnancy, especially those with pulmonary obstruction. Strict cooperation between obstetrician, neonatologist, paediatric cardiologist and paediatric cardiac surgeon is essential for their survival.

368: SECONDARY QT PROLONGATION DUE TO EXOGENIC STRENGTH TO PRECORDIAL REGION MAY INDUCE COMMOTIOCORDIS

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Background: Commotiocordis (CC) is defined as the combination of ventricular fibrillation (Vf) and sudden cardiac death secondary to blunt chest wall impact, and it occurs especially in healthy children. We have recently experienced 10 patients who exhibited typical QT prolongation (LQT) in electrocardiogram (ECG) after receiving external force against thea thoraco-abdominal region. We studied the clinical, electrocardiographic and laboratory features to clarify whether there is a relationship between CC and secondary LQT.
Methods: We reviewed the charts and ECGs of 10 patients (two girls and eight boys, six to 15 years) who showed LQT in the first ECG following blunt trauma against the chest and abdomen, and analysed their clinical backgrounds, laboratory data and ECGs.
Results: Clinical features: the cause of the hard knocks was as follows: traffic accident (four patients), collision during sport (three patients), and a fall (three patients). Five patients suffered from liver, spleen and intestinal tract damages, three patients showed a skull fracture, one exhibited hemi-pneumothorax, and another showed severe dyspnoea.
ECG findings: the first ECG on arrival disclosed long QTc (442–503) and two peaks of T wave in V2–3 leads. The second ECG demonstrated normal QTc (396–429) and normal T wave forms. No ECGs of the patients showed Vf or cardiac arrest.
Laboratory data: laboratory studies on arrival showed increased blood glucose concentrations (132–222) and decreased potassium levels (2.8–3.9), which both subsequently normalised.
Conclusions: CC usually results from Vf following a mechanical stimulus on the precordial region. The over-excited sympathetic nervous system after a severe shock causes over-secretion of adrenaline. This condition could increase the intra- and extracellular gradient of potassium ions, which results in LQT. Children may incur sudden death following blunt trauma to the chest from TdP due to secondary LQT.

374: PROGNOSTIC IMPLICATIONS OF TWO-DIMENSIONAL, M-MODE AND DOPPLER ECHO INDICES OF RIGHT VENTRICULAR FUNCTION IN CHILDREN WITH PULMONARY ARTERIAL HYPERTENSION

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Background: Right ventricular (RV) function may be a key determinant of mortality in paediatric idiopathic pulmonary artery hypertension (iPAH) and that associated with congenital heart disease (cPAH), but echo indices of RV function have not been adequately studied.
Methods: Children (0–18 years) with iPAH and cPAH were retrospectively studied. RV function indices (indexed RV end-diastolic area, fractional area change, tricuspid annular excursion, right atrial volume) were analysed at diagnosis and at last follow up. Indices were compared between iPAH and cPAH patients at baseline and follow up. Living iPAH patients (group 1) were compared with deceased/transplanted patients (group 2) at the time of the study. Cut-off points predictive of survival were generated (ROC) and Kaplan–Meier survival as analysed.
Results: Fifty-four patients [36 cPAH (7.5 ± 5.9 years; M:F 12:24); 18 iPAH (8.9 ± 5.7 years; M:F 7:11); group 1 'alive', $n = 12$, group 2 'dead\lung transplant', $n = 6$) were studied. Median follow up was 4.3 (0.2–7.4) years. In iPAH patients, RV indices were similar between groups 1 and 2 at presentation. However, at follow up, despite similar pulmonary artery pressures and PVRi, RV function was significantly worse in group 2. A small pericardial effusion was seen in three patients in group 2 vs 0 in group 1. RV function was

significantly decreased in iPAH vs cPAH patients during follow up. In iPAH patients, survival was significantly different based on RV function cut-off points (RV fractional area changes < than 15.5%, RV end-systolic area > than 18.4 cm^2/m^2, RV end-diastolic diameter in cm *z*-score > than 4.8 and TAPSE *z*-score < than –4.3).
Conclusion: Conventional echo RV function indices including RV end-diastolic area, fractional area of change and tricuspid annular excursion appear to be useful for prognosis in children with PAH.

375: ARTERIAL SWITCH OPERATION: TIMING OF SURGERY BASED ON NEUROIMAGING

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Objectives: We examined the relationship between duration of pre-operative hypoxaemia and prevalence of postoperative brain injury in neonates with TGA who were managed with two different strategies.
Methods: From September 2009 to March 2012, 93 newborns with TGA underwent brain MRI. Patients were divided in two groups according to ASO time. Group A included 35 patients who underwent surgical repair in the first hours of life (4 ± 2 hours). MRI was performed pre-operatively in patients aged 3.6 ± 1.9 hours, 30 of them had postoperative MRI at 10.8 ± 3.2 days of age. Group B included 58 patients with a conventional surgical approach who underwent ASO at the age of 8.1 ± 4.0 days; in 43 patients BAS was performed prior to surgery. All of them had pre-operative MRI at age 6.6 ± 4.0 days. Correlations between MRI findings and systemic arterial oxygen saturation were analysed.
Results: Parenchymal brain damage was diagnosed in 40% of patients (n = 12) from gGroup A in comparison to 57% (n = 33) from group B. Postoperative parenchymal brain damage was limited to WMI. There was no focal arterial brain stroke or basal ganglia injury in group A. In group B arterial stroke was diagnosed in 26% of patients (n = 15). We found a correlation of risk of brain injury with level and duration of systemic hypoxaemia. In patients with WMI, the average value of SaO_2 was 60.2 ± 13.7% compared to 77.5 ± 8.4% in patients without brain injury ($p < 0.001$). The mean age in newborns with WMI was 7.25 ± 4.2 days and –4.6 ± 3.1 ($p < 0.001$) without it. Mean age for BAS in neonates with brain stroke was 6.7 ± 2.9 days, and without stroke 2.5 ± 1.5 days.
Conclusion: Prolonged systemic hypoxaemia is associated with higher risk of brain injury. Our brain MRI-based research shows that early TGA repair enables prevention of brain injury associated with prolonged systemic hypoxaemia.

378: CORRELATION BETWEEN QRS DURATION, PULMONARY ARTERY REGURGITATION AND RIGHT VENTRICLE PERFORMANCE IN TOTALLY CORRECTED TETRALOGY OF FALLOT PATIENTS

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Background: Tetrallogy of Fallot (TOF) is the most common cyanotic congenital heart disease and total correction has a good prognosis. Despite this progress, pulmonary artery regurgitation and progressive disorders of the right ventricle affect the long-term prognosis after total corrective surgery by causing fatal arrhythmias and death. In this study we reviewed the correlation between QRS duration with pulmonary artery regurgitation and right ventricle performance.
Methods: We reviewed 57 TOF patients with total corrective surgery who returned to the paediatric cardiology clinic at the Children's Medical Centre of Tehran. They had 13-lead ECGs to calculate QRS duration, continuous Doppler echocardiography to calculate pulmonary artery regurgitation (total time of pulmonary artery insufficiency/diastolic time), and Tei index to assess performance of the right ventricle. QRS duration was measured in milliseconds (ms) in the D2 lead and the patients were categorised into the groups. Group 1 was patients with normal QRS and QRS duration < 120 ms. Group 2 was patients with long QRS and duration between ≤ 120 and < 160 ms. Group 3 patients had QRS duration ≥ 160 ms. Pulmonary artery regurgitation was measured as a percentage and its index (PRi) calculated. The group with mild regurgitation was PRi ≥ 70% and the group with severe regurgitation was PRi < 70%.
Result: Twenty-one of 57 patients were in group 1 (36.8%). Among the 36 patients with long QRS, 24 patients (42.1%) were in group 2 and 12 patients (21.1%) in group 3, and had a QRS duration > 160 ms. Only one patient had a QRS duration > 180 ms. PRi in group 1 was 80 ± 12% and PRi in groups 2 and 3 were 75 ± 8.8 and 63 ± 15 respectively, which was significantly different ($p < 0.001$). Right ventricular function (RV MPI) in group 1 was 0.27 ± 0.13, in groups 2 and 3, it was 0.32 ± 0.12 and 0.34 ± 0.15, respectively, which was not significantly different ($p = 0.143$).
Conclusion: Increments in QRS duration of more than 120 ms in totally corrected TOF patients could be relevant to indicate increases in pulmonary artery regurgitation.

380: DEXTROCARDIA, SITUS INVERSUS AND CYANOTIC CONGENITAL HEART DISEASE WITH MULTIPLE CARDIAC DEFECTS IN A NIGERIAN INFANT: A CASE REPORT

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Background: The association of dextrocardia with situs inversus is reported to be 1:2 500 to 1:20 000; dextrocardia with situs solitus being less common. The incidence of congenital heart disease varies from 10% in situs inversus to up to 90% in situs solitus. We report an unusual case of dextrocardia, situs inversus, and cyanotic congenital heart disease with multiple heart defects.
Methods: We carried out a retrospective review of case notes.
Results: A seven-month-old Nigerian male infant presented at the children's out-patient clinic with symptoms of recurrent cough and bluish discolouration of the lips. On examination, he was found to be centrally cyanosed with grade 3 digital clubbing and a right-sided apical impulse. He had widely spaced nipples but no other dysmorphic features, and weighed 7.6 kg. His respiratory rate was 40 cycles per minute. He had normal volume pulses with a heart rate of 140 beats per minute, the first heart sound was normal while the second was single and soft with no murmur.

He was the product of term gestation delivered by spontaneous vertex to a 32-year-old woman. There was no history of exanthematous febrile illness, ingestion of herbal concoctions or exposure to irradiation in pregnancy. Yhere was no history of hypertension or heart disease in the family. His birth weight was 3.2 kg. There was no history of feeding difficulties but occasional respiratory distress.

Chest radiograph revealed dextrocardia with situs inversus. Abdominal ultrasonography confirmed a left-sided liver and gall bladder with a right-sided spleen. Echocardiogram revealed atrial situs inversus, a common atrio-ventricular valve with moderate regurgitation, non-restrictive ventricular septal defect, biventricular hypertrophy, an atretic pulmonary artery and a patent ductus arteriosus. This child has been referred for urgent open-heart surgery
Conclusion: This case shows that dextrocardia when associated with situs inversus can be associated with multiple congenital cardiac anomalies.

386: PATIENTS WITH UNBALANCED ATRIO-VENTRICULAR SEPTAL DEFECT AND SMALLER LEFT VENTRICLE: THE OUTCOME

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Background: Borderline unbalanced atrio-ventricular canal defect and borderline small left ventricle (uCAVC-small LV) is characterised by an atrio-ventricular valve index (AVVI) of 0.30–0.67 and left–right ventricle long-axis ratio (LV/RV) < 0.65 by echocardiography. The aim of this study was to compare long-term outcome of these patients after biventricular repair versus univentricular palliation.
Patients: Between 1992 and 2012, 18 patients (eight male, 10 female) with uCAVC-small LV underwent surgery; seven patients (group 1) had univentricular palliation due to associated atrio-ventricular valve (AV) malformations and/or large ventricular septal defect. In 11 patients (group2) biventricular repair was possible.
Methods: Differences at time of surgery and prospective follow up (FU) with repeated clinical and echocardiographic evaluation were performed. Differences in outcomes after biventricular repair versus univentricular palliation were evaluated.
Results: Group 1/group 2 at time of surgery (SURG): median age 8.9/2.7 months ($p = 0.01$). Two early deaths were present in: group 1 on postoperative day 306 (sepsis); group 2 on day 40 (cardiac failure). Sixteen patients underwent regular FU, group 1/group 2: median length 8.6/5.9 years ($p = 0.19$). Comparing progress according to patients' weight: all patients were under the 3rd percentile at SURG, not changing during FU in group 1 ($p = 0.2$), but reaching normal weight five years after operation in group 2 ($p = 0.042$). More than mild AV/mitral valve regurgitation in group 1/group 2 was present: in two (28.6%)/four (36.4%) patients ($p = 0.07$). Late re-operation for severe regurgitation was performed in two patients, with no difference between group 1 and group 2 ($p = 0.94$): AV valvuloplasty in one (14.3%) vs mitral valvuloplasty in one (9.3%), 2.8/4.6 years postoperatively. Mitral valve (group 2) showed normal long-term growth [median *z*-score at SURG –2/+0.26 at the last visit ($p = 0.02$)].
Conclusions: When possible, in patients with uCAVC-small LV biventricular repair should be done. During FU these patient generally do better and the mitral valve reaches normal values. On the other hand, mitral as well as AV valve regurgitation may represent a severe problem after both types of correction.

388: HOW DOES AGE AFFECT LEFT VENTRICULAR TWIST IN CHILDREN?

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Background: The recently introduced method, speckle tracking echocardiography, represents a simplified, objective and angle-independent modality for quantification of regional myocardial deformation. As previously published, there was no significant change in LV torsion with aging. There may however be some difference in LV rotation at the base and apex with aging.
Objective: The purpose of this study was to assess the relationship of LV rotation for torsion twist with aging in children.
Methods: Forty healthy children were recruited and divided into two groups of 20 pre-school children (age 2–6 years) and 20 school-age children (7–12 years). After obtaining conventional echocardiographic data, apical and basal short-axis rotations were assessed with speckle tracking echocardiography. LV rotations in the basal and apical short-axis planes were determined on six myocardial segments along the central axis.
Results: There was no significant change in apical and basal LV rotation with age between preschool and school-age children. However, there was a change between the two age groups with each basal and apical rotation. With basal and apical rotation, the values of pre-school children were higher than those of school-age children at the six anteroseptal, anterior, lateral, posterior, inferior and septal segments.
Conclusion: There was a trend of higher rotation values in pre-school children than in school-age children, with decreased rotation and torsion values with aging during childhood from two to 14 years old. Although there was no statistically significant age-related change in LV torsion from the rotation data, the decreasing trend with aging for rotation and torsion twist during childhood warrants further investigation.

393: A FEASIBILITY STUDY OF LEFT VENTRICULAR ROTATION AND TORSION BY TWO-DIMENSIONAL ECHO SPECKLE TRACKING DURING SEMI-SUPINE CYCLE EXERCISE IN CHILDREN

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Background: Rotation (R) of the LV apex and base produces torsion (T), an important component of the active phase of the cardiac cycle. Ventricular dysfunction leads to inadequate R and abnormal T. Changes in these aspects of ventricular function during exercise can be used to assess myocardial function. Echo speckle tracking can be used to measure LV rotation from which T can be calculated. We sought to determine the feasibility of measuring R and T in controls (CON) and paediatric transplant patients (PT) during incremental semi-supine cycle exercise (SSCE).
Methods: Fourteen CON (median age: 11.1 years) and five PT (median age: 14.8 years) exercised to volitional fatigue. 2D echo basal and apical short-axis views were obtained at rest, at each stage of SSCE, immediately and three minutes post-exercise. R and T were obtained by standard techniques. Each variable was measured three times and averaged. Coefficients of variation (CV) were calculated.
Results: Data acquisition was increasingly difficult with increasing exercise intensity. At peak exercise, it was possible to obtain data in only 5/14 CON and 2/5 PT; however, sub-maximal exercise data could be obtained in 11/14 CON and 5/5 PT and immediately post-exercise data in 9/14 CON and 5/5 PT. The CV was as high as 50%.
Conclusions: This preliminary study shows that measurement of R and T is feasible at rest and during sub-maximal exercise, but difficult to measure with increasing exercise intensity during SSCE. Failure to document an increase in R and T during exercise may reflect the technical difficulties of this method and individual measurement variability.

394: NON-INVASIVE ASSESSMENT OF VENTRICULAR-VASCULAR COUPLING AND HYDRAULIC EFFICIENCY IN POST-OPERATIVE CONGENITAL AND ACQUIRED HEART DISEASE

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Background: Ventricular-vascular coupling (V-VC), the ratio of LV elastance (E_{LVI}) to arterial elastance (E_{AI}), measures the optimum cardiovascular interaction. Hydraulic efficiency (HE), the ratio of mean to total power, is another measure of V-VC. We sought to determine E_{AI}:E_{LVI} and HE in healthy controls and patients with post-operative congenital and acquired heart disease and the relationship between these measurements.
Methods: Subjects consisted of 20 healthy controls (CON), 17 Marfan, 17 tetralogy of Fallot (TOF), 14 coarctation (CoA), and six transposition of the great arteries (TGA) patients. Systolic blood

pressure (SBP) was used to calculate LV end-systolic pressure (LVESP). LV end-systolic volume (LVESV) and stroke volume (SV) were calculated using standard M-mode dimensions and indexed for BSA. E_{AI} (LVESP/SV_i) and E_{LVI} (LVESP/$LVESV_i$) were calculated. HE was calculated from carotid pulse applanation tonometry tracings and Doppler flows.
Results: Age, BSA and SBP were similar between groups. E_{AI} was lower in CoA vs TOF patients ($p = 0.027$). E_{LVI} was higher in CoA vs TGA patients ($p = 0.048$). E_{AI}:E_{LVI} was lower in CoA vs Marfan ($I < 0.001$), TOF ($p = 0.001$) and TGA patients ($p = 0.005$). There was no correlation between HE and E_{AI}, E_{LVI} or E_{AI}:E_{LVI}.
Conclusion: V-VC appears to be optimal in CoA and less so in Marfan, TOF and TGA patients. These groups may have to work at suboptimal V-VC to maintain HE and this may contribute to the ventricular dysfunction seen in these groups.

395: LEAD REMOVAL IN YOUNG PATIENTS WITH A CONGENITAL HEART DISEASE IN VIEW OF LIFELONG PACING

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Aims: In children and young patients with or without a congenital heart disease, transvenous leads for pacemakers or implantable cardioverter defibrillators can cause later vascular obstruction or infection. Removal of non-functional leads is controversial as it bears the risk of vascular disrupture and embolisations. Early lead removal in our clinic was evaluated retrospectively.
Methods: Over the last six years in 22 patients with a mean age of 12.9 years (range: 3.6–29.5 years) removal of 30 transvenous leads (mean lead age: 5.1 years) was attempted. The main indications for removal were vascular obstruction, increased threshold, and lead dislocation. Commercially available retraction tools were used, if necessary. Twenty-seven leads (90%) were retrieved with clinical success, of which 23 (77%) were removed with complete procedural success. In three leads the lead tips were retained, while three leads could not be retrieved. No major complications occurred. Additional interventions such as recanalisation, balloon dilation, or stent implantation were performed as indicated. Procedure and X-ray times could be correlated to the implant age of the leads.
Conclusion: Using only mechanical techniques (no electro or laser sheaths), transvenous lead removal could be performed with a clinical success rate of 90%. In the case of vessel obstructions, lead replacement should be performed early, as the older the lead, the more prolonged and more hazardous the extraction procedure becomes. The use of new leads and precautionary implantation techniques may facilitate later lead removal.

396: TELEMONITORINGIN CHILDREN WITH A CONGENITAL HEART DISEASE AND ELECTRONIC DEVICES

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Aim: In children and young adults self-perception and self-responsibility is not fully developed. An automated cardiac monitoring system can assist to early diagnose clinical problems or to anticipate device failure in the seriously affected patients with congenital heart diseases (CHD).
Population: Out of 200 patients, 65 received a pacemaker (PM) or defibrillator (ICD) with the home monitoring (HM) option. Patients' age at implantation ranged from five weeks to 37.6 years (median 12.4 years). The individual follow-up time from the daily monitoring data was seven days to 7.3 years (mean 2.1 years).
Results: The evaluation period summarises up to 80 patient years with successful transmission on 72% of the days; 17% of all messages were categorised as 'emergency' with the need to immediately react to the incoming data. Consequences were system or lead revisions, electro-physiological studies, reprogramming of parameters, modifications in medication and sport, or to further observe. Transmitted intracardiac electrograms (IEGM) reflect the proper function of the system as well as the actual cardiac electric performance. In 14 patients with an ICD, tachycardia with the need to treat was found in seven patients. Five patients had 19 episodes with anti-tachycardic pacing (ATP).
Conclusion: The day-to-day transmission of data routinely and continuously monitored in every PM or ICD markedly improves safety and reliability of electronic device therapy in young patients. High transfer rates increase the probability of early event detection and offer the chance for early intervention. Despite some impact on our clinical workload and legal aspects regarding liability and organisation of procedural steps, this system improves therapy in our most critical patients.

398: NORWOOD PROCEDURE: A SUCCESS STORY FOR THE CHILD OR THE SURGEON, AND FOR HOW LONG?

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Introduction: The Norwood procedure is a series of three open-heart surgeries that gradually improve certain life-threatening forms of congenital heart disease. The first successful use of the procedure was reported by Norwood and colleagues in 1981. It is used most often to treat hypoplastic left heart syndrome, but variations of the procedure may also be used to treat other congenital heart diseases in which one or both of the lower chambers of the heart (ventricles) are defective. Each of the three surgeries is done at a different age, beginning from infancy and spanning through the toddler years. The first two surgeries (stage I and stage II) are used to temporarily relieve blood flow problems to and from the lungs. The third surgery (stage III) is used to further improve circulation. The Norwood procedure cannot cure the underlying heart defects.
Methods: This was a retrospective review of our unit's experience with more than 20 patients who underwent the Norwood procedure between 1 January 2010 and 30 June 2012. We also reviewed significant numbers of cases done abroad but regular follow ups done locally.
Results: Although the results of the modified Norwood procedure as palliation for the hypoplastic left heart syndrome have improved considerably, in-hospital mortality remains high (up to 46%) while one-year survival is only 1%.
Conclusion: Our study and supporting data from the literature have shown that the Norwood procedure, despite being life saving for infants less than week old with HLHS, has failed to give strong evidence of long-term survival. The poor surgical outcome also has social implications, which may necessitate a religious input as to the validity of such a procedure in light of the overall results.

399: THE IMPACT OF CONGENITAL HEART DISEASE ON OUTCOMES OF INFANTS WITH OESOPHAGEAL ATRESIA

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Background: The presence of associated congenital anomalies in children with oesophageal atresia (EA) with or without tracheo-oesophageal fistula (TEF) is well described, but few studies have examined the impact of congenital heart disease (CHD) on the

outcome of EA/TEF. Our aim was to evaluate the impact of CHD on timing of surgical repair and outcome of children with EA/TEF in the current era.
Methods: A retrospective review of patients with EA/TEF treated at two academic institutions from 1995 to 2011 was performed. The presence of cardiac defects, other anomalies, surgical intervention and outcome were recorded.
Results: Among 231 babies with EA, 117 (51%) had CHD. Neonates with EA/TEF and CHD had lower gestational age ($p = 0.04$), birth weight ($p = 0.001$), and were more likely to be syndromic ($p = 0.0002$) than patients without CHD. VSD and ASD were most common, followed by anomalies of the systemic/pulmonary veins, tetralogy of Fallot, aortic arch anomalies, atrio-ventricular valve abnormalities, and double-outlet right ventricle. There was no difference between EA/TEF alone and EA/TEF with CHD in the age of oesophageal surgery, surgical approach, days of mechanical ventilation, and the length of hospital stay, although CHD neonates had a higher incidence of pre-operative mechanical ventilation ($p = 0.006$). Overall mortality was 9%, 6/114(5%) in EA/TEF without CHD, 16/117(14%) in EA/TEF with CHD ($p = 0.04$). However, only 5/22 deaths were the direct result of CHD; the remainder were due to other anomalies or respiratory disease.
Conclusions: CHD did not influence surgical strategy or morbidity in this series, although mortality was higher in the presence of CHD. Our data indicate that CHD was not directly responsible for death, and mortality may therefore have been multifactorial. Newborns with EA/TEF should be evaluated for CHD, but with improvements in surgical and neonatal care the co-occurrence of EA/TEF and CHD does not preclude a good outcome in the majority of patients.

403: SILVER DRESSINGS FOR STERNOTOMY INCISION CARE IN PAEDIATRIC CARDIAC PATIENTS TO DECREASE SURGICAL SITE INFECTIONS

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Background: The consequences of surgical site infections (SSIs) can be significant and range from discharge delays to mediastinitis. Meticulous wound care is important to reduce SSIs. Our study team hypothesised that the use of silver-impregnated dressings on post-operative paediatric cardiac surgery patients could reduce SSIs.
Methods: Institutional review board approval was obtained to examine the infection preveniton qualities of silver-impregnated dressings on children post-sternotomy for congenital heart defects. Our population included infants to adolescents < 19 years of age and cardiac diagnoses ranging from RACHS-1 score 1 to 6. The final sample was 122 children (silver = 62; standard = 60). The sample size was chosen for a clinically important effect size of 0.5 in the detection of differences in SSI rates with > 75% power at a level of 0.05. Appropriate dressings were applied in the operating room, and children were followed for five days post-operatively or until discharge. The ASEPSIS wound score is a validated wound assessment tool; it was utilised to assess wound infections. The study team supervised all dressing changes during the study period. Thirty-day follow up for SSIs was performed. This randomised controlled trial evaluated silver-impregnated dressings versus our standard dressings on SSI rates.
Results: There were no SSIs in either study group. We found that there was no difference in type of dressing utilised, on SSI rates.
Conclusions: Our study does not support the use of silver-impregnated dressings in children after congenital heart surgery as a SSI prevention method.

404: MYOCARDITIS IN CHILDREN "CLINICAL PROFILE AND OUTCOME

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Aim: To study the demographic profile, clinical presentation and outcome of children diagnosed with acute myocarditis.
Methods: Records of 38 children diagnosed with acute myocarditis were analysed.
Results: There were 44.7% of patients below one year of age with male:female ratio being 1.4:1. Congestive heart failure, breathlessness, viral prodrome and fever were the common presenting symptoms. The initial clinical diagnosis was congenital heart disease, CCF, unexplained tachycardia and pneumonia in 78%. The duration of symptoms before diagnosis was more than five days in 85%. Tachycardia, gallop, bradycardia, hypotension, hyperdynamic precordium and cardiogenic shock were the presenting signs. Fulminant myocarditis was diagnosed in five patients (13.1%). Anaemia, hypocalcaemia, and altered liver enzymes were the biochemical abnormalities noted. Anaemia correlated with a poorer outcome on follow up ($p < 0.05$). High CPK:CPK-MB was seen in 31 (81.5%); 34 (84.7%) had an abnormal X-ray and 26 (68%) had abnormal ECGs. Two-dimensional echocardiogram revealed moderate to severe left ventricular dysfunction (LV) in 19/38 (50%). Furosemide and/or captopril were initial medications used with 11 (28.9%) needing an inotrope and eight (21%) requiring immune modulators. Hospitalisation ranged from < five days in 7.8% to > 15 days in 2%. CCF was difficult to treat in five patients (13.1%). Two patients died during the acute phase and four in the sub-acute phase of illness. At the three-month follow up, those with fulminant myocarditis had earlier normalisation of LV function than those with acute myocarditis ($p = 0.048$);. 26 children (68.4%) had complete recovery with six (15.7%) having a partial recovery.
Conclusion: Acute myocarditis often presents with non-cardiac symptoms, thus delaying clinical diagnosis. Fulminant myocarditis has better long-term outcomes than acute myocarditis.

405: RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE: AN URBAN STUDY

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Aims: To study the demographic**,** clinical and echocardiographic profile at presentation and a two-year follow up of children diagnosed with rheumatic heart disease (RHD)/acute rheumatic fever (ARF).
Methods: Records of 69 patients diagnosed with ARF/RHD were analysed. Psycho-social evaluation and compliance were assessed with a separate questionnaire
Results: Male:female ratio was 2.1:1. Mean age was 9.5 years with 2/69, being less than three years. Over-crowding was noted in 62.3 and 80% of patients belonged to the lower socio-economic class. Carditis (78.2%), polyarthritis (56.5%), chorea (7.2%), fever (68%) and arthralgia in 62.3% were the presenting symptoms. ASLO titre was positive in 82.6%, while leucocytosis, elevated ESR and positive CRP was seen in 49, 60.8 and 78%, respectively. Compliance with penicillin prophylaxis was noted in 86.5%. Non-availability of injection (24.6%), self-omission (13%) and switching to alternative medicine (11.6%) were the reasons cited by the defaulters. Initial 2D echocardiogram/Doppler revealed mitral affection in 92.7%, aortic valve in 1.4%, dual valve involvement in 8.6% and silent carditis in 8.6%. On follow-up echocardiogram, 36.2% showed improvement, 20.3% worsened, 43.5% remained unchanged with six patients requiring valve surgery. Compliance with penicillin prophylaxis was 90.3% in the improved group and 71.2% in those who worsened ($p < 0.05$). Parents were concerned about poor scholastic performance (32%), sub-optimal health and activity (78%), financial burden of therapy (33.3%), future marital and reproductive life concerns (100%), with 6% of young adults reporting employment issues.
Conclusion: Carditis and arthritis were the commonest presentation. Intermittent non- availability of drugs was the commonest reason for poor compliance. The disease and treatment hampers scholastic performance, increases economic burden and lowers the self-esteem of the child.

415: A LONGITUDINAL STUDY OF VENTRICULAR CONTRACTILE FUNCTION IN HYPOPLASTIC LEFT HEART SYNDROME PRIOR TO FONTAN

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Background: The long-term prognosis in hypoplastic left heart syndrome (HLHS) is limited by progressive ventricular dysfunction. The objective of this longitudinal study was to determine changes in HLHS ventricular function across staged palliative surgeries using speckle tracking echocardiography.
Methods: Twenty HLHS patients who survived to pre-Fontan surgery assessment were prospectively studied. Two-dimensional echocardiograms were performed through palliation at: (1) pre-Norwood (6 ± 7 days), (2) pre-BCPA (bidirectional cavo-pulmonary anastomosis, 5 ± 2 months), and (3) pre-Fontan (2.6 ± 0.6 years) stages. Speckle tracking echocardiography measured global and segmental four-chamber longitudinal and basal circumferential strain, strain rate (SR), post-systolic strain index [PSSi = (peak strain–peak systolic strain)/peak strain], rotation, myocardial dyssynchrony index (MDI = standard deviation of time to peak strain in 12 segments), and longitudinal:circumferential strain ratio. Differences across the three stages were analysed using one-way ANOVA for repeated measures with post hoc testing ($p < 0.05$).
Results: Both longitudinal and circumferential SR were decreased at pre-BCPA and pre-Fontan when compared to pre-Norwood ($p < 0.0001$). Rotation also declined after the pre-Norwood stage ($p = 0.02$). PSSi was greatest at pre-BCPA stage (longitudinal, $p = 0.0002$; circumferential, $p = 0.03$). Although global strain had no detectable change between stages, longitudinal:circumferential strain ratio decreased between pre-Norwood and pre-BCPA ($p = 0.01$). Interestingly, MDI was significantly greater at pre-Norwood compared to pre-Fontan ($p = 0.02$). Fractional area change was unchanged across the stages.
Conclusions: Ventricular ejection appears to be preserved in HLHS patients who survived to pre-Fontan assessment. However, at the pre-BCPA assessment, there was a significant detrimental change in ventricular contractility, coupled with an increase in PSSi, a potential marker of myocardial ischaemia. We also observed reductions in the longitudinal:circumferential strain ratio and ventricular rotation. Whether this is a single RV adaptive process to chronic afterload or evidence of subtle RV decline remains unclear. Except for PPSi, recovery in the ventricular functional parameters was not observed at pre-Fontan, despite volume unloading with BCPA.

416: PROTEIN-LOSING ENTEROPATHY AFTER FONTAN OPERATION: GASTROINTESTINAL EVALUATION OFFERS INSIGHT INTO THE PATHOPHYSIOLOGY

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Background: Protein-losing enteropathy (PLE) remains an enigmatic ailment seen after Fontan operation (FO). While the haemodynamic disturbances of elevated central venous pressure and diminished cardiac output inherent in the Fontan circulation are likely inciting factors, characterisation of the gastrointestinal (GI) tract may offer clues to a better understanding of the pathophysiology. Sulfated glycosaminoglycans (s-GAG) play an important role in maintaining integrity of the intestinal mucosa. Congenital deficiency of intestinal s-GAG, and acquired deficiency in disorders of glycosylation lead to PLE. Altered s-GAG expression is also noted in patients with kwashiorkor, a condition with features similar to PLE after FO.
Objective: To describe the GI histological findings in patients with PLE after FO with focus on intestinal s-GAG expression.
Methods: Twenty-eight combined upper GI endoscopy/colonoscopy studies were performed in 26 patients with PLE after FO. Gross histology was assessed. In a random select group of 10 patients, immunohistochemistry s-GAG staining was performed on tissue specimens from the duodenum and terminal ileum.
Results: Age at study was 15.8 ± 5.6 years; albumin was 2.8 ± 0.9 gm/dl. Of the 28 GI studies, tissue histology revealed inflammation (oesophagitis, gastritis, small bowel inflammation, cryptitis or colitis) in 15 (53%), lymphangiectasia in 12 (43%), and eosinophilia in four (14%). Seven of 10 patients demonstrated variable degrees of s-GAG deficiency, with more prominent depletion seen in the terminal ileum than in the duodenum.
Conclusions: Intestinal lymphangiectasia and inflammation are common in PLE after FO. Enteric mucosal s-GAG deficiency is evident and may be a commonly shared molecular pathway to PLE among various conditions. Our findings support the model that circulatory stressors in combination with inflammation trigger change at the enteric mucosa in predisposed individuals, which leads to enteric protein loss after FO. Effective treatment strategies should target modification of circulatory stressors, reduce inflammation, or replenish tissue s-GAG (i.e. heparin sulfate).

421: ARE WE IGNORING APICAL NON-COMPACTION OF BOTH VENTRICLES

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Background: Isolated left ventricular non-compaction (LVN) is reported extensively but apical non-compaction (ANC) of both ventricles and the inter-ventricular septum (IVS) is not reported much in literature.
A**im:** The aim of our study was to analyse and evolve echocardiographic (TTE) diagnostic criteria for apical non-compaction.
Methods and Results: From January 2011 to July 2012, 60 consecutive cases that fulfilled standard echocardiographic criteria for LVNC, and the additional criteria: (1) Swiss-cheese appearance of IVS, or looking like the delta of a river, and (2) non-compaction of the right ventricle (RV) on TTE, comprised the study material. The diagnosis of ANC was totally missed by the echocardiographer and either only specific lesions or Swiss cheese VSD were diagnosed. The diagnosis was made after review. Age ranged from three days to 35 years, with 36 males and 24 females. The associated lesions were present in all 60 cases; 52 patients had acyanotic heart disease (86.7 %) and eight had cyanotic heart disease (13.3 %). Sixteen of 60 cases had pump failure (26.7 %), seven had LV dysfunction, seven had RV dysfunction and two had biventricular dysfunction, 28 patients (46.7 %) had pulmonary hypertension, and two (3.3 %) had thrombus. Three cases of VSDs were post-operative residual shunts. ANC is known to happen in the chicken heart, but thus far there has been no evidence to suggest a similar mechanism in humans.
Conclusion: For the first time worldwide, we are presenting the largest series of ANC. ANC is probably ignored and is invariably associated with other serious congenital cardiac malformations, which worsen the pump failure.

424: IMPACT OF THE PERINATAL TRANSITION ON CARDIOVASCULAR FUNCTION AND CEREBRAL DOPPLER PROFILES IN HYPOPLASTIC LEFT HEART SYNDROME

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Background: Rapid changes in myocardial loading occur during the transition from foetal to postnatal circulation. In healthy neonates previous studies have shown that the LV output acutely doubles within hours of delivery but decreases at 24 hours to levels only slightly more than that of the foetus. Little data exists regarding the

perinatal transition in congenital heart disease. We sought to explore the impact of the perinatal transition on myocardial function and cerebral blood flow in hypoplastic left heart syndrome (HLHS).
Method: Pregnancies with foetal HLHS were prospectively enrolled (n = 8). Echocardiography was longitudinally performed at prenatal (38 ± 0.5 weeks) and postnatal (4 –12 hours, 24 hours, 48 hours, 3–5 days) time points. We assessed stroke volume (SV), heart rate (HR), cardiac output (CO), and middle cerebral artery pulsatility index (MCA-PI).
Results: In late gestation, mean CO in foetal HLHS was 416 ± 86 ml/kg/min and did not change at 4–12 hours after birth (414 ± 49 ml/kg/min). However, at 24 hours, 48 hours and 3–5 days after birth a significant increase in CO occurred (530 ± 103, 597 ± 142, 649 ± 111 ml/kg/min, respectively) due to increases in both SV and more so HR ($p < 0.05$). CO correlated positively with PaO_2 ($r = 0.39$, $p < 0.05$). During the same period, MCA-PI progressively increased from the foetal stage through all time points ($p < 0.05$).
Conclusion: In HLHS, in contrast to the normal perinatal transition, CO progressively increased from levels comparable to that in the foetus at 4–12 hours, to more than 150% by 3–5 days. This was likely secondary to decreasing pulmonary vascular resistance in the presence of a patent ductus arteriosus. Changes in pulmonary vascular resistance may also contribute to increasing MCA-PI. Ongoing investigations into the perinatal transition in HLHS with comparison to normal neonates will provide further insight into how the myocardium adapts in HLHS to postnatal demands and its influence on peripheral circulation.

426: BRAIN ABSCESS IN CYANOTIC CONGENITAL HEART DISEASE: A FIVE-YEAR REVIEW

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Background: Brain abscess in cyanotic congenital heart disease (CCHD) accounted for 5 to 10% of all kinds of brain abscess in children. The prolonged uncorrected lesions, chronic hypoxaemia and polycythaemia were the most common risk factors for brain abscess in CCHD.
Objective: To report the cases of brain abscess with CCHD in our institution.
Methods: A retrospective study was done of all patients admitted to our institution with the diagnosis of brain abscess and cyanotic congenital heart disease from July 2006 to July 2010.
Results: There were nine patients, ranging in age from three to 11 years, of whom six were males and three females. The clinical presentations were fever, vomiting, headache and seizure. Two patients developed hemiparesis and five had seizures. The duration of fever before admission was from 11 days to one month. Diagnosis was confirmed by CT scan. The most common location of abscess was the parietal lobe of the cerebral hemisphere. Multiple lesions were detected in six cases and solitary lesions in three cases. The types of CCHD were tetralogy of Fallot, complex cyanotic CHD with right isomerism, TGA and TAPVD. *Burr hole* aspiration was performed in five cases, pus culture in one case revealed *Acinetobacter calcoaceticus*, and four cultures were negative. Two patients died due to brain herniation. BT shunt and total correction was performed in the patients with tetralogy of Fallot.
Conclusion: Uncorrected cyanotic CCHD after two years of age, or delayed repair of CCHD in children can potentially lead to development of brain abscess, with poor outcome and prognosis.

435: THREE-DIMENSIONAL SPECKLE TRACKING ECHOCARDIOGRAPHY IN THE ASSESSMENT OF LEFT VENTRICULAR VOLUME AND FUNCTION IN NORMAL CHILDREN: A COMPARISON WITH CARDIAC MAGNETIC RESONANCE IMAGING

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Background: Three-dimensional speckle tracking echocardiography (3DSTE) is a potential clinical tool for rapid assessment of cardiac function and volume. Currently, CMRI is the gold standard for functional and volumetric assessment of children with congenital heart disease. 3DSTE may provide equivalent information on cardiac function and volume, which would remove the need for general anaesthesia, and offers a more cost effective and accessible method of imaging in children.
Methods: Fifty-five healthy children averaging nine years of age were investigated using CMRI and transthoracic echocardiography to assess LV function and volumes. Three-dimensional images were acquired using a Phillips IE33 echocardiographic machine and an X7-2 probe and stored for off-line analysis using Tomtec 4D LV analysis software. Parameters derived from speckle tracking analysis included global longitudinal strain (GLS), torsion, EF, EDV and ESV, which were correlated with CMRI-derived indices.
Results: LVEDV derived from 3DSTE correlated significantly with that derived from CMRI (63.4 ± 11.6 vs 72.2 ± 11.7, $r = 0.43$, $p < 0.001$). These volumes were significantly different with 3DSTE tending to underestimate LV volume. The LVESV derived from the two methods also showed a significant correlation (30.7 ± 7.6 vs 25.5 ± 6.2, $r = 0.5$, $p < 0.001$). There was a poor correlation with stroke volume (32.8 ± 6 vs 46.6 ± 7.8, $r = 0.215$, $p = 0.12$). EF derived from 3DSTE showed a significant correlation with that derived from CMRI (51.8 ± 6.7 vs 64.7 ± 5.1, $r = 0.29$, $p < 0.05$). GLS showed a better correlation with CMRI EF than 3DSTE EF ($r = –0.38$, $p < 0.005$). Torsion showed no correlation with CMRI derived parameters of function or volume.
Conclusion: 3DSTE shows significant correlation with CMRI-derived parameters of cardiac function and volume. 3DSTE tends to underestimate ventricular volume and function in comparison to CMRI-derived values. This technique has potential to be developed as a rapid assessment tool in children with congenital heart disease in the future.

436: EARLY REPAIR OF TETRALOGY OF FALLOT DOES NOT PROTECT AGAINST LATE AORTIC ROOT DILATATION

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Introduction: Tetralogy of Fallot (TOF) is associated with late aortic root dilatation following primary repair. The pathophysiology of this is thought to be related to volume loading of aortic outflow prior to repair and/or due to an intrinsic aortic wall abnormality. We sought to determine whether early repair of TOF would protect against late aortic root dilatation.
Method: Patients with repaired TOF who had undergone CMRI were retrospectively reviewed. The size of the aorta at the level of the sinuses was compared with that predicted for age and BSA. Details with regards to timing of surgery and initial palliation were collected.
Results: A total of 93 patients were included. Mean age was 23 years (1–74). The mean age at repair was 30 months (1–792). Measurements at the level of the aortic sinuses were 33.9 ± 7 mm (n = 60) in the group with infant repair vs 35.8 ± 5 mm in the group with later repair (n = 33) ($p = 0.17$). Measurements were also compared in patients who initially required palliative procedures vs those who did not [34.1 ± 7 mm (n = 20) vs 34.6 ± 6 mm (n = 71), $p = 0.77$]. Patients who had primary repair at less than six months did not show a significant difference from those performed later [34.8 mm ± 6 (n

= 69) vs 33.8 ± 6 (n = 24), p = 0.52]. The measurements of the aortic sinuses in all patients was compared with the predicted size for age and BSA, this was 34.6 ± 6 mm vs 30.9 ± 2.9 mm (p < 0.001.)
Conclusion: This study confirms progressive aortic root dialatation in patients with TOF. The mechanism of this appears to be complex, as early repair, type of palliation or primary repair does not provide protection against late aortic dilatation. Routine follow up of post-operative patients with TOF for aortic root dilatation and the role of preventive therapy needs further evaluation.

438: INCREASED REGIONAL DEFORMATION OF THE LEFT VENTRICLE IN CHILDREN WITH A RAISED BODY MASS INDEX: IMPLICATIONS FOR FUTURE CARDIOVASCULAR HEALTH

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Background: The prevalence of obesity continues to increase in the developing world. The effects of obesity on the cardiovascular system include changes in systolic and diastolic function. More recently obesity has been linked with impairment of longitudinal myocardial deformation properties in children. We sought to determine the effect of a raised body mass index (BMI) on cardiac deformation in a group of children taking part in the population-based Southampton Women's Survey.
Methods: A sample of 68 children aged nine years old had assessments of longitudinal myocardial deformation in the basal septal segment of the left ventricle using two-dimensional speckle tracking echocardiography. Parameters of after-load and pre-load, which may influence deformation, were determined from cardiac magnetic resonance imaging. BMI was determined from the child's height and weight at the time of the echocardiogram.
Results: A higher BMI was associated with an increase in longitudinal myocardial deformation or strain in the basal septal segment of the left ventricle (r = 0.41, p < 0.001), but was not related to contractility or strain rate in this part of the heart (r = 0.04, p = 0.75). The end-diastolic volume of the left ventricle increased with increasing BMI (r = 0.33, p = 0.011).
Conclusion: Regional deformation in the left ventricle increases significantly with increasing BMI, while normal contractility is maintained. This may be explained by the increased pre-load of the left ventricle due to increased somatic growth. The long-term implications of this altered physiology need on-going follow up.

439: IMPAIRED RIGHT VENTRICULAR CONTRACTILE RESERVE LATE AFTER SURGICAL CLOSURE OF ISOLATED ATRIAL SEPTAL DEFECT

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Background: Impaired aerobic exercise capacity and abnormally elevated right ventricular systolic pressure during exercise have previously been demonstrated in asymptomatic adolescents after surgical closure of isolated atrial septal defect early in life. We studied right ventricular contractile reserve during incremental exercise in this patient group. The study hypothesis was that differences in aerobic exercise capacity and pulmonary pressure response to exercise might be combined with differences in right ventricular systolic function during exercise.
Methods: Seventeen asymptomatic patients (age 15–23 years, 12 females, median age at defect closure 53 months) and 22 age-matched healthy control subjects were studied by echocardiography at rest and during recumbent bicycle exercise until a target heart rate of 160 bpm. M-mode images and colour-coded tissue Doppler recordings from apical four-chamber view were analysed offline.
Results: Patients had lower tricuspid annular peak systolic excursion (TAPSE) (14.2 ± 3.1 mm) at rest compared to controls (22.3 ± 2.9, p < 0.001). Correspondingly, the maximal TAPSE during exercise was reduced in the patient group (20.5 ± 4.5 vs 31.4 ± 4.1, p < 0.001). Peak systolic tricuspid annular velocity (S') was significantly lower in the patient group both at rest (patients 6.8 ± 1.8 cm/s, controls 9.7 ± 1.6 cm/s, p < 0.001) and as the highest measured S' during exercise (11.7 ± 2.8 cm/s vs 15.3 ± 2.7 cm/s, p < 0.001). Isovolumetric right ventricular acceleration (IVA), measurable in the tricuspid annulus of n = 13/22, was reduced in the patient group at pre-exercise (1.1 ± 0.5 vs 1.8 ± 0.6c m/s^2, p < 0.001, median heart rate 88/91.5), but tended towards equalisation for the highest measured IVA during exercise (3.5 ± 1.4 vs 4.2 ± 1.2 cm/s^2, p = 0.145, median heart rate 159.5/149.5 at peak IVA).
Conclusions: Asymptomatic adolescent patients with surgically closed isolated atrial septal defect have impaired right ventricular contractile reserve, most markedly demonstrated in reduced longitudinal shortening.

440: QTC PROLONGATION PRIOR TO ANGIOGRAPHY PREDICTS POOR OUTCOME AND ASSOCIATES SIGNIFICANTLY WITH LOWER LEFT VENTRICULAR EJECTION FRACTIONS AND HIGHER LEFT VENTRICULAR END-DIASTOLIC PRESSURES

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Background: QT prolongation on the surface ECG is associated with sudden cardiac death. The cause of QT prolongation in ischaemic heart disease (IHD) patients remains unknown, but may be due to a complex interplay between genetic factors and impaired systolic and/or diastolic function through, as yet, unexplained mechanisms. It was hypothesised that QT prolongation before elective coronary angiography is associated with an increased mortality at six months.
Methods: Complete records of patients (n = 321) who underwent coronary angiography were examined for QT interval corrected for heart rate (QTc) (Bazett's formula), left ventricular ejection fraction (LVEF), left ventricular end-diastolic pressure (LVEDP) and correlated with triple-vessel coronary artery disease (TVCAD) and other known IHD risk factors (hypercholesterolaemia, diabetes mellitus, smoking, hypertension or a family history of IHD). Patients were designated LQTc when they had prolonged QTc intervals, or NQTc when the QTc interval was normal. Patients with atrial fibrillation, bundle branch blocks, no ECG in the 24 hours before angiography, or a creatinine level > 200 µmol/l were excluded. Survival was determined telephonically at six months.
Results: Twenty-eight per cent of the total population had a LQTc. During follow up 15 patients (4.7%) died suddenly, 73% of whom had a LQTc. LQTc was significantly associated with mortality (LQTc: 12% vs NQTc: 1.7%; p < 0.01), and with lower, but normal, LVEF (LQTc: 52.9 ± 15.4% vs NQTc: 61.6 ± 13.6%; p < 0.01), higher LVEDP at LVEF > 45% (LQTc: 19.2 ± 9.0 mmHg vs NQTc: 15.95 ± 7.5 mmHg; p < 0.05), hypercholesterolaemia and a negative family history of IHD.
Conclusion: In patients with sinus rhythm and normal QRS width, QTc prolongation before coronary angiography predicts increased mortality at six months. QTc also strongly associates with left ventricular systolic and diastolic dysfunction, hypercholesterolaemia and a negative family history of IHD.

441: EXERCISE CAPACITY AND QUALITY OF LIFE IN ADOLESCENTS AFTER SURGICAL CLOSURE OF ATRIAL OR VENTRICULAR SEPTAL DEFECT DURING CHILDHOOD

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Background: Patients with congenital heart disease generally report good quality of life but they also show reduced exercise capacity of varying degrees depending on the type of congenital heart defect. The study hypothesis was that aerobic exercise capacity would impact on self-reported quality of life (QoL) in adolescents after surgical closure of isolated atrial (ASD) or ventricular septal defect (VSD) during childhood.
Methods: In 32 asymptomatic patients (18 ASD, 14 VSD, median age 18, 13–25 years, 20 females, median age at surgical closure 57, 1–229 months) and 103 healthy control individuals (61 females, median age 17.5, 12–24 years) we assessed maximum oxygen uptake (VO_2 peak). Assessment of quality of life was achieved with either the PedsQL™ questionnaire (< 18 years) or the SF-36™ questionnaire (> 18 years) with 100% response rate in both groups. All test scales ranged from 0 to 100.
Results: The VO_2 peak *z*-score for the patient group (n = 31) was –1.80 ± 1.54 and for the control group (n = 103) 0.27 ± 1.64 (p < 0.001). The results from quality-of-life assessment and group comparisons were calculated. By linear regression analysis we found no statistically significant relationship between the group-wise *z*-score of VO_2 peak and any of the subscale results or total score results from the quality-of-life assessment.
Conclusions: Adolescents operated on for isolated heart septal defects during childhood reported normal quality of life compared to the healthy population. Patients younger than 18 years of age reported better quality of life than estimated by their guardians. Our patients had lower aerobic exercise capacity compared to normal controls, but we found no relationship between VO_2 peak and any of the reported subscale scores or total scores of quality of life for either the patient or control groups.

452: ARE THERE CLINICALLY IMPORTANT RACIAL DIFFERENCES IN ATRIAL SEPTAL DEFECT MORPHOLOGY?

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Background: Sinus venosus defects are considered uncommon, representing only 5–10% of interatrial communications, with the inferior subtype being particularly rare and more difficult to diagnose. Suspecting and correctly identifying this unusual anatomy facilitates appropriate repair.
Methods: A retrospective review was undertaken of patients who underwent repair of interatrial communications at our Central American centre between October 2006 and June 2012 to determine the anatomical subtype. Partial atrioventricular canal defects were excluded.
Results: Fifty-four patients (53 children and one young adult, age range 14 months to 23 years) underwent surgery for interatrial communication. Sinus venosus defects with partial anomalous pulmonary venous connection were encountered in 10 patients (19%, p = 0.025), with half of these (five patients, 9.2% of the total operated) being inferior subtype. The remaining 44 patients (81%) had secundum atrial septal defects, six of whom (11%) had an additional diagnosis of pulmonary stenosis requiring valvotomy, and one of whom had a small patent ductus arteriosus. One patient (1.9%) died as a result of a ruptured oxygenator during bypass; the remaining patients had an uneventful postoperative course, spending one night in intensive care and being discharged at a median of two days postoperatively. All surviving patients had postoperative echocardiograms revealing no residual shunt and no obstruction to systemic or pulmonary venous drainage.
Conclusions: Our Central American patient population demonstrates a higher than expected proportion of sinus venosus defects and in particular of the inferior subtype. Data from major centres in North America and Western Europe may not be representative of the majority of the world's children with congenital heart defects, and if the effect is truly racial (i.e. genetic) rather than environmental these findings may also apply to immigrant populations.

455: PRENATAL DIAGNOSIS IMPROVED THE POSTNATAL CARDIAC FUNCTION IN POPULATION BASED COHORT OF INFANTS WITH HYPOPLASTIC LEFT HEART SYNDROME

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Background: Prenatal diagnosis of hypoplastic left heart syndrome (HLHS) enables planning of perinatal care and is known to be associated with more stabile pre-operative haemodynamics. The aim was to determine whether prenatal diagnosis of HLHS has an impact on postnatal myocardial function.
Methods: We reviewed a consecutively encountered cohort of 66 HLHS infants born between years 2003 and 2010 in Finland. Postnatal global and segmental right ventricular fractional area change (FAC), strain rate (SR) and myocardial velocity (V) were analysed from apical four-chamber views using the Velocity-Vector-Imaging technique (Syngo USWP 3.0, Siemens). Intra- and inter-observer correlations were good (r > 0.7, p < 0.05). Pre-operative haemodynamic status and end-organ damage measurements were the lowest arterial pH, highest lactate, alanine aminotransferase and creatinine. Early mortality was studied until 30 days after the Norwood procedure.
Results: Twenty-five infants (38%) had a prenatal diagnosis. Prenatally diagnosed infants had better cardiac function: FAC 27.9 ± 7.4 vs 21.1 ± 6.3%, p = 0.0004; SR 1.1 ± 0.6/1.3 ± 1.0 vs 0.7 ± 0.2/0.7 ± 0.3 1/s, p = 0.004/0.003; V 1.6 ± 0.6/2.0 ± 1.1 vs 1.3 ± 0.4/1.4 ± 0.4 cm/s, p = 0.0035/0.0009, respectively. In segmental analysis, the difference was global. Mechanical dyssynchrony was similar in both groups (p > 0.3). Infants diagnosed prenatally had less acidosis (pH 7.30 vs 7.25, p = 0.005) and end-organ dysfunction (alanine aminotransferase 33 ± 38 vs 139 ± 174 U/l, p = 0.0001; creatinine 78 ± 18 vs 81 ± 44 mmol/l, p = 0.05). No deaths occurred among the prenatally diagnosed infants but four deaths were recorded among postnatally diagnosed infants (p = 0.15).
Conclusions: A prenatal diagnosis of HLHS is associated with improved postnatal right ventricular function, reduced metabolic acidosis and end-organ dysfunction. Prenatal diagnosis is important for optimal prognosis of these infants.

461: PREVALENCE OF A POSITIVE SCREENING SCORE FOR ATTENTION DEFICIT HYPERACTIVITY DISORDER IN CHILDREN AFTER EARLY REPAIR OF CONGENITAL HEART LESIONS

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Background: Adverse neurodevelopmental outcomes, such as attention deficit hyperactivity disorder (ADHD), are recognised as important contributors to chronic morbidity in paediatric cardiac surgery populations. The SNAP-IV is a validated, parent-completed screening test used to identify children at risk of ADHD. The objective of this study was to determine whether children who underwent congenital cardiac repair with open-heart surgery at less than one year of age are more likely than healthy controls to have a positive screening score suggestive of ADHD.
Methods: Eligible cases were identified from the IWK Paediatric Cardiology database and were included if they were aged seven to 15 years and underwent open-heart surgery at less than one year of age. Patients were excluded if they had a known genetic disorder, multiple congenital abnormalities, or a head injury. Age-matched healthy controls were recruited from volunteers. Parents of consenting participants completed a SNAP-IV questionnaire and a study-specific demographic questionnaire. Case subject charts were reviewed for baseline characteristics and potential risk factors. Frequencies of those meeting the threshold for suspicion of ADHD were compared using Fisher's exact test. Regression analysis was used to identify potential predictors of higher screening scores.
Results: Questionnaires were completed by 57 of 170 eligible case subjects (response rate of 34%). Responders did not differ from non-responders in baseline characteristics. Of case subjects, 17/57 (30%) had a positive ADHD screening score in at least one domain, compared with 3/60 (5%) of controls ($p < 0.001$). Cases and controls differed significantly for average combined SNAP-IV scores ($p < 0.001$) and for both hyperactivity and inattention component scores ($p < 0.001$).There was no correlation between operative factors and SNAP-IV scores. No significant predictors of a higher score were identified.
Conclusion: Children who have open-heart surgery at less than one year of age are more likely than healthy controls to have a positive screening score for ADHD.

466: RECURRENT GIANT LEFT VENTRICULAR ANEURYSM OF TUBERCULOUS AETIOLOGY IN A CHILD

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Case report: A 10-year-old girl was admitted with a six-month history of multiple joint pain and decline in overall health status. Pericardial effusion was observed and pericardiocentesis performed, which failed to yield pericardial fluid. An echocardiogram 20 days later was suggestive of antero-apical aneurysm. Left ventriculography confirmed dyskinetic saccular formation in the antero-apical region and surgical intervention was recommended. Aneurysmectomy was performed using normothermic cardiopulmonary bypass on a beating heart. The patient's intra- and postoperative courses were uneventful, and she was discharged 18 days after surgery. Pathological examination revealed chronic myocardial inflammation in the reparative phase. Laboratory tests performed to shed light on the polyserositis condition were inconclusive. The aetiology of the aneurysm was not clearly elucidated.

Eight months after surgery the girl was brought back with dyspnoea. A chest radiograph revealed right-sided pleural effusion. Cytological examination of pleural fluid revealed lymphocyte predominance. The Mantoux test was strongly positive. The patient was started on antituberculous therapy. Fourteen months after surgery the girl was re-admitted with atelectasis of the left upper lobe. Bronchoscopic examination showed 50% narrowing of the left main bronchus due to extrinsic compression. Left ventriculography disclosed an antero-apical giant aneurysm. Re-operation was performed. After an uneventful recovery, the patient was discharged.

Pathological examination revealed cardiomyocyte hypertrophy and replacement myocardial fibrosis, confirming the diagnosis of true left ventricular aneurysm. Given the overall clinical picture, tuberculosis was considered as the likely cause of myocardial involvement. A review of the pathological specimens from the first surgery showed: inflammatory process with micro-abscesses, granulomas with central caseous necrosis surrounded by epithelioid cells in a palisade arrangement, and multinucleated giant cells, corroborating our clinical reasoning. Three aspects make this report unique: a rare diagnosis of cardiac aneurysm caused by tuberculosis, especially in a child; its recurrence, which is even rarer; and successful surgical treatment.

469: ATHEROSCLEROSIS RISK AND CAROTID INTIMA–MEDIA THICKNESS AFTER KAWASAKI DISEASE IN MEXICAN CHILDREN

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Background: Kawasaki disease (KD) is an acute febrile illness characterised by systemic vasculitis of unknown aetiology. Recent studies have shown that even after resolution of the disease, endothelial dysfunction persists and may progress to atherosclerosis. Carotid intima–media thickness (cIMT) is a well-established indicator for atherosclerosis in both paediatric and adult patients.
Objectives: To assess whether patients after Kawasaki disease (KD) have increased risk factors and abnormalities suggestive of early atherosclerosis by measuring the cIMT compared with healthy control subjects.
Methods: Fifty-seven patients with KD aged 9.02 ± 3.98 years (2–21 years after acute illness) and 83 age-matched healthy control subjects were examined for medical and dietary history, serum markers of atherosclerotic risk and inflammation and carotid intimal–medial thickness (CIMT) with vascular ultrasound scanning.
Results: Patients and control subjects were similar in age, gender, family and dietary history, body mass index and blood pressure. We found no difference in the levels of triglycerides and glucose. And the levels of total cholesterol (162 ± 39.2 vs 150 ± 37.4), low-density lipoprotein cholesterol (102.57 ± 32.3 vs 89.6 ± 33.5), and high-density lipoprotein cholesterol (47.38 ± 17.65 vs 39.5 ± 17.54) were slightly higher, with no statistical significance. The cIMT was slightly higher in the KD group (0.48 ± 0.1 vs 0.45 ± 0.15) We found higher levels in the lipid profile and in the cIMT in children with or with regression of coronary aneurysms compared with children without coronary aneurysms.
Conclusions: There was no clear evidence of increased atherosclerosis in Mexican children with KD, but there was evidence of an altered lipid profile and cIMT in patients with KD with coronary lesions compared with children with KD without coronary lesions. This warrants further study

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477: FAMILIAL ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA: MAGNETIC RESONANCE IMAGING RETROSPECTIVE STUDY IN CHILDREN

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Purpose: To evaluate cardiac magnetic resonance imaging (MRI) findings of familial arrhythmogenic right ventricular dysplasia (ARVD) in a paediatric population and to correlate MRI data with other 2010 ARVD task force criteria.
Methods: This wasw a retrospective study (January 2001 to June 2012) of 70 MRIs in 36 children (one month to 20 years old, mean 10.7 years) from 22 families with proven first-degree relatives with ARVD. Mean number of MRIs by patient was 1.9; 21 patients had two or more MRIs for a mean follow up of 2.3 years.
Results: MRI examinations were normal in 50, non-diagnostic in two and abnormal in 18 (11 patients). Anomalies consisted of abnormal cardiac contractility: dyskinesia (n = 5), akinesia (n = 7) and hypokinesia (n = 6), with only one patient with RV dilatation. Abnormal contractility was noted in the RV free wall: apex (n = 18), inlet (n = 2), outlet (n = 0). There was no RV fatty infiltration and no LV anomaly. Mean age of the positive cases was 13.6 years. Only three patients were symptomatic (cardiac arrest, syncope, ventricular tachycardia). Of the other 2010 ARVD task force criteria, two patients also had epsilon waves in the right precordial leads on ECG. Six patients underwent implantable defibrillators, with no appropriate shocks, and one with inappropriate shocks.
Conclusion: According to the 2010 ARVD task force, isolated RV wall contractility abnormalities, in addition to familial context, lead to a diagnosis of ARVD. In our small series, contractility abnormalities preceded global dilatation and alteration of function. Since most positive MRIs were found in adolescents, screening seems optimal at that age. Larger cohorts are needed to confirm our results.
Clinical application: Regional RV free wall dyskinesia/akinesia adjacent to the apex is the first manifestation of familial ARVD and becomes conspicuous in adolescence.

478: PREVALENCE OF PULMONARY HYPERTENSION IN CHILDREN WITH ADENOID OR ADENOTONSILLAR HYPERTROPHY AT THE KENYATTA NATIONAL HOSPITAL

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Background: Adenotonsillar hypertrophy is a common condition in childhood, whose serious complications of pulmonary hypertension and cor-pulmonale are common and devastating but local prevalence is unknown.
Methods: This was a cross-sectional descriptive study in children aged 0–12 years attending ENT clinics and general paediatric wards at KNH, with clinician-diagnosed adenoid hypertrophy confirmed on lateral neck radiography. Eligible patients were consecutively recruited into the study between September and November 2011. The patients were evaluated for symptoms, physical findings (adenoid, tonsil and airway sizes on lateral neck radiography) and Doppler echocardiographic assessment of systolic pulmonary artery pressure (sPAP). Pulmonary hypertension was defined as mean pulmonary arterial pressure of ≥ 25 mmHg estimated by the Chemla equation (0.61 sPAP + 2 mmHg).
Results: The prevalence of pulmonary hypertension in children with adenoid or adenotonsillar hypertrophy at KNH was 21.1% (95% CI: 14.3–29.4%). Independent factors associated with pulmonary hypertension included daily hyperactivity (OR = 0.22, 95% CI: 0.06–0.87, p = 0.03), oxygen saturation (OR = 0.72, 95% CI: 0.54–0.97, p = 0.03) and palpable P2 (OR = 9.84, 95% CI: 3.2–55.4, p = 0.01). Daily mouth breathing singly or in combination with restless sleep on history showed the highest sensitivity (88.5%) and negative predictive value (86.4%) for pulmonary hypertension in these children.
Conclusion: Clinical screening and echocardiography evaluation is vital in children with adenoid or adenotonsillar hypertrophy for early identification of pulmonary hypertension.

479: DOCUMENTED CORONARY ARTERY DILATATION DURING ACUTE VIRAL MYOCARDITIS

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Introduction: Detecting coronary artery (CA) dilatation is essential in the diagnosis and follow up of Kawasaki disease (KD). Myocarditis however, is almost always present in acute KD.
Purpose: To investigate whether myocarditis causes CA dilatation, we sought to use viral myocarditis as a clinical model.
Methods: A retrospective series of children with acute viral myocarditis were reviewed to confirm the diagnosis. CA diameters of the proximal right and the left CA were measured at onset and during the first two years of follow up. CA z-score was calculated based on our published equations. CA dilatation was defined as a z-score > 2.5. Occult CA dilatation was defined as a z-score variation of > two standard deviation points along the follow up in those with z-score always < 2.5. All other cases were labelled without CA involvement.
Results: There were 11 girls and three boys between 2000 and 2006 who met the selection criteria. KD was not in the differential diagnosis of any case. Age was 1.67 ± 3.22 years at diagnosis (range 0.02–9.45 years), with a follow-up duration of 16.2 ± 16.4 months. Microbial laboratory tests/cultures confirmed the diagnosis in 11 patients (78.5%), whereas the history of familial/personal acute infectious illness was present in the remaining. Cardiac enzymes were elevated in nine patients, normal in two and not available in three. CA involvement was detectible in 9/14 (64.3%) cases; dilatation in three (21%) and occult dilatation in six (42.9%). Peak CA z-score was at the onset of the disease in 7/9. Maximum CA z-score was 1.56 ± 0.8 vs 0.42 ± 0.9 for cases with or without CA involvement respectively; p = 0.036.
Conclusion: CA dilatation is not uncommon in acute myocarditis. Our findings represent a potential challenge to the diagnostic significance of the clinical criteria of KD especially when 'supported' by the finding of a dilated CA.

480: NATRIURETIC PEPTIDE RELEASE IN ACUTE KAWASAKI DISEASE PREDICTS GAMMAGLOBULINS RESISTANCE AND CORONARY ARTERY INVOLVEMENT

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Background: We recently reported on the diagnostic values of B-type natriuretic peptide (NT-proBNP) in the diagnosing Kawasaki disease (KD), with a diagnostic odds ratio of 26.7:1 (95% CI: 8.6–82.5) compared to febrile control children. There are also independent reports indicating that hyponatraemia is associated with severe KD and suggesting an inappropriate secretion of the antidiuretic hormone (ISADH).
Objectives: We hypothesised that increased serum NT-proBNP correlates with the severity of KD in terms of resistance to gammaglobulins (IVIG) and risk of coronary artery (CA) involvement.
Methods: Serum NT-proBNP concentrations were measured upon suspicion of KD (n = 74; 3.79 ± 2.92 years; diagnosed at 6.58 ± 2.82 days of fever), and correlated with serum Na and urine specific gravity. Data related to cardiac involvement were also analysed.
Results: NT-proBNP level was inversely proportional to serum sodium concentration (r = 0.39, p < 0.001), whereas urine specific gravity correlated proportionally with lower serum Na concentration (r = 0.18, p = 0.12). Since ISADH would increase urine specific gravity and lower serum Na concentrations, our observation refutes the previously suggested ISADH theory. IVIG resistance was higher

in cases with elevated NT-proBNP (18–20.5 vs 3.5–8%; $p = 0.01$–0.12), with a trend towards an increased incidence of CA dilatation (15–16.4 vs 5.3–5.8%; $p = 0.08$). After identifying the upper quartile related to serum NT-proBNP level, 8/10 subjects had clinical and echocardiographic findings suggestive of myocardial dysfunction.
Conclusion: NT-proBNP is most likely the cause of hyponatraemia during acute KD. It is associated with higher resistance to IVIG, and higher incidence of CA lesions. The use of NT-proBNP may become a cardinal biochemical marker in predictive scoring for IVIG resistance, coronary outcome, and the extent of myocardial involvement.

481: SURGICAL CONGENITAL HEART DISEASE CATEGORIES AND GENDER: DIFFERENT INCIDENCE, BUT SIMILAR LONG-TERM SURVIVAL

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Background: Gender differences exist in the incidence of different types of congenital heart disease (CHD), but there are few data on differences between males and females in long-term survival after corrective surgery.
Methods: Between 1971 and 2010, the 6 769 patients aged 0–16 years who underwent corrective CHD surgery at Oslo University Hospital, Rikshospitalet (80% of all CHD surgery in Norway), were prospectively enrolled in the study. Primary CHD diagnoses (ICD –9 or –10) were categorised according to a consensus-based severity hierarchy. Date of first operation was registered. Complete all-cause mortality data were recorded until 1 January 2012.
Results: Of the total number of patients, 3 256 (48.1%) were female. Median age at first operation was 0.8 years (mean 2.8, SD 3.8). The incidence of atrial septum defect and patent ductus arteriosus was highest in females (61.7 and 62.5%, respectively, $p < 0.001$). The incidence of transposition of the great arteries (66.3% males), tetralogy of Fallot (58.4%), double-outlet right ventricle (65.2%), coarctation of the aorta (60.0%), interrupted aortic arch (56.6%) and single-valve surgery (64.5%) was highest in males ($p < 0.001$). Incidence of pulmonary atresia, truncus arteriosus, univentricular hearts, ventricular septum defect and atrioventricular septum defect was similar in males and females ($p > 0.05$). During up to 40 years of follow up after surgery, female patients with coarctation of the aorta had a slightly higher mortality rate than males (15.7 vs 10.1%, $p = 0.05$). Survival in the other diagnostic groups was similar.
Conclusion: Despite substantial differences between males and females in the incidence of different CHD types, survival after initial corrective surgery within each diagnostic group was remarkably similar.

483: SURGICALLY TREATED PULMONARY STENOSIS: 50 YEARS OF FOLLOW UP

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Background: The natural history of surgically relieved pulmonary artery stenosis is not well described, since balloon valvuloplasty has replaced surgery. We hypothesised that the incidence of morbidity and mortality increase with increasing time and that more than 30% of patients have been re-operated due to pulmonary regurgitation before the age of 40 years.
Methods: A retrospective follow-up study was done of the population treated surgically for pulmonary artery stenosis at Aarhus University Hospital from 1 January 1957 to 31 December 2000. Patients were identified by their personal identification number; their hospital records were reviewed and present status noted.
Results: Of 87 patients, six were excluded due to missing information. The age at primary surgery was less than 15 years in more than 70% ($n = 58$) of the study population of 81 patients. Valvular stenosis was the most common pathology, 84% ($n = 68$), the rest had an infundibular stenosis. Three patients died (age 0 and 40 years, and one unknown) resulting in a mortality of 2.4%. The mean age for all patients at follow up was 32 years (range 4–79 years). At a maximum follow up of 54 years and a mean follow up of 25 years, 18 (24%) required at least one re-intervention. Pulmonary valve replacement due to pulmonary regurgitation was the most common re-intervention (60%). Out of 19 patients older than 40 years and 15 patients between 30 and 40 years of age, 47 and 28% respectively, have had a re-operation.
Conclusion: With a mean long-term follow up of 25 years (maximum 54 years), re-intervention was necessary in 24% and almost half of the patients reaching the age of 40 years had a re-operation. This retrospective study shows that there is a call for prolonged follow up and re-intervention in patients operated for simple pulmonary stenosis.

484: CONGENITAL HEART LESIONS ASSOCIATED WITH IMPORFORATED ANUS

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Objectives: We aimed in our study to determine the incidence and type of associated congenital cardiac anomaly (CCA) in newborns with imperforated anus (IA), the outcome of anorectal surgical reconstruction in our centre, and to determine the need for performing echocardiography in all patients with IA.
Methods: The pre-operative echocardiography reports of all cases born with imperforate anus and managed at King Abdulaziz University Hospital, Jeddah, Saudi Arabia over a period of 11 years (January 2000 to December 2010) were reviewed. The average annual delivery rate of this hospital is 5 500.
Results: During the study period, 61 patients with IA were diagnosed, which is an incidence of one per 992 live births. The rate of CCA among IA subjects was 15 (24.6%). In 12 patients (19.6%), the associated CCA was mild and reconstruction of the anorectum went smoothly. Three patients (4.9%) had significant CCA and died.
Conclusion: The incidence of imperforated anus in our hospital is one per 992 live births and the association with CCA is 24.6%. The majority of associated CCA with IA were of the mild type.

486: SINUS NODE DYSFUNCTION DUE TO MYOCARDITIS

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Introduction: The most common ECG findings associated with myocarditis are non-specific T-wave changes and tachyarrhythmias, including sinus tachycardia, ventricular tachycardia and ventricular fibrillation. Sinus node dysfunction in viral myocarditis has not been reported in the literature. We report a case of viral myocarditis complicated by sinus node dysfunction (SND), which resolved after a few days with intravenous immunoglobulin (IVIG) treatment.
Case report: A 17-year-old Caucasian male was hospitalised after a four-day history of progressively worsening chest pain and dyspnoea. On physical examination his vital signs were normal. Cardiovascular system examination revealed no murmur or pericardial rub and no signs of congestive heart failure. ECG showed ST elevation on the lateral chest leads. Echocardiogram revealed mild pericardial effusion, LV dilatation and mild–moderate depressed cardiac function (EF 45%). Laboratory data were consistent with myocarditis. He had an elevated creatinine kinase level (1 113 U/l), and CK MB was 84.9 ng/ml, troponin-T 4.900 ng/ml and myoglobin 211 ng/ml. Cardiac MRI revealed diffuse myocarditis, global hypokinesia, and low ejection fraction. Adenoviral and Coxsackie B_1 antibody titres were mildly elevated. IVIG was started in the first day of hospitalisation.

On day three, telemetry showed badycardia with a rate of 28 and sinus pauses of 6.7 s; 24-hour Holter revealed bradycardia with the lowest rate 27, with frequent sinus pauses up to 6.1 s. On Day 10 the

sinus pauses resolved. Subsequently the patient showed improvement in cardiac function. Telemetry showed normal sinus rhythm without sinus pause for five consecutive days prior to discharge. A repeat 24-hour Holter monitor after three weeks revealed a maximum sinus pause of 1.68 s.
Discussion: SND can happen in children with heterotaxia or after CHD cardiac surgery. In myocarditis, acute inflammatory processes trigger arrhythmogenic activity. This may cause transient conduction block of the AV node. SA node inflammation in this patient led to severe bradycardia and sinus pauses. As with other conduction abnormalities or arrhythmias in myocarditis, it may resolve once the inflammation improves.

488: ECHO DOPPLER ASSESSMENT OF ARTERIAL STIFFNESS IN PAEDIATRIC PATIENTS WITH KAWASAKI DISEASE

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Background: There is growing evidence to suggest there is increased arterial stiffness in patients with history of Kawasaki disease (KD). Pulse-wave velocity (PWV) is the most validated measure of arterial stiffness.
Methods: The aortic stiffness and impedance indexes were derived using an echocardiography Doppler method. The KD cohort were identified using our echocardiography database (2002–2012) for any patient who had had KD follow up of more than one year and included 42 patients (age 9.7 ± 2.0 years), compared to 44 age-matched control subjects recruited in an ongoing prospective manner. Our primary outcome measure was aortic PWV. Secondary outcome measures included characteristic impedance (Zc), input impedance (Zi), the elastic pressure–strain modulus (Ep), beta stiffness index (β-index), and measures of systolic function [shortening fraction (SF), ejection fraction (EF), mean velocity of circumferential fibre shortening (MVCFc) and peak systolic wall stress (σps)].
Results: Physical characteristics were similar between the two groups. The PWV was higher among KD patients compared to controls (458 ± 153 vs 370 ± 61 cm/s, $p = 0.0008$). The Zc, (Ep), and β-index were slightly higher among KD patients; however, the difference was not statistically significant. LV dimensions, M-mode derived EF, SF and MVCFc were all within normal limits with no difference in values between the two groups. The KD patients had lower σps compared to controls ($p = 0.01$). There was no significant correlation between the arterial stiffness indexes (PWV or Zi or Zc or Ep or β-index) and patient age, interval from time of diagnosis or fever duration. Logistic regression analysis of coronary artery involvement class showed no significant correlation with any of the arterial stiffness indices.
Conclusions: Arterial stiffness was increased in children after Kawasaki disease. There was no association between coronary artery involvement and PWV.

495: WORLD HEART FEDERATION ECHOCARDIOGRAPHIC CRITERIA FOR RHEUMATIC HEART DISEASE ALLOWS FOR REPRODUCIBLE DIAGNOSIS WORLDWIDE

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Background: Different echocardiographic definitions of rheumatic heart disease (RHD) have been used for screening for RHD. This led to the 2012 evidence-based World Heart Federation (WHF) echocardiographic criteria for RHD. The objective of this study was to determine whether the WHF criteria allow for consistent and reproducible differentiation of normal echocardiographic findings from mild RHD and therefore to assess the usefulness of the diagnostic criteria as a clinical and epidemiological tool.
Methods: Participants consisted of 15 international cardiologists/physicians with considerable RHD experience. A set of 100 echocardiograms was collated from population-based surveys of high-risk school-aged children of Australia and New Zealand. Echocardiograms were uploaded for blinded web-based reporting. Inter-observer variability in categorising echocardiograms as normal, borderline or definite RHD, as per WHF criteria, was measured by comparing the individual readings made by 15 participants with a reference reading.
Results: Of the 100 echocardiograms, 99 were considered suitable for reporting. A total of 1 485 reports were analysed. The reference readings distribution of cases was: 33 borderline RHD, 20 definite RHD and 46 normal or congenital heart disease. Overall agreement in categorising echocardiograms as normal, borderline and definite RHD (primary endpoint) was good, kappa 0.68 (95% CI: 0.65–0.72) with overall accuracy of 76.77% (95% CI: 0.75–0.79). The agreement over secondary endpoints, the presence of pathological degrees of aortic and mitral valve regurgitation were excellent, kappa of 0.87 (95% CI: 0.8–0.90) and 0.83 (95% CI: 0.79–0.86) respectively.
Conclusions: WHF echocardiographic criteria for RHD allows for reasonably consistent and reproducible diagnosis of RHD when used by experienced physicians. The ability of less experienced physicians and community health workers to diagnose RHD by echocardiography needs to be further evaluated if echocardiographic screening is to have a role in RHD control in resource-poor settings. Intra-observer studies of the WHF criteria are in progress.

496: WRESTLING MANOEUVRE AS THE CULPRIT IN ACUTE SEVERE AORTIC REGURGITATION

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A 16-year-old Hispanic male was seen in ER for facial petechiae after 'choke-hold' during wrestling. He had no chest pain, syncope, palpitation, dizziness or shortness of breath. The examination findings revealed a new onset three-quarter diastolic murmur and blood pressure of 174/38 mmHg with bounding peripheral pulses. There was no evidence of any diastolic murmur or elevated blood pressure during his last primary physician visit, one month prior to the episode. Past medical history was insignificant for any evidence of infective endocarditis, rheumatic heart disease, cardiac catheterisation, central line placements or blunt chest trauma. The patient underwent an echocardiogram which revealed severe aortic regurgitation, central aortic valve coaptation defect, severe left ventricular dilatation and normal ventricular contractility. He was then admitted to our hospital. Laboratory findings were unremarkable including cardiac enzymes, acute phase reactants and blood cultures. Since the initial attempt to surgically repair the aortic valve was unsuccessful, the Ross procedure was performed, with excellent results. His aortic valve pathology result showed slightly thickened valve cusps without evidence of vegetation of microorganisms. Since the new examination findings and symptoms developed immediately after wrestling, we surmised that our patient developed acute severe aortic regurgitation secondary to sudden increase in afterload caused by 'choke-hold' application. To our knowledge this is the first case of acute severe aortic regurgitation caused by wrestling 'choke-hold' manoeuvre.

512: ADJUSTABLE BILATERAL PULMONARY ARTERY BANDING FOR HYPOPLASTIC LEFT HEART AND ITS VARIANTS

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Object: Bilateral pulmonary artery banding (BPAB) is effective for high-risk hypoplastic left ventricle syndrome and its variants. However, deformation or stenosis of the pulmonary artery is a serious concern as a result. Recently we performed an adjustable BPAB with ePTFE suture (CV-0) and absorbable sutures to prevent this problem and examined its efficacy.
Method: From April 2003 to January 2012, we retrospectively evaluated 17 children who underwent BPAB and received the Norwood procedure or definitive repair (nine in the adjustable BPAB group and eight in the non-adjustable BPAB group). CV-0 was used as a band in the adjustable BPAB. The band was fixed with absorbable sutures (7-0 PDS) to the appropriate diameter according to the pulmonary venous flow and/or O_2 saturation; along with non-absorbable sutures (5-0 prolene) a few millimeters apart from the absorbable sutures. In the non-adjustable BPAB, a 2-mm-wide Teflon tape and non-absorbable sutures were used.
Results: The average interval until the next operation was 101 days in the adjustable BPAB group and 109 days in the non-adjustable BPAB group. There was no stenosis of the pulmonary artery in the adjustable group in the next operation, whereas pulmonary artery angioplasty was needed in four children in the non-adjustable group.
Conclusion: Adjustable BPAB is effective in preventing stenosis of the pulmonary artery and allows for pulmonary artery growth.

516: OUTCOME OF PRIMARY REPAIR OF ATRIOVENTRICULAR SEPTAL DEFECTS IN CHILDREN: AN EXPERIENCE FROM THE RED CROSS CHILDREN'S HOSPITAL IN SOUTH AFRICA

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Background: There is evidence to question the rationale for performing pulmonary artery banding (PAB) prior to repair of a complete atrio-ventricular septal defect (AVSD) in resource-limited settings. However, data on primary repair of a complete AVSD in these settings are scarce.
Objective: We examined the outcome of primary repair of a complete AVSD among children at the Red Cross Children's Hospital (RCCH) in South Africa, to determine whether this approach is justifiable as the first line of management in a developing country.
Methods: A retrospective review was performed on 31 children who underwent primary repair of a complete AVSD at RCCH between January 2009 and December 2010. We determined the surgical result, mortality and current follow-up status. The minimum follow-up period was one year.
Results: Median age was 8 (3 to 26) months (nine aged < 6 months, 10 aged 6–12 months and 12 aged > 12 months. Mean weight was 6 ± 2.4 kg. The majority (23/31) had Down syndrome (DS). Twelve children required pre-surgical cardiac catheterisation. An acceptable surgical result was achieved in 90% (28/31). There were three re-operations; two right and one left atrioventricular valve annuloplasty. Early (30-day) mortality was 13% (4/31); three in-hospital deaths attributable to infection and one 'cot death' at home. Overall mortality was 29% (9/31); the majority (6/9) of deaths occurred after initial hospital discharge. Out of 18 children followed up at RCCH, 13 are free of anti-failure treatment and there is no anticipated re-operation.
Conclusion: Primary repair of a complete AVSD was successfully carried out with low incidence of re-operation and in-hospital mortality. Late surgery was common, translating to increased costs required for cardiac catheterisation to assess operability. We conclude that timely primary surgery should be advocated for, even in resource-limited settings. Factors that reduce survival following successful surgery and discharge from hospital should be addressed.

523: STUDY ON THE DIAGNOSIS AND TREATMENT OF CHILDHOOD SUPRAVENTRICULAR TACHYCARDIA WITH INTRACARDIAC ELECTROPHYSIOLOGY:REPORTS OF 50 CASES

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Background: The aim of this study was to clarify the electrocardiographic characteristics of supraventricular tachycardia (SVT) in children and improve the technique of intracardiac electrophysiology (EPS) and radio-frequency catheter ablation (RFCA) in children.
Methods: Fifty patients with SVT were enrolled in the study from December 2007 to July 2012. The ECG data and the results of RFCA (ablation success, complications and recurrence) were studied retrospectively.
Results: Among the 50 patients, there were 29 cases (58%) with atrial ventricular re-entrant tachycardia (AVRT), 10 cases (20%) with atrial ventricular node re-entrant tachycardia (AVNRT), six cases (12%) with atrial tachycardia (AT), two with AVRT accompanying AVNRT, and one case with both AVRT and AT. The procedure was abandoned in two patients because of the risk in one case (AVRT and accessory pathway location near His bundle), and one case not induced by EPS. No recurred tachycardia and complications were found in all 50 cases up to the present.
Conclusions: RFCA is a safe procedure for tachycardia management, with high success rate and low rate of complications but the indications for RFCA should be carefully considered in young patients.

536: A CONCEPTUAL FRAMEWORK FOR COMPREHENSIVE RHEUMATIC HEART DISEASE CONTROL PROGRAMMES

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Background: The World Health Organisation (WHO), World Heart Federation (WHF) and other organisations recommend comprehensive control programmes for rheumatic fever (RF) and rheumatic heart disease (RHD). However, advice on components of control programmes tend to be simple linear lists, with little guidance on programme structure or priorities. In particular, there are limited recommendations on 'stepwise' implementation with few guidelines on which programme components should take temporal priority. An evidence-based framework for describing, prioritising and implementing comprehensive RF/RHD control programmes is needed. A unified framework approach would provide a structure for international collaboration and comparison. Providing guidance on programme priorities would be beneficial for emerging RHD control programmes, particularly those spurred on by scale-up of echocardiographic screening or delivery of tertiary interventions.
Methods: A literature review of existing RF/RHD control programme recommendations generated a list of programme components. Descriptions and analysis of RF/RHD control programmes informed temporal prioritising of component parts. Relevant programmatic research from other vertical disease control programmes was reviewed for generalisable implementation experiences.
Results: Twenty-four individuals' components of comprehensive RF/RHD control programmes were identified. These fell into 'baseline' programme requirements (including burden of disease data, treatment guidelines and human resources), and requirements for

providing primary, secondary and tertiary interventions. Primordial prevention and research priorities were overarching themes. These components were developed into a conceptual framework scheme.
Conclusions: Existing literature contains considerable lessons on the design and implementation of comprehensive RF/RHD control programmes. Fashioning these guidelines and programmatic experiences into a conceptual framework scheme benefits clinicians, policy makers and RHD advocates. Extending the framework may yield a model to isolate and evaluate individual programme components.

542: IMPLANTABLE CARDIOVERTER DEFIBRILLATOR THERAPY FOR PREVENTION OF SUDDEN CARDIAC DEATH IN CHILDREN AND YOUNG ADULTS IN MEXICO

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Background: Children at risk for sudden cardiac death have a wide variety of underlying cardiac diseases such as the broad spectrum of congenital heart disease, inherited arrhythmogenic diseases, and hypertrophic or dilated cardiomyopathy. Current data on paediatric implantable cardioverter defibrillator therapy are derived from small studies and retrospective multicentre studies. This single-centre study was undertaken to review our experience with ICD implantation in children and young adults with relatively different aetiologies.
Methods: We retrospectively reviewed the records of the paediatric patients who underwent cardioverter-defibrillator implantation at the InstitutoNacional de Cardiología Ignacio Chávez between January 2005 and February 2011.
Results: A total of 20 patients who underwent ICD implantation during this period were included in this study. The median age was 15 years. Most of the patients had cardiomyopathy (n = 13) or ion channel diseases (n = 6). Devices were implanted for either secondary (n = 15) or primary (n = 5) prevention. The selected ICD generator type was dual chamber in 12 patients, single chamber in seven patients and biventricular in one. Six patients received 30 shocks. Four of 15 patients (26%) from the secondary prevention group experienced at least one appropriate shock during a median period of 3.4 years (range: 1.2 months to 6.5 years). Eight inappropriate shocks were delivered in two patients from the secondary prevention group during the median period of 3.4 years. The most important reason for inappropriate shocks was T-wave over-sensing. No acute or chronic complications occurred.
Conclusions: The ICD was safe and effective in interrupting malignant arrhythmias in children and young adults with a high risk of sudden cardiac death. The occurrence of lead-related acute or chronic complications was zero and the incidence of inappropriate shocks was low. Careful programming is mandatory to reduce inappropriate shocks.

547: CARDIAC RESYNCHRONISATION THERAPY IN CHILDREN IN MEXICO

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Background: Cardiac resynchronisation therapy (CRT) is an important management tool in adults with congestive heart failure (CHF). The role of CRT in children is unclear. This study was undertaken to review our initial experience of CRT in children.
Methods: We initiated the CRT programme in children on January 2011; only two children have been included.
Results: Case 1: a five-year-old girl was transferred because of progressive LV dysfunction. At one year of age she underwent a ventricular septal defect closure. One year later a permanent VVI pacemaker was implanted for the treatment of AV block. Echocardiography revealed cardiac asynchrony, altered diastolic filling, mitral and tricuspid valve regurgitation, severe ventricular dilatation and an ejection fraction of 16%. We implanted an epicardial lead in the right atrial appendage and an epicardial lead in the left ventricle as an upgrade to the existing epicardial right ventricular lead. After 18 months of follow up no clinical improvement has been seen. She is on a cardiac transplant list.

Case 2: a 16-year-old girl was transferred with the diagnosis of idiopathic dilated cardiomyopathy and severe CHF. The NYHA class was IV. Echocardiography showed severe left ventricular dilatation, ejection fraction of 17% and cardiac asynchrony. We implanted a biventricular pacemaker. Three endocardial leads were placed on the right atrial appendage, the RV apex and on the LV posterolateral wall via coronary sinus. After 14 months the NYHA class is I, the ejection fraction is 40%, left ventricular volume and diameter have reduced and asynchrony values have improved.
Conclusions: CRT may have an important role in select paediatric patients. Further work is necessary to delineate in which underlying anatomical and pathophysiological condition it would be more effective. In less developed countries there are financial restrictions preventing this treatment and other new therapies from being offered to all patients who could benefit from them.

548: EXTENSIVE MYOCARDIAL INFARCTION IN AN 11-YEAR-OLD GIRL: CASE REPORT

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Myocardial infarct in childhood is very rare. Mostly it is associated with structural, hereditary or acquired disease. We report on an 11-year-old girl who presented after a near-drowning episode. The child was in cardiopulmonary shock and had to be ventilated. Despite volume and starting on catecholamines, her blood pressure remained low. Primary echocardiogram revealed depressed myocardial function. Initial blood work showed a white blood cell count of 27 000, abnormal I/T ratio, CK 7 000 U/l, CK-MB 688 ug/l, troponin 12 ug/l. The girl's history was unremarkable. Repeated echocardiogram after four hours showed a severely reduced left ventricular function (EF 25–30%) with dyskinesia/hypokinesia along the free left ventricular wall. The right ventricular systolic function remained normal. ECG showed sinus tachycardia, deep Q waves in leads V3–V5, and ST elevation in V1–V2. Based on these findings, we primarily diagnosed acute myocarditis (DD acute vasculitis with coronary vasospasm, hypoxic ischaemia after near drowning).

Within the following two days her LVEF recovered (EF 50–55%). A sudden episode of nausea and vomiting on day three was followed by ventricular tachycardia and ventricular fibrillation. Resuscitation and rescue extracorporeal membrane oxygenation (ECMO) was initiated. Coronary angiogram revealed an abnormally widely spread thin network of coronary branching along the left coronary artery with a discrete narrowing of the main stem. CT scan confirmed the diagnosis of a coronary anomaly with the left coronary artery coming from the acoronary sinus.

Corrective surgery with an 'unroofing' of the intramural part and creation of a neo-ostium of the LCA was performed. The patient was weaned from ECMO on day five. However, despite regained normal coronary flow, the LV function remained depressed (EF 20–25%). After syx months, the patient underwent heart transplantation.
Conclusion: The presence of segmental myocardial dyskinesia/hypokinesia always implies further diagnostic imaging regardless of the patient's age.

552: SICK CHILD OR SICK SINUS: REPEATED SYNCOPE IN EARLY INFANCY

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Introduction: Syncope is often observed in paediatrics. The majority of patients are teenagers. In contrast to adults, a cardiac origin is only observed in 2–5% of cases. Regardless of the reported low incidence of cardiac origin for sudden loss of consciousness (LOC) in childhood, detailed cardiac evaluation is mandatory as the rare organic causes are all potentially dangerous (arrhythmia, cardiomyopathies, structural heart disease, pulmonary hypertension).
Patient report: A 19-month-old girl with repeated LOC since the age of six months (two to three times per month) presented with developmental delay. The mother had ablation at the age of 18 years for WPW. Her other family history was unremarkable. Detailed neurological evaluation (two EEGs, psychomotor skills) was unremarkable. At the age of 18 months the frequency of LOC increased after an event-free period of three months. Echocardiogram revealed a large atrial septal defect. The standard ECG and 24-hour Holter examination was unremarkable but a seven-day ECG recording revealed two episodes with sinus pauses of up to 3.4 seconds, both during the daytime when the toddler was awake. The diagnosis of a sick sinus syndrome was made. A VVI pacemaker (back-up heart rate 80 beats/min) was put in place and ever since, no episodes of sudden LOC have been reported. The closure of the atrial septal defect has been postponed to the age of four to five years.
Conclusion: This case illustrates the importance of detailed cardiac assessment for sudden LOC in childhood, even for toddlers with obvious neurological issues (i.e. developmental delay), as this might be due to recurrent hypoxic encephalopathic events. The incidence of sick sinus syndrome in paediatric patients is very low, which makes it more difficult to diagnose. In our patient neither Holter recordings nor standard ECG showed any sign of arrhythmia.

553: DOES THE 12-LEAD ELECTROCARDIOGRAM IMPROVE DIAGNOSTIC DETECTION OF ATRIAL SEPTAL DEFECTS DURING POPULATION-BASED SCREENING?

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Background: Controversy continues regarding the diagnostic utility of the electrocardiogram (ECG) for the diagnosis of congenital heart disease (CHD) and cardiomyopathy. Unlike other forms of clinically significant CHD, patients with atrial septal defects (ASDs) often have few symptoms and may, under some circumstances, have a nearly normal examination. We sought to evaluate the added diagnostic yield of ECGs in the diagnosis of ASDs.
Methods: During a humanitarian screening in two rural provinces in Mongolia (Bayan-Ulgi and Khovd), children from birth to 20 years underwent a cardiac physical examination by American and Mongolian paediatricians and nurse practitioners. All children also underwent a 12-lead ECG read by a paediatric electrophysiologist. A potentially pathological murmur on examination or an ECG consistent with ASD (QRS duration > 100 ms with rSR' in lead V_1) prompted echocardiographic screening (ECHO) by a paediatric cardiologist.
Results: CHD was identified in 47 of 1 615 patients (3.0%) screened over five days, 14 (29.8%) of whom had an ASD by echo. All patients with ASD had an abnormal examination, with 12 having ECG findings consistent with the diagnosis (85%). No ASDs were identified when the ECG was the only feature consistent with ASD. Overall, an ECG was abnormal in 51 patients, of whom 31 had an abnormal echo.
Conclusions: ECG was not found to improve the diagnostic yield for ASDs over the physical examination alone, even when non-cardiologists were employed. In addition, the ECG does not appear to be more sensitive than the physical examination. Given the added time and resources required to perform them, the use of an ECG for population-based screening does not seem justified.

565: POST-FONTAN COMPLETION. USE OF WARFARIN OR ASPIRIN: 12.5-YEAR EXPERIENCE FROM A SINGLE PAEDIATRIC CARDIAC CENTRE

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Introduction: Thromboembolic events are known complications after Fontan operations, with the published literature attempting to distinguish the most effective preventative measure. Some articles have shown no difference in efficacy between warfarin and aspirin. We focused on the side effects of warfarin and aspirin in our experience.
Methods: We retrospectively reviewed all patients undergoing Fontan completion between January 2001 and June 2012.
Results: Over the 12.5 year period, 134 patients underwent Fontan completion. The median age at Fontan completion was 5.5 years (range 2.9 to 16.5 years). Median follow up was 4.3 years (2 days to 11.9 years). Of these, 120 of 134 patients were started on warfarin, while only 10 were started on aspirin. Four patients died in the early postoperative period before establishment on warfarin or aspirin.

In the warfarin group, four patients converted to aspirin. One patient had significant extradural haematoma following a bicycle accident, which needed drainage. One patient developed postoperative haemorrhagic stroke, while two patients converted out of choice. Of the remaining 116 patients on warfarin, seven had minor complications with self-limiting epistaxis (four patients), superficial bruising (one patient), bleeding into pleural cavity (one patient), and clot in the left atrium (one patient due to low INR). One patient developed a clot on the fenestration plug device and was put on warfarin and aspirin. Seven of these eight patients continued with warfarin. In the aspirin group, there was no documented complication. The limitations of this retrospective review are that the patients were non-randomised into warfarin or aspirin therapy and we had not actively looked for thrombus in asymptomatic patients.
Conclusions: There is no increased risk of bleeding complications from the use of warfarin post-Fontan surgery. Our future aim is to prospectively randomise patients to warfarin or aspirin to assess complications.

566: NOVEL APPLICATION OF REAL-TIME MAGNETIC RESONANCE ANGIOGRAPHY: THE END OF DIAGNOSTIC CATHETERISATION IN NEONATES?

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Background: Magnetic resonance (MR) imaging for delineating extracardiac vasculature in newborns with congenital heart disease is not widely used. Fast circulation times mean current MR angiographic techniques lack the temporal resolution to assess complex cardiac anatomy within a single breath-hold. We report on the use of four-dimensional time-resolved angiography with keyhole (4D TRAK) to confirm diagnoses in newborns not fully resolved by echocardiography. 4D TRAK has the advantage of rapid sequence acquisition, providing high temporal resolution, three-dimensional datasets, with dynamic multi-phase visualisation.
Methods: A retrospective review of neonates (< 28 days old) undergoing cardiac MR imaging with 4D TRAK from July 2011 to July 2012 was performed. All underwent an initial reference scan, with

subsequent dynamic images acquired within a single breath-hold. Indication for referral, diagnosis made from the MR scans and correlation with surgical findings were assessed. All MR scans were performed on a commercial 1.5T scanner (Achieva; Philips Healthcare, Best, the Netherlands).
Results: Nine neonates had 4D TRAK MR sequence performed under general anaesthetic after injection of a contrast agent (Gadopentolate 0.1 mmol/kg). Median age was five days, range two to 23 days. Mean weight was 3.1 kg, range was 2.1–4.5 kg. Seven patients proceeded to surgery based on the MR, where findings were confirmed; one required no further intervention, and one required diagnostic catheterisation to assess multiple aorto-pulmonary collateral arteries (MAPCAs).
Conclusions: MR angiography with keyhole permits rapid acquisition of 3D datasets with high temporal resolution. Within a single breath-hold, the sequential filling of arterial and venous vessels can be visualised, overcoming the limitations of temporal resolution imposed by existing MR angiography. The use of 4D TRAK confers high diagnostic accuracy vital for surgical planning. 4D TRAK is appropriate where diagnostic uncertainty remains following echocardiographic assessment and should be considered in place of invasive diagnostic cardiac catheterisation or X-ray-dependent computed tomography.

569: PREDICTORS OF OUTCOME IN PAEDIATRIC IDIOPATHIC CARDIOMYOPATHY

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Objectives: Idiopathic dilated cardiomyopathy (IDCM) is a severe illness with a high mortality rate in the paediatric population. The purpose of this study was to highlight our experience on the clinical course and outcome of IDCM.
Methods: Patients' files were reviewed retrospectively for diagnosed cases of IDCM in the paediatric cardiology unit of King Abdulaziz University Hospital, Jeddah, Saudi Arabia from January 2003 to June 2011. Data on full history, clinical examinations and investigations were recorded and grouped according to outcome as alive and well (group 1), alive and symptomatic (group 2), and worsened or dead (group 3). Data was subjected to descriptive analysis. Chi-square and Student's paired *t*-test techniques were used where appropriate. Spearman rank correlation and survival analysis was done.
Results: Eighty-three patients were included, with presenting age median 14 months (range 2 months to 12 years), and female predominance: 53 patients (63.9%). On presentation, cardiomegaly was noted in 72 patients (86.7%) with increased lung vascularity in 45 (54%). Sixty-one (74%) patients had ST-segment and T-wave changes on electrocardiogram while the same number had left ventricular hypertrophy, and 15 (18%) had arrhythmias. Echocardiography records on presentation and at last follow up showed significant differences in several areas. Group 1 had 40 (48.2%), group 2 had 23 (27.7%) and group 3 20 (24.1%), including nine cases who died. Survival rate over three years was 78%. Older the age worse was the outcome (Spearman's rho = 0.3, $p = 0.04$).
Conclusion: The majority of subjects presented during the first year of life; three-year survival rate was 78%. Favourable outcome was correlated with younger age at presentation.

579: CARDIAC LESIONS IN NEONATES WITH GASTROINTESTINAL MALFORMATIONS

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Introduction: The association between congenital structural cardiac lesions and major gastrointestinal malformations, for example, trachea-oesophageal fistula, omphalocoele, gastroschisis and anorectal abnormalities, has been well described in the literature. Most of the descriptions have been made in a series of articles by RD Greenwood who started in 1975 by documenting an incidence of 23% of cardiac lesions in patients with congenital diaphragmatic hernia, 12% in those with imperforate anus, 19% in those with omphalocoele and 15% in neonates with trachea-oesophageal fistula. It was decided to do an institutional retrospective analysis of this association at an African tertiary care centre.
Methods: This study was a retrospective descriptive analysis of cardiac lesions that were found in these neonates. Secondarily, it will also give an indication as to the type of cardiac lesions these neonates are likely to have, determine whether or not a gender preponderance exists and document the geographical distribution of these neonates.
Results: An average of 20 000 to 25 000 live births per year occur in the neonatal unit at Chris Hani Baragwanath Hospital. Over the five-year period between 1 July 2006 and 31 July 2011, 129 neonates with the above-mentioned gastrointestinal malformations were referred for echocardiography. The study revealed an incidence of 27% of cardiac lesions in neonates with gastrointestinal malformations. Most lesions occurred in neonates with anorectal malformations (27%) and omphalocoele (38%). The commonest lesions were ventricular septal defect, complex cyanotic heart defects and right-sided aortic arch. There seemed to be no significant male or female preponderance. Most of the neonates fell within the southern areas of the referral regions of Chris Hani Baragwanath Hospital cachment area, which includes southern Gauteng and the North West Province.
Conclusion: The findings were similar to the world literature. It is important to make the association between GIT malformations and cardiac lesions. The association can impact on the outcome of both the GIT and cardiac lesions.

580: DYSSYNCHRONY AND VENTRICULAR FUNCTION IMPROVE FOLLOWING CATHETER ABLATION OF NON-SEPTAL ACCESSORY PATHWAYS IN CHILDREN

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Introduction: Prior studies assessing ventricular dyssynchrony in children with Wolff–Parkinson–White syndrome (WPWs) have focused on septal or paraseptal pathways and described paradoxical or hypokinetic septal motion. Data regarding non-septal pathways are limited, as these have generally been perceived to carry lesser risk for ventricular dysfunction.
Methods: We characterised the degree of left ventricular (LV) dyssynchrony and LV function by echocardiographic analyses with tissue Doppler imaging (TDI) prior to and following catheter ablation of accessory pathways.
Results: Sixteen children, age 14.2 ± 3.7 years, weight 53 ± 17 kg, were assessed. All had WPWs, structurally normal hearts, and underwent successful ablation (cryoenergy in four; radiofrequency in 12). Septal/paraseptal pathways were present in six (37.5%) and non-septal pathways in 10 (62.5%): left lateral ($n = 5$), right lateral/anterolateral ($n = 3$), left posterior ($n = 2$). Following ablation, LV ejection fraction (EF) (Simpson's method) increased by 4.9 ± 2.1% ($p = 0.038$) from a baseline value of 57.0 ± 7.8%, with a decrease in the difference between aortic and pulmonary preejection times (11.0 ± 3.3 ms, $p = 0.017$). By TDI, the interval from QRS onset to peak systolic velocity decreased from 33.0 [interquartile range (IQR 20.0, 18.0)] to 18.0 (IQR 5.0, 24.0) ($p = 0.013$). No significant change in septal-to-posterior wall motion delay or diastolic parameters was noted. LVEF increased to a greater degree following ablation of non-septal (5.9 ± 2.6%, $p = 0.023$) versus septal (2.5 ± 4.1%, $p = 0.461$) pathways. The four patients with LVEF < 50% prior to ablation, two of whom had left lateral pathways, improved to > 50% post ablation. Similarly, the

magnitude of improvement in LV dyssynchrony was more marked in patients with non-septal versus septal pathways, e.g. difference between septal and lateral wall motion delay before and after ablation of 20.6 ± 7.1 ms ($p = 0.015$) versus 1.4 ± 11.4 ms ($p = 0.655$).
Conclusion: LV systolic function and dyssynchrony improved after ablation of antegrade conducting accessory pathways in children, with greater changes for non-septal pathways.

589: LEFT VENTRICULAR SUBMITRAL ANEURYSMS

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Objective: A retrospective institutional review was done of the pathology, classification and surgical management of left ventricular submitral aneurysms (LVSMA). This is a well-recognised but relatively rare disease, commonly found in patients of African ancestry.
Methods: The series compromised 31 patients treated surgically at two institutions from 2001 to 2012. Patients were of African ancestry ($n = 25$) and mixed ancestry ($n = 6$). Natural history, clinical presentation, histopathological findings, aetiology, and operative techniques are presented.
Results: There were 19 male and 12 female patients, mean age was 19 ± 6 (range 8–45) years. Patients were grouped according to the degree of posterior mitral annulus involvement. In group I ($n = 18$) a single neck, in group II ($n = 3$) multiple necks, and in group III ($n = 10$) involvement of the entire annulus was found. Mean age of group III (31 ± 7 years) was older than groups I and II (15 ± 5 years) ($p = 0.001$). This is suggestive of progression of disease with age. An intracardiac surgical approach was used in 21 patients, an extracardiac approach in two and a combined approach in nine patients. Failure to control the neck of the aneurysms ($n = 3$) and failure of the mitral valve repair ($n = 3$) resulted in subsequent re-operation. Operative mortality ($n = 1$).
Conclusion: The aetiology of LVSMA is thought to be congenital due to an inherent weakness of the posterior mitral annulus. In our study the majority ($n = 23$) of patients had no histological evidence of contributing aetiology, but the study confirms multi-factorial aetiology. A new classification is proposed based on pathological findings.

600: BIOPHYSICAL PROPERTIES OF THE AORTA IN ADOLESCENT FEMALES WITH ANOREXIA NERVOSA

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Background: Patients with anorexia nervosa (AN) have altered physiological responses to exercise. This study aimed to determine the differences in exercise capacity and haemodynamic parameters with exercise in patients with AN.
Methods: This was a retrospective case–control study. Sixty-six adolescent females with AN and 21 adolescent female controls exercised on a semi-recumbent ergometer in three-minute, 20-watt incremental stages to volitional fatigue. Heart rate (HR), blood pressure (BP) and echo-Doppler indices were measured pre-, at each stage, immediately and three minutes post-exercise. Fractional shortening (FS), peak aortic velocity (PAoV), mean velocity of circumferential fibre shortening (MVCFc), wall stress (WS), cardiac index (CI), and systemic vascular resistance (SVR) were calculated. Peak oxygen consumption (VO_2), minute ventilation (VE), respiratory exchange ratio (RER), and arterial–venous oxygen difference (a-vO_2) were determined using open-circuit spirometry.
Results: Patients with AN had a significantly lower BMI (16.7 vs 19.7 kg/m^2, $p < 0.001$), total work (1 126 vs 1 914 J/kg, $p < 0.001$), total test duration (13.8 vs 20.8 min, $p < 0.001$), peak VE (47.4 vs 72.0 l/min, $p < 0.001$), and VO_2 (31.3 vs 39.7 ml/min/kg, $p < 0.001$) and higher RER (1.14 vs 1.06, $p = 0.001$) when compared to controls. Systolic BP, diastolic BP, and PAoV were lower at pre-exercise, increased with exercise, and were lower at peak exercise in ANsubjects vs controls. HR, FS, MVCFc and CI showed no difference at pre-exercise, increased with exercise, and were lower at peak exercise in AN subjects vs controls. WS decreased with exercise and was lower in ANsubjects vs controls at pre-exercise and peak exercise. SVR pre-exercise was lower in AN subjects, decreased with exercise, and there was no difference at peak exercise. The a-vO_2 increased with exercise and showed no differences between groups.
Conclusions: Adolescent patients with AN had decreased exercise capacity and abnormalities in their haemodynamic parameters and myocardial performance during exercise compared with control subjects.

621: KAWASAKI DISEASE ANALYSES IN A REFERENCE PAEDIATRIC HOSPITAL IN SOUTHERN BRAZIL FROM 1980 TO 2012

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Background: Kawasaki disease is a systemic and acute vasculitis with unknown aetiology and is actually considered the main cause of acquired heart disease in children in developed countries. The clinical presentation is typically characterised by five days or more of fever, associated with at least four of the following: conjunctivitis, oral changes, extremity changes, rash and cervical adenopathy, and these manifestations usually appear in a sequence, but without a defined order. Because there is no specific laboratory test, the diagnosis is purely clinical, and may be confirmed by indirect tests of inflammatory activity. Delayed treatment of patients increases the risk of developing cardiac abnormalities, so early diagnosis is the key to a better prognosis.
Objectives: To identify and analyse epidemiological, clinical and therapeutic aspects of the disease in patients at the Joana de Gusmao Children's Hospital, Florianópolis, Santa Catarina, from 1980 to 2012.
Methods: Using data from medical records, cases were analysed with regard to age, gender, race, origin, year and season of occurrence, clinical manifestations and diagnostic criteria, laboratory tests, cardiac involvement, treatment, complications and death.
Results: During the study period, 60 cases occurred, with higher frequencies after 2001. Patients were predominantly white male children under five years old from Florianópolis. Of the patients, 70% fulfilled the diagnostic criteria. Anaemia, leukocytosis with neutrophilia, thrombocytosis, increased ESR and CRP were frequent findings. Cardiac involvement occurred in 53.3%, predominantly coronary changes. Intravenous immunoglobulin was used in 98.3% of children, and ASA in 100% of them. There were complications in 6.6% of the cases and no deaths.
Conclusions: The increased number of cases in recent years may reflect greater attention to the clinical features, even though the diagnosis is delayed. Cardiac compromise was found to be frequent, especially in the coronary arteries, probably due to the long time period until its identification.

624: LONE ATRIAL FIBRILLATION IN AN ADOLESCENT

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We report on a 15-year-old adolescent girl with good previous health, who presented with sudden onset of palpitation and dizziness. Examination revealed fast, irregular apical beats (170 beats/min) with normal blood pressure. Electrocardiogram showed typical fast atrial fibrillation (AF). Transthoracic echocardiogram confirmed there was no structural abnormality, but the cardiac function was suboptimal with biventricular hypokinesia. No intracardiac thrombus was identified. Successful synchronised cardioversion was performed without anticoagulation, and the rhythm was converted to sinus without any thromboembolic events.

The girl was put on new-generation anticoagulant and antiarrhythmic medication, dabigatran and dronedarone, respectively, for a short period of time, and there was no recurrence of AF. Lone AF is rare in the paediatric population. The thromboembolic risks are exceedingly low. Therefore, aggressive rhythm control would be the approach in its management, and anticoagulation before cardioversion may not be indicated. Genetic predisposition has become a new trend in the study of young-onset lone AF. The updated evidence of managing lone AF in children and adolescents is discussed, including the use of dabigatran and dronedarone.

625: VENTRICULAR HYPERTROPHY WITH OUTFLOW TRACT OBSTRUCTION VERSUS DILATED CARDIOMYOPATHY IN NEUROBLASTOMA

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Catecholamine-associated hypertension secondary to neuroblastoma can occasionally be demonstrated, but cardiomyopathies caused by neuroblastoma have been rarely reported. We report two cases of neuroblastomas with different extremes of severe cardiomyopathic changes and complications.

The first case was a two-month-old girl with a retroperitoneal mass, which was incidentally found in an ultrasonic examination during a postnatal evaluation of antenatal hydronephrosis. Hypertension was detected and echocardiogram showed severe concentric left ventricular hypertrophy with mid-ventricular obstruction, with a peak pressure gradient of 51 mmHg, simulating hypertrophic obstructive cardiomyopathy (HCOM). The second case was a 30-month-old girl who presented with abdominal discomfort, subsequently developed refractory hypertension and heart failure. Echocardiogram revealed a markedly dilated left ventricle with poor cardiac contractility. Computed tomographic scan indicated a huge abdominal mass.

Both girls were diagnosed with neuroblastoma, and high levels of urinary catecholamine and its metabolites were identified. Choice of antihypertensives was different. Beta-blocker was used in the first case to promote left ventricular filling, while angiotensin converting enzyme inhibitor was used in the second case. The anthracycline group of chemotherapy was avoided in the initial phase of treatment. Chemotherapy and subsequent removal of the tumour led to successful normalisation of blood pressure and regression of abnormal cardiomyopathic changes. HOCM-like features secondary to catecholamine-secreting neuroblastoma is exceedingly rare. The very early onset of HOCM suggested that the remodelling of the heart had already started during her foetal period. Updated management of catecholamine-induced cardiomyopathy associated with neuroblastoma is also discussed.

627: ROUTINE HEAD ULTRASOUND SCANS ARE NOT INDICATED IN THE PRE-OPERATIVE EVALUATION OF INFANTS WITH CONGENITAL HEART DISEASE

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Background: Head ultrasound scans (HUS) are a routine part of the pre-operative evaluation of young infants with CHD in many institutions. HUS have the advantage of being inexpensive, easily performed and readily available. However, the utility of HUS in the cardiac population is unknown. More recently, MRI has become a valuable tool in the early detection of brain injury in infants with CHD. The purpose of this study was to assess the utility of pre-operative HUS in a cohort of young infants who also underwent pre-operative MRI as part of a prospective research study of brain injury in infants undergoing surgery for CHD.
Methods: One hundred and sixty-seven infants born at 35 weeks' gestation or greater and due to undergo surgery for CHD were included in this three-centre study. None of the patients had any clinical signs or symptoms of pre-operative brain injury and all received HUS and brain MRI. All imaging was reported by experienced neuroradiologists who were blinded to any specific clinical details of the study participants. The findings were compared to evaluate for the presence of brain injury.
Results: Pre-operative brain injury was found on HUS in five infants (3%) and MRI in 44 infants (26%) ($p < 0.001$). The abnormalities on HUS were: intraventricular haemorrhage in four that was not confirmed on MRI, performed within a few days after HUS, and periventricular leukomalacia in one patient. The predominant MRI abnormality was white matter injury (32 infants). Other findings included infarct (14), haemorrhage (five), and lesion or infarct of deep nuclear grey matter (two).
Conclusions: Pre-operative brain injury on MRI was present in 26% of infants with CHD, but only 3% had any evidence of brain injury on HUS. Among positive HUS, 80% were false positives. Our findings suggest that routine HUS are not indicated in asymptomatic term or near-term neonates undergoing surgery for CHD.

629: CONGENITAL HEART DISEASE IN MILLER–DIEKER SYNDROME

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Background: Miller–Dieker syndrome (MDS) is a contiguous gene deletion syndrome caused by deletion of 17p13.3. It is characterised by severe lissencephaly, characteristic facial features, severe neurological abnormalities, and occasionally other congenital anomalies such as renal, gastrointestinal and cardiac defects. The lissencephaly in MDS is due to deletion of the *LIS1* gene. The dysmorphisms and other features are believed to be due the deletion of genes distal to *LIS1*. The incidence of CHD among individuals with MDS has been

estimated to be 65%. The frequency of various form of CHD in MDS patients has not been established.
Methods: Records of patients diagnosed with MDS and CHD between 2008 and 2012 at our institutions were reviewed. A literature review included searching MEDLINE and Google Scholar. Inclusion criteria included a clinical or genetic diagnosis of MDS and a specific CHD diagnosis. Cases with ambiguous descriptions of cardiac anatomy and infants with only a PFO and/or PDA, which could be physiological, were excluded from the analysis.
Results: We report four new cases of CHD in MDS. Their cardiac diagnoses were TAPVR, VSD, VSD and pulmonary stenosis (PS) and TOF. The literature review identified 16 cases of CHD in MDS: TOF-PA (four), TOF (three), PS (two), and one each of Ebstein anomaly, PA/IVS, ASD and DORV.
Conclusions: This is the first report of TAPVR in a patient with MDS. In addition, this study found that among MDS patients with CHD, right-sided lesions were common. TOF-PA was particularly frequent. Genes in the region of chromosome 17 associated with MDS have not previously been reported to be involved in CHD. The results of this study highlight the need for further study of the impact of genes from this region on cardiac development.

644: CARDIAC AUTONOMIC FUNCTION IN ADOLESCENTS OPERATED BY ARTERIAL SWITCH SURGERY

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Background: Children with transposition of the great arteries, in whom an arterial switch operation (ASO) is performed, have been shown to have an increased incidence of sudden death, which may be due to cardiac autonomic imbalance and repolarisation instability. We hypothesised that (1) cardiac norepinephrine (NE) kinetics, and (2) arterial baroreflex sensitivity (BRS), reflecting sympathetic activity and vagal function, respectively, are altered in this group.
Methods and Results: Seventeen children (15.8 ± 1.6 years) with ASO surgery in the neonatal period were studied; 17 had cardiac BRS assessed by spontaneous fluctuations of systolic blood pressure and RR interval, and repolarisation was measured as QT variability index. Matched healthy subjects were controls. Cardiac vagal function and repolarisation pattern were unchanged following ASO surgery. At cardiac catheterisation, we infused tritiated $^{[3H]}$NE into eight of these children to examine total body and cardiac sympathetic function at baseline and following five minutes of adenosine infusion to induce reflex sympathetic activation. Blood was sampled simultaneously from the aorta and coronary sinus. Cardiac fractional extraction of $^{[3H]}$NE was substantially lower in operated children, being 56 ± 10 vs 82 ± 9% ($p = 0.0001$). Following i.v. adenosine in the operated group, NE total body spillover doubled vs baseline ($p < 0.002$) and the coronary venous–arterial concentration of $^{[3H]}$dihydroxyphenylglycol increased four-fold ($p = 0.04$).
Conclusions: The arterial switch operation performed neonatally appeared to leave cardiac vagal function intact and, although cardiac sympathetic activation in response to adenosine occurred, cardiac neuronal NE re-uptake was impaired. This may have been pro-arrhythmic by reducing removal capacity of NE from the cardiac synaptic cleft.

654: PRENATAL RISK FACTORS ASSOCIATED WITH CONGENITAL HEART DISEASE

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Background: Congenital heart disease (CHD) affects 1% of live births. We previously identified advanced parental age, maternal smoking and medication use during pregnancy to be associated with risk for CHD in offspring.
Objective: To assess the interactions between prenatal risk factors and CHD risk.
Methods: Patients with CHD and healthy controls enrolled in the Heart Centre Biobank were studied. Prenatal exposure data and CHD anatomical subtypes were obtained from questionnaires and medical records. The cardiac phenotype was compared by prenatal risk-factor exposure using chi-squared analysis.
Results: The study cohort included 2 345 CHD patients and 193 control patients; 19% reported advanced maternal age, 11% advanced paternal age, 21% maternal smoking, 31% non-fertility medication use, and 6% fertility medication use during pregnancy; 53.6% had no risk factor, 18.8% had one risk factor, and 27.5% had ≥ two risk factors. Advanced paternal age was associated with lower frequency of left heart lesions (LHL) ($p = 0.003$). Medication exposure during pregnancy was associated with higher frequency of septal defects ($p = 0.003$), endocardial cushion defects ($p = 0.009$), right heart lesions ($p < 0.001$), and thoracic vessel anomalies ($p = 0.001$), and a lower frequency of LHL ($p = 0.018$). Other prenatal risk factors did not show a predilection for specific CHD subtypes.
Conclusions: There is a high burden of environmental risk factors in CHD causation. Advanced paternal age and non-fertility medications were associated with specific CHD phenotypes. Additional studies will evaluate whether genetic factors increase the susceptibility to development of foetal CHD in pregnancies with environmental exposures.

659: GENETIC ASSOCIATIONS WITH ANTHRACYCLINE CARDIOTOXICITY IN PAEDIATRIC CANCER PATIENTS

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Background: Anthracycline cardiotoxicity (ACT) is the third leading cause of death in cancer patients and is associated with left ventricular (LV) wall thinning and dysfunction. Several genetic variants that regulate anthracycline pharmacokinetics have been identified and variants in pharmacodynamic pathways on ACT require further study.
Objective: To study the relative contribution of genetic variants involved in ACT.
Methods: One hundred and thirty-nine paediatric cancer patients were prospectively enrolled through the Heart Centre Biobank; 77 patients were genotyped for three hypoxia-inducible factor 1-alpha (HIF1A) SNPs, rs11549465 (1744C > T), rs2057482 (45T > C) and rs10873142 (-145C > T). Serial echocardiograms were reviewed to obtain markers of LV function and wall thinning, including LV ejection fraction (LVEF), end-diastolic dimension and posterior wall thickness (LVPWT), and these were compared with patient genotype.
Results: Of the 139 anthracycline-exposed patients, 59% were male, 64.7% white, 26.6% Asian, and 8.6% other. Mean age at enrolment was 12.7 years. Mean duration of follow up was 5.8 years; 14% of patients had at least one abnormal measure (≤ 55%) of LVEF at any time point. All three SNPs were in Hardy–Weinberg equilibrium. For -145C > T (rs10873142), the LVPWT *z*-scores were significantly lower in patients with the CT+CC genotype compared with the TT genotype (–0.78 vs –0.03, $p = 0.008$).
Conclusions: The HIF1A-145CT/CC genotype was associated with LV posterior wall thinning during follow up. Additional candidate SNPs are being genotyped and will help for early identification of patients with a genetic susceptibility to ACT that can guide pre-emptive risk-reduction measures.

663: GENETIC VARIANTS ASSOCIATED WITH PROGRESSIVE RIGHT VENTRICULAR REMODELLING IN TETRALOGY OF FALLOT

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Background: The predictors for progressive right ventricular (RV) remodelling following surgical repair of tetralogy of Fallot (TOF) are not well known.
Objective: To identify genetic variants associated with progressive RV remodelling following TOF repair.
Methods: Patients with repaired TOF were enrolled prospectively through the Heart Centre Biobank. Eight single-nucleotide polymorphisms (SNPs) in six genes identified in association with TOF in a genome-wide association study were chosen. RV remodelling was assessed by RV dilation *z*-scores and qualitative RV measurements obtained through echocardiographic reports, RV ejection fraction (RVEF), and RV end-systolic volume index (RVESVi) scores from MRI at their last follow up.
Results: Of the 141 patients included, 53.9% were male (all Caucasian; mean age 14.07 years). The mean age at initial repair was 2.1 years and 23 patients subsequently underwent pulmonary valve replacement at a mean age of 14.16 years. Mean age at last follow up for echocardiograms was 11.9 years, and for MRI it was 13.2 years. All SNPs were intronic and in Hardy–Weinberg equilibrium. SNP c.462+181G > A in CHD1L gene was associated with lower RVEF (mean = 40.8 ± 5.3%) versus the GG genotype (mean = 49.7 ± 6.4%, $p < 0.0007$). SNP c.127+3084C > T in CHD1L was associated with lower RVEF (mean = 40.8 ± 5.3%) compared to CC genotype (mean = 50.2 ± 6.3%, $p < 0.0004$). SNP c.344-1874G > A in TNNI3K gene was associated with higher RVESVi (mean = 94.5 ± 40.3 ml/m^2) versus the GG genotype (mean = 73 ± 1.4 ml/m^2, $p < 0.0036$).
Conclusion: Genetic variants associated with TOF causation also influenced RV remodelling after TOF repair. Genotype knowledge may help early identification of at-risk patients for optimisation of medical and/or surgical management.

673: SPECTRUM OF CONGENITAL HEART DISEASE AMONG CHILDREN PRESENTING TO THE UGANDA HEART INSTITUTE, KAMPALA, UGANDA

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Congenital heart diseases are among the commonest birth defects globally. The echo diagnoses of children who presented to the Uganda Heart Institute were reviewed over a period of five years (January 2007 to December 2011). The Uganda Heart Institute is a super speciality centre located within Mulago Hospital, the country's main referral hospital. It is the main centre that offers peadiatric cardiac services in the country.
Objectives: (1) To describe the common congenital cardiac lesions among children in Uganda, (2) to establish a registry for congenital heart disease. To date no large-scale database exists.
Methods: An ongoing registry has been compiled since 2007 by generating echo reports and storing them digitally. These records were retrieved and analysed. Data were compiled in simple tabular form and percentages.
Results: Out of a total of 3 849 children with an echo diagnosis of heart disease, 2 663 children were found to have a congenital cardiac defect. VSD was the commonest acyanotic heart defect (26.5%) and tetralogy of Fallot was the most common cyanotic heart defect (7.0%). There was a much higher prevalence of truncus arteriosus in our series (4.6%) compared to the reported prevalence of about 1–2% in most studies. Coarctation of the aorta was almost non-existent (0.26%).
Conclusion: The prevalence of congenital heart disease is as common as that reported elsewhere. There may be regional differences in the type of defects seen. Further studies are needed to ascertain whether genetic or environmental factors account for these variations.

681: CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES: SINGLE-CENTRE EXPERIENCE

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Background: There are several rhythm and conduction disturbances associated with congenitally corrected transposition of the great arteries (ccTGA). The purpose of this study was to determine the incidence of rhythm and conduction disturbances in ccTGA patients with two adequate-sized ventricles.
Methods: Retrospective analysis of the records of 49 patients from a single centre were reviewed to determine long-term results of ccTGA patients.
Results: The study comprised 49 patients (15 girls, 34 boys). The median age of the patients at initial presentation was three months (one day to 34 years) and mean period of follow up was 4.5 ± 1.8 years (one month to 22 years). Forty-seven of them had associated heart anomalies. The most common associated lesion was VSD (38 patients). Pulmonary valve abnormalities were second most common lesion. Pulmonary stenosis was more common than pulmonary atresia (17 vs six patients). As usual, Ebstein anomaly and tricuspid regurgitation were quite common among our patients. During the follow-up period, 18 patients had a total of 22 operations. Systemic-to-pulmonary circulation shunts were the most common procedures (nine patients). Conventional biventricular repair and double-switch procedure were performed equally (5/5 patients). Tricuspid valve replacement was performed in two patients. At initial examination, two patients had first-degree AV block, one second-degree AV block and one congenitally complete AV block, Additionally, one patient had atrial ectopic rhythm, and one left bundle branch block. Supraventricular tachycardia was detected in three patients. At follow up, complete AV block developed in five patients after intracardiac surgery. Pacemaker implantation was required for these patients and one patient with congenitally complete AV block.
Conclusions: Patients diagnosed as ccTGA should be followed lifelong. During the disease course they may need different types of surgical procedures and ccTGA may complicate with different types of rhythm and conduction disturbances at any time.

688: THE UTILITY OF TRANSIENT ELASTOGRAPHY TO ASSESS FOR HEPATIC FIBROSIS IN PAEDIATRIC FONTAN PATIENTS

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Background: Hepatic fibrosis (HF) is a recognised complication following Fontan surgery and heralds long-term risk for cirrhosis, with significant morbidity. While liver biopsy is considered the gold standard to assess for HF, it is invasive and potentially life-threatening. Transient elastography (elastography) is a novel diagnostic tool that offers a rapid, non-invasive method for monitoring HF. The device measures liver stiffness by transmitting a vibration to determine the velocity of an elastic shear wave propagated through liver tissue. For the diagnosis of HF, elastography cut-off values range from 7.1–8.8 kPa. Few reports have examined elastography in post-operative paediatric Fontan patients.
Objective: To measure and compare liver stiffness in post-operative paediatric Fontan patients with age-matched controls utilising elastography.
Methods: Fontan patients (n = 14) and controls (n = 28) were enrolled at cardiology and GI clinics at British Columbia Children's Hospital. Demographic information, echocardiography and cardiac catheterisation data were collected. Elastography measurements using age and size-appropriate imaging probes were obtained.

Results: The age of the Fontan cohort was 11.9 (5.9–16.7) years. The interval from Fontan surgery to the elastography scan was 8.2 (1.0–13.5) years. None of the Fontan patients were in cardiac failure, only one had a persisting fenestration, and six had hepatomegaly. All had an extra-cardiac repair. Liver enzyme values were higher in the Fontan group (ALT 30 vs 16 U/l; $p < 0.003$ and GGT 49 vs 11 U/l; $p < 0.0001$). Elastography values were significantly higher in Fontan patients compared with controls (17.1 (11–39) vs 4.7 kPa (3-6), $p < 0.0001$). There was no association between elastography values and patient age or time since Fontan surgery.
Conclusions: Elastography is a feasible non-invasive method to assess liver stiffness in children following Fontan surgery. Paediatric Fontan patients have markedly elevated liver stiffness, suggesting a high risk for advanced HF. Elastography has important utility in the follow up of paediatric Fontan patients.

693: OUTCOMES OF PAEDIATRIC PULMONARY HYPERTENSION: DATA FROM THE SPANISH REGISTRY FOR PAEDIATRIC PULMONARY HYPERTENSION (REHIPED)

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Objectives: There is sparse data on epidemiology and survival in paediatric pulmonary hypertension.
Methods: From January 2009 to June 2012, 21 referral and non-referral centres collected data of 225 patients, with mean pulmonary artery pressure (mPAP) > 25 mmHg and pulmonary vascular resistance index (PVRI) > 3 WU/m^2. We included all Dana point (DP) aethiologies (groups I to V) and analysed differences in survival according to DP aethiological group, age at diagnosis, functional class (FC), syncopes, chromosomopathy, right atrial pressure (RAP), mPAP, cardiac index (CI), PVRI, and PVRI/SVRI.
Results: Patients were classified as: group I (PAH, $n = 142$, 61%), II (left heart disease, $n = 31$, 14%), III (respiratory disease, $n = 41$, 18%, half of them with bronchopulmonary dysplasia), IV (thromboembolic PH, $n = 2$, 1%), V ($n = 10$, 4.5%, mostly inherited metabolic diseases), but 31% had multifactorial PH. Median age at diagnosis was 4.3 ± 4.9 years (50% < 2 years); 21% were premature, and chromosomopathies/syndromes were present in 38%. Functional class at diagnosis was III/IV in 53%, without significant differences in mPAP, RAP, PVRI/SVRI or CI between the different DP groups. For the whole cohort, one- and three-year survival was 80 and 74%. Mortality risk factors (univariate analysis): DP aethiology group (three-year survival, 81% for PAH, 60% for left heart disease PH, 58% for lung disease PH, and 15% for Dana point group V, $p < 0.001$), functional class at diagnosis ($p < 0.001$), RAP ($p = 0.006$), and age at diagnosis (three-year survival, 60% for < 2 years, 92% between 2 and 8 years, and 85% for > 8 years, $p < 0.001$). Multivariate analysis: aethiology ($p < 0.001$), age at diagnosis ($p < 0.001$), FC at diagnosis ($p < 0.001$) and RAP ($p = 0.002$).
Conclusions: Age at diagnosis was a significant risk factor: there was worse survival in younger patients (often frequently missed in referral centre-based registries). In severe paediatric PH, prognosis was better in PAH than in the other DP aethiological groups.

698: CHALLENGES FOLLOWING SURGERY IN CONGENITAL HEART DEFECTS: INCREASING INCIDENCE AND IMPROVED SURVIVAL IN COMPLEX LESIONS

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Background: Few congenital heart defects (CHD) are now beyond the reach of surgical treatment. There are limited data on the impact of recent surgical development on the composition of the CHD population.
Methods: Between 1971 and 2010, the 7 018 patients aged 0–16 years undergoing corrective CHD surgery at Oslo University Hospital, Rikshospitalet (80% of all CHD operations in Norway), were prospectively enrolled. Primary CHD diagnoses were categorised according to a consensus-based severity hierarchy. Date of first operation was noted, and complete all-cause mortality data were recorded up to 1 January 2012.
Results: Number of patients increased from 1 233 in 1971–1980 to 2 286 in 2001–2010. The proportions of patients with tetralogy of Fallot (TOF; 7.7% in 2001–2010), transposition of the great arteries (TGA; 7.9%), univentricular hearts (UNI; 5.1%) and ventricular septal defect (VSD; 16.3%) did not change. However, there was a marked increase in the proportions of patients with hypoplastic left heart syndrome (HLHS), atrioventricular septal defect (AVSD), pulmonary atresia (PA) and interrupted aortic arch (IAA) (3.6% in 1971–1980, and 25.4% in 2001–2010), and a decrease in surgical treatment of anatomically simpler defects. Overall 30-day postoperative survival improved from 89.3% in 1971–1980 to 97.5% in 2001–2010, with particularly marked changes in groups with complex lesions. Among the 1 011 patients operated in 1971–1980 who survived beyond 16 years, 50 (4.5%) had HLHS, AVSD, IAA, PA or UNI. In contrast, among the 1 682 patients operated in 1991–2000 surviving beyond 16 years or until 1 January 2012, 344 (17.6%) had such complex defects.
Conclusion: The number and proportion of patients with complex CHD surviving childhood are increasing rapidly, since more of these defects are treated surgically, and with substantially improved survival.

699: MANAGEMENT AND CLINICAL FEATURES OF PAEDIATRIC PULMONARY ARTERIAL HYPERTENSION IN SPAIN: DATA FROM THE SPANISH REGISTRY FOR PAEDIATRIC PULMONARY HYPERTENSION (REHIPED)

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Objectives: There is sparse information on paediatric PAH, but referral centre-based registries offer data on selected populations. We collected data on clinical features, management and outcomes of paediatric PAH in Spain from the REHIPED registry.
Methods: From January 2009 to June 2012, 21 centres (referral and non-referral) included cases, aged two months to 18 years at diagnosis, with mean pulmonary arterial pressure mPAP > 25 mmHg, pulmonary vascular resistance index (PVRI) > 3 WU/m^2, and pulmonary wedge pressure < 15 mmHg.
Results: We included 142 patients: congenital heart disease (PAH/CHD, $n = 105$, 74%), idiopathic/familial (IPAH, $n = 32$, 21%), portopulmonary ($n = 3$, 2.1%), HIV infection ($n = 1$, 0.7%), connective tissue disease ($n = 1$, 0.7%); 42% were prospective cases. Mean age at diagnosis: 5.2 ± 4.8 years (36% patients < 2 years.), age at inclusion: 8.7 ± 6.5 years. Female/male ratio: 1.2. Syndromal anomalies were present in 44%; 51% cases were in functional class (FC) III/IV at diagnosos. Haemodynamic data: mPAP 46 ± 18 mmHg, PVRI 8.7 ± 7.8 WU/m^2, PVRI/SVRI 0.7 ± 0.5, cardiac index (CI) 4.6 ± 2l /min/m^2. Only 6% of IPAH children were responders in

the vasoreactivity test. Of the whole group, 92% received PH drugs (46% in combination, 23% prostanoids), 7% oral anticoagulation and 14% anti-aggregation. For the whole group, one-, three- and five-year survival was 89, 85 and 79%, respectively, without differences between IPAH and PAH/CHD. Mortality risk factors (univariate analysis): younger age at diagnosis ($p = 0.013$), FC III/IV ($p < 0,001$), CI ($p = 0.005$) and RAP ($p = 0.014$). Multivariate analysis: age at diagnosis [HR 0.84 (95% CI: 0.73–0.96), $p = 0.009$], FC ($p < 0.001$), IC [HR 0.46 (95% CI: 0.3–0.7), $p < 0.001$], RAP (HR 1.2 (95% CI: 1.06–1.38), $p = 0.012$].

Conclusions: REHIPED registry patients had a younger age at diagnosis, higher percentage of prospective cases, and lower percentage of IPAH responders than other referral-centre registries, but similar survival, with high prevalence of combination therapy. In paediatric PAH, besides the already known risk factors (FC, CI, RAP), younger age at diagnosis was also a risk factor for mortality.

704: CLINICAL EXPERIENCE OF SUBCUTANEOUS AND TRANSVENOUS IMPLANTABLE CARDIOVERTER-DEFIBRILLATORS IN YOUNG PATIENTS

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Background: Subcutaneous ICDs do not require a lead to be in contact with the heart. Potential advantages over transvenous ICDs are the elimination of vascular complications and reduction of lead fractures. We compared the short-term performance of subcutaneous and transvenous ICDs in patients < 20 years old.

Methods: The study was a retrospective, observational study of all patients < 20 years of age who received an ICD over a four-year period (July 2007 to July 2011) in the west of Scotland. Baseline characteristics, complications and ICD therapy were recorded. The primary outcome measure was survival. The secondary outcome measure was survival free from inappropriate ICD therapy or system revision.

Results: Nine patients received a subcutaneous ICD and six patients received a transvenous ICD. Underlying diagnoses and age at implant were similar for the two groups. There were no implant complications with subcutaneous ICDs, but one patient sustained a pneumothorax during transvenous ICD implantation. Median follow up was 20 months (range 12–32 months) for subcutaneous ICDs and 36 months (range 24–55 months) for transvenous ICDs. Appropriate shocks for VF occurred in two patients with a subcutaneous ICD and one with a transvenous ICD. No subcutaneous ICD required revision, but two transvenous ICDs were extracted due to infection ($n = 1$) and lead fracture ($n = 1$). Survival was 100% in both groups. Survival free from inappropriate ICD therapy or system revision was 89% for subcutaneous ICDs and 25% for transvenous ICDs (log rank test, $p = 0.0237$).

Conclusions: In our series of young patients, subcutaneous ICDs performed well on short-term follow up, with a lower incidence of inappropriate shocks and complications requiring system revision, compared to transvenous ICDs. In the absence of randomised trials, subcutaneous ICDs should be compared prospectively with transvenous systems in large multi-centre registries with comparable periods of follow up.

705: PREVALENCE OF MYOCARDIAL FIBROSIS BY CARDIAC MAGNETIC RESONANCE IMAGING PREDICTS CLINICAL PRESENTATION AND OUTCOME IN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY

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Background: Patients with hypertrophic cardiomyopathy (HCM) frequently demonstrate non-ischaemic pattern myocardial fibrosis (MF). Cardiac magnetic resonance (CMR) imaging with late gadolinium enhancement (LGE) can detect a small and focal MF. The aim of the study was to assess the occurrence of MF in children with HCM and to evaluate its clinical significance.

Methods: Fifty-six children with HCM underwent CMR from January 2010 to June 2012. All 56 patients were divided into two groups: group I: 30 (54%) children, mean age 12.7 years with myocardial fibrosis by LGE and group II: 26 (46%) patients, mean age 11.3 years without MF. Patients' demographics, clinical symptoms as well as the results of LGE CMR were analysed and compared between the groups.

Results: The patients in the fibrosis group more often had symptoms such as syncopes (13 vs 4%), presyncopes (37 vs 23%), chest pain (43 vs 23%), and fatigue (80 vs 65%). Among children with MF, LV wall thickness (mean 22 vs 14 mm; $p = 0.0001$) and LV mass index (mean 105 vs 86 g/m²; $p = 0.003$) were significantly greater compared with those without MF. In 79% patients in group I and in 36% in g roup II, LV mass was increased ($p = 0.003$). In group I, nsVT episodes were more frequent than in group II (10 vs 4% of patients). Patients in both groups differed significantly regarding the occurrence of major risk factors for SCD (67 vs 31%; $p = 0.016$). LV wall thickness ≥ 30 mm and cardiac arrest were observed only in group I (20 and 7%, respectively; $p = 0.025$), and family history of SCD was more frequently positive (30 vs 15%). In group I, 13% of patients underwent surgical myectomy compared with 4% in group II. ICD was implanted only in children with MF (33%; $p = 0.001$), in two patients as secondary prevention and in eight as primary prevention.

Conclusion: The presence of myocardial fibrosis was related to an unfavourable clinical course of HCM in children. Visualising LGE in CMR may contribute to better risk stratification of SCD and may help in taking therapeutic decisions in HCM patients.

707: PULMONARY HYPERTENSION IN BRONCHOPULMONARY DYSPLASIA: CLINICAL FINDINGS, ASSOCIATED CARDIOVASCULAR ABNORMALITIES AND OUTCOMES

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Background: Severe pulmonary hypertension (PH) worsens the prognosis of bronchopulmonary dysplasia (BPD). Clinical characterisation, incidence of cardiovascular abnormalities (CVA) and outcome in this setting have not yet been fully established.

Methods: This was a retrospective study of the clinical records, CT imaging and catheterisation data of 29 patients with BPD and PH referred to our pulmonary hypertension unit from March 2006 to December 2011. Median gestational age was 27 weeks (IQR 24–27.3) and median birth weight 740 g (IQR 620–830). Sixteen were male and 13 female (ratio 1.2).

Results: PH diagnosis was made at a median age of 4.5 months (IQR 2.4–7.8), in 48% after initial hospital discharge, with echocardiography estimating the median right ventricular pressure/systemic pressure ratio of 70% (IQR 60–80%). CT scan was performed in 21 patients and catheterisation in 13, finding CVA in 19 patients (65.5%): nine systemic-to-pulmonary collaterals, seven pulmonary vein stenosis, four ASD, one restrictive VSD and nine PDA. Haemodynamic data were (median and IQR): pulmonary arterial systolic pressure 78.5% of systemic (57.8–94.7), PVR 4.6 UW/m²

(2.7–7), PVR/SVR 0.48 (0.3–0.8), transpulmonary gradient 32 mmHg (20–40.5). At a median follow up of 35 month (IQR 17–84): seven patients had undergone closure of shunts, 21 received specific drug treatment for PH (PH improvement or resolution in nine), three showed spontaneous improvement of their PH, and eight (26%) died.
Conclusion: Severe PH in BPD carries a bad prognosis and its diagnosis is sometimes delayed. Prompt diagnosis, accurate detection of CVA, early shunt closure and aggressive specific drug therapy can improve the outcome of these patients.

710: REDUCING THE PAIN OF BENZATHINE PENICILLIN INJECTIONS IN THE RHEUMATIC FEVER POPULATION

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Background: In the Counties Manukau District Health Board (CMDHB) region, 405 rheumatic fever (RF) patients were offered pain management with their monthly benzathine penicillin injections. It was hypothesised this would reduce the pain and fear associated with this injection.
Aims: To evaluate the effectiveness of pain management for benzathine penicillin injections.
Methods: A BUZZY vibrating cold pack device and 0.25 ml of 2% lignocaine, mixed with the benzathine penicillin prior to administration, were offered to patients. A survey was conducted evaluating pain scores at four time points (delivery, 2 min post, 1 hour post, next day). Fear of the injection and duration of that fear were also evaluated.
Results: Just under half of patients (49%) responded. Pain at injection delivery and fear of injection scores were significantly higher for patients under 16 years compared with older patients. Paired data pre- and post-intervention were available (n = 119). Mean pain score at delivery changed from 5.4/10 pre-intervention to 2.4/10 post-intervention ($p \leq 0.001$). Pain scores were significantly reduced at all four time points, as was fear of the injections. Lignocaine plus BUZZY resulted in a greater improvement in pain score than lignocaine alone during injection delivery. A separate file audit conducted five months after the study found that 66% of 405 RF patients were using lignocaine, 43% were using BUZZY and 73% were using one or both interventions.
Conclusions: Offering analgesia with benzathine penicillin injections has been popular with the RF population in CMDHB and is associated with reduced pain and fear.

711: THE USEFULNESS OF SCINTIGRAPHY WITH ^{99M}TC ANTI-GRANULOCYTE ANTIBODY FOR DIAGNOSIS AND FOLLOW UP IN CHILDREN WITH MYOCARDITIS

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Background: Clinical diagnosis of myocarditis is difficult due to variable presentation ranging from asymptomatic cases to acute heart failure. Endomyocardial biopsy (EMB) is an invasive procedure. Therefore, a non-invasive diagnostic method is highly desirable, especially in children. The aim of the study was to investigate whether scintigraphy with ^{99m}Tc-anti-granulocyte antibody (^{99m}Tc-AGA) would be useful for diagnosis and follow up in children with myocarditis and to determine its correlation with EMB and clinical features.
Methods: From 2005 to 2010, 11 children, aged 6.6 to 17 years, mean 13 ± 8 years, presenting with symptoms of myocarditis were evaluated at the time of initial presentation and six, 12 and 24 months after the first study. Patients' demographics and clinical symptoms, as well as results of echocardiography, electrocardiography, EMB and scintigraphy with ^{99m}Tc-AGA were analysed. In all patients, myocardial scintigraphy was performed with estimation of antigranulocyte antibody uptake. EMB was done in 10 patients at the time of the initial presentation and in eight patients after six months.
Results: Out of 11 patients, in 10 (91%), positive antigranulocyte uptake was observed, with EMB confirming myocarditis in eight (80%) children. With scintigraphy, after six months, positive uptake was found in nine (82%) patients, with EMB performed in eight patients showing persistent myocarditis. After 12 months, scintigraphy indicated positive uptake in seven (64%) and after 24 months only in four (36%) patients. The mean LVEF was 59 ± 12% at initial presentation and 61 ± 10, 65 ± 12 and 66 ± 7% after six, 12 and 24 months respectively.
Conclusion: (1) In 80% of patients with positive scintigraphy results, biopsy-proven myocarditis was observed. (2) The positive antigranulocyte uptake correlated with clinical features at diagnosis and at follow up. (3) The control scintigraphy performed at follow up after six, 12, and 24 months allowed us to evaluate resolved or persistent myocarditis. (4) Myocardial scintigraphy results indicate that the inflammatory process in the myocardium decreased significantly after 12 months of the onset of the disease.

714: USE OF A HANDHELD ECHOCARDIOGRAPHY MACHINE IN LARGE-SCALE SCREENING OF NEWBORNS FOR CONGENITAL HEART DEFECTS

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Background: Early identification of congenital heart disease (CHD) is desirable in international settings where limited surgical resources necessitate longer planning periods. Use of large-scale screening would facilitate this goal, but is hampered by limited resources, lack of expertise and infrastructure.
Methods: A convenience sample of consecutive newborns within a six-day window was screened at Duhok Teaching Hospital (Kurdistan, Iraq) as part of a Ministry of Health-directed demonstration project. All infants were first screened with a VScan® handheld device (VS; assessment of 2D and colour-flow data) and then using a Seimen's Cypress® echocardiogram machine (SC; assessment of 2D and colour Doppler data – real-time and spectral). Diagnostic accuracy of the handheld device was sought. Patent ductus arteriosus (PDA) and patent foramen ovale (PFO) were considered normal findings.
Results: A total of 178 newborns were screened out of 276 live births (64.5%) with both ultrasound machines. VS diagnosed 16 newborns with an abnormal screen, compared with 24 for SC. VS and SC found 150 and 154 PDAs respectively. Both diagnosed 167 of 178 newborns with PFOs. VS diagnosed two of three infants with muscular ventricular septal defects (VSD) identified by SC. Both machines diagnosed five infants with trivial aortic insufficiency. VS sensitivity for diagnosing a PDA and PFO was 97 and 100%, respectively. PDAs missed by VS screening were considered trivial. VS sensitivity for diagnosing muscular VSD was 67%. The positive and negative predictive values

for an abnormal screening ultrasound were 100 and 95%, respectively. VS sensitivity for diagnosing a shunting lesion was 97%. No infants were found with significant CHD in this evaluation.
Conclusions: VS screening appears to have sufficient specificity to exclude significant CHD in experienced hands; sensitivity for surgically relevant defects beyond the PDA requires additional study.

715: INCIDENCE OF CONGENITAL HEART DEFECTS IN DUHOK, IRAQ

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Background: In Iraq, there have been various reports of an increased incidence of congenital heart defects above the historical world average of 0.8% of all live births. However, there have been no systematic studies to capture a true incidence of congenital heart disease (CHD) in Iraq.
Methods: During the week of 19 to 25 May 2012, a convenience sample of 176 newborns was screened at Azadi Teaching Hospital in Duhok, Iraq, by two paediatric cardiologists, with a complete echocardiogram (echo) as part of a demonstration project under the Ministry of Health. Patent foramen ovale (PFO) and patent ductus arteriosus (PDA) were considered normal and not recorded as defects.
Results: A convenience sample of 176 of 276 live-born infants (64%) was screened during the study period. Of those screened, 24 (13.6%) were found to have at least one abnormality on echo, some had up to three. There were nine structural defects (5.1%), which included three cases of tiny muscular ventricular septal defect (VSD) (1.7%), two cases of atrial septal defect (ASD) (1.1%), three cases with dysplastic aortic valves (1.7%), one aneurysmal atrial septum (0.6%), one dysplastic tricuspid valve (0.6%), and one bicuspid aortic valve (0.6%). None of these defects were considered haemodynamically significant.
Conclusion: This study is the first of its kind to attempt to screen newborns with echo in a single hospital to obtain a true incidence of CHD in Iraq. The overall incidence of structural congenital heart defects was found to be 5.1%. If only VSDs and ASDs were counted, an incidence of 2.8% is clearly higher than would be expected. Systematic efforts at population-based screening in newborns are warranted, both to define the true incidence of CHD as well as to delineate any type-specific variations in expected disease rates.

717: EPIDEMIOLOGY OF PULMONARY ARTERIAL HYPERTENSION IN A SPANISH PAEDIATRIC POPULATION: DATA FROM THE SPANISH REGISTRY FOR PAEDIATRIC PULMONARY HYPERTENSION (REHIPED) AND SPANISH REGISTRY FOR PULMONARY ARTERIAL HYPERTENSION IN ADULTS (REHAP)

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Nowadays there is a lack of data regarding the epidemiology of paediatric pulmonary arterial hypertension (PAH). In order to provide valuable epidemiological data about paediatric PAH in Spain, we collected clinical data of patients diagnosed with PAH from two months to 18 years old from the REHIPED and REHAP registries.
Methods: Voluntary reporting (from referral and non-referral centres) of their paediatric patients (aged 2 months to 18 years) diagnosed with PAH was used. Consecutive cases diagnosed from January 2009 to June 2012 were selected to provide data on average annual incidence from 2009 to 2012, and point prevalence (June 2012). PAH was defined by a mean pulmonary artery pressure (PAP) > 25 mmHg, pulmonary vascular resistance (PVR) > 3 Wood units (WU) and pulmonary capillary wedge pressure < 15 mmHg.
Results: A total of 205 PAH patients (mean age 8 ± 5.7 years) was reported in both registries: iPAH (idiopathic, n = 57), PAH/CHD (congenital heart disease PAH, n = 135), veno-occlusive (n = 2), portopulmonary (n = 4), HIV (n = 1), connective tissue disease (n = 2), others (n = 4). Yearly incidence rates for PAH diagnoses were 2.56 ± 0.25 cases/million/year. For iPAH and CHD-PAH these rates were respectively 0.49 ± 0.29 and 1.87 ± 0.18 cases/million/year. We estimated a point prevalence (June 2012) of 14 cases/million for PAH, 2.97 cases/million for iPAH, and 10. cases/million for HAP/CHD.
Conclusions: The estimated incidence and prevalence of PAH, IPAH and PAH/CHD in the paediatric Spanish population was comparable to those provided for this age range by other European countries with mandatory registration. Congenital heart disease is the most common cause of PAH in patients from two months to 18 years.

718: DETERMINATION FACTOR OF PULMONARY-TO-SYSTEMIC FLOW RATIO IN ATRIAL SEPTAL DEFECT

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Background: Left ventricular (LV) diastolic stiffness has been postulated as an important determinant of pulmonary-to-systemic flow ratio (Qp/Qs) in atrial septal defect (ASD). However, to date, there are no data to directly support this. Therefore, the present study tested the hypothesis that increased LV diastolic stiffness is an independent determinant of increased Qp/Qs in ASD.
Methods and Results: Study subjects were 192 consecutive ASD patients who underwent catheter closure of ASD with the Amplatzer septal occluder. Patients' age ranged from four to 78 years old, and the average Qp/Qs was 2.38 ± 0.90. The size of the defect was determined as the diameter of the deployed device, and was normalised to body surface area of the patient. LV diastolic stiffness was calculated as follows: (LV end-diastolic pressure – LV diastolic minimum pressure)/stroke volume index. Univariate analysis showed that Qp/Qs was significantly and positively correlated with the size of defect (r = 0.49, $p < 0.0001$) and LV diastolic stiffness (r = 0.29, $p < 0.0001$). Multivariate analysis also demonstrated that the size of ASD (β = 0.45, $p < 0.0001$), LV stiffness (β = 0.28, $p < 0.0001$), RV stiffness (β = –0.21, p = 0.001) and age (β = 0.25, p = 0.001) were independent determinants of Qp/Qs (r = 0.65, $p < 0.0001$).
Discussion: The present study for the first time confirmed that increased LV stiffness independently increases Qp/Qs in ASD. Because increased Qp/Qs due to an increased LV stiffness causes RV dilation and further increase in the LV stiffness via a ventricular interaction, this could explain the progressive nature of this disease.

721: NEUROCOGNITIVE OUTCOMES OF INFANTS WITH SINGLE-VENTRICLE PHYSIOLOGY SEEN IN A COMPLEX CONGENITAL HEART DISEASE CLINIC

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Background: Earlier intervention and exposure to various perioperative techniques and care have contributed to improved survival rates, but may also adversely impact on future developmental processes and outcomes. Specific aims were: (1) determination of which medical characteristics of the baby are predictive of which

neurodevelopmental outcomes, (2) determination of changes in an individual baby's neurodevelopment over time, and (3) description of the neurodevelopmental outcomes of an aggregate population of infants with congenital cardiac disease and early surgical repair, followed by timely BSID at CCHDC.
Methods: This study retrospectively reviews the charts of infants with single-ventricle physiology after surgical repair. The purpose of the study was to identify the influence of peri-operative procedures, including time on ECHMO post-operatively, length of stay, certain patient characteristics on neurocognitive outcomes, as well as oral feeding development, oxygen saturations and weight gain.
Results: A linear mixed-effects model will be used to model BSID subscale scores and their trend over time. Predictor variables will include subscale measured (cognitive, language or motor), time (or a non-linear function thereof), medical predictors such as diagnosis or co-morbid conditions, and demographic covariates. Any significant interactions among these predictors will also be included in the final model. A covariance structure suitable to model the within-subject and within-subscale dependence will be chosen at the time of analysis based on model fit.
Conclusions: Since the inception of our clinic, all infants with single-ventricle physiology have been followed closely for medical recovery as well as weight gain, feeding development and neurocognitive outcomes. The final mode will provide insight into the neurocognitive outcomes of this vulnerable population.

727: UTILITY OF PULSE OXIMETRY AND BEDSIDE ECHOCARDIOGRAPHY IN IDENTIFYING CONGENITAL HEART DISEASE IN NEWBORNS

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Background: Congenital cardiac malformations (CCM) in newborns may be difficult to diagnose clinically. Pulse oximetry has been shown to have better sensitivity and specificity, but echocardiography remains the gold standard. We carried a prospective study to determine the utility of pulse oximetry in identifying CCM in newborns in a community hospital setting.
Methods: All 20 305 (male-to-female ration 1:0.9) consecutive neonates born over specific period in a large community hospital were included in the study. Recruitment was done within 48 hours of birth. We noted cyanosis, murmurs or abnormal heart sounds, femoral pulse and respiratory distress. The arterial saturation was measured non-invasively by pulse oximetry. All the neonates underwent a screening bedside echocardiogram using a portable machine.
Results: Of 20 305 neonates screened, 151 were found to have significant CHD by echocardiography (7.4/1 000, 95% CI: 6.3–8.6/1 000). An additional 851 babies had insignificant CHD, small muscular VSD being the commonest lesion seen in 663 babies. Major cardiac defects needing early intervention, including transposition of the great arteries, hypoplastic left heart, large VSD and pulmonary atresia were identified in 64 neonates (3.1/1 000 live births); 1 599 (7.9%) newborns had a resting arterial saturation of < 92%. The sensitivity and specificity of clinical examination for diagnosing CHD was 14 and 97%, respectively.
Conclusion: The prevalence and spectrum of significant CHD in a community-level hospital in India is not very different from that reported from the West. In our study, clinical examination had a low sensitivity for diagnosis of CHD in newborns. Oximetry performed within 24 hours of life also had a poor sensitivity for diagnosing critical CHD. However sensitivity improved markedly in cases where oximetry was done beyond 24 hours of life.

729: LÖFFLER MYOCARDIOPATHY IN A NINE-YEAR-OLD BOY

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Background: Löffler myocardiopathy is a restrictive cardiomyopathy associated with eosinophilia. Eosinophilic states that may occur in association with Löffler myocardiopathy include hypereosinophilic syndrome, eosinophilic leukaemia, carcinoma, lymphoma, drug reactions or parasites.
Case report: We present a nine-year-old male patient who was admitted to Instituto Materno de Medicina Integral Professor Fernando Figueira (IMIP) in June 2005 with pallor of a month's duration, lower back ache for one week and fever for five days prior to admission. His vital signs included heart rate = 140 bpm, respiratory rate = 28 bpm, blood pressure = 90/60 mmHg. On physical examination his general state was found to be compromised, and he had tachypnoea (+/4+), pallor (3+/4+). There was no sign of cyanosis and pulses were present with normal amplitude. Heart rhythm was irregular due to extrasystoles (20/minute), with no murmurs or thrills. The lungs were clear to percussion and auscultation. The abdomen was soft and non-tender, with normal bowel sounds and hepatomegaly palpable 6 cm from the right costal margin. Joint examination was normal. Laboratory findings included microcytic and hypochromic anaemia with haemoglobin of 5 gm/dl, thrombocytopenia with platelet count of 47 000 /mm^3 and leukocytosis with white blood cells 74 000 /mm^2 and 83% eosinophils.

Myelography revealed hyperplasia of the eosinophil lineage and 15% lymphoblast infiltration. Chest radiography showed cardiomegaly with normal pulmonary flow and electrocardiography a diffuse alteration of the ventricular repolarisation. The echocardiogram revealed an enlarged right atrium and inferior vena cava with mild to moderate pericardial effusion, mild tricuspid regurgitation and a mass in the right ventricle suggestive of a thrombus. Magnetic resonance revealed imaging consistent with Löffler myocardiopathy in its biventricular fibro-thrombotic state. He was submitted for another myelography which was compatible with hypereosinophilic syndrome due to acute lymphocytic leukaemia. Anticoagulation with warfarin was started and he was referred to paediatric oncology to begin chemotherapy.

733: ECHOCARDIOGRAPIC CHANGES DURING AN EPISODE OF ACUTE RHEUMATIC FEVER

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Background: The purpose of this study was to evaluate the usefulness of echocardiography, including three-dimensional echocardiography, to assess the structural and functional changes in mitral valve apparatus during an episode of acute rheumatic fever.
Methods: Twenty-two consecutive patients with acute rheumatic fever (mean age 12.15 ± 4.2 years) with carditis, satisfying WHO diagnostic criteria, were enrolled in the study. Annular diameters, leaflet surface area, tent height, tent volume, leaflet thickness, leaflet volumes and submitral volumes were measured. The follow-up echocardiograms were repeated after four weeks of steroid therapy. Age-matched chronic rheumatic mitral regurgitation patients were included as controls.
Results: Mitral regurgitation was severe in eight (36%), moderate in 12 (55%), and mild in two (9%) patients. Three-dimensional echocardiography showed a diffuse nodular appearance of mitral leaflet surface during activity. The functional class improved on follow up after treatment with steroids. However, there were no significant changes in echocardiographic parameters except the LV end-diastolic dimensions, which decreased from 54.17 ± 9.75 mm at baseline to 52.5 ± 10.18 mm on follow up ($p = 0.02$). The thickness the anterior/

posterior leaflets at the tip, middle and base of during acute activity was higher than in the controls. The thickness of the mid-part of the anterior mitral leaflet was 5.1 ± 0.6 mm in the study group initially and 4.9 ± 0.96 mm on follow up, while the control group had a thickness of 3.87 ± 0.8 mm ($p = 0.008$).
Conclusions: Three-dimensional echocardiography complements 2D echocardiography in the evaluation of patients presenting with rheumatic fever. Nodular appearance and leaflet thickening are important echocardiograpic features of rheumatic carditis. The treatment with steroids may result in some favourable changes on left ventricular dimensions measurable by echocardiography.

734: LEFT ATRIAL MIXOMA MIMICKING REUMATHIC FEVER

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Background: Left atrial myxoma is rare in childhood and may present in a variety of forms, including a fever of unknown origin, symptoms related to peripheral embolisation, acute congestive heart failure or symptoms indistinguishable from collagen-vascular diseases. Sometimes signs and symptoms of mitral valvar disease dominate the clinical presentation.
Case report: We present an 11-year-old female admitted to Instituto de Medicina Integral Professor Fernando Figueira with intermittent fever associated with dyspnoea on moderate exertion for six months. Progressive symmetric arthralgia in the knees, followed by the ankles, impaired her walking. She had asthenia and diffuses abdominal pain. She did not have any history of repeated episodes of tonsilitis. The patient had had a diagnosis of rheumatic fever in another city and had used benzathine penicillin for six months before referral to us.

On admission she had fever, pallor, finger clubbing and watch-glass nails. Heart rhythm was regular with a murmur of mitral insufficiency and mitral stenosis. The abdomen examination showed splenomegaly. She had oedema and pain with any movement of her knees and left ankle. Electrocardiography presented signs of left atrial overload and interatrial block. Echocardiography showed a mild enlarged left atrium, moderated mitral regurgitation and a tumour adhered to the left surface of the interatrial septum, protruding into the left ventricle during diastole.

The patient underwent surgical resection of the mass and the histopathology was compatible with a myxoma. She was discharged from hospital on medication. Four months after the surgery she was re-admitted with heart failure. Echocardiography showed both left atrial and ventricle overload and thickened mitral valve with severe mitral regurgitation. She was submitted for an implant of a prosthetic valve. Histopathology showed fibrosis with areas of calcification and neutrophilic exudation. Postoperative course was uneventful and the patient was discharged from hospital seven days after surgery.

737: PROPRANOLOL IN INFANTS WITH VENTRICULAR SEPTAL DEFECT AND HEART FAILURE: VSD-PHF (PROPRANOLOL FOR HEART FAILURE) TRIAL

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Background: Infants with ventricular septal defect (VSD) may develop congestive heart failure in spite of conventional medical therapy. We investigated the effects of additional beta-blockade in such infants in a randomised controlled trial.
Methods: In this single-centre, open-label, randomised, controlled trial, infants with VSD and heart failure were randomised to propranolol and no propranolol in addition to conventional treatment. A total of 80 patients were enrolled, 40 in each group. Median follow up was seven months (range 1–32 months). The primary endpoint was a composite endpoint of death, hospitalisation and referral for surgery.
Results: Fourteen (35%) patients in the conventional arm and 10 (25%) in the beta-blocker arm had reached primary endpoint (p = ns). Worsening of heart failure occurred more commonly in the conventional treatment arm compared with the propranolol-treated arm (27.5 vs 5% respectively; $p = 0.015$). Two patients in the conventional treatment arm and one in the propranolol arm died. No episodes of bradycardia or bronchospasm were reported with propranolol treatment.
Conclusions: The addition of propronolol was well tolerated by infants with VSD and heart failure. Addition of beta-blocker over and above the conventional treatment leads to symptomatic improvements and reduces worsening of heart failure. However, there was no difference in the death rate, hospitalisation or need for surgery.

739: RIGHT VENTRICULAR OUTFLOW MASS IN A NEWBORN WITH PULMONARY ATRESIA: ECHOCARDIOGRAPHY AND HISTOPATHOLOGICAL ASPECT

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Background: Typical tumours in the paediatric age group are fibroma, rhabdomyoma and teratoma. A non-neoplastic mass may consist of thrombus and infection, which can be defined by a pathological examination. We describe a five-month-old female child who had suffered from progressively increasing cyanosis since birth. At five months a murmur was noticed during a paediatric consultation and she was referred for paediatric cardiology at the Instituto de Medicina Integral Professor Fernando Figueira.
Case report: At admission her weight was 6.0 kg, her height was 61 cm, heart rate was 110 bpm, O_2 saturation was 73%, normal breathing, cyanosis (+++), normal pulses, systolic murmur in left sternal border, and no hepatomegaly. Electrocardiogram showed left ventricular overload. Chest radiography showed increased cardiac area and decreased pulmonary blood flow. The echocardiographic findings were pulmonary atresia, mild hypoplasia of the tricuspid valve with moderate regurgitation, right atrium enlargement, mild enlargement of the right ventricle and severe hypertrophy. The pulmonary valve was atresic. A mass (17 × 12 mm) was seen beneath the pulmonary valve with no mobility. The ductus arteriosus was patent and the pulmonary arteries were well developed.

The child underwent surgery to open the pulmonary valve and excise the mass from the right ventricle outflow. The pathological findings were a necrohaemorragic material with foci of calcification consistent with thrombus. The child was discharged from hospital 20 days after surgery in a good condition. We have shown here the association between cyanotic heart disease and thrombus, which is related to slow pulmonary blood flow rather than degree of cyanosis or coagulation abnormalities.

742: CAUSES OF HAEMOPTYSIS IN EISENMENGER SYNDROME: A CT ANGIOGRAPHY STUDY

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Background: Haemoptysis is a common cause of morbidity in patients with Eisenmenger syndrome. We aimed to analyse the predictors of haemoptysis in patients with Eisenmenger syndrome and determine the underlying causes using computerised tomographic pulmonary angiogram (CTPA).
Methods: Forty-one patients of Eisenmenger syndrome were studied; among them 24 had no haemoptysis and 17 patients had haemoptysis. The mean age of the patients was 23.7 ± 7.9 years with a range from 13–50 years. The patients with haemoptysis underwent CTPA within one week of their index bleed.
Results: No significant difference was found between patients with and without haemoptysis in baseline demographic characteristics, diagnosis, complexity of lesion, functional class, symptoms, and parameters before and after 6 MWT. The only statistically significant finding was the reduced 6 MWD in patients with haemoptysis (323.8 ± 81.7 m) compared to patients without haemoptysis (385.2 ± 92.6 m) ($p = 0.03$). The CTPA was abnormal in 13 patients and normal in four. The most common extraparenchymal lesion was the presence of collaterals (five patients). The more described cause of haemoptysis, pulmonary thrombus, was seen in only one patient. One patient had a pseudo-aneurysm from a branch of the left pulmonary artery, which was closed with a coil. One patient was diagnosed to have miliary tuberculosis. The most common intraparenchymal lesion was the presence of mosaic pattern suggestive of recent pulmonary haemorrhage. Overall, seven patients underwent a therapeutic procedure based on the finding of CTPA.
Conclusion: Haemoptysis remains a major cause of morbidity in patients with Eisenmenger syndrome. Haemoptysis occurs more frequently in patients with greater exercise limitation. CT pulmonary angiogram immediately following an episode of haemoptysis could identify a potentially treatable cause in nearly half of the patients.

745: MITRAL VALVE REPAIR IN CHILDREN AND ADOLESCENTS WITH RHEUMATIC HEART DISEASE

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Background: In Brazil, most of the interventions on the mitral valve are secondary to rheumatic heart disease. Mitral valve repair, compared with valve replacement, has many advantages. This work aimed to analyse the results of mitral valve repair in the correction of mitral regurgitation in children and adolescents with rheumatic heart disease.
Methods: Medical records from 29 patients with mitral regurgitation submitted to mitral valve repair between 2002 and 2011 in the Instituto de Medicina Integral Professor Fernando Figueira (IMIP) were reviewed and supplemented by physical examination and echocardiography. The age ranged from six to 16 years with a median of 10.9 years. The median postoperative period was 6.21 years (range at least one to no more than 10 years).
Results: Four (13.8%) patients still had severe mitral regurgitation postoperatively, two of them requiring mitral valve replacement and the other two are in clinical follow up. Two other patients required valve replacement, one due to several mitral stenoses and other due to major haemolysis. With the exception of four patients re-operated, all others were in New York Heart Association functional class I. Six (20.7%) patients had moderate mitral regurgitation and 17 (58,6%) mild mitral regurgitation. In eight (27.6%) patients we detected mild mitral stenosis, eight (27.6%) showed moderate mitral stenosis and two had moderate to severe. The left ventricular systolic function was decreased in four (13.8%) patients. Mild or moderate pulmonary hypertension was present in 10.3%. There was no report of death.
Conclusions: The present study demonstrates that valve repair is a good option for surgical treatment of mitral regurgitation in rheumatic heart disease, showing an improvement in the clinical pattern even in the late postoperative period.

747: MEASUREMENT OF EXERCISE CAPACITY AND ECHOCARDIOGRAPHIC LEFT VENTRICULAR FUNCTION DURING SEMI-SUPINE STRESS-CYCLE ERGOMETRY IN PATIENTS WITH ANOREXIA NERVOSA

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Background: Patients with anorexia nervosa (AN) have altered physiological responses to exercise. This study aimed to determine the differences in exercise capacity and haemodynamic parameters with exercise in patients with AN.
Methods: This was a retrospective case–control study. Sixty-six adolescent females with AN and 21 adolescent female controls exercised on a semi-recumbent ergometer in three-minute, 20-watt incremental stages to volitional fatigue. Heart rate (HR), blood pressure (BP), and echo Doppler indices were measured pre-, at each stage, immediately and three minutes post-exercise. Fractional shortening (FS), peak aortic velocity (PAoV), mean velocity of circumferential fibre shortening (MVCFc), wall stress (WS), cardiac index (CI), and systemic vascular resistance (SVR) were calculated. Peak oxygen consumption (VO_2), minute ventilation (VE), respiratory exchange ratio (RER), and arterial–venous oxygen difference (a-vO_2) were determined using open-circuit spirometry.
Results: Patients with AN had a significantly lower BMI (16.7 vs 19.7 kg/m^2, $p < 0.001$), total work (1 126 vs 1 914 J/kg, $p < 0.001$), total test duration (13.8 vs 20.8 min, $p < 0.001$), peak VE (47.4 vs 72.0 l/min, $p < 0.001$), and VO_2 (31.3 vs 39.7 ml/min/kg, $p < 0.001$) and higher RER (1.14 vs 1.06, $p = 0.001$) when compared to controls. Systolic BP, diastolic BP, and PAoV were lower at pre-exercise, increased with exercise, and were lower at peak exercise in AN vs control subjects. HR, FS, MVCFc and CI showed no difference at pre-exercise, increased with exercise, and were lower at peak exercise in AN patients vs controls. WS decreased with exercise and was lower in AN patients vs controls at pre-exercise and peak exercise. SVR pre-exercise was lower in AN, decreased with exercise, and there was no difference at peak exercise. The a-vO_2 increased with exercise with no differences between groups.
Conclusions: Adolescent patients with AN have decreased exercise capacity and abnormalities in their haemodynamic parameters and myocardial performance during exercise compared to controls.

751: CLINICAL ASPECTS OF 112 PATIENTS WITH ACUTE RHEUMATIC CARDITIS IN A PAEDIATRIC HOSPITAL

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Background: Rheumatic fever is a universal disease and primarily affects children in developing countries. The authors describe the clinical aspects of 112 children and adolescents admitted to the cardiology ward of the Hospital Instituto de Medicina Integral Professor Fernando Figueira during the period 2004 to 2010.

Methods: A survey was conducted of medical records for data collection. Median age was 10.4 years old and the median length of stay for acute rheumatic fever hospitalisation was 16 days.
Results: Evidence of previous streptococcus infection, which is very important for diagnosis was present in a few cases (21% had tonsillitis and 32% had increased antistreptolysin). Minor criteria such as fever were observed in 60 patients (53%), prolongation of the PR interval on the electrocardiogram in 86 (77%) and raised erythrocyte sedimentation rate or C reactive protein in 48 (36%) patients. Because it is a reference hospital for paediatric cardiology, there was a high incidence of carditis (96 cases, 85% of patients) and heart failure (57 cases, 51% of patients) with 13% of the patients requiring the use of amines during hospitalisation. Another cardiac parameter observed was mitral regurgitation in 29 (26%) patients, nine (8%) with aortic regurgitation, and 45 (40%) with both lesions. The incidence of arthritis 33 (29%) was low and only seven patients (6.25%) had chorea. Subcutaneous nodules were rare, with only two cases (1.8%). Most of the patients received corticosteroid therapy 68 (61%) and only 13 (12%) received acetylsalicylic acid. One patient died during the acute phase (0.89).
Conclusion: From this study one can conclude that rheumatic fever with cardiac involvement still remains a major cause of heart disease in developing countries, bringing high economic and social costs to these countries.

754: PRE-NATAL DIAGNOSIS OF COMPLEX CONGENITAL CARDIAC DEFECT: IS THERE ANY NEURO-DEVELOPMENTAL BENEFIT?

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Objectives: High pre-operative lactate levels have been associated with early childhood neuro-developmental delay. Our objective was to determine whether pre-natal diagnosis (PreN) has neuro-developmental benefit in patients with complex congenital cardiac defects compared to post-natal diagnosis (PostN).
Methods: A single-centre retrospective review was carried out of all patients with complex congenital cardiac defects who had complex surgery before six weeks of age, between 2001 and 2010 (n = 101). Prospective neuro-developmental assessments using the Bayley scales of infant development-II (n = 25) and, in 2005 and after, the Bayley scales of infant and toddler development-III (n = 50) were performed between 18 and 24 months of age, excluding children with chromosome anomalies or syndromes affecting development (n = 13). Two-year mortality was 14.8% (12/88). One patient was lost to follow up. Grouped scores for mental and motor delay [> 2SD below mean (< 70)] were compared between PreN (n = 47) and PostN-groups (n = 28). Peri-operative variables were collected. Propensity match analysis was performed (15 patients per group) controlling for surgical procedure and significant peri-operative variables.
Results: The PreN-group had trended to lower 30-day [1.9% (1/52) vs 5.6% (2/36), p = 0.56 (Fisher's test)] and 2-year mortality [9.6% (5/52) vs 19.4% (7/36), p = 0.31 (Fisher's test)].The Pre-group had lower mean pre-operative lactate levels [2.3 mmol/l (SD 1.6) vs 4.4 mmol/l (SD 5.0), p = 0.024 (t-test)] and lower median cardiopulmonary bypass flow rates > 10 min [100 ml/kg/min (IQR = 54–126) vs 125 ml/kg/min (IQR = 100–131), p = 0.020 (Mann-Whitney test)]. Other peri-operative variables were similar between groups. Fewer patients in the PreN group had combined delayed mental scores [4.3% (2/47) vs 25% (7/28), p = 0.01 (Fisher's test)], although mean scores were similar. There was no difference in motor delay. No NDO were significantly different after propensity match analysis.
Conclusion: Patients with a pre-natal diagnosis of complex congenital cardiac defects had significantly lower pre-operative lactate levels with a lower prevalence of cognitive delay. Longer follow up and larger cohorts might lead to more significant beneficial effects of pre-natal diagnosis on ND.

765: PULMONARY HYPERTENSION IN THE PRESENCE OF AN AORTOPULMONARY WINDOW, IN PATIENTS LIVING AT HIGH ALTITUDE, DOES NOT AFFECT MORBIDITY–MORTALITY OUTCOMES

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Background: Aortopulmonary window is a rare abnormal congenital communication. Early surgical correction should be offered before the development of irreversible pulmonary hypertension. The aim of this study was to present the morbidity–mortality rates in the surgical correction of this pathology in patients with and without pulmonary hypertension who live at high altitudes.
Methods: Case reviews were examined of 17 patients who underwent surgery between January 1990 and March 2012 at 2 640 m above sea level. Continuous variables are presented in means or medians with their SD or IQR, categorical variables in absolute and relative frequencies; Mann Whitney and Fisher's exact test were used to compare patients with or without pulmonary hypertension. All patients had an echocardiogram, which was diagnostic in 41.2% of patients (7/17).
Results: The global median age was 10.5 months (IQR 1.4–211), females made up 58.8% (10/17), pulmonary hypertension was present in 41.2% (7/17), and PDA was the most frequent cardiac-associated anomaly in 47.1% of patients (8/17). Mean time on pump was 64 ± 18.4 min, an aortotomy was carried out in 76.5% (13/17), a goretex patch was placed in 35.3% (6/17), and low cardiac output and arrhythmias were observed in 52.9% of patients (9/17). The mortality rate was 5.9% (1/17). Survival at 30 days was 100%, and at the 10-year follow up it was 37.5% (6/16). No statistical differences were found in pre- or postoperative variables in patients with or without pulmonary hypertension.
Conclusions: Pulmonary hypertension in the presence of an aortopulmonary window lesion in children did not differ from that in other types of lesions with left-to-right shunt. The mortality rate was low and apparently not related to these findings.

768: INFECTIVE ENDOCARDITIS: IMPROVING POSITIVITY OF MODIFIED DUKE'S CRITERIA IN DEVELOPING COUNTRIES

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Introduction: Successful management of infective endocarditis (IE) depends on timely accurate diagnosis, both being very challenging in developing countries due to multiple factors.
Objective: This study was undertaken to assess the usefulness of additional modification of Duke's criteria in diagnosing infective endocarditis in children in the developing world.
Methods: A retrospective 10-year chart review was done of children younger than 14 years of age, suspected to have or diagnosed with infective endocarditis (IE).
Results: Fifty patients were diagnosed to have IE. Mean age was 8.4 ± 5.6 years. Fever was present in 94%. Congenital heart diseases were predisposing in 74% and rheumatic heart disease in 16%, and

10% had invasive lines; 80% had a history of prior antibiotic use. A single blood culture was positive in 42%, and second and third blood cultures were positive in 30 and 10%, respectively. On echocardiography, 84% had vegetations. Thrombocytopaenia was seen 28%. Modified Duke's criteria were positive in 62%, probable in 16%, and rejected in 22%. Including C-reactive protein and ESR (raised in 80 and 60%, respectively) improved the diagnostic sensitivity. We further modified Duke's criteria to include one positive blood culture and thrombocytopaenia to evaluate if it further improved positivity; the positivity of Duke's criteria improved to 70%, and the possibility and rejection reduced to 10 and 18%, respectively. All nine rejected by the modified Duke's criteria responded to antibiotic therapy, based on high clinical suspicion. Surgical intervention was required in 8% and the overall mortality was 8%.
Conclusion: Diagnosis of IE is challenging in developing countries due to prior antibiotic use. High clinical suspicion and echocardiography remain the mainstay of diagnosis. Modified Duke's criteria may have to be further relaxed to improve diagnostic application in such situations, and we have found that using one positive blood culture with a known organism and thrombocytopaenia may be helpful.

782: HIGHER INCIDENCE OF ENDOCARDITIS IN BOVINE JUGULAR VEIN GRAFTS COMPARED WITH CRYOPRESERVED HOMOGRAFTS

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Background: Both cryopreserved homografts (CH) and bovine jugular vein grafts (BJVG) are used to reconstruct the right ventricular outflow tract (RVOT). We sought to compare the incidence of endocarditis (EC) in patients receiving the BJVG conduit to those receiving a CH.
Methods: We reviewed retrospectively all available clinical data in patients receiving either BJVG or CH implanted between 2000 and 2012. Endocarditis was defined as new conduit vegetation visualised on echocardiography +/– positive blood cultures (BC) or accelerated conduit deterioration and positive BC.
Results: We implanted 378 conduits (BJVG 244, CH 134) in 298 patients (median age 43 months, range 1 day – 50 years), median follow up 40 months (range 2 days – 12 years). Indications for surgery were pulmonary atresia/ventricular septal defect (46%), aortic stenosis (20%), truncus arteriosus (16%), other (18%). There were 22 cases of EC, 21 associated with BJVG (8.6%) and one with CH (0.75%; $p = 0.0009$) occurring at a median age of 12 years (range 6–21) and median time post conduit implantation 44 months (20 days – 10 years). BC were positive in 17 patients (11 *Staphylococcus aureus*, two *Streptococcus viridans*, two *Cardiobacterium hominis*, one *Staphylococcus epidermididis*, one *Haemophilus parainfluenzae*). Conduit replacement was required in 14/22, three patients had recurrent EC of the revised conduit. EC (+) patients had significantly reduced freedom from re-operation at one, five and seven years: EC(–): 95.7 ± 1.2%, 80.0 ± 2.7% and 69.2 ± 3.5%, respectively; EC(+) 95.5 ± 4.4%, 63.9 ± 1.2%, 32.0 ± 1.2%, respectively ($p = 0.003$).
Conclusions: The use of the BJVG conduit, compared to CH for reconstruction of the RVOT was associated with a significantly higher incidence of bacterial endocarditis. Furthermore, endocarditis of the BJVB was associated with conduit deterioration and more frequent re-operation. This information may be useful in the decision about which conduit to use for RVOT reconstruction in children with congenital heart disease.

783: CARDIAC FUNCTION AFTER ANATOMICAL REPAIR AND FUNCTIONAL REPAIR IN CORRECTED TGA AND TGA IN THE LONG TERM

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Background: In patients with congenitally corrected transposition of the great arteries (ccTGA), it is still unknown whether anatomical repair is better than functional repair in the long term. Furthermore, the physiological mechanism of failure of the systemic right ventricle (sRV) in ccTGA is not fully understood.
Objective: To evaluate cardiac function in patients with ccTGA long after anatomical and functional repair.
Methods: A retrospective review of catheterisation data between 2005 and 2011 was carried out in consecutive patients older than 16 years of age with ccTGA and complete TGA. Patients were divided into four groups; in TGA patients, an atrial switch group ($n = 12$) and an arterial switch goup; and in ccTGA patients, a double-switch group ($n = 8$) and conventional Rastelli group ($n = 6$). Unpaired *t*-test and one-way ANOVA were used for statistical analysis.
Results: Central venous pressures, cardiac index (CI), systemic pressures, ejection fraction (EF) of the systemic and pulmonary ventricle, and end-diastolic pressure (EDP) of the pulmonary ventricle were significantly different between the four groups (ANOVA, $p < 0.05$). Regarding EF of the systemic ventricles, EF in the arterial switch group (anatomical left ventricle) was significantly better than that in the atrial switch group (anatomical LV) in TGA (60 ± 6 vs 44 ± 77%, $p < 0.01$). In ccTGA patients, however, EF of the LV in the DSO group was not significantly different from that of the RV in the conventional Rastelli group (53 ± 7 vs 52 ± 18%). When EF of the pulmonary ventricle was compared, EF of the RV in the DSO group was significantly lower than that in the arterial switch group (45 ± 8 vs 61 ± 11%, $p = 0.017$). CI was also significantly lower in the DSO group compared to the arterial switch group (2.3 ± 0.4 vs 3.2 ± 0.6, $p < 0.05$).
Conclusion: LV and RV function after DSO were compromised. Systemic RV function in the conventional Rastelli group was also compromised.

784: COMPARATIVE STUDY: RIGHT VENTRICULAR ASSESSMENT IN POST TETRALOGY OF FALLOT REPAIR PATIENTS BY ECHOCARDIOGRAM WITH CARDIAC MRA

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Background: Post TOF repair patients should be continuously evaluated for cardiac function, especially the right ventricle (RV). Pulmonary regurgitation (PR) is a major cause of RV failure. Currently, cardiac magnetic resonance angiography (MRA) is considered the clinical reference method for RV assessment. The echocardiogram is an alternate tool for evaluating cardiac anatomy and function.
Objective: Assessment the RV parameters using the echocardiogram in comparison with cardiac MRA.
Methods: Twenty patients (mean age 14 ± 2 years) after TOF repaired were recruited from June 2011 to March 2012. The RV was evaluated by cardiac MRA followed by echocardiography. The echocardiographic parameters were tricuspid annular plane systolic excursion (TAPSE), fractional area change (FAC), area of right ventricle end-diastolic index (area RVEDi), RV free wall myocardial performance index (MPI) and severity of PR. The cardiac MRA parameters were right ventricular ejection fraction (RVEF), right ventricular end-diastolic volume index (RVEDVi) and severity of PR. Comparative analyses were assessed by Pearson's sample correlation coefficient, Crosstab kappa, sensitivity and specificity of area of

RVEDi from ROC curve analysis.
Results: There were correlations between RVEDVi and area of RVEDi ($r = 0.768$, $p < 0.01$), RVEF with FAC ($r = 0.759$, $p < 0.01$) and RVEF with TAPSE ($r = 0.688$, $p < 0.01$); and 100% correlation of moderate-to-severe PR assessment by echocardiogram vs cardiac MRA (Crosstab kappa = 0.912). Abnormal MPI by echocardiogram was not correlated with NYHA classification, chest X-ray and EKG (Crosstab kappa = –0.10, 0.15, –0.04). The area RVEDi ≥ 20.43 cm^2/m^2 from echocardiography was correlated with RVEDVi ≥ 160 ml/m^2 from cardiac MRA (sensitivity 64% and specificity 83%) from ROC curve analysis.
Conclusions: The echocardiogram is an effective tool for RV evaluation in TOF with PR. Measurement of area of RVEDi, FAC, TAPSE and degree of PR correlated well with cardiac MRA parameters.

787: RESIDUAL PULMONARY STENOSIS HAS THE ABILITY TO PREVENT PULMONARY REGURGITATION AND RIGHT VENTRICULAR DILATATION IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

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Background: Pulmonary regurgitation (PR) and resultant right ventricular (RV) dilatation are important determinants of long-term outcome in patients with repaired tetralogy of Fallot (TOF). While residual pulmonary stenosis (PS) acts as a pressure load on RV, it may help reduce PR and prevent RV dilatation. To test this hypothesis, we employed wave-intensity (WI) analysis, which provides information about wave-front behaviour based on ventricular–vascular interaction.
Methods and Results: The study subjects were 53 patients with repaired TOF and 39 control subjects. WI of peripheral pulmonary arteries was computed as an instantaneous product of simultaneously measured pressure and flow velocity. WI yielded three major components: (1) systolic compression and acceleration wave (W1), which reflects RV ejection performance, (2) negative reflection wave (NR), which represents intensity of wave reflection, and (3) end-systolic expansion and deceleration wave (W2), which denotes the speed of regression in the antegrade blood flow and encompasses ventricular sucking effects, inertia of flow, and regurgitation. Pressure gradient across the PS in the repaired TOF group was 24.6 ± 22.8 (0–109) mmHg. While there was no significant difference in W1 between the two groups, both NR and W2 were markedly higher in repaired TOF patients than in the controls, consistent with the increased wave reflection and PR in repaired TOF patients. In the repaired TOF patients, multivariate regression analysis demonstrated that pulmonary wedge pressure and PR independently increased W2, while pulmonary stenosis reduced W2 (standardised β: 0.40, 0.39, –0.30, $p = 0.0024$, 0.012, 0.016, respectively).
Conclusions: The results indicated that residual PS served to reduce PR, while left ventricular diastolic dysfunction, represented by the high pulmonary wedge pressure, increased it. These data raise a potential caveat to perform angioplasty to relieve PS, and also suggest the importance of preservation of left ventricular diastolic function to improve long-term outcome of repaired TOF with residual PR.

799: PAEDIATRIC THREE-DIMENSIONAL ECHOCARDIOGRAPHY FACILITATES DECISION MAKING IN MANAGEMENT OF CONGENITAL HEART DISEASES

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Background: The improved image quality and ease of use of the current real-time 3D echocardiographic (3DE) system promotes incorporation of this study into routine use. Which condition would benefit most from 3DE are evolving and being defined. We describe conditions for which 3DE provides information that has an impact on decision making in the management of congenital heart diseases.
Methods: During 2009 to 2012 at the university referral medical centre, the patients were studied for 2DE and additional 3DE using the Philips iE33 3DE system with either an X7-2 or X5-1 probe, where appropriate, for body size. Younger and uncooperative infants were controlled with moderate sedation. The decision to perform additional 3DE was due to inconclusive or questionable information from standard 2D echocardiography. The images were acquired in live narrow-sector, zoom, full-volume, and full-volume with colour flow Doppler modes.
Results: Among 4 500 conventional 2DE studies, 3DE was performed for more information in the questioned 5% of studies. The patients were male (55%) and were in the age ranging from birth to 15 years old. Incremental information obtained by 3DE included clarification of septal morphology and relationship to adjacent structures, AVSD valve morphology and location of regurgitation, valvular regurgitation, confirmation and extent of aortopulmonary window, ventricular outlet morphology, identification of circumferential subaortic membrane, and morphology of complex single ventricle. The impacts of 3DE on decision making included cancelling unnecessary cardiac catheterisation, cancelling unnecessary further investigation, planning for valve repair, planning for subpulmonary resection and patch, and planning for appropriate arterial bypass cannulation. 3DE findings were in concordance with surgical findings in 98% of cases. 2DE missed the important information in 7% of studies.
Conclusion: Paediatric 3DE provides incremental information to facilitate decision making in the management of congenital heart disease.

801: CHANGES OF DIMENSIONS AND LEFT VENTRICLE FUNCTION IN CHILDREN WITH CONGENITAL HEART DISEASE (LEFT-TO-RIGHT SHUNT) AND HEART FAILURE POST CARVEDILOL THERAPY

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Background: Heart failure in children due to congenital heart disease (CHD) and left-to-right shunt lead to the activation of compensatory mechanisms. If these mechanisms are excessively activated, cardiac remodelling will occur, which is characterised by changes in dimensions and function of the left ventricle. Carvedilol, a novel third-generation nonselective β-blocker, can postpone this ventricular remodelling.
Objective: To determine changes in dimension and function of the left ventricle after carvedilol administration in children with heart failure due to CHD left-to-right shunt.
Methods: This was a double-blind, randomised, controlled trial. Children with VSD and PDA were divided into carvedilol and control groups, and observed using echocardiography for three months. The evaluation consisted of LVIDs, LVIDd, LVPWd, IVSd, volume, mass, ejection fraction, shortening fraction and E/A ratio.
Results: There were 30 children, 19 (63.3%) boys and 11 (36.7%) girls. The mean age was 57.6 (43.57) months. Twenty (70%) children had VSD and nine (30%) had PDA. There were significant differences between the two groups. The mean of changes in LVIDs: 18 (SD 0.37) vs 0.04 (0.35), $p = 0.04$; LVIDd: 0.25 (0.43) vs 0.20 (0.58), $p = 0.04$; LVPWd: 0.04 (0.10) vs 0.04 (0.10), $p = 0.03$; IVSd: –0.11 (0.14) vs 0.01 (0.21), $p = 0.04$; volume: 7.85 (14.74) vs 7.78 (22.87), $p = 0,01$; mass: –15.87 (13.38) vs 19.48 (51.52), $p = 0.03$; ejection fraction: 3.50 (5.96) vs –1.54 (6.17), $p = 0.03$; and the shortening fraction: 3,17 (5.43) vs –0.95 (4.89), $p = 0.04$. There was no difference in E/A ratio 0.07 (0.32) vs 0.03 (0.31), $p = 0.71$ between the two groups.
Conclusion: There were significant changes in LVIDs, LVIDd, LVPWd, IVSd, volume, mass, ejection fraction, and the shortening fraction, without any difference in E/A ratio, in children with CHD left-to-right shunt and heart failure post carvedilol therapy.

806: EFFECT OF ANTI-HEART FAILURE THERAPY ON VENTRICULAR END-DIASTOLIC PRESSURE IN CHILDREN WITH SINGLE-VENTRICLE CIRCULATIONS

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Background: The systemic ventricle in children with single-ventricle circulations is exposed to chronic volume loading. Heart failure medications such as angiotensin converting enzyme inhibitors (ACEI) and beta-blockers are frequently used despite a paucity of evidence supporting their efficacy in this population. Our aim was to determine the effect of ACEI ± carvedilol on ventricular end-diastolic pressure (EDP).
Methods: Subjects with single-ventricle physiology who underwent cardiac catheterisation between 1991 and 2011 were identified from the departmental database. Subjects were included if they underwent repeat catheterisation either pre- or post-Fontan and who were commenced on ACEI ± carvedilol, or had doses increased between catheterisations. Those who underwent interventions influencing loading conditions between catheter assessments were excluded.
Results: Sixteen patients were identified. Seven/16 (44%) had a systemic right ventricle. There were 10 patients pre-Fontan and six post-Fontan completion who had had repeated assessment. All patients were treated with ACEI; two post-Fontan patients were also commenced on carvedilol. The mean interval between assessments was 14.8 months (range 2.5–41.6 months). ACEI ± carvedilol were instituted or increased over 11.9 months (3–33 months) before haemodynamics were re-measured. Overall, a reduction in EDP from a median of 13 mmHg (IQR 12–15.8 mmHg) to 10 mmHg (IQR 8.3–12.5 mmHg) was demonstrated ($p = 0.001$), as well as a fall in mean atrial pressure from 11.5 mmHg (IQR 9.9–13.1 mmHg) to 9.2 mmHg (IQR 7–10.6 mmHg) ($p = 0.01$). A reduction in cardiothoracic ratio ($p = 0.04$) was also observed.
Conclusions: Ventricular diastolic function is an important determinant of optimal flow in a Fontan circuit. Our data demonstrated a significant reduction in EDP and mean atrial pressure in patients with single-ventricle physiology following treatment with ACEI ± carvedilol. Given that a raised ventricular filling pressure is a risk factor for failure of Fontan palliation, these findings suggest the positive haemodynamic effects of anti-heart failure therapy may impact on clinical outcomes in this population.

810: WHAT IS NORMAL? ECHOCARDIOGRAPHIC FINDINGS IN LOW-RISK CHILDREN LIVING IN A REGION WITH HIGH RATES OF RHEUMATIC HEART DISEASE

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Background: Echocardiographic screening for rheumatic heart disease (RHD) is becoming more widespread, yet there are uncertainties around the significance of mild valvular regurgitation or morphological abnormalities, and few studies in low-risk children. The aim of this study was to describe the echocardiographic findings of healthy school-aged children in northern Australia, and to apply the new 2012 World Heart Federation (WHF) criteria for the echocardiographic diagnosis of RHD.
Methods: Portable echocardiography was performed on 1 087 predominantly Caucasian children aged five to 15 years in urban Darwin and Cairns. Abbreviated echocardiograms were performed, followed by comprehensive studies in those with pre-determined indicators of possible abnormalities. Screening echocardiograms were subsequently reported in a blinded standardised fashion by cardiologists.
Results: Of the 1 087 children screened, 106 (9.8%) had a comprehensive echocardiogram; 32 (2.9%) had at least one morphological abnormality of the mitral valve (MV). A thickened anterior MV leaflet (defined as ≥ 3 mm) was most common (19 children); 214 children (19.5%) had some degree of mitral regurgitation (MR) with the majority reported as trivial. Eleven children had MR jets ≥ 2 cm seen in at least one view**,** and only two children had MR which met all four WHF criteria for significant MR. Aortic regurgitation (AR) was found in 23 (2.1%). Five children had AR jets ≥ 1 cm, three of whom had bicuspid aortic valves. There were no cases of mitral or aortic stenosis. Congenital abnormalities were detected in 16 children (1.5%). No children met the WHF criteria for definite RHD, and five children met the criteria for borderline RHD.
Conclusion: Trivial MR is common in healthy school-aged children, but significant regurgitation and morphological valvular abnormalities associated with RHD are rare. The absence of any cases of definite RHD detected in this low-risk cohort suggests that the WHF diagnostic criteria for RHD are appropriately specific.

815: PREVALENCE OF RHEUMATIC HEART DISEASE IN HIGH-RISK CHILDREN IN NORTHERN AUSTRALIA: APPLICATION OF THE 2012 WORLD HEART FEDERATION CRITERIA

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Background: Indigenous Australian children have among the highest prevalence rates of rheumatic heart disease in the world, as documented through local registers of clinical cases, but an epidemiological survey has never been undertaken. This study aimed to establish the prevalence of RHD in high-risk indigenous Australian children using the new 2012 World Heart Federation diagnostic criteria.
Methods: Portable echocardiography was performed on 4 158 predominantly Aboriginal and Torres Strait Islander children aged five to 15 years living in remote communities of northern and central Australia. Abbreviated echocardiograms were performed, followed by comprehensive studies in those with pre-determined indicators of possible abnormalities. Screening echocardiograms were reported off-site in a blinded, standardised fashion by cardiologists.
Results: Of the 4 158 children screened, 591 (14.2%) required a comprehensive echocardiogram, the majority of which (80%) were considered normal after secondary evaluation. Using the 2012 WHF criteria, 30 children had definite RHD and 49 had borderline RHD. Congenital abnormalities were detected in 57 children (1.3%).

Overall prevalence of definite RHD was 7.2/1 000 indigenous children (95% CI: 4.9–10.5) with a median age of 10 years and a slight female predominance (58.6%). A significantly higher rate of 14/1000 was observed in the Top End region of the Northern Territory ($p < 0.05$). The majority of children with definite RHD had isolated mitral valve disease. The prevalence of borderline RHD was 12.2/1 000 (95% CI: 9.2–16.4), with one in three of these having isolated significant aortic regurgitation.

Conclusions: The prevalence of definite RHD in indigenous children in northern Australia is similar to that of developing countries and is consistent with previous register-based clinical estimates, suggesting that the WHF criteria are appropriately sensitive and specific. These data will be critical in determining the cost-effectiveness of routine screening in Aboriginal children.

816: WESTERN AUSTRALIA'S RECENT EXPERIENCE IN DELINEATING PATENT DUCTUS ARTERIOSUS MORPHOLOGY BY ECHOCARDIOGRAPHY PRIOR TO DEVICE CLOSURE

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Background: The Children's Cardiac Centre in Perth, Western Australia uses high-quality cardiac ultrasound equipment in its day-to-day practice. Interventional management of PDAs is now widely practised with a wide range of devices readily available. Choice of a device is dependent on the anatomical detail of the duct – all of this detail has traditionally been delineated using angiography performed at the time of the interventional procedure. However, it has been noted that this anatomical detail could be accurately outlined using detailed echocardiography and therefore an accurate prediction of the probable device type can be made prior to the interventional procedure.

Methods: Retrospective review was done of 2D echocardiograms of all patients with PDAs who underwent interventional closure between January 2006 and July 2012. Appropriate echocardiographic images were collated side by side with lateral angiographic stills. Based on the echo image alone, devices thought suitable were selected and compared to the actual device deployed.

Results: We retrospectively reviewed the echocardiograms as well as the angiographic images of 74 patients with PDAs deemed suitable for interventional closure. Imaging quality of the duct in the pre-procedural echocardiogram was of sufficient quality in 31 patients to accurately predict the choice of a device to interventionally occlude the duct.

Conclusion: Modern ultrasound equipment almost always enables accurate delineation of anatomical detail of PDAs. Decisions regarding choice of appropriate device can almost always be made prior to proposed interventional procedure. This has important implications with regard to limiting stock of expensive interventional devices required, as well as the process of informed consent when discussing the procedure with patients and their carers.

822: A CASE OF ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY PRESENTING WITH ACUTE MYOCARDIAL INFARCTION AND CARDIOVASCULAR COLLAPSE

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Background: Although a rare clinical entity, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a common cause of myocardial infarction in children. Unrecognised and untreated, it leads to progressive left ventricular dilatation and systolic dysfunction.

Clinical case: We present the case of a 10-week-old infant who had been seen a month earlier in our cardiology service with fever, cough and difficulty in breathing. His echo showed dilated left ventricle with poor systolic function that was attributed to a myocarditis or dilated cardiomyopathy. Four weeks later he presented to the in-patient unit with marked restlessness and irritability. He was inconsolable, had marked respiratory distress, cool extremities, and central and peripheral cyanosis. The radial and brachial pulses were absent. The mean arterial pressure was 65 mmHg, heart rate of 160 beats per minute with a third heart sound. Laboratory tests showed an elevated CK-MB of 112.5 U/l. Other laboratory tests were normal. ECG revealed deep Q waves in leads I, aVL, V5, V6 with ST elevation in the anterolateral leads. Echo showed a dilated left ventricle with paradoxical septal motion, severe LV systolic dysfunction, LV anterolateral wall echo brightness and flow reversal in the left coronary artery with its origin from the pulmonary trunk. He was admitted to the coronary care unit, given fluid resuscitation, dopamine, standard management of heart failure and was discharged six days later.

Conclusion: A combination of a high index of suspicion, typical ECG and echocardiographic findings in a young infant presenting with LV dysfunction could lead to an earlier diagnosis of ALCAPA.

835: TREATMENT STRATEGIES IN PATIENTS WITH EISENMENGER SYNDROME ASSOCIATED WITH COARCTATION OF THE AORTA: CASE SERIES

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Aim: To establish the treatment strategies in patients with Eisenmenger syndrome and coarctation.

Methods: A review was carried out of all cases that had established pulmonary hypertension and coarctation between January 2008 and January 2012.

Results: Three patients were identified.

Case 1: A 22-year-old female was referred with differential cyanosis and large PDA. The echocardiogram showed coarctation (COA). The RV pressure was systemic. Finally she underwent stent angioplasty of the COA. During the procedure the RV pressure was systemic. The COA was stented with a covered CP stent, which closed the PDA. Immediate RV pressure was systemic. She was started on sildenafil. On follow up her RV pressure normalised and the sildenafil was stopped.

Case: 2. An eight-year-old was referred for differential cyanosis and he was noted to have a large PDA with COA. He underwent stent angioplasty of the COA as in the previous patient. His PA pressure was systemic, which normalised on follow up.

Case: 3. A seven-year-old was referred with large VSD and COA. Her saturations on admission were 80%. The echocardiogram showed large VSD with bidirectional flow. She underwent stent angioplasty of the COA. Postoperatively she was stable initially but later developed severe R-L shunt across the VSD (50% sats). She was started on bosentan and sildenafil. The saturations on discharge were 80%, but on follow up her PA pressure remained the same and she is currently being medically managed.

Conclusion: PDA with COA is a protected circulation as the pulmonary blood is shunted into the descending aorta and thus the pulmonary vasculature is not exposed to high blood flow. Therefore it is safe to close the PDA and stent the COA. VSDs with coarctation have increased pulmonary blood flow and cause irreversible pulmonary hypertension early. However treating the coarctation will help but patients need careful pre- and postoperative management.

837: OUTCOME OF DILATED CARDIOMYOPATHY IN DUTCH CHILDREN

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Background: Dilated cardiomyopathy (DCM) in children is a severe disease with a grave prognosis. However, a subgroup may recover completely or do well for years. In this retrospective study we sought to describe outcome and to identify predictors of outcome.
Methods: Children presenting with DCM between 2005 and 2010 were included. Data at presentation and in the month before reaching an endpoint or closing the study were retrospectively analysed. Three subgroups were defined, (1) those reaching a primary endpoint [death, heart transplantation (HTx) or mechanical circulatory support (MCS)]; (2) recovering patients [left ventricular end-diastolic dimension (LVEDd) and SF < p95] or (3) those with ongoing disease.
Results: One hundred and eight children were included [median follow up 1.8 years (range 0–5.4)]; 25 (23%) children reached a primary endpoint (10 died, three HTx, 11 MCS), 33 (31%) recovered and 50 (46%) had ongoing disease. The time (median, IQR) to a primary endpoint was significantly shorter [0.2 years (0.03–1.1)] than the time to recovery [0.8 years (0.3–2.5)] ($p < 0.05$). Fifty-four (50%) children had idiopathic DCM, 21 (19%) had myocarditis. Fifteen (28%) children with idiopathic disease reached a primary endpoint, compared to only one (5%) with myocarditis (log rank $p < 0.05$). At presentation, LVEDd (SD) was larger in children reaching a primary endpoint [*z*-score +6.9 (± 3.9)] than in those recovering [*z*-score +5.0 (± 2.5)] ($p < 0.05$). During follow up, LVEDd *z*-score further increased in children reaching a primary endpoint (+0.5/year), in contrast to those who recovered (–2.9/year) ($p < 0.01$). At presentation, weight for height (WFH) was similar in all subgroups, but during follow up children reaching a primary endpoint lost weight (–0.6 WFH SDS/year) in contrast to those not reaching a primary endpoint (+0.4 WFH SDS/yr) ($p < 0.05$).
Conclusion: One-fourth of children with DCM reached a primary endpoint. Adverse outcome was related to (at presentation) idiopathic disease and large LV, and (during follow up) further LV dilatation and weight loss. Favourable outcome was related to myocarditis and reverse remodelling during follow up.

841: OXIDATIVE PHOSPHORYLATION DISORDERS AMONG CHILDREN WITH SEVERE CARDIOMYOPATHY

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Background: Cardiac involvement occurs in 17 to 40% of children with oxidative phosphorylation (OXPHOS) disorders, but the incidence of OXPHOS disorders among children with primary cardiomyopathy is unknown.
Methods: We analysed data of all children with documented cardiomyopathy who underwent OXPHOS testing between 1984 and 2012. Testing on heart, skeletal muscle and/or liver was performed because of the suspicion of a mitochondrial condition, the use of circulatory support, cardiac transplantation or death. Children with severe non-cardiac organ dysfunction were excluded. The diagnosis of an OXPHOS disorder was based on results of enzymology in conjunction with established diagnostic criteria. Cardiomyopathy type was characterised by a single cardiologist.
Results: There were 62 patients (50% female). The median (IQR) age at presentation was 1.29 (0.31–7.43) years. The median (IQR) duration of follow up was 1.03 (0.12–7.37) years. During this time 25 (40.3%) children received a transplant and 21 (33.9%) died. Eleven (17.7%) patients had a definite OXPHOS deficiency, one (1.6%) was considered probable and seven (11.3%) were considered possible. These included 13 of 38 (34.2%) children with dilated cardiomyopathy, five of 11 (45.5%) children with hypertrophic cardiomyopathy, one of five (20%) children with left ventricular non-compaction and none out of eight (0%) children with restrictive cardiomyopathy. Of 51 children without any documented extra-cardiac abnormalities, eight (15.7%) were considered definite and six (11.8%) possible. The predominant clinical findings in this group at presentation were congestive heart failure ($n = 37$, 72.5%) and arrhythmias ($n = 5$, 9.8%). There was no difference in age and signs at presentation between those with and without an OXPHOS disorder. Transplant-free survival was similar in both groups (9/20; 45% vs 7/17; 41%, respectively).
Conclusions: OXPHOS disorders may have a variable cardiomyopathy phenotype, and were common in this cohort of children with severe cardiomyopathy.

847: RESULTS OF CARDIAC CATHETERISATION AND TREATMENT OF PROTEIN-LOSING ENTEROPATHY IN CHILDREN WITH HLHS AFTER THE FONTAN PROCEDURE

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Objective: Protein-losing enteropathy (PLE) occurs in three to 15% of patients with Fontan circulation. All require adequate medication, cardiac catheterisation (CC) and some re-intervention. We reviewed the cardiac catheterisation laboratory database at the University Children's Hospital of Cracow, Poland, to identify HLHS patients after a Fontan procedure (FP), who underwent CC between January 2001 and July 2012.
Results: At that time, 330 HLHS children were palliated using FP, with only one postoperative death. Of 21 patients subjected to CC, in 10 (50%) patients operated when *x*–2.9 ± 1 years old and catheterised when *x*–5.8 ± 2.5 years old, the cause was PLE confirmed by blood and stool alpha-1 antitrypsin levels. The time from FP to CC ranged from three months to 7.5 years, *x*–2.6 years. In only one patient (with PLE recognised three months post-FP), no structural cardiac changes were found. Two patients revealed narrowed interatrial communication and required re-operation, seven had significant left, and one bilateral pulmonary artery branch stenosis demanding balloon pulmonary angioplasty (BPA) (in five patients with stents). Before BPA, the dimension of the most narrowed pulmonary artery branch ranged from 1.5–6.2 mm and after, 5.8–12 mm, *x*–9.1 mm. Systemic venous pressure ranged from 13–22 mmHg, *x*–16 mmHg, RVEDP: 4–13, *x*–7.6 mmHg, Sa02: 88–99%, *x*–95.1%. The follow up ranged from 0.5–7.5 years, *x*–2.2 years. All patients were treated with diuretics (furosemide, spironol, hydrochlorothizide), ACE inhibitors (enarenal, captopril), and aspirin (one was also on warfarin), and received a specific high-protein MCT product-enriched diet, three additionally received steroids and sildenafil, two steroids, and one sildenafil. Significant improvement was achieved in 8/10, with protein levels increasing from *x*–42.4 ± 6.5 g/l to 53.5 ± 9.5 g/l.
Conclusions: Post-FP, the majority of HLHS and PLE patients may reveal various structural changes that impede Fontan circulation. Management of PLE requires elimination of such changes and complex, mostly symptomatic treatment.

849: COMPUTATIONAL FLUID DYNAMICS IN FONTAN PATIENTS TO EVALUATE ENERGY LOSS DURING SIMULATED EXERCISE

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Background: In most patients with a functional single ventricle, a total cavopulmonary connection (TCPC) is created. Exercise intolerance is common in Fontan patients. It has been suggested that energy loss (Eloss) inside the TCPC may play an important role in reduced exercise performance. The aim of this study was to establish whether Eloss inside the TCPC plays a significant role during (simulated) exercise.
Methods: In 15 patients a three-dimensional reconstruction of the TCPC was created using anatomical data from cardiac magnetic resonance imaging (CMR). Phase-contrast CMR was used to obtain through-plane flow rates from the inferior vena cava (IVC) and superior vena case (SVC) during rest and during exercise, simulated with dobutamine (7.5 µg/kg/min) intravenously. Patients underwent cardiopulmonary exercise testing to assess maximal oxygen uptake. CFD simulations were performed using the flow rates obtained by CMR. Total energy loss (Eloss) inside the TCPC structure was calculated for both conditions in each patient. A condition where only IVC flow was increased two-fold compared to the resting condition was added to the study to reflect a more natural flow distribution, which would be expected during supine exercise.
Results: Eloss was higher during simulated exercise in all patients but one. Mean Eloss was 0.62 ± 0.37 mW during rest, 1.07 ± 0.64 mW during simulated exercise and 2.97 ± 2.49 mW with two-fold IVC flow. The correlation between cardiac index and Eloss was exponential ($r$sq = 0.393, $p < 0.000$). The increase in Eloss depended on the specific anatomy of each patient. No correlation was found between the increase in Eloss during dobutamine infusion and exercise capacity as assessed by maximum oxygen uptake.
Conclusions: Eloss inside the TCPC structure was limited but increased with (simulated) exercise. This was due to increased blood flow, but also depended on patient-specific anatomy of the TCPC. We did not observe a correlation between Eloss and exercise capacity.

850: CROSSED PULMONARY ARTERIES: REPORT OF 20 CASES WITH EMPHASIS ON CLINICAL FEATURES WITH GENETIC, CARDIAC AND EXTRACARDIAC ABNORMALITIES

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Background: Crossed pulmonary arteries (CPA) is a rare abnormality in which the ostium of the left pulmonary artery originates superiorly and to the right of the right pulmonary artery. Recognition of this rare pathology is important because it is usually accompanied by other congenital heart disease, extracardiac anomalies, and some genetic problems. To date, there have been few reported cases. Most are case report with complex cardiac abnormalities.
Methods: We have detected 20 cases due to increased awarenes of this anomaly in the last three years. Approximately 9 250 echocardiaograms were performed in this period. We describe 20 cases with this anomaly with an emphasis on the clinical features with genetic, cardiac and extracardiac abnormalities.
Results: The ages of the patients ranged from one day to 13 years at the time of initial diagnosis. There were 10 female and 10 male patients. Four patients had complex cardiac pathology such as TOF, truncus, TGA and CAVSD. Eleven of the 20 patients had ventricular septal defect. ASD and PS were detected in 12 of 20 patients (60%). Aortic arch abnormalities such as right aortic arch and coarctation were seen in six patients. One patient had persistent left superior vena cava; 45% of the cases were associated with genetic syndromes (Di-george, Noonan and Holt Oram syndrome, VACTER anomalies). These syndromes were diagnosed on the basis of clinical features. Karyotypes and FISH for 22Q deletion were studied in 11 patients. All had normal karyotypes and FISH results. Six patients underwent succesfull operations. Three of 20 patients died during long-term follow up. The remaining cases were clinically stable and were being followed without surgery.
Conclusions: CPA may be related to different congenital heart diseases. Detection of CPA may be an important clue to the presence of structural heart disease and chromosomal abnormalities.

851: RARE CASES OF LEFT VENTRICULAR ANEURYSM IN CHILDREN: A SINGLE-CENTRE EXPERIENCE

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LV aneurysms in children are very rare and often result from a congenital defect in the posterior portion of the mitral valve annulus. We present two unique cases of children with tuberculosis (TB) and acquired sub-mitral aneurysms.
Case report: The first case was an afebrile two-year-old girl with progressive dyspnoea, CHF and systolic murmur at the apex for eight days. CXR showed marked cardiomegaly with pulmonary venous congestion. Echocardiogram revealed a huge LV aneurysm at the sub-mitral valve area, causing moderate MS, severe MR and TR. EF was 50%. MRI showed a large pseudoaneurysm at thte posterior wall of the LV. Additional non-contrast CT scan revealed multiple calcified mediastinal lymph nodes (caseous nodes). The lung parenchyma was normal. Mitral valve replacement was performed due to complete destruction of the posterior leaflet and annulus. LV aneurismorhaphy and tricuspid valve repair were also done. The post-operative course was uneventful. Pathological examination suggested necrotising granulomatous inflammation. AFB from the lymph node was negative.

The second case was a 2.5-year-old boy with a history of TB contact, who presented with prolonged fever and dyspnoea. Physical examination revealed no signs of CHF or murmur. CXR showed mild cardiomegaly. He suddenly collapsed after complaining of severe epigastric pain immediate after administration. CPR was unsuccessful. The autopsy revealed rupture of a huge LV aneurysm at the sub-mitral valve area, with a massive intrapericardial blood clot, generalised lymphadenopathy, pleural effusion and ascites. Even though AFB from myocardial tissue was negative, pathological findings suggested granulomatous inflammation.

TB endocarditis has been reported sporadically. Most cases presented as LV aneurysm at the sub-mitral valve area. Patients with LV aneurysm are at potential risk for life-threatening conditions leading to sudden death. Echocardiography and MRI are crucial modalities to define the morphology and extent of the aneurysm and surgical planning. Management should be individualised and mainly directed toward early diagnosis to prevent moribund outcomes.

854: KAWASAKI'S DISEASE PRESENTING WITH DILATED CARDIOMYOPATHY: A CASE REPORT

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Background: Kawasaki's disease is a mucocutaneous lymph node syndrome of unknown aetiology with an estimated incidence of 9–19 per 100 000 children younger than five years. It is speculated to be due to immune system abnormalities initiated by an infectious insult. Kawasaki's disease is more prevalent in the Asian community with peak incidence between the ages of one and two years, and 80% of the cases present before the age of four years. Cardiovascular complications include pericardial effusions, coronary artery aneurysms and myocardial infarction. Early treatment with intravenous immunoglobulin and aspirin form the mainstay of treatment. Serial echocardiography is indicated to monitor the development of coronary artery aneurysms. Occasionally coronary artery angiography and bypass surgery might be indicated.
Case presentation: A nine-year-old Zimbabwean boy presented with congestive cardiac failure. There was no recent history of a febrile disease. On echocardiography he was found to have dilated cardiomyopathy and dilated left main coronary artery. A diagnosis of Kawasaki's disease was suspected. He was started on anti-failure treatment, aspirin and warfarin. Cardiac catheterisation showed an aneurysmal origin of the left coronary artery, with complete occlusion of the left anterior descending branch. A further area of stenosis and aneurysm was found in the right coronary artery. A few days post catheterisation he presented with a right-sided hemiplegia and aphasia. On echocardiography he had left ventricular thrombi and on CT brain, an embolic cerebrovascular incident involving the left middle cerebral artery and basal ganglia was confirmed. Coronary artery bypass surgery was considered, but on myocardial isotope scanning, there was no viable tissue in the left anterior descending artery territory. On follow up, he remains with left ventricular dysfunction.
Conclusion: A rare case of Kawasaki's disease with dilated cardiomyopathy, due to a previously undiagnosed myocardial infarction is presented. With early treatment, these complications are rarely seen.

855: CONSANGUINITY AND LONG QT SYNDROME: EXPERIENCE FROM SAUDI ARABIA

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Introduction: Congenital long QT syndrome (LQTS) is usually an autosomal-dominant inherited cardiac arrhythmia disorder. Patients are predisposed to ventricular tachyarrhythmias and fibrillation leading to recurrent syncope and/or sudden cardiac death. Not much is known about the prevalence, clinical severity and genetics of LQTS in communities with high rates of consanguinity.
Objective: We performed clinical and genetic investigations in eight Saudi Arabian families with a history of unexplained sudden death in children. Additionally, we also investigated the pathology of repeated intra-uterine foetal deaths in two families.
Methods and Results: Clinical symptoms, ECG phenotypes and genetic findings led to the diagnosis of LQT1 in four families (recessive) and LQT2 in four families (three recessive and one dominant). Onset of arrhythmia was more severe in all recessive carriers and occurred during early childhood in all recessive LQT1 patients. Arrhythmia originated at the early intra-uterine stages of life in the recessive LQT2 patients. LQT1 causal mutation c.387 -5 T > A in the *KCNQ1* gene was detected in three families. LQT2 causal mutation c.3208 C > T (p.Q1070X) in the *KCNH2* gene were identified in two families. In one family with sudden death of five siblings, we identified an unclassified variant c.1179 G > T (p.K393N) in the *KCNQ1* gene. This variant was present in heterozygous form in about 2% of the healthy Arabs, but not in healthy Caucasian controls.
Conclusion: Mutations detected in this study are novel, founder mutations in the Assir province of Saudi Arabia. Due to the high rate of consanguineous marriages in the Assir province, we could speculate that the mutations in *KCNQ1* (c.387 -5 T > A) and *KCNH2* (c.3208 C > T; p.Q1070X) could be quite frequent in LQTS pathogenicity and could be used as a first line of genetic investigation before proceeding to comprehensive screening for all LQTS causal genes. Further, c.1179 G > T (p.K393N) in *KCNQ1*, although present in heterozygous form in about 2% of the healthy Arabs, we suspect this variant could be highly deleterious when present in homozygous form. We are currently conducting studies to establish the pathogenicity of the p.K393N (KCNQ1) variant.

860: NATRIURETIC PEPTIDES IN MYOCARDIAL DYSFUNCTION IN NEWBORNS WITHOUT STRUCTURAL HEART DISEASE

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Background: Myocardial dysfunction is increasingly recognised in sick newborns without structural heart disease.With increasing availability of biomarkers for congestive heart failure, we analysed the role of serum proBNP (pro brain natriuretic peptide) levels in newborns with suspected myocardial dysfunction, in addition to ECG and echocardiography.
Methods: All newborns with suspected myocardial dysfunction had a 12-lead ECG, serum proBNP and a detailed echocardiogram done. Modified inotrope score (MIS) was calculated for the first five days in babies who needed inotropes (threshold value 1 000)
Results: Forty babies were included in the study. Mean gestational age was 36 weeks, mean birth weight 2 480 g; 50% of babies were born by C-section and antenatal stress factors were present in 85% of the babies. ECG abnormalities included low voltage, T-wave abnormalities and ST depression; 26 newborns with poor systolic and diastolic function had ECG abnormalities and only two with normal function had the same ($p = 0.008$). ECG was a poor predictor of isolated diastolic functional abnormalities.
Conclusions: The serum natriuretic peptides are a useful tool to assess cardiac failure in the sick newborn and need to more widely used. Available but underutilised tests such as ECG can also help in the early diagnosis of cardiac failure in the newborn.

862: EVALUATION OF CARDIAC AUTONOMIC FUNCTION USING HEART RATE VARIABILITY IN CHILDREN WITH ACUTE CARBON MONOXIDE POISONING

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Background: Carbon monoxide (CO) poisoning may cause myocardial toxicity and cardiac autonomic dysfunction, which may contribute to the development of life-threatening arrhythmias. Autonomic nervous system function can be measured by heart rate variability (HRV), a non-invasive index of autonomic controls of the heart. We investigated the potential association between CO exposure and cardiac autonomic function as measured by HRV.
Methods: This study included 40 children (18 boys and 22 girls) aged between one and 17 years admitted to the emergency department with acute CO poisoning, and 40 healthy age- and gender-matched controls. Carboxyhaemoglobin (COHb) and cardiac enzymes were measured at admission. Twenty-four-hour Holter electrocardiographies were digitally recorded; HRV was analysed in both the time

domain and frequency domain. Mean heart rate, P wave, P dispersion (Pd), QT, QTc, QTd and QTcd intervals were measured from electrocardiogram on admission and at discharge.
Results: Time-domain indices (SDNN, SDANN, rMSSD, SDNN index and NN50) were similar between patient and control groups ($p > 0.05$). Among the frequency-domain indices, mean HF level in the CO poisoning group was higher than in the controls ($p = 0.018$) while LF levels were similar ($p = 0.636$). LF/HF ratio was significantly lower in the CO poisoning group ($p < 0.001$). CoHb levels were negatively correlated with LF/HF ratio ($r = -0.351$, $p < 0.05$). On admission, mean heart rate, QTd, QTcd, and Pd values were higher in the CO poisoning group ($p < 0.05$). On discharge, QTd and QTcd were still longer in the CO poisoning group than in the controls ($p < 0.05$).
Conclusions: Frequency-domain indices recorded within the first five minutes on admission, especially the LF/HF ratio, are useful for evaluating cardiac autonomic function. Decreased LF/HF ratio reflects a balance of autonomic nervous system which shifted to parasympathetic components. These results suggest that exposure to CO may alter the balance of cardiac autonomic control, and thus may increase the susceptibility of high-risk patients to adverse cardiac events.

870: DOES MEASURMENT OF OXYGEN SATURATION IMPROVE THE DIAGNOSTIC DETECTION OF CONGENITAL HEART DISEASE DURING POPULATION SCREENING?

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Background: No consensus exists regarding the most efficient means to conduct population-based screening of children for undiagnosed congenital heart disease (CHD). A similar lack of consensus exists in the USA regarding the benefit of screening oxygenation saturation (O_2Sat) prior to newborn discharge. We sought to determine whether assessing O_2Sat would increase the sensitivity of CHD diagnosis during humanitarian, population-based paediatric screening.
Methods: During a humanitarian screening in two rural provinces in Mongolia (Mandal Gobi and Dalanzagdad), children from one month to 20 years underwent a cardiac physical examination by American and Mongolian paediatricians and nurse practitioners. All children also underwent evaluation of oxygen saturation with pulse oximetry. A potentially pathological murmurs on examination or oxygen saturations less than 94% prompted echocardiographic screening (echo) by a paediatric cardiologist.
Results: Previously unrepaired CHD was identified in 61 of 822 patients (7.0%) screened over four days, with 133 echos performed (16.1%). Identified cardiac defects included: 19 haemodynamically insignificant ventricular septal defects (VSDs), nine haemodynamically significant VSDs, 10 patent ductus arteriosi, eight atrial septal defects and two tetralogy of Fallot (ToF) patients. Remaining lesions were only minor valve abnormalities; 49 patients (6%) had O_2Sat < 94% on screening. Of these patients, seven had CHD (sensitivity for pulse oximetry to diagnose CHD = 11.5%). A reading ≥ 94% carried a higher specificity and negative predictive value (94.5 and 93%, respectively). Only one patient had oxygen saturation < 94% with a negative auscultatory examination (child with ToF and O_2Sat 72%). Of the 42 patients without CHD and O_2Sat < 94%, all had normal examinations.
Conclusions: While specificity is high, O_2Sat appears to have sufficiently low sensitivity and adds little to the diagnostic accuracy of physical examination alone, to warrant its use as a tool for population-based screening of paediatric patients.

873: EFFECTIVENESS OF INDOMETHACIN IN FULL-TERM INFANTS WITH SYMPTOMATIC PATENT DUCTUS ARTERIOSUS

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Background: Common treatment methods for patent ductus arteriosus (PDA) include surgical ligation and, recently, catheter intervention. Inhibiting prostaglandin synthesis seems effective for the non-surgical closure of PDA and indomethacin has been widely used with a reported efficacy of 70–80% in preterm neonates with significant PDA. However, there is a controversy on the effectiveness of indomethacin in full-term neonates with significant PDA. Therefore we evaluated the effect on indomethacin treatment on patients with significant PDA among full-term infants with birth weight (BW) ≥ 2 500 g and a gestational age (GA) ≥ 37 weeks.
Methods: We retrospectively reviewed 29 infants with significant PDA and a BW of 2 500 g or more and a GA of 37 weeks or more who were admitted to Chonnam National University Hospital between 2007 and 2009. During indomethacin therapy, feeding was prohibited and water intake restricted (60–80 ml/kg/day). Indomethacin (0.25 mg/kg/day) was intravenously administrated as a single dose at 12- to 24-hour intervals. Patients were classified as responders if there was complete closure of the DA and as partial responders if there was incomplete closure of the DA and clinical symptoms improved. The remaining patients were classified as non-responders.
Results: In indomethacin-treated patients, 13 (44.8%, responders) of 29 patients with PDA were completely closed. Eight (27.6% partial responder) were incompletely closed but clinical symptoms such as congestive heart failure improved without any particular treatment.
Conclusions: Indomethacin therapy may be a useful medical treatment option prior to considering surgery for PDA in full-term infants.

874: RHEUMATIC HEART DISEASE IN NAMIBIA: A PRELIMINARY REPORT FROM THE NATIONAL REGISTRY

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Background: The burden of rheumatic heart disease (RHD) in Namibia is unknown and there is no epidemiological data with which to inform public policy. The aim of this registry was to collect data to assist resource development and distribution with the end objective a reduction in prevalence, morbidity and mortality rates from a much-neglected disease.
Methods: This was a prospective, national, hospital-based registry of patients referred to the RHD Clinic at Windhoek Central Hospital. Questionnaires documented patients' presentation, clinical course, investigations, complications, management and demographics at enrollment. The study was initiated in July 2010 in collaboration with the Global Registry for RHD (REMEDY).
Results: One hundred and ninety-three patients were enrolled, with the distribution of cases reflecting regional population density; 58% were male and 42% female, 81% between 10 and 40 years and 5% under 10 years. Thirty-two per cent had severe disease (NYHA III–IV). The mitral valve was most commonly affected (77% with MR, 40% with MS), followed by tricuspid then aortic valve disease. Nineteen per cent hade atrial fibrillation, 6% stroke and 13% previous surgery; 38% of patients are receiving secondary penicillin prophylaxis. Of those needing anticoagulation, 44.7% were receiving warfarin and of those, 38% were aware of the target INR; 73% had no INR analysis in the preceding six months.
Conclusion: Patients are referred late with advanced disease. The low number of patients on secondary prophylaxis reflects a lack of awareness of the disease among the general public and health workers. Poor compliance with anticoagulation protocols indicates health workers are poorly informed about heart disease and reflects weakness in organisation of laboratory services. The RHD burden is highly significant. Gaps have been identified in the organisation and delivery of care and point to the urgent need for a national programme for the prevention and control of rheumatic heart disease.

876: ECHOCARDIOGRAPHIC PATTERN AND SEVERITY OF VALVE ABNORMALITIES IN CHILDREN WITH RHEUMATIC HEART DISEASE SEEN AT UGANDA HEART INSTITUTE, MULAGO HOSPITAL

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Background: Rheumatic heart disease (RHD) is the commonest acquired heart disease in children worldwide but in Uganda, data are scarce regarding its morbidity and mortality. The disease has a progressive course and patients will usually require valve surgery.
Objectives: To describe the frequency of the different heart valves affected, and the relationship of the disease severity with age and gender.
Methods: This was a retrospective descriptive study done at the Uganda Heart Institute. Echo reports of children 15 years and below with a diagnosis of RHD, done from January 2007 to December 2011, were retrieved from the data base and analysed.
Results: A total of 376 children had RHD. The mean age (± SD) of the children was 11.0 ± 2.7 years (range 4–15 years) and 216 (57.5%) were females. Severe mitral regurgitation (MR) was the commonest lesion seen in 277 (73.7%) of the children, 28 (7.4%) had severe aortic regurgitation (AR), 22 (5.9%) had severe mitral stenosis (MS), 32 (8.5%) had severe tricuspid regurgitation (TR), while only one (0.3%) had severe aortic stenosis. Twenty-seven of the 28 children with severe AR had concomitant severe MR. Severe MR was found to be significantly more prevalent in females ($p = 0.04$), while severe AR was significantly more common in males ($p = 0.007$). There was no difference in disease severity between children below 10 years and those 10 years and above.
Conclusions: The commonest severe valvular abnormality was MR (74%). Girls were more likely to have severe MR whereas boys were more likely to have severe AR.

877: THE EFFECT OF BETA-BLOCKER THERAPY ON LEFT VENTRICULAR VOLUME IN PAEDIATRIC FAMILIAL HYPERTROPHIC CARDIOMYOPATHY

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Background: We have previously shown that paediatric patients with hypertrophic cardiomyopathy (HCM) had significantly reduced left ventricular volumes compared with age- and gender-matched normals. We aimed to study how cardiac end-diastolic volumes and cardiac output at rest were affected by high-dose beta-blocker therapy in children and adolescents with HCM.
Methods: The study included 14 patients (two females), mean age 9.2 (range 1.8–17.7) years, with familial HCM and moderate-to-severe hypertrophy. They were paired in seven untreated and treated pairs (treatment: metoprolol or propranolol 5–20 mg/kg) according to age, gender and severity of hypertrophy, using septum-to-cavity (sepcavr), and left ventricular wall-to-cavity ratios (lvcavr) as age-independent measures of hypertrophy at diagnosis. The beta-blocker group had received treatment for at least one year. Ultrasound examinations were performed with Philips IE33 and analysed with 3-D QLab advanced software (version 7). We took 3-D four-chamber images and measured end-diastolic and end-systolic volumes and recorded heart rate. Measurements were compared with Wilcoxon signed-rank.
Results: There were no significant differences between the untreated and treated patients in degree of cardiac hypertrophy before treatment was commenced, with sepcavr 0.58 (0.30–0.86 and 0.63 (0.31–1.28; $p = 0.51$) and lvacvr 0.22 (0.17–0.68) and 0.27 (0.17–0.31; $p = 1.0$), respectively. The group receiving beta-blockers had a 30% higher left ventricular end-diastolic volume/m^2 BSA, untreated: 31.9 (22.3–43.6) ml/m^2, beta-blocker group: 41.4 (30.2–55.2; $p = 0.022$), and stroke volume showed a somewhat smaller increase, 21.9 (14.3–31.4) versus 27.9 (18.4–39.4), which did not reach significance ($p = 0.08$). Resting heart rates were not different, 76 (53–95) versus 76 (56–94). Calculated resting cardiac output was non-significantly 32% higher in the beta-blocker group, 2.13 (1.41–3.31) versus 1.61 (1.13–2.01) l/m^2 BSA for the untreated group ($p = 0.07$).
Conclusions: Beta-blocker therapy did not reduce cardiac output at rest, and the trend for improvement suggests that the increase in resting left ventricular volumes was beneficial.

878: OUTCOMES OF PATIENTS WITH TETRALOGY OF FALLOT WITH ABSENT PULMONARY VALVE SYNDROME: 37 YEARS OF EXPERIENCE

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Background: Absent pulmonary valve syndrome (APVS) is associated with varying degrees of aneurysmal dilatation of the pulmonary arteries and compression of the tracheobronchial tree, and may lead to significant respiratory compromise. We describe the outcomes of patients with APVS who underwent surgery in our unit.
Methods: A retrospective review of 51 patients with APVS who underwent surgical correction between August 1975 and August 2012 was conducted. The median age and weight at repair were 0.9 years (4 days –24.2 years) and 6.9 kg (1.8–56 kg), respectively. Pre-operative intubation was required in 15 (30%) patients and 21 (41%) required urgent surgery. The pulmonary valve was replaced with valved conduit (15, 30%) or monocusp valve (16, 31%). No valve repair was performed in 20 patients (39%). Pulmonary artery reduction/plication was performed in 38 (75%); two (4%) underwent a Lecompte manoeuvre.
Results: Operative mortality was 14% (7/51); 1975–1989, 19% (3/16); 1990–2000, 19% (4/21), and 2001–2012, 0% (0/14). Late mortality was 6.8% (3/44); 1975–1989, 15% (2/13); 1990–2000, 0% (0/17); 2001–2012, 7% (1/14). There were more Contegra valved conduits (5/14, 36%) and less valveless repairs (4/14, 29%) performed during 2001–2012 compared with earlier eras (1975–1989, 0%, 50%; 1990–2000, 0%, 38%). Less pulmonary artery reduction/plication surgery was performed in the latest era (50 vs 88 and 81% in the early and middle eras), with better antenatal diagnosis (64 vs 6 and 19%, respectively). Overall survival at five, 10 and 20 years was 81.4 ± 5.6%. On multivariate analysis, pre-operative ventilation ($p = 0.009$), prematurity ($p = 0.04$) and repair using a homograft ($p = 0.009$) were risk factors for overall mortality. Freedom from late re-operation at five, 10 and 20 years was 79.7 ± 6.9%, 69.4 ± 8.2% and 52.1 ± 9.8%, respectively. No difference was found between conduit, monocusp or valveless techniques. Risk factors for late re-operation include prematurity ($p = 0.001$) and neonatal primary repair ($p = 0.007$). Longer postoperative ventilation periods were predicted by pre-operative ventilation ($p < 0.0001$) and surgery during infancy ($p = 0.008$).
Conclusion: Long-term survival for APVS has improved significantly over the last decade. Pre-operative ventilation predicted longer postoperative ventilation and mortality.

879: COMPREHENSIVE IMAGING TOOLS: AN UNUSUAL CASE OF RIGHT ISOMERISM WITH COARCTATION

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Background: The incidence of isomerism in children with congenital heart disease (CHD) is 2.3%. Due to its complexity, right isomerism (RI) remains one of the most challenging heart lesions with a high mortality rate. RI often presents in association with severe cardiac malformations, isomeric arrangement of the bronchi and abnormalities of the abdominal organs. Frequently echocardiography is sufficient for diagnosis, but in complex cases cardiac MRI is complementary.
Case report: A 10-day-old newborn was transferred with a diagnosis of complex CHD after presenting in cardiogenic shock at another hospital. Despite several echocardiographies the morphology of the dominant ventricle had not been confirmed, and the exact anatomy of the pulmonary veins and aortic arch was unclear. A cardiac MRI confirmed RI with dextrocardia, unbalanced AVSD with a small posterior left ventricle, DORV and a small anterior aorta, hypoplastic right aortic arch with coarctation, supracardiac TAPVC draining via a mildly obstructed vertical vein to a single LSVC with Qp:Qs of 2:1. In the setting of this unusual case of RI with obstructed TAPVD in combination with left-sided outflow tract obstruction, the typical postnatal management needed to be modified.

Several treatment options were discussed, including a Norwood procedure with TAPVD correction and extended arch reconstruction. Other possibilities entertained were primary heart transplantation and compassionate care. The patient was listed for HTx, and a hybrid procedure with PDA stenting and banding of the pulmonary branches was performed. Pulmonary vein stenting was deferred to see how the patient responded to the hybrid. Two weeks later, there was increased pulmonary venous obstruction. By this time, the parents requested switching to a palliative approach, declining any further intervention.
Conclusions: While echocardiography is often able to provide a detailed cardiac imaging, MRI is superior to identify arch anomalies, the course of pulmonary veins and, in difficult cases, the ventricular morphology.

881: EXOME SEQUENCING AS PART OF A STRATEGY TO IDENTIFY IMPORTANT VARIANTS IN CONGENITAL HEART DISEASE

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Introduction: Exome sequencing is emerging as a strategy to identify potentially causal variants in families with two or more members affected by congenital heart disease. Lessons from this approach will inform future efforts to investigate the genetic contribution to sporadic forms of congenital heart disease, which constitute the majority of cases.
Methods: Commercial exome sequencing with screening-out of common variants was performed using usual bio-informatics approaches. Families with two or more affected individuals were identified from our DNA bank. Permission for exome sequencing was specifically sought.
Results: The majority of potentially damaging variants identified were in genes known as being relevant to cardiac development or previously identified as being responsible for cardiac malformations in humans or experimental mouse models. In a number of families, potentially causal variants have been identified in known gene families and pathways and are being modeled *in vitro* to assess functional impact.
Conclusions: Emerging sequencing technologies need to be paired with powerful bio-informatics resources. Translational teams including clinical geneticists and cardiac clinicians are required. Logical extension of these approaches will support personally informative genetic counselling as well as providing insights into cardiac development.

885: CLINICAL IMPLICATION OF SERUM N-TERMINAL PRO-HORMONE BRAIN NATRIURETIC PEPTIDE IN THE PREDICTION OF OUTCOME IN PAEDIATRIC DILATED CARDIOMYOPATHY

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Background: Serum levels of N-terminal pro-hormone brain natriuretic peptide (NT-proBNP) are known to be related to cardiac function. This study aimed to investigate serial changes in NT-proBNP as a prognostic factor for outcomes of paediatric dilated cardiomyopathy (DCMP) in a single tertiary centre.
Methods: Retrospective reviews included 69 DMCP patients who were treated at Samsung Medical Centre from 2004 to 2011. Echocardiographic measurements and NT-proBNP levels were serially analysed at the time of diagnosis, three months, six months, and at the last follow up. They were classified into three groups according to the last follow-up LV function: group I ($n = 35$) with left ventricular ejection fraction (LVEF) > 55%, group II ($n = 18$) with LVEF 35–55%, and group III ($n = 16$) with LVEF < 35% or mortality or heart transplantation.
Results: The median age at diagnosis was 30 months and median duration of follow up from diagnosis was 45 months. The causes of DCMP were idiopathic (69.6%), myocarditis (10.1%), tachycardia (7.2%), familial (1.4%), mitochondrial disease (7.2%), and anthracycline-induced cardiomyopathy (4.3%). There was no significant difference in NT-proBNP level between groups according to cause of DCMP. NT-proBNP was correlated to LVDD and LVSD *z*-score, LVEF at each point. Serial NT-proBNP levels showed statistically significant differences between the three groups. The levels of NT-proBNP were significantly higher in group III at three month, six month, and the last follow up, except at diagnosis. The degree of improvement in NT-proBNP level from diagnosis to any point was worse in group III. On multivariate Cox analysis, the degree of improvement in NT-proBNP level from diagnosis to six months ($p = 0.040$) was a significant predictor of adverse outcome. Patients with the degree of improvement from diagnosis to six months < 76.7% were at increased risk of severe LV dysfunction or cardiac death ($p = 0.001$).
Conclusions: The degree of improvement in NT-proBNP level at six month from diagnosis could predict adverse outcomes in paediatric DCMP and could be used a guide for long-term treatment plan.

886: CHEST ULTRASOUND IN EVALUATION OF POSTOPERATIVE LUNG OEDEMA IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Postoperative management of the patient with congenital heart disease (CHD) is influenced by pulmonary complications, such as lung oedema (LE), which may interfere with lung mechanics and worsen hypoxaemia. Precise assessment of LE by regular or phase-contrast X-ray, and serial assessment of lung compliance is difficult. Lung water content (LWC) can be probed using chest ultrasound (C-u/s) to measure wedge-shaped echodense reflections (B-lines) impinging in the echo window below the pleural demarcation. This method has been useful to diagnose high-altitude pulmonary oedema and transient tachypnoea in the newborn. Accordingly, we explored whether C-u/s can be used to assess postoperative LWC in patients with CHD.
Methods: We studied six patients with TGA and eight with acyanotic shunt defects (0.2–0.7 and 2.0–6.2 months of age, respectively). The measurements were done at < two and < 30 hours postoperatively. Static lung compliance (L-Cst) was measured under anaesthesia by the double-occlusion technique (Labmanager 4.52i; Erich Jaeger

GmbH; Germany). C-u/s was performed using an 18-MHz linear transducer (Esaote, Italy) at bilateral transverse sections of three designated segments. The digital images were blinded, the proportional area of the B-lines in each section was graded from one (0%) to five (> 75%), and a mean chest sonographic score (C-SS) of the six measurement points was used for the statistics. Statistical differences were studied using the Mann-Whitney *U*-test.
Results: TGA patients had significantly more B-lines on C-u/s postoperatively ($p = 0.01$) and on first postoperative day ($p = 0.01$) than patients with shunt defects. L-Cst did not differ significantly between patient groups.
Conclusions: Measurement of LE by ultrasound is a quick, easy and safe procedure, which may prove to be useful in postoperative evaluation of patients with CHD. Complex open-heart surgery or significant hypoxaemia, or both, may increase and prolong postoperative LWC.

890: INSIGHT INTO DYNAMIC THREE-DIMENSIONAL MITRAL VALVE GEOMETRY AND ANNULAR FUNCTION IN NORMAL CHILDREN, ADOLESCENTS AND YOUNG ADULTS: A NOVEL METHODOLOGY

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Background: We have previously reported dynamic mitral annular function in children. However this relationship to the leaflets and papillary muscles was not possible with earlier software.
Aim: To demonstrate dynamic relationships between the leaflets, the papillary muscles (PMs), and the mitral annular function throughout the cardiac cycle in a young population.
Methods: Forty healthy volunteers, with the mean age of 15.7 (3.4–38.4) years, underwent apical left ventricular full-volume imaging with real-time three-dimensional echocardiography (RT3DE) at a frame rate of 30–40 FPS. RT3DE data set was cropped into 15 slices (spaced 24 degrees) around the centre of the mitral valve. Data analysis was performed using prototype software (TomTec Inc). Leaflets and PMs were manually traced at each slice during mid-systole (MS), late-systole (LS) and late-diastole (LD), and were reconstructed as a 3D graph using our customised software (MathWorks Inc). Measurements included annular area, bending angle and height, and tethered and prolapsed volume of the leaflets, and anterolateral (APM)/posteromedial PM (PPM) angle.
Results: There was a strong correlation between the annular bending angle and the height throughout all phases of the cardiac cycle (range of p values 0.007 to < 0.001). There was a correlation between the annular area and the height during MS and LS ($p = 0.001$ and 0.004). On the other hand, there was no correlation between the PM angle and the bending angle at MS and LD, but a weak correlation between them at LS ($p = 0.04$). Both 'normal' tethered and prolapsed volume of the leaflets had a correlation with the APM angle ($p = 0.01$), but not with the PPM angle.
Conclusion: Dynamic mitral annular functions could be assessed quantitatively. In particular, the angle between the PMs and the mitral annulus was constant throughout the cardiac cycle.

894: RESOLUTION AND COMPLICATIONS OF CORONARY ARTERY ANEURYSMS AFTER KAWASAKI DISEASE

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Background: Coronary artery aneurysms (CAA) that fail to resolve by returning to a normal luminal dimension after Kawasaki disease (KD) are at high risk for both thrombotic and stenotic complications.
Methods: We reviewed the case records of 169 patients with KD and CAA (1999–2012). CAA were classified as small ($z > 2.5$–5.0), large ($z > 5.0$–10.0) or giant ($z > 10.0$) using previously published criteria. Time to resolution ($z < 2.5$) and freedom from thrombosis or stenosis were determined.
Results: For small CAA ($n = 102$), at one/five years after acute KD, the proportion of CAA showing resolution were: 57–78% and 90–100% depending on the coronary branch. For giant CAA ($n = 51$), the proportion of CAA showing resolution at one, five and 10 years were 0–7%, 15–34%, and 40–73%, respectively. Patients with small/large CAA were not at risk for either thrombosis/stenosis. Patients with giant CAA were at substantial risk (20% at two months, 33% at five years and 47% at 10 years) despite 82% on anticoagulation; 43% of thromboses had important consequences (seven thrombolytics, three myocardial infarctions, one death). Freedom from stenoses was 96/77% at one/five years. Factors associated with CAA resolution included smaller maximum CAA z-score (HR: 0.928/z, $p = 0.02$), smaller CAA longitudinal area (length × diameter × 0.8) (HR: 0.850/cm^2, $p = 0.03$) and younger age at diagnosis (HR: 0.847, $p = 0.02$). Low albumin level, high erythrocyte sedimentation rate, C-reactive protein and neutrophil levels at three months and one year after acute KD were associated with CAA persistence. Factors associated with increased risk of thrombosis were higher maximum CAA z-score (HR: 1.072/z, $p < 0.001$) and higher CAA longitudinal area (HR: 1.050/cm^2, $p = 0.02$). Factors associated with increased risk of stenoses were higher maximum CAA z-score (HR: 1.071/z, $p = 0.003$), smaller CAA longitudinal area (HR: 1.037/cm^2, $p = 0.05$) and the presence of complex (vs isolated) CAA) (HR: 9.0, $p = 0.04$).
Conclusions: Resolution of CAA is prevalent and is influenced by the location, maximum size and extent of involvement, and is more likely in younger patients. Despite aggressive thromboprophylaxis strategy, patients with giant CAA continue to be at substantial risk of thrombotic and stenotic complications.

900: PROTHROMBOTIC GENE POLYMORPHISMS IN PATIENTS WITH CONGENITAL HEART DISEASE WITH AND WITHOUT TRISOMY 21

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Background: Children with congenital heart disease represent the paediatric patient population at the highest risk of thrombosis, mainly due to exposure to multiple pro-thrombotic factors. Limited evidence regarding a genetic predisposition to thrombosis in these patients is available. We sought to determine whether patients with congenital heart disease have an increased prothrombotic polymorphism burden and whether the subpopulation with trisomy 21 has magnified risk.
Methods: A total of 381 patients with congenital heart disease were reviewed. Ninety-six SNPs on 53 genes involved in the coagulation/fibrinolysis pathways were assayed using the the Illumina GoldenGate® custom SNP panel; genotyping was successful for > 99% of SNPs. Minor allele frequency was compared to population average; a difference of 14% was considered statistically significant (corresponding to a p-value < 0.005 adjusted for multiple testing). Differences between patients with trisomy 21 and those with no known genetic abnormalities were compared with bootstrap resampling (1 000 samples, > 50% reliability) for SNP selection.
Results: Population-based minor allele frequency was available for 91 SNPs; 11 (12%) SNPs had minor allele frequency differences from population-based averages. These included seven SNPs known to be associated with increased venous thrombosis risk, including one known to affect fibrinogen levels and three associated with coagulation factor activity. Trisomy 21 was present in 27 (7%) patients, 17 (4%) had other genetic syndromes. Patients with trisomy 21 had increased frequency of TT polymorphism in coagulation factor

XIII rs5982 (19 vs 6%, $p = 0.04$), GG polymorphism in fibrinogen alpha chain rs2070006 (62 vs 38%, $p = 0.02$), CC polymorphism in coagulation factor V rs3753305 (31 vs 17%, $p = 0.05$), and AG/GG polymorphism in plasminogen rs13231 (44 vs 65%, $p = 0.04$).
Conclusions: Patients with congenital heart disease have an imbalance of prothrombotic gene polymorphisms that is magnified in the subset of patients with trisomy 21. Clinical significance of these genetic changes regarding thrombosis risk and thromboprophylaxis effectiveness should be investigated further.

902: EISENMENGER IN INFANCY: IS IT TRIGGERED BY COMBINED PRESSURE-VOLUME PULMONARY BLOOD FLOW RATHER THAN BY INCREASED PULMONARY VENOUS PRESSURE ALONE? IMMEDIATE AND MIDTERM NORMALISED PULMONARY ARTERY PRESSURES IN A SIX-YEAR-OLD CHILD WITH COR TRIATRIATUM AFTER REPAIR: A CASE REPORT

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Introduction: Eisenmenger disease describes a condition with fixed pulmonary hypertension. Mostly congenital heart malformations with increased pulmonary arterial pressure and blood flow (VSD, ASD, PDA) or elevated pulmonary venous pressure (mitral stenosis, cor triatriatum, obstructed pulmonary veins) are thought to be responsible for the irreversible remodelling of the pulmonary vasculature. If such a condition is left untreated for approximately two years, failure of normal regression of the intimal smooth muscles occurs. We report on a six-year-old child from Togo with severe pulmonary hypertension due to an untreated cor triatriatum. At rest he had slight tachypnoea of 40–45/min, thoracic deformation (cardiac voussure), and saturation in room air > 96%.
Methods: Echocardiography gave a diagnosis of a cor triatriatum with severe pulmonary arterial hypertension (TI gradient 150 mmHg, BP 100/45 mmHg, gradient over membrane of cor triatriatum 55/15 mmHg). There was no atrial or ventricular septal defect. At mild exertion (walking to out-patient clinic) there was immediate desaturation to 80% in room air, with fatigue.
Results: After surgical repair the patient showed immediate recovery from pulmonary hypertension: 1/3 pulmonary arterial pressure while coming off bypass circulation with Milrinon but without antihypertensive treatment (NO, prostacyline). After six weeks, there was improved physical performance with no desaturation while walking. Echocardiography showed no evidence of pulmonary hypertension.
Conclusion: Excessive high pressure-volume pulmonary blood flow is most harmful for the pulmonary vascular bed and leads to early fixed pulmonary hypertension. This case illustrates that increased pulmonary venous pressure alone related to obstructive lesions such as a cor triatriatum behave haemodynamically similar to severe mitral stenosis in adults. In contrast to the high pressure-volume state in large shunt lesions that develop usually over a period of 24 to 48 months, in fixed pulmonary hypertension, these patients obviously have a greater potential to recover from pulmonary hypertension regardless their age.

903: INCREASE IN USE OF POOLED HUMAN ANTITHROMBIN REPLACEMENT THERAPY IN PAEDIATRIC PATIENTS

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Background: Antithrombin is an essential part of the coagulation system. A number of paediatric patients, including those with congenital heart disease, may have low antithrombin activity. For patients with critically low antithrombin activity, endogenous human antithrombin from pooled donors is available for replacement therapy. We sought to determine current trends in the use of antithrombin supplementation in paediatric patients.
Methods: Hospital records of all patients who received antithrombin supplements at the Hospital for Sick Children between 2002 and 2011 were reviewed. Indication for antithrombin use was classified as replacement therapy for cardiac patients [non-extracorporeal membrane oxygenation (ECMO)], patients supported on ECMO and for non-cardiac/non-ECMO patients.
Results: A total of 551 paediatric patients received 1 912 courses of antithrombin replacement therapy, of which 315 (57%) were cardiac patients not on ECMO, 116 (21%) were patients supported on ECMO and 121 (22%) were non-cardiac patients. Nearly half (48%) of all patients receiving antithrombin were neonates (< 31 days), 32% infants (31 days – 1 year), 10% young children (1–**9** years) and 10% adolescents (10–18 years). A higher proportion of neonates were in the cardiac, non-ECMO patient group (52 vs 43%, $p = 0.03$). Median baseline blood antithrombin level was 46% (25th–75th percentiles: 32–61%). Number of patients receiving antithrombin increased from 33 in 2002 to 81 in 2011 (+5 patients/year, $p = 0.002$). During this period, use of antithrombin for cardiac patients not on ECMO remained stable (+0.6 patient/year, $p = 0.38$) as did use for non-cardiac patients (+0.4 patient/year, $p = 0.16$). However, use of antithrombin supplementation in the setting of ECMO significantly increased (+3 patients/year, $p = 0.02$) during the study period.
Conclusions: Antithrombin use has been increasing in recent years, primarily in patients on ECMO, despite the lack of high-quality studies evaluating safety and efficacy. Future studies are needed to determine proper indications and outcomes in these populations.

905: INTRACARDIAC THROMBUS IN PAEDIATRIC PATIENTS WITH DILATED CARDIOMYOPATHY

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Introduction: Intracardiac thrombosis (ICT) in patients with dilated cardiomyopathy (DCMO) is a serious complication with associated significant morbidity and potential mortality. This study investigates the incidence, risk factors and outcome of ICT in children with DCMO. A retrospective review of clinical records was performed at two tertiary centres for all children with DCMO.
Method: This was a **r**etrospective review of paediatric cardiology databases. All DCMO patients between 0 and 14 years seen between January 1983 and December 2010 at Chris Hani Baragwanath Academic Hospital (CBAH), Johannesburg, and between January 2003 and December 2011 at Inkosi Albert Luthuli Central Hospital (IALCH), Durban, both tertiary institutions in South Africa, were assessed.
Results: An ICT was found in 39 (10.8%) of the 361 patients seen at CBAH and in 13 (10.5%) of the 123 patients seen at IALCH. The ICT was located within the left ventricular cavity in the majority of patients. Only one patient had ICT in the right ventricular outflow tract. All patients had poor left ventricular systolic function with fractional shortening (FS) below 18%; 32% presented with an acute thromboembolic event. All patients with ICT were anticoagulated with heparin initially before changing to warfarin.
Discussion: The development of ICT in patients with DCMO occurs with stasis of blood from impaired left ventricular function, an abnormal pro-coagulant endocardial surface, arrhythmias and heritable hypercoagulable states. Clinical studies report a frequency of 4–16% of ICT in patients with DCMO, with a much higher incidence of 43–57% in post mortem reports. Our retrospective review shows a similar high incidence of ICT in patients presenting with DCMO who are at risk of thromboembolic complications.

Conclusion: Patients with DCMO with poor left ventricular function are at high risk for ICT and should undergo routine echocardiographic surveillance. While no firm paediatric guidelines exist, anticoagulation has been recommended for patients with FS below 20%.

906: SPECTRUM OF RHEUMATIC FEVER AND HEART DISEASE IN A SPECIALIST CARDIAC SERVICE IN THE UNITED KINGDOM

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Introduction: Acute rheumatic fever and rheumatic heart disease remains a public health concern in the developing countries, however, with population migration, it is not unusual in developed countries.
Methods: We reviewed all the patients referred to the Cardiac Unit and diagnosed and treated as acute rheumatic fever or recurrence over a 10-year period diagnosed by modified Jones criteria.
Results: Over a 10-year period, we saw 33 cases of acute rheumatic fever or recurrence of rheumatic activity. Mean age 10 years (range 5–14 years), with a male preponderance (eight girls, 25 boys); 25 patients were of a different ethnic background with the majority being South Asian or British Asian (11). All patients had carditis of varying degrees, manifest predominantly as valvulitis. Mitral and/or aortic regurgitation was the most common lesion, with mitral stenosis being rare; 55 % had polyarthritis, 13 % had chorea and none had subcutaneous nodules. Four patients required surgery in the form of mitral valve repair or replacement (four patients) and one had concomitant aortic valve repair. All patients with clinical carditis were treated with bed rest, oral steroids and then salicylates for a total period of eight weeks. Twelve patients had been undertreated with steroids before. Secondary prophylaxis with oral penicillin over a follow-up period of a mean of 4.5 years (range 2–12) led to no rheumatic recurrences.
Conclusion: Although the prevalence of rheumatic heart disease is high in certain parts of the developing world, with population migration, the disease is still seen in low-prevalence areas. Carditis and arthritis are common with chorea and subcutaneous nodules are rare.

997: HETEROTAXY SYNDROME: IS A PROPHYLACTIC LADD PROCEDURE NECESSARY IN ASYMPTOMATIC PATIENTS?

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Background: Complex congenital heart disease and abnormalities of intestinal rotation are commonly associated with heterotaxy syndrome (HS). Malrotation is the most worrisome intestinal rotation abnormality (IRA) due to the risk of bowel ischaemia and infarction. There is controversy whether asymptomatic infants with HS require screening for IRA and if present, whether a prophylactic Ladd procedure is indicated. As this population grows due to continuing advances in cardiovascular care, it is vital to better understand the natural history of IRA and devise an evidence-based treatment model. The first objective of this study was to determine institutional practice in the management of asymptomatic infants with HS and IRA. The second was to prospectively observe a cohort with HS and evaluate their long-term outcomes.
Methods: We have begun a prospective, multi-centre, observational study using a web-based database to follow infants with HS to five years of age. Data collection includes screening methods used for the detection of IRA, management of IRA, cardiac diagnosis, cardiac interventions, long-term complications and outcomes. Patient management will not be dictated by the study protocol, given the observational design.
Results: Ethics and scientific approval has been obtained at two centres in Canada and six patients have been enrolled to date. Fourteen other centres across North America and the United Kingdom are currently obtaining ethics approval. Additional centres are being actively recruited.
Conclusions: Results from this study may change general surgical practice in the management of this complex group of patients. Establishment of this cohort will facilitate future studies of the HS population with regard to gastrointestinal and cardiovascular outcomes.

909: AN EVALUATION OF THE REPRODUCIBILITY AND INFLUENCE OF PROCESS FACTORS ON AORTIC INTIMA–MEDIA THICKNESS MEASUREMENTS BY TRANS-ABDOMINAL ULTRASOUND IN YOUNG INFANTS

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Background: Aortic intima–media thickness (aIMT) is a novel parameter increasingly used as a marker of early atherosclerosis. The reproducibility of aIMT measurement by trans-abdominal ultrasound in infancy has been established in small, tertiary hospital-based studies, but there are no data from trained staff in a community-based setting. In addition, there are no data on whether environmental and infant behavioural factors influence the reproducibility of aIMT measurements.
Methods: The Barwon Infant Study (n = 1 250) is a regional non-selected Australian birth cohort. Aortic IMT is measured on four-week-old infants by two trained research staff using trans-abdominal ultrasound (GE vivid I™ with vascular probe). A subset of babies has aIMT performed by both staff to assess inter-observer consistency. Data were collected on infant behaviour, sleep/wake cycle, recent feeding, sucrose use, and presence of siblings. Two analysts, blinded to the other's measurements, quantified image quality and aIMT using Echopac software. Results were assessed for inter-observer consistency between (1) sonographers and (2) analysts using Pearson's correlations, Bland-Altman plot and $2 \times n$ Chi^2 analysis.
Results: Among the first 292 babies, aIMT approximated a normal distribution (mean 0.564 mm, SD 0.06 mm). The inter-observer correlation of aIMT measurements from infants scanned by both sonographers (n = 17 to date) was 0.8449, $p < 0.001$. The inter-operator correlation between measurements of aIMT made by both analysts (n = 115 to date) was 0.8783, $p < 0.001$ (mean difference 0.004 mm, SD 0.028). Infant and environmental factors did not affect image quality.
Conclusions: In the setting of a large-scale population-based study, aIMT measurement by trans-abdominal ultrasound in young infants was reproducible and unaffected by environmental and behavioural factors examined. (These are interim data. Data on 500 subjects will be available by February 2013.)

915: COMPARISON OF ECHOCARDIAGRAPHIC AND ELECTROCARDIOGRAPHIC RISK FACTORS FOR THE PREDICTION OF SUDDEN DEATH IN PAEDIATRIC HYPERTROPHIC CARDIOMYOPATHY

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Background: Hypertrophic cardiomyopathy (HCM) is the most common cause of sudden unexpected death in childhood and among athletes. Success at attempting to prevent sudden arrhythmia death rests on identifying individuals at increased risk, but most clinically used strategies for risk stratifications are based on research in adult HCM patients
Methods: From the complete Swedish national material on patients who have died suddenly due to HCM, patients diagnosed before 20 years of age, and with sudden death (SuD) occurring before 30 years of age were retrieved (SuD group; $n = 26$) and compared with a well characterised complete regional cohort of paediatric HCM survivors from west Götaland region (Surv group; $n = 46$). Previously published risk features were compared.
Results: The groups were comparable in terms of age at diagnosis (SuD group 9.7 ± 5.9 years; Surv group 9.1 ± 6.5, mean ± SD), and duration of follow up was not significantly different (7.4 ± 5.9 vs 10.2 ± 8.8 years). The adult criterion of wall thickness of ≥ 3 cm has a low sensitivity of 33%, and a relative risk of 2.9 (95% CI: 1.5–5.2). Two paediatric wall thickness criteria are much better: a wall thickness ≥ 2 cm, relative risk 8.2 (3.1–32.3), sensitivity 91%; and a septal thickness of ≥ 190% of 95th centile value for age: relative risk 7.1 (2.3–21.9) and sensitivity of 84%; the latter has a better specificity of 74% compared to 63% for ≥ 2 cm. Among electrocardiographic risk markers, an electrocardiographic risk score ≥ 6 points (European Heart J 2010; 31:439) gives a relative risk of 21.2 (3.0–148), a sensitivity of 96% and a specificity of 78%. A 12-lead QRS amplitude × duration product ≥ 2.2 mV/s has a relative risk of 74.7 (4.3–1303), a sensitivity of 100% but a somewhat less good specificity of 59%.
Conclusions: The best electrocardiographic risk markers discriminate better than any wall thickness criteria for the risk of sudden arrhythmia death in paediatric HCM.

916: CONGENITAL HEART DISEASE IN NIGERIAN CHILDREN: A MULTICENTRE EXPERIENCE WITH 605 CHILDREN

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Background: Congenital heart disease (CHDx) is among the leading causes of morbidity and mortality in childhood with a global incidence of 3.5 to 11.3 per 1 000 live births. We report the findings on the spectrum of echocardiographically diagnosed CHDx from three different centres across Nigeria.
Methods: Over a period of 42 months, children who were referred for echocardigraphic evaluation in the centres located in three large metropolitan cities were consecutively recruited if they were confirmed to have identifiable CHDx. The data was collected on age, gender, types of CHDx and analysed using SPSS 16 (Chicago IL, USA).
Results: A total of 605 children were recruited. Of these 296 (48.9%) were males and 494 (81.7%) had acyanotic congenital heart disease (ACHDx). The mean age of the study population was 2.1 ± 3.5 (range 0–17) years. Almost half of the children (42.5%) were diagnosed within the first year, only 17% within the neonatal period. The gender was evenly distributed. The commonest CHD was VSD (46.6%) followed by ASD (11.7%), PDA (10.6%) and TOF (7.8%). Over half (55.0%) of the VSD were peri-membranous, and the outlet variety accounted for 24.5%. Complex CHDx accounted for 7.7%.
Conclusion: VSD is the commonest CHDx, as has been previously reported. There is a high proportion of the outlet variety of VSD in our study population. There is increasing awareness, availability and use of diagnostic facilities as mirrored in the age distribution of the children. However access to definitive surgery is poor and draws attention to the urgent need for affordable surgical facilities in the country.

924: THE SUCCESSFUL MANAGEMENT OF FAUCIAL DIPHTHERIA TOGETHER WITH MYOCARDITIS AND SEPTIC DIPHTHERIA AS COMPLICATIONS

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Background: Severe diphtheria epidemics with high mortality rates have been recorded since the 16th century and the implementation of diphtheria immunisation has led to marked decreases in some countries. But in 2011, Indonesia had the second highest number of cases.
Objective: To describe a comprehensive management of faucial diphtheria with myocarditis and septic diphtheria as complications in an unimmunised child.
Case report: A five-year-old girl was referred from Bangil General Hospital because of seizures, parotid gland enlargement due to a suspected diphtheria infection and encephalitis. The physical examination found her delirius with nasal flares, a bull neck and pseudo membranes on both enlarged tonsils. Initial laboratory findings showed slight anaemia and severe neutropenia with lymphocyte dominance. For early treatment she was put on a ventilator, given ADS 100 000 IU, dexamethasone intravenously and an intramuscular of penicillin procaine injection 1.5 million IU. On the fourth day, she had myocarditis as a complication. Methylprednisolone 4 mg/kg/day was administered and then continued at 3 mg/kg/day for a week. The first routine throat swab culture was positive for *C diphtheriae* and her father was a carrier. After 11 days of treatment, the methylprednisolone was tapered off. The results of the second to fourth routine throat swab culture showed no more *C diphtheriae*. The FNAB from the persistant bull neck found a neck abscess. Incision and drainage were performed. After one month of treatment, she was given DPT immunisation and discharged.
Conclusion: A complicated case of diphtheria required comprehensive management to achieve an optimum outcome.

927: PERSISTENT FIFTH AORTIC ARCH: A CLINICAL CONUNDRUM

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Background: Persistence of the embryonic fifth aortic arch is a rare and enigmatic condition with variable anatomical forms and physiological consequences. As such, it may be significantly underdiagnosed as a primary pathology.
Methods and Results: A search of the surgical database revealed five cases of persistent fifth aortic arch (PFAA). Four cases were diagnosed with congenital heart disease antenatally; one case presented at six weeks of age. In two cases there was isolated PFAA with significant left-to-right shunting causing heart failure. In another two cases the PFAA provided the only source of pulmonary blood flow in obstructive right heart lesions. In these four cases the PFAA mimicked the arterial duct, which was absent in all. The PFAA was sensitive to prostaglandin E_1 in one case with pulmonary atresia. The two cases with isolated PFAA were treated by surgical ligation avoiding the need for cardiopulmonary bypass and the two cases with pulmonary atresia have been successfully repaired. The PFAA in the fifth case was wide and co-existed with a patent arterial duct, interruption of the aortic arch ('type A'), and severe pulmonary stenosis.

Being rather wide, the PFAA communication was thought initially to be an aorto-pulmonary window. The patient died from pulmonary infarction several days after surgical repair.
Conclusions: Although rare, recognition of PFAA in all its various forms is important for clinical management. The PFAA in our first four cases was a vessel arising from an unusually proximal position from the aorta to connect to the pulmonary artery. This vessel could be mistaken for an arterial duct but it has unpredictable reactivity to prostaglandin E_1. The differential diagnosis for the fifth case was an aorto-pulmonary window but its length and pulmonary communication at the bifurcation were against this diagnosis. The co-existence of arch interruption in this case made diagnosis particularly challenging.

944: OUTCOMES OF PATIENTS UNDERGOING OPEN-HEART SURGERY AT THE UGANDA HEART INSTITUTE, MULAGO HOSPITAL

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Background: Heart disease is a disabling condition and the necessary surgical service is often lacking in many developing countries. A novel approach in which open-heart surgeries are conducted locally by visiting teams alongside skills transfer to build local capacity has been adopted at the Uganda Heart Institute (UHI).
Objectives: We evaluated the progress of open-heart surgery at the UHI, to describe the postoperative outcomes and challenges faced in conducting these surgeries.
Methods: Medical records of patients undergoing open-heart surgery at the UHI from October 2007 to June 2012 were reviewed.
Results: A total of 124 patients underwent open-heart surgery during the study period. The commonest conditions were VSDs in 35.5% (44/124), ASDs in 34.7% (43/124) and TOF in 10.5% (13/124). Foreign charities funded 82.3% (102/124) of the operations, and only four patients (3.2%) paid for their surgeries. A local NGO funded the rest. There was increasing complexity in cases operated from predominantly ASDs and VSDs at the beginning, to more complex cases such as TOFs and TAPVR later on. The local team independently operated on 19 patients (15.3%). Postoperative morbidity was low, with left ventricular dysfunction and re-operations occurring in six (4.8%) patients each. Sepsis occurred in only two cases (1.6%). The 30-day mortality was 3.2 %
Conclusion: Open-heart surgery, although expensive, is feasible in a developing country. Governments and local charities should direct funding to support treatment of more children with heart disease locally as opposed to referral abroad to increase access to the service.

945: PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE IN PAEDIATRIC AND ADULT SPANISH POPULATIONS: DATA FROM THE SPANISH REGISTRY FOR PAEDIATRIC PULMONARY HYPERTENSION (REHIPED) AND SPANISH REGISTRY FOR PULMONARY ARTERIAL HYPERTENSION IN ADULTS (REHAP)

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Background: Pulmonary arterial hypertension is a serious complication of congenital heart disease (PAH/CHD), but there is not enough data about clinical profiles and survival of these adult and paediatric patients. Our objective was to analyse clinical and survival data in adult and paediatric Spanish patients with PAH/CHD.
Methods: Voluntary reporting was used of 338 adults diagnosed with PAH/CHD from January 1998 to April 2012 from the REHAP programme and 105 children diagnosed from January 2009 to June 2012 from REHIPED. Clinical classification for PAH/CHD was used: (1) Eisenmenger, (2) non-restrictive shunt with high PVR, (3) small shunt with high PVR , (4) corrected CHD without residual shunt. Kaplan-Meier curves were analysed, censoring patients at death/transplantation.
Results: We included 443 patients. Mean age was 31 ± 17 years in the adult registry and 5.5 ± 5 years in the paediatric one. Paediatric patients had worse functional class (FC) ($p = 0.003$), and more comorbidities ($p < 0.05$), but they showed higher cardiac index ($p = 0.015$), and lower mean pulmonary arterial pressure ($p < 0.001$). The most frequent forms in adults were Eisenmenger (68%) and operated shunts (20%), while in paediatrics, non-restrictive shunts with high PVR (33%) or operated shunt (33%) were the most common forms ($p < 0.001$). Nevertheless, survival was similar in paediatric and adult patients: 91, 85, and 80% at one, three and five years from diagnosis. For the whole population, we found significant differences in five-year survival according to FC at diagnosis (90% in FC I/II, 74% in FC III, 50% in FC IV, $p < 0.001$), and in Eisenmenger versus operated CHD (94 vs 72%, respectively, $p = 0.038$).
Conclusions: (1) Clinical profiles in CHD-PAH differ significantly between paediatric and adult patients, but survival was similar in both groups. (2) In the whole population, predictors for worse survival were FC III/IV at diagnosis and operated CHD.

951: THE EPIDEMIOLOGICAL AND CLINICAL PROFILE OF KAWASAKI DISEASE IN WESTERN AUSTRALIA: A 30-YEAR POPULATION-BASED STUDY

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Background: The current epidemiology of Kawasaki disease (KD) in Australia is poorly understood. Previous enhanced national surveillance (1993–1995) gave an estimated incidence of 3.7/100 000 children aged 0–4 years. In non-Asian countries, the incidence varies between 3.6 in Denmark, 8.1 in the UK, 17.1 in the USA and 26.2 in Canada, although many data are regional rather than national.
Materials and Methods: We identified all patients hospitalised in western Australia (population ~2.2 million, of whom 3.4% self-identified as indigenous) between 1979 and 2009 with an ICD discharge diagnosis of KD. We retrieved demographic, clinical, laboratory and echocardiographic data from individual patient files. Age-specific population estimates were from national census data; 95% confidence intervals were calculated assuming a Poisson distribution.
Results: We identified 353 KD cases. Male-to-female ratio was 1.7:1 and the median age was three years (IQR 12, 60 months). The mean annual incidence increased from 2.82 per 100 000 children aged 0–4 years (95% CI: 1.93–3.99) in 1980–1989, to 8.04 (6.55–9.76) in 1990–1999, to 9.66 (8.01–11.55) in 2000–2009. The highest incidence in the 0–4 age group was 15.7 in 2005. Incomplete KD was diagnosed in 40/353 (11.4%) patients. Of the 288 children who had complete echocardiographic studies, 42/288 (14.6%) had coronary artery (CA) ectasia/dilatation and 26/288 (9%) had coronary aneurysms. The only significant risk factor for CA involvement was Asian ethnicity. There were no KD cases in indigenous children (5.8% of WA population).

Conclusions: KD epidemiology in WA mirrors that in other industrialised, predominantly Caucasian populations. The rising incidence reflects both improved ascertainment and a real increase in disease burden. The current Australian incidence of KD is two- to three-fold higher than previously reported and comparable to that in the UK and US. The CA outcomes, which include the pre-IVIG era, are comparable to those reported elsewhere.

958: MIDTERM RESULTS OF MITRAL VALVE REPAIR FOR RHEUMATIC HEART DISEASE IN CHILDREN

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Objective: To analyse the techniques and midterm results of mitral valve repair in children with rheumatic mitral valve disease.
Methods: The study population includes 67 patients who underwent mitral valve repair for rheumatic mitral valve disease in the paediatric age group of under 18 years in our institution. The group included 36 female and 31 male patients. The age varied from six to 18 years with a mean age of 14 years; 43 patients had only mitral regurgitation, 18 had mixed lesions, and five had pure mitral stenosis; 71 % of patients were in NYHA class III or IV.
Discussion: All patients underwent mitral valve repair under CPB with aortic and bicaval cannulation, with the core cooled to 28°C. The approach to the mitral valve included both left atrial and superior septal. We performed annuloplasty using rings, Teflon strip and suture only in 63 patients. Neo-chordae was created in 22 patients using PTFE suture. Chordal transfer was done in 11 patients. Open mitral valvotomy was done in 13 patients. Intra-operatively, the valve was checked by saline injection and TEE. Two years follow-up study was done for those patients. Six patients were lost to follow up. Postoperative echo revealed severe MR in three patients, moderate MR in nine, mild MR in 31 and trivial MR in 18 patients. Four patients were re-operated.
Conclusion: Mitral valve repair in rheumatic patients can be performed with acceptable mortality and good midterm results. We believe valve repair should be preferred over valve replacement for rheumatic patient in the paediatric age group to avoid problems related to anticoagulation and prosthetic valve function.

975: LONG-TERM SURVIVAL AND FONTAN COMPLETION RATES AFTER SURGERY FOR UNIVENTRICULAR HEART DEFECTS

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Background: To analyse long-term outcome after surgery for univentricular heart defects.
Methods: All 320 patients (133 girls, 187 boys) operated due to univentricular heart defects in our institution before the age of 18 years from 1 January 1994 to 1 January 2010 were included. Patient files were studied and cross-checked as of 1 January 2012 against the National Population Registry in Sweden, allowing for reliable and complete data on survival. Two patients (0.6%) emigrated and were lost to follow up three and six years after Fontan completion.
Results: Median age and weight at first surgery was 11 days (0–15.1 years) and 3.6 kg (1.2–30). Median age of survivors at follow up was 10.8 years (2.1–30.7); 87 deaths (27.1%) occurred with a median age at death of 69 days (3 days – 22.7 years). Median survival time in deceased patients was 29 days (0–11.3 years) after the last major surgery. In all 276 patients with their first surgery in 1994–2009, mortality was 53/124 (42.7%) in patients with classic hypoplastic left heart syndrome, unbalanced atrial septal defect or critical aortic stenosis with left ventricular hypoplasia (A), 12/94 (12.7%) in patients with tricuspid atresia, double-inlet left ventricle or pulmonary atresia with intact septum (B) ($p < 0.001$). Mortality rate in children with other complex heart defects (C) was 19/68 (27.9%). In these subgroups the number of patients alive with completed Fontan circulation was 59/124 (47.6%) (A), 73/94 (77.7%) (B), 38/68 (58.9%) (C). Nineteen patients had a heart transplantation with four late deaths.
Conclusions: Overall survival was 72.9%. Patients with a morphological left ventricle as systemic ventricle had a high survival (87.3%) and Fontan completion rate (77.8%). The corresponding figures were less favourable in patients with a systemic right ventricle (57.3 and 47.6%) and in those with other complex univentricular heart defects (72.1 and 58.9%).

982: ANATOMY OF THE CORONARY ARTERIES AND AORTIC ARCH AND ITS RELATION TO TRUNCAL VALVE DYSPLASIA

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Background: Surgical correction of the common arterial trunk (CAT) requires knowledge of the particularities of the malformation, such as the anatomy of the truncal valve, coronary arteries and aortic arch. The truncal valve may show dysplastic leaflets. Anomalies of the coronary arteries and of the aortic arch are also described as frequent findings in CAT. In this study, we sought to check for possible associations of truncal valve dysplasia with coronary and aortic arch anatomy.
Methods: Forty-three heart specimens were analysed with CAT. Coronary orifices were expressed regarding their number, location and shape. Anatomy of the aortic arch was described as normal, interrupted or coarcted, double, right-sided and presenting an aberrant subclavian artery. The truncal valve was described regarding the number of leaflets and, according to its thickness and symmetry, classified as mildly dysplastic, moderately dysplastic, severely dysplastic or without dysplasia.
Results: Thirty-one (72%) specimens presented at least a mild degree of dysplasia, three (6%) presented an abnormal number of coronary orifices (two single and one triple ostium) and 17 (40%) presented at least one type of aortic arch anomaly. Truncal valve dysplasia was associated with the number of leaflets other than three ($p = 0,022$). Anomalous shape of the coronary orifices was predominantly described as slit-like for the right coronary orifices and as funnelled for the left coronary orifices. Anomalous shape of the right coronary orifice was associated with both dysplasia and abnormal number of leaflets ($p = 0.038$ and $p = 0.025$, respectively). Specimens with abnormal anatomy of the aortic arch also showed a trend to present abnormal numbers of leaflets ($p = 0.072$).
Conclusion: Anomalies of the coronary ostia and aortic arch seemed to be related to the dysplasia of the truncal valve. This correspondence should alert both the echocardiographist and the cardiac surgeon to the diagnosis and management of associated lesions.

988: AN INFANT WITH MITRAL VALVE ENDOCARDITIS CAUSED BY SACCHAROMYCES SP: AN UNUSUAL CASE AND SUCCESSFUL MEDICAL TREATMENT

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Background: Invasive fungal infection (IFI) is an emerging complication in paediatric population. Its incidence has recently increased,

especially in patients admitted to neonatal and paediatric intensive care units (NICU/ PICU). Fungal endocarditis is an IFI with high risk of complications and death. Several risk factors such as prolonged hospital stay, use of central venous catheters and broad-spectrum antibiotics, and the particular vulnerability of the immunological immaturity of the infants, especially preterm newborns, have been identified. In these cases, even non-pathogenic strains might be identified with related risk factors.
Methods: We reviewed paediatric case report with unusual fungal mitral valve endocarditis treated medically in Colombia.
Results: We present a two-month-old male infant with a history of complications associated with 28 weeks' prematurity, prolonged hospitalisation in the NICU/PICU, episodes of early and late neonatal sepsis, necrotising enterocolitis complicated with short-bowel syndrome transiently treated with probiotics. He was managed with broad-spectrum antibiotics, and had a central venous catheter. He developed mitral valve endocarditis caused by *Saccharomyces* sp, which was isolated from several blood cultures. He was treated initially with fluconazole and changed to deoxicolato of amphotericin B based on laboratory susceptibility, with complete clinical and microbiological recovery. Surgical management was not performed and differed due to the patient's condition.
Conclusions: Isolation of this non-pathogenic yeast, widely used as probiotic, as a cause of endocarditis in a patient with multiple risk factors such as our patient's case has been infrequently reported. Nevertheless, it suggests that there are certain circumstances in which these probiotics should be used with caution.

989: LONG-TERM SURVIVAL AFTER SURGERY IN PATIENTS WITH CONGENITAL HEART DEFECTS AND DOWN SYNDROME

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Background: To collect reliable and complete data for evaluation of long-term survival after paediatric cardiac surgery in patients with Down syndrome.
Methods*:* All 288 patients (131 girls, 157 boys) operated due to congenital heart defects in our institution before the age of 18 years, from 1 January 1994 to 1 January 2009 were included. During the study period, nearly 50% of all paediatric cardiac surgery in Sweden was performed at our institution. Patient files were cross-checked as of 1 Janury 2012 against the National Population Registry in Sweden, allowing for reliable and complete data on long-term survival. Two patients emigrated and were lost to follow up.
Results: Median age at first surgery was 0.44 years (0.01–15.69 years); 142 (49%) had surgery for atrioventricular septal defects (AVSD), 72 (25%) for ventricular septal defects (VSD), 13 (5%) for isolated tetralogy of Fallot and seven for AVSD and tetralogy. Six patients had univentricular heart palliations. Eleven of the patients with AVSD/VSD had a pulmonary artery banding as the first operation. Median age of survivors at follow up was 10.6 years (3.3–32.6 years); 18 deaths (6.3%) occurred with a median age at death of 1.6 years (0.06–21.36 years). The cause of death was pulmonary hypertension in four, and septicaemia in five. One patient operated for a VSD at the age of four months also had biliar atresia and had a liver transplant. He died at the age of three years. One patient died in an accident. There were seven cardiac deaths. Fifteen patients developed complete AV-block postoperatively, needing a pacemaker. Eleven patients developed significant mitral valve regurgitation and had a mechanical valve prosthesis inserted.
Conclusions: With 93.7% of the patients alive at follow up, long-term survival was favourable. The majority of deaths (78%) occurred after 30 days postoperatively, reflecting the need for long-term follow up.

990: THE AMAZING RIGHT BUNDLE BRANCH BLOCK: A VANISHING ACT WITNESSED IN MOTION

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We report a three-year-old male with VSD and aortic insufficiency who acquired a RBBB after surgical repair and we have captured its real-time resolution on surface ECG during a follow-up clinical visit. The initial postoperative visit 12-lead ECG showed a complete RBBB with a QRS duration of 160 ms. A 12-lead ECG at a subsequent follow-up visit showed normal sinus rhythm with rSR' pattern noted in V1, slurred S in lead I, V1 QRS duration of 160 ms, and T wave that was deflected opposite the terminal deflection of the QRS complex defining the patient's right bundle branch block. Interestingly, the RBBB changed morphology during the electrocardiogram on the V1 rhythm strip to an incomplete RBBB pattern (QRS duration = 80 ms).

This ECG captures the resolution of a RBBB after VSD surgical repair in real time. Subsequent ECGs confirmed the resolution of the RBBB. The mechanism behind a RBBB secondary to surgical VSD repair is usually due to operative injury to the proximal right bundle, Purkinje fibre network, or distal branch or branches of the right bundle from where the patch is introduced. This type of damage is unlikely to resolve. Transient RBBB in postoperative VSD patients is most likely due to a different mechanism such as local inflammation or oedema. This case demonstrates the need to closely follow ECGs after surgical VSD repair and that damage to the conduction system may not be permanent.

Since RBBB may eventually cause diastolic dysfunction in this group of patients, resolution of the block is not trivial in nature. The resolution of this conduction delay shows that inflammation and oedema may play a role in RBBB, even months after surgery.

1000: MIDTERM FUNCTIONAL OUTCOME AFTER REPAIR OF ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA)

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Background: Treatment options for ALCAPA repair include direct implantation of the origin of the left coronary artery into the aorta, and the Takeuchi procedure, which involves creation of an aortopulmonary window and a tunnel that directs blood from the aorta to the anomalous left coronary artery. The patient outcome and the incidence of residual and new lesions after the two types of ALCAPA repair were analysed.
Patients: From 1995–2012 a total of 21 patients (76.2% female) underwent ALCAPA repair at our institution. Four (19%) had a Takeuchi procedure and 17 (81%) direct implantation of the LCA. Concomitant procedures included mitral valve repair (n = 2) and ASD closure (n = 1). The mean patient age at time of surgery was 8.3 months (range: first day of life – 41.4 months).
Results: Surgical and late mortality rate was zero. The mean length of follow up after surgery was 6.2 years (range: 0.4–10.9 years). Mean postoperative LV fractional shortening at last follow up was 38.3 ± 5.6% and mean LVEDD was within normal limits. Reasons for re-operations included residual mitral regurgitation (MR) (n = 1) and baffle leaks (n = 2). The freedom from re-operation was 81% at five years. Mild main pulmonary artery stenosis was documented in three patients after Takeuchi repair. Three patients had a moderate degree of residual mitral regurgitation; the remaining patients were free of MR or had only a mild degree.
Conclusions: ALCAPA repair can be performed with low surgical mortality. In the majority of patients after ALCAPA correction, both ventricular function and mitral valve regurgitation normalise over time. Therefore concomitant mitral reconstruction may not be required upon initial repair. Coronary transfer is our preferred

technique today because the Takeuchi procedure is associated with a higher rate of re-operations and residual lesions.

1006: ANOMALOUS ORIGIN OF CORONARY ARTERIES FROM RIGHT PULMONARY ARTERY

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Background: Anomalous origin of coronary arteries from the right pulmonary artery AOCARPA i**s** a rare variant of anomalous left coronary artery from the pulmonary artery AlCAPA. It is essential to recognise this anomaly since it causes higher morbidity/mortality of the associated congenital cardiac lesions; also correction of the associated congenital cardiac defect could reduce left coronary artery flow.
Methods: We describe the cases of AOCARPA that were diagnosed at a tertiary care referral cardiac centre during 1990–2012.
Results: Forty-five patients had ALCAPA, three patients (7%) were diagnosed with AOCARPA. All underwent surgical re-implantation of AOCARPA; patient two had uneventful convalescence in spite of being diagnosed late postcoarctation repair, patient three died due to sepsis postoperatively, and patient one with a single coronary from RPA died during surgery.
Conclusions: AOCARPA is a rare subset of ALCAPA where diagnosis is suspected by clinical/echocardiographic findings and confirmation requires cardiac angiography. It is usually associated with other congenital heart disease and carries higher mortality rates.

1016: EXERCISE TRAINING IN CHILDREN AND YOUNG ADULTS WITH A CORRECTED TETRALOGY OF FALLOT: PRELIMINARY RESULTS OF THE TOFFIT STUDY

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Objective: To study whether exercise training in children and young adults with corrected tetralogy of Fallot can improve exercise capacity, physical activity and ECG markers.
Methods and Results: The study is a multicenter, randomised, controlled trial. After randomisation the participants were assigned to an interventional or control group. In total, 92 participants were recruited from five participating university medical centres. All participants underwent a cardiopulmonary exercise test, an electrocardiogram and wore a five-day activity monitoring device. The interventional group followed 12 consecutive weeks of supervised aerobic exercise training, three times a week, for an hour at the level of 60–70% of heart rate reserve. The results of the first 26 participants were included in an interim analysis. Baseline characteristics of the intervention group consisting of 14 participants were: age 16.1 ± 2.4 years, BMI 21.3 ± 3 kg.m^2, 10 males; baseline characteristics of the control group consisting of 12 participants were: age 17.6 ± 3.0, BMI 20.6 ± 3.2 kg/m^2, nine males. VO_{2peak}/kg of the intervention group improved significantly (34 ± 6 vs 38 ± 8 ml/kg/min, $p < 0.03$) whereas the control group did not show any change (35 ± 7 vs 34 ± 5 ml/kg/min). Activity levels did not change in either group. No significant ECG changes were seen (QRS duration intervention group 128 ± 27 vs 127 ± 26 ms; control group 128 ± 24 vs 125 ± 27 ms, QTc duration intervention group 432 ± 29 vs 427 ± 27 ms, control group 431 ± 19 vs 425 ± 22 ms). Moderate to vigorous activity levels in percentage of total recorded time did not change in either group (intervention group: 15 ± 6 vs 15 ± 5; control group 14 ± 6 vs 14 ± 6).
Conclusion: Exercise training did improve VO_{2peak}/kg of children and young adults with a corrected tetralogy of Fallot without changing ECG markers and physical activity.

1021: NORMAL VALUES OF ARTERIAL VALVE DIAMETERS IN NEONATES

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Background: Normal values of various cardiovascular diameters are needed to facilitate proper diagnosis and decision making on treatment of children with congenital heart defects. The larger the group analysed, the more accurate and useful the conclusions and norms are. The aim of this study was to assess the normal values of aortic and pulmonary valves in healthy, term neonates and to correlate this with age (1–30 days), weight and body surface area.
Methods: We reviewed 771 transthoracic echocardiographic examinations performed between 2002 and 2008, and data were collected retrospectively. All examinations were performed on healthy, term neonates without any structural heart defect. Measurements were taken from the short (pulmonary valve) and long (aortic valve) parasternal axis in diastole. Statistical analysis was performed using Statistica 10 software.
Results: The average age at examination was 10.6 days (SD 7.8) and weight was 2.6 kg (SD 0.7). Aortic valve diameter [average 7 68 mm (SD 0.95)] was statistically significantly correlated with age ($p < 0.001$, $r = 0.29$), BSA ($p < 0.001$, $r = 0.47$) and weight ($p < 0.001$, $r = 0.54$). In the case of the pulmonary valve, the average diameter was 9.05 mm (SD 1.24). It was also significantly correllated with age ($p < 0.001$, $r = 0.4$), BSA ($p < 0.001$, $r = 0.58$) and weight ($p < 0.001$, $r = 0.62$). In both of the valve diameters, the strongest correlation and clinical significance was observed with weight. Because all observed correlations were medium or weak, we calculated the normal values of aortic (6.3–9.7 mm) and pulmonary (7–11.4 mm) valves for the whole study group, which gave values between the fifth and 95th percentile.
Conclusion: The diameters of the aortic and pulmonary valves in the neonatal period did not correlate strongly with age, weight or BSA, despite the fact that it was statistically significant in all cases. This allows us to use the common normal value range for the whole neonatal period. In case of borderline values, weight should be taken into consideration, because it has the strongest correlation with arterial valve diameters.

1028: PULMONARY HAEMOSIDEROSIS SECONDARY TO SEVERE MITRAL STENOSIS IN PATIENTS UNDERGOING VALVULOPLASTY AT TYGERBERG HOSPITAL

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Aim: ulmonary haemosiderosis (PH) secondary to mitral stenosis (MS) is considered rare. This study aimed to determine if PH is more common than currently thought, as well as determining factors contributing to its development, impact on lung function and reversibility.
Methods: Chest X-rays (CXRs) of patients who underwent mitral valvuloplasy (17/01/1997–10/02/2012) were reviewed for PH. Data collected included date of birth, date of valvuloplasty and pre-procedural echocardiography reports. Patients with PH on CXR were invited to participate in a prospective trial. Patients without PH were selected as controls. In the prospective trial a clinical evaluation, ECG, echocardiography, CXR and lung functions were performed.

Results: Sicty (24.8%) of 242 patients who underwent valvuloplasty at Tygerberg Hospital had CXRs available. The incidence of PH in this cohort of patients with severe MS was 19%. Patients with PH had higher pulmonary pressures (71.1 vs 55.7 mmHg; $p = 0.04$) and had significantly smaller mitral valve areas (0.85 vs 0.99 cm^2; $p = 0.09$). Patients with PH also had higher peak transmitral pressure gradients (30.1 vs 24.37 mmHg; $p = 0.10$). Although the mean transmitral pressure gradients were higher in the patients with PH, this difference was not significant (17.43 vs 15.02 mmHg; $p = 0.23$). Patients with PH were younger at the time of valvuloplasty (29 vs 38 years; $p = 0.02$). No difference was seen when comparing the lung functions in terms of ventilatory and diffusion impairment. The data regarding radiological reversibility was inconclusive.
Conclusion: PH occurs more commonly in severe MS than currently thought. Compared to patients without PH, they had more severe MS and underwent valvuloplasty at a younger age. The data suggest that PH is an irreversible process with little impact on lung function but the numbers studied prospectively were too small to provide conclusive evidence regarding the impact on lung function and reversibility.

1030: SCORE FOR QUANTIFICATION OF THE MORPHOLOGICAL AND FUNCTIONAL ECHOCARDIOGRAPHIC FEATURES OF MITRAL VALVE IN PATIENTS WITH RHEUMATIC HEART DISEASE

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Background: Rheumatic heart disease (RHD) still challenges all those involved in its diagnosis and in this context, a more accurate assessment by echocardiography has added valuable information about the functional and anatomic subtract of valvar and subvalvar mitral apparatus (MV).
Objective: To present an echocardiographic score to support the evaluation of MV lesions.
Methods: Out of 298 patients seen during the period of 2009–2010, 93 patients [duration of the disease: $\bar{x}$ (mean) = 8.0 ± 5.8 years; age at first episode: $\bar{x}$ = 9.2 ± 2.9 years] with a definitive diagnosis of RHD, followed since the first episode and without surgical intervention, were selected and underwent echo screening. The echo tapes were reviewed by two other echocardiographists and the kappa statistic was used for the assessment of inter-observer variability [kappa: 0.875 (95% CI: 0.775–0.974)].
Results: The patients' data were scored on a rating scale (1–15), to quantify the severity of the mitral valve involvement. The quantification included five groups of morphological and functional variables and three degrees of severity. In the comparative analysis, the degrees established for the classification of the scores of morphological/functional features were associated with the corresponding degrees of severity of the haemodynamic findings for mitral regurgitation ($p = 0.00$) and stenosis ($p = 0.02$).
Conclusion: Considering the characteristic aspects of the morphological features of the mitral valve in patients with RHD, the stratification by degree of commitment could represent additional support for the echocardiographic evaluation.

1035: IMPROVING THE DIAGNOSTIC YIELD OF THE ECG IN TACHYARRHYTHMIAS BY ROUTINELY PERFORMING LEWIS LEAD RECORDINGS

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Background: Determining the origin of tachyarrhythmias is a skill required by all clinicians. Identifying the P wave is integral to making a diagnosis in tachyarrhythmias. A diagnostic tool that is proposed as an aid in this regard is doing Lewis leads, but there is little evidence for this in the literature. As with any diagnostic tool one would expect that its value is influenced by the users' knowledge and experience.
Methods: We prospectively studied patients presenting with a tachycardia to the division of cardiology of Tygerberg Hospital over a six month period. Both standard ECGs and Lewis leads were performed in all cases. These ECGs were then analysed by three sets of participants, namely cardiologists, registrars and students. An initial diagnosis was made on the standard ECG only. A revised diagnosis was then given based on the added information provided by the Lewis lead in addition to the standard ECG. These answers were compared to the definitive diagnoses and classified as correct, incorrect and unsure (where a differential diagnosis was given).
Results: More correct diagnoses were obtained in all groups with the addition of Lewis leads. Cardiologists displayed a trend towards improvement with Lewis leads (87–96%; $p = 0.07$). The registrar group had the most marked improvement (64–82%; $p = 0.0026$). Students did not improve significantly (52–54%; $p = 0.8$). Lewis leads were of assistance to students in patients with regular rhythms but not in irregular rhythms.
Conclusion: Lewis leads are a valuable diagnostic tool to enhance ECG interpretation. It aids clinicians in making accurate diagnoses in tachyarrhythmias. The amount of benefit is dependent on the user's knowledge and expertise. Cardiologists and registrars may benefit from routinely registering Lewis leads in tachyarrhythmias. Inexperienced individuals should be wary of using Lewis leads in irregular tachyarrhythmias. Students would benefit from specific training in assessing Lewis leads.

1040: IMPACT OF AGE AND GENDER ON CARDIAC PATHOLOGY IN CHILDREN AND ADOLESCENTS WITH MARFAN SYNDROME

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Background: Cardiac pathologies are the major aspect in the treatment strategies of Marfan syndrome (MFS). In this progressive disease, less is known about manifestation and progression of cardiovascular symptoms in children. To define a certain decision regarding therapeutic options, knowledge about onset of cardiovascular findings in children is essential.
Methods: From 1998 to 2011 patients with clinical features of MFS were subject to a standardised diagnostic programme. Cardiovascular findings were analysed concerning age at first clinical manifestation, prevalence and gender differences, morbidity, mortality and treatment during follow up.
Results: MFS was diagnosed in 82 patients (46 male, mean age at diagnosis 9.0 ± 5.7 years). At first presentation, aortic root dilation and mitral valve prolapse was found in 56.1 and 31.7%, whereas pulmonary artery dilation and tricuspid valve prolapse were detected in 22.0 and 17.1%. Aortic valve (2.4%) and mitral valve regurgitation (22.0%) were significantly correlated with aortic root dilation ($p < 0.01$) and mitral valve prolapse ($p < 0.05$) but without relevant progression during childhood and adolescence. Because of progressive aortic root dilation, medication was initiated in 41.5% of patients (mean age 8.0 ± 4.5 years). Aortic dissection did not appear. Aortic root surgery in case of severe progression of aortic root dilation was needed in 3.7% of patients (mean age 9.7 ± 2.3 years). Gender-specific differences concerning cardiovascular findings, progression of disease or treatment did not appear.
Conclusion: Comparable with adults, aortic root dilation was the most frequent cardiovascular finding and was associated with relevant morbidity and the aim of early prophylaxis. While aortic and mitral valve regurgitation is of minor clinical relevance, manifestation at an early age and slow progression of cardiovascular findings in childhood underline the necessity of repeated echocardiographic examinations in cases of suspected MFS for early diagnosis and an early start to prophylactic treatment.

1041: CARDIOMYOPATHY IN PATIENTS WITH THE AMISH AND MENNONITE VARIANT OF PROPIONIC ACIDAEMIA

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Background: Propionyl-CoA carboxylase deficiency (propionic acidaemia, PA) caused by a *PCCB* c.1606A > G variant is prevalent among Amish and Mennonite (Plain) people. Systemic metabolic crises are infrequent, but they remain at risk for life-threatening cardiomyopathy, which may result from loss of anaplerotic propionyl-CoA flux into the myocardial tricarboxylic acid (TCA) pool.
Methods: Thirty-three patients (10.2 ± 5.6 years) homozygous for *PCCB* c.1606A > G were followed longitudinally at a single centre for 250 aggregate patient-years. We studied cardiac morphology, function and outcome using ultrasound and cardiac magnetic resonance (CMR) equipped with quantitative T1 mapping software (Siemens Healthcare). Two brothers with symptomatic cardiomyopathy were treated with a dietary anaplerotic mixture designed to supply 2-, 4-, and 6-carbon substrates to the TCA cycle.
Results: Cardiomyopathy (left ventricular ejection fraction, LVEF < 3 SD below age-matched controls) developed in 12 (36%) patients. (EF 58–7%), and was lethal in three children. In PA patients, LV EF (64 ± 11%) was lower than controls (73 ± 5%; $p < 0.0001$). Two brothers with symptomatic cardiomyopathy when treated with the TCA anaplerotic mixture had resolution of symptoms over four to six months. LV EF increased from 18 to 56% and 44 to 63%. CMR during the acute phase showed biventricular dysfunction and increased end-diastolic volumes but no evidence of tissue fibrosis or oedema (myocardial interstitial volume 26%). Following anaplerotic treatment, biventricular function and cardiac volumes normalised with no tissue scarring (myocardial interstitial volume 29%).
Conclusion: Cardiomyopathy is common in patients with PA who are otherwise metabolically stable and is the major cause of untimely death resulting from the *PCCB* c.1606A > G variant. PA should be considered in patients of any age who present with 'idiopathic' cardiomyopathy, even when standard metabolic screening tests are normal. The cardiomyopathy of PA may be fully reversible by supporting the TCA anaplerotic function normally served by propionyl-CoA.

1042: CHANGES IN CLINICAL SIGNS OF HEART FAILURE AFTER CARVEDILOL THERAPY IN CHILDREN WITH LEFT-TO-RIGHT SHUNT CONGENITAL HEART DISEASE

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Background: Heart failure (HF) in children with left-to-right shunt congenital heart disease (L-R shunt CHD) causes heart remodelling, making the symptoms worse. Tachycardia, tachypnoea and hepatomegaly are the cardinal signs of HF. Carvedilol, a non-selective beta-blocker, has shown good results for HF treatment in adults with tolerable side effects, but there is little evidence in children. The objective of this study was to determine the changes in heart rate, respiratory rate, and hepatomegaly after three months' carvedilol additional therapy on top of standard HF therapy in children with L-R shunt CHD.
Methods: A randomised, controlled trial was designed for 30 subjects with paediatric heart failure score (PHFS) > 2 due to L-R shunts CHD. Subjects were divided into a carvedilol group ($n = 15$) and placebo group ($n = 15$). Two group received standard therapy for HF, ACEI and a diuretic. The study lasted for three months. The changes in heart rate, respiratory rate and hepatomegaly were observed. Statistical analysis was done using paired *t*-test and independent samples *t*-test with confidence intervals of 95%, McNemar test and Chi-square test, CI 95%.
Results: Nineteen (63.3%) patients were boys, 11 (36.7%) were girls. Mean age was 57.6 (SD 43.57) months. Twenty-one had VSD (70%), nine had PDA (30%). Heart rate and respiratory rate were significantly decreased in the carvedilol group compared with the placebo group ($p < 0.0001$). No hepatomegaly was found. Yjere was no adverse effect during the study.
Conclusion: Additional therapy with carvedilol on top of standard therapy for HF due to L-R shunt CHD significantly improved the clinical signs of HF. Carvedilol was effective and safe as additional therapy in addition to standard HF therapy.

1047: ERYTHROCYTE DEFORMABILITY MECHANISM IN TETRALOGY OF FALLOT PATIENTS WITH IRON DEFICIENCY

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Background: Iron deficiency in patients with tetralogy of Fallot (TF) is an adverse condition related to erythrocyte deformability and the incidence of fatal complications of the disease. However, the mechanism of erythrocyte deformability in TF patients with iron deficiency so far remains unexplained.
Objective: To discover the mechanism of erythrocyte deformability in TF patients with iron deficiency. This was an observational study involving four groups: TF patients with iron deficiency (I), TF patients without iron deficiency (II), non-TF patients with iron deficiency (III), and non-TF patients without iron deficiency (IV). The variables of erythrocyte deformability used in this study were SaO_2, transferrin saturation, H_2O_2 molecule levels in erythrocytes, and spectrin-denatured ethrocytes, as well as the number of erythrocytes passing through a device membrane.
Results: The reduction in SaO_2 level resulted in a decrease in transferrin saturation, followed by an increase in H_2O_2 molecule levels, leading to a rise in the number of spectrin-denatured erythrocytes, reduced erythrocyte deformability and a reduction in the number of erythrocytes passing through a device membrane. Iron therapy may increase transferrin saturation, thereby reducing the erythrocyte H_2O_2 molecular level, and lead to further reduction in the number of spectrin-denaturated erythrocytes, improved deformability of erythrocytes and increase in the number of erythrocytes passing through a device membrane.
Conclusion: This study produced a model describing the contribution of each discriminator. The discriminator model of the occurrence of erythrocyte deformability in TF patients with and without iron deficiency can be applied as a novel basic concept to underscore and optimise the management of TF patients who have no opportunity to have cardiac corrective surgery.

1050: USEFULNESS OF N-TERMINAL PRO-B-TYPE NATRIURETIC PEPTIDE AS BIOMARKERS FOR CONGESTIVE HEART FAILURE TREATMENT WITH CONGENITAL HEART DISEASES

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Background: The aim of this study was to determine plasma N-terminal pro-B-type natriuretic peptide (NT pro-BNP) levels in infants with congenital heart disease (CHD) and congestive heart failure (CHF) before and after decongestive treatment, and correlate this with the heart failure score.
Methods and Results: The study comprised 46 infant with CHD aged from 21 day to 26 months who were categorised into CHF mild, moderate and severe, according to the modified Ross scoring system. The patients were evaluated before and 10 and 30 days after decongestive treatment. Before the treatment, 15 patient had mild CHF, 27 had moderate CHF and four severe CHF. Mean NT-proBNP

level was 4 983.4 ± 6 326 pg/ml. On the 10th day after treatment, 27 had (59%) mild, 18 (39%) moderate, and one (2%) severe CHF. Mean NT-proBNP level was 2 177.1 ± 2 629.8 pg/ml. On the 30th day after the treatment, 41 patients (89%) had mild CHF, and five (11%) had moderate CHF. None had severe CHF. Mean NT-proBNP level was 1 701.8 ± 2 126.4 pg/ml. NT-proBNP levels decreased with decongestive therapy ($p < 0.05$). NT-proBNP levels were lower on the 10th day of therapy than before the therapy, and lower on the 30th day of therapy than on the 10th day of therapy. There was no significant correlation between NT-proBNP level and Ross scoring on the 10th day of therapy. There was a significant correlation between NT-proBNP level and Ross scoring on the 30th day of therapy.
Conclusion: Plasma NT-proBNP levels were elevated in infants with CHD with left-to-right shunt before treatment, and decreased with decongestive therapy. Nevertheless clinical evaluation is more important to determine the severity of CHF.

1051: PROFILE OF THE PRESENTATION AND EVOLUTION OF RHEUMATIC FEVER IN CHILDREN AND ADOLESCENTS

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Background: Rheumatic fever (RF) and rheumatic heart disease (RHD) represent a health burden worldwide. As a condition carried throughout life, it has repercussions at all ages and accounts for an important number of repeated hospitalisations and deaths.
Objectives: To analyse the profile of the clinical and epidemiological presentation and its relationship with the severity and evolution of cardiac involvement.
Methods: This cohort study was carried out on 823 consecutive patients aged between 2.7 and 18.9 years and with a mean follow up of 7.6 ± 2.8 years (1984–2004). The following variables were analysed: age at first attack, gender, clinical manifestations and recurrences, family antecedents, previous pharyngotonsillitis and pattern of severity.
Results: The first episode was most frequent at the age of 6–15 years ($\bar{x}$: 9.2 ± 3.1 years), without gender predisposition, except for chorea (F/M: 1.7/1.0; $p = 0.0013$). Previous pharyngotonsillitis was reported at 54.9%. The prevalence of RF in patients' families (14.2%) was higher than among relatives of children without the disease ($p = 0.0000$). At the first attack, 96.4% of patients presented with MR, isolated (44.2%) or associated with AR. Out of those with RHD, 97.6% showed mitral and/or aortic involvement (isolated AR: 2.4%) with regurgitation in 78.8% and mixed lesions in 21.2%, without patients with AS. Severe carditis was more prevalent in children with two or more recurrences, and valvar sequelae more significant in those with severe carditis ($p = 0.0001$); 34.8% of patients showed complete resolution of cardiac findings, mostly without recurrences, and presented with mild regurgitant lesions but none with severe valvar involvement. Significant decrease was seen in the occurrence of severe carditis, surgery and death after the control of recurrences ($p = 0.0000$).
Conclusions: The valvar sequelae were influenced by the severity of the carditis and by the number of recurrences. Considering the difficulties in primary prevention, the authors reinforce the need for effective strategies of secondary prophylaxis to reduce morbidity and mortality rates.

1055: INNSBRUCK EXPERIENCE WITH KAWASAKI DISEASE

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Background: We describe long-term follow up of three out of 89 patients with Kawasaki disease seen in our institution in a 33-year period, with different clinical courses of cardiac or coronary artery involvement.
Methods: A retrospective description including clinical investigation, echocardiography and angiography was performed.
Results: Patient 1 had a myocardial infarction in the acute phase of Kawasaki disease 33 years ago. At that time, aneurysms in both carotids, subclavian, renal and mesenterial arteries were detected by sonography and angiography. Last heart catheterisation six years ago revealed two aneurysms of the left ventricle (one at the apex and one at the base, each 3 cm in diameter) and a hypokinetic left ventricle. He is in stable clinical condition and denied any therapy.

Patient 2 developed a 17 × 11-mm aneurysm of the left coronary artery. Twelve years later, a 90% stenosis of the left coronary artery and a 70% stenosis of the left anterior descending artery, as well as the ramus circumflex were detected by angiography. He underwent aortocoronary bypass with a bilateral arteria thoracica interna bypass. He is in excellent clinical condition on treatment with a platelet aggregation inhibitor.

Patient 3 was diagnosed with a 9 × 6-mm aneurysm of the right coronary artery during the acute phase of Kawasaki disease. Spontaneous regression was observed within 12 years. The right coronary artery shows no signs of stenosis or thrombosis in angiography, ventricle function is normal and the patient is in an excellent clinical condition, receiving clopidogrel.
Conclusions: In a 33-year period, two out of 89 patients evolved giant coronary artery aneurysms; one patient therefore underwent aortocoronary bypass surgery. One patient developed two aneurysms of the left ventricle, and multiple aneurysms in the great arteries affecting multiple central arteries.

1056: MYOCARDIAL RESPONSE TO EXERCISE AFTER PAEDIATRIC HEART TRANSPLANT: A BICYCLE EXERCISE STUDY

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Background: Data on myocardial systolic and diastolic response to exercise of the transplant heart are limited. We used semi-supine cycle ergometry (SSCE) stress echocardiography to evaluate left ventricular (LV) systolic and diastolic reserve in paediatric heart-transplant (P-HTx) recipients and compared the exercise response to healthy controls.
Methods: Forty-three P-HTx and 24 age- and gender-matched controls were included. A stepwise SSCE protocol was used. Peak systolic and early diastolic tissue Doppler velocities were measured in the lateral and septal basal segments and values were expressed versus heart rate. LV myocardial acceleration during isovolumic contraction (IVA) was measured in all the subjects at incremental heart rates to evaluate the force–frequency relationship (FFR). LV longitudinal strain was also quantified at rest and during exercise.
Results: At rest early diastolic tissue Doppler velocities (E') were reduced in the P-HTx group in the lateral LV wall (11.1 vs 13.7 cm/s, $p = 0.001$) and the basal septum (8.1 vs 11.1 cm/s, $p < 0.001$). Lateral and septal S' values did not differ significantly between the groups. At peak, all S' (8.1 vs 11.1 cm/s, $p < 0.001$) and E' (8.1 vs 11.1 cm/s, $p < 0.001$) velocities were lower in P-HTx. The change in E' and S' values from baseline to peak was lower P-THx compared to controls. Also the E/E' ratio was higher in P-HTx in the lateral wall and in the septum. The contractile response as studied by the FFR was blunted in P-HTx. LV longitudinal peak systolic strain values increase during exercise in both groups, but the P-HTx had lower strain value than the controls.
Conclusions: P-HTx patients showed a reduced systolic contractile response as well as a reduced diastolic response to exercise compared to the controls. This was not related to the heart rate response. The clinical and prognostic implications of these findings require further study.

1058: ASSESSMENT OF PULMONARY VASCULAR VOLUME AND LUNG PERFUSION IN PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME IN FONTAN CIRCULATION USING CARDIOVASCULAR MAGNETIC RESONANCE IMAGING

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Introduction: Angiographic data indicate that children with hypoplastic left heart syndrome (HLHS) in Fontan circulation have a smaller anatomical size of the central pulmonary arteries. We utilised novel MRI techniques to assess pulmonary vascular volume and lung perfusion to investigate whether HLHS patients had lower values than lung healthy controls.
Methods: Thirty-one children with HLHS (4.9 ± 2.3 years) and eight lung healthy controls (9.8 ± 6.4 years) were studied. A modified contrast-enhanced dynamic MR angiography sequence was used to assess pulmonary vascular and total volume of the right and left side of the lung. With the use of dedicated software we measured the lung volume, relative pulmonary vascular volume, the rate of parenchymal contrast enchancement ('up-slope'), as well as the parenchymal mean transit time (MTT) in order to assess pulmonary vascular status and perfusion. Standardised cardiopulmonary exercise testing on a treadmill was performed in all patients and compared to a cohort of paediatric volunteers.
Results: The indexed pulmonary total lung volume and the relative vascular volume in HLHS were significantly reduced compared to the control group (634 ± 105 vs 945 ± 274 ml/m², $p < 0.05$; 6.5 ± 3.7 vs 9.5 ± 2.4%/m², $p < 0.05$). Lung perfusion in children with HLHS was also impaired compared to healthy children (MTT: 10.8 ± 2 vs 6.7 ± 3 s, $p < 0.01$; up-slope: 0.32 ± 0.24 vs 1.12 ± 0.305 $s^{-1}m^{-2}$; $p < 0.01$ $s^{-1}m^{-2}$; $p < 0.01$). Relative vascular volume showed no difference between the right and left lung and correlated significantly with oxygen uptake measured by standard spirometry (Spearman's $r = 0.78$; $p < 0.01$).
Conclusions: Advanced MRI methods enabled the assessment of pulmonary perfusion in HLHS patients in Fontan circulation. HLHS patients had a reduced total lung volume, pulmonary vascular volume, and an impaired lung perfusion, compared to controls. The reduced vascular volume in HLHS patients appears to be an important limiting factor for pulmonary function. Long-term follow-up investigations are important to detect a potential further decline of values during growth and which implication may occur for the function of the Fontan circulation.

1062: CONGENITAL LONG-QT SYNDROME: A SINGLE-CENTRE EXPERIENCE

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Background: Congenital long-QT syndrome (LQTS) is characterised by prolonged ventricular repolarisation (QT interval prolongation) and a propensity for syncope and sudden death secondary to torsades de pointes.
Methods: In this report we identified 16 congenital LQTS patients who were followed at Department of Paediatric Cardiology, Cukurova University between 2000 and 2012. The mean age of the patients was 104 months, the mean follow-up period was 24 months, and mean corrected QT interval was 0.52 ms.
Results: At pre-diagnosis, nine patients (56%) had a history of syncope, convulsions or cardiac arrest, and three patients were treated with anti-epileptic drugs as they were misdiagnosed as epileptics. During the follow-up period, two patients received implantable cardioverter defibrillators (ICD). One was a boy with symptomatic bradicardia on Holter ECG record and history of cardiac arrest of a first-degree family member, and the other was a girl with recurrent sustained ventricular tachycardia with syncope even though she was on medical therapy. All patients were treated with beta-adrenergic blockade as soon as LQTS was diagnosed, and potentiated through maximal heart rate on a treadmill exercise test. During the follow-up period, no patient had cardiac arrest. We diagnosed LQTS in two patients' asymptomatic first-degree relatives, just with ECG screening.
Conclusion: Being a survivor of sudden cardiac arrest, having a history of recurrent syncope, or history of sudden cardiac arrest in a family member, and with a diagnosis of LQTS in relatives, genetic determination must take into consideration during clinical follow up of a congenitally LQTS patient. Beta-adrenergic blockade, avoidance of triggering factors, ICD or permanent pacemaker implantation for high-risk patients, and sympathetic ganglion blockade are treatment choices for LQTS patients. Physicians must screen family members with an ECG, and must overemphasise the history of recurrent syncope attacks, convulsions or sudden cardiac arrest in patients with suspicion of a LQTS diagnosis.

1063: VALUE OF RADIOFREQUENCY ABLATION VERSUS CRYO-ABLATION FOR ATRIO-VENTRICULAR NODAL RE-ENTRY TACHYCARDIA IN CHILDREN

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Background: Radiofrequency ablation (RFA) and cryo-ablation are two strategies for treating atrio-ventricular nodal re-entry tachycardia (AVNRT) in children, but it is unclear which strategy may offer better value.
Methods: We performed a retrospective cohort study of all AVNRT ablation cases for children (age ≤ 18 years) from 1 July 2009 to 30 June 2011 at a single institution. Cost data included fixed cost, miscellaneous hospital costs, and labour costs. Outcomes data regarding the acute long-term success (six months) of the ablations were collected through chart review. *T*-test and regression analysis were conducted to investigate any association between ablation procedure type (RFA, cryo-ablation, or combination of both), and the cost and long-term success rate of the ablation procedures, adjusting for patient (age, BSA) and provider (experience) characteristics.
Results: Of 96 unique cases, 48 were cryo-ablation only, 42 RFA only, and six were a combination (four starting with RFA, two starting with cryo-ablation). Acute success was 100% for the cryo-ablation only and RFA only cases and 83% for the combination cases. There were no notable adverse events. The average total cost was US$9 636 for cryoablation cases, US$9 708 for RFA cases, and US$10 967 for combination cases. The difference in the cost between cryo-ablation only cases and RFA only cases was not statistically significant. The unadjusted long-term success rate was 79.1% for cryo-ablation only cases, 92.8% for RFA only cases, and 66.7% for combination cases ($p < 0.05$ for cryo-ablation only vs RFA only). After adjusting for patient and provider characteristics, the difference between cryo-ablation only and RFA only was not considered significant.
Conclusions: There was no difference in costs or short-term success rate for cryo-ablation vs RFA for treatment of AVNRT in children. Differences in long-term success rates may be attributed to multiple factors other than the choice of procedure and may lead to differences in long-term costs if children are referred for repeat ablation.

1064: IMPACT OF BT SHUNT SIZE ON TRICUSPID REGURGITATION IN HYPOPLASTIC LEFT HEART SYNDROME

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Background: The prognosis for hypoplastic left heart syndrome (HLHS) has greatly improved over the past years, but there is still a significant risk of inter-stage mortality, particularly between stages 1 and 2 of surgical palliation. Tricuspid regurgitation (TR) is known to be a risk factor for postoperative and inter-stage mortality. We hypothesised that a smaller BT shunt would lead to a reduction in ventricular volume loading and also a reduction in TR. This study aimed to evaluate the effect of BT shunt size on the severity of TR.
Methods: A retrospective review of all patients with HLHS who had a Norwood procedure in our institution between January 2006 and December 2011 was performed. Patient demographic, echocardiograms, MRI, operative and postoperative data were reviewed. The impact of BT shunt size on severity of TR at the time of BCPS was analysed.
Results: In this time period, 64 neonates with HLHS underwent a Norwood procedure with a modified BT shunt at our institution. Thirty-two (32/64) had a 3.5-mm BT shunt (group 1) and 32/64 had a 3.0-mm BT shunt (group 2). Survival to BCPS was 53/64 (83%) overall, with 25/32 (78%) in group 1 and 28/32 (87%) in group 2. The prevalence of significant TR (≥ moderate) on pre-operative assessment in preparation for BCPS was 9/25(36%) in group 1 and 2/28(7%) in group 2 ($p = 0.01$). Tricuspid valve repair was performed on 7/25(28%) in group 1 and 4/28(14%) in group 2 ($p = 0.21$). The rate of non-fatal serious adverse events was similar in the two groups.
Conclusion: A smaller-sized modified BT shunt at the initial Norwood procedure reduces the prevalence of significant tricuspid regurgitation. Further investigation and long-term follow up is required to determine other potential complications of this surgical strategy.

1065: EFFECT OF BERAPROST ON PULMONARY ARTERY PRESSURE IN PULMONARY HYPERTENSION DUE TO LEFT-TO-RIGHT SHUNT CONGENITAL HEART DISEASE

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Background: The late diagnosis and surgical management of left-to-right shunt congenital heart disease (L-R shunt CHD) causes pulmonary hypertension. To date there is no effective drug to decrease pulmonary artery systolic pressure. Beraprost, an oral prostacyclin analogue that works as a pulmonary artery vasodilator, was expected to decrease PASPS in children with L-R shunt CHD.
Methods: A pretest–postest study was conducted on 17 subjects aged two months to 16 years old, with VSD, ASD, PDA or a combination, who developed pulmonary hypertension. Patients with other structural cardiac defects or with pulmonary infection were excluded. Echocardiography examinations were performed before and after administration of beraprost 1 µg/kgBB/day for three months.V_{max} TR , V_{max} L-R, and PASP were measured and M-mode measurements of the pulmonary valve, including a wave, EF slope and mid-systolic notch were examined. Adverse reactions were noted, and monitoring was performed every two weeks.
Results: A V_{max} TR decrease [0.7 ± 0.79 m/s ($p = 0.004$)], Vmax L-R increase [0.6 ± 0.64 m/s (p = 0,018)] and PASP decrease [18.3 ± 21.90 mmHg ($p = 0.003$)] were observed. There were no significant changes in wave, EF slope and mid-systolic notch.
Conclusions: There was a decrease in pulmonary artery systolic pressure after administration of beraprost in children with L-R shunt CHD who develop pulmonary hypertension.

1067: DELAYED UPTAKE AND WASHOUT OF CONTRAST IN INFARCTED MYOCARDIUM SHOWN WITH 4D CT IN PIGS

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Background: Assessment of ischaemic and potentially viable myocardium plays an important role in the planning of revascularisation of patients with critical coronary atherosclerosis. Cardiac computed tomography (CT) is being used increasingly in the functional and morphological analysis of cardiovascular diseases and specifically in the screening and evaluation of atherosclerotic lesions in coronary artery disease. The aim of the present study was to investigate the pattern of contrast distribution over time in pigs with experimentally induced anterior myocardial infarctions, to evaluate the feasibility of using simple CT enhancement as a viability marker.
Methods: This study was conducted as an experimental animal follow-up study. Twelve female pigs weighing 50 kg were subjected to an ischaemic myocardial lesion induced by 60 minutes of balloon occlusion of the left anterior descending artery immediately distal to the first diagonal branch, followed by removal of the balloon and reperfusion. After six weeks, CT was performed using intravascular contrast agent. Measurements of radiodensity in Hounsfield units in the infarct zone and the non-ischaemic lateral wall were performed at 20 seconds, one, three, five, eight and 12 minutes after contrast injection.
Results: We found highly significant differences in radiodensity between the two zones at all time points except $t = 1$ minute. The healthy myocardium showed a more rapid uptake and washout of contrast compared to the infarct zone. In particular, the ratio between early and late uptake was a strong marker of viable myocardium.
Conclusion: The delayed uptake and washout of contrast agent in the infarct zone compared to the zone of healthy myocardium corresponded well with existing knowledge from MRI and other methods and most likely reflects the impairment of the vascular bed in the infarcted myocardium. The present study shows promise for future clinical application of CT in a combined assessment of coronary anatomy and myocardial viability.

1069: THREE-DIMMENTIONAL ROTATIONAL ANGIOGRAPHY IN PATIENTS WITH SINGLE VENTRICLE AFTER TOTAL CAVO-PULMONARY CONNECTION

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Background: Three-dimensional rotational angiography (3DRA) is an emerging technology that could be a beneficial adjunct to fixed-plane angiography and could enhance diagnostic capabilities in patients with single ventricle after various stages of palliation. We report our total experience using 3DRA in patients after total cavo-pulmonary connection (TCPC).
Methods: A retrospective analysis of all patients after TCPC who underwent 3DRA was performed. Philips Allura system was used to acquire non-gated, breath-held images. During a 240-degree, 4.1-second isocentric rotation, 122 angiographic images were acquired and automatically reconstructed in real time.
Results: Between 09/2010 and 06/2012, we performed 41 3DRAs in 21 patients after TCPC. All but one patient underwent total of 32 interventions. Twenty-four 3DRAs were performed prior to the intervention, with 17 remaining to assess the results of percutanous treatment. Median age and weight was 9.13 years (4.1–24.1) and 24.8 kg (13–56), respectively. Median contrast dose for 3DRA and for total study was 1.45 ml/kg (0.9–2.4) and 4 ml/kg (1.5–9.8), respectively. Median area dose for the whole study, time of fluoroscopy and total time of study was 330.2 cGy/cm^2 (144.8–1325.2), 15.2 min (9.3–54.4) and 70 min (55–165), respectively. Rotational angiogra-

phy provided a median of 96 projections (78–105) and the following 3D reconstruction was successful in all studies. Both allowed for precise visualisation of the external tunnel, superior vena cava and pulmonary arteries. Overall quality of 3DRA images was graded as good in 32 (78%) studies and satisfactory in nine (22%). None of the 3DRAs was graded as poor, however eight additional angiographies (three 3DRA, five fixed-plane) were performed to better visualise the left pulmonary artery.
Conclusions: In patients after TCPC, 3DRA provided a large number of projections with relatively small amounts of contrast and allowed for perfect visualisation of the Fontan circuit.

1083: ASSESSMENT OF TRICUSPID ANNULAR-PLANE SYSTOLIC EXCURSION IN PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME IN FONTAN CIRCULATION

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Background: The tricuspid annular plane systolic excursion (TAPSE) is an established echocardiographic parameter to assess right ventricular (RV) systolic long-axis function in biventricular hearts. The aim of this study was to investigate long- and short-axis function in patients with hypoplastic left heart syndrome **(**HLHS) utilising MRI.
Methods: 40 children (5.8 ± 2.5 years) with HLHS in Fontan circulation and 10 healthy controls (10.6 ± 5.2 years) underwent a comprehensive cardiac MRI study to evaluate global and regional RV function. We used CMR cine-imaging (TR/TE/α = 1.1/1.6/60, FOV: 240 × 260) for analysis of global (EF, and cardiac index: CI) and regional ventricular function. TAPSE was analysed with an in-house developed software that allowed 3D reconstruction of ventricular geometry.
Results: HLHS patients had a lower TAPSE and cardiac index compared with healthy subjects. In contrast with healthy subjects, TAPSE and RV-EF did not show significant correlation in HLHS patients with a rudimentary LV. In HLHS patients without a visible LV cavum we found a significant correlation between RV-EF and TAPSE (r = 0.62; p = 0,01). Furthermore, HLHS patients with a rudimentary LV had a significantly reduced septal wall motion and a cardiac index compared with HLHS patients without a rudimentary LV (4.6 ± 1.5 vs 6.9 ± 2.1 mm; p = 0.001; 3.4 ± 0.8 vs 2.8 ± 0.9 ml/m^2/min; p = 0,01).
Conclusion: Patients with HLHS had impaired long-axis RV function compared to healthy controls. A rudimentary LV impaired in particular, contraction in the septal segment, resulting in a reduced long-axis and global RV function. This may be of prognostic significance for the long-term outcome in HLHS patients.

1090: CAN MINOR CONGENITAL MITRAL VALVE VARIANTS BE DIFFERENTIATED FROM RHEUMATIC MITRAL VALVE DISEASE?

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Background: The echocardiographic features of congenital mitral valve prolapse and severe mitral rheumatic heart disease (RHD) are well established. Mild forms of these entities are harder to differentiate. A frequent clinical scenario in regions with high prevalence RHD is whether mild mitral regurgitation is of congenital or rheumatic aetiology.
Methods: Fifteen echocardiograms with mild mitral regurgitation (MR) were reported by two experienced observers and two trainee cardiologists without knowledge of the patient demographics. The aetiology of the MR was judged congenital if there was classical mitral valve bi-leaflet prolapse, accessory mitral valve leaflet or scallop, diastolic excessive movement, or mitral valve cleft. Five echocardiograms had minor congenital mitral valve features previously defined by a panel of three cardiologists.
Results: The range of congenital mitral anomalies diagnosed was one to four of the five positive studies (kappa 0.25–0.46).
Conclusions: Minor congenital mitral valve variations may cause mitral valve regurgitation but they are not easily differentiated from MR due to RHD. Minor congenital mitral valve anomalies can cause a false-positive diagnosis of RHD.

1102: LONG-QT MOLECULAR AUTOPSY: RESULTS OF SIX YEARS OF A POPULATION-BASED CLINICAL SERVICE FOR AUTOPSY-NEGATIVE SUDDEN DEATH IN 1–24 YEAR OLDS

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Background: Families and forensic pathologists seek a diagnosis with urgency following the tragedy of a sudden, unexpected and unexplained death in children and youth. We report the results from six years of a national molecular autopsy service in New Zealand (population 4.3 million) focusing on this age group.
Methods: Following a national protocol, pathologists save DNA at all sudden unexpected natural deaths in young people, and refer for cardiac genetic opinion and molecular autopsy when the standard autopsy and ancillary tests are uninformative.
Results: Between April 2006 and February 2012, DNA was preserved on 132 1–24 year olds who had died suddenly and unexpectedly without an overt cause at initial autopsy. In 67 cases, ancillary tests (histology, toxicology, microbiology, etc) revealed a cause of death (mostly infection and poisoning). The remaining 65 DNA samples underwent genetic sequencing of six genes linked to long-QT syndrome or Brugada syndrome; 39/65 (68%) were of European ancestry and 17/65 (26%) Maori. Rare variants were identified in 13 of 65 cases (20%), seven in SCN5A, two in KCNQ1 and one each in KCNH2, KCNE1, KCNE2 and KCNJ2. Nine of these 13 deaths (69%) were nocturnal. Uncertainty remains over the pathogenicity of five of these variants. Familial evidence of cardiac ion channel disease on cardiological testing is present in five families thus far.
Conclusions: Sudden unexpected natural death in 1–24 year olds occurs with a minimal incidence of five per million general population per year, half of which remain unexplained after standard autopsy. Abnormalities in genes linked to long-QT and Brugada syndromes occur in 20% of these, with SCN5A being the commonest involved, and with most deaths occurring during sleep. Interpretation of the molecular genetic results remains a significant challenge and involves a multidisciplinary approach, including engaging with the family early in the process.

1103: DETECTION OF CORONARY ARTERY LESIONS LATE AFTER THE ARTERIAL SWITCH OPERATION FOR TRANSPOSITION OF THE GREAT ARTERIES IN CHILDREN AND ADOLESCENTS: USE OF LOW-DOSE MULTIDETECTOR COMPUTED TOMOGRAPHY (MDCT)

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Background: One of the most catastrophic late sequelae of the arterial switch operation (ASO) for transposition of the great arteries (TGA) is the development of coronary artery stenosis in approximately 7.2% of cases. The 'gold-standard' imaging modality to detect

these complications is conventional invasive X-ray angiography. Non-invasive retrospective triggered 64-MDCT has been shown to be safe and accurate in some centres, albeit with a higher effective radiation dose than conventional angiography (4.5 vs 3.1 mSv). Our objective was to assess the feasibility of low-dose MDCT in detection of coronary artery lesions with good image quality late after ASO for repair of TGA.

Methods: This was a prospective study of 14 patients operated on for transposition of the great arteries with the arterial switch operation who underwent MDCT angiography. Ethical approval to carry out this study was obtained from our institutional ethics committee, and all patients signed informed consent. Examinations were performed using prospective ECG-triggered study at a dual-source 64-row MDCT. No beta-blockers were used for these patients. Two independent investigators analysed MDCT angiography images for quality and the ability to detect coronary artery stenosis, anatomy and course. The effective dose was derived from the product of dose-length product (DLP) and conversion coefficient for the chest based on patient age using the Shrimpton and Wall method.

Results: The median age was 15 years (IQR 13.8–16.4) with 78.6% males and 21.6% females. Proximal coronary arteries were visualised in all patients despite step artifact in 2/14. Left coronary artery origin stenosis was identified in 2/14 patients. The mean dose-length product (DLP) was 40 ± 9.4 mGycm and mean effective radiation dose was 0.55 ± 0.13 mSv. The average heart rate was 74/minute.

Conclusion: Prospective ECG-triggered MDCT angiography provided good quality and interpretable images with an added advantage of lower radiation dose in coronary artery imaging in adolescents.

1113: MANAGEMENT OF HAEMOSTASIS IN PAEDIATRIC PATIENTS AFTER PLACEMENT OF A VENTRICULAR ASSIST DEVICE: REPORT FROM THE BERLIN EXCORÂ PAEDIATRIC VENTRICULAR ASSIST DEVICE IDE STUDY

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Background: Efficacious ventricular assist device (VAD) support in paediatric patients depends on successful haemostasis management, balancing the risk of thromboembolic complications with potential life-threatening bleeding. The experience with the haemostasis management algorithm developed for the EXCOR™ Paediatric IDE study is described.

Methods: All patients enrolled in the study were included. Coagulation parameters and thromboembolic and haemorrhagic events were reviewed for relatedness to antithrombotic therapy. All events were centrally adjudicated. Evaluation of adherence to the haemostasis management algorithm was based on mean results of primary (PTT, INR, anti-factor Xa) and secondary (thromboelastogram, platelet mapping) laboratory tests at pre-specified times.

Results: One hundred and nine patients in primary study cohorts 1 and 2 (n = 48), continued-access patients (n = 20) and compassionate-use cohort (n = 41) at IDE sites were included. Adherence to antithrombotic guidelines was acceptable. Pump change occurred in 52% of all patients; half were hypercoagulable based on laboratory or prior thrombotic history. Major bleeding occurred in 46% of all patients. Mean haemostatic parameters in patients with major bleeding trended to higher anti-factor Xa, PTT, and arachidonic acid platelet inhibition. Bleeding events were probably related to haemostasis management in 18% of the total IDE group events. Neurological events occurred in < 30% of all patients; mean haemostatic parameters at the time of the event trended toward lower anti-factor Xa and PTT on UFH, and higher net ADPG. Neurological dysfunction was probably related in 9% and possibly in 42% of events to antithrombotic therapy intensity.

Conclusions: Implementation of a haemostatic algorithm is crucial to minimise VAD-related adverse events. Adherence to an algorithm was possible in the setting of a multi-centre study. Incidence of significant bleeding and thromboembolic events is acceptable when balanced against potential life-saving benefits of the VAD. Additional algorithm refinements and a standardised approach may further improve safety and efficacy of haemostatic management.

1115: RELIABILITY OF ECHOCADIOGRAPHIC PARAMETERS IN PREDICTING THE GRADE OF PULMONARY REGURGITATION IN PATIENTS WITH TETRALOGY OF FALLOT: ECHO COMPARED TO CMRI

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Introduction: In tetralogy of Fallot the extent of pulmonary regurgitation (PR) and right ventricular (RV) volume load are the main criteria for long-term assessment. Today the gold standard for evaluation is cardiac magnetic resonance imaging (cMRI). Reliable echocardiographic parameters for assessing PR are lacking.

Methods: Analysis was carried out of 219 sequential cMRIs and echocardiographic examinations in 118 patients over one year (53 female, 65 male, mean age 15.7 ± 5.1 years, mean BSA 1.6 ± 0.3 m²). Data were obtained within the German competence network for congenital heart disease. cMRI: RV and left ventricular volumes and ejection fractions were measured, PR was defined as mild (≤ 25%), moderate (> 25 ≤ 40%) and severe (> 40%). Echocardiography: RV diameters in M-mode, tricuspid valve annulus diameter, tricuspid annular plane systolic excursion (TAPSE), tricuspid regurgitation, right ventricular pressure (RVP), pressure half time (PHT), ratio of the PR jet width to the pulmonary valve annulus (PR/PVA) and the ratio of the regurgitation time divided by the diastolic time interval (PRi) were assessed.

Results: PR severity correlated with RV end-diastolic volumes (p < 0.001). TAPSE showed a significant difference between mild and moderate (p = 0.033) as well as mild and severe PR (p = 0.004). PHT showed similar results comparing the PR group of mild to severe (p = 0.001) and moderate to severe (p = 0.018). Analogical results were obtained by the PRi with p = 0.028 (mild/severe) and p = 0.022 (moderate/severe) and the PR/PVA ratio between mild and severe PR (p = 0.049).

Conclusion: With increasing severity of PR there was a decline in RV function measurable by TAPSE. A short PHT and PRi indicated a more rapid equilibration of RV and pulmonary artery pressure and consequently a shorter PR time and PR/PVA ratio. TAPSE, PHT, PRi and PR/PVA ratio are reliable and reproducible echocardiographic tools to assess the severity of PR.

1117: ELECTROCARDIOGRAPHIC CHANGES AFTER CARVEDILOL THERAPY OF CONGESTIVE HEART FAILURE IN CHILDREN WITH LEFT-TO-RIGHT SHUNT CONGENITAL HEART DISEASE

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Background: Heart failure (HF) is the most common complication of left-to-right shunt congenital heart disease (L-R shunt CHD). Sympathetic activity and stimulation of the renin–angiotensin–aldosterone system are the compensation. Excessive and prolonged compensations leads to heart remodelling. Medical treatment is necessary for optimising the heart function. Carvedilol is a non-selective beta-blocker, used as an additional drug in HF. The purpose of this study was to determine the effects of carvedilol on changes in heart rate, R-wave amplitude in V6, S wave amplitude in V1 and R/S ratio in V1.

Methods: This was a randomised, controlled trial study where 30 HF L-R CHD patients were divided into two groups. The treatment group consisted of 16 children with carvedilol + standard therapy (ACEI and diuretic). The control group comprised 14 children with standard therapy. ECG was performed before and after therapy. The changes in heart rate, R-wave amplitude in V6, S-wave amplitude in V1 and R/S ratio in V1 were measured. Adverse effects were monitored. The study duration was three months.
Results: Heart rate decreased significantly in the treatment group compared to the control group (17.7 vs 4.1%; $p = 0.002$). R-wave amplitude in V6 decreased significantly in the treatment group compared to the control group (23.6 vs 7.5%; $p = 0.03$). S-wave amplitude in V1 decreased significantly in the treatment group compared to the control group (27.9 vs 10.1%; $p = 0.008$). R/S ratio in V1 increased significantly in the treatment group compared to the control group (51.6 vs 15%; $p = 0.31$). No adverse effects were found.
Conclusions: Carvedilol was effective in improving ECG parameters and safe as additional therapy to the standard therapy of CHF in children with L-R shunt CHD.

1120: MANAGEMENT OF HAEMOSTASIS IN PATIENTS AFTER PLACEMENT OF A VENTRICULAR ASSIST DEVICE: USE OF POINT-OF-CARE INR MONITORING TO MANAGE VITAMIN K ANTAGONISTS

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Vitamin K antagonists (VKA), required for haemostasis management in patients with ventricular assist devices (VADs), must be regularly monitored to avoid both thrombotic and bleeding complications. The gold-standard monitoring method is the prothrombin time (PT) testing of plasma collected via venipuncture and expressed as an INR. The point-of-care POC INR monitor is convenient for patients, uses a capillary whole blood sample and produces an INR within minutes. The POC INR monitor is demonstrated to be accurate in adults and children requiring VKA therapy facilitating more frequent INR testing; a requirement for VAD patients.
Methods: This was a prospective cohort study including all patients with a VAD treated with VKA to determine the accuracy of the CoaguChek XS®, by comparing whole-blood INR results from the CoaguChek XS® with plasma laboratory INR results. Meters were loaned to patients for use. Prior to patient use, comparisons between POC INR and laboratory INR were performed; 158 meter validations were performed from two time points from 16 patients receiving warfarin, by drawing a venous blood sample for laboratory PT INR measurements and simultaneous INR using the POC INR meter. Agreement between CoaguChek XS® and laboratory INR was assessed using the concordance correlation coefficient (CCC).
Results: VKA therapy for VAD thromboprophylaxis was received by six children, median age 4.1 years (range 3.1–15.5) and 10 adults, median age 51.2 years (range 18.6–63.0) who had $n = 5$ EXCOR, $n = 1$ Heartware, $n = 1$ EXCOR, and $n = 9$ Heartmate VADs, respectively. There were a mean of 13 (range 2–27), six (range 4–14), and nine INR tests per patient for the Berlin Heart, Heartmate and Heartware, respectively. The CCC was 0.65 and 0.72 in children and adults, respectively.
Conclusions: The CoaguChek XS® meter readings showed a moderate to good agreement when compared to laboratory INR test results.

1125: NATRIURETIC PEPTIDE (proBNP) AS A MARKER OF CARDIOVASCULAR DISEASE AND CONATAL INFECTION IN PRETERM INFANTS

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Introduction: Natriuretic peptide (proBNP) is a molecule secreted by the myocardium in response to pressure or volume overload.
Objective: To determine the relationship between proBNP levels and the presence of cardiovascular disease and/or perinatal infection in preterm neonates.
Methods: A prospective study including preterm neonates (< 32 weeks and/or < 1 500 g) was conducted at the neonatal unit of Clinica Las Condes between June 2011 and June 2012. ProBNP levels were determined between 48 and 72 h after birth by electrochemiluminescence assay. Echocardiogram, CBC, CRP and proBNP tests were performed. Patients were divided into four groups according to the presence of cardiovascular disease (PDA > 1 mm, CHD, cardiomyopahty and/or pulmonary hypertension) and/or conatal infection. The study was approved by the local IRB. Logistic regression and ROC analysis were performed to determine the predictive value and a cut-off point for proBNP.
Results: Fifty-three patients were recruited with a median gestational age of 29.6 weeks (range 24–33) and a mean birth weight of 1 316.4 g (750–1890). The median proBNP levels were 5 215 pg/ml (662–100 080) for the entire study group; 17 691 pg/ml (3 761–100 080) for patients with cardiovascular disease and 2 855 pg/ml (662–6750) ($p < 0.005$) for patients without cardiovascular disease. ProBNP levels of 17 730 pg/ml corresponded to patients with associated infection and 3 184 pg/ml ($p < 0.005$) to those without infection. To detect cardiovascular disease, proBNP levels with 80% sensitivity, 79% specificity, a positive predictive value of 60% and a negative predictive value of 91% were obtained based on the ROC curve analysis and a cutoff value of 9 970 pg/ml. With regard to using proBNP to detect perinatal infection, 92% sensitivity and 73% specificity were obtained with a cut-off value of 7 522 pg/ml.
Conclusions: The results in this study suggest the need for proBNP measurement to accurately predict cardiovascular disease in premature newborns. Possible perinatal infection of the patient should be considered.

1131: SYSTEMIC VENOUS ANOMALIES IN THE MIDDLE EAST

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Introduction: The purpose of this study was to evaluate the incidence of systemic venous anomalies in patients requiring congenital heart surgery in the region of the Middle East.
Methods: From September 2010 to March 2012, 155 consecutive patients undergone surgery for congenital heart defects were reviewed for the presence of the following systemic venous anomalies: persistent left superior vena cava (PLSVC); inferior vena cava (IVC) interruption; and retro-aortic innominate vein. The modalities utilised for the diagnosis were also reviewed: echocardiography, computed tomography (CT) scan or intra-operative.
Results: Systemic venous anomalies were identified in 28/155 patients (18.1%). PLSVC was present in 21 patients (13.5%), median age 4 months (range 3 days – 18 years), median body weight 6 kg (range 2.6–34.0 kg). IVC interruption was identified in five patients (3.2%), median age 2 months (range 30 days – 26 years), median body weight 3.7 kg (range 2.3–68.0 kg). Retro-aortic innominate vein was diagnosed in three patients (1.9%), mean age 5 years (range 10 months – 5 years), mean body weight 12 kg (range 4.5–14 kg). The diagnosis was established pre-operatively in 22/28 (78.6%) patients; in 14/28 (50%) with echocardiography, and in 8/28 (28.6%) with CT scan. In 6/28 (21.4%) patients the diagnosis was intra-operative.
Discussion: The incidence of systemic venous anomalies in this study resulted in higher values than previously reported in the litera-

ture, for a total incidence of systemic venous anomalies = 18.1% (vs 4–14% in the literature), with the presence of PLSVC = 13.5% (vs 4–11%), IVC interruption = 3.2% (vs 0.6–2%), and retro-aortic innominate vein = 1.9% (vs 0.2–1%).
Conclusions: This study showed a higher incidence of systemic venous anomalies in the Middle East population with congenital heart defects than in the previous literature reports. In a substantial percentage of patients (21.4%) the diagnosis was intra-operative. Better pre-operative screening should be performed in all patients with congenital heart defects to identify all systemic venous anomalies for a more precise planning of the surgical approach.

1134: ECHOCARDIOGRAPHIC RIGHT VENTRICULAR-TO-LEFT VENTRICULAR RATIO IN SYSTOLE CORRELATES WITH CARDIAC MRI MEASUREMENTS IN CHILDREN WITH PULMONARY HYPERTENSION

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Background: Pulmonary hypertension (PH) increases right ventricular (RV) pressure resulting in RV dilation and ventricular septal shift towards the left ventricle (LV). A ratio of systolic RV/LV diameter seeks to combine these effects into a single measure and has been shown to correlate well with pulmonary vascular resistance in children with PH. In this study we sought to validate the measure by comparing this ratio to the identical ratio by cardiac MRI (CMR) as well as CMR indices of biventricular volume and function.
Methods: Seventeen children with PH, median age 12 years, (4–23), had 18 echocardiograms and CMRs within 72 hours. Echocardiographic data included: RV end-systolic diameter (RVESD), LV end-systolic diameter (LVESD) (parasternal short axis) and RV/LV ratio. CMR data included: RVESD, RV end-diastolic volume, RV end-systolic volume, RV ejection fraction (RVEF), LVESD, LV end-diastolic volume, LV end-systolic volume, LV ejection fraction (LVEF), and RV/LV ratio. Echocardiographic measures were correlated with CMR.
Results: Echocardiographic RV/LV ratio correlated significantly with CMR RV/LV ratio. However, systolic RV/LV ratio does not correlate with CMR indices of RV or LV size or systolic function. Echocardiographic RVESD correlated significantly with CMR RV volume and function. Although echocardiographic LVESD had a negative correlation with CMR LVEF ($r = –0.62$, $p < 0.001$), there were no significant correlations between echocardiographic LVESD and CMR LV volumes.
Conclusion: There was good agreement between the systolic RV/LV ratio obtained by echocardiography and CMR. Interestingly, echocardiographic RVESD in isolation appears to be an easily obtainable and accurate descriptor of RV size and function in children with PH.

1137: CLINICAL PERFORMANCE OF SMALL-CALIBRE HIGH-VOLTAGE IMPLANTABLE CARDIOVERTER DEFIBRILLATION (ICD) LEADS IN CHILDREN AND YOUNG ADULTS

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Recent reports have called attention to ICD lead-related adverse events with small-calibre leads. This study sought to define the incidence of lead-related adverse events of small-calibre ICD leads at a single-site paediatric centre.
Methods: Clinical and lead performance information was collected retrospectively on patients ≤ 30 years of age with small-calibre right ventricular (RV) ICD lead implantation between 1995 and 2011. Small-calibre ICD leads were defined as lead diameter ≤ 7.6 French, and standard ICD leads were of diameter ≥ 8.6 French.
Results: Out of 142 patients with RV ICD lead implantation, 37 (26%) had small-calibre ICD leads, and 85% of these were single-chamber ICDs. The most frequent small-calibre ICD lead implanted was Medtronic Sprint Fidelis 6931 (Medtronic, Minneapolis, MN) in 43% ($n = 16$), followed by Medtronic Sprint Fidelis 6949 in 24% ($n = 9$), Riata 1582 (St Jude Medical, St. Paul, MN) in 19% ($n = 7$), and Riata ST 7002 in 14% ($n = 5$). In addition, 105 patients had standard-calibre ICD leads. The mean age at ICD implantation was 15.8 ± 5.7 years, with a mean follow-up time of 3.4 ± 1.6 years. All implantations were associated with acceptable lead performance at initial implant. Lead fractures occurred in 13 (35%) patients at an average of 3.1 ± 1.2 years after ICD implantation. Medtronic Sprint Fidelis 6931 and 6949 leads were most frequently affected (92% of all conductor fractures). When compared to standard-sized leads (five lead failures), small-calibre ICD leads had a significantly increased complication rate: 35 vs 5% ($p < 0.05$).
Conclusion: ICD lead-related complication rates were higher with small-diameter ICD leads than standard-calibre ICD leads. The lead-related adverse events were more frequent in the Sprint Fidelis leads when compared to the Riata leads, emphasising that lead design in addition to diameter size may play a significant role in lead performance.

1139: CARDIOVASCULAR MAGNETIC RESONANCE IN PATIENTS WITH REPAIRED TETRALOGY OF FALLOT: THE GOAL STANDARD IN ASSESSMENT OF INJECTABLE PULMONARY VALVE IMPLANTATION AND FOLLOW UP

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Background: Severe pulmonary regurgitation, progressive dilatation and dysfunction of the right ventricle are the most frequent causes of late morbidity post tetralogy of Fallot repair. Pulmonary valve replacement is often indicated in these patients. Bio-integral injectable pulmonary valve (IPV) is an innovative and less invasive technique, often done off cardiopulmonary bypass (CPB). Cardiovascular magnetic resonance (CMR) is fundamental to assess patient suitability for IPV insertion and to control the follow up.
Methods: From January 2006 to June 2012 we performed 10 pre-operative CMRs. Of these, five patients also underwent a CMR three months to six years post IPV insertion. We measured the diameters of the right ventricular patch, pulmonary valve and pulmonary bifurcation and the length of the pulmonary trunk.
Results: Ten patients were implanted with an IPV. Three months to six years post IPV insertion, CMR showed an improvement in the right ventricle end-diastolic volume. The IPV was continent and the mean transvalvular gradient was lower than that of a traditional pulmonary valve prosthesis.
Conclusions: CMR is a safe and effective method. It is necessary before IPV insertion to exclude contraindications and to determine the need for CPB. In the follow-up CMR, the pulmonary valve efficiency, the transvalvular gradients and the right ventricular function must be measured. IPV is also better detected by CMR than traditional prosthetic valves that present focal artefacts that can obscure small jets.

1143: DOUBLE-CHAMBERED RIGHT VENTRICLE. CLINICAL AND ECHOCARDIOGRAPHIC CHARACTERISTICS OF A SERIES OF 11 CASES

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Introduction: The double-chambered right ventricle (DCRV) is an uncommon congenital heart disease; it represents 0.5% of these congenital cardiopathies.
Objective: A series of 11 cases with diagnosis of double-chambered right ventricle is presented from the paediatric age to adulthood (2–64 years).
Methods: The median of age of the studied patients was 10 years (range: 2–64); 36.45% of these cases were asymptomatic and the diagnosis was made because of an incidental finding of cardiac murmur. In other cases patients did not thrive in the breatfeeding stage and in older children a worsening of NYHA functional class was observed. The echocardiogram showed double-chambered right ventricle as seen by a muscular band in all patients. The most frequent associated anomalies were: ventricular septal defect, patent foramen ovale, subvalvular aortic stenosis, and tetralogy of Fallot. Seven patients had surgical treatment and six are now in NYHA functional class I.
Conclusions: This study has special importance because is the largest series of patients with double-chambered right ventricle in Mexico and the results are comparable to studies reported in the literature. Worsening of NYHA functional class was the predominant symptom. Clinical manifestations can present from infancy to adulthood and the degree of obstruction may be progressive. Echocardiography is the method of choice in the diagnosis of these patients. The 91% of patients with DCRV who had surgical treatment had excellent haemodynamic and functional results at mid-follow up.

1144: MORTALITY AND MORBITY IN YOUNG PACEMAKER AND INTRACARDIAC DEFIBRILLATOR CARRIERS: A FOCUS ON EPICARDIAL VS ENDOCARDIAL STRATEGY IN LONG-TERM FOLLOW UP

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Background: The choice of the most appropriate procedural technique for paediatric cardiac pacing is still debated.
Aim: To evaluate the outcome, in terms of mortality and morbidity, of young PM or ICD carriers.
Methods: All PM or ICD carriers in our centre less than 18 years were included. Indications for device therapy, peri-procedural complications and long-term follow up were revised. An echocardiogram was performed in patients with isolated congenital atrioventricular block (iAVB) focused on the presence of aortic dilation.
Results: We followed 34 patients, 28 with PM and six with ICD, for a median time of 7.8 and 2.4 years, respectively. The overall survival in patients carrying an ICD was 100% (93 ± 5) in the PM group. All four deaths were complex congenital heart disease patients (CHD). The major complications were lead failures (19%) and infections (12%). The complication-free survival was 57 ± 12% at 10 years in all patients. Infections were responsible for a worse outcome in CHD. Time to first complication was significantly shorter for epicardial pacing systems (3.35 years vs 5.63 years, $p = 0.016$). The difference between epicardial and endocardial systems was not significant in terms of complications and electrical parameters. iAVB was associated with significant dilation of the ascending aorta (median z-score 2.32, Q1–Q3: 0.55–3.73, $p = 0.047$).
Conclusions: PM/ICD carriers experienced a significant number of complications. Systemic infections were responsible for the majority of deaths, especially in patients with CHD and endocardial leads. For this reason, epicardial leads used as long as possible in CHD patient might be a reasonable choice. The challenge for the future is to increase longevity of pacing systems and to reduce number of re-intervention. The positive correlation between dilation of the ascending aorta and iAVB in stimulated hearts needs further investigations.

1145: THE UTILITY OF REMOTE MONITORING OF IMPLANTABLE CARDIAC DEFIBRILLATORS AND PACEMAKERS IN A PAEDIATRIC TERTIARY CARE CENTRE

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Background: The number of paediatric and adult congenital heart disease patients with implantable cardiac defibrillators and pacemakers continues to grow on a yearly basis. Cardiovascular implantable electronic devices (CIED) require regular follow up to ascertain technical integrity. There is a paucity of published literature in the paediatric population regarding remote monitoring in patients with CIED.
Methods: A single-centre, retrospective, observational chart review was performed. Data were obtained and reviewed for patients who have CIED and were followed at the Children's Hospital of Philadelphia from 1 January 1991 to 1 November 2011. The primary objective was to describe the use of remote monitoring in paediatric patients with CIED at a large tertiary care children's hospital. The secondary objective was to determine the potential clinical benefit to patient safety and care, and determine if remote monitoring decreases the number of hospital visits due to CIED related issues.
Results: Fifty patients were included: 15 with pacemakers, 35 with implantable cardiac defibrillators. The mean age was 19 years old. The data sent by the remote monitoring system was monitored for 12 months for determination of clinically actionable events (CAE). Of 633 total transmissions, 41 resulted in a CAE in 19 patients. The CAE comprised the following: 29 arrhythmias, five lead malfunctions, and seven other issues. In the pacemaker group, the alerts that were most frequently activated were those concerning atrial tachyarrhythmia, battery exhaustion, lead impedance, as well as sensing and threshold measurements. In the ICD therapy group, the alerts nearly always activated were those concerning a detection set-off, battery exhaustion, or critical values of impedance.
Conclusions: Remote monitoring is a reliable method for following up patients with CIEDs. Accurate alert settings personalised to the patient's clinical features are essential for effective management and avoidance of excessive flow of data.

1146: UNUSUAL ATRIO-FASCICULAR ACCESORY PATHWAY IN AN ADOLESCENT WITH PRE-EXCITATION AND LONG-TERM TACHYCARDIA

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Background: Mahaim fibres are rare decremental and antegrade-only fibres connecting the right atrium to the right ventricle. We present an extremely rare case of an atrio-fascicular fibre with proximal insertion in a para-Hissian position.
Case presentation: A 17-year-old patient with pre-excitation and history of tachycardia leading to heart failure at age 2 was referred with recurrence of the arrhythmias despite therapy. The electrophysiology study showed at baseline a negative HV interval in sinus rhythm. Decremental atrial pacing lead to progressive AH prolongation and loss of pre-excitation with normalisation of the HV interval. Dual AV nodal physiology was excluded. Atrial stimulation induced a usual wide-complex tachycardia with persistence of a retrograde His. Late atrial extrastimuli during tachycardia advanced the QRS complex proving pre-excitation of the ventricle via the AP. Atrial mapping on sinus rhythm indentified the earliest signals in the right antero-septal position. On the ventricular site, the earliest potentials were identified on the moderator band. Ventricular pacing excluded retrograde accessory pathway conduction. Cryo and RF ablation of the atrial insertion was unsuccessful. We finally cryo-

ablated successfully the ventricular insertion over the right bundle potential, leaving the patient with a right bundle branch block on the surface ECG but otherwise normal His and infraHis conduction. Unsurprisingly, given the chosen technique, pre-excitation recurred overnight. The patient requested a six-month period before attempting a distal insertion RF ablation.
Conclusion: The case of para-Hissian atrio-fascicular connection represents an extremely rare finding and a further challenge for the electrophysiologist in terms of correct diagnosis as well as therapy. In our patient, a precise diagnosis was done by pacing manoeuvres. Recurrence after cryo-ablation of the moderator band confirms once again that this approach is of little long-term success in the setting of accessory pathways.

1147: ARRHYTHMIAS IN PATIENTS WITH TETRALOGY OF FALLOT: A NATIONAL DATABASE STUDY

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Objective: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease and the incidence is 0.6/1 000 live births in Taiwan. Although the surgical outcome is generally good, long-term morbidity and mortality rates are increasingly recognised. A population study from Taiwan, a country with national health insurance, easily accessible medical care and child health index similar to that in the USA, would reflect adequately the long-term risk.
Methods: We retrieved the data of TOF patients from our national health insurance 2000–2010 database.
Results: From an average 23 million population, there were 6 073 TOF patients: a prevalence rate of 44.2/100 000 (95% CI: 42.4–45.9) for the paediatric population (aged < 18 years) and 9.5/100 000 (95% CI: 9.1–10.0) for adults. Among them, 224 (3.8%) patients had arrhythmias during the study period spanning 11 years, including 165 (74%) patients with tachycardia and 59 (26%) with bradycardia. Male dominance was noted for all types of arrhythmias, particularly for ventricular tachycardia and sudden cardiac death. The mean age of each type of arrhythmia was oldest for atrial fibrillation (incidence = 0.7%, $n = 45$, 44.4 ± 15.6 years), followed by bradycardia–tachycardia syndrome (0.07%, $n = 4$, 28.9 ± 10.9 years), atrial flutter (0.18%, $n = 11$, 29.7 ± 17.8 years), sick sinus syndrome (0.13%, $n = 8$, 27.9 ± 16.6 years), ventricular tachycardia (0.46%, $n = 28$, 24.4 ± 15.3 years), supraventricular tachycardia (1.19%, $n = 72$, 16.4 ± 15.8), and atrio-ventricular block (0.77%, $n = 47$, 15.8 ± 13.9 years). Arrhythmias intervention was performed in 17 patients during the study period (RFCA in 12, ICD in three and pacemaker in 26), giving an annual risk of arrhythmia intervention at 0.028%.
Conclusion In this Asian national cohort spanning 11 years, arrhythmias occurred in 3.8% of the TOF patients, particularly in males. Tachycardia accounted for almost three-quarters of the arrhythmias and was a growing problem during long-term follow up.

1148: DEXTROCARDIA AND ASSOCIATED CARDIAC MALFORMATIONS: EXPERIENCE FROM A TERTIARY CENTRE IN SAUDI ARABIA

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Introduction: Dextrocardia is an abnormal congenital positioning of the heart in the chest, with the apex pointing towards the right side. Isolated dextrocardia without other cardiac lesions does not require treatment. Dextrocardia with situs solitus or situs ambiguous is usually associated with complex congenital heart disease and requires attention.
Objective: To determine the frequency of dextrocardia and associated cardiac defects in children presenting to our service.
Methods: This was a retrospective study. All patients with diagnosed dextrocardia referred to our tertiary cardiac centre from January 2008 to July 2012 were included in the study and their charts and echocardiograms were reviewed.
Results: A total of 8 648 patients were seen during this period**.** The number of new patients was 2 657 (30.72%) of the total. Dextrocardia was found in 30 (0.35%) of the total cohort. Of the dextrocardia cases, 14 (46.7%) had situs solitus, 14 (46.7%) situs inversus and two (6.6%) situs ambiguous. The predominance was males, with male-to-female ratio 60:40. The majority (25, 83.4%) of patients had associated congenital heart disease while five (16.7%) had no congenital heart disease. Of those with structural heart defects, 13 (52%) had situs solitus, 10 (40%) situs inversus, and two (8%) patients were in the situs ambiguous group.
Conclusion: The frequency of dextrocardia was 0.35% in our study, which is in accordance with the rest of the world (0.2–0.8%). In addition, congenital heart disease was more common in patients with dextrocardia with situs solitus or situs ambiguous.

1152: RISK OF ADVERSE CARDIAC EVENTS IN CHILDREN AND ADOLESCENTS WITH SEVERE RHEUMATIC LEFT-SIDED VALVAR LESION

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Background: Rheumatic heart disease (RHD) and rheumatic fever (RF) continue to be a scourge in developing nations. A number of patients who need valvar surgery cannot afford the intervention. With limited resources, government hospitals need to prioritise patients needing immediate surgery. This study aimed to determine the demographic and echocardiographic variables that are risk factors for the development of adverse cardiac events (ACE) in children with severe rheumatic left-sided valvar lesions.
Methods: We reviewed 376 echocardiograms of paediatric (< 19 years old) patients with RHD done at the Philippine General Hospital from January 2002 to December 2003. Based on the latest echocardiograms of 296 patients, they were grouped as to the left-sided valve most severely affected. Demographic and echocardiographic parameters of patients with ACE (death, AF, thrombus formation, and ≥ two admission/two-year period) were compared with those without ACE among patients with the same left-sided valvar lesion. A p-value < 0.05 was considered significant.
Results: The following factors were found to increase the risk of ACE: (1) severe mitral stenosis ($p < 0.0001$); (2) severe MR with concomitant moderate MS, severe TR, LVESD ≥ 3.5 cm (RR = 2.44; 1.27–4.68, $p = 0.006$), or LVEDD ≥ 5.5 cm (RR = 2.19; 1.15-4.14 p = 0.01); (3) severe AR with an EF < 60% (RR = 5; 1.15–21.78, $p = 0.03$), or LVESD ≥ 4.5 cm (RR = 4.17; 1.17–14.8, $p = 0.04$); and (4) moderate TR or PR in patients with combined severe MR and AR. Patients with severe MR and ACE were also found to be older (14.35 ± 2.42 vs 12.65 ± 3.3) and had larger LA size (5.18 ± 1.05 vs 3.99 ± 1.14). The absence of PR in patients with combined severe MR and AR conferred significant freedom ($p < 0.05$) from ACE. Based on these findings, among the 178 paediatric RHD patients with severe left-sided valvar lesion/s who were still alive during the time of the study, at least 88 patients (49.4%) were in immediate need of surgery/intervention**.**
Conclusion: The risk of adverse cardiac events in children and adolescents with RHD appeared to be associated with the severity and nature of valvar involvement and, depending on the valvar lesion, on certain demographic and echocardiographic variables, which could guide the clinician on the timing of surgery.

1155: DILATED CARDIOMYOPATHY WITH SEVERE LEFT VENTRICULAR SYSTOLIC DYSFUNCTION IN A PATIENT ON HIGHLY ACTIVE ANTIRETROVIRAL THERAPY

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Background: Cardiovascular dysfunction is a recognised complication of HIV infection in children. Cardiac complications of HIV usually occur late in the course of the disease or may be associated with drug therapy and hence become more common as therapy and survival rates improve. Left ventricular (LV) dysfunction at baseline is a risk factor for death independent of the CD_4 cell count, HIV viral load, and neurological disease.
Clinical case: We present the case of a 15-year-old girl diagnosed with HIV and subsequently started on highly active antiretroviral therapy (HAART) for the previous five years, on zidovudine, lamivudine and Nevirapine, who presented with a two-month history of cough, orthopnoea and haemoptysis. Clinically she had pedal oedema, a tachycardia of 118 beats per minute, BP = 106/79 mmHg, a laterally displaced apex, a third heart sound, basal crepitations and a tender hepatomegaly. She had a total lymphocyte count of 1 624 cells/ml, ESR of 103 mm/h and the CRP was 132 mg/dl. Her CD_4 count was 51 cells/ml. The viral load was 36 869 copies/ml. ECG showed a sinus tachycardia with diffuse ST-T segment changes and LVH with strain. Echo revealed dilated left heart chambers with severe LV systolic dysfunction and a small pericardial effusion. She was started on heart failure medicines. Two weeks later her symptoms had markedly improved but she was experiencing transient weakness of her left limbs. A repeat echo showed an intracardiac thrombus in the LV apex. Anticoagulation was started. Her HAART regimen was switched to abacavir, lamivudine and lopinavir. Six months after diagnosis, she was asymptomatic with normal LV size and normal systolic function on echo. Her CD_4 count had risen to 360 cells/ml.
Conclusion: Early cardiology referral of patients on HAART with suspected cardiac dysfunction could lead to better treatment outcomes.

1157: A WIDE QRS COMPLEX TACHYCARDIA WITH GROUP BEATING IN A PAEDIATRIC PATIENT WITH HEART FAILURE

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Background: Idiopathic sustained ventricular arrhythmias are a rare cause of cardiomyopathy in the paediatric age. We present a peculiar case of primary sustained ventricular tachycardia with an unusual group beating pattern and heart failure at diagnosis.
Case presentation: A previously healthy 11-year-old girl was referred following a pre-syncopal episode while playing sport. On arrival, a 12-lead ECG showed a sustained wide complex tachycardia with a left inferior axis and VA concordance. A prolonged strip showed an incessant arrhythmia, regularly interrupted by narrow QRS complexes. A transthoracic echocardiography demonstrated a left ventricular ejection fraction of 35%. A dose of adenosine confirmed loss of retrograde concordance for one beat without interruption of the arrhythmia.

The patient was urgently taken to the EP laboratory. The intracardiac recordings revealed a repetitive pattern of three wide beats followed by a narrow complex. Interestingly, this pattern was accompanied by progressive prolongation of the retrograde VA activation. We discovered that the ventricular activity was conducted retrogradely in an alternative pattern by a fast pathway and a decremental slow pathway. The third retrograde P wave was reconducted anterogradely by the fast pathway and depolarised the ventricular septum, creating a fusion beat. This last ventricular event blocked retrogradely and gave place for the phenomenon to start again. The ventricular tachycardia was successfully ablated at the lateral free wall of the right ventricular outflow tract. After ablation, the EP study confirmed a dual-AV node physiology. Incremental ventricular pacing at the right ventricular apex reproduced the observed phenomenon of a retrograde Wenckebach in the slow pathway without induction of supraventricular tachycardia despite aggressive protocol. No accessory pathway was found. One month after the procedure, the left ventricular ejection fraction normalised.

1165: CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES: OUTCOMES OF SURVIVORS FROM DIFFERENT SURGICAL APPROACHES IN 46 PATIENTS

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Background: Outcome of congenitally corrected transposition of the great arteries (CCTGA) depends on the associated intracardiac defects. Different surgical repairs have been proposed. Late outcomes are still poorly understood.
Objectives: To compare the outcomes of different surgical techniques in 46 patients with CCTGA.
Methods: Between 1998 and 2012, 46 patiens (mean age 47.5 months) underwent surgery. Group I (nine patients): single-ventricle repair (Glenn + extracardiac conduit); group II (11 patients): anatomical correction (four double switch + VSD closure, four Senning + Rastelli and three Mustard + Rastelli) and group III (26 patients): conventional surgery (five VSD closure, three tricuspid valve replacement and 18 conventional Rastelli). Mean follow up was: group I, 53.3; group II, 36; group III, 105 months. Tricuspid regurgitation (TR), aortic regurgitation (AR), systemic ventricle dysfunction, and AV block were assessed.
Results: Group I: (19.5%) AV-valve regurgitation was mild and non-progressive in 18%. All patients had normal ventricular function and NYHA class I. Second-degree AV block, one patient (11.1%). Survival was 83% at 120 months. Group II: (24%) TR was moderate–severe in four patients (36%). After surgery it improved in three. Six patients developed transient ventricular dysfunction (54.5%). Four patients had mild–moderate AR. Neither AV block nor residual lesions was found in the atrial switch repair. Survival was 89% (120 months). Four patients needed re-operation. Group III: (56.5%) the main complications were progressive TR in 16 patients ($p = 0.001$), associated with mild ventricular dysfunction in three patients and AV block in 10 ($p = 0.002$). Seven patients required a pacemaker implantation postoperatively and three during follow up. Six patients required re-operation (26%). Survival was 87% at 120 months.
Conclusions: Results for conventional repair were satisfactory, despite TR progression during follow up. AV block and pacemaker implantation were relevant in this group. Anatomical correction showed decreasing TR, without AV block complication, but increased risk for AR at mid-term follow up. Survival rates were similar among the three groups.

1173: INITIAL EXPERIENCE WITH A NOVEL ECHO-BASED MAGNETIC TRACKING SYSTEM FOR RECONSTRUCTION OF RIGHT VENTRICULAR VOLUMES AND FUNCTION IN ADULTS WITH CONGENITAL HEART DISEASE

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Background: Long-term monitoring of RV function in ACHD patients is important and continuous. It is critical for timing the intervention. CMR is the gold standard for this but remains a time-consuming, costly resource and impossible in some patients. Technology has developed a system that combines magnet-based

tracking with bedside echocardiography, which can be performed in much less time and more cost effectively than CMR. We hypothesised that this could be used in the majority of ACHD patients with average echo windows.
Methods: We used an industry-developed tracking system and patented software installed on a standard computer, linked up with a standard 2D echocardiography machine. Key anatomical points acquired in expiration were then processed through the software and sent securely via the internet to the industry server, where the system generated a three-dimensional model within seconds, providing volumetric measurements and ejection fraction of the RV, to the user. Twenty-one (nine male) patients, mean age 27.9 years (range 17–64 years), diagnosis: repaired tetralogy of Fallot 16; pulmonary stenosis five, underwent standard echocardiography. Key anatomical points were placed and image reconstruction undertaken. In eight patients, CMR data were available for comparison.
Results: Fifteen scans were suitable for reconstruction. Limited acoustic windows made reconstruction difficult, but poor delineation of valve planes and RV apex was even more important. In six of eight patients the echo and CMR-derived RVEF were within 6%. In two patients with limited echo images, the RVEF differed by > 10%.
Conclusions: In the majority of ACHD patients with ToF, this novel and rapid technique could be performed in out-patients to assess the RV. Reconstruction can be performed in less than 10 minutes; accuracy depends on visualising the right heart valves and the apex. The use can be extended to other CHD conditions such as systemic RV. This will greatly reduce costs and waiting times. Further experience and data are required.

1175: NURSE-LED ECHOCARDIOGRAPHIC SCREENING FOR RHEUMATIC HEART DISEASE IN FIJI: RESULTS FROM A PILOT STUDY

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Background: Rheumatic heart disease causes significant morbidity and mortality in Fiji, where the prevalence of RHD in children is approximately 1%. Antibiotic prophylaxis is most effective for mild cases, many of whom are asymptomatic. Screening with echocardiography is the most sensitive screening approach, but is limited by lack of resources, particularly experienced cardiologists and technicians. Training nurses in basic echocardiography for screening is a potential solution.
Hypothesis: We hypothesised that with simple training and supervised field experience in echocardiographic screening for RHD, nurses will have high sensitivity in the identification of children requiring referral for RHD, when compared with diagnosis by a cardiologist.
Methods: We designed an eight-week training syllabus for primary health workers in basic echocardiography for RHD, using a simplified screening protocol and referral criteria based on significant mitral or aortic regurgitation. Seven Fijian nurses participated, and were assessed in a pilot study at the end of the training programme. Nurses screened 16 children, including selected cases with known RHD. A technician performed an extended echocardiogram on all children. All echocardiograms were reported by a paediatric cardiologist.
Results: Four subjects met the echocardiographic criteria for definite or borderline RHD, and all were correctly identified for referral by all nurses (sensitivity 100%). The overall specificity was 45% (range 0–67%); 55 of 56 (98%) nurse-screened echocardiograms were of adequate quality for diagnosis.
Conclusions: The high sensitivity observed in this pilot supports the feasibility of training nurses in echocardiographic screening. The study has been expanded to a larger fieldwork phase involving screening 2 000 Fijian school-children, and is powered to formally assess the sensitivity of nurse-led echocardiographic screening. These results will assist resource-poor countries to evaluate the utility of this novel screening strategy.

1177: COMPUTER-BASED SURGICAL PLANNING AND THE Y-GRAFT: THE NEXT INNOVATION OF FONTAN'S PROCEDURE

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Background: The total cavopulmonary connection (TCPC) for single-ventricle lesions creates adverse haemodynamics that are hypothesised to negatively impact on long-term outcomes. Patient-specific computational modelling may provide novel means to improve blood flow characteristics in these patients. This study reviews our experience with two novel advances: pre-operative surgical modelling and a bifurcated Fontan baffle ('Y-graft').
Methods: Cardiac magnetic resonance (CMR) images are segmented to create patient-specific TCPC models and provide flow information. A specially designed anatomy editing tool can be used to accurately mimic Fontan surgery by simulating baffle placement. Blood flow simulations using computational fluid dynamics characterise haemodynamic metrics (i.e. power loss and hepatic flow distribution) and compare connection performances.
Results: Computer-based surgical planning has been prospectively used for 16 patients. The primary indication has been pulmonary arterio-venous malformations, which are believed to form in the absence of hepatic nutrients in the blood reaching pulmonary arterial segments. Therefore the modelling objective was to optimally distribute hepatic blood flow to the lungs. In five of 16 patients, a Y-graft Fontan baffle was the modelling recommendation based on beneficial flow-distribution characteristics, and was surgically implemented. Follow up in select patients using arterial oxygen saturations, four-dimensional phase velocity CMR, and post-operative simulations have shown favourable clinical outcomes and consistency between model predictions and operative results. Based on hypothetical energetic improvements, the Y-graft was used for 11 separate patients. It is too early to conclude on energetic characteristics compared to traditional extracardiac connections, however operative outcomes and simulated flow distribution results are both positive.
Conclusions: Computer-based surgical planning is an exciting new paradigm for patients with single-ventricle lesions, with the potential to deliver patient-specific benefit. Findings have motivated the use of the Y-graft Fontan, which may provide both improved energy efficiency and flow distribution for the TCPC.

1178: CLINICAL CLUES TO IDENTIFY ANOMALOUS ORIGIN OF LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY IN PATIENTS DIAGNOSED WITH CONGENITAL MITRAL REGURGITATION OR DILATED CARDIOMYOPATHY

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Background: Children presenting with isolated mitral regurgitation (MR) or dilated cardiomyopathy (DCM) may have anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA).
Methods: A 20-year review of ALCAPA from three centres was undertaken.
Results: There were 41 patients (27 females and 14 males). Age ranged from one month to 17 years (median 5 months). It took from one week to eight years to diagnose ALCAPA in 11 patients. The commonest reasons for referral were cardiac failure in 23 (56%), respiratory distress or asthma in 21 (51%), failure to thrive in 15 (37%), and incidental murmur in 11 (27%) patients. The initial echocardiographic diagnosis was DCM in 29 (70%), congenital MR in 10 (24%) and PDA in two (5%). MR was considered as the primary pathology in eight (18%), hence its association with ALCAPA was overlooked for as long as five years; 18 patients (43%) exhibited dilated right CA, and in seven (17%) the origin of the left CA could not be demonstrated. In 15 (37%) patients abnormal retrograde flow in the pulmonary artery was noted. The commonest ECG findings were non-specific T-wave or ST-segment changes in 24 (59%) and 18 (43%) patients, respectively; 22 (54%) patients had pathological Q waves in lead aVL. Ninteen (46%) patients required cardiac catheterisation to confirm ALCAPA. In two patients, ALCAPA was suspected after PDA ligation or during mitral valve replacement; 39 (95%) patients had surgery immediately after diagnosis, but two were not operated on. Post-surgical echocardiograms showed resolution of MR and left ventricular dysfunction in 29 (70%). Three (16%) patients died after surgery.
Conclusion: Isolated MR and left ventricular dilatation are important and frequent findings in ALCAPA syndrome. Any child with echocardiographic diagnosis of congenital MR or DCM should prompt a careful search for ALCAPA.

1179: FOETAL AND POSTNATAL OUTCOME OF 22Q11 DELETION AND ASSOCIATED CONGENITAL HEART DEFECTS

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Background: 22q11.2 deletion is the most common chromosomal microdeletion syndrome. It has a strong association with conotruncal heart defects and may exert negative influence on the clinical outcomes.
Methods: A review was done of patients with 22q11.2 deletion who either presented to the University Hospital of Wales with congenital heart disease or reported to the Congenital Anomaly Register and Information Service for Wales (CARIS) between 1990 and 2011.
Results: During the study period, 102 cases of 22q11.2 deletion were reported, giving a gross prevalence of 1.2 per 10 000 total births (one in 8 335 total births); 95 cases had both 22q11.2 deletion and at least one congenital heart defect (CHD) (93%). Seven cases had a normal heart. In 18 cases, diagnosis was established in foetal life: pregnancy was terminated in four cases, still birth was found in one and in 13 cases pregnancy resulted in a live birth (72%). Eighty-four patients were diagnosed at the postnatal period. Mean follow-up duration was 16.2 ± 11.4 years (range 1–52 years, median 14.5 years). Eight patients died after birth, seven in the first year of life, and one at age 3.8 years.
Conclusions: Of patients with 22q11.2 deletion, 93% exhibited CHD. In spite of the antenatal detection rate being low, survival was similar for all cases irrespective of the presence of CHD. Most deaths occurred within the first year of life but beyond infancy survival was favourable

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1191: A NEW METHOD FOR MANUFACTURING FLEXIBLE REPLICAS OF CONGENITAL HEART DISEASE USING STEREOLITHOGRAPHY AND VACUUM-CASTING TECHNIQUE: REHEARSAL OF SURGICAL OPERATION

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Background: Stereolithography is a rapid prototype technology whereby an ultraviolet laser beam selectively polymerises and solidifies photosensitive and polymeric liquid plastic. By using this technique, 3D volumetric image data of multi-slice (MS) CT can be converted into plastic models that enhance our spatial perception of real-life anatomy and pathology. However, the materials of the stereolithography are limited to several photosensitive plastics or urethane, which are not satisfactory for simulation surgery. Recently, a vacuum-casting method has been developed, where more delicate models with different stiff materials can be manufactured.
Methods: Three-dimensional volumetric data sets of MSCT angiography of congenital heart disease (CHD) were converted into standard triangulated language files to make stereolithographic biomodels representing both the outer and inner surface of the heart. Then, urethane with appropriate stiffness was injected using the vacuum-casing method. After solidification of the urethane, the casts were carefully removed.
Results: We have made replicas of CHD including VSD, ASD, TOF, DORV, SV, criss-cross heart, and ccTGA. The vacuum casting in association with stereolithography enabled us to manufacture replicas with similar texture and structure to the real heart. This technique also allowed the surgeon to cut and suture, facilitating the simulation of the surgical operation.
Conclusion: The vacuum casting in association with stereolithography is a promising technique for the pre-operative practice and simulation of individual surgery, and planning of novel and innovative surgical procedures of CHD. This technique could be helpful for rescuing children with CHD.

1192: DAMUS-KAYE-STANSEL ANASTOMOSIS MITIGATES THE RISK OF SYSTEMIC OUTFLOW TRACT OBSTRUCTION IN CHILDREN WITH SINGLE VENTRICLE CARDIAC ANOMALIES

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Background: Children with various single-ventricle (SV) pathologies are at risk of developing systemic ventricular outflow tract obstruction (SVOTO) following volume unloading with cavo-pulmonary connection (CPC). We aimed to evaluate the efficacy of Damus-Kaye-Stansel anastomosis (DKS) at time of CPC in decreasing late SVOTO risk.
Methods: A retrospective review of SV patients who underwent DKS concurrent with CPC between 1997 and 2012 was performed. Clinical, echocardiographic and angiographic outcomes were analysed.

Results: Thirty-six children with SV underwent DKS at the time of Glenn bidirectional CPC (n = 29) or Fontan total CPC (n = 7). The underlying anatomy was double-inlet left ventricle (n = 19), double-outlet right ventricle (n = 7), unbalanced atrio-ventricular septal defect (n = 4) and other (n = 6). Prior palliation included pulmonary artery band (n = 35), atrial septostomy (n = 9), and coarctation/arch repair (n = 10). Median age at time of DKS was 8.7 months (range 3.5 months – 8.9 years) and median weight was 6.3 kg (range 5–27 kg). Overall survival was 87% at five years. Deaths were unrelated to DKS or SVOT complications. Pressure difference between the systemic ventricle and the aorta decreased from 14 ± 15 mmHg pre-operatively to 0 after DKS (p = 0.005). At last follow up, none of the patients developed recurrent SVOT gradient; 73% of them had zero or trivial aortic/neo-aortic valve regurgitation while 20 and 7% had mild or moderate regurgitation, respectively. None of the patients had evidence of compression of the left pulmonary artery or bronchus. None of the patients had heart block requiring permanent pacemaker insertion; 86% of the patients have reached or are suitable candidates awaiting final palliation stage.
Conclusions: DKS can be safely performed in conjunction with CPC without added mortality hazard. It is very effective mitigating the SVOTO risk with good durable semilunar valve function. Our data support an aggressive approach of performing concurrent DKS with CPC in children with SV pathologies at risk of developing SVOTO.

1197: DEVELOPMENT OF THE VANGUARD DERMONSTRATION SITE FOR RHD AMONG SCHOOL-AGED LEARNERS IN THE WESTERN CAPE, SOUTH AFRICA

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Background: In South Africa, anecdotal information suggests that rheumatic heart disease (RHD) is still the leading acquired heart disease in young adults. Recent data on the prevalence of RHD in school-aged learners remain scanty, with two earlier studies suggesting an estimate around seven per 1 000. However this figure is thought to be an underestimation given that the screening was performed using auscultation, which is less sensitive than echocardiography.
Objective: We have developed as part of ASAP, an echocardiography-based RHD screening programme for learners within the Vanguard community of the Western Cape. Outcomes include determining the prevalence of RHD, monitoring the disease progression in scree-positve participants referred for appropriate follow up, and evaluating the cost effectiveness of such a programme.
Methods: We present the issues involved in the establishment of the demonstration site, the lessons learned, and the ongoing challenges facing the research team, having screened in excess of 2 500 participants. In brief, we detailed our experience in five areas: engaging the community on all levels; completing a situational analysis and gathering background data, including sampling frame and the random-sampling procedure; consent process; on-site data collection and management; post-screening responsibility and continued community involvement.
Conclusion: We concluded that screening for RHD is feasible in the community setting using schools as the sample frame. We firmly believe that our experience will serve as a meaningful resource for other similar research programmes.

1200: EFFECTS OF ADRENERGIC RECEPTOR BLOCKER TREATMENT ON RIGHT HEART FAILURE

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Background: The late diagnosis and surgical management of left-to-right shunt congenital heart disease (L-R shunt CHD) causes pulmonary hypertension. To date there is no effective drug to decrease pulmonary artery systolic pressure. Beraprost, an oral prostacyclin analogue which works as a pulmonary artery vasodilator, was expected to decrease PASPS in children with L-R shunt CHD.
Methods: A pretest–postest study was conducted on 17 subjects aged two months to 16 years old, with VSD, ASD, PDA or a combination, who developed pulmonary hypertension. Patients with other structural cardiac defects or with pulmonary infection were excluded. Echocardiographic examinations were performed before and after administration of beraprost 1 µg/kg/day bid for three months. V_{max} TR, V_{max} L-R, and PASP were measured and M-mode echo of the pulmonary valve, including a wave, EF slope and mid-systolic notch, was done. Adverse reactions were noted, and monitoring was performed every two weeks.
Results: V_{max} TR decreased (0.7 ± 0.79 m/s, p = 0.004), V_{max} L-R increased (0.6 ± 0.64 m/s, p = 0.018) and PASP decreased (18.3 ± 21.90 mmHg, p = 0,003). There was no significant change in wave, EF slope and mid-systolic notch.
Conclusions: There was a decrease in pulmonary artery systolic pressure after administration of beraprost in children with L-R shunt CHD who develop pulmonary hypertension.

1202: BICYCLE STRESS ECHOCARDIOGRAPHY IN CHILDREN: FEASIBILITY, SAFETY AND DETERMINATION OF INTER-OBSERVER VARIABILITY

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Background: In the adult population, exercise-stress echocardiography (ESE) is a well-validated technique providing a dynamic evaluation of myocardial structure and function. In children, its use is rather limited due mainly to a precipitously rapid drop in peak heart rate after exercise. The aim of this study was to assess the feasibility, safety and reproducibility of ESE using on-line scanning in a semi-supine cyclo-ergometer protocol in a wide spectrum of children.
Methods: Between July 2008 and April 2012, 42 patients (mean age 14.1 ± 2.9) underwent a bicycle ESE. Two independent observers without knowledge of any patient data interpreted all stress studies, grading quality of each acquired image and presence of regional wall motion abnormalities (RWMA).
Results: ESE was successfully performed and well tolerated by all patients. Rest HR was 82 ± 13.6, peak HR was 153.4 ± 19.7 (161.5 ± 18.3 excluding heart transplant patients and patients on beta-blockers). Among 464 views acquired, the visualisation was optimal in 403 (87%), suboptimal in 32 (7%) and inadequate in 29 (6%). Among 10 patients with hypertrophic cardiomyopathy, we were able to assess a significant left ventricular outflow tract gradient increase (> 25 mmHg) during exercise in three patients (33%). ESE was performed in 29 patients with congenital or acquired coronary abnormality (Kawasaki disease, heart transplant recipients, congenital coronary abnormalities, transposition of the great arteries after arterial switch operation). In this group, RWMA were revealed in eight patients (28%). The agreement between observers showed a κ-index of 0.7276 (95% CI = 0.6497–0.8055) for the image quality and a κ-index of 0.5125 (95% CI = 0.4782–0.5468) for the RWMA analysis.
Conclusions: Bicycle-stress echocardiography performed by on-line scanning during exercise is a feasible, safe and reproducible modality in children. Further data to assess its diagnostic accuracy are however needed.

1204: ASSOCIATED PULMONARY ATRESIA IS A SIGNIFICANT RISK FACTOR FOR MORTALITY IN SYMPTOMATIC NEONATES WITH EBSTEIN'S ANOMALY

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Background: Ebstein's anomaly (EA) presenting in the severely symptomatic neonate remains a complex entity with high mortality rates. There is no accepted standard approach to surgical repair, and opinions vary between complete biventricular repair and single-ventricle (Fontan) pathways. More than half of neonates with EA have associated anatomical pulmonary atresia (PA). The outcome in this subset is not well defined. We report on our experience with the early and intermediate outcome of EA/PA.
Methods: From 1994 to 2011, 35 consecutive patients (26 neonates, nine young infants) with EA were operated on by a single surgeon (CKC). Of the 26 neonates, 16 had associated anatomical pulmonary atresia (61%, 16/26) and seven had functional PA. For the entire cohort ($n = 35$), follow up was complete in 93% of patients, extending to 18 years (mean 6.4 ± 4.6).
Results: Thirty-two of the 35 patients had a complete two-ventricle repair, and three patients had a single-ventricle palliation (BT shunt, $n = 2$, Starnes palliation, $n = 1$). Hospital mortality for the group was 20% (7/35). For patients with associated PA, mortality was 37.5% (6/16) compared to 5.2% (1/19) for patients without PA ($p < 0.05$). The hospital mortality for patients with EA/PA undergoing biventricular repair was 46% (6/13), and the late mortality was 22% (2/9). In our more recent experience (2005–2011) we altered our management of patients with EA/PA to include either a RV-PA valved conduit or a single-ventricle palliation ($n = 7$) with no early mortality. Kaplan-Meier 15-year survival estimates were 79 ± 15% for neonates without pulmonary atresia, and 40 ± 15% for those with pulmonary atresia ($p = 0.03$).
Conclusions: The surgical management of EA with pulmonary atresia in neonates continues to evolve. The early survival of neonates with EA/PA is improved when a RV-PA conduit is included in the repair or the patient is stratified to a single-ventricle palliation.

1206: RARE COMPLICATIONS OF CONGENITAL BICUSPID AORTIC VALVE DISEASE, POSSIBLE CORONARY ARTERIOPATHY

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We present a case of a patient with unusual complications post Bentall repair and aortic valve replacement, possibly due to arteriopathy involving the proximal coronary trees. A 46-year-old well woman was referred for assessment of palpitations. She was found to have a bicuspid aortic valve with mild aortic stenosis, mild-to-moderate aortic regurgitation, but significantly dilated aortic root and left ventricular dilatation, all of which increased in size under surveillance. CTCA showed normal coronary arteries. She was referred for cardiac surgery, which was complicated by emergency vein graft to the LAD as the patient did not come off-pump easily.

She was initially well, then four months later presented with acute chest pain. A small 4-mm defect in the anterior wall of the prosthesis with a large pseudo-aneurysm was found on echo, confirmed on angiography and CTA, and subsequently closed with an Amplatz device. Angiography then showed mild RCA (FFR0.91) and left main coronary artery stenosis. The vein graft was occluded. The patient subsequently presented two months later with crescendo angina, and was found to have critical 95% + left main coronary artery stenosis, and was stented successfully. The RCA stenosis was confirmed but did not merit PCI.

This patient seems to have developed rapid severe intimal hyperplasia, possibly triggered by instrumentation on the substrate of bicuspid aortopathy/arteriopathy. Bicuspid aortic valves are known to be associated with cystic medial necrosis and premature coronary artery disease, abnormal vascular reactivity, coarctation, and cranial aneurysms but this patient developed severe coronary disease within a few months of having normal coronary anatomy, suggesting possible rapid intimal hyperplasia. We have not found any similar reports in the literature. Echo and three-dimensional CT images will be displayed.

1208: PULMONARY VENO-OCCLUSIVE DISEASE IN CHILDREN

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Background: Pulmonary veno-occlusive disease (PVOD) is a rare lung disease and accounts for five to 10% of all cases with pulmonary hypertension (PH) of unknown aetiology. Incidence, prevalence and aetiology of PVOD in children are not well defined. The mortality remains high, which is partly related to limited treatment options.
Methods and Results: This retrospective analysis (1985–2011) summarises symptoms, associated factors, treatment and outcomes of nine paediatric patients (five female, four male) with histological confirmation of PVOD. PH was diagnosed at a mean age of 13.5 years (range 8–16 years), followed by the definitive diagnosis of PVOD at a mean age of 14.3 years (range 10–16 years). Symptoms, such as decreased exercise tolerance ($n = 6$) and/or shortness of breath ($n = 9$) preceded the diagnosis by two years on average; the mean survival time after diagnosis was 14 months (range 0–47 months). Computed tomography (CT) of the lungs showed typical radiological features. Treatment included home-oxygen ($n = 5$), diuretics ($n = 9$), warfarin ($n = 4$) and pulmonary vasodilators ($n = 4$). Four children were listed and three successfully lung transplanted. Eight patients died, including two after lung transplant. One transplant patient survived with good quality of life.
Conclusions: PVOD is an important differential diagnosis for patients with PH. CT is a valuable tool to image lung abnormalities; the definitive diagnosis can be made only by lung biopsy, which subjects the patient to additional risk. Early listing for lung transplantation is essential, as the mean survival time is only 14 months.

1212: ANOMALOUS ORIGIN OF A CORONARY ARTERY FROM THE AORTA: OUTCOMES OF TREATMENT STRATEGIES

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Background: Anomalous origin of a coronary artery from the aorta (AOCA) is often an incidental finding but can be associated with myocardial ischaemia and sudden death, especially in athletes. Because AOCA is rare and the natural history unclear, there are no established management guidelines. We sought to investigate outcomes of various treatment strategies.
Methods: We reviewed records of patients with the diagnosis of AOCA seen in our hospital between 1995 and 2012. We included all patients with coronary origin from an inappropriate sinus or single coronary ostium, and excluded patients with other congenital or acquired coronary abnormalities or a CHD. Diagnosis, presentation, cardiac testing results, treatment, and outcome were recorded.
Results: We identified 114 patients, 72 (63%) male, ages 0 to 49 years (mean 9.7 ± 9.7 years). The LCA arose from the right sinus in 20 (18%), the RCA from the left sinus in 73 (64%), and the

LCA from the non-coronary sinus in three (3%). There was a single coronary ostium in 18 (16%). Fifty-three (46%) were symptomatic at presentation. Diagnosis was made by echocardiography in all and confirmed by TEE (53%), cardiac MRI (37%), catheterisation (19%), or angio CT (17%). Surgery was performed in 48 (42%) patients, including unroofing (41), re-implantation (four), CABG (two), and excision of a prominent sinotubular ridge (one). There were no surgical deaths but 13% of patients had complications. The mean CICU stay was two days (range 1–16) and the mean hospital stay was five days (range 3–20). There were no deaths or significant morbidities during follow up (mean 33 months). Ten patients reported continued symptoms, five after surgical repair. Fifty-two patients were physically active.

Conclusions: Death or other significant clinical events are rare in young patients with AOCA. Surgical correction can be performed with very low risk but symptoms can persist regardless of treatment strategy.

1216: DIFFUSION TENSOR MAGNETIC RESONANCE IMAGING TRACTOGRAPHY DETECTS MYOFIBRE ARCHITECTURE IN DEVELOPING HUMAN FOETAL HEARTS

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Background: The timing and mechanism of development of the helical myo-architecture of the human heart is largely unknown. Understanding normal development is essential for detection of alterations in myofibre architecture associated with congenital and acquired heart disease. Diffusion tensor MRI (DTI) tractography exploits the preferential diffusion of water along the direction of muscle fibres to define myofibre structure and orientation non-destructively.

Hypothesis: We hypothesised that DTI tractography could delineate myofibre architecture during heart development.

Methods: Normal human foetal hearts ranging from 10 to 19 weeks' gestational age (GA), stored in 10% formalin were studied. DTI data were obtained using a 4.7-Tesla magnet and 24 diffusion-encoding directions. The primary eigenvectors of the diffusion tensor field were integrated into streamlines and colour-coded by their helix angle (HA). Hearts were then sectioned for histology parallel to the LV free wall and stained with H&E. Fibre orientation (DTI tractogram) by MRI at multiple levels through the LV wall was compared with the registered histology sections.

Results: Fibre orientation by DTI correlated well with histology. At 10 weeks the myocardium resembled an isotropic tissue without distinct myofibre patterns. However, by 19 weeks of gestation, LV myo-architecture closely resembled that seen in adult hearts. The helix angle varied smoothly from a left-handed helix (0° to –90°) in the subepicardium (epi) to a right-handed helix (0° to 90°) in the subendocardium (endo). Fibres in the midwall (mid) were circumferential (± 0°). The characteristic crossing helical pattern was present but less well developed at 14 weeks.

Conclusions: DTI tractography shows that developing human myocardium is relatively isotropic at the end of embryogenesis (10 weeks), develops microstructural anisotropy by 14 weeks, and essentially resembles adult myocardium by mid-gestation (19 weeks).

1222: RHEUMATIC FEVER: A SOUTH AFRICAN PERSPECTIVE A PILOT STUDY ASSESSING ADHERENCE, NOTIFICATION AND SECONDARY PROPHYLAXIS

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Hypothesis: (1) The notification system for acute rheumatic fever (ARF) is underutilised and inadequate to establish a central data base of rheumatic fever patients. (2) Adherence to secondary prophylaxis is poor. (3) The factors underlying adherence is multifactorial and can be addressed by the healthcare system.

Methods: Thirty-four patients with previous ARF (modified Jones criteria) or established rheumatic heart disease (RHD) (echocardiography) were subjected to semi-structured interviews. Adherence was gauged against provincial guidelines as a gold standard. Echocardiograms were assessed for morphological and functional abnormality. All patient names were cross correlated with provincial notification data to evaluate the effectiveness of the notification system.

Results: The notification system for ARF at TBH is ineffective, with utilisation figures during the past nine years of only 12.67%. Adherence figures were found to be inadequate: 33.8% in the high-risk period (< 25 years) and 11.06% in the total recommended period. Patients' knowledge of their diagnosis, information transfer at the time of diagnosis and the presence of a reminder system were some of the strongest predictors of adherence. Xhosa-speaking patients living far from healthcare facilities had a poor adherence. Lack of social support, the absence of indoor toilet facilities and long waiting times for treatment were significant predictors of poor adherence.

Conclusion: Gross under-reporting of ARF occurs in TBH. Adherence to secondary prophylaxis is poor. Factors that are modifiable by the healthcare system, which predicted adherence included patients' knowledge of their condition, information transfer at the time of diagnosis, patient perception of the need for prophylaxis and the presence of a reminder system.

1223: PROGNOSTIC VALUE OF SUBMAXIMAL CARDIOPULMONARY EXERCISE PARAMETERS FOR CARDIAC MORBIDITY IN FONTAN PATIENTS

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Background: Submaximal exercise parameters are associated with an increased risk of hospitalisation in patients with heart failure, but the implication in patients with Fontan circulation remains unknown. Therefore, we investigated the prognostic value of submaximal exercise parameters in Fontan patients, in whom maximal exercise effort is often limited.

Methods and Results: Fifty-two patients who had received Fontan completion at least 12 months earlier underwent cardiopulmonary exercise tests at our institute. We evaluated two maximal parameters [peak oxygen consumption (VO_2) and heart rate reserve (HRR)] and two submaximal parameters [oxygen uptake efficiency slope (OUES) and minute ventilation (VE) to carbon dioxide elimination (VCO_2) slope]. Peak VO_2 and the OUES were expressed as the percentage of predicted values. During a median follow up of 22.7 months, 11 patients (21%) exhibited cardiac morbidity, defined as cardiac-related hospitalisation. Time-dependent receiver operating characteristic curve analysis demonstrated that submaximal parameters were related to two-year cardiac morbidity (area under the curve for the OUES = 0.781, $p = 0.018$; for the VE/VCO_2 slope = 0.714, $p = 0.04$), whereas peak VO_2 and HRR were not. The optimal threshold value for the percentage of the predicted OUES was 45%, and for the VE/VCO_2 slope, it was 37%. Furthermore, the OUES conveyed independent prognostic information beyond resting oxygen saturation and a history of heart failure or protein-losing enteropathy.

Conclusion: Submaximal exercise parameters provided superior prognostic information to maximal exercise data for predicting cardi-

ac morbidity in Fontan patients. Moreover, the association between the OUES and cardiac morbidity was independent of relevant baseline clinical information.

1230: LONG-TERM MANAGEMENT OF CO-MORBIDITIES ASSOCIATED OBESITIY IN CHILDREN WITH PREADER-WILLI SYNDROME

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Background: Prader–Willi syndrome (PWS) is the most common syndromic form of obesity, caused by deletion in the 15q11-13. Management of obesity and its complications is a critical part of care.
Objective: To present long-term management of co-morbidities associated with obesity in children with PWS.
Case: A seven-year-old girl was admitted to Saiful Anwar General Hospital due to dyspnoea on effort. She was born to non-cosanguineous and healthy biological parents. Her developmental progress was delayed. She had mild mental retardation, speech articulation defect and global developmental delay. She developed feeding difficulties in infancy but became preoccupied with food, accompanied by a compulsion to eat at three years old. At five years she had morbid obesity. Physical examination showed dysmorphic features. Genetic testing indicated deletion in the 15q11-13. We found dilated cardiomyopathy and type 2 diabetes mellitus as a complication of obesity. Echocardiography showed decreased ejection fraction and mild MI/MR. Long-term management included monitoring and management of the co-morbid conditions, such as medication for the cardiomyopathy, a well-balanced, low-calorie diet, regular exercise, environmental modification and antidiabetic agents. Early intervention and special education, followed by supportive employment, were appropriate to address the developmental disabilities. Physical, occupational and speech therapy were started after the establishment of a diagnosis. This resulted in a decrease in BMI, improvement in the cardiomyopathy, improvement in developmental and behavioural problems, including food-seeking behaviour, thus improving the quality of life.
Conclusion. Comprehensive management of Prader–Willi syndrome resulted in improvement in the quality of life of the patient.

1234: THE USE OF COMPUTED TOMOGRAPHY ANGIOGRAPHY (CTA) IN THE EVALUATION OF CONGENITAL HEART DISEASE

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Background: While echocardiography is the commonest non-invasive imaging modality used in the diagnosis of congenital heart disease, CT angiography may be useful to further elucidate anatomical detail that is crucial to accurate surgical planning.
Methods: From 2009 to the present, at our institute CT angiography has been used selectively to evaluate pulmonary artery size, pulmonary venous drainage, aortic arch abnormalities and coronary artery abnormalies in patients with congenital heart disease. Furthermore, select patients who underwent corrective or palliative surgery were evaluated by CTA.
Results: The use of CTA as an adjunct to echocardiography has enabled accurate pre-operative assessment and facilitated surgical decision-making.
Conclusions: The frequent use of CTA has enabled cardiologists, congenital cardiac surgeons and radiologists alike to familiarise themselves with the anatomical details evident on CT angiography, and correlation with the intra-operative findings enhances the interpretation of the study.

1236: INTERVENTION FOR RE-COARCTATION IN THE SINGLE-VENTRICLE RECONSTRUCTION TRIAL: INCIDENCE, RISK AND OUTCOMES

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Objectives: To determine the incidence of re-coarctation (re-CoA), risk factors and outcomes in patients with single right ventricle lesions after the Norwood procedure.
Methods: The cohort included subjects randomised to right ventricle–pulmonary artery shunt (RVPAS) or modified Blalock–Taussig shunt (MBTS) in the Single Ventricle Reconstruction (SVR) Trial. Re-CoA was defined by intervention, either catheter-based or surgical. Univariate analysis and multivariable Cox proportional hazard models were performed adjusting for centre.
Results: Of the 549 SVR subjects, 97 (18%) underwent 131 interventions (92 balloon aortoplasty; 39 surgical) for re-CoA. Intervention typically occurred at pre-stage II catheterisation (n = 71, 54%) or at stage II surgery (n = 38, 29%). Median (range) age and catheterisation gradient at first intervention were 4.9 months (1.1–10.5) and 17 mmHg (0–60). Centre intervention rates varied from 0–50%. In multivariable analysis, re-CoA was not associated with assigned shunt type, but was associated with actual shunt type received (HR 2.0 for RVPAS vs MBTS, p = 0.02), and Norwood discharge peak echo-Doppler arch gradient (HR 1.07 per 1 mmHg, $p < 0.01$). No other demographic, anatomical or surgical variables predicted intervention. Subjects with re-CoA demonstrated co-morbidities at pre-stage II evaluation, including higher pulmonary arterial pressures (15.4 ± 3.0 vs 14.5 ± 3.5 mmHg; p = 0.05), higher PVR (2.6 ± 1.6 vs 2.0 ± 1.0 WU × m²; p = 0.04) and increased echocardiographic volumes (end-diastolic volume: 126 ± 39 vs 112 ± 33 ml/BSA$^{1.3}$; p = 0.02). There was no difference in 12-month post-randomisation transplant-free survival for those with and without re-CoA (91 vs 93%; p = 0.7).
Conclusions: Intervention for re-CoA was common and varied by centre. Intra-operative crossover due to complex arch anatomy may explain the association with receipt of an RVPAS. Although those undergoing intervention demonstrated co-morbidities prior to stage II, there was no difference in one-year transplant/mortality. Further evaluation is warranted to evaluate effects of the morbidity rate of re-CoA.

1239: FACTORS ASSOCIATED WITH SERUM B-TYPE NATRIURETIC PEPTIDE LEVELS IN INFANTS WITH SINGLE VENTRICLE

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Background: Data regarding the value of B-type natriuretic peptide (BNP) in infants with single-ventricle (SV) physiology are lacking. The objective of this analysis was to describe the changes in BNP in infants with SV physiology before and after superior cavopulmonary connection (SCPC) surgery.
Methods: BNP levels were measured by a core laboratory pre-SCPC (5.0 ± 1.6 months) and at age 14 months during a multicentre trial of ACE inhibition therapy in infants with SV. Associations between BNP, clinical and laboratory variables were assessed using log-transformed BNP. To identify the predictors of BNP, longitudinal analysis was performed for each set of grouped variables (echo, catheterisation, growth), and multivariable analysis was performed using patient characteristics at both visits and neurodevelopmental predictors at the 14-month visit.

Results: BNP was significantly higher ($p < 0.01$) at pre-SCPC ($n = 173$) compared to 14 months ($n = 134$); median (IQR) 80.8 pg/ml (35–187) vs 34.5 pg/ml (17–67). BNP > 100 pg/ml was present in 72 (42%) of subjects pre-SCPC and 21 (16%) at 14 months. Patients with BNP at both visits ($n = 117$) had a median (IQR) decrease of 32 (1–79, $p < 0.01$). In longitudinal analysis, higher levels of BNP were associated with increased echocardiographic end-systolic volume *z*-score ($p = 0.01$), greater degree of AV valve regurgitation ($p < 0.01$), lower weight *z*-score ($p < 0.01$), and lower length *z*-score ($p = 0.02$). In multivariable analyses at 14 months, higher BNP level was associated with presence of arrhythmia post-SCPC surgery ($p < 0.01$), prior Norwood procedure ($p < 0.01$), and longer length of hospital stay post-SCPC surgery ($p = 0.04$), as well as lower Bayley Psychomotor Developmental Index ($p = 0.015$).
Conclusion: BNP decreased in infants with SV after SCPC surgery. Elevations in BNP were associated with increased ventricular dilation, increased atrio-ventricular valve regurgitation, and poorer growth and neurodevelopmental outcomes. Therefore, BNP may be a useful seromarker in identifying infants with SV at risk for worse outcomes.

1240: MAGNETIC RESONANCE ASSESSMENT OF MYOCARDIAL SCARRING AND VENTRICULAR FUNCTION BEFORE AND AFTER REPAIR OF ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

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Background: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare cardiovascular anomaly. Left ventricular (LV) dilatation and dysfunction (DCM) evolves due to diminished myocardial perfusion caused by coronary steal phenomenon. Myocardial scarring has been shown in ALCAPA patients late after repair, however, the incidence of myocardial scarring before surgery and its impact on the postoperative course is unknown. Accordingly, the aim of the study was to assess myocardial function and viability using CMR imaging before and in the short-term follow up after ALCAPA repair.
Methods: Eight patients (mean age 10.0 ± 5.8 months) with diagnosed ALCAPA underwent CMR before and after (mean time interval 4.9 ± 2.5 months) coronary re-implantation procedures (six direct re-implantation, two Takeuchi method). CMR included functional analysis and late gadolinium enhancement (LGE) for detection of myocardial scars.
Results: Severe LV dilatation (mean indexed LVEDV 171 ± 94 ml/m^2) and dysfunction (mean LV-EF 22 ± 10%) were present in all patients and improved significantly after surgery in seven of eight patients (mean LVEDV 68 ± 42 ml/m^2, p = 0.02; mean LV-EF 58 ± 19%, $p < 0.01$). Myocardial scarring (one apical subendocardial, one midventricular transmural) was present in two of the eight patients (25%) and did not predict postoperative course or functional recovery. Early follow-up CMR showed an unchanged degree of infarcted myocardium in these patients. Although functional recovery was sufficient, transmural scarring not evident before re-implantation was found at follow-up CMR in two patients.
Conclusions: Despite often severely compromised LV function preoperatively, myocardial scarring was only present in the minority of our patients. According to our experience, myocardial dimension and function recovered quickly after coronary re-implantation, independently of myocardial scarring. Further studies are needed to elucidate the endogenous repair mechanisms that are responsible for the re-remodelling of the LV in patients with ALCAPA.

1244: INTER-PARAMETRIC CORRELATION BETWEEN ECHOCARDIOGRAPHIC MARKERS IN PRETERM INFANTS WITH PATENT DUCTUS ARTERIOSUS

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Background: Various echocardiographic parameters are studied in the assessment and evaluation of patent ductus arteriosus. In isolation, many of these parameters have low sensitivity and specificity for ductal significance when compared to ductal size.
Aim: To correlate various echocardiographic parameters with ductal size (transductal diameter) in infants with symptomatic patent ductus arteriosus, and ascertain the sensitivity, specificity, positive and negative predictive values and likelihood ratios of various parameters to predict a duct ≥ 3mm in size.
Methods: Preterm infants less than 32 weeks' gestation who were evaluated for the presence of a patent ductus arteriosus were assessed. The following echocardiographic parameters were measured: transductal diameter (TDD), ductal velocity, end-diastolic left pulmonary artery (LPA) flow, ductal diameter-to-left pulmonary artery ratio (TDD:LPA), left atrial-to-aortic root ratio (LA:Ao), left ventricular output-to-superior vena cava flow ratio (LVO:SVC), transmitral E/A ratio and isovolumic relaxation time (IVRT).
Results: Fifty-two infants were enrolled with a mean gestational age of 26 ± 2 weeks and mean birth weight of 837 ± 240 g. The mean transductal diameter was 2.8 ± 0.5 mm. Transductal diameter correlated significantly with ductal velocity, end-diastolic LPA flow, TDD:LPA, LA:Ao and LVO:SVC ratio. No significant correlation with transmitral indices was noted. Among the parameters, LVO:SVC ratio had the highest specificity (0.83) and sensitivity (0.95) to detect a duct of ≥ 3 mm. The area under the curve was 0.95 (95% CI: 0.85–0.99), indicating a 95% probability that a randomly selected patient with LVO:SVC ratio ≥ 4 will have a ductal size ≥ 3 mm.
Conclusions: Significant correlations between ductal size and surrogate markers of pulmonary over circulation were noted. A combination of echocardiographic markers may help assess the magnitude of the haemodynamic impact of a patent ductus arteriosus.

1245: MARFAN SYNDROME IN AN EIGHT-YEAR-OLD BOY

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Background: Marfan syndrome is a systemic disorder of the connective tissue associated with mutations. Early diagnosis and advances in medical technology have improved the quality of life for people with this syndrome. It can be determined by multisystem disorder with manifestations typically involving the cardiovascular, skeletal and ocular systems. Rupture of the aorta was a serious complication of Marfan syndrome.
Objective: An eight-year-old boy weighing 27 kg, height 138 cm came to the hospital on 20 March 2009 for endocrinology counselling. The boy looked healthy. He was taller than expected for his genetic background. A craniofacial physician found a small chin and malar flattening. There was no chest deformity and in the extremities, a wrist and thumb sign. An ophtalmologist diagnosed simple ectopia lentis. Echocardiography revealed elongation of the aorta. He is the second child, and his father went blind about three years ago. This patient fulfills the Ghent criteria (three out of five criteria). He was prescribed a beta-blocker to prevent progression of the elongation of the aorta, followed by serial echocardiography regularly every six months.
Conclusions: This diagnosis of Marfan syndrome was done using clinical manifestations (Ghent criteria) without genetic testing. Medication with a beta-blocker and serial echocardiography was necessary to detect elongation of the aorta, which may cause spontaneous rupture of the aorta.

1250: CLINICAL COURSE AND PROGNOSIS OF HYPERTROPHIC CARDIOMYOPATHY IN EGYPTIAN CHILDREN

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Background: Hypertrophic cardiomyopathy (HCM) is an important cause of disability and death in patients of all ages. Egyptian children may differ from Western and Asian patients in the pattern of hypertrophic distribution, clinical manifestations and risk factors.
Objectives: The present work aimed to register the clinical characteristics and outcomes of our children with HCM over a period of seven years, and to determine whether reported adult risk factors for SCD are predictive of outcome in these affected children.
Methods: This was a retrospective study that reviewed the clinical data of 128 paediatric HCM patients. These data included personal and family history, physical examination, baseline laboratory measurements, ECG, Holter and echocardiographic results. Logistic regression analysis was done for detection of risk factor of death.
Results: Fifty-one out of 128 patients died during the period of the study. Extreme LVH [interventricular septal wall (IVS) thickness or posterior wall (PW) thickness *z*-score > 6, presence of sinus tachycardia and supraventricular tachycardia (SVT)] was an independent risk factor for prediction of death in patients with HCM.
Conclusions: In our Egyptian tertiary care centre, HCM has a relatively bad prognosis. Infants have a worse outcome than children presenting after the age of one year. A poorer prognosis in HCM is predicted by extreme LVH, presence of sinus tachycardia and SVT.

1251: INITIAL EXPERIENCE WITH CARDIAC RESYNCHRONISATION THERAPY IN PAEDIATRIC PATIENTS

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Introduction: Cardiac resynchronisation therapy (CRT) is an effective intervention in adults with heart failure who do not improve after optimisation of medical therapy. Experience in paediatrics is limited in terms of patient numbers and long-term follow up. We present our experience with two paediatric patients on CRT at our institution.
Case reports: Patient 1 was diagnosed with an ASD and a large VSD that was surgically corrected at one year of age. The surgery was complicated by complete heart block, necessitating the insertion of an epicardial pacemaker. Over the next three years he developed dilated cardiomyopathy (DCMO), which did not improve on optimal medical therapy. A CRT pacemaker system was inserted epicardially at the age of four years. He now displays improved exercise tolerance on a six-minute walk test, with improved LV function.
Patient 2 presented as a neonate with complete heart block. An epicardial pacemaker was inserted. Over the next five years she developed DCMO despite optimal medical therapy. A CRT pacemaker system was inserted epicardially at the age of five years. She now has improved symptoms and LV function.
Discussion: CRT has emerged as an effective treatment strategy for pacemaker-induced DCMO, as chronic RV stimulation can adversely influence LV function over time. The indications, procedures and outcomes for CRT in paediatrics are evolving. CRT has been shown to improve symptoms, exercise capacity and quality of life.
Conclusion: CRT was associated with improved outcomes in our two patients with pacemaker-induced DCMO.

1257: SYNDROMIC PHENOTYPES OF UGANDAN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: This study determined the occurrence and pattern of recognisable genetic syndromes in Ugandan children with congenital heart disease.
Methods: A medical geneticist, genetic counsellor and cardiologists from the Uganda Heart Institute (UHI) at Mulago Hospital, and Children's National Medical Centre in Washington, DC, evaluated children with congenital heart disease. Cardiac diagnoses were confirmed by echocardiography. Phenotypic assessment was performed and family history obtained.
Results: The 124 children with congenital heart disease were evaluated over a one-week period in February 2012. Mean age was 5.5 ± 5.5 years. The most common diagnoses were ventricular septal defect (n = 40), tetralogy of Fallot (n = 28), pulmonary stenosis (n = 12), atrial septal defect (n = 11), double-outlet right ventricle (n = 8), patient ductus arteriosus (n = 7), and truncus arteriosus (n = 6). By phenotypic evaluation, 26 (20%) of these children had a suspected genetic syndrome: 22q11 deletion (n = 10), Noonan (n = 5), Down (n = 4), Turner (n = 3), Kabuki (n = 2), Holt-Oram (n = 1) and CHARGE (n = 1). The majority of patients phenotypically positive for 22q11 deletion syndrome (9/10) had a conotruncal abnormality: tetralogy of Fallot (seven), D-transposition of the great arteries (one), or truncus ateriosus (one). The majority with a Noonan's syndrome phenotype (four/five) had pulmonary stenosis. An additional 15 patients (12%) had a pattern of dysmorphic features suggesting an unrecognisable syndrome. Five children (4%) had a family history of one or more first- or second-degree relatives with congenital heart disease.
Conclusions: This is the first phenotypic survey of CHD in an East African population. Our data suggest a substantial number of children with known congenital heart disease have underlying genetic abnormalities. Truncus arteriosus constitutes a larger-than-expected proportion of children with congenital heart disease. Coarctation of the aorta is under-represented. Genotypic studies are needed to evaluate the full spectrum of genetic variation seen in this population.

1259: GENETIC SUSCEPTIBILITY TO ENDOMYOCARDIAL FIBROSIS

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Background: Endomyocardial fibrosis (EMF) is the most common form of restrictive cardiomyopathy. Cases have been reported from tropical and subtropical regions around the equator and have repeatedly been shown to cluster both within families and within select ethnic groups. Eosinophilia is an independent risk factor for EMF, suggesting that both genetic and environmental factors play a role in disease development. This study was designed to investigate the genetic susceptibility to EMF, and possible links with eosinophilia. It sought associations between EMF and inherited alleles in the human leukocyte antigen (HLA) system, including antigens HLA-A, -B, -C, -DR, and -DQ. Alleles in the HLA-A1 region have previously been associated with predisposition to allergic and hypersensitivity reactions.
Methods: Fifty patients with EMF and 50 controls were recruited from the Uganda Heart Institute and the cardiology ward at Mulago Hospital. EMF was confirmed by echocardiography. Blood was obtained from peripheral venipuncture and sequence-based typing was used to identify HLA class I (HLA-A, -B, -C) and class II (DRB1 and DQB1) genes. Chi-square analysis was used to identify any difference in class I and class II HLA alleles between cases and controls.
Results: HLA-A*02:02 appeared more frequently at site A1 in the cases than in controls (29% in cases compared to 4% in controls).

In the light of this difference, we performed a secondary analysis distinguishing between those with the allele and those without. In this new analysis, there was a significant association ($p = 0.005$) between EMF and presence of the HLA-A*02:02 allele.
Conclusions: HLA-A*02:02 at site A1 was more commonly found in patients with EMF than in controls. Further investigations are needed to more fully understand the role of the HLA system in EMF development and the link between genetics and eosinophilia.

1260: TETHERED PRE-NORWOOD TRICUSPID VALVE IN HYPOPLASTIC LEFT HEART SYNDROME IS ASSOCIATED WITH MEDIUM-TERM TRICUSPID VALVE REGURGITATION

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Background: Tricuspid regurgitation (TR) is a risk factor for morbidity and mortality in hypoplastic left heart syndrome (HLHS). Our group previously described the association between HLHS medium-term TR and laterally displaced anterior papillary muscle (APM). This study aims to determine whether congenital position of the APM is an early risk factor for TR in HLHS.
Methods: Neonates with HLHS were prospectively assessed prior to stage-1 palliation (S1P) with two-dimensional (2DE) and three-dimensional echocardiograms (3DE). 2DE was used to assess TR, RV fractional area change, sphericity index and end-diastolic area. Neonates with moderate or greater TR prior to S1P were excluded. 3DE datasets were analysed off-line to extract spatial coordinates of TV annulus, leaflets and PM (TomTec Inc, Germany), with annulus size, leaflet areas, prolapse and tethering volumes calculated using MatLab-based software (MathWorks Inc, Mass).
Results: Forty neonates were assessed pre-S1P and at mean follow up of 23 ± 19 months. Patients were divided by degree of TR at follow up; 15 patients had moderate or greater TR (group A) whilw 25 had mild or less TR (group B). Group A had an increased risk of death or cardiac transplant (60 vs 28%, $p < 0.05$) and rate of TV surgery (33 vs 0%, $p < 0.01$). Group A and B had similar RV size, shape and function. No difference was found using 3DE assessment of the APM position. However, group A patients had increased TV tethered volume (0.69 vs 0.47 ml/m^2, $p < 0.05$), increased 3D tenting height ($p < 0.05$), with a trend for greater leaflet area (8.5 vs 7.2 cm^2/m^2; $p = 0.06$). No differences in annular area and TV prolapse were present

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1267: OBJECTIVE ESTIMATION OF LEFT VENTRICULAR FUNCTION IN CHILDREN WITH AORTIC INSUFFICIENCY ON EXERCISE STRESS TEST

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Method: LV function was evaluated in 56 children (31 boys and 25 girls) with congenital AI, NYHA 1–II. Median age was 14.9 years (10–18). Group 1 (36 children) had mild AI (P1/2t 485.04 ± 16.85 ms). Group 2 (20 children) had moderate AI (P1/2t 324.74 ± 39.41 ms). Echo and stress-echo were performed (Phillips HD11XE) to evaluate the LV in all patients. The stress agent was a step-test. Power was calculated by the formula: W (kg/min) = weight (kg) × height of step (m) × number of raises/min × 1.33 (1.33 = coefficient: 6 kg/min/kg on the step for 3 minutes). Thirty normal children were evaluated as controls. An echo-Doppler study recorded LV end-diastolic (EDD, EDV) and end-systolic (ESD, ESV) dimensions, volumes, ejection fraction (EF) pressure gradient on the aortic valve.
Results: Patients with mild congenital AI had no significant differences in LV parameters after the stress test compared to healthy children ($p > 0.05$): EDD (4.06 ± 0.13 sm), ESD (2.23 ± 0.06 sm), EDV (79.05 ± 3.95 ml), ESV (17.84 ± 1.19 ml) compared to the control group (4.15 ± 0.06 sm, 2.16 ± 0.04 sm, 77.22 ± 2.55 ml, 15.90 ± 0.78 ml). EF in group 1 (77.56 ± 1.09%) did not differ from that in healthy children (79.56 ± 0.63%) ($p > 0.05$). In children with moderate AI there were significantly higher LV parameters after the stress test: ESD (2.41 ± 0.11 sm), EDV (90.03 ± 4.13 ml), ESV (21.97 ± 1.96 ml) compared with the same indices in healthy children ($p < 0.05$). EF in this group (73.84 ± 2.18%) was lower than in the healthy children group ($p < 0.05$).
Conclusion: Estimation of LV function during stress-echo in patients with AI detected early signs of dysadaptation for developing heart failure. This is very important for children with moderate AI as an objective indication for surgical correction.

1271: VALVE SURGERY IN BRAZILIAN CHILDREN AND ADOLESCENTS WITH RHEUMATIC HEART DISEASE: TIME TO OPERATION AND ASSOCIATED FACTORS

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Background: Rheumatic heart disease (RHD) remains the most common acquired heart disease in children and adolescents worldwide, being responsible for a high rate of morbidity and mortality, often demanding valve surgery in acute or chronic phases of the disease.
Hypothesis: Valve surgery is associated with variables/factors from three distinct dimensions: sociodemographic, clinical, and echocardiographic in children and adolescents with RHD.
Methods: A longitudinal observational study of a hospital-based population was done, using the Kaplan–Meier method for time estimation and Cox regression model for hazard risk (HR) evaluation of associated variables. The cohort was three- to 20-year-old patients, registered between 1986 and 2006, and followed up until 2011. RHD was confirmed by Doppler echocardiography. The variables evaluated at the patient's first visit were grouped into three dimensions: sociodemographic (gender, age group, skin colour, residential region, decade of diagnosis); clinical (disease status, NYHA functional class, number of previous rheumatic episodes, secondary prophylaxis, infectious endocarditis, atrial fibrillation); and echocardiographic (valve lesion and severity, left atrium diameter, systolic left ventricle diameter, left ventricle function, pulmonary hypertension, rupture of mitral chordae). For the database, ACCESS 2000 was used. For statistical analysis, R program. Significance was defined as $p < 0.05$.
Results: Of the 348 patients (58% female), 39% underwent valve operations. Median age at registration was 12.5 years. Median follow-up time was 9.0 years (2–21 years). Median time until surgery was 22.3 years. Univariate analysis: all variables except residential region ($p > 0.5$) presented significance (HR > 1). Multivariate analysis: the final model included decade of diagnosis (HR = 1.36), NYHA functional class II–IV (HR = 1.97), number of prior episodes (HR = 1.72–2.15), endocarditis (HR = 2.01), valve lesion and severity (HR = 2.15–2.89), left atrium diameter ≥ 40 mm (HR = 2.67), systolic left ventricle diameter ≥ 45 mm (HR = 2.67) and rupture of mitral chordae (HR = 2.68).
Conclusions: Valve surgery was associated with sociodemographic, clinical and echocardiographic factors in Brazilian children and adolescents. Although most patients underwent surgery in first two years, long-term surveillance of this population was mandatory.

1273: ASSESSMENT OF SYSTEMIC ENDOTHELIAL FUNCTION IN CHILDREN AND ADOLESCENTS WITH PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary endothelial function is known to be affected in patients with pulmonary arterial hypertension (PAH). We hypothesised that PAH is a progressive panvasculopathy, affecting both pulmonary and systemic vascular beds, and that systemic endothelial dysfunction correlates with pulmonary vascular changes.
Methods: Systemic endothelial function was assessed by the vascular response to reactive hyperaemia and was recorded non-invasively by peripheral arterial tonometry (EndoPAT) under standardised conditions. Digital reactive hyperaemic index (RHI) was examined in 25 children and adolescents (mean age 21.1 ± 10.1 years; 14 females) with pulmonary arterial hypertension (IPAH, $n = 13$; PAH-CHD, $n = 12$; eight of them with Eisenmenger syndrome). Measurements were compared with functional data assessed by echocardiography and cardiac catheterisation.
Results: Mean tricuspid annular plane systolic excursion (TAPSE) was 20.9 ± 4.6 mm. Mean pulmonary artery pressure (MPAP) of all patients was 68 ± 27.4 mmHg, and ratio of pulmonary vascular resistance to systemic vascular resistance 1.0 ± 0.6. Vasodilator response to acetylcholine (mean pulmonary flow ratio) was 2.1 ± 2.2. Mean RHI was lower in IPAH (1.6 ± 0.6) and Eisenmenger patients (1.5 ± 0.4) compared to PAH-CHD without Eisenmenger patients (2.1 ± 0.7) ($p = 0.04$). RHI correlated with TAPSE ($p = 0.001$). There was no correlation between RHI and pulmonary endothelial function assessed by vasodilator response to acetylcholine.
Conclusions: Systemic endothelial function may be impaired in children and adolescents with IPAH and Eisenmenger syndrome and correlates with RV systolic function. However, according to our data there was no relation between systemic vascular changes and pulmonary endothelial function, therefore, different mechanisms may contribute to their pathogenesis and progression.

1279: CONTRIBUTION OF ATRIOVENTRICULAR PLANE DISPLACEMENT TO LEFT AND RIGHT VENTRICULAR STROKE VOLUME IN HEALTHY SUBJECTS AND PATIENTS WITH PULMONARY REGURGITATION

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Background: Atrio-ventricular plane displacement (AVPD) contributes to 60% of left ventricular (LV) stroke volume (SV) and 80% of right ventricular (RV) SV in healthy subjects. However, there are no data in patients with RV volume overload. The aim was to quantify the physiological effect of RV volume overload on the contribution of AVPD to ventricular pumping.
Methods: MRI was performed in 29 healthy subjects and 22 patients with moderate-to-severe pulmonary regurgitation (PR) due to surgically corrected tetralogy of Fallot or pulmonary stenosis (mean age 23.5 ± 13.1 years). LVSV and RVSV were calculated by delineation of the endocardium of both ventricles in diastole and systole. The longitudinal contribution to the SV was calculated for both ventricles using a previously described method, using AVPD and the epicardial area of the basal part of the ventricle.
Results: In the patient group, regurgitant fraction was 44 ± 10% and RV end-diastolic volumes (EDV) were increased (278 ± 84 ml) compared to LVEDV (154 ± 56 ml, $p < 0.0001$). The contribution of the AVPD to the LVSV did not differ between healthy subjects (59 ± 2%) and patients with PR (56 ± 13%, $p = 0.244$). However, the contribution of AVPD to the RVSV was significantly lower in patients with PR (46 ± 8%) compared to healthy subjects (78 ± 2%, $p < 0.0001$).
Conclusion: Patients with PR had normal longitudinal contribution to LVSV, but decreased longitudinal contribution to RV pumping compared to healthy subjects. This was caused by decreased AVPD in volume overloaded RVs, which only in part was compensated for by an increased RV area. Instead, increased septal movement towards the RV and radial contraction of the free wall contributed to the major part of the RVSV in patients with volume overload due to PR.

1280: CARDIAC PACING IN PATIENTS WITH 'YE OLD' FONTAN-KREUTZER ATRIOPULMONARY CONNECTION

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Background: Atrial arrhythmias are frequent and represent an important complication in the long-term follow up of patients with atrio-pulmonary connection. The loss of A-V synchrony is clearly associated with a worsening of the functional status and failure of the univentricular circulation.
Methods: From 1975 to December 2011, 75 patients with atrio-pulmonary connection were followed up at our Institution; 48 patients (64%) developed arrhythmias. Eleven patients (range 5–41 years) needed a pacemaker because of sinus node disfunction (five patients), A-V block (three) and atrial tachyarrhythmias (three).
Results: A DDDR pacemaker was implanted in five patients, VVIR pacemakers in four and an AAI pacemaker was implanted in two patients. Seven devices were endovenously placed and four were epicardial. The measured mean parameters at implantation in the atrium and the ventricle, respectively, were: threshold 1.18 and 0.925 mV at 0.5 ms; P wave 1.64 and R wave 11.18; impedance 615 and 725 Ohms. For ventricular stimulation, four intravenous leads were placed in the coronary sinus, three bipolar contact epicardial leads and two unipolar contact epicardial leads. Regarding the atrial leads, five were endocavitary active fixation leads and two bipolar contact epicardial leads. Mean follow up was 7.7 years (9 months to 23.7 years). There has been one late death in a conversion procedure to TCPC. Functional status has improved in all the remaining cases.
Conclusions: The use of the appropriate devices to control frequent atrial arrhythmias in these patients improved the functional class, thus delaying the need for a conversion procedure or heart transplant. It is mandatory to use adequate techniques to achieve the goal of maintaining A-V synchrony in these patients.

1284: MITRAL VALVE REPAIR IN CHILDREN WITH RHEUMATIC HEART DISEASE

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Introduction: Rheumatic heart disease (RHD) is common in rural India. The mitral valve is the most common valve involved in RHD. Children with rheumatic mitral valve disease present with either mitral regurgitation or stenosis. Mitral valve repair and its results have been studied at our centre.
Methods: This was a retrograde single-centre study from March 2009 to June 2012. The total number of patients was 90, with 41 male and 49 female, ranging in age from from five to 18 years. Mean age was 10.6 years; 79 patients had mitral regurgitation while 11 had mitral stenosis; 76 patients underwent mitral valve repair while 14 required mitral replacement. Mitral repair: 11 patient had chordal shortening, 17 had neochordal formation, and 25 required thining of the leaflet. Suture annuloplasty was done in 32 patients while 35 had ring annu-

loplasty. All patients underwent epicardial or transoesophageal echo postoperatively. Mean CPB time was 130 min. Mean aortic cross clamp time was 78 min. Five patients had grade II MR on TEE, which was expected as the left atrial pressure was low. No early or late mortalities were found. Mean hospital stay 7.4 days. On discharge eight patients had mild MR while five had grade II MR. We have complete follow up for two years. Three patients required mitral valve replacement after two years, while three developed new grade III MR.
Conclusion: Mitral valve repair is a good option for children with rheumatic mitral valve disease. With modification of surgical techniques, most mitral valves can be repaired, with good surgical results.

1291: RETROSPECTIVE EVALUATION OF PATIENTS WITH KAWASAKI DISEASE

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Background: The diagnosis of Kawasaki disease requires the presence of five days of fever and at least four of the five principal clinical features, including bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash and cervical lymphadenopathy. The most serious complication is coronary artery aneurysm.
Methods: In this study, 18 patients diagnosed with Kawasaki disease in the Department of Paediatrics, Eskişehir Osmangazi University Hospital between 1996 and 2012 were evaluated.
Results: At admission, the mean age of the patients was 40.6 ± 22.5 months (8–90 months) and the duration of fever was 8.8 ± 4.4 days (3–20). All patients had high fever and the second most commonly seen finding was changes in lips and oral mucosa (94.4%). The other findings were changes in the extremities (83.3%), rash (72.2%), non-exudative conjunctivitis (55.5%) and cervical lymphadenopathy (27.7%). Twelve (66.7%) of the cases were diagnosed with complete and six (33.3%) with incomplete Kawasaki disease. Coronary artery dilatation was observed in sic patients (33.3%), mitral insufficiency in four (22.2%), pericardial effusion in one (5.6%), and increased end-diastolic diameter of the left ventricle in one (5.6%) patient. Sixteen patients were given intravenous immunoglobulin (IVIG) and acetylsalicylic acid (ASA). Two patients, one complete and one incomplete, could be given only ASA. Coronary artery pathology was not seen in these two patients. One patient was given a second dose of IVIG because of the persistent fever. During follow up, coronary artery aneurysm was observed in four (25%) and coronary artery stenosis in one (6%) of the patients given IVIG.
Conclusion: Early diagnosis and treatment of Kawasaki disease is important to prevent coronary artery complications. It is essential to suspect Kawasaki disease and to perform echocardiographic evaluation for cardiac involvement in patients with fever persisting longer than five days.

1293: EVALUATION OF VENTRICULAR SEPTAL DEFECT WITH REAL-TIME THREE-DIMENSIONAL ECHOCARDIOGRAPHY

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Background: As a dynamic three-dimensional structure, the heart can be hard to understand in conventional two-dimensional (2D) plane imaging. Hence, real-time 3D (RT-3D) imaging can be used for assessing cardiac structures and intracardiac lesions. This study assessed a RT-3D platform for delineating ventricular septal defect (VSD) geometric characteristics.
Methods: Nine patients with VSD (four female and five male) enrolled in this study. The VSD types included three perimembranous cases, three inlet cases, and three outlet cases. The Philips IE 33 system was used to acquire both the 2D and the RT-3D echocardiography images. These patients' lesions were subsequently diagnosed and analysed for clinical comparison.
Results: The 2D VSD diameter was 6.78 ± 1.55 mm (range: 4.2–10.1 mm). The RT-3D mean maximum and minimum VSD diameters were 7.27 ± 1.74 and 6.37 ± 1.66 mm, respectively. All patients underwent VSD surgical repair; the respective intra-operative maximum and minimum VSD diameters were 7.29 ± 1.83 mm (range: 5.0–11.5 mm) and 6.17 ± 1.98 mm (range: 3.0–10 mm). The correlation coefficient between 2D and RT-3D mean maximum diameters was $r = 0.966$, between surgical diameters it was $r = 0.967$, while it was $r = 0.945$ between the RT-3D mean maximum and intra-operative diameters.
Conclusion: 'Real-time' 3D echocardiography can be a good diagnostic tool to clearly delineate the size, position and size of a VSD. In the future, it may pre-operatively assess VSD and device closures.

1301: COST EFFECTIVENESS OF ECHO-BASED RHEUMATIC HEART DISEASE SCREENING

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Background: Rheumatic heart disease (RHD) is a leading cause of mortality and morbidity in young citizens of low- and middle-per capita income settings. Echocardiography-based screening approaches dramatically expand the number of children identified at risk of progressive RHD. We developed a Markov model to project the cost-effectiveness of this nascent screening approach.
Methods: A Markov model was constructed comparing a No-Screen to Echo-Screen approach. Both scenarios commit staff to provide secondary prophylaxis, prophylaxis transportation, consumables, heart failure medications, anticoagulation and monitoring, general practitioner and/or cardiology follow up appropriate to the severity of RHD, valve replacement in a fraction of compromised patients including operative costs, consumables, valve, and post-operative stay, and severity-appropriate lost wages to patient/parent. The screen scenario posits technician-driven limited screening echo followed by detailed cardiology evaluation in screen-positive children. The screen scenario entailed one-time costs for staff, transportation, echo machine, and a single day's lost wages. RHD-related states were categorised as well (utility weight 0.9), dead (utility 0), silent RHD defined as visible on echocardiography but silent by auscultation (utility 0.75), auscultation-audible RHD (utility 0.75), previously audible but now resolved RHD (utility 0.75), RHD with functional compromise in activities of daily living (utility 0.58), and RHD post valve replacement (utility 0.58). Sensitivity analyses varied echo accuracy, surgical availability, disease prevalence, and screening-associated costs. Results are denominated in Australian dollars and future utilities and costs were discounted by 3.5% per year.
Results: The Echo-Screen strategy may be cost-effective, and under certain circumstances, dominated the No-Screen strategy. This result appears insensitive to screening costs, surgical availability, and echo accuracy, but did appear to be affected by RHD prevalence.
Conclusions: Contrary to our expectation, a two-stage echo screening approach in a health system committed to providing secondary prophylaxis may be robustly superior under a variety of circumstances.

1302: THE NEW GHENT CRITERIA FOR MARFAN SYNDROME: CLINICAL IMPLICATIONS

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Background: Marfan syndrome (MFS) is a connective tissue disorder with major features in cardiovascular, ocular and skeletal systems. The diagnostic criteria were revised in 2010 and more weight was given to the aortic root dilatation, ectopia lentis and genetic study.
Methods: This was a retrospective study of every patient evaluated for MFS in our centre over the last 15 years. We aimed to analyse the practical clinical implications of the revised nosology in a paediatric population.
Results: The study included 18 patients (nine girsl), with the first evaluation in paediatric cardiology at the mean age of 6.7 years (6 days –16.8 years). There was a positive family history in four cases. The diagnosis of MFS by the classical Ghent nosology was established in 12 patients; eight patients at the first consultation and in the rest, 2.8 years later, confirming that MFS has an evolving phenotype. With the revised criteria, the diagnosis of MFS remained in 11/12 patients and two new patients were diagnosed. The diagnosis of the five rejected patients were: two as MASS phenotype (in the absence of aortic dilatation), two as mitral valve prolapse syndrome and one as potential MFS in patients under 18 years. The diagnosis was not confirmed because the *z*-score of the aortic root was < 2 or 3.
Conclusions: The revised Marfan nosology led to a different diagnosis in three cases; one of the MFS patients was reclassified as MASS; conversely, two were reclassified as MFS in the presence of aortic dilatation. The diagnosis of MFS was rejected mostly because of the absence of aortic root dilatation defined as *z*-score ≥ 2 or 3. All patients suspected of MFS maintain follow up in paediatric cardiology, considering the possibility that they may develop the classic MFS with time.

1308: THE RELATIONSHIP OF A LEFT ATRIAL MYXOMA WITH THE MITRAL VALVE IS BEST EVALUATED BY 3D ECHOCARDIOGRAPHY

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Background: Seventy-five per cent of myxomas are located in the left atrium, the majority originating from the interatrial septum. They can come in contact with vital structures. We used intra-operative 3D TEE to reassess a myxoma of the left atrium just before surgery.
Method and Results: A 51-year-old woman consulted the Aswan Heart Centre for progressive mild dyspnoea. Her physical findings were compatible with bronchial reactivity without signs of cardiac pathology. Her chest X-ray and ECG were normal. The TTE showed a sessile mass of 13 × 18 mm compatible with a myxoma attached to the left side of the inter-atrial septum without other anomalies. She was brought to the operating room for elective resection of the myxoma. A real-time 3D TEE was performed pre-operatively, followed by a full-volume acquisition with a systematic cropping of the 3D TEE dataset in multiple plans. Reconstructions showed a tumour measuring 14 × 19 mm attached to the lower part of the interatrial septum in close contact with the anterior leaflet of the mitral valve as well as the mitral annulus. The space between the mitral annulus and the myxoma was estimated at 5 mm. Transmitral blood flow was normal. There were echolucent areas in the tumour compatible with necrosis. After cardiopulmonary bypass and cardiac arrest with blood cardioplegia, a right atriotomy was performed and the myxoma completely resected without traumatising the mitral valve. The septum was then reconstructed with an autologous pericardial patch. The postoperative course was simple and the patient discharged on day six.
Conclusion: 3D echocardiography facilitated description of a left atrial myxoma better than 2D. It provided a better understanding of the relationships with adjacent structures, especially regarding the space between the tumour and vital structures, and should be considered as first-line examination in order to help plan surgery and avoid complications.

1314: PERSISTENT RETINAL VASCULAR CHANGES IN KAWASAKI SYNDROME: POTENTIAL ROLE IN CORONARY RISK STRATIFICATION

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Background: We hypothesised that the inflammatory effects of Kawasaki syndrome (KS) causes longstanding changes in the retinal vasculature akin to traditional coronary risk factors such as diabetes mellitus and hypertension.
Methods: We compared the retinal vascular dimensions of a cohort with a history of KS to matched controls. All subjects underwent high-resolution digital retinal photography in which the diameters of all arterioles and venules coursing through a specified area one-half to one disc diameter from the optic disc were measured with a computer program (IVAN), according to a published standardised protocol. Central retinal arteriolar equivalent (CRAE), central retinal venular equivalent (CRVE), and arteriole-to-venule ratio (AVR) were calculated for each retinal photo.
Results: Thirty-two subjects with a history of KS and 138 controls were examined. KS subjects had a mean CRAE of 149.77 µm, a mean CRVE of 211.61 while controls had a mean CRAE of 147.25 µm (p = 0.300), and a mean CRVE of 221.11(p < 0. 001). Cases were matched for age, gender, ethnicity, body surface area and also controlled for calibre of neighboring vessels.
Conclusions: KS results in independent, persistent and substantial narrowing of the retinal venules. Such an effect may be related to endothelial dysfunction and serve as a potential marker for incipient coronary vasculopathy.

1320: TWO CASES OF POST-FONTAN WARFARIN-INDUCED TRACHEOBRONCHIAL CARTILAGE CALCIFICATION

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Background: This study identifies tracheobronchial cartilage calcification in children with congenital heart disease. Calcification of the tracheobronchial airways has previously been found to be more common in adults taking warfarin, and has been described in children who are receiving warfarin following mitral valve replacement.
Methods: A nine-year-old female who had undergone a Fontan repair six years previously underwent a cardiac CT to further image her pulmonary arteries. An incidental finding of extensive tracheobronchial cartilage calcification was noted. We then conducted a retrospective review of all the paediatric Fontan patients who had undergone cardiac CT to look for calcification of the tracheobronchial cartilage.
Results: Ten paediatric Fontan patients had undergone cardiac CT scan. Two patients with extensive calcification of the tracheobronchial airways were identified. A nine-year-old female with a cardiac diagnosis of hypoplastic left heart syndrome had undergone a staged repair with Fontan completion at age three years. A 16-year-old male with a cardiac diagnosis of tricuspid atresia underwent staged repair and Fontan completion at age 3.5 years. They had received continuous warfarin for six and 13 years, respectively. Other common causes of airway calcification were excluded.
Conclusions: We describe warfarin-induced tracheobronchial calcification in patients following the Fontan procedure. This finding has potential implications for airway growth and vascular calcification.

1322: HIV+ MALAWIAN CHILDREN WITH DECREASED EXERCISE PERFORMANCE AND NORMAL CARDIAC STRAIN

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Background: Traditional measures of cardiac function are often normal in HIV+ children on antiretroviral therapy (ART). Previously, strain analysis has shown subtle cardiac dysfunction in HIV+ adolescents and young adults. We hypothesised that, using speckle tracking, we would detect subtle cardiac dysfunction in this larger cohort of Malawian children.
Methods: In this prospective observational study of children aged 4–18, we recruited 241 HIV+ youth, and 95 HIV– controls. An echocardiogram and six-minute walk test (6MWT) were performed. CD_4 count and HIV viral load were performed on HIV+ subjects. Ejection fraction, strain and strain rate were measured. Within the HIV+ group, analysis of covariance was implemented to compare means of cardiac function and performance on the 6MWT in HIV+ children on and off ART as well as HIV– children, controlling for age, gender and BMI. Multiple linear regression models evaluated whether cardiac function was related to CD_4 count or log viral load. *T*-tests based on linear contrasts were used to compare function measures between those with detectable and undetectable viral loads.
Results: The HIV– subjects performed better on the 6MWT (p = 0.001). HIV+ patients on ART averaged 470 m, HIV+ patients off ART averaged 460 m, and HIV– patients averaged 500 m. Cardiac function and strain were normal for all groups. EF and strain were not related to CD_4 count or log viral load. Among HIV+ participants, children with undetectable viral loads had more negative (better) global circumferential strain (GCS) compared to those with detectable viral loads (p = 0.02).
Conclusions: HIV+ children had decreased exercise performance but did not exhibit decreased strain compared to controls. Detectable viral loads were associated with worsened GCS, suggesting that viral suppression may delay the onset of cardiac dysfunction. Although these children do not currently exhibit clinical cardiac dysfunction, long-term evaluation is warranted.

1324: HAND-HELD ECHOCARDIOGRAPHY: INITIAL EXPERIENCE IN USE FOR ASSESSMENT OF RHEUMATIC HEART DISEASE IN INDIGENOUS CHILDREN IN THE KIMBERLEY REGION OF WESTERN AUSTRALIA

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Background: ARF and RHD remain a major problem in indigenous children in Western Australia (WA) particularly in the remote communities in the Kimberley. It is difficult to triage the care of children presenting with signs of ARF without some echocardiographic assessment. This is difficult in WA as the geography and population distribution means there is very limited access to any paediatric echo skills in remote areas. With the recent introduction of hand-held echocardiographic equipment (HHE), it was decided to trial the use of such technology in the Kimberley.
Methods: An HHE was taken to outreach clinics in the Kimberley over a six-week period and over 30 patients with RHD and a number with signs suggestive of ARF had an echo study performed using HHE. This included 2D echo images in long-axis, short-axis and four-chamber views and colour Doppler images from the same views. This was followed by a more formal complete study using standard portable echo equipment (SPE). The images between the studies were compared to assess whether there was good correlation between the degree of mitral and/or aortic regurgitation.
Results: There was very good correlation between the HHE and SPE studies. It was possible using HHE to detect even trivial to mild valvar regurgitation. Chamber sizes and significant valvar morphological changes could be reasonably defined, although this was more limited than in the SPE studies.
Conclusions: This initial experience using HHE has been encouraging and warrants further scientific exploration. HHE could be a useful tool to train non-cardiology staff in remote WA to triage management of patients with possible ARF, in consultation with paediatric cardiologists. HHE may be useful for quick screens of patients with known RHD to select patients for more complete echo studies.

1325: EPIDEMIOLOGY OF ACUTE RHEUMATIC FEVER IN MANITOBA, CANADA: A 10-YEAR RETROSPECTIVE STUDY

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Objective: The aims and objectives of this study were to determine the incidence of acute rheumatic fever (ARF) between the first-nation (FN) and non-first nation (n-FN) children in the province of Manitoba, Canada.
Methods: This was a retrospective study conducted at the Children's Hospital of Winnipeg and Variety Heart Centre, Winnipeg, MB, Canada. Children with a discharge diagnosis of ARF were identified through the Children's Hospital of Winnipeg medical records and the Electronic Echocardiography data based at the Variety Heart Centre. The diagnosis of ARF was based on the revised Jones criteria.
Results: Over a 10-year study period (2000–2010), a total of 61 patients meeting the Jones criteria for the diagnosis of ARF were included in this study. The overall annual incidence rate of ARF in the province of Manitoba was 1.613/100 000 with 4.748/100 000 for FN and 0.618/100 000 for the n-FN children ($p < 0.001$). The overall incidence of ARF among girls was 1.562/100 000 (FN girls 4.984/100 000, n-FN girls 0.475/100 000, $p < 0.001$). Overall incidence of ARF among boys was 1.661/100 000 (FN boys: 4.522/100 000, n-FN: 0.754/100 000, $p < 0.001$).

1327: ACUTE EFFECT OF INHALED ILOPROST IN CHILDREN WITH PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE

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Objective: To evaluate the safety and efficacy of inhaled iloprost in children with pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) using measurements of haemodynamic parameters.
Methods: The study selected children hospitalised with severe PAH associated with systemic-to-pulmonary shunt-type CHD in the Department of Paediatric Cardiology of Beijing Anzhen Hospital between June 2007 and October 2011. All of the children underwent left- and right-sided cardiac catheterisation, during which iloprost was administered by atomisation inhalation. Changes in haemodynamic parameters before and after iloprost administration were recorded. Adverse events were also recorded.
Results: In total, 89 children with severe PAH associated with systemic-to-pulmonary shunt-type CHD (mean age 10.8 ± 4.7 years) were enrolled. Following inhalation of iloprost, the mean pulmonary arterial pressure decreased from 79 ± 11 to 74 ± 12 mmHg ($p < 0.01$), while the pulmonary-to-systemic blood flow ratio (Qp/Qs) increased from 1.15 ± 0.45 to 1.71 ± 1.36 ($p < 0.01$). Pulmonary vascular

resistance index (PVRI) decreased from 19.73 ± 9.31 Wood U/m² prior to inhalation to 15.49 ± 8.87 Wood U/m² following inhalation ($p < 0.01$), but there was no significant change in systemic blood pressure (prior to inhalation 85 ± 13 mmHg, following inhalation 83 ± 11 mmHg, $p > 0.05$).
Conclusion: Inhalation of iloprost during cardiac catheterisation in children with severe PAH associated with systemic-to-pulmonary shunt-type CHD can lower pulmonary vascular resistance, while causing no significant change in systemic blood pressure and without producing any obvious adverse reactions.

1328: OUTCOME OF BOSENTAN TREATMENT IN PAEDIATRIC PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE

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Objective: To describe the outcome of paediatric patients with pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) treated with bosentan, which is known to be safe and effective in patients with idiopathic pulmonary hypertension (IPAH).
Methods: In this monocentre, open-label, uncontrolled, observational study, 23 patients with PAH associated with CHD were treated with bosentan, The mean age was 9.12 ± 3.6 years (range 2.1–14.7) and they were treated for a mean of 13.3 ± 7.5 months (range 6–31). Six-minute walking test, percutaneous blood oxygen saturation (SpO_2%), New York Heart Association functional class (NYHA) and cardiac catheterisation data were compared before and after bosentan treatment.
Results: After treatment, 6MWT ($n = 17$) improved from 458 ± 16 to 496 ± 69 m ($p = 0.035$). SpO_2% increased from 89 ± 5 to 91 ± 5% ($p = 0.009$). In addition, the NYHA class improved. One patient in NYHA class IV moved to class III, and one in class III moved to class II, others in class II were stable ($p = 0.001$). Twelve patients had cardiac catheterisation before and after bosentan treatment, Qp/Qs increased from 0.97 ± 0.40 to 1.16 ± 0.40, PVRI decreased from 20.8 ± 8.8 to 18.1 ± 7.7 Wood unit/m², but the changes had no statistical significance. Bosentan was well tolerated by all patients, except one patient had temporary lower gastrointestinal tract bleeding.
Conclusion: Bosentan was safe and effective. Bosentan caused significant improvements in 6MWT SpO_2 and NYHA functional class and improvements in Qp/Qs and PVRI in children with PAH associated with CHD.

1331: SUBVALVAR ANEURYSMS IN CHILDREN AT A SOUTHERN AFRICAN TERTIARY CARE CENTRE

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Introduction: Subvalvar aneurysms, which may be subaortic or submitral, were described originally in young Africans in the 1960s, but later reports have included other race groups. Proposed aetiologies include infections, particularly tuberculosis, post traumatic, post surgery and ischaemia. An underlying congenital or developmental weakness between the muscular ventricular wall and the fibrous valve annulus is postulated. Complications such as rupture of the aneurysm, coronary artery compression, thromboembolism, and sudden death support the need for surgical intervention.
Methods: Children with subvalvar aneurysms were sourced from a paediatric cardiology computerised database compiled over a 20-year period at a southern African tertiary care centre.
Results: Nine children with an average age of 8.8 years (range 1–16) were diagnosed with submitral aneurysms using echocardiography. The male-to-female ratio was 1:2 and all were black. Five presented in heart failure and with varying degrees of mitral valve incompetence, one with an incidental murmur, one with syncope, one with a pneumonia and one with abdominal TB and human immunodeficiency virus infection. Two patients were found to have additional left ventricular cavity aneurysms and two aneurysms were found at surgery to have ruptured into the left atrium. Four patients had positive Mantoux tests and one was confirmed to have myocarditis and non-compaction of the myocardium on MRI scan. Surgery was undertaken in six patients. Histological examination of operative specimens showed peri-cardiac tuberculosis in two, features suggestive of rheumatic fever in one, a false aneurysm in one, and non-specific changes in the remaining two. Two deaths were recorded. One patient died suddenly pre-operatively and one immediately post-operatively.
Conclusion: Subvalvar aneurysms are a rare cause of mitral regurgitation and heart failure in children. Diagnosis is readily confirmed on echocardiography. Tuberculosis is a frequent association, but the aetiology remains inconclusive. There may be an underlying congenital predisposition.

1340: CONGENITAL CENTRAL HYPOVENTILATION SYNDROME (UNDINE SYNDROME) WITH RECURRENT HYPERCAPNIA AND HYPOXAEMIA IS LIKELY TO ACT AS ENDOTHELIAL PRECONDITIONING

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Introduction: Undine syndrome (US) is a rare disease with severely impaired central autonomic control of the breathing and dysfunction of the autonomous nervous system. The incidence is estimated to be one in of 200 000 livebirths. Due to reccurent hypercapnia and hypoxia we hypothesised that these patients have higher risk for pulmonary arterial hypertension (PHT) and long-term systemic vascular dysfunction. We examined seven patients with US at baseline and high altitude with regard to pulmonary artery pressure and systemic vascular disease and compared them with six age- and gender-matched subjects.
Methods: Seven patients with US (five female, two male, mean age 19 ± 3 years) and six age- and gender-matched healthy controls (three female, three male, mean age 20 ± 2 years) were examined at 550 m and at 3 883 m above sea level with echocardiography (to measure pulmonary artery pressure). Vascular function was measured at 550 m by flow-mediated dilatation with and without oxygen.
Results: All US patients had mild-to-moderate PHT at 550 m above sea level. PHT was more pronounced in males than in females (41 ± 5 vs 27.4 ± 3 mmHg) while no PHT was found in the controls. All US patients had high-normal systemic vascular function while controls had normal systemic vascular function. At 3 883 m above sea level, all US subjects showed only mild increase in PHT with regard to baseline while controls developped moderate-to-severe pulmonary hypertension (38 ± 7 vs 55 ± 17 mmHg).
Conclusion: Despite recurrent hypercapnia, US patients showed high-normal vascular function. We speculate that endothelial preconditioning (recurrent hypercapnia) could be a stimulus. Until recently, recurrent hypercapnia was believed to be the most harmful for endothelial functional properties. The presence of PHT at 550 m above sea level was not significantly aggravated by high altitude in comparison with healthy controls. This underlines the hypothesis of endothelial preconditioning and identifies environmental hypoxia as a key trigger for PHT in these patients.

1341: RHEUMATIC HEART DISEASE IN A TERTIARY HOSPITAL IN MALAYSIA

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Background: Acute rheumatic fever (ARF) and chronic rheumatic heart disease (CRHD) remain one of the leading causes of acquired heart disease in children, especially in developing countries. Malaysia, being a developing country, should share the same disease burden. A literature search revealed several articles on rheumatic heart disease from Malaysia. Two retrospective reviews in a single centre reported 42 patients over four years and 313 patients over 30 years.
Methods: We conducted a retrospective review of the records of all patients with a diagnosis of ARF or CRHD in our hospital from July 2011 to July 2012.
Results: There was a total of 20 patients. The male-to-female ratio was 1.5:1. The racial breakdown was 18 Malay, one Chinese and one Indian. A total of 12 patients had ARF, one patient had Sydenham chorea with mild mitral regurgitation on echocardiographic examination, and seven patients had CRHD. Of the 12 patients with ARF, two were under five years old, seven were five to nine years and three were over 10 years old. The patient with Sydenham chorea was a Malay boy of 9.5 years. Of the 12 patients with ARF, six had their first attack of ARF with carditis, five had acute-on-chronic rheumatic heart disease, and one patient had ARF with no carditis. Of the 11 patients who presented with acute rheumatic carditis, seven (63.3%) were in NYHA functional class I or II, one (9.1%) in class III and three (27.3%) in class IV. One patient required intubation and ventilation. All 11 patients had mitral valve involvement, five with severe mitral regurgitation. Eight patients also had aortic valve involvement, two with severe aortic regurgitation on echocardiographic examination.
Conclusions: Rheumatic heart disease is not uncommon in Malaysia. A quarter of patients with acute rheumatic carditis presented with severe congestive cardiac failure.

1342: POOR EFFICACY OF DRUG THERAPY FOR PDA CLOSURE IN INDIAN PRETERM BABIES

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Background: Ibuprofen is being used as an alternative to indomethacin in the medical management of PDA. Due to a paucity of data on the efficacy of these drugs for PDA closure in India, a study was designed to assess and compare their efficacy for PDA closure in preterm Indian neonates.
Methods: A prospective, randomised, controlled study on clinically diagnosed and echocardiographically confirmed haemodynamically significant PDA in preterm neonates was designed. Patients were randomly assigned to receive either oral ibuprofen at a dosage of 10, 5, 5 mg/kg every 24 hours or three doses of oral indomethacin (0.20–0.25 mg per kg 24 hourly) starting on the third day of life or when diagnosed. A second course of ibuprofen/indomethacin was given if the PDA failed to close within 48 hours of the first course. Surgical ligation was considered if the PDA did not close after two courses of treatment. Patients were monitored for complications such as oliguria, bleeding, NEC, IVH, oxygen dependency and gastrointestinal side effects.
Results: The baseline characteristics were comparable in both groups. Of the 83 children with PDA, 57.8% received oral ibuprofen and 42.1% received oral indomethacin. The overall closure rate of PDA was 60 and 65.7% in the ibuprofen and indomethacin groups, respectively, however in babies with postnatal age three to five days, the closure rate was 69.2 and 100%, respectively. Complications were similar in both the groups.
Conclusion: The efficacy of both drugs was similar. However, the overall closure as well as the closure in the subset of patients with postnatal age three to five days in the ibuprofen group was much lower than reported in the international literature. Cytochrome P_{450} enzyme causes increased plasma clearance of the drug, which is negligible at birth and increases with postnatal age. Poor closure in our study could have been because of genetic differences in the in the Indian population affecting the pharmacokinetics of the drugs.

1343: LONG-TERM FOLLOW UP (40 YEARS) OF PATIENTS POST NEONATAL INTERVENTION FOR CONGENITAL CRITICAL AORTIC STENOSIS AT A SINGLE CENTRE

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Background: Survival with congenital critical aortic valve stenosis (CAS) can be successfully achieved by surgical or catheter interventions. Long-term outcome and follow up remains uncertain.
Methods and Results: A 40-year (1970–2010) review was carried out of 96 patients requiring intervention for neonatal (< 30 days) CAS. Surgery (SX) was undertaken in 61 patients and balloon dilation (BD) in 35 patients. Before 2000, SX was more common (56/63 cases), while BD was favoured after 2000 (28/33 cases). Follow-up data were available for 88 patients. There were 29 (32.9%) reported deaths, early death (< 90 days) accounted for 22 (25%) patients. Early death accounted for four of the five deaths since 2000. Twenty (22.7%) patients died without any further intervention, one patient had a heart transplant and another had a single-ventricle palliation.
Discussion: Overall free survival rate at five, 10 and 20 years was 73, 73 and 61%, respectively. Of the 57 remaining survivors, 52 (87%) had at least one re-intervention, with further re-interventions required in 21 (36.8%) patients. There were 33 aortic valve replacements (AVR) in 32 patients, 31 (35.2%) patients had a Ross or Ross-Konno procedure. Freedom from AVR at five, 10 and 20 years was 78, 63, and 44%, respectively. The median time interval between initial procedure and AVR for the BD group was 1.32 years (mean 3.9 ± 4.7, range 1 day – 14.4 years). This was shorter than for the SX patient group: 8.9 years (mean 7.24 ± 5.1, range 1 day – 16.5 years). For patients presenting for the initial procedure since 2000, AVR was undertaken in 14/34, with a median time to AVR of 1.3 years.
Conclusions: This study highlights the acute and long-term mortality associated with neonatal CAS. Catheter-based intervention has become more common but is still associated with early mortality. Re-intervention is highly likely in survivors, over 50% of survivors requiring surgical AVR, and a trend towards earlier surgical AVR at our institution in recent years.

1344: ASSOCIATION OF MODIFIED BLALOCK–TAUSSIG SHUNT AND SCOLIOSIS IN CHILDREN WITH CONGENITAL HEART DISEASE

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Background: Previous literature reviews have shown congenital heart disease highly related to scoliosis. The operative methods such as median sternostomy and lateral thoracotomy were predisposed to the development of scoliosis. Nowadays, long-term follow up for these patients is possible with the improved survival rate of congenital heart disease patients. The role of Blalock–Taussig shunt in scoliosis has not yet been reported as a risk factor in these patients.
Methods: A retrospective review of spinal deformity in congenital heart disease patients who received Blalock–Taussig shunt, open-heart surgery or lateral thoracotomy was performed in the National Cheng Kung University Hospital from December 1989 to December 2005. Serial chest roentgenograms were taken before and after

the operation in 221 patients, to assess for spinal deformity. The curvature of the spinal deformity was defined by Cobb's angle. Comparison of the scoliosis was made between the operative methods and type of congenital heart disease. Follow up was done up to five years or longer.
Results: A total of 45 patients had scoliosis, with curvature greater than 10 degrees. Most of them (93.3%) were cyanotic heart disease patients. The patients who received Blalock–Taussig shunts were relatively high if compared to median sternostomy and lateral thoracotomy. There was a clinically significant correlation ($p < 0.05$).
Conclusions: Blalock–Taussig shunt may be one of the risk factor for scoliosis in patients with congenital heart disease. The prevalence of scoliosis increased in patients with cyanotic heart disease and treated with modified Blalock–Taussig shunt.

1352: ECHOCARDIOGRAPHIC ANALYSIS BY 3D, TISSUE DOPPLER, AFI, STRAIN, SR OF THE RIGHT AND LEFT VENTRICULAR DYSFUNCTION IN SURGICALLY REPAIRED PATIENTS WITH TETRALOGY OF FALLOT

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Objectives: Our study focuses on echocardiographic assessment of the right and left ventricular systolic function in response to volume overload resulting from pulmonary regurgitation in surgically treated patients with tetralogy of Fallot.
Methods and results: We included 63 patients with severe pulmonary regurgitation after surgical correction of tetralogy of Fallot. Automatic function image (AFI) by two-dimensional speckle tracking, left ventricular longitudinal strain and strain rate in right (RV) and left (LV) ventricle were recorded. X age: 14 ± 4 y; surgical correction was done at X:2.7 ± 1 y; follow-up X:11.8 ± 6 y, dividing them into 3 groups: 1) RV < 100 ml/m^2, 2) RV 100-120 ml/m^2, 3) RV:> 120 ml/m^2 The first group showed a decrease in RV medial tissue – Doppler: velocity systolic (S') and diastolic (E') waves and IVA m/s2, minimal dysfunction in AFI RV, and depressed strain and SR medial RV. The second group showed basal and medial dysfunction of RV and basal of LV. The third group: RV > 120 ml/m^2 tissue Doppler S' < 0.05 m/s, and E' wave and IVA m/s2 included inversion E/A ratio, IVA m/s^2, RV and LV decrease. AFI RV and LV show severe dysfunction, strain and SR RV basal and medial positive and LV basal and medial the same.
Conclusions: Echocardiographic analysis offers great possibilities for assessment of right and left ventricular dysfunction, identifying in particular, as well as timing and selection of patients for re-intervention.

1353: PERI-OPERATIVE RISK FACTORS FOR IN HOSPITAL DEATH OR RETRANSPLANTATION IN PAEDIATRIC HEART TRANSPLANT RECIPIENTS

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Introduction: While advances in surgical techniques and immunosuppression regimens have contributed substantially to the success of paediatric heart transplantation (HTx), increased understanding of peri-operative risk factors associated with death or retransplantation (ReTx) could potentially improve survival to hospital discharge.
Methods: Peri-operative risk factors were explored in 226 paediatric HTx recipients between 1995 and 2010.
Results: Mortality prior to hospital discharge occurred in 20 patients (9%), a further five patients (2%) underwent ReTx for early primary graft failure of whom one died peri-operatively. Death or ReTx in nine patients (36%) occurred < 48 hours of HTx secondary to primary graft failure ($n = 4$), operative complications ($n = 3$) or multisystem organ failure ($n = 2$), including 4/5 ReTx. Death or ReTx > 48 hours was secondary to primary graft failure ($n = 6$), infections ($n = 4$), multisystem organ failure ($n = 3$), rejection ($n = 2$) and postoperative complications ($n = 1$). In a multivariable regression model, factors associated with increased hazard of peri-operative death or ReTx were earlier year of HTx (HR: 1.2, $p = 0.001$), peri-operative use of factor VIIa (HR: 32.9, $p = 0.001$), postoperative chest re-opening (HR: 11.0, $p = 0.001$), postoperative use of extracorporeal life support (HR: 7.7, $p < 0.001$), rejection prior to discharge (HR: 6.0, $p = 0.03$), donor negative rhesus factor (HR: 6.2, $p = 0.001$), and higher donor BMI (HR: 1.033/kg/m^2, $p = 0.001$). UNOS status 1 (vs status 2) at the time of HTx was associated with increased hazard of death or ReTx from causes other than primary graft failure (71 vs 27%, $p = 0.05$), implying that patients on life support are at risk of death from non-cardiac causes despite receiving a HTx.
Conclusions: Important peri-operative risk factors for early death or ReTx post paediatric HTx include surrogates of poor graft function or bleeding, and early rejection. Understanding risk factors that impact on peri-operative outcomes may help with difficult decisions around the timing of listing for primary HTx and appropriateness of relisting for ReTx.

1354: A SMARTPHONE APP TO OPTIMISE INTER-STAGE HOME MONITORING FOR INFANTS WITH SINGLE VENTRICLE

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Background: While the surgical results of the Norwood procedure have improved dramatically over the past two decades, post-discharge interstage morbidity and mortality remain a significant problem. While interstage home monitoring is widely employed to try to minimise adverse interstage events, the need for data entry and interpretation by the family lead to a system that is imperfect at many levels. Advances in technology may facilitate improvements in automating home monitoring. We explored the use of an iPad and wireless technologies to help build an optimal solution for interstage monitoring.
Methods: We started with standard devices that are in current usage for interstage monitoring (pulse oximeter and weight scale). We constructed an iPad application that uses Bluetooth and other wireless technologies to automate the upload of data from these devices to the iPad. The iPad camera is used to obtain a 10-second video of the patient. Family members are responsible for entering intake and output manually. All data is automatically transmitted to the hospital servers and integrated with the electronic medical record (EMR). Machine learning algorithms that have been developed within the EMR help trend measurements and to triage clinical scenarios. The ability to view videos of the patient in a non-linear manner promises to yield important and previously unavailable insights as well as be of educational value for both providers and families.
Results: The prototype application has been developed and is functional. Its integration with an EMR system (Cerner) has been validated; validation with other EMR systems is in progress.
Conclusions: The ability to use a consumer device for automated interstage home monitoring could be an important advance in the care of these fragile and high-resource infants. Such technology could also be used for other high-risk children with congenital or acquired heart disease.

1357: A POPULATION-BASED STUDY OF PAEDIATRIC IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION FROM AUSTRALIA AND NEW ZEALAND

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Background: Idiopathic pulmonary arterial hypertension (IPAH) in children is an uncommon and progressive condition with a poor natural history. Currently available therapies have not been trialled in children and little information exists about long-term survival in treated children.
Methods: A multicentre, web-based Australian and New Zealand registry for children with PAH was established to better understand the profile of children with IPAH, aiming to describe demographics, presenting features, clinical course and outcomes of children aged three months to 18 years at the time of the PAH diagnosis, from 1 January 2002. A group of nine historic controls with IPAH diagnosed between 1970 and 1990 were used for comparison.
Results: Of the 24 patients enrolled with IPAH, 54% were female and 75% were Caucasian. The mean (SD) age at diagnosis was 9.3 (4.90) years with 50% in the WHO functional class III at presentation. During follow up, 20 (83%) were treated with an ERA, (exclusively Bosentan monohydrate), 17 (71%) with PDE5 inhibitor (sildenafil) and 10 (42%) with either epoprostenol or iloprost; 19 (79%) received warfarin. The mean duration of follow up was 3.45 years, with an annualised mortality of 6%. Study end-points of death/transplant at nine years post PAH diagnosis was 25% for patients in the current era compared to 60% for historic controls ($p < 0.007$). Freedom from death, transplant or epoprostenol initiation for current era patients was 30% at seven years after diagnosis. The severity of PAH from cardiac catheterisation was similar between patients and controls, except for a lower mean CI of 2.57 ± 0.93 in current era group versus 3.85 ± 1.85 ($p < 0.05$) for the historic controls.
Conclusions: Multiple medical therapies, in particular a high utilisation of epoprostenol, have resulted in improved survival for children with IPAH. A better understanding of childhood IPAH will facilitate medical care, permit standardisation of therapeutic guidelines, and allow better representation in the healthcare arena.

1358: ECHOCARDIOGRAPHIC LONGITUDINAL STUDY IN CHILDREN WITH ACQUIRED IMMUNODEFICIENCY SYNDROME

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Background: Cardiovascular abnormalities in children with acquired immunodeficiecy syndrome is slow and progressive in occurence. The most frequent echocardiographic abnormalities are left ventricular dilation and dysfunction.
Aim: To study prospectively the cardiovascular profile of children with acquired immunodeficiecy syndrome (AIDS) by echocardiography.
Methods: We studied 46 HIV 1-positive children born to HIV-infected mothers who progressed to AIDS, 25 were males and 21 females, and age ranged from four months to 11 years (average 6 years). The patients were classified according to CDC's 1994 revised classification system and were longitudinally studied by two-dimensional and Doppler echocardiography from 1995 to 2012. The mean follow-up period was six years for each child.
Results: Cardiac abnormalities were encountered in 12 children (26%): mild tricuspid regurgitation: 16.6%; mild mitral regurgitation: 8.3%; mild aortic regurgitation: 8.3%; severe dilation of ascending aorta: 8.3%; mild dilation of left ventricle: 8.3%; pericardial effusion with cardiac tamponade: 16.6%; mild-to-severe left ventricular dilation and dysfunction: 33.3%; severe pulmonary hypertension with right ventricular dysfunction: 8.3%. Eight cases were in an advanced clinical immunological stage (C3) and the others had moderate signs and symptoms of AIDS. There was one case (C3) of reversibility of severe left ventricular dilation with dysfunction and the patient is doing fine at the age of 17 years.
Conclusions: Cardiac abnormalities initially appeared in children with moderate symptoms (B1) of AIDS and were more frequent in the ones with severe symptoms (C3). The most frequent abnormality was dilated cardiomyopathy and it was reversible in one case.

1361: 10-YEAR EXPERIENCE OF IMPLANTATION OF CONTEGRA GRAFT IN YOUNG PATIENTS: A SINGLE-CENTRE STUDY OF OUTCOME

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Background: It has been shown that the use of a bovine jugular vein graft (Contegra) to reconstruct the RVOT is currently, in the mid-term perspective, the best alternative to homografts. Recent studies also report on a sustained freedom from re-intervention in the long-term perspective for graft sizes 16 mm and larger. Here, we investigate the outcome in a group that consisted of predominantly young patients, and consequently also a higher number of smaller-sized grafts. We also investigated the value of post-implantation right ventricular and pulmonary arterial pressure to predict future stenosis.
Methods: A total of 94 grafts had been implanted between 2002 and 2012. Mean patient age was 3.3 years (3 days – 18 years, median 1.25 years). Total follow-up time was 226 patient years (25 days – 10 years, median 1.84 years). Diagnoses included: 37 pulmonary atresia, VSD with or without MAPCA (39%), 15 tetralogy of Fallot (16%), 14 truncus arteriosus (15%), 10 transpositions of the great arteries (11%), and 18 other diagnoses (19%).
Results: Implanted graft sizes ranged from 12 to 22 mm, with a predominance of 12-mm (9%), 14-mm (27%) and 16-mm (38%) grafts. There were thrree mortalities, all postoperatively in hospital, and none related to the graft. Freedom of re-intervention was 67% for graft sizes 12–14 mm and 93% for graft sizes 16 mm or larger. Post-implantation RV pressure could not predict future risk of stenosis development, 39/4 vs 36/2 mmHg, $p > 0.05$, stenosis vs normal, respectively.
Conclusion: Compared to previous reports, our study group was made up of younger patients and a higher proportion of smaller grafts. Nevertheless, our data support previous studies that have shown that smaller graft size, 12 and 14 mm, is the main independent risk factor for re-intervention. The Contegra grafts continue to be a reliable and readily accessible alternative to homografts.

1364: THE ROLE OF REFLUX OESOPHAGITIS IN PRECIPITATING ISCHAEMIC SYMPTOMS

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Background: A link has been postulated between ischaemic heart disease (IHD) and gastric-oesophageal reflux disease (GORD). To what extent GORD may precipitate angina is not clear. In 1985, Davies suggested that reflux may precipitate angina events. We designed a study to investigate the role of GORD in precipitating angina.
Hypothesis: It is possible that reflux symptoms may precipitate angina events. The objectives were to determine whether the presence of GORD may serve as a trigger for symptoms of ischaemia in subjects with coronary artery disease (CAD) and to document the electrocardiographic (ECG) changes that occur during reflux.
Methods: Patients with endoscopically confirmed GORD, as well as recent MI admitted to CCU, constituted the study group. Patients with GORD underwent endoscopy and acid installation to determine whether GORD could induce ischaemia. Hydrochloric acid (0.1 N) was instilled during endoscopy and the ECG simultaneously recorded. Ischaemia was detected by ST changes during ECG monitoring. Nuclear imaging with methoxyisobutylisonitrile (MIBI) scanning was performed to look for objective evidence of ischaemia. Twenty

normal volunteers served as control subjects.
Results: To date 23 subjects with GORD have been studied. ST-segment changes were noted in 12 out of 23 and five showed ischaemia on MIBI scanning. In the IHD group, 21 out of 35 showed ST changes, with 33 of the 35 showing changes consistent with ischaemia on MIBI scanning. Other ECG changes were frequent during acid installation. Only three subjects showed no changes on the ECG monitoring.
Conclusion: Acid reflux produced ischaemic changes on the ECG recording and may have lowered the threshold for angina and coronary events. We present evidence to suggest that coronary vascular changes may arise from oesophageal stimulation and may lead to the development of myocardial ischaemia.

1377: SUCCESSFUL SURGICAL CORRECTION OF TYPE II TRUNCUS ARTERIOSUS AT THREE YEARS OF AGE

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Background: Truncus arteriosus is an uncommon congenital cardiac malformation. The condition presents in early infancy with features of pulmonary overcirculation as the neonatal pulmonary hypertension regresses. We report the clinical case of a girl from rural Bengal whose first presentation to a paediatric cardiac facility took her three long years after birth.
Methods: Initial clinical examination and investigations showed cardiomegaly, with biventricular enlargement and pulmonary plethora. On echocardiography she was diagnosed to have type II truncus arteriosus with mild truncal valve regurgitation.Cardiac catheterisation was done for operability, which showed pre-oxygen pressures of the aorta and pulmonary artery to be 100/56/75 and 84/49/69 mmHg. Post oxygen aortic and pulmonary artery pressures were 105/60/80 and 72/40/50 mmHg, respectively, which showed a drop of 20 mmHg in the mean PA pressures post oxygen, depicting operability.

She underwent corrective surgery with closure of VSD with a flap technique and reconstruction of the right ventricular outflow tract with a 16-mm valved conduit. Postoperatively she was managed with inhaled nitric oxide, sildenafil and bosentan. She required prolonged ventilation and inotropic support. Her recovery was good and echocardiography on the 27th postoperative day documented good biventricular contractility, estimated pulmonary arterial systolic pressure of 34 mmHg and mild truncal valve regurgitation.
Conclusion: Most of the world literature on surgical correction of truncus arteriosus report surgery at a very young age, which is explained by the early irreversible change of pulmonary vasculature these patients develop if they remain uncorrected. Our patient along with few other published reports testify to the fact that meaningful surgical outcome is still possible in a select subset of patients with this condition who present late, if they are chosen carefully on clinical grounds.

1378: VASCULAR MECHANICS AT REST AND DURING EXERCISE AFTER ARTERIAL SWITCH OPERATION FOR COMPLETE TRANSPOSITION OF THE GREAT ARTERIES

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Background: Progressive neo-aortic root dilatation and regurgitation after arterial switch operation (ASO) for complete transposition of the great arteries (TGA) are well documented. We tested the hypothesis that neo-aortic stiffness is increased and is associated with neo-aortic dilatation in patients after ASO. We further explored the changes in mechanics of the neo-aorta during exercise stress.
Methods: Thirty patients (22 males) aged 16.2 ± 2.1 years and 22 healthy controls (15 males) were studied. Central and peripheral arterial pulse-wave velocity (PWV), carotid (c-AI) and radial (r-AI) augmentation indices and central systolic blood pressure (CsBP) were assessed by applanation tonometry. Dimensions of the aortic annulus, sinus, sinotubular junction, ascending aorta, and right carotid artery dimensions were determined at rest and during exercise by two-dimensional echocardiography. Aortic strain, distensibility, aortic and carotid stiffness indices were calculated.
Results: At rest, patients compared with controls had higher c-AI, heart–carotid PWV, CsBP, and r-AI (all $p < 0.05$), while brachial–ankle arterial PWV were similar. During rest and exercise, patients had significantly lower aortic strain and distensibility, and greater systolic blood pressure, and aortic and carotid stiffness were significantly different ($p < 0.05$). Aortic root dimensions at all levels were significantly greater in patients compared with controls (all $p < 0.05$). Patients with aortic dilatation had higher CsBP and aortic stiffness at rest, and lower aortic strain and distensibility at rest and at submaximal exercise (all $p < 0.05$). Linear regression models identified resting aortic distensibility ($\beta = -0.57$, $p = 0.005$) and age at operation ($\beta = 0.40$, $p = 0.005$) as significant determinants of aortic sinus *z*-score. Significant aortic regurgitation was identified in 18.8% (6/32) of patients, in whom significantly higher *z*-scores for the aortic annulus and sino-tubular junction were found (both $p < 0.05$).
Conclusions: In adolescents late after ASO for TGA, aortic root dilatation and regurgitation were prevalent and were associated with stiffening of the central arteries at rest and during exercise.

1379: PRESENTATION AND TREATMENT OUTCOME OF TAPVC IN AN OLDER POPULATION: A SINGLE-CENTRE EXPERIENCE FROM INDIA

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Background: TAPVC is a rare congenital heart disease that is diagnosed in the neonatal period or in infancy. This study investigated the spectrum of presentation and treatment outcome of an Indian cohort who presented after five years of age with TAPVC.
Methods: Isolated TAPVC cases were identified from our institutional database between 2003 and 2012. We reviewed the medical records of 12 patients who belonged to the age group specified, and analysed the data.
Results: A total of 98 patients were diagnosed with TAPVC. Of these, 12 patients (12.24%) presented after five years. Median presenting age was 13.25 years (range 5–30 years) with eight males (66.66%) and four females (33.33%). Unobstructed supracardiac TAPVC was the most common diagnosis, and shortness of breath on exertion the most common presenting complaint. All patients were investigated with ECG, chest X-ray, and echocardiography pre- and postoperatively. All were in sinus rhythm pre- and postoperatively except one who had transient atrial fibrillation in the immediate postoperative period. TAPVC with moderate pulmonary arterial hypertension was the most common echocardiographic diagnosis pre-operatively. All the patients underwent rerouting of the pulmonary veins and closure of the ASD with the flap technique. One patient died 40 days after the operation from sepsis and multi-organ failure. Mean duration of ventilation was 20 hours. Median duration of hospital stay after surgery was 10.5 days. The postoperative period was uneventful for all except one who required longer duration of inotropic support and ITU stay. Only one patient required oral sildenafil for PAH postoperatively. The median duration of follow up was 25.5 months (range 1–83 months). Three patients were lost to follow up. All patients were asymptomatic during follow up and were in sinus rhythm.
Conclusion: Patients diagnosed late with TAPVC were those who are naturally selected for better survival. Operative outcome was good with complete reversal of PAH in the majority of patients.

1387: ADENOSINE IMPROVES THE EFFICACY AND COST-EFFICIENCY OF TRANSOESOPHAGEAL ELECTROPHYSIOLOGY STUDIES TO RISK-STRATIFY PATIENTS WITH ASYMPTOMATIC WOLFF–PARKINSON–WHITE SYNDROME

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Introduction: All patients with Wolff–Parkinson–White (WPW) syndrome, even if asymptomatic, are at risk of sudden death during atrial fibrillation (AFib). This risk can be determined by measuring the shortest pre-excited R-R interval during AFib. This study evaluated adenosine's potential to induce AFib during transoesophageal electrophysiological studies (TEEPS) when atrial-pacing modalities alone failed to do so, and analysed its cost-saving potential during the work-up of asymptomatic WPW.
Methods: A retrospective review was carried out of patients with WPW who had had a TEEPS performed. Inclusion criteria: evidence of WPW on electrocardiogram, no history of arrhythmias or associated symptoms, age < 18 years, and failure to induce AFib by initial TEEPS protocol. When TEEPS protocol was unsuccessful, adenosine 0.2 mg/kg was administered via rapid intravenous push during atrial-burst pacing. AFib was considered successfully induced if persisting > 20 s. If induced, patients were classified as at risk of sudden death if the shortest pre-excited R-R interval during AFib was < 250 ms or at no risk if ≥ 250 ms. Using 2011 and 2012 Medicaid re-imbursement data, the cost of adenosine during TEEPS was compared to proceeding directly to a transvenous electrophysiological study (TVEPS).
Results: Inclusion criteria were met by seven patients. Adenosine and atrial-burst pacing induced AFib in four of these patients (57%). Of those induced, three (75%) had no risk and one (25%) had risk. No complications occurred. The average cost of TEEPS was $999, TVEPS $4 524, and adenosine $84.21. The average cost of the adenosine-decision arm was $2 669.35, resulting in an average cost saving of $1 854.65.
Conclusions: To improve the efficacy and cost-efficiency of TEEPS to risk-stratify patients with asymptomatic WPW, we recommend rapid intravenous adenosine administration during atrial-burst pacing when prior modalities have failed to induce AFib.

1389: REFERENCE ECHOCARDIOGRAPHIC MEASUREMENTS IN LOW-BIRTH WEIGHT INFANTS IN A DEVELOPING COUNTRY

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Aim: To obtain a set of reference echocardiographic values in a group of low-birth weight infants in central South Africa as none exist for sub-Saharan Africa.
Methods: Over a 12-month period, 290 infants were included. Standardised M-mode, two-dimensional and systolic functional assessments [shortening fraction (SF), myocardial performance index (MPI)] were carried out according to the guidelines of the American Society of Echocardiography. Studies were performed by three experienced echocardiographers and reviewed by a paediatric cardiologist. A longitudinal study was also included to examine changes in these indices from days 1–28 of life.
Results: Median weight was 1.36 kg (range: 0.69–2.50) with a median gestational age of 31 weeks (range: 26–38). Eighty-seven (29%) infants were small for gestational age (SGA). Body surface area (BSA) and weight had a near-perfect correlation (r = 0.98). Inter-observer variation was less than 6%. Cardiac dimensions increased with increase in body weight. SF and MPI for left and right ventricles were 34.6 ± 6%, 0,29 ± 0.14 and 0.23 ± 0.15, respectively. Dimensions of SGA infants did not differ from other infants of comparable weight. Longitudinal data showed that all dimensions from birth to 28 days remained within the ranges determined by the study. Comparison with international reference ranges showed that local interventricular septal and posterior wall thicknesses as well as left atrial dimensions were significantly larger ($p < 0.01$).
Conclusion: Cardiac dimensions increased parallel to increase in body weight. BSA and weight correlated excellently and either can be used. Longitudinal data indicated that reference values obtained are applicable to infants from 0–28 days of age. The differences in certain cardiac dimensions from international reference values demonstrate that regional differences exist and emphasise the need for development of local reference ranges.

1395: CONGENITAL COMPLETE HEART BLOCK: REVIEW OF CASES SEEN AT CHRIS HANI BARAGWANATH ACADEMIC HOSPITAL FROM 1986–2012

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Introduction: Congenital complete heart block is a rare conduction anomaly, which may either be isolated or associated with congenital structural heart disease. Prenatal diagnosis, identification of high-risk neonates and early planned management is associated with improved outcomes.
Methods: This was a retrospective clinical review of 30 cases between 1986 and 2012 presenting to the Division of Paediatric Cardiology at the CH Baragwanath Academic Hospital, which is a southern African tertiary care institution. Data related to clinical features, diagnosis, pacemaker interventional procedures and follow up were collected.
Results: Thirty-four patients (18 males and 16 females) were diagnosed: 33 (97.1%) postnatally, and one antenatally. Eight patients were delivered by Caesarian section for foetal distress, including six premature babies. Presenting features included bradycardia, congestive cardiac failure, respiratory distress, cardiac murmur, cardiomyopathy with poor left ventricular function and metabolic acidosis. Median age at diagnosis was two days. Antibodies Ro/Lo were positive in 21 patients (61.8%). The major associated cardiac lesion was patent ductus arteriosus in 15 patients, four of whom had left ventricular non-compaction. Four patients had heterotaxia associated with atrio-ventricular septal defects. Other cardiac defects found in two patients were valvar pulmonary stenosis and a secundum atrial septal defect. The median ventricular rate was 50 beats/min. The median atrial rate was 150 beats/min. Four patients (11.8%) had QTc interval > 460 ms. Two patients had a wide QRS duration greater than 120 ms. Thirteen patients (38.2%), including six neonates had permanent pacemaker insertion at median age of five weeks. Thirteen patients (38.2%) died prior to permanent pacemaker placement, including three patients with heterotaxia. Three patients died after permanent pacemaker insertion.
Conclusion: Congenital heart block in our setting has a high mortality rate and is linked with premature delivery. The majority of patients were associated with autoimmune antibodies.

1397: INVESTIGATING SYNCOPE IN CHILDREN, WHERE TO DRAW THE LINE

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Background: Syncope is the temporary loss of consciousness and postural tone resulting from an abrupt transient decrease in cerebral blood flow. Its death-like quality makes it an alarming presentation for parents and investigating it is troublesome, expensive and often fruitless. The present work aimed at determining the yield of diagnostic tests in paediatric syncope at a tertiary paediatric referral

centre and to report on the prevalence of cardiac diagnoses among these patients.
Methods: The work was a retrospective study conducted at a tertiary referral arrhythmolology service. The clinical charts of 1 896 paediatric patients presenting with a primary complaint of syncope with an average age of 8.3 ± 2.1 (3.4–17) years were reviewed by the investigators. Statistical Package of Social Science (SPSS) version 14 was used for the analysis of data. A complete history, physical examination, 12-lead ECG, EEG, echocardiography and Holter monitoring were done for the whole study group.
Results: The commonest trigger for syncope in the study population was early following exercise ($n = 526$) and the commonest prodrome was palpitation, noted in 190 patients. A murmur was present in 147 of our patients (7.7%) while 107 (5.6%) had abnormal ECGs. Echocardiography revealed a cardiac cause in 109 (5.7%) of them, two were missed by ECG. The most common cardiac cause was HOCM. All patients were offered ambulatory 24-hour ECG. One patient with sick sinus syndrome was diagnosed only with Holter. EEG diagnosed a neurological cause for syncope in 88 (4.6%) patients, all suspected through history taking.
Conclusions: Twelve-lead ECG and history taking are the most cost-effectivel investigations in diagnosing syncope in children. Transthoracic echocardiography, Holter monitoring and EEG have a low yield in paediatric syncope and should be reserved for when an abnormality is suspected from the history or 12-lead ECG.

1398: THE YIELD OF AMBULATORY 24-HOUR HOLTER MONITORING IN PAEDIATRIC PATIENTS

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Background: The diagnosis of arrhythmias may be a tedious process, due to the episodic and often transient nature of the disease, hence the need for longer periods of ECG recording. Holter monitoring (HM) has been established as one of the most effective non-invasive clinical tools in the diagnosis, assessment and risk stratification of cardiac patients. However, studies on its value in children remain debatable. The present work aims at determining the value of HM in the diagnosis and management of children.
Methods: The work was a retrospective study conducted at a tertiary referral arrhythmolology service. Holter records of 4 015 paediatric patients (52% males and 48% females) with an average age of 4.8 ± 2.9 years (2 days – 16 years) were reviewed. The indications for which Holter monitoring was done were analysed as well as all the abnormalities diagnosed and factors that may increase Holter yield.
Results: The most common indications were palpitations (23%), syncope (16%), cardiomyopathy (14%), chest pain (8%), evaluation of anti-arrhythmic therapy (8%), postoperative assessment (3.7%) and complete AV block (4.5%). A total of 602 Holter recordings were found abnormal with a total diagnostic yield of 15%. The highest contribution to diagnosis was in postoperative assessment (37%), cardiomyopathy (28%) and palpitations (14%), where the most common abnormalities were frequent supraventricular/ventricular premature beats, supraventricular tachycardia, ventricular tachycardia and AV block. Diagnostic yield was lowest in patients with syncope (0.4%) and chest pain (0%). An abnormal ECG was significantly associated with a higher diagnostic yield ($p = 0.0001$).
Conclusions: HM plays an extremely valuable role in the assessment of high-risk patients (postoperative and cardiomyopathy) and those with palpitations. However in children with syncope and chest pain, HM had a low yield. In this group of patients an abnormal ECG is more likely to be associated with abnormal Holter recordings.

1401: TELECARDIOLOGY PARTNERSHIP BETWEEN WASHINGTON, DC AND MARRAKECH, MOROCCO: SUPPORTING A GROWING PAEDIATRIC CARDIOVASCULAR SERVICE IN THE DEVELOPING WORLD

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Background: Only 7% of the world's population has access to modern paediatric cardiology, resulting in nearly six million children with treatable conditions who are denied care. Telemedicine has the potential to help bridge this gap by providing remote consultation and distance education.
Methods: The Children's National Medical Centre developed a telemedicine partnership with Marrakech, Morocco (4–5 hours time difference) to augment the skills of the paediatric cardiovascular team. Videoconferencing units and satellite dishes were installed in 2009 with subsequent training in 2010.
Results: Live monthly videoconferences were started in 2009, increasing to weekly in 2011 between cardiovascular teams in Washington (CS, SS) and Marrakech (DB, YB). Patient data and echocardiograms were reviewed in real time. The Children's Hospital technical team managed conferences remotely; 38 conferences occurred in the last 12 months and 14 were cancelled due to scheduling conflicts (no technical difficulties). Ninety-five cases/73 patients were presented; 22 were discussed more than once. Most common diagnoses were tetrology of Fallot ($n = 14$), transposition of the great arteries ($n = 10$), double-outlet right venticle ($n = 9$), atrio-ventricular canal ($n = 8$), and ventricular septal defect ($n = 6$). Mean age was 4.8 years (3 days – 30 years); 44 cases were under two years old. Mean oxygen saturation was 83%; 22 ≤ 80%. Additional imaging was recommended in 22 patients; improvement in echocardiography skills was observed. Cardiac surgery was performed in 25% of patients, half had a difference in approach as a result of the teleconference. Three operations (tetralogy of Fallot, atrioventricular canal, D-transposition) were performed sucussfully in infants for the first time. Meetings with US and Moroccan government officials contributed to ongoing support. Focus on barriers including technology, satellite availability, language, funding and time difference has contributed to the sustainability of the project.
Conclusions: Telemedicine is an innovative and practical means to augment the skills of paediatric cardiovascualar surgery teams in the developing world.

1402: INTERMEDIATE-TERM RESULTS FOLLOWING POST-CARDIOTOMY EXTRACORPOREAL MEMBRANE OXYGENATION (ECMO) SUPPORT IN CONGENITAL HEART SURGERY(CHS) PATIENTS

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Background: There are considerable data regarding in-hospital results of CHS patients requiring post-cardiotomy ECMO; however there is limited information on mid-term outcomes.
Methods: A retrospective, single-institutional review of 25 consecutive CHS patients who survived to hospital discharge following post-cardiotomy ECMO between January 2003 and June 2008 was completed. Primary endpoint was survival at last follow up. Secondary endpoints included evidence of neurological deficits, renal injury, respiratory failure, and unplanned cardiac re-interventions and hospitalisations.
Results: Median age at ECMO support was four months. Primary indications for ECMO included cardiac arrest (12.48%), low cardiac output (7.28%), failure to wean from CPB (5.20%), and hypoxia (1.4%). There were 24 survivors; one death occurred within 48 hours of hospital discharge. Median follow up was 3.4 years (IQR: 1.7–6.0). Kaplan–Meier patient survival was 95% at three years (95% CI: 88–99%). Neurological deficits were present in seven

patients (cognitive deficits in five; motor deficits in three). No patients had evidence of renal dysfunction. At hospital discharge, three patients had respiratory failure requiring tracheostomy and at the latest follow up one remained dependent on mechanical ventilation. Echocardiogram in 23 patients demonstrated normal systemic ventricular (SV) function in 19 and mild–moderate SV dysfunction in four. Unplanned cardiac re-interventions were required in 13/19 patients (68%): both percutaneous and operative in four (21%), solely percutaneous in six (32%) and solely operative in three (16%). Unplanned hospital re-admissions were documented in nine patients.
Conclusions: Post-hospital discharge outcomes of CHS patients requiring post-cardiotomy ECMO were encouraging with regard to survival, respiratory and renal assessments. While SV function was largely preserved in the mid-term, the rate of unplanned cardiac re-interventions was high. Neurological impairment remained a concern with evidence of both cognitive and motor deficits.

1405: REPAIR OF ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY: OUTCOMES AND FOLLOW UP

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Backround: We reviewed early and late surgical results of children with anomalous left coronary artery from the pulmonary artery (ALCAPA).
Methods: Between July 1994 and June 2012, 40 patients with ALCAPA were repaired in our hospital. The median age was 4.5 months (range 1–156), mean weight 6 kg (range 3–63). Patients were divided in two groups: (I) direct coronary transfer (n = 34) and (II) coronary ligation (n = 6). Three simultaneous annuloplasties were performed. The mean follow up was 4.12 years (range 0.25–16.1).
Results: There were two hospital deaths: group I: 1/34 (2.9%), group II: 1/6 (16.6%) and one late sudden death in each group. Both techniques were equally effective regarding left ventricular shortening fraction (LVSF) (long-term follow up: 38% vs pre-operative: 23%) and mitral valve competence (mitral regurgitation (MR) was absent or mild in 76% of patients in long-term follow up). One patient required mitral valve replacement two months after coronary transfer. Four patients underwent a redo: second mitral annuloplasty, coronary artery bypass grafting, mitral valve re-replacement and pulmonary supravalvular stenosis repair (one, 10, 11 and 13 years after surgery). All survivors remain asymptomatic. Sixteen patients were catheterised during follow up. In four patients, the left coronary artery was occluded, all had normal LVSF and two had a normal excercise test. This was performed in 15 patients: five were abnormal, two however had normal coronary angiography.
Conclusions: A dual coronary system could be stabilised safely in children with ALCAPA. Early mortality was related to the severity of pre-operative left ventricular dysfunction. Late results were satisfactory, with marked improvement of left ventricular LVSF and mitral valve competence. Echocadiographic assesement and exercise test are useful tools to evaluate potential myocardial injury, however these two methods did not predict patency of the coronary arteries. Therefore, a coronary angiography should be included in the follow-up evaluation.

1410: CARDIAC RESYNCHRONISATION THERAPY (CRT) IN PAEDIATRIC CARDIOLOGY: A NEW TENDENCY OR THE EXCEPTION

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Background: Biventricular pacing was introduced in the middle 1990s as a new modality of treatment for patients with drug-refractory dilated heart failure (FE < 0.3) and ECG evidence of dissynchronous ventricular contraction. Initially the third lead was exclusively placed by thoracotomy in the left ventricle wall (epimyocardial). In 2000 the first system was introduced to place left ventricular leads through the coronary sinus. This study reports on our clinical experience with cardiac resynchronisation therapy per year since 1997, in patients under 18 years of age.
Methods and Results: We reviewed our case reports on CRT, from the first surgical procedure in 1997 to 2012. Since 1997 we have implanted 1 135 CRTs but only 16 patients were under 18 years old (1.41%). In both groups there was a prevalence of males; 68% in adults (group I) and 75% in the under-18s (group II). Five patients were complete congenital heart block with pacemaker VVI who developed dilated myocardial insufficiency from two months to eight years and elected to CRT.
Discussion: In children, CRT implantations are still limited by generator size, and lead diameter incompatibility with vascular dimensions. In adults epicardial leads are the exception, they are more commonly used in children. CRT is indicated in very specific cardiological conditions and even in those cases, the most optimistic expectations show three-quarters respond to the therapy, but it has a low impact on survival rates. Despite the growing indication for and feasibility of CRT in infants and neonates, we lack evidence and a consistent rationale for carrying out the procedure on them.
Conclusion: Despite our experience with adults, and other trials on CRT, in paediatric cardiology there is not enough evidence to know when, how and why to do this operation on infants.

1411: LATERAL TUNNEL GROWTH: MAGNITUDE AND IMPACT ON THE TOTAL CAVOPULMONARY RESISTANCE

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Background: The total cavopulmonary connection (TCPC) is usually performed with a lateral tunnel (LT) pathway or an extracardiac (EC) conduit. Growth potential of the LT pathway has been hypothesised as one of the main benefits of this strategy compared to an EC conduit. However, the nature, magnitude and haemodynamic implications of LT growth have not been characterised.
Methods: LT (n = 7) and EC (n = 3) patient-specific TCPC anatomies and flows were reconstructed from retrospective serial cardiac magnetic resonance images. Vessel centrelines were extracted using the Vascular Modeling Toolkit for the Fontan pathway (FP), superior vena cava (SVC), descending aorta (dAo) and pulmonary arteries (PA). Parameters computed included: minimum, mean, and maximum diameters, FP volume, Nakata index and McGoon ratio. To understand the impact of growth on TCPC resistance (R_{TCPC}), computational fluid dynamic simulations were performed for the LT group by simulating both serial changes (S1, S2) and absence of growth (S3).
Results: Absolute and normalised diameters increased in all vessels in the LT patients; in the EC patients' vessels, growth was less than expected. Changes in the FP volume were one order of magnitude larger in the LT than in the EC patients. Haemodynamically, average R_{TCPC} was seen to increase serially by 20%, while in absence of growth it increased 57%, indicating that TCPC growth helps limiting the R_{TCPC} increase while flow rate changes in time.
Conclusions: Geometric changes over an average span of almost five years showed cross-sectional and volumetric growth of the LT pathway. Based on simulations, growth had a positive effect on haemodynamic efficiency by reducing R_{TCPC} even when some growth was accounted for. These findings support the rationale of LT TCPC

growth potential, and may have implications for understanding the relative benefits of different approaches to the Fontan procedure.

1412: DISTRIBUTION OF HEART DISEASE IN A MULTI-CENTRE PAEDIATRIC CARDIAC REGISTRY

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Background: The incidence of congenital heart disease is generally eight per 1 000 live births. The University of Minnesota's multi-centre Paediatric Cardiac Care Consortium (PCCC) has enrolled children undergoing cardiac catheterisation, operation or autopsy examination since 1982. The study comprises all registered children born from 1991–2002 to state residents of Arkansas (AR), Minnesota (MN) and Missouri (MO). We hypothesised that age-stratified incidence of heart disease could be established for these states to investigate paediatric cardiac case finding and management.
Methods: De-identified PCCC records for children born from 1991–2002 in AR, MN and MO were analysed and categorised by diagnosis to calculate birth rate-adjusted incidence. Conditions with abnormal oximetric saturation that were detected by three months of age are 'oximeter-screenable' anomalies.
Results: The rate of registered heart disease for subjects born from 1991–2002 for the three states was 46.3 per 10 000 live births (1.8 million births). For PCCC registrations within the first three months of life, 17.1/10 000 were oximeter-screenable and 13.7/10 000 were potentially cyanotic (e.g. PS, CAVC), compared with the total incidence of 37.0/10 000. MO showed significantly higher registration rates for CAVC (Poisson distribution, $p = 0.012$), coarctation ($p = 0.0015$), TOF ($p = 0.028$) and VSD ($p = 0.0029$). AR registered more than half of left-to-right shunts within the first three months [9.4/10 000 for < 3 months vs 18.5/10 000 all ages (Poisson distribution, $p = 0.00064$)].
Conclusions: PCCC-registered overall heart disease rates showed no significant differences among states. Although other anomalies often present with desaturation, only 46% of heart disease registered within three months of life is oximeter-screenable. Therefore, additional screening methods are needed to optimally manage paediatric heart disease. Incidence and timing of presentation of specific anomalies and categories from a large cohort were analysed to allow insights into detection and management of paediatric heart disease and can be used to investigate mechanisms of environmental and genetic causation.

1415: DIASTOLIC FILLING IMPAIRMENT DURING EXERCISE LIMITS EXERCISE CAPACITY IN FONTAN PATIENTS

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Background: Better imaging techniques are needed to assess haemodynamic changes that limit exercise performance in Fontan patients. This study aimed at evaluating changes in end-diastolic and end-systolic volumes (EDV and ESV) using a novel cardiac magnetic resonance (CMR) methodology during mild, moderate and strenuous exercise.
Methods: Seven Fontan patients (five male, age 19 ± 5 years) underwent CMR at rest and during supine exercise on a programmable cycle ergometer. Systemic ventricular volumes were obtained at rest (heart rate 77 ± 13 bpm) and during mild (105 ± 10 bpm), moderate (129 ± 15 bpm) and strenuous (144 ± 13 bpm) exercise. Images were acquired using an ungated, free-breathing real-time CMR sequence (12–18 contiguous 8-mm slices). Software was developed to allow for synchronisation of short- and long-axis images with compensation for respiratory phase translation. Endocardial borders were delineated using a bi-plane model. Simultaneously, pulmonary artery pressures were measured during exercise using a fluid-filled catheter.
Results: Cardiac output (CO) increased continuously during exercise (6.6 ± 1.9 vs 9.4 ± 1.8 vs 11.1 ± 3.5 vs 11.5 ± 3.4 l/min; $p < 0.0001$). The increase in CO depended on a 94 ± 40% increase in heart rate. Stroke volume (SV) did not change from rest to mild exercise (87 ± 22 vs 90 ± 20 ml; $p = 0.458$) and decreased during moderate and strenuous exercise (90 ± 20 vs 85 ± 22 vs 79 ± 17 ml; $p < 0.0001$). EDV increased from rest to mild exercise (162 ± 39 vs 170 ± 43 ml; $p = 0.040$), but decreased during moderate and strenuous exercise (170 ± 43 vs 164 ± 47 vs 158 ± 45 ml; $p = 0.004$) whereas ESV did not change during exercise (74 ± 28 vs 80 ± 32 vs 78 ± 35 vs 80 ± 38 ml; p = NS). Mean pulmonary artery pressures increased during exercise (10 ± 4 vs 14 ± 4 vs 18 ± 4 vs 22 ± 5 mmHg; $p < 0.0001$).
Conclusions: In Fontan patients, CO augmentation during exercise was predominantly dependent upon increasing heart rate. Impaired ventricular filling caused a decrease in SV already evident at moderate exercise, suggesting that decreased preload reserve is an important determinant limiting exercise capacity.

1417: POPULATION STUDY OF 332 CONSECUTIVE NEWBORNS WITH HYPOPLASTIC LEFT HEART SYNDROME (HLHS): A SINGLE-CENTRE EXPERIENCE

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Background: Our hospital is one of the leading centres for HLHS treatment in Poland.
Aim: The aim of the study was the estimation of perinatal and anatomic data of consecutive newborns with HLHS operated at our institution.
Methods: This was a retrospective analysis of perinatal data and anatomical findings of 332 newborns with HLHS (226 boys: 68% and 106 girls: 32%) treated by staged Norwood operation at our institution from 1992 to 2011.
Results: Prenatal diagnosis was made in 58% of patients (68–75% in last five years), mean pregnancy duration was 39.4 ± 1.8 (31–43) weeks. Mean mother's age was 26 ± 5 (17–43) years. In 147 cases (44%) the child with HLHS came from the first pregnancy. Mean Apgar score was 8.7 ± 1.4 (1–10), mean birth mass was 3236 ± 505.6 g (1 995–4 430 g). Ten patients (3%) came from twin pregnancies. Five patients (1.5%) had coexistent extracardiac malformations. In four cases (1.2%) our patient was the second child with HLHS from the same parents. In six families (1.8%) cardiac or extracardiac malformations in the HLHS patient's siblings were confirmed (in two families TGA in a sibling). In one patient Turner syndrome (45, X) was diagnosed. Anatomical subtypes of HLHS: MA/AA in 123 patients (37%); MS/AA in 113 patients (34%); MS/AS in 93 patients (28%); MA/AS in three patients (1%). Right ventricle myocardial performance index (RV-MPI) was 0.521 ± 0.18 (0.2–0.968) vs 0.3 ± 0.078 (0.183–0.445) in the control group of 50 healthy newborns. Restrictive atrial communication was confirmed in 33 patients (10%). Mean ascending aorta diameter was 3.8 mm (1–7.5 mm). Severe tricuspid regurgitation was diagnosed in 40 patients (12%).
Conclusions: Patients with HLHS were usually male, well developed, full-term delivery newborns of a young mother. HLHS rarely coexisted with other malformations or genetic disorders. In our material MA/AA, MS/AA, MS/AS subtypes occurred with similar frequency, MA/AS was very rare. RV-MPI for HLHS patients was significantly higher compared with healthy neonates.

1421: ASSOCIATION OF TEMPORARY COMPLETE AV BLOCK AND JUNCTIONAL ECTOPIC TACHYCARDIA AFTER SURGERY FOR CONGENITAL HEART DISEASE

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Junctional ectopic tachycardia (JET) is a postoperative complication with a mortality rate of up to 14% in patients after surgery for congenital heart disease. This study evaluated the association of intra- and/or postoperative temporary third-degree AV block and the occurance of junctional ectopic tachycardia.
Methods: Data were collected retrospectively from 1 158 consecutive patients who underwent surgery for congenital heart disease in the period 2006 to 2010.
Results: The overall JET incidence was 2.8%. Temporary third-degree AV block occured in 2.8% of cases. Ongoing third-degree AV block requiring pacemaker implantation occured in 1.2% of cases. Postoperative JET was significantly correlated with temporary AV block ($p < 0.001$), occuring in 56% of patients with temporary AV block. No case of postoperative JET was reported in a patient with ongoing AV block. Furthermore, bivariate regression analysis showed a statistically significant correlation between postoperative JET and age at operation ($p < 0.001$), cardiopulmonary bypass time ($p < 0.001$) and aortic cross-clamping time ($p < 0.004$).
Conclusion: There was an association between temporary complete AV block and postoperative JET. In addition, ongoing complete AV block seemed to be negatively correlated with postoperative JET.

1426: QUALITY OF LIFE IN FAMILIES WITH A CHILD AFTER STAGED TREATMENT FOR HYPOPLASTIC LEFT HEART SYNDROME (HLHS)

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Background: In spite of increasing numbers of survivors with HLHS after a staged Norwood operation, data on quality of life in this group of patients and their families are limited.
Aim: The aim of the study was to assess the quality of life of children with HLHS and their families.
Methods: Mothers of 54 children with HLHS completed the questionnaire to assess subjectively quality of their children's life and impact of the child's illness on the family. All children (age 4–16 years) were operated on at our institution. In 35%, the child with HLHS was the only child in the family.
Results: Problems with physical activity in HLHS patients were reported in 79%, emotional problems in 25%, and educational in 9%. Development estimated as normal was reported in 89% of patients; 79% of patients attended normal schools or kindergartens. Good tolerance of frequent hospitalisations was reported in 75% of cases. Childhood illness is connected with strong parental stress (73%), and negative emotions such as sadness (41%), fear and helplessness (42%). Own family support, support groups of parents and religious faith were considered most helpful. Only 13% of mothers looked for professional psychological care; 94% of responders assessed the familial atmosphere as good, and in 67% the child's illness strengthened the parental marriage. The impact of the child's illness on the family's material situation was assessed as significantly negative in 79% . In 59% of families the father was the only working parent.
Conclusion: Patients with HLHS are active members of the society, they attend normal schools and kindergartens although their physical activity is limited. The family functioning is good but the child's illness is a cause of parental stress and indicates material problems. Increasing numbers of HLHS survivors indicates the need for a continuation of these studies concerning neurodevelopmental outcome, quality of life and family functioning in this group of patients.

1427: PAEDIATRIC CARDIAC INTERVENTION PROGRAMME IN UPPER EGYPT: MORE BENEFITS AND FEWER RISKS

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Background: The high prevalence of congenital heart disease among developing nations due to their high birth rate and the highly qualified medical staff needed to deal with such patients makes the development of sustainable cardiac centres for the treatment of children and young people with heart diseases, in countries where the facilities for such treatments are unavailable, a top priority.
Methods: The study included all children suffering from congenital heart disease who were referred for elective percutaneous cardiac intervention in Aswan Heart Centre over a period of six months. Patients and procedural data were recorded, including age, gender, weight, height, underlying heart disease, type of the procedure, procedure duration, outcome as well as any intra- or postprocedural complications and the severity level of these complications.
Results: Eighty-four children were included in the study; their mean age was 4.4 ± 4.3 years; the youngest patient was two months old while the oldest was 18 years. Patients underwent a wide range of percutaneous interventional procedures, most of which were balloon pulmonary valvuloplasty ($n = 18$), ASD device closure ($n = 18$), and PDA device closure ($n = 32$). Five patients underwent two interventional procedures in the same setting. The mean procedure duration was 52.7 ± 32.7 minutes and the maximum hospitalisation period was less than 48 hours. The success rate was 97.6% ($n = 82$) and the mortality rate was 0%. Periprocedural complications occurred in 19 patients, none of which were life threatening and the majority were self limiting intra-procedural arrhythmias.
Conclusion: The development of a specialised paediatric cardiac interventional centre in remote area that lacks this service is very cost effective. Such a centre will not only provide excellent medical service to children in need but will also free up the available surgical theatres, allowing these centres to deal with more complex and challenging cases.

1430: QUALITY OF CARE AND OUTCOMES AMONG CHILDREN UNDERGOING CARDIAC CATHETERISATION: A CHARITY-BASED PROGRAMME IN UPPER EGYPT

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Background: The inhabitants of Aswan and neighboring Upper Egypt amount to approximately 24 million people. Currently there are no local specialised cardiac services for this large population. This tremendous need motivated Prof Yacoub and the Chain of Hope team to stimulate the development of a new unit to offer cardiac facilities at the highest level.
Methods: All children with congenital heart disease who were referred for elective cardiac catheterisation in Aswan Heart Centre over a period of two years were subjected to a baseline 2D echocar-

diogram and a follow-up echocardiogram after device closure or valvuloplasty. Percutaneous cardiac interventions were done by a paediatric cardiologist with a minimum of five years' experience. A dedicated paediatric anesthetist was responsible for the general anesthesia/deep sedation throughout the procedure. The patients were admitted in a specialised post-catheterisation care unit for 24 to 48 hours post procedure. All demographic data as well as periprocedural hospital period including the procedural outcome and any complications were recorded and analysed.
Results: The study included 214 patients; 146 patients underwent 157 interventional procedures while the remaining 68 underwent diagnostic catheterisation. The interventional procedures included, ASD device closure (n = 50), PDA device closure (n = 42), valvuloplasty (n = 38), VSD device closure (n = 4) and coarctation stent (n = 8). The mean age was 4.7 ± 4.1 years; the youngest patient was two months old while the oldest was 18 years old. The success rate of the interventional procedures was 96.6% (n = 141) and the mortality rate was 0%.
Conclusion: Establishing a specialised paediatric cardiac catheterisation centre in a remote area with results comparable to highly qualified international centres is feasible with the help of experienced local and international teams as long as continuous funding and development plans can sustain such centres.

1447: THE USE OF IMPLANTABLE CARDIOVERTER DEFIBRILLATORS IN PAEDIATRIC PATIENTS

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Background: The implantation of devices in children is always a challenge. Implantable cardioverter defibrillator (ICD) therapy has been indicated in 14 children from March 2003 to June 2012 at Santa Casa de Sao Paulo Hospital. Ages ranged between eight and 16 years old. The objective was to analyse all pathologies, techniques, medical treatment and events related to these children.
Methods: The following diagnoses were observed: long QT syndrome (n = 2), hypertrophic cardiomyopathy (n = 3), Brugada syndrome (n = 1), LV non-compaction (n = 1), congenital heart disease – postoperative (n = 3), dilated cardiomyopathy (n = 1), catecholaminergic ventricular tachycardia (n = 1), rhythmogenic right ventricular tachycardia (n = 1) and idiopathic VT (n = 1). Syncope (n = 4), ventricular tachycardia (n = 6), or recovery from sudden death (n = 4) was the indication for ICD. In all cases an endovascular endocardial approach for implanting ICDs was used. The prostheses were located below the left pectoralis major muscle, in 12 patients and below the rectus abdominis muscle in two patients. The defibrillation threshold was distributed as follows: 15 J (n = 1), 20 J (n = 11), and 36 J (n = 2).
Results: The children were followed from one month to nine years. Each patient received pharmacological treatment for the arrhythmias with specific drugs. Seven patients had no events. Inappropriate shocks occurred in six patients. Three of them needed ablation due to atrial tachycardia. One patient had appropriate shocks. Two patients had lead dysfunction and needed replacement.
Conclusion: Cardioverter defibrillator implantation was successfully done by endovascular approach in our paediatric patients. The follow up of this group has showed that ICDs are the solution for those children with tachyarrhythmia whose medical treatment has failed.

1449: ANTENATAL DIAGNOSIS OF CONGENITAL HEART DISEASE IN NOVA SCOTIA: A 20-YEAR RETROSPECTIVE ON SURVIVAL AND SURGICAL OUTCOMES

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Background: Effective antenatal diagnosis of congenital heart disease can improve outcomes and survival in certain conditions. Our aims were to examine antenatal detection rates for hypoplastic left heart syndrome (HLHS), transposition of the great arteries (TGA), and tetralogy of Fallot (TOF) among live births at our tertiary care centre, and measure outcomes in infants with antenatal diagnoses. We hypothesised high, moderate, and low detection rates for HLHS, TOF and TGA respectively.
Methods: Charts for live births between July 1989 and December 2010 were identified from the Nova Scotia Atlee Perinatal Database and reviewed retrospectively. Stillbirths and infants with extra-cardiac abnormalities were excluded.
Results: Of the 215 618 live births in Nova Scotia during this time, 23 infants were born with HLHS, 25 with TGA and 40 with TOF. The rates of antenatal diagnosis of HLHS, TOF and TGA among live births were 57.7, 60.0 and 68.9%, respectively. The number of HLHS live births declined to zero by 2008. One-year survival for all infants born with HLHS was zero, regardless of the timing of diagnosis. A prenatal diagnosis of TOF was not associated with increased survival or decreased morbidity rates. A prenatal diagnosis of TGA was associated with significantly shorter time to NICU admission (1.1 vs 29.5 hours, $p < 0.03$). There were no significant differences between maternal or neonatal factors between cohorts. The commonest morbidities at most recent follow up were ADHD and behavioural difficulties.
Conclusions: The low incidence of antenatal HLHS diagnosis with a decline in HLHS live births suggests an increase in terminations. Incidence and benefit of antenatal diagnosis of TOF remains moderate. Preliminary data suggest that antenatally diagnosed infants born with TGA receive definitive management faster. Further review of TGA outcomes is pending. The notable incidence of ADHD could reflect latent hypoxic sequelae or chronic disease-related psychosocial issues.

1451 DONOR–RECIPIENT SIZE MATCHING IN PAEDIATRIC HEART TRANSPLANTATION: IS WEIGHT THE MOST APPROPRIATE PARAMETER TO PREDICT OUTCOMES IN ALL AGE GROUPS?

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Background: The aim was to retrospectively review our practice of donor–recipient (D/R) size matching by weight criteria alone and simultaneously reassess and compare post-transplant outcomes using the additional variables of height and body surface area (BSA).
Methods: We retrospectively reviewed the medical records of 211 patients who underwent orthotopic heart transplantation at Boston Children's Hospital over the past 20 years. The patients were divided into the following age groups: < one year (n = 30), one to 12 years (n = 107) and > 12 years (n = 74). Donor–recipient weight, height and BSA ratios were determined for each age group at the date of transplantation. Outliers were identified for each variable and analysed separately. We compared the average and median of continuous outcomes such as length of stay, need for open chest, post-transplant filling pressures and survival.
Results: In our cohort the median D/R weight ratio was 1.276 (0.2–5.0), BSA ratio was 1.172 (0.2–2.0) and height ratio1.061 (0.3–2.0). There was no difference in post-transplant survival when comparing outcomes by weight, BSA or height ratios. Increased D/R size ratio was associated with significantly increased likelihood of delayed chest closure, ICU and total hospital length of stay at all ages. The odds ratio was highest for the youngest age group with disproportionate number of outliers (33.3%) but this effect was less significant when D/R matching was done by BSA. The younger patients were more likely to receive size-mismatched hearts than older recipients.
Conclusions: The current practice of using weight for donor–recipient size matching in paediatric heart transplantation does not seem

to impact on long-term outcomes. In younger patients, an increase in donor-to-recipient size ratio increases postoperative recovery. However, the use of BSA is a more reliable tool for matching heart sizes in all age groups.

1453: CLINICAL AND SURGICAL EVOLUTION OF TRICUSPID ATRESIA AT A PAEDIATRIC HOSPITAL IN ARGENTINA

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Aim: The follow up of patients with tricuspid atresia (TA).
Methods: A retrospective review was done of 45 clinical records of patients with TA who were assessed at Pedro de Elizalde Children's Hospital from August 2005 to August 2012. They were grouped according to Edwards and Burchel classification. Analysis was based on clinical examination, radiology (Rx), electrocardiography (EKG), echocardiography, catheterisation records and surgical treatment. Statistical analysis: multivariable test $p < 0.05$.
Results: There were 44 patients with TA, 44 patients with situs solitus (SS), and one patient was indeterminate (Si). Type I: 43 patients had ventricle–arterial concordance (95.5%). Type IA: three patients had decreased pulmonary flow (PQ) (7%). Treatment: two systemic pulmonary anastomosis (SPA), one Glenn shunt. Complications: one patient with stenosis of the left pulmonary branch, one patient needed occlusion of the inferior vena cava. Type IB (58%): 24 patients had normal PQ. Treatment: two patients had Park septectomy, 14 had SPA, five had Glenn shunt, one had Glenn pulsatile, three had atrio-pulmonary connection, and three had extracardiac cavopulmonary connection. Complications: one patient needed mitral anuloplasty; one had endocarditis; one AV dissociation, and required a pacemaker; one had W-P-White syndrome. Type IC (32%): 12 patients had increased PQ. Diagnosis: all in the first month: four patients with pulmonary hypertension. Treatment: three patients had SPA; later: two had Glenn, one Glenn pulsatile, and two atrio-pulmonary connection. Complications: fibrillo-flutter four years later in one patient and AV block in one.

Type II: ventricle–arterial discordance (5%). Type IIA: two patients had decreased PQ, Si, RBVF, severe pulmonary stenosis and mitral insufficiency. Treatment: one patient had SPA that needed a stent in the pulmonary artery branch, Glenn dysfunction.
Type III: left-sided right ventricle (one loop) No patients. Follow up only in 23 patients.
Conclusion: Type I was the most frequent (95%) [IB (56%) and IC (33%)]. Cyanosis was the most common clinical manifestation (66%). EKG: superior axis was found in 19/45 patients, left ventricle hypertrophy in 82.9% and left anterior hemiblock in 70% of the patients. Regarding surgical procedure: 21 patients needed systemic pulmonary anastomosis, 12 Glenn shunt (nine non-pulsatile, three pulsatile), six atrio-pulmonary connection, and four extracardiac tube connection.

1461: DETERIORATING PAEDIATRIC CARDIAC WARD PATIENTS: A 12-MONTH REVIEW

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Background: To identify deteriorating patients and to describe characteristics of the events and mortality.
Methods: We retrospectively analysed data of all children from our paediatric cardiac ward requiring secondary emergency response team (SERT) attendance, and/or unplanned paediatric intensive care unit (PICU) admission between January and December 2010. Data were extracted from medical charts and the ward's clinical information system.
Results: Thirty-six events were observed in 28 patients; 16 (44%) were SERT calls, which included four (11%) episodes of CPR, and 20 (56%) separate PICU reviews. Overall 27 (75%) required transfer to PICU. Most common reasons for triggering SERT calls or PICU transfer were cyanotic episodes (28%), low-cardiac output state (23%), bradycardia (11%), and arrhythmia (SVT/VT, 11%). There was a statistical significance between cause of deterioration and whether the child was categorised as either medical or surgical ($p \leq 0.05$). Of the children that deteriorated, 86% were aged less than 12 months. Medical children were the most likely to deteriorate (57%) and it was more likely to occur in the close observation unit within the ward (69%) rather than in the general ward area. A slow deterioration as opposed to an acute deterioration occurred in 64% of cases. Timing of deterioration varied with 18% occurring within 24 hours of PICU transfer and 36% within 72 hours. More events (72%) occurred outside of office hours. Of the 28 patients there was a 24% mortality rate.
Conclusions: Deteriorating cardiac patients on the paediatric cardiac ward carry a high mortality rate long term. The implementation of a clinical early warning tool, specifically designed for cardiac ward patients, may be more efficient in detecting the deteriorating patient.

1468: LONGEVITY OF PALLIATION PROVIDED BY NEONATAL DUCTAL STENTING BEFORE DEFINITIVE SURGICAL CORRECTION FOR CONGENITAL HEART DISEASES WITH DUCT-DEPENDENT PULMONARY CIRCULATION

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Background: Ductal stenting (DS) in duct-dependent pulmonary circulation involves less morbid than neonatal Blalock–Taussig shunt. We aimed to study whether DS provided adequately long palliation.
Methods: A prospective interventional clinical follow-up study of consecutive infants followed after successful DS was done from two tertiary referral centres. Patients were divided into three anatomical groups. A: following pulmonary valvotomy for critical pulmonary stenosis and atresia, for continuing duct dependency; B: tetralogy of Fallot with pulmonary atresia where palliation is needed until conduit replacement; and C: univentricular hearts where ductal stent is needed until bidirectional Glenn shunt surgery. Interstage mortality was studied.
Results: Among a total of 24 infants, four patients in group A followed for 26–54 months had adequate oxygen saturations and no residual gradient. Tricuspid valve, pulmonary annulus and right ventricle grew with age. After discontinuing antiplatelets, two stents were patent after one year. In eight out of 11 group B patients, corrective biventricular repair using conduits was done after 5–14 months (body weight 5–7.5 kg). One patient awaits conduit repair. Bidirectional Glenn shunt and confluence repair was done in seven of nine group C patients after 8–15 months (weight 6–8.5 kg). The hilar pulmonary artery growths in B and C groups were adequate for surgical repair. No patient needed stent redilatations or additional shunts. Four patients had sudden death at home and no autopsy was done.
Conclusions: Duration of palliation by DS was sufficient to allow adequate somatic growth before the next surgery. DS was acceptable palliation after pulmonary valvotomy and univentricular hearts where short-term patency is sufficient. In patients needing biventricular repair with conduits, a longer term of palliation may be desirable and ductal stent did not provide adequate pulmonary flows for a long time. There was an interstage mortality of 16% of unknown causes after DS.

1472: CMR AS A NON-INVASIVE DEFINITIVE DIAGNOSTIC TOOL FOR COMPLEX CYANOTIC CONGENITAL HEART DISEASE

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Aim: To study the feasibility of CMR as a useful advanced diagnostic tool in the evaluation of CCHD in our institute during the period Junuary 2011 to May 2012.
Methods: We enrolled 12 patients with CCHD in the age group 8–50 years, female:male ratio 1:1. All patients had thorough clinical examinations along with 2D echo evaluation. CMR was the final decision-making tool for all patients, none had CT/cardiac cath for delineation of anatomy. After confirmation of the diagnosis, of the 12 patients, nine underwent surgical correction. Two patients refused surgery; one DORV with severe PAH was inoperable.
Results: Of our 12 patients, tetralogy of Fallot was found in three patients, DORV in its various forms in four patients, CCTGA in two patients, unbalanced AV canal defect with reduced pulmonary flow in one patient, DILV L-TGA with PS in one patient, and tricuspid atresia with failed Glenn in one patient. Two patients with TOF had total correction, one DORV with pulmonary atresia, MAPCAS had unifocalisation with central shunt, one DORV was identified with non-confluent PAS and had total correction, one DORV with PS with non-routable VSD had a Fontan, DILV and unbalanced AV canal had single staged fontan, TA also had a Fontan, one CCTGA underwent double-switch surgery. All these patients who were taken up for surgery had on-table assessment of PA pressures, and all were found to be fit for final surgery. The one who we predicated to be not suitable for surgery was confirmed by cardiac cath.
Conclusion: We conclude that CMR is a dynamic modality which can give all the necessary information to help us plan the management strategy in CCHD with decreased pulmonary blood flow, thus avoiding the need for catheterisation studies or CT angiogram in most of the complex congenital heart diseases.

1475: FUNCTIONAL INDEXATION OF RV PARAMETERS IN REPAIRED TOF PATIENTS: A BETTER REFLECTION OF CLINICAL STATUS?

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Background: Routine cardiac magnetic resonance imaging (MRI) is a well-accepted imaging method for assessment of right ventricular (RV) and left ventricular (LV) functional parameters and degree of pulmonary insufficiency after repair of tetralogy of Fallot (TOF). Poor correlation with clinical status as expressed by New York Heart Association (NYHA) class has however been reported. We hypothesised that a combination of degree of RV dilatation [expressed by RV end-diastolic volume (RV_{EDV}) and end-systolic volume (RV_{ESV})] indexed for LV function [expressed by LV ejection fraction (LV_{EF})] may better correlate with clinical status than these parameters individually.
Methods: Wer studied 227 repaired TOF patients (mean age 12.0 years ± standard deviation 4.6 years) with routine cardiac MRI. RV_{EDV} and RV_{ESV} were indexed for body surface area[1] and LV_{EF}[2], respectively. Clinical status as expressed by NYHA class was derived from clinic visit at around the time of MRI. A univariable model was used for statistical analysis.
Results: RV volumes (mean ± standard deviation) were: RV_{EDV}[1]: 144.96 ± 38.49 ml/m^2, RV_{EDV}[2]: 2.60 ± 0.81, RV_{ESV}[1]: 76.74 ± 28.25 ml/m^2, RV_{ESV}[2]: 1.39 ± 0.59. Other parameters were: LV_{EF}: 56.81 ± 6.18%, RV_{EF}: 48.07 ± 9.24%, $LV_{stroke\ volume}$: 46.86 ± 9.03 ml, $RV_{stroke\ volume}$: 68.09 ± 17.67 ml, LV_{EDV}[1]: 84.28 ± 19.52 ml/m^2, LV_{ESV}[1]: 36.35 ± 9.71 ml/m^2, pulmonary regurgitation fraction (PRF): 32.51 ± 13.61%. RV_{ESV}[2] and RV_{EDV}[2] showed much higher correlation (0.0002 and 0.007, respectively) with a higher NYHA score than RV_{ESV}[1] and RV_{EDV}[1] (0.80 and 0.03, respectively), or LV_{EF} (0.43), RV_{EF} (0.05) and PRF (0.11).
Conclusions: RV volumes indexed for LV_{EF} showed a much higher correlation to clinical status than all routinely used RV and LV parameters as indexed by BSA. RV volume indexation for LV_{EF} that combines degree of RV dilatation and LV (dys-)function may therefore be more useful in clinical follow up of TOF patients.

1477: HEART RATE-CORRECTED PULMONARY ARTERY ACCELERATION TIME CORRELATES WITH PULMONARY CAPACITANCE AND RIGHT VENTRICULAR STROKE WORK IN CHILDREN WITH PULMONARY HYPERTENSIVE VASCULAR DISEASE: A SIMULTANEOUS ECHOCARDIOGRAPHIC AND CARDIAC CATHETERISATION STUDY

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Background: Pulmonary capacitance calculated at cardiac catheterisation is a strong independent predictor of mortality in pulmonary hypertensive vascular disease (PHVD). However cardiac catheterisation is invasive, risky and is unsuited for frequent assessments. There is a pressing need for non-invasive correlates of catheter-obtained haemodynamic parameters. We sought to investigate relationships between Doppler-derived indices and invasively obtained haemodynamic measurements.
Methods: We performed transthoracic echocardiograms with Doppler interrogation on children undergoing cardiac catheterisation with PHVD after induction of anesthesia. We measured tricuspid regurgitation velocity (TRV) and heart rate-corrected pulmonary artery acceleration time (PAAT). From cardiac catheterisation data we calculated pulmonary capacitance index (PCI) and right ventricular stroke work index (RVSWi).
Results: We studied 17 consecutive patients (11 males, median age six years, range 0.4–15). Mean PA pressure was 40 ± 20 mmHg and mean Rp was 9.9 ± 6 WU/m^2. Peak TR velocity correlated with systolic pulmonary artery pressures (PAp) ($r = 0.79$, $p < 0.01$). Heart rate-corrected PAAT correlated negatively with PCI ($r = –0.58$, $p = 0.03$) and RVSWi ($r = –0.68$, $p = 0.01$).
Conclusion: Increased heart rate-corrected PAAT was associated with reduced PCI and RVSWi, both measures of the efficiency of the RV performance and pulmonary artery–right ventricular coupling. Heart rate-corrected PAAT is measured non-invasively by Doppler. Heart rate-corrected PAAT is easy to obtain, reproducible and may prove useful in the assessment of children with PHVD.

1481: PULMONARY VEIN STENOSIS IN EX-PREMATURE INFANTS

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Background: Pulmonary vein stenosis (PVS) is a rare disorder that may occur as an isolated lesion or in association with congenital heart defects. It can be acquired following cardiac surgery or interventions around the pulmonary veins. However, a group of ex-premature infants who developed PVS has been identified, although the aetiology and association with prematurity remains poorly understood.
Methods: We reviewed all available clinical and laboratory data in

patients with a diagnosis of pulmonary vein stenosis. We excluded patients with total anomalous pulmonary venous drainage, atrial isomerism and gestational age ≤ 35 weeks.
Results: We identified 11 patients with pulmonary vein stenosis, eight were male, median gestational age was 27 weeks (25–34), median birth weight was 860 g (432–2 100 g), and three patients were of twin pregnancies whose twin siblings were unaffected. Most patients were diagnosed with chronic lung disease and needed significant respiratory support after birth. In nine/11 initial neonatal echocardiograms did not report abnormal pulmonary vein flow. The median age at diagnosis was 5 months (3 months –2 years) the diagnosis was most often made by or suspected by echocardiography because of apparent worsening of chronic lung disease. Eight/11 patients underwent CT scan or MRI. The left pulmonary vein was the most commonly stenosed (91%) and all patients underwent a sutureless surgical repair. Median survival after pulmonary vein surgery was six months (4–10). In seven/11 PVS recurred and four/11 patients died.
Conclusion: Pulmonary vein stenosis should be considered if an ex-premature baby has late deterioration of chronic lung disease or evidence of pulmonary hypertension by echocardiogram. Further imaging by CT scan or MRI may be required to complete the diagnosis. Pulmonary vein stenosis appears to develop postnatally but the aetiology remains unknown. The response to surgery and late recurrence appear to be similar to pulmonary vein stenosis in infants born at term.

1483: SERIAL MEASURES OF SYSTEMIC-TO-PULMONARY ARTERIAL COLLATERAL FLOW IN PATIENTS WITH SUPERIOR AND TOTAL CAVOPULMONARY CONNECTIONS

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Background: We have described a method of quantifying systemic-to-pulmonary collateral (CollF) flow in patients with superior (SCPC) and total (TCPC) cavopulmonary connections using magnetic resonance phase-contrast velocity mapping (PC-MRI). Cross-sectional data suggests that CollF decreases with time after TCPC completion, but may take years. We wished to examine more acute changes in physiology and CollF from SCPC to TCPC by comparing serial (paired) PC-MRI data.
Methods: We retrospectively reviewed PC-MRI data to identify patients who had CollF quantified at both SCPC and TCPC. CollF, systemic blood flow (Qs = total caval flow), and pulmonary blood flow (Qp = total pulmonary vein flow) were quantified as previously described. Fenestration flow was quantified in TCPC by the difference in caval and pulmonary artery flow.
Results: Of 104 SCPC and 113 TCPC CollF studies, 26 patients had both SCPC and TCPC studies performed on average five months before and seven months after TCPC completion. Indexed CollF was 1.4 ± 0.7 l/min/m^2 at SCPC and 1.7 ± 1.1 l/min/m^2 at TCPC (p = 0.12), which was a significantly higher fraction of aortic flow in TCPC compared to SCPC (37 ± 18 vs 31 ± 14%, p = 0.03). There was a significant decrease in Qs from 3.2 ± 0.7 l/min/m^2 at SCPC to 2.8 ± 0.8 l/min/m^2 at TCPC (p = 0.004). Q_p increases significantly, primarily as a result of increased cavopulmonary flow. Fenestration flow averaged 48 ± 42% of the inferior vena caval (IVC) flow at TCPC, with a significant decrease in right-to-left shunt from SCPC to TCPC. However, in four patients, fenestration flow equaled or exceeded IVC flow with flow reversal in the Fontan baffle above the fenestration in two patients.
Conclusions: CollF did not decrease after TCPC completion and as a fraction of aortic flow initially increased, despite a significant reduction in right-to-left shunt. Q_s decreased and Q_p increased significantly thereafter.

1486: CORONARY ARTERY DYSPLASIA WITH PERSISTENT SINUSOIDS: RARE CAUSE OF FATAL MYOCARDIAL INFARCTION IN AN INFANT

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Introduction: Myocardial infarction in infancy is a very rare and serious event. Coronary artery dysplasia with persistent sinusoids is an extremely rare defect. We report on a previously healthy infant with a normal heart, who developed a massive myocardial infarction and died secondary to severe coronary artery dysplasia.
Case report: A two-and-a-half-month old boy was transferred to intensive care unit of our hospital with a brief history of feeding intolerance and respiratory distress leading to cardiorespiratory arrest and requiring cardiopulmonary resuscitation with inotropic infusion. Other than congenital abnormalities such as mildly dysplastic left hip, multiple cortical renal cyst, undescended left testis and Talipes left foot, he had an asymptomatic postnatal course. Initial arterial blood gas showed severe lactic acidosis. Chest roentogram showed cardiomegaly with pulmonary congestion. ECG revealed persistent pulseless electrical activity. Echocardiogram demonstrated structurally normal heart with severely depressed biventricular function. Prolonged cardiopulmonary resuscitation was ceased after failure to gain any significant cardiac output and he was pronounced deceased. Micro-organism investigations including bacterial culture and nucleic acid analysis for viruses were all negative. Post-mortem examination showed a significantly enlarged heart, acute and subacute ischaemic changes, widespread myocardial fibrosis, normal coronary artery origin and course, and most importantly, persistent myocardial sinusoids. Histology confirmed marked coagulative necrosis with acellular fibrosis in the subendocardial region of the ventricles, dystrophic calcification, and persistent sinusoids. There were no inflammatory infiltrates suggestive of myocarditis. A coronary specimen revealed intimal thickening and myxoid changes, duplication and fragmentation of the internal elastic lamina and hypertrophy of media smooth muscle, along with the presence of myocardial bridging pattern.
Conclusion: Physicians and pathologists should be aware of unusual coronary artery dysplasia when managing infants with severe ventricular dysfunction.

1487: AORTIC DISEASE AND RECURRENCE OF CONGENITAL HEART DISEASE IN FIRST- AND SECOND-DEGREE RELATIVES OF PATIENTS WITH PERSISTENT DUCTUS ARTERIOSUS

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Aims: Persistent ductus arteriosus (PDA) and familial thoracic aneurysm and dissection (TAAD) are genetically heterogeneous. Data on the prevalence of aortic disease including TAAD, the familial recurrence of PDA, and other congenital heart disease (CHD) provide essential information for the investigation of genetic factors of these diseases.
Methods and Results: This was a retrospective patient-based study using a questionnaire assessing the prevalence of cardiovascular disease in family members of children with PDA. A thorough family history provided information on three generations. The data were analysed by a binominal test. The prevalence in the general population was derived from a Dutch population-based study. Over the last 30 years, 320 patients older than three months with isolated PDA have been treated in our institution. Data on 175 patients and their 2 336 family members were available. The prevalence of aortic disease was higher in the study group compared to the general Dutch population. Aortic disease was reported in four/590 = 0.7% of first-degree relatives and 29/1339 = 2% of second-degreee relatives. In second-

degree relatives, the prevalence of TAAD was increased to 0.7% (9/1339) compared to 0.016% in the general population ($p < 0.001$). The recurrence rate of CHD was higher (2.7%) in first-degree relatives than in second-degree relatives (0.79%).
Conclusion: These data provide evidence for a genetic association between PDA and aortic disease. The prevalence of PDA and other CHD is increased in first-degree relatives of the index patient, suggestive of common genetic factors in the determination of CHD and aortic disease.

1488: THREE-DIMENSIONAL EN FACE ECHOCARDIOGRAPHIC RECONSTRUCTION OF SIMPLE AND COMPLEX VENTRICULAR SEPTAL DEFECTS ON RIGHT VENTRICULAR SEPTALSURFACE USING A SIMPLE BEDSIDE PROTOCOL

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Background: Increasing clinical use of three-dimensional echocardiogram (3DE) has enabled volume rendering of ventricular septal defects (VSD) on the right ventricular septal surface to guide understanding of the cardiac morphology. This will help decision making on surgical or interventional closure. A simple 3DE protocol to show all VSDs from right ventricular surface in a uniform format will aid understanding among all team members.
Methods: We studied 40 patients with simple (single or multiple VSD) and complex VSD (tetralogy of Fallot, atrio-ventricular canal, double-outlet right ventricle, transposition of the great arteries and truncus arteriosus) using 3DE enface reconstruction. We adopted a uniform subxiphoid live 3DE acquisition of a sector width that covers the entire interventricular septum. This acquisition gives good reproducible images in most patients under 10–12 years using x7-2 matrix probe (Philipsmedical, Andover, MA). The probe is positioned in the coronal plane in subxiphoid window to display the entire ventricular septum from right ventricular inflow to outflow. Using volume rendering, an enface reconstruction of the right ventricular septal surface is presented to give anatomical location of the ventricular septal defect.
Results: Out of 40 patients, good 3DE acquisition was achieved in all except one with optimal images. Twenty-two patients had different types of simple VSD (single in 17 and multiple in five), four patients had atrio-ventricular canal, two had tetralogy of Fallot, six had double-outlet right ventricle, three had truncus arteriosus and two had transposition of the great arteries. The 3DE reconstruction was accurate in showing the VSD location to the interventionist (two perimembranous VSD and two muscular VSD) and surgeons.
Conclusion: A simple right ventricular septal surface enface reconstruction of the VSD by a bedside live 3DE greatly enhances understanding of the location of the VSD and aids in planning closure.

1490: MYOCARDIAL FUNCTION FOLLOWING REPAIR OF ANOMALOUS ORIGIN OF LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA) IN CHILDREN

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Background: We hypothesised that speckle tracking may improve detection of myocardial dysfunction and provide new pathophysiological insights in anomalous coronary artery from the pulmonary artery (ALCAPA).
Methods: Echocardiography including speckle tracking was performed in 22 children with ALCAPA (eight males, median age at surgery 0.4 years; IQR: 0.21–1.05) pre- and postoperatively and in 22 healthy controls. Measurements included global and segmental longitudinal, radial and circumferential peak deformation (strain) and synchronicity index (SI) defined as agreement of time to peak strain measurements between segments per subject, summarised using intraclass correlation coefficient.
Results: Global strains were lower in unoperated patients than controls (longitudinal: –46 vs –123; $p < 0.001$; circumferential: –48 vs –118; $p < 0.001$, radial: 110 vs 357; $p < 0.001$) and improved postoperatively (longitudinal: –46 pre vs –82 post; $p = 0.002$, circumferential: –48 vs –96; $p = 0.012$, radial: 110 vs 317; $p = 0.001$). Unoperated patients with normal 2D function ($n = 8$) had significantly impaired strain. Global dyssynchrony significantly improved postoperatively (longitudinal SI 0.93 pre vs 0.94 post, circumferential 0.85 pre vs 0.9 post, radial 0.71 pre vs 0.88 post). Global time to peak shortened (longitudinal 2 236 pre vs 1 589 post; $p < 0.001$, circumferential 2 037 vs 1 447; $p = 0.005$, radial 2 169 vs 1 602; $p = 0.01$ ms). Despite overall global improvement some abnormalities remained. Strain improved in the majority of segments but apical septal and anterolateral segments remained abnormal. Post-systolic index improved in some segments but pre-systolic stretch persisted.
Conclusions: Both global contractility (strain) and global synchrony (coordinated contraction) improved after repair of ALCAPA, suggesting recovery of hibernating myocardium. Contractility in some segments supplied by the anomalous left coronary artery failed to improve following ALCAPA repair, suggesting a degree of irreversible myocardial damage. 2D speckle tracking identified impairment of function not revealed by standard echocardiography.

1494: AN UNUSUAL CASE OF COARCTATION THE AORTA AND PATENT DUCTUS ARTERIOSUS

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Background: We describe a highly unusual variant of juxta-ductal coarctation of the aorta (CoAo) and the lessons learned by our team.
Case report: The patient, M, was a five-month-old female, who was hospitalised with bilateral pneumonia and referred for cardiac evaluation. Echo showed marked biventicular hypertrophy, CoAo gradient 45 mmHg, patent ductus arteriosus (PDA) 3 mm, pulmonary hypertension (PH) 75 mmHg. ECG suggested biventricular hypertrophy. Cardiac catheterisation: Asc Ao = 142/60 mmHg, RV = 110/10 mmHg, PA = 110/50 mmHg. Thoracotomy revealed CoAo and PDA. During PDA ligation with simultaneous aortic clamping there was severe bradycardia and collapse, without signs of ischaemia on ECG. Inotropes were ineffective. The patient recovered with direct cardiac massage, declamping of the aorta and removal of PDA ligature. Our interpretation was vagal stimulation. After stabilisation, PDA ligation was repeated with resection of coarctation with 'end-to-end' anastomosis. Further temporary haemodynamic instability occurred during chest closure but she was stabilized. However, three hours after surgery, the child suddenly developed refractory cardiac arrest. Post mortem study: PDA ligated, anastomosis of the aorta 'end-to-end'. A single coronary artery was found on the front of the pulmonary trunk in 0.7 cm from the valve, which divided into anterior and posterior descending branches and circumflex branch. No coronaries were present from the aorta. Right ventricular wall: 8 mm, left: 11 mm.
Discussion: We suppose that ischaemic symptoms of anomalous origin of the coronary supply were not seen in this case due to CoAo with PDA resulting in high PA pressure maintaining high coronary perfusion pressure. After correction of CoAo and PDA, PA pressure fell and led to an acute fall in coronary perfusion. Following post mortem we reviewed the pre-operative cardiac catheterisation and aortogram, which clearly showed the absence of coronary arteries from the aorta, which were not seen by our team focusing on the aortic arch and ductus.

1495: COMPARATIVE EFFECTIVENESS AMONG DIFFERENT IMMUNOGLOBULIN FOR KAWASAKI DISEASE

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Background: Kawasaki disease is the leading cause of acquired heart disease among children in most industrialised countries. Using immunoglobulin to treat Kawasaki disease, only a few studies have ever evaluated the comparative effectiveness among immunoglobulin from different manufacturing processes. Moreover, those studies were limited by small case numbers and lack of longitudinal follow up. The aim of this study was to evaluate the comparative effectiveness of immunoglobulin from different manufacturing processes.
Methods: The data come from the National Health Insurance Research Database of Taiwan. From 1997 to 2008, patients who were admitted with the diagnosis of Kawasaki disease and receiving immunoglogulin therapies for the first time were enrolled in the research cohort. The immunogloblulin manufacturing process was divided into β-propiolactonation, acidification and containing IgA. The comparative effectiveness was evaluated by portion of treatment failure, acute aneurysm formation, chronic aneurysm formation and recurrence.
Results: Totally 3 830 children were enrolled in the cohort. For treatment failure, β-propiolactonation had a relative risk of 1.45 (95% CI: 1.08–1.94). However, acidification and containing IgA were non-significant. For acute aneurysms, acidification had a relative risk of 1.49 (95% CI: 1.17–1.90), but β-propiolactonation and containing IgA were non-significant. For chronic aneurysm, β-propiolactonation had a relative risk of 1.44 (95% CI: 1.18–1.76) and acidification could protect it, with a relative risk of 0.82 (95% CI: 0.69–0.97). Containing IgA was non-significant. For recurrence, all three factors were non-significant.
Conclusions: This is the first and largest longitudinal study evaluating immunoglobulin effectiveness. β-propiolactonation immunoglobulin had higher risks for treatment failure and chronic coronary aneurysm. Acidification may increase the risk for acute coronary aneurysm. Further prospective studies may be needed to elucidate the real causal relationship between manufacturing process and outcomes.

1496: EPIDEMIOLOGY OF GROUP A STREPTOCOCCAL PHARYNGITIS IN THE VANGUARD COMMUNITY DEMONSTRATION SITE, SOUTH AFRICA

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Background: Incidence rates of group A streptococcal (GAS) swab-positive pharyngitis from 3.9–95/100 child-years have been reported. However, data from South Africa are scant and an understanding of the incidence of GAS pharyngitis among children within a local context is an important component of any acute rheumatic fever and rheumatic heart disease (RHD) control programme. Treating all symptomatic GAS sore throats in susceptible individuals in the community with a course of oral or parenteral penicillin presents the opportunity for primary intervention of RHD. The primary aim of this study was to determine the crude incidence rate of GAS pharyngitis-associated disease burden among children with sore throat.
Methods: We conducted a prospective, clinic-based study from 2008–2012. Following enrolment, participants with pharyngitis, aged 5–15 years were examined physically, before rendering a throat-swab specimen for microbiological culture. SA census data were used to estimate crude incidence rates.
Results: We enrolled 840 participants with a mean age of 8.17 years, presenting with pharyngitis as a symptom. GAS was positive in 181 patients (21.6%). The crude incidence of pharyngitis and GAS pharyngitis was estimated to be 837 and 180.4 cases/10^5 per year, respectively. A significantly higher incidence of GAS pharyngitis was observed in the younger age group (IRR 2.265, 95% CI: 1.61–3.21), a trend seen throughout each year of the study period.
Discussion: This is the first community-based prospective incidence study of GAS-positive pharyngitis in Africa. While it is likely that our prevalence and incidence rates are an underestimate, our data nevertheless confirm that the complaint of GAS pharyngitis is common, particularly among younger children. Our data call attention to the need for vigilance as to the correct management of sore throat.

1497: ISOLATION OF THE RIGHT INNOMINATE ARTERY WITH RETROGRADE FLOW INTO THE PULMONARY ARTERY

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Isolation of the innominate artery is a rare congenital vascular defect. It is often associated with connection to the pulmonary artery. The majority of patients described have right aortic arch and isolation of the left innominate artery. A literature review by J Manner *et al*. in 1997 showed only one of 16 patients had an isolated right innominate artery. We present a patient with a left aortic arch and isolation of the right innominate artery.
Case report: The patient is a six-year-old girl, diagnosed at birth with a large ventricular septal defect and pulmonary hypertension and 22q11 deletion. She had surgical repair of the VSD at three months of age. Postoperative echocardiogram showed no residual ventricular septal defect but the presence of a small patent ductus arteriosus. A cardiac catheterisation was done with the intention to close the duct. The angiogram showed a left aortic arch with absent right innominate artery. The left carotid, left vertebral and left subclavian arteries were seen. There was no patent ductus arteriosus seen in the usual position. Retrograde flow following the aortogram shows the right innominate artery draining via a duct into the main pulmonary artery. Surgical correction is considered in view of the arterio-venous steal. Manner *et al*. proposed that the pathogenesis of this condition is related to derangement of the embryonic aortopulmonary septation due to abnormal migration of neural crest cells.
Conclusion: Isolated right innominate artery is a very rare condition. This was seen in an asymptomatic patient with chromosome 22q11 deletion, unsuspected at the time of surgical repair of VSD, and can be a challenge diagnose.

1501: EVALUATION OF CARDIAC FUNCTION USING SPECKLE-TRACKING ECHOCARDIOGRAPHY AND TISSUE DOPPLER IMAGING IN THALASSAEMIA MAJOR PATIENTS

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Background: Speckle-tracking echocardiography (STE) and tissue Doppler imaging (TDI) are new techniques available for objectively quantifying the movement of individual cardiac segments. Increasing data suggest that STE and TDI provide reliable estimates of cardiac contractility and provide more detail than conventional echocardiography (CE).
Objective: To clarify the value of STE and TDI in the early detection of myocardial dysfunction in chronically transfused thalassaemia major patients.

Methods: Multi-centre study using STE, TDI and CE to examine 66 thalassaemia patients (incl. 14 children) and 132 age-, gender-, height- and weight-matched controls. STE will provide strain and strain rate for each of the 16 heart segments. All data will be compared across iron overload stratifications and between patients and healthy controls. STE performed before and after the blood transfusion (usually 4 units of packed red blood cells) in all patients will help to quantify the effect of the increase of cardiac pre-load on STE.

Data acquisition is performed on Philips IE33 machines and is currently under way. Each echocardiography study is de-identified and will be assessed by 3 different experienced cardiologists utilising the latest release of the Philips QLab Software (version 9) to test for inter-observer variability. The examiners will save their results in an electronic worksheet (Ms Excel), which will be analysed automatically. A custom-made software package will extract relevant data (i.e. global and regional, longitudinal and radial, end-systolic strain and peak systolic strain rate) and prepare for statistical analysis (SPSS). All measurements will be correlated with T2* magnetic resonance imaging (MRI) data for all adult patients. T2* MRI measures myocardial iron-load.

Results/conclusions: Available in January/February 2013.

1507: RHEUMATIC MITRAL VALVE DISEASE IN CHILDREN: A BIG PROBLEM IN A DEVELOPING COUNTRY

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Rheumatic valvular heart disease remains a serious problem of public health in developing countries, being a common cause of morbity and mortality and responsible for half of the cardiovascular internments among children in Brazil.

Objective: This study was to assess the long-term results of mitral valve repair in children with chronic rheumatic heat disease.

Method: From January 2002 through July 2012, 163 patients underwent mitral valve surgery at our institution;70 (42.94%) underwent mitral valve reconstruction, 62 (38.05%) submitted to mitral valve replacement with a bioprosthesis, and 31 (19.21%) underwent a second replacement. The age varied from 3 to 16 years (mean age at surgery was 11.5 ± 3.5 years. Ninety-two (56.44%) children were female; 88 children (54%) were in the New York Heart Association functional Class III and 49 (30%) in Class IV. Reparative procedure included posterior leaflet extension with a pericardial patch in 46 (65.7%), and 24 patients (34.3%) received a Carpentier ring.

Results: In the groups of valve repair there was one early death caused by a metabolic disturbance and two children needed reoperation. One had a rupture of the plasty at 24 hours after surgery and another presented with severe haemolysis in the postoperative period; both were given mitral valve replacement. The period of follow-up extended from 6 months to 10 years. In the group of valve replacement the durability of the bioprosthesis was 16–60 months (mean 33 months).

Conclusion: Mitral valve repair in children with rheumatic heart disease is feasible and provides acceptable long-term results. In spite of the short bioprosthesis durability, in our poor population the socio-economic level must be considered in the choice of the valve substitute.

1511: ESTABLISHING RHEUMATIC HEART DISEASE CONTROL PROGRAMME IN SUDAN: ACHIEVEMENTS AND CHALLENGES

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Introduction: Rheumatic heart disease (RHD) continues to have a significant burden on the health of young people in Sudan with a prevalence of 10 per 1 000. Control programmes were initiated by the World Health Organization based on awareness and secondary prevention in 1990.

Materials and methods: The authors, inspired by the experience of the Pan African Society of Cardiology (PASCAR) and the RHD Group of the World Heart Federation, last meeting in Dubai May 2012, initiated a RHD control programme initiative based on the Awareness, Surveillance, Advocacy and Prevention (ASAP) initiative of the PASCAR.

Results: Primary prevention protocol is based on research done locally that validated a clinical algorithm for diagnosis of bacterial pharyngitis. The programme included protocols for primary and secondary care physicians as well as health assistants, conducting workshops for their training. We initiated awareness programmes for physicians, medical students, school staff and the public, and initiated a local registry with more emphasis on the primary prevention aspect. The project was approved after long discussions with Ministry of Health and the scientific societies (Sudan Heart Society and the Sudanese Association of Pediatricians). It is included in the School and Adolescent Health Program.

Conclusion: The paper throws light on the achievements and difficulties of this project.

1513: UNDERDIAGNOSED CARDIOMYOPATHIES IN THE PAEDIATRIC AGE

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Introduction: The commonest type of paediatric cardiomyopathy (CMP) is the dilated type (DCMP) which constitutes 70–80% of cases.

Materials and methods: In this report we reviewed 2 series of patients with other types of CMP which are not commonly reported but we believe that they are underdiagnosed.

Results: The first is known as tropical restrictive CMP or endomyocardial fibrosis (EMF). We found that EMF represented 18% of cases of CMP seen at a tertiary centre. We reviewed the clinical and echocardiographic features of patients with EMF and discussed differentiation of this entity from diseases that can mimic it. The second type of CMP is non-compaction CMP (NCCMP) which has recently gained a lot of attention and is thought to be underdiagnosed. We describe a series of 52 patients in Arab/African patients and it was found that NCCMP represented at least 20% of cases of CMP. Within this disease we reported unique clinical and echocardiographic associations. Female preponderance, a relapsing course and mitral valve regurgitation were distinctive features of NCCMP in our area.

Conclusion: We need to increase the awareness of physicians about these types of CMP so that appropriate treatment and counselling can be instituted.

1520: CLINICAL AND ECHOCARDIOGRAPHIC FEATURES OF CHILDREN WITH RHEUMATIC HEART DISEASE AND THEIR SERUM CYTOKINE PROFILE

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Introduction: Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) constitute important public health problems in developing countries.

Materials and methods: Children with ARF and RHD seen at Children's Hospital, Sudan from May 2008 to 2009 were examined

clinically and by echocardiography. Blood cytokines (interleukin-10 (I-10), tumour necrosis factor alpha (TNF-alpha) and interferon gamma (IFN-gamma) were done.
Results: Thirty six children were enrolled; 63% had established RHD, and 37% ARF. Mitral regurgitation (MR) was the most common lesion (94%). Ninety five per cent of the valve lesions were severe. The serum I-10 level ranged between 3 and 6 pg/ml. TNF-alpha levels were 9–100 pg/ml in 12 patients (40%), 101–1 000 pg/ml in 10 patients (33%), and more than 1000 in 8 patients (26%). The level of IFN-gamma ranged between 2 and 7 pg/m in all patients except 2 (84 and 135 pg/ml).
Conclusion: RHD manifests with severe valvular lesions and a high TNF-alpha indicating an ongoing inflammation.

1531: THE ARISTOTLE COMPREHENSIVE COMPLEXITY SCORE PREDICTS MORTALITY AFTER SURGICAL LIGATION OF PATENT DUCTUS ARTERIOSUS IN PRETERM INFANTS

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Background: The outcome measure after surgical ligation of patent ductus arteriosus (PDA) in preterm infants is often complicated by prematurity-associated co-morbidities. The Aristotle comprehensive complexity score (ACCS) has been proposed as a useful tool for complexity adjustment in the analysis of outcome after congenital heart surgery. The aims were to define preoperative risk factors for mortality and to demonstrate the usefulness of ACCS to predict mortality after surgical ligation of PDA in the preterm.
Material and methods: Included were 49 preterm babies (< 35 weeks' gestation), from May 2009 to July 2012, who had surgical ligation of PDA. Mean (range) gestational age and birth weight were 28 weeks (23–35 weeks) and 1 102 g (520–2 280 g), respectively. Mean (range) age and weight at operation were 16 days (4–44 days) and 1 199 g (400–2 880 g), respectively. Initial oral ibuprofen was ineffective in 24 patients and contraindicated in 25. All surgical ligations were done at bedside in the neonatal intensive care unit. Preoperative clinical and laboratory profiles were reviewed and ACCS was derived.
Results: Eight out of 49 (16.3%) died after a median of 14 days (2–73 days) after PDA ligation. Patients who had contraindications for oral ibuprofen (odds ratio (OR) 8.94; $p = 0.049$), coagulopathy (OR 12.13; $p = 0.025$), renal dysfunction (OR 28.88; $p = 0.003$), intraventricular haemorrhage ≥ grade III or seizure within 48 hours of operation (OR 34.00; $p = 0.002$), and ACCS > 15 (OR 415.00; $p < 0.001$) were significantly associated with an increased risk for mortality. At multivariate logistic regression analysis, ACCS > 15 was the only independent risk factor (OR 103; $p = 0.023$) for mortality.
Conclusions: The ACCS, especially procedure-independent complexity factors, is a useful tool to predict mortality after ligation of PDA in the preterm.

1532: COARCTATION OF THE AORTA IN INFANTS: CLINICAL, THERAPEUTIC AND PROGNOSTIC IN A SERIES OF 52 CASES

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Objective: To describe the clinical and therapeutic characteristics of 52 infants undergoing surgical repair of coarctation of the aorta.
Material and methods: A total of 52 infants under 1 year of age underwent surgical correction of coarctation of the aorta from July 2000 to January 2012.
Results: Thirty-nine (75%) children were male. The median age at diagnosis was 30 days, ranging from 20 days to 1 year. Regarding the onset of clinical manifestations, in 20 (51.9%) cases they occurred within the first month of life, in 13 (25%) cases from the first to the sixth month of life, and in 19 (36.5%) cases between the sixth month and the first year of life. The most significant finding was absence or reduction of arterial pulses in the lower limbs in 52 patients and all clinical signs of congestive heart failure in 46 (88.5%). Cardiomegaly was a finding common to all children. There was a predominance of right ventricular overload in half the cases. The echocardiographic study showed an association of coarctation of the aorta and other intracardiac abnormalities in 24 (46.1%) of cases, with the bicuspid aortic valve and ventricular septal defect (VSD) more frequent. Ventricular systolic dysfunction was observed in all children. Regarding the surgical procedure, angioplasty of the left subclavian artery flap was performed in 46 (90.2%) cases, which is associated with pulmonary artery banding in four (7.8%) and angioplasty with bovine pericardium was performed in four (7.8%) cases. Paradoxical hypertension was observed in 44 (84.6%) cases. The mortality rate was 11.5%.
Conclusion: In our series there was a predominance of congestive heart failure as the initial manifestation of the disease, underscoring the importance of pulse palpation in the lower limbs as a means of early detection of this anomaly.

1533: IMPROVING CARDIAC PATIENT EDUCATION IN A MULTI-LINGUAL SOCIETY

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Background: Steve Biko Academic Hospital is the cardiac referral centre for four of the nine provinces in South Africa, including two with large rural populations, namely Limpopo and Mpumalanga provinces. As such, our patients speak a wide selection of the eleven official languages. The occasional foreign national also finds their way to our doors and it is not unusual for as many as eight different languages to be spoken on any outpatient clinic day. Many of the parents and caregivers we see are also illiterate or only have primary level literacy skills.

This provides a challenge to the cardiologist who has to explain often very complex congenital cardiac defects to patients and parents who do not speak his/her home language or languages. Adequate informed consent for procedures must also be obtained and patient compliance with treatment schedules and dosages, which often change from visit to visit, must be ensured in order to offer the child the best care and management possible. Skilled, informed interpreters are not always readily available, so we have had to find other methods of getting our message across. We present some of the teaching aids and materials that we use in our unit to improve communication with our patients and their caregivers.

1536: GEOGRAPHICAL INFORMATION SYSTEMS (GIS) AS AN AID TO INVESTIGATING CONCORDANCE PATTERNS AMONG RHEUMATIC HEART DISEASE (RHD) PATIENTS: A PILOT PROJECT

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Background: Lack of concordance to monthly international normalised ratio (INR) monitoring in RHD patients is considered to be a contributor to stroke and other complications. Various factors for this non-concordance have been suggested including limited access to health care facilities. We hypothesise that the rate of default in patients is correlated with the travel distance from the patient's residence to the health care facility to which they have been referred. GIS

enables the visual representation of data so as to establish patterns and trends between patients' follow-up visits for INR monitoring and their geographical context. The aim of this study was, therefore, to investigate these associations by developing a spatial display instrument, mapping the residential addresses of patients against their respective designated referral clinics.
Methods: Patient data including residential addresses, referral clinics and concordance patterns were obtained from the REMEDY database. REMEDY is an RHD registry targeting 3 000 patients in its pilot phase. Addresses were converted to geographical coordinates and ArcGIS 9.3.1® was used for mapping and spatial analyses. The Geocoded addresses were further checked in ArcGIS for errors in coordinate data. The travel distances between individual residential addresses and referral clinics were calculated and compared to concordance patterns of patients.
Results: RHD patients (n = 26) reside between 1.2 and 26.2 km from their referred clinics (mean 9.68 ± 8.1 km). The average period between clinic visits was 32 days (range 18–43 d). Preliminary results suggest that concordance to monthly INR monitoring is not associated with patients' travel distance.
Discussion: This is the first attempt at the application of GIS within this area of RHD. Distance from referred clinic does not impact upon patients' attendance at INR monitoring. Nevertheless, GIS presents as an ideal tool to visual relationships between RHD patients' follow-up visits for INR monitoring and their geographical context.

1538: RISK FACTORS ASSOCIATED WITH THE DEVELOPMENT OF SYSTEMIC-PULMONARY COLLATERAL FLOW IN SINGLE VENTRICLE PATIENTS WITH SUPERIOR CAVOPULMONARY CONNECTIONS

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Background: Systemic-pulmonary collateral flow (CollF) is common in patients after superior cavopulmonary connection (SCPC). Risk factors associated with CollF are unclear. We sought to identify risk factors for CollF in a cross-section of patients with SCPC.
Methods: A retrospective chart review of events from birth to the time of study was performed for SCPC patients who had CollF quantified by cardiac magnetic resonance imaging (CMRI). CollF was reported as indexed flow (l/min/m^2), as a percentage of aortic flow (CollF/Ao), and as a percentage of pulmonary venous flow (CollF/PV).
Results: From 4/08 to 8/11, 96 SCPC patients at 2.6 ± 1.1 years of age and 799 ± 400 days from SCPC had CollF measured at 1.6 ± 0.7 l/min/m^2, comprising 33.5 ± 11.1% of aortic flow and 48.3 ± 15.9% of pulmonary venous flow. Two of three CollF indices were higher in patients with a prior BT shunt vs no prior BT shunt (pulmonary artery band, right ventricle-pulmonary artery conduit, or no initial palliation) (CollF 1.7 ± 0.8 vs 1.4 ± 0.4 l/min/m^2, p = 0.04; CollF/Ao: 35.2 ± 11.6 vs 28.6 ± 7.6%, p = 0.009) and in those with a hemi-Fontan vs bidirectional Glenn (CollF/Ao 38.5 ± 13.5 vs 32.2 ± 10%, p = 0.02; CollF/PV: 57.6 ± 20.1 vs 45.8 ± 13.8%, p = 0.003). With Spearman testing, positive correlations exist between CollF indices and indicators of perioperative morbidity. We did not find associations between CollF and pulse oximetry, haemoglobin, or haemodynamics at pre-stage 2 catheterisation.
Conclusion: CollF occurs commonly in patients with SCPC and is related to surgical history, chest tube and hospital duration. These data support the hypotheses that perioperative morbidity and pleural inflammation may play a role in the development of CollF.

1540: POSTMORTEM IMAGING IN PAEDIATRIC CARDIOLOGY

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Background: Postmortem imaging (PMI), used widely in forensic pathology, could be substituted for autopsy in paediatric cardiology patients, increasing effective autopsy rate and facilitating quality improvement.
Materials and methods: Paediatric cardiology patients who died during the study period and in whom an autopsy was planned were eligible. PMI included magnetic resonance imaging (MRI) of heart, brain and abdomen (table), and multi-detector computed tomography (MDCT) angiography, using iodinated contrast diluted 1:8 with PEG, injected in the inferior vena cava (8 ml/kg) and aorta (5 ml/kg).
Results: During a 4-month period 5 patients underwent PMI followed by autopsy; both MDCT and MRI in 4 and MDCT only in 1 because of metal ECMO cannulas. Cardiac anatomy, condition of surgical repair, and abdominal anatomy by PMI correlated perfectly with autopsy. All major intracranial findings (intracranial haemorrhage, parietal infarct, white matter atrophy) were diagnosed by both PMI and autopsy. Cause of death by PMI and autopsy was congruent in 3 cases: not determined in 2 cases and intracranial haemorrhage with herniation in 1 case. PMI (MDCT only) showed severe bilateral pulmonary injury, found to be infarction at autopsy, but missed left ventricle and pulmonary artery thrombosis. PMI missed diffuse left and right ventricular infarction seen by histology in 1 case. MRI and MDCT were complementary, with MRI better for delineation of cardiac and abdominal anatomy, and brain abnormalities, but MDCT was superior for delineation of central vasculature, especially the coronary arteries, and pulmonary disease. Position of lines and tubes, condition of the body wall and skin, and skeletal anatomy were demonstrated by the MDCT 3D data set.
Conclusions: Our preliminary study in human cadavers shows that PMI is feasible in a paediatric hospital environment. The small sample size precludes analysis of diagnostic accuracy. Nevertheless, we are encouraged by the diagnostic performance of PMI.

1542: POSTMORTEM IMAGING OF ANTEMORTEM MYOCARDIAL ISCHAEMIA

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Background: Postmortem magnetic resonance imaging (PMRI) has been used in forensic pathology to detect antemortem ischaemia but the sensitivity is unknown. We tested the survival time required for detection of antemortem ischaemia by PMRI in a pig carcass model.
Materials and methods: Nine pigs (7–35 kg) underwent surgical ligation of the distal left anterior descending (LAD) (8) and/or right coronary (RCA) branch (4) and were euthanised 1–6 hours after ligation. PMRI (T1, T2, PD, and spin echo 3D volumetric sequences) was performed 12–48 hours after euthanasia. Images were inspected, and signal intensity of 17 myocardial segments was measured serially. Heart sections were submitted for histology.
Results: Ligation produced discoloration and dyskinesis of the target segment(s) in all cases. MRI T2-weighted sequences (TE 102, TR 4000) showed the ischaemic area as hyperintense in 4/4 LAD ligations with ≥4 hours of ischaemic time and in 0/4 with < 4 hours.

Histological evidence of ischaemia was present in 4/4 after 4 hours. Right ventricular ischaemia was visible on MRI after 6 hours of ischaemic time in 1 animal. The right ventricular myocardium is thinner than the left ventricular myocardium, making it difficult to distinguish myocardium from epicardium. Intraobserver variability for MRI signal intensity of the myocardium was 0.93 (95% confidence interval (CI) 0.91–0.94) for the LV and 0.18 (95% CI 0.03–0.37) for the RV. The percentage difference in signal intensity between ischaemic and normal myocardium was greatest on T2-weighted images up to 12 hours after death and then decreased with increasing time, approaching that of the non-ischaemic areas by 24–48 hours.
Conclusions: Ischaemic lesions of the LV, but not the RV, at least 4 hours old can be reliably detected as hyperintense areas on T2 weighted PMRI. By 24–48 hours after death, surrounding myocardial density approaches that of the ischaemic area obscuring it.

1544: SPECTRUM AND OUTCOME OF PAEDIATRIC PATIENTS UNDERGOING RIGHT VENTRICULAR PACING

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Background: Pacemaker implantation (PMI) in the paediatric population is uncommon with implantation performed in selected patients with congenital heart block or following cardiac surgery.
Aim: To evaluate the spectrum and outcomes of patients undergoing right ventricular (RV) pacing in the centre's paediatric population.
Method: A retrospective chart review of all paediatric patients undergoing PMI since 2003 .
Results: Seventeen patients were identified; 12 had congenital complete heart block (CCHB), of whom 2 had associated structural abnormalities and 5 were heart block post cardiac surgery. Age ranged from 7 days to 13 years. All implants were single-chamber rate-responsive devices with 82% epicardially placed and 17.6% endocardially. Epicardial placement was initially on the RV free wall except for the last 3 which were placed on the RV outflow tract. Four early mortalities in neonates with CCHB were recorded with one late mortality. Associated morbidity in these patients included: 1) hydrops fetalis in a patient with left isomerism and *total anomalous pulmonary venous connection* (TAPVC); 2) long QT syndrome with poor ventricular function post pacemaker implantation; and 3) Gram-negative sepsis 9 days after PMI. Sudden unexpected death shortly after PMI occurred in one neonate. Late mortality occurred from pneumonia in a 14-month-old child with right isomerism/AVSD who underwent PMI and pulmonary artery banding at 4 months of age. Two patients with progressive heart failure at follow-up attributed to pacemaker-induced cardiomyopathy were upgraded to biventricular pacing with good results.
Conclusion: While patient numbers were insufficient to reach any clear conclusion, patients undergoing RV PMI had good outcomes, the exception being those with heterotaxy syndrome and neonates with comorbidity. Alternative pacing techniques to avoid or manage pacemaker-induced cardiomyopathy may be indicated and requires further investigation.

1545: QUANTITATIVE GATED SPECT VALUABLE TO EFFECTIVENESS OF CARDIAC RESYNCHRONISATION THERAPY IN FUNCTIONALLY SINGLE VENTRICLE

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Background: Cardiac resynchronisation therapy (CRT) is effective treatment for severe left heart failure. Recently, CRT has increasingly been used in children and congenital heart disease (CHD) patients, but published data about effectiveness are limited in comparison with data on adults with left heart failure. Furthermore, for CHD patients except cardiomyopathy, it is hard to evaluate the outcome.
Methods: We describe a case of a 35-year-old woman with functionally single ventricle who started CRT 4 years ago because of failing Fontan. At baseline and after CRT, clinical status by New York Heart Association (NYHA) class, ejection fraction (EF), ventricular dyssynchrony using quantitative gated single-photon emission computed tomography (QGS) from Cedars-Sinai QGS software was evaluated. Improvement of NYHA class and EF was considered response. With 99mTc-sestamibi, end-diastolic volume (EDV), end-systolic volume (ESV), ventricular EF, summed motion score (SMS), and summed thickening score (STS) were calculated with QGS.
Results: We could find improvement of EDV (210 ml to 186 ml), ESV (173 ml to 120 ml), EF (18% to 35%), SMS (32 to 7), STS (27 to 13) and uptake of nuclide to the myocardium. Phase analysis derived from QGS polar map was indicated as heterogeneous distribution in pre-CRT, and it was improved at follow-up. This case showed improvement in NYHA class and EF.
Conclusions: The radionuclide study is a useful examination to evaluate synchrony because it is reproducible, simple and visual.

1562: SEVERE LEFT VENTRICULAR INFLOW OBSTRUCTION IN AN INFANT - AN UNUSUAL COMPLICATION OF DILATED CORONARY SINUS: A CASE REPORT AND REVIEW OF LITERATURE

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Introduction: Dilated coronary sinus secondary to persistent left superior vena cava is a common entity. However, dilated coronary sinus causing severe left ventricular inflow obstruction is an infrequent finding. We report on a 4-month-old infant who presented with congestive cardiac failure, secondary to marked enlargement of coronary sinus with consequent obstruction of left ventricular inflow.
Case report: A 4-month-old infant was referred to our paediatric tertiary hospital with history of poor weight gain, breathlessness during feeding and frequent sweating. Cardiovascular examination revealed right parasternal heave with loud diastolic murmur at mitral area. Electrocardiograph (ECG) showed right axis deviation with marked right ventricular hypertrophy. Chest roentgenograms showed mild cardiomegaly and passive pulmonary venous congestion. 2D echocardiogram suggested diagnosis of cor triatriatum. However, acquisition of stereoscopical 3D volume dataset identified the exact nature of the obstruction and revealed persistent left superior vena cava and markedly dilated coronary sinus causing severe narrowing of inflow pathway of left ventricle. Operative finding confirmed the diagnosis with presence of markedly dilated coronary sinus causing narrowing of left ventricular inflow and absence of any membrane in left atrium. He underwent reduction plasty of coronary sinus which included longitudinal incision of coronary sinus, excision of a segment of its wall and reconstruction of the dilated coronary sinus wall. Excellent result was achieved with no residual left ventricular inflow obstruction at follow-up.
Conclusion: Dilated coronary sinus is a rare but possible aetiology for left ventricular inflow obstruction. Reduction plasty of coronary sinus is a safe and effective procedure for treating this entity. In addition, stereoscopical 3D is an effective alternative to conventional catheterisation for diagnosing this entity.

1565: INVESTIGATION OF IDIOPATHIC DILATED PULMONARY ARTERY: THE ROLE OF DIFFERENT NON-INVASIVE IMAGING MODALITIES: A CASE REPORT

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Case description: A 14-year-old Chinese girl with Noonan syndrome, mild mental retardation and left renal agenesis was followed up for idiopathic pulmonary artery dilatation. At 7 years old, her echocardiogram and right heart catheterisation showed idiopathic pulmonary artery dilatation.

Trans-thoracic echocardiogram was repeated at 14 years of age. It showed dilated main pulmonary artery, measuring 3.38 cm in diameter and dilated left coronary artery. The left main coronary artery measured 6.2 mm and the left anterior descending artery measured 5.2 mm in diameter. There were multiple low velocity turbulent jets in the mid right ventricular free wall. There was no evidence of pulmonary hypertension from Doppler pressure gradient assessment of pulmonary regurgitation.

Left and right cardiac catheterisation was performed with a suspicion of coronary arteriovenous fistula to the right ventricle. Left coronary angiogram did not show any fistula but retrograde contrast flow to the right coronary artery. Attempt to cannulate the right coronary artery was unsuccessful. Detailed review of the angiogram showed suspicious anomalous origin of right coronary artery from the main pulmonary artery but we could not confirm the diagnosis by manipulating a catheter in it.

A computed tomography (CT) coronary angiogram with 3D reconstruction was later performed. It showed beautifully the anomalous origin of the right coronary artery from the main pulmonary artery. Magnetic resonance imaging (MRI) perfusion scan with adenosine stress test showed no evidence of myocardial ischaemia. The patient was treated conservatively.

Conclusion: One should be careful with the diagnosis of idiopathic pulmonary artery dilatation. In this case, the combined role of non-invasive imaging including echocardiogram, CT coronary angiogram and MRI perfusion scan can replace traditional invasive cardiac catheterisation to reveal the true underlying cause.

1571: COURSE OF THE DFT IN THE MIDTERM FOLLOW-UP AFTER ICD-IMPLANTATION USING THE EXTRACARDIAC TECHNIQUE

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Introduction: Up to now, there is limited experience in *implantable cardioverter defibrillator* (ICD) implantation in infants and small children. The previously described extracardiac technique offers an effective and safe ICD implantation technique in these patients. However, data about midterm follow-up, especially the course of the defibrillation thresholds (DFT), during the growth of the patients are lacking.

Patients and methods: An extracardiac ICD-system was implanted in 19 patients (mean age: 5.6 [range 0.2–9.8] years) as previously described. In brief: Under fluoroscopic guidance a defibrillator lead was tunnelled subpleurally along the course of the 2-6th rib until almost reaching the vertebral column. After a partial inferior sternotomy, bipolar steroid-eluting sensing and pacing leads were sutured to the atrial wall in selected patients and to the anterior wall of the right ventricle in all patients. The ICD device was implanted as 'active can' in the upper abdomen. Sensing, pacing, and DFT as well as impedances were verified intraoperatively, 3 months later and then every 12 months.

Results: In all patients, intraoperative DFT between the extracardiac lead and device was < 15 J (mean 11.2 J). No serious complications were noted. After a mean follow-up of 1.8 years DFT remained stable or below < 20 J in 15/19 patients. Revision was required in 3 patients because of unacceptable DFT > 20 J. Using multiple regression analysis a significant correlation between the size of the patient and mean DFT was noted. In 3 patients inadaequate ventricular sensing was noted. Revision of the system could be avoided in 2/3 patients by an alternative sensing option between the ventricular leads and subpleural shock electrode.

Conclusions: The extracardiac technique offers a safe and effective approach for ICD implantation in infants and small children. However, regular DFT testing is mandatory to recognise failure of the system.

1573: RISK FACTORS AND NATURAL HISTORY OF ISOLATED NON-COMPACTION MYOCARDIOPATHY IN CHILDHOOD

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Background: Left ventricular non-compaction (LVNC) is an uncommonly reported genetic disorder of endocardial morphogenesis. Clinical and prognostic heterogeneity is described. The purpose of this study was to identify the clinical characteristics and risk factors of children with LVNC.

Methods and result: We retrospectively reviewed 24 children with LVNC evaluated at the Garrahan Children's Hospital from January 1996 to December 2011. The median age at presentation was 2 years (range 15 days to 13 years). Median follow-up was 4 years (2–15 years). Seventeen patients (70%) had electrocardiograph (ECG) abnormalities, 6 patients left bundle branch block (LBBB), two patients Wolff-ParkinsonWhite syndrome (WPW). Both ventricles were involved in 8 patients (33.3%) and only the left ventricle in 16 patients (69.1%). Left ventricular systolic function was depressed in 12 patients (50%), with a median ejection fraction of 30% (20–66%) at diagnosis. Of 9 patients who presented with depressed left ventricular contractility, 66% had functional recovery. During follow-up, 8 patients (33.3%) suffered an adverse event. Five patients with biventricular and three with left ventricular compromise were put on the heart transplantation programme and two required a ventricular assistance device.

Conclusion: In this cohort of patients with LVNC 69.9% remain in functional class I–II. Risk factors for adverse events, heart transplantation or death were biventricular involvement ($p < 0.05$) and conduction disturbances ($p = 0.001$).

1574: PERIPHERAL MICROVASCULAR FUNCTION IS AFFECTED IN YOUNG INDIVIDUALS AT RISK FOR HCM AND CORRELATES WITH FINE CHANGES IN MYOCARDIAL FUNCTION AND ELECTROPHYSIOLOGY

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Background: Recent data from our centre suggested that advanced electrocardiograph (ECG) analysis combined with tissue Doppler (TD) has increased sensitivity in identifying individuals at risk for developing hypertrophic cardiomyopathy (HCM), and in distinguishing HCM from athlete's heart. Adult patients with HCM appear to have functional abnormalities not only in coronaries but also in peripheral vessels. Whether the latter are present in young individuals, and whether they can be used as an additional index for stratifying the risk for HCM have not been yet studied.

Methods: The study population, part of a cohort from an ongoing prospective study, consisted of children and adolescents (age 4–27, median 13.9 years) with familial HCM heredity, either without left

ventricular hypertrophy (LVH) (HCM-risk: z-score for IVS and/or PW < 2.5 SD; $n = 10$) and HCM-patients with LVH (HCM: z-score > 2,5 SD; $n = 10$), and compared to age- and gender-matched healthy controls ($n = 20$) and athletes ($n = 10$, endurance physical exercise > 10 hours/week, z score > 2 SD). Conventional ECHO, TD, advanced 12-lead ECG (A-ECG) for calculation of spatial mean QRS-T angle by Kors (Cardiax®, IMED Co Ltd, Budapest/Hungary and Houston/USA) were used. Cutaneous microvascular responses to acetylcholine (Ach) and sodium nitroprusside were assessed by laser Doppler with iontophoresis.

Results: As compared to controls and athletes, the micro-vascular responses to Ach were increased in patients with HCM and LVH, and also in those at risk for HCM (i.e. HCM heredity without LVH; $p = 0.09$). In this latter group, micro-vascular responses to Ach correlated significantly with both spatial mean QRS/T angle by Kors and E/E´ ratio by TD ($p < 0.05$, $r > 0.3$). With the exception of left ventricular mass (LVM) and myocardial thickness, which were increased in both athletes and HCM patients, there were no differences in any other measured variable between controls and athletes.

Conclusion: Independent to the presence of LVH the presymptomatic HCM-risk individuals and HCM patients demonstrated enhanced peripheral micro-vascular reactivity, probably reflecting compensatory vasoactive mechanisms in response to HCM-related noxious factors. The enhanced peripheral micro-vascular reactivity is present in both presymptomatic HCM-risk individuals and HCM patients, but not in athletes or normal controls, and seem to be associated with fine abnormalities in myocardial electrophysiology and function.

1575: COMPLICATION CAPTURE TO ENHANCE PAEDIATRIC CARDIAC SURGERY OUTCOMES

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Background: With current low mortality rates in paediatric cardiac surgery, we can now focus on reducing complications to enhance patient outcomes. In the absence of a risk-adjusted score for morbidity, we have developed a system of tracking programme-wide complications. The system generates a flag at the time of any excess observed:expected (O:E) complications.

Methods: Consecutive procedures and 30-day complications have been recorded prospectively at the IWK Heart Centre since 2009, using the Multi Institutional Database Committee definitions. Inclusion criterion was a procedure with an assignable risk adjusted congenital heart surgery (RACHS) category, which includes 70% of cases. A morbidity score was generated for each procedure factoring in frequency and severity of complications. Observed outcomes were collected (2011–12) and expected outcomes were calculated based on historic complications (2009–2011). An overall O:E plot was generated case-by-case. Complications were also stratified by RACHS category and organ-system. Flags were generated to indicate unexpected complication frequency or severity.

Results: In 2011–12, 116 procedures were performed on 106 patients, 86 of whom had an assignable RACHS category. The operative mortality was 5/116 (4.3%). Ninety-two complications occurred in 31 procedures. The overall O:E plot was flagged 4 times as a result of 9 complications (arrhythmia, neurologic and pulmonary). The arrhythmia flags were generated by spontaneously resolving arrhythmias in 3/4 cases. The single neurologic flag resulted from a subdural haematoma in a procedure requiring preoperative ECMO. Pulmonary flags were generated in 4 neonatal procedures (1 reintubation and 4 long-term intubation). There was excess morbidity in RACHS category 4 and 6.

Conclusions: This complication capture and stratification system allows real-time identification of excess complication occurrence. We anticipate that the ability to detect complications will allow focused changes to improve patient outcomes.

1576: DUAL ANTIPLATELET THERAPY IN PAEDIATRIC CARDIOVASCULAR PRACTICE

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Introduction: Antiplatelet drugs are increasingly used in paediatric patients to prevent thromboembolic episodes, as an alternative to warfarin. Aspirin remains the most frequently used antiplatelet agent; however there has been an increased interest in the use of newer agents. Clopidogrel is an antiplatelet drug that acts by irreversibly modifying the adenosine diphosphate (ADP) purinergic platelet receptor. Clopidogrel use in children has been reported principally by American centres, predominantly in patients with congenital cardiac disease. However, the dosage, duration and indications vary widely. Experience in the UK remains very limited.

Objective: The aim of the study was to assess the indication, dosage, duration of therapy and safety of dual antiplatelet therapy with aspirin and clopidogrel in paediatric patients with cardiovascular disease.

Methods: The study included 21 consecutive children (with cardiovascular disease, presenting over a 2-year period with an indication for anticoagulation (excluding mechanical valve replacement), who were treated with aspirin and clopidogrel. Patient demographics (12 male), median (range) (age 4.08 (0.02-13.61) years), diagnosis, indication, dose (clopidogrel 0.3 (0.18-1.12) mg/kg; aspirin 4.46 (1.6-9.44) mg/kg), duration of treatment (23.14 (0.57-101.3) weeks) and adverse events were recorded. Indications for combination use were systemic-pulmonary artery shunt (7), Fontan (7), Glenn shunt (3), stent (3) and others (3).

Results: There were 2 adverse events:

- 4-year-old boy with gastrointestinal bleed, 7 days after starting clopidogrel at 0.8 mg/kg/day. An endoscopy confirmed a gastric ulcer. He was also taking aspirin 75 mg (4.17 mg/kg) and has now been recommenced on warfarin.
- 6.5-year-old girl with haemorrhagic cerebrovascular accident 5 days after starting clopidogrel at 0.2 mg/kg/day. MRI showed arteriovenous malformations in her brain. Her aspirin was restarted at 50 mg (2.83 mg/kg) with no further adverse events.

Discussion: Combined use of aspirin and clopidogrel is increasing among children. Compared with warfarin, dual antiplatelet therapy has advantage of ease of administration, with no monitoring required. However there is limited knowledge of the bleeding risks associated with dual antiplatelet therapy in children. This study highlights the need for caution and vigilance in treating children with clopidogrel in combination with aspirin, especially at higher clopidogrel doses.

1578: CHANGES IN MESENTERIC FLOW, HEART RATE VARIABILITY AND SYSTEMIC MICROVASCULAR FUNCTION AFTER TOTAL CAVOPULMONARY CONNECTION-RELATIONSHIP TO PULMONARY HAEMODYNAMICS AND POSTOPERATIVE PLEURAL EFFUSION

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Background: Prolonged pleural effusion (PE) after total cavopulmonary connection (TCPC) remains a serious complication with important clinical and economic impact. Downstream pulmonary action of mediators released in response to abnormal circulation in the gut along with changes in cardiac autonomic nervous activity have been suggested among possible mechanisms. We aimed to assess this hypothesis by studying in a prospective manner the profiles of mesenteric flow, systemic microvascular function, and cardiac autonomic function derived from the analysis of heart rate variability (HRV).

Methods: Fifteen patients (median age at surgery: 2.1 yrs) with univentricular heart physiology and previous Glenn surgery were

assessed serially before and after TCPC (extracardiac conduit in all patients), until discharge, for superior mesenteric flow velocities, systemic micro-vascular function via laser Doppler (baseline and reactive skin perfusion in response to acetylcholine (Ach) and sodium nitroprusside), and for certain HRV indices (standard deviation of the normal-to-normal RR intervals (SDNN), LF and HF; BioMedical Inc, Taiwan).
Results: The mesenteric diastolic flow VTI and the micro-vascular perfusion and reactivity to Ach decreased postoperatively especially on day 1 after surgery ($p < 0.05$). HRV LF and HF were reduced postoperatively at all time points ($p \leq 0.05$). The mesenteric diastolic flow velocity on day 1 correlated with the preoperative transpulmonary gradient ($p = 0.02$, $r = -0.7$), and both with the duration of PE ($p < 0.05$, $r > 0.4$). Lower preoperative values of SDDN were associated with higher preoperative transpulmonary gradients ($p = 0.01$, r = -0.8), and to some extent with PE duration ($p = 0.06$, $r = -0.6$). Neither the demographic variables (age at surgery, time between Glenn and TCPC, weight) nor the heart-lung machine duration were associated with PE duration.
Conclusion: TCPC is ensued by abnormalities in mesenteric flow, systemic micro vascular function and cardiac autonomic function. Although several of these variables show some association with PE duration, given the small sample size, further studies are needed to validate these findings and to interrogate the mechanisms.

1581: HOW IS THE PERI-PATCH MYOCARDIUM IN VENTRICULAR SEPTAL DEFECT (VSD) PATCH REPAIR

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Introduction: Strain rate (SR) and strain (ε) in tissue Doppler imaging provide new non-invasive measurements of myocardial function, independent of heart motion. This study assesses the extent of peri-patch regional myocardial function after patch repair of ventricular septal defect (VSD).
Method and results: Myocardial SR and ε were recorded from the peri-patch myocardium and remote septum from patch area in 20 patients (1 month to 10 years of age, mean 2.6 years). Distance between the patch and the point of returning to remote normal ε profile was measured. Compared to the remote myocardial region, peri-patch myocardium had decreased peak longitudinal SR ($-3.3 \pm 1.8\ s^{-1}$ vs $-4.8 \pm 3.3\ s^{-1}$, $p < 0.05$), delayed time to peak longitudinal SR (151 ± 81 ms vs 119 ± 56 ms, $p < 0.05$), decreased peak ε (longitudinal $-20 \pm 8\%$ vs $-28 \pm 10\%$; radial 20 ± 14 % vs $34 \pm 22\%$, $p < 0.01$), and delayed time to peak ε (longitudinal 316 ± 76 ms vs 241 ± 67 ms; radial 341 ± 94 ms vs 269 ± 81 ms, $p < 0.0001$). The mean distance from the patch to the remote patch ε curve was 2.65 ± 0.77 mm.
Conclusion: Peri-patch myocardium after repair of VSD has delayed and diminished contraction as compared to more remote normal myocardium.

1595: GEOMETRIC CHARACTERISATION OF 100 PATIENT-SPECIFIC TOTAL CAVOPULMONARY CONNECTIONS AND THEIR RELATION TO HAEMODYNAMIC OUTCOMES

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Background: The total cavopulmonary connection (TCPC) anatomies are complex and have great patient-to-patient variability. The geometric characteristics can impact the haemodynamic outcomes, namely: (1) TCPC energy dissipation which can affect ventricular function; and (2) unbalanced hepatic flow distribution (HFD) which increases the risk of pulmonary arteriovenous malformations (PAVM). In this study, we present a large-scale cohort geometric characterisation of different TCPCs and explore how the anatomical differences translate to the resulting haemodynamic outcomes.
Methods: A total of 100 patient-specific TCPC 3D anatomies were reconstructed from magnetic resonance images (MRI). Diameter, shape factor (SF), minimum/maximum diameter and tortuosity (vessel length/shortest distance between the two ends of a vessel) were quantified at each vessel. Angles between connecting vessels and caval offset (distance between the superior vena cava (SVC) and the Fontan pathway) were also quantified. Computational fluid dynamics simulations were carried out using time-averaged flow boundary conditions (obtained from phase contrast MRI) to quantify the resulting TCPC energy dissipation index (TCPC-EDI) and HFD, which were then correlated to the geometric parameters; $p < 0.05$ was considered significant for all statistical correlations.
Results: TCPC-EDI correlated negatively with normalised diameters at all TCPC vessels, and also SFs of SVC, RPA (right pulmonary artery) and the Fontan pathway. Cardiac index was found to have positive significant correlations with Fontan pathway SF, and normalised SVC and Fontan pathway diameters. HFD was found to correlate with the pulmonary flow distribution, normalised PA diameter, caval offset and angles across the connections. No significant correlations were found between tortuosity and the haemodynamic endpoints.
Conclusions: Small vessel diameters and SFs are correlated with higher TCPC-EDI and lower cardiac index. This highlights the importance of dilating stenosis in any associated vessel of the connection. Also, PA diameter, caval offset and connection angles together can impact HFD.

1599: MANAGEMENT OF DEXTROCARDIA, SITUS INVERSUS TOTALIS, MIXED TYPE TAPVC, COMPLETE AV CANAL DEFECT, COMMON ATRIUM, BILATERAL SVC, PDA, SEVERE PAH'

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Objective: To present the management of dextrocardia, situs inversus totalis, TAPVC – mixed, complete atrioventricular (AV) canal defect, common atrium, bilateral SVC, patent ductus arteriosus (PDA), severe pulmonary arterial hypertension (PAH).
Diagnosis: This 9-month-old girl baby girl was admitted with failure to thrive and respiratory distress. Echo evaluation revealed the above-mentioned diagnosis. After preoperative evaluation, surgery was performed. Operative finding included mixed type TAPVC – right superior and middle pulmonary veins and left inferior pulmonary vein were draining into common venous chamber. A vertical vein from the common chamber was joining RSVC, and a left superior vein to LSVC. The right upper lobe vein joined the RSVC. Morphologic right atrium (RA) was on right side receiving RSVC and hepatic vein. Morphologic left atrium (LA) was on the left side receiving LSVC and IVC. Other findings were: unroofed coronary sinus; complete AV canal defect – Rastelli Type A with large inlet ventricular septal defect (VSD); severe mitral regurgitation (MR) – anterior mitral leaflet (AML) cleft; common atrium.
Results: The procedure included TAPVC repair & AV canal repair. Rerouting of pulmonary veins was done with wide anastomosis between the common venous chamber and mLA. Vertical vessel was ligated .Since innominate vein was present, LSVC was interrupted, hence diverting left superior vein to mLA. Regarding AV canal repair, a double patch technique was used. VSD closure, MV repair with AML cleft repair, tricuspid valve repair, atrial septal defect (ASD) closure with Pericardia patch created an atrial baffle, diverting IVC, RSVC and hepatic vein to mRA. Core cooling was

done to 28°C. Total CPB time was 325 min and aortic cross clamp time 235 min. Total ventilatory support was given for 86 hours and inotropes for 6 days. Intensive care unit (ICU) stay was until the 8th post-operative day (POD).
Conclusion: Baby was discharged on 16th POD with stable haemodynamics and on normal oral feeds.

1602: DELAYED RECOGNITION OF UNILATERAL PULMONARY VEIN OBSTRUCTION AFTER REPAIR OF COR TRIATRIATUM

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Background: Cor triatriatum is a rare condition where the left atrium (LA) is subdivided by a membrane. Although cor triatriatum can be an isolated lesion, in many cases, it is associated with other congenital cardiovascular anomalies: atrial septal defect, a left superior vena cava (SVC), and pulmonary venous anomalies, etc. We report a case of unilateral pulmonary vein stenosis diagnosed 7 years after the repair of cor triatriatum.
Methods: A 2-month-old boy was diagnosed with cor triatriatum and severe pulmonary hypertension (PAH). The LA was divided by an obstructing membrane with 2 mm opening. The membrane was excised and the postoperative course was uneventful. Seven years after the operation, we recognised that the vascular markings and the volume of the left lung were markedly decreased. Therefore, an examination was done to obtain detailed information.
Results: On the echocardiogram, the left pulmonary artery (LPA) was relatively small (7 mm, z = -3), but there was no PAH. The lung perfusion scan showed the markedly decreased perfusion on the left (95% vs 5%). In the cardiac catheterisation, LPAwas small and left pulmonary vein is not recognised. Right ventricle (RV) pressure was 25/0/7, MPA 25/15/13, wedge pressure of RPA 10, LPA 15 and mean LA pressure was 10 mmHg. The 3D CT showed small LPA and no left pulmonary veins.
Conclusion: In this case, unilateral pulmonary vein obstruction was hard to diagnose because pulmonary hypertension and symptoms did not appear. We assume that the pulmonary vein stenosis may have been associated with cor triatriatum at the time of surgery, and progressed slowly during the follow-up period. Thus, particular care should be taken to identify such anomalies in patients with cor triatriatum before and after repair.

1615: END POINT SEPARATION BETWEEN MITRAL VALVE AND SEPTUM OVER THE DIASTOLIC LEFT VENTRICLE DIAMETER (EPSS/LVDD) OF THE LONG-AXIS VIEW OF ECHOCARDIOGRAPHY IS AN EASY AND RELIABLE PARAMETER FOR ASSESSMENT OF GLOBAL LV DYSFUNCTION IN PATIENTS OF THALASSAEMIA MAJOR

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Background: Myocardial dysfunction is a common complication in patients of thalassaemia major (TM). Although echocardiography is a convenient instrument, none of its parameters so far has been considered as an ideal surrogate for prediction of impending heart failure in TM. However, our previous study of using the long-axis, M-mode echocardiographic measurements in TM patients with acute heart failure proved that the distance of end point separation between mitral valve and septum (EPSS) and the ratio of EPSS divided by diastolic diameter of the LV (EPSS/LVDd) was significantly correlated with their clinical and laboratory improvements. This study intended to extend the clinical application of EPSS and EPSS/LVDd to TM patients of long-term follow up.
Materials and method: From January 2003 through July 2012, the thalassaemia registry of China Medical University Hospital, Taiwan, enrolled 34 TM patients for daily oral iron chelator therapy and echocardiographic studies, which were performed every 6 months to 1 year by two double-blinded technicians. The normal ranges of EPSS (2.5 ± 1.7 mm) and EPSS/ LVDd (0.08 ± 0.06) were used as references. Compared to the baseline data at enrolment, the trends of changes in EPSS, LVDd and EPSS/LVDd were assessed by the generalized estimation equation (GEE).
Result: Their average ages were 13.4 ± 3.7 years at enrolment and the average follow-up duration was 7.2 ± 2.0 years. In accord with improvements of EPSS/LVDd, favourable response to iron chelation occurred in 24 of the 34 patients (13 with direct improvement, 5 stayed as before (normal) and 6 showed delay in improvement after initial worsening), and unfavourable response occurred in 10 patients (5 remained as before (abnormal) and 5 even deteriorated). GEE analysis showed a general trend of rising EPSS/LVDd after 5 years of follow up ($p < 0.01$).
Conclusions: EPSS/LVDd is a convenient parameter to monitor the global cardiac function of TM patients.

1616: OUTCOMES OF IMPLANTABLE CARDIOVERTER-DEFIBRILLATOR USE IN PAEDIATRIC AND CONGENITAL HEART DISEASE PATIENTS - THE NORTH WEST EXPERIENCE

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Objective: We reviewed our experience over 20 years of the use, complications and outcomes of implantable cardioverter defibrillators (ICD) in a large paediatric population.
Method: Retrospective analysis was done of 35 patients requiring ICD insertion from 1993 to 2012 at a single tertiary paediatric cardiac centre. Indications for implant were long QT syndrome (n = 16), catecholaminergic polymorphic ventricular tachycardia (n = 5), hypertrophic obstructive cardiomyopathy (n = 5), idiopathic ventricular tachycardia (VT) (n = 3), ventricular tachycardia (VT) with underlying congenital heart disease (n = 4), Brugada (n = 1), arrhythmogenic right ventricular dysplasia (n = 1).
Results: There were 38 ICD implantations (19 males). Median age at presentation was 10 years (range 5–20) and median weight of 37 kg (range 21–70). The presentation was 15 with cardiac arrest, 17 with syncope, 1 with pre-syncope, 1 with palpitations and 1 with a seizure. Concomitant antiarrhythmic drug therapy was with beta-blocker alone in 10, beta-blocker and amiodarone in 2 and amiodarone alone in 1. A single chamber ICD was used in 18 and dual chamber in 20. Median age at implant was 11 years (range 6–23). At follow-up (median 4 years, range 0.5–12 years), monitoring revealed 7 episodes (18%) of appropriate shocks and 9 (24%) of inappropriate shocks as a result of lead failure (n = 4), oversensitivity (n = 2) and sinus/atrial tachycardia (n = 3). Twenty-two (58%) patients did not receive any shocks. There were 9 lead replacements, 3 of which were elective as a result of box change. There were 2 episodes of infection, one minor and one needing box change. Two (5%) patients had died, 1 with a head injury after collapse secondary to VT successfully cardioverted and 1 with catecholamine storm and repeated cardioversions.
Conclusion: ICD implantation in the paediatric population can be life-saving but is associated with a significant incidence of inappropriate shock and requirements for generator and electrode replacement.

1619: AETIOLOGY OF PULMONARY HYPERTENSION IN AFRICA - PRELIMINARY DATA ANALYSIS AFTER ONE YEAR OF RECRUITMENT: THE PAN AFRICAN PULMONARY HYPERTENSION COHORT STUDY (PAPUCO)

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Background: Pulmonary hypertension (PH) is a devastating, progressive disease, with increasingly debilitating symptoms and, usually, shortened overall life expectancy. The epidemiology of PH in Africa has not yet been determined, but limited reports suggest that the incidence is higher than that reported from developed countries. Many known factors for PH are hyperendemic in this part of the world, including HIV/AIDS, rheumatic heart disease, schistosomiasis and sickle cell disease. We aim to describe the aetiology of PH in Africa.
Materials and methods: A prospective observational study of patients with newly diagnosed and previously untreated PH based on echocardiography. Fifteen cardiovascular centres from sub-Saharan Africa participate in this study. Preliminary data analysis after 1 year of recruitment is presented.
Results: A total of 63 patients were recruited within the first year of the study. Median age was 40 years (range 11–86) and 62% were female. Twenty-four per cent of patients were living below the WHO poverty line of one US$ per day; 37% of patients were known to be HIV positive with a median CD4 count of 401 cells/µl (interquartile range (IQR) 226–537 cells/µl) and 83% were on antiretroviral therapy at presentation. Thirty-two per cent of patients had documented previously treated TB with site of TB being pulmonary in 85% of documented cases; 50% of those had at least two or more documented episodes of TB. At presentation, median pulse at rest was 94 bpm (IQR 78–100 bpm) and pulse oximetry was 95% (IQR 91–98%); respiration rate was 26 bpm (IQR 21–32 bpm). On echocardiography, median right ventricular systolic pressure (RVSP) was 56 mmHg (IQR 48–68 mmHg), tricuspid annular plane systolic excursion (TAPSE) 14 mm (11–17 mm) and left ventricle ejection fraction (LV-EF) 50% (35–65%). Thirty-three per cent of patients died within 6 months of presentation.
Conclusion: PH in Africa is an acquired cardiovascular disease of high mortality and of multiple aetiologies. HIV seems to be an important risk factor for PH in Africa.

1620: IMPACT OF SURGERY BY A VISITING TEAM ON QUALITY OF LIFE INDICATORS IN PATIENTS WITH CONGENITAL OR ACQUIRED HEART DISEASE IN NATIONS WITH LIMITED HEALTHCARE RESOURCES

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Hypothesis: Surgery by a visiting team improves quality of life indicators in patients with congenital or acquired heart disease in nations with developing healthcare programmes.
Method: CardioStart International's visiting surgical teams interviewed patients via translator between February 2008 until November 2011 (n = 198) in Peru, Ghana, Vietnam, and Grenada. Ages ranged from 24 days to 80 years old. Data were collected regarding symptoms, income, perception of health, and emotional status. Parents/guardians were interviewed if the child was unable to answer questions. Follow-up interviews were conducted with patients who had undergone surgery in prior years (n =33).
Results: For surgical candidates, the most common symptoms were shortness of breath (46.5%, 92/172) and fatigue (46.0%, 91/172); 57.1% (76/133) of patients had their symptoms for more than one year. Half (50.4%, 71/141) reported a household income loss of 50% or greater since diagnosis of the heart condition. In comparison with others, 51.6% (79/153) reported their health as fair or poor. Emotionally, 20.6% (29/141) felt fears and 36.4% (43/118) felt concern for the future related to the heart condition ('often' or 'almost always'). Follow-up data: After surgery, 63% (17/27) revealed no symptoms; 11.1% (3/27) reported shortness of breath; 11.1% (3/27) reported fatigue. For income, 72% (18/25) reported no loss or an improvement in income since the operation. Health perception: 81.5% (22/27) reported their health as 'good', 'very good', or 'excellent' compared to others, while emotionally, 8.7% (2/23) felt fears and 20% (4/20) felt concern for the future related to the heart condition ('often' or 'almost always').
Conclusions: Surgery to correct congenital and acquired heart disease has the potential to improve quality of life in these diverse populations. Lack of health care infrastructure in these countries made it difficult to obtain follow-up data. More follow-up data are needed to make reliable inferences, and a more efficient implementation of follow-up is in progress.

1626: PREVALENCE AND CLINICAL IMPACT OF MAJOR AORTOPULMONARY COLLATERALS IN POSTOPERATIVE PATIENTS WITH TRANSPOSITION OF GREAT ARTERIES

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Background: Transposition of great arteries with intact ventricular septum (TGA) is rarely associated with clinically significant aortopulmonary collateral arteries (MAPCA). The known associated complications are: congestive heart failure, pulmonary volume overload, left ventricular dysfunction and respiratory failure. The aim of this study is to evaluate the prevalence and clinical impact of MAPCAs in postoperative patients with TGA.
Methods: All patients with TGA admitted after arterial switch procedure for a period of 7 years from July 2005 to August 2012 were included in the study. Data was analysed from PICU database regarding duration of ventilation, length of stay and outcome. Data from the catheter laboratory was obtained to identify infants with TGA who had undergone catheter occlusion of MAPCA.
Results: A total of 48 cases were identified to have TGA. Three haemodynamically significant cases requiring catheter occlusion of MPACA were identified on echocardiography. Two patients had difficulty in weaning from ventilation and 1 presented with pulmonary haemorrhage. Outcome of TGA with (3) vs without MAPCA (45) in relation to ventilation, PICU stay and mortality is as follows: ventilation: 10 (5–14) vs 3.8 (1–6) days, PICU stay: 14 (12–22) vs 5 (2–9) days; and mortality 0 vs 1. The mean duration of ventilation post coiling was 2 days (1–3 days). No coronary abnormality or ischaemic changes were identified on ECG in any of the 48 cases, except in one who died.
Conclusion: TGA with MAPCAs is associated with prolonged ventilation, pulmonary hypertension (PHT) and pulmonary haemorrhage. There is a higher than expected incidence of clinically significant MPACA in our series and we report the first case of postoperative pulmonary haemorrhage in this setting. TGA with prolonged venti-

lation, persistent PHT, or pulmonary haemorrhage warrants careful evaluation for MAPCAs.

1633: PULMONARY VALVE GROWTH FOLLOWING VALVE SPARING TETRALOGY OF FALLOT REPAIR

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Background: There has been increased focus on pulmonary valve (PV) preservation in tetralogy of Fallot (TOF) repair. Valve sparing (VS) techniques utilise a combined transatrial and transpulmonary approach, a limited right ventriculotomy, sharp commissurotomies and valvar/annular balloon dilation. The objective of this study is to describe the VS TOF repair experience in terms of valve growth and performance.
Materials and methods: We did a retrospective review of pre-operative echocardiograms in all patients who underwent repair of isolated TOF from 2004 to 2012. PV annulus (PVA) dimensions were assessed on immediate post-repair and most recent echocardiogram studies in those undergoing a VS approach.
Results: Ninety-seven patients were identified. Fifty-nine (61%) underwent a VS repair at a median age of 4.1 months and 38 had a transannular patch (TAP) at a median age of 3.5 months ($p = 0.5$). Mean pre-operative PVA diameter was 7.4 mm (z-score, -1.7) in the VS group versus 5.5 mm (z-score, -3.2) in the TAP group ($p < 0.0001$). In the VS group, the mean post-operative PVA was 9.7 mm (z-score of -0.2; $p < 0.0001$). Mean post-operative peak RVOT velocity (Vmax) was 2.1 m/s and 93% had < mild pulmonary insufficiency (PI). Intra-operative pulmonary balloon valvuloplasty was performed in 10 patients. In this group, mean PVA increased from 6.9 mm (z-score, -2.1) to 9.1 mm (z-score, -0.8) and post-operative Vmax was 2.3 m/s with 80% having < mild PI. Nineteen patients are followed at this institution with a mean follow-up of 3.7 years. In this subset, the mean post-operative PVA grew from 9.6 mm to 14.9 mm ($p < 0.0001$), while maintaining normal z-scores (-0.3 to -0.4), and no increase in the minimal pulmonary stenosis (PS) or PI.
Conclusions: VS TOF repair maintains the integrity of the PV and allows ongoing annular growth with minimal PS or PI. This technique may prevent the long-term sequelae of right ventricular volume overload secondary to chronic pulmonary insufficiency.

1643: CONCERNING OCULAR SIDE-EFFECT POSSIBLY ASSOCIATED WITH PDE5 INHIBITOR IN AN INFANT AFFECTED BY COMPLEX CONGENITAL HEART DISEASE

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We report a case of bilateral blindness possibly associated with PDE5-inhibitor intake (sildenafil) in a 7-month-old baby affected by a complex congenital heart defect.

Cavopulmonary anastomosis was performed at 4 months of age for double inlet left ventricle, hypoplastic right ventricle, bulbo-ventricular foramen and moderate subpulmonary obstruction. The post-op period was uneventful, except for the occurrence of chylothorax, which was treated with multiple drainages, parenteral nutrition and diuretics. Echocardiographic evaluation showed satisfactory results at the anastomosis level, with good left ventricle and atrioventricular valve function. Nonetheless, intermittent episodes of chylothorax were still present after 2 months. Thus, despite the lack of obvious echographic signs of pulmonary hypertension, we raised the hypothesis that a relative increase of pulmonary vascular resistances might have been involved in the aetiology of the persistent chylothorax. Then, *ex adiuvantibus*, we decided to start sildenafil at 0.2 mg/kg TDS.

Four weeks later a lack of visual focus on moving objects was noticed, with poor pupillary light reflex. An ophthalmologic examination revealed bilateral pendular nystagmus, absent papillary reflex, transparent crystalline lens, light pallor papilla, arterial venous tortuous vessels, peripapillary retinal haemorrhages and macular exudation. The sudden onset of visual loss associated with light pallor optic disc, in the absence of other neurological and orbital abnormalities, prompted the diagnosis of posterior ischaemic optic neuropathy (PION).

Sildenafil was immediately suspended, while a trial with methyprednisolone intravenously was attempted for the treatment of PION.

One month later, complete reduction of retinal haemorrhages, macular exudation and arterial venous tortuosity was noticed; however visual evoked potentials were absent. At one year follow-up no visual recovery was observed, with further worsening of bilateral optic disc atrophy. This alarming case raises questions about the safety of sildenafil in young infants and the necessity of ongoing ophthalmological evaluations before and during therapy with PDE5 inhibitors.

1647: OVERVIEW OF PAEDIATRIC DILATED CARDIOMYOPATHY IN A TERTIARY HOSPITAL

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Introduction: Dilated cardiomyopathy (DCM) is the most common form of cardiomyopathy in the paediatric population and an important cause of heart transplantation in children.
Objective: To characterise the paediatric population diagnosed with DCM in our paediatric cardiology referral centre in the north of Portugal.
Methods: We performed a retrospective review of patients diagnosed with DCM from January 2005 to June 2012, including demographic data, clinical presentation, aetiology, treatment and outcome.
Results: We identified 61 patients with DCM (37 female; 24 male). Median age at diagnosis was 16 months. Heart failure was present in 83.6% ($n = 51$) of patients and 44.3% ($n = 27$) needed intensive care. Median left ventricular ejection fraction at clinical presentation was 32.0% (SD 13.0%). The most prevalent causes were idiopathic ($n = 29$ [47.5%]), viral myocarditis ($n = 11$ [18.0%]) and inherited metabolic diseases ($n = 7$ [11.5%]). In viral myocarditis parvovirus B19 was the most common identified agent. Anticongestive agents, angiotensin-converting enzyme inhibitor and anti-aggregant therapy were used in the majority of patients. Overall mortality was 16.1% ($n = 10$) and 5 patients underwent heart transplantation. Age under 1 year was associated with higher mortality ($p < 0.001$) and increased need of intensive care ($p < 0.05$). A higher rate of transplantation was observed in children over 1 year of age ($p < 0.05$) and with intensive care admission ($p < 0.05$). We report no mortality in the 5 patients who underwent heart transplantation, after 1 year of follow-up.
Discussion: The most important prognostic factors identified were age at DCM diagnosis and requirement for intensive care. Although the majority of patients had an idiopathic form of DCM, a significant number of patients presented with inherited metabolic diseases. This reinforces the importance of considering them in the differential diagnosis of paediatric DCM, as an appropriate approach to inherited metabolic diseases could alter the outcome of affected patients.

1655: THE EFFECT OF SPIRONOLACTONE, FUROSEMIDE AND CAPTOPRIL ON BLOOD ELECTROLYTE LEVELS IN CONGENITAL HEART DEFECT INFANTS WITH UNCONTROLLED CONGESTIVE HEART FAILURE

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Objective: The aim of this study was to evaluate the effect of spironolactone, furosemide and captopril on blood electrolyte levels in congenital heart defect infants with uncontrolled congestive heart failure.
Material and method: We examined 27 infants with congestive heart failure who had been treated with furosemide and captopril for at least 1 month in the Cukurova University Department of Pediatric Cardiology. Before the spironolactone treatment and after the first week of the treatment blood sodium, potassium, urinary creatinine levels were checked. The dosage of spironolactone was determined as 1–3 mg/kg/day. Patients treated with the dosage of spironolactone ≤ 2 mg/kg/day were Group 1, and with the dosage of 2–3 mg/kg/day were group 2.
Results: Seventeen patients (63%) were in Group 1 and 10 (37%) in Group 2. After the first week of the treatment, mean blood sodium level was 134 mmol/l, and mean blood potassium level was 4.6 mmol/l. Hyponatraemia occurred in 12 patients. There were statistical differences in mean blood sodium levels between group 1 and 2 ($p < 0.02$). There was no statistical difference in development of hyponatremia between two groups. There was no statistical difference in potassium levels between the groups and no hyperpotassaemia.
Conclusion: There is no safe dosage range or safe combination protocol in congestive heart failure treatment in paediatric patients. Because of the development of natriuresis and hyperpotassaemia, clinical and laboratory monitoring is needed in combination therapy. We did not encounter to hyperpotassaemia at different spironolactone dosages. There was no statistical upgrade of potassium levels between groups with different spironolactone dosages. Hyponatraemia improved quickly with reduction or stopping the spironolactone dose. In this prospective pilot study there was no statistical difference in potassium levels with spironolactone treatment; hyperpotassaemia did not occur even with high dosages. But the occurrence of hyponatraemia in 44% of the patients was a remarkable result. In conclusion, frequent electrolyte monitoring is needed with the treatment of congestive heart failure when spironolactone is added to furosemide and captopril.

1657: PERMANENT CARDIAC PACING IN CHILDREN - CHOOSING THE OPTIMAL PACING-SITE: A MULTI-CENTRE STUDY

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Purpose: We evaluated the effects of pacing-site on left ventricular (LV) synchrony and function in children requiring permanent pacing.
Methods: A total of 178 children (age < 18 years) from 21 centres with complete AV block and a structurally normal heart undergoing permanent pacing were cross-sectionally studied. Median age at evaluation was 11.2 (inter-quartile range (IQR) 6.3–15.0) years. Median pacing duration was 5.4 (IQR 3.1–8.8) years. Data were analysed in a core lab. Pacing-sites were the free wall of the right ventricular outflow tract (RVOT) ($n = 8$), lateral RV (RVLat) ($n = 44$), RV apex (RVA) ($n = 61$), RV septum (RVS) ($n = 29$), LV apex (LVA) ($n = 12$), LV mid-lateral wall (LVLat) ($n = 17$) and LV base (LVB) ($n = 7$).
Results: LV synchrony, pump function (ejection fraction (EF), end-systolic volume index and change in shortening fraction as compared to pre-implantation values) and contraction efficiency were significantly affected by pacing-site and were superior in children paced at LVA/LVLat. LV dyssynchrony assessed by radial strain correlated inversely with LV EF (r = 0.80, $p = 0.031$). Pacing from RVOT/RVLat predicted decreased LV function (LV EF < 45%; odds ratio (OR) 5.19, confidence interval (CI) 1.74-15.50, $p = 0.003$) whereas LVA/LVLat pacing was associated with preserved LV function (LV EF > 55%; OR 6.97, CI 2.21-22.00, $p < 0.001$). Age at implantation, pre-implantation LV size and function, duration of pacing, DDD mode, QRS duration and presence of maternal auto-antibodies had no significant impact in a multivariable analysis.
Conclusions: LV mechanical synchrony, pump function and contraction efficiency may significantly deteriorate with RVOT/RVLat pacing and are best preserved with LVA/LVLat pacing.
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1663: OUTCOME OF CHILDREN AND ADOLESCENTS UNDERGOING INVASIVE TESTING FOR ASYMPTOMATIC WOLFF-PARKINSON-WHITE PRE-EXCITATION

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Objective: Results of a risk stratification strategy in asymptomatic Wolff-Parkinson-White (WPW) pre-excitation in the young were evaluated.
Methods: Eighty-five consecutive patients aged < 18 years with a WPW pattern (with persistent pre-excitation at maximum exercise) undergoing an invasive electrophysiological study for risk stratification were retrospectively evaluated. Potentially dangerous accessory pathway (AP) properties were defined as any of the following: antegrade effective refractory period ≤250 ms, shortest pre-excited RR interval during atrial fibrillation/rapid atrial pacing ≤250 ms, inducible atrioventricular re-entrant tachycardia (AVRT) or presence of multiple APs.
Results: Age at evaluation was median 14.9 (interquartile range (IQR) 12.5–16.6) years. Eighty-two patients had a structurally normal heart and 3 had hypertrophic cardiomyopathy. A single manifest AP was present in 80 patients, one manifest and 1 concealed AP in 4 and two manifest APs in 1 patient. Potentially dangerous AP properties were present in 33/85 patients (38.8 %) at baseline and in an additional 16/44 (36.4 %) of those subjected to isoproterenol challenge. Ablation was performed in 41/49 patients with a potentially dangerous AP and deferred in the remaining 8 because of proximity to the atrioventricular node. In addition, 18 of low-risk patients were ablated based on patient's/parental decision.
Conclusion: 39 % of the evaluated patients with asymptomatic WPW pre-excitation persisting at peak exercise exhibited potentially dangerous AP properties at baseline. Isoproterenol challenge yielded an additional 36% of those tested at risk. Ablation was performed in 69% of patients.

1664: A SINGLE INSTITUTIONAL EXPERIENCE OF USING CARDIAC MAGNETIC RESONANCE AS THE PRIMARY INTER-STAGE DIAGNOSTIC SCREEN, PRIOR TO SURGICAL COMPLETION OF TOTAL CAVOPULMONARY CONNECTION FOR PATIENTS WITH FUNCTIONALLY SINGLE VENTRICLES

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Background: Cardiac magnetic resonance (CMR) is proposed as a sensitive diagnostic tool to guide decision-making after bidirectional cavopulmonary connection (BCPC) and prior to total cavopulmonary connection (TCPC). Our unit has adopted a protocolled, CMR-based screening method, performed under general anaesthetic with transduced central venous pressure (CVP) measurement from the internal jugular vein.
Methods: We did a retrospective review of single-centre, medium-term TCPC experience, in relation to pre-operative CMR data. Non-parametric statistical methods were used. Results were expressed as median (interquartile range).
Results: The cohort included 192 local patients undergoing BCPC since 2005. Of those, 86 had undergone TCPC at age 4.0 (3.3–4.6) years; 47 (55%) had right ventricular dominance. Weight at surgery was 15.1 (13.6–16.9) kg. Fifty-six (65%) were non-fenestrated at the time of initial surgery.Seventy-six (88%) operated patients are alive without heart transplantation, with duration of follow-up 2.2 (1.1–3.7) years. Interstage CMR was carried out in 55 (64%) of the 86 operated patients. The remaining patients required early investigation and intervention following BCPC, and underwent out-of-protocol CT scan or cardiac catheterisation. These patients experienced a 2.9-fold higher mortality; 6/31(19.4%) deaths occurred in this group, compared with 4/61(6.6%) deaths in the CMR group. Of those undergoing CMR median CVP was 12 (11–14) mmHg. Patients with CVP > 13 had a greater proportion of pulmonary venous return contributed by systemic to pulmonary collateral flow (median 32% vs 42%, $p < 0.05$). The CVP measured at CMR was not significantly related to pulmonary artery stenosis/hypoplasia, and did not predict length of hospital stay or mortality. However, mortality following TCPC was strongly associated with prominent venous channels visible on MR angiography, and off-loading superior vena cava (SVC) into inferior vena cava (IVC) territory ($p < 0.05$).
Conclusions: Interstage CMR prior to TCPC offers a comprehensive assessment of morphology and physiology, can elicit risk factors of postoperative outcome, and may identify confounding factors for pulmonary artery pressure, such as SVC to IVC collateralisation.

1669: SPOT THE DIFFERENCE: CAN YOU TRANSFER 1.5 T REFERENCE VALUES TO THE 3T ERA?

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Background: Cardiac MRI is important in the treatment of children with congenital heart disease. It is the reference standard for the assessment of ventricular dimensions and function. Most published reference values were obtained by 1.5 T MR scanner.
Methods: Quantitative volumetric cardiac MR measurements were performed on a 3T TX MRT (Philips ACHIEVA) and a 1.5T MRT (Philips Intera) using a multi-slice multi-phase steady-state free precision gradient-echo acquisition in breath hold (TR/TE/flip = 2.9 ms/1,45 ms/40°; Matrix = 1.4–1.5 × 1.5–0.7 mm²; 22–30 phases, 5 mm slice thickness). Patient sample included 17 healthy persons (7 male, 10 female, mean age 13.5 ± 4.3 y; range 6–20). Calculated stroke volume was controlled by flow derived stroke volumes using phase-contrast MRI. Data were quantified by a single expert.
F-test and unpaired t-test were performed.
Results: There were no significant differences for both left and right ventricle.
Conclusion: There is no relevant difference in ventricular size when using a 1.5T or 3T scanner.

1672: SINGLE-CENTRE EXPERIENCE OF DEVICE CLOSURE OF CONGENITAL VENTRICULAR SEPTAL DEFECTS

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Background: Routine transcatheter closure of perimembranous ventricular septal defects (VSD) is controversial because of atrioventricular nodal conduction disturbances. Muscular and postoperative VSD do not pose such risks, but are rare.
Methods: In a retrospective review of 80 patients who underwent VSD device closure, 59 had perimembranous VSD, 11 had muscular defects, two had intraconal outlet VSD, one had subpulmonary VSD, and five had residual defects after surgery. Indications for closure were symptoms, pulmonary hypertension and cardiac enlargement. Asymmetric devices were chosen in membranous VSD without aortic margin, intraconal and subpulmonary VSD. Other devices were used in membranous defects with septal aneurysm.
Results: A total of 78 procedures were successful. Age ranged from 8 months to 50 years and body weight from 6 to 79 kg. Two patients had echocardiographic guided perventricular closure; others had closure in the catheterisation lab. Asymmetric VSD occluder was used in 17 patients, symmetric VSD occluder in 2, muscular VSD occluder in 2, atrial septal occluder in 1, standard duct occluder in 30 and Amplatzer duct occcluder II in 26 patients. With the exception of hybrid closures and duct occluder II devices, all others needed formation of arteriovenous loop. Two perimembranous defects needed two duct occluder II devices. One perimembranous VSD measuring 13 mm procedure failed and was closed in surgery. Another patient with postoperative residual VSD refused repeat surgery. None had complete atrioventricular nodal block or need for pacing. One postoperative patient with aortic regurgitation and ventricular dysfunction had closure of residual large VSD with atrial septal occluder but died of progressive heart failure after 2 years. All others are free of any residual shunt at a mean follow-up of 2 years.
Conclusion: Closure of VSD with devices in various locations is safe and feasible. Atrioventricular nodal disturbances are not seen when duct occluders are used.

1673: SURGICAL CORRECTION OF TOTAL ANOMALOUS PULMONARY VEIN CONNECTION: A SINGLE-CENTRE EXPERIENCE

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Background: Total anomalous pulmonary venous connection (TAPVC) is a rare cardiac anomaly often associated with other cardiac malformations and with a poor prognosis without surgical treatment. We present a 15-year single-centre experience in surgical treatment of different types of TAPWC.
Methods: Between January 1998 to May 2012, 58 patients underwent TAPVC surgery at Children's City Hospital #1 St Petersburg, Russia. Data collection occurred retrospectively. Mean age at the time of surgery was 7.2 days. TAPVC was supracardiac in 30 patients (51.7%), intracardiac in 19 (32.8%), infracardiac in 4 (6.9%) and mixed in 5 patients (8.6%). TAPVC obstruction was confirmed by

echocardiography and was evident clinically in 8 patients (13.8%). Median CPB time was 64 minutes (40–106 min). DHCA was used in all cases with a median time of 29 minutes (24–37 min).
Results: Hospital mortality was 1.7% ($n = 1$). Cause of death in this case was low cardiac output due to heart failure. Follow-up was available for all of the operative survivors for a median period of 6.8 years (2.5 months to 14 years). Five patients underwent repeat surgery; 3 of them had obstruction and 2 had residual anomalous pulmonary vein connection. The times to reoperation for anastomotic stricture ($n = 3$) were 13 days, 3 months, 4 years. Late mortality was 1.7% ($n = 1$); this patient died of heart failure due to PV obstruction. The overall mortality of this series was 3.4% ($n = 2$). Hospital mortality has not been observed since 2008 in all age groups.
Conclusion: TAPVC is a rare congenital heart lesion, which still remains a surgical challenge. Despite the good short-term results of surgical correction, we should focus on long-term results, analysis of which will help us to reduce the number of late complications, the need for re-interventions and improve the quality of life.

1675: ANOMALOUS LEFT CORONARY ARTERY TO PULMONARY ARTERY (ALCAPA) AT RED CROSS WAR MEMORIAL CHILDREN'S HOSPITAL (RCWMCH), CAPE TOWN

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Introduction: Dilated cardiomyopathy (DCM) has a poor prognosis in the developing world where ICU beds, mechanical assist devices and cardiac transplants are limited. ALCAPA presents similarly to DCM but is surgically treatable; therefore ALCAPA should be considered when investigating DCM.
Aim: To audit patients admitted to RCWMCH with a new diagnosis of ALCAPA.
Method: We did a retrospective folder review of ALCAPA patients admitted between July 2004 and August 2012.
Results: Twenty-five newly diagnosed ALCAPA patients with median age of 5.3 (range 0.5– 30) months at presentation were selected. Presenting symptoms were tachypnoea in 96%, cardiomegaly 92%, coughing 88%, failure to thrive 60%, feeding difficulty 52%, and gallop in 44%. Median length of symptoms was 24 days (range 1–300). The diagnosis was made on echocardiography in 68%. Echocardiography findings were pathological MR and dilated LV in 79%, and bright papillary muscles in 78%. Median ejection fraction on admission was 26%. Median time from RCWMCH admission to surgery was 5 days (range 1–20). All received coronary reinplantation. Median cross clamp and bypass time was 71 and 140 minutes, respectively. Twenty-four per cent had delayed sternal closure. Median length of hospital stay, ICU stay, ventilation and inotropic support was 21, 10.8, 7.4 and 9.3 days, respectively. Median Wernovsky inotrope score was 32 (5–85). Perioperative complications included sepsis 68%, pleural effusion 24%, arrhythmias needing pacing 20%, bleeding 16% and renal replacement therapy 12.5%; 92% (23/25) survived to hospital discharge, 1 patient died pre surgery and 1 during surgery. Three patients died during subsequent admission for intercurrent chest infection.
Conclusion: RCWMCH can expect 3 new ALCAPA cases per year presenting in congestive cardiac failure. Treatment is successful with mortality rates comparable to the developed world. The major complication is perioperative-related sepsis.

1683: SINGLE- VERSUS TWO-STAGE REPAIR FOR PATIENTS WITH PULMONARY ATRESIA AND MAJOR AORTOPULMONARY COLLATERALS

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Background: Both single and staged approaches are described for patients with pulmonary atresia (PA) and variable availability of major aortopulmonary collaterals (MAPCAs) for unifocalisation. We compared the outcomes between patients who underwent single-stage versus staged repair for this pathology at our institution.
Materials and methods: Between 2007 and 2012, 61 patients who underwent procedures for ventricular septal defects (VSD), PA and MAPCAs were reviewed. Preoperative computed tomographic angiogram was done in all patients to evaluate pulmonary vasculature and MAPCAs. Twenty-five patients (group I) who underwent a staged repair (VSD was not closed in the 1st stage) were compared with 36 patients (group II) who underwent single-stage complete repair (VSD closed). The first stage included a systemic to PA shunt in 19 patients, shunt and unifocalisation in 6 and shunt and MAPCA ligation/coil embolisation in 8. All patients of group I underwent VSD closure and right ventricle (RV) to PA conduit interposition after a minimum follow-up period of 6 months. The number of MAPCAs unifocalizsed per patient ranged from 1 to 4(median 2) in both groups.
Results: There was no significant difference in the overall early complication rate between the two groups. Early mortality was significantly higher in patients who underwent staged procedures ($n = 8$) versus those who underwent single stage complete repair ($n = 0$) ($p = 0.01$). At a median follow-up period of 70 months, late mortality was similar between the two groups ($p = 0.45$).
Conclusions: The data demonstrate that single-stage complete repair has yielded good early and midterm results. The high mortality of the shunt group indicates the need for an alternative strategy to improve pulmonary blood flow other than a systemic to PA shunt. A palliative RV to PA conduit can be considered in these patients.

1684: A NOVEL SYNDROME OF HUMAN PACEMAKING DYSFUNCTION: LESSONS FROM A RARE DISEASE FOR PERSONALIZED MEDICINE

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Background: Lifelong rhythmic contractions are the hallmark of the human heart and gut. They result from the pacemaking activity of the specialised tissue, namely the sinoatrial node in the heart and interstitial cells of Cajal in the gut. Here, we describe a new syndrome characteried by progressive loss of normal sinoatrial node function (sick sinus syndrome (SSS)) and chronic intestinal pseudo-obstruction (CIPO) in French-Canadians.
Methods: Detailed chart review, family history, physical exam, ECG and echocardiography were performed in 14 carriers with SSS/CIPO. Whole exome sequencing was performed in three affected patients and analysed under a model of recessive inheritance.
Results: Age at onset of SSS was between 6 and 27 years of age, with all features of bradycardia-tachycardia syndrome. Pacemaker implantation was required in 5 individuals (age 6–21 years). Gastrointestinal symptoms appeared between 5 and 14 years of age, but not in any particular order to SSS/CIPO. Unexpectedly, we found that a mutation in shugoshin-like 1 (*SGOL1)*, a component of the cohesin complex, leads to this hitherto undescribed syndrome. All surviving affected patients are homozygous carriers of a founder mutation in *SGOL1* (c.67 T> C [p.Lys23Glu]) which results in a dramatic change of a highly conserved amino acid. Haplotype and genealogical analysis point to the introduction of a founder mutation at least 220 years ago. None of the unaffected first-degree relatives

was homozygous for the mutation, and the mutation was not detected in 360 French-Canadian controls. A 6-year-old homozygous mutation carrier developed signs of SSS/POIC during the course of the study.
Conclusion: Genetic analysis of the novel SSS/CIPO syndrome demonstrates a fundamental new link between the two main pacemakers in humans. A predictive value of *SGOL1* testing was found in one case and highlights the need to develop preventive strategies based on molecular findings.

1685: PRESENCE AND PATTERNS OF MYOCARDIAL GADOLINIUM ENHANCEMENT IN CHILDHOOD DILATED CARDIOMYOPATHY

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Background: Different patterns of myocardial gadolinium enhancement (MGE) including mid-wall fibrosis have been reported in up to 42% of adult patients with non-ischaemic dilated cardiomyopathy (NI-DCM). In these studies, MGE was associated with pronounced LV remodelling and predicted adverse cardiac outcomes. Accordingly, the purpose of our study was to determine the presence and patterns of MGE in children with NI-DCM.
Methods: Patients presenting with severe congestive heart failure who were admitted for evaluation of heart transplantation at our centre underwent CMR examination on a 3T system that consisted of ventricular functional analysis and assessment of MGE for detection of myocardial scars. Ischaemic DCM was excluded by coronary angiography and right ventricular endomyocardial biopsies ruled out ongoing myocarditis.
Results: Fifteen infants and children (mean age 9.3 ± 7.6 months) with severe LV dilatation (mean indexed LVEDV 160 ± 55 ml/m^2) and LV dysfunction (mean LV-EF 16 ± 7%) were examined. MGE was detected in 2 of the 15 patients (13%) appearing in patterns characterised as focal patchy and transmural respectively. None of the patients exhibited 'classic' mid-wall enhancement.
Conclusions: Despite the small cohort size, the observed differences in frequency and type of myocardial fibrosis compared to adult patients might influence the therapeutic strategy in childhood NI-DCM. However, it remains unclear whether these findings reflect preserved endogenous repair mechanisms that enable favourable remodelling. Larger trials are needed to assess the prognostic implications of MGE in childhood NI-DCM and to determine whether MGE might be used for risk stratification as demonstrated in adult NI-DCM.

1687: MID-TERM OUTCOMES OF A HANDMADE TRILEAFLET VALVED RIGHT VENTRICLE TO PULMONARY ARTERY CONDUIT

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Background: We have been using a new technique to make bovine pericardial valved conduits to overcome the shortage of cryopreserved homografts for right ventricle (RV) to pulmonary artery (PA) reconstructions. Gore Preclude pericardial membrane (0.1 mm thickness) is used for making the trileaflet valve. We reviewed our experience to analyse postoperative outcomes and their mid-term results.
Materials and methods: Between 2007 and 2012, 203 patients (71.9 % male) underwent primary right ventricle outflow tract reconstruction using bovine pericardial valved conduits in our centre. Their ages ranged from 6 days to 42 years. Diagnoses included 'ventricular septal defect (VSD), pulmonary atresia (PA), MAPCAs'(n = 61), 'VSD, PA'(n = 52), 'truncus arteriosus' (n = 20), 'double outlet right ventricle, VSD, pulmonary stenosis (PS)' (n = 18), 'tetralogy of Fallot/absent pulmonary valve' (n = 12), 'corrected transposition, VSD, PS' (n = 11), 'transpostion of great arteries, VSD, PS' (n = 7) and 'Ross procedure' (n = 11). Fifty-four (26.6%) patients had undergone a prior palliative shunt procedure. The sizes of the conduits implanted ranged from 12 to 24 mm (median 18).
Results: Conduit-related early complication rate was 0.5% (n = 1) (conduit revision for early pulmonary artery thrombosis). The mean hospital stay was 13.2 ± 31.6 days. Early mortality was 7.4 % (n = 15). The mean follow-up period was 61 months. Twenty-three (11.3 %) patients underwent conduit replacement. Of these, acquired distal conduit stenosis was observed in 10.8 % (n =22) and 0.5 % (n =1) developed infective endocarditis. Late mortality was 0.5 % (n = 1). A gradient > 30 mmHg was detected in 13.4 % (n = 27) patients, while 15.3 % (n 31) developed conduit valve regurgitation greater than 2+.
Conclusions: The handmade valved conduit seems to offer equivalent results as compared to other conduits available today at midterm follow-up. Long-term results are awaited. Easy availability and low cost are additional advantages of this conduit.

1690: CONGENITAL HEART DISEASE AND THE EMISSION OF DEVELOPMENTAL TOXICANTS IN ALBERTA, CANADA

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Background: Congenital heart disease (CHD) is a significant global public health issue affecting 1% of all live births and the most common lethal congenital abnormality in infancy. Although CHD may occur in the presence of aneuploidy and single gene disorders, in most affected children the cause is unknown. The role of environmental pollutants has recently received attention. We sought to explore the association of developmental toxicants (DTs) from industrial sources and CHD in Alberta, Canada through an interdisciplinary multi-step study.
Materials and methods: In this ecological study we collected the following data: (1) Chemical emissions between 2003-2010 from Canada's National Pollutant Release Inventory; (2) CHD cases (born between 1/06/04 and 31/08/11) from Stollery Children's Hospital Xcelera database; and (3) annual births from Statistics Canada/ Alberta Reproductive Health. We used Scorecard criteria to identify emitted DTs and corresponding toxic equivalent potential values in order to normalise emissions (risk score). The location of the emitting facilities and CHD cases were determined using the longitude and latitude coordinates. Data was aggregated by year and cases were assigned to the year when pregnancy occurred. Statistical analysis was done using STATA 12.
Results: We identified 1 903 cases of CHD and 17 DTs emitted to air (99% of all emissions) during the study period. The average rate of CHD was 5.8 ± 1.09/1 000 live births with the most commonly encountered including septal (47.9%), left ventricular outflow tract obstruction (15.2%) and conotruncal (12.2%) defects. The average DTs emissions were 7 817 417 ± 570 380 tonnes. Annual sulphur dioxide, ethylene oxide, 1.3-butadiene, hexachlorobenzene and carbon disulphide average risk scores strongly correlated with CHD rates (r = 0.89; r = 0.89; r = 0.87; r = 0.84 and r = 0.84, respectively, $p \leq 0.01$).
Conclusions: These findings suggest that DTs emitted to air in Alberta could have an impact on the development of CHD. An

in-depth analysis using sophisticated statistical methods, modelling and GIS mapping is in progress.

1691: COMPARISON OF SHEAR STRESS RATE DISTRIBUTION AFTER NORWOOD WITH RIGHT VENTRICLE-TO-PULMONARY ARTERY CONDUIT AND BLALOCK-TAUSSIG SHUNT FOR HYPOPLASTIC LEFT HEART SYNDROME: MATHEMATIC MODELLING OF HAEMODYNAMICS

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Objective: The introduction of right ventricle to pulmonary artery (RV-PA) conduit instead of Blalock-Taussig shunt (BTS) for Norwood procedure for hypoplastic left heart syndrome resulted in a higher survival rate in many centres. Exact mechanisms and long-term results are still under investigation. The main objective of this study was the comparison of shear stress rate (SSR) distribution in two models of Norwood physiology regarding different types of pulmonary blood flow sources.
Method: Based on anatomical details obtained from echocardiographic assessment and angiographic studies, two three-dimensional computer models of post-Norwood physiology with RV-PA 5 mm conduit and Blalock-Taussig shunt (BTS) 3.5 mm shunt were developed. The finite-element method was applied for computational simulations. Shear stress distribution was analyded in both models at basal level of pulmonary and systemic vascular resistances and also with decreased systemic vascular resistance and increased pulmonary vascular resistance.
Results: The highest values of SSR were observed in shunts both in RVPA and BTS Norwood models during the peak systolic phase. The imperfect reconstruction of the aorta influences the local distribution of SSR. Changes of systemic and pulmonary vascular resistance do not change significantly the distribution patterns of SSR in both models.
Conclusions: The sources of pulmonary blood flow are the areas of the highest SSR. The way of reconstruction of the aorta influences the local distribution of SSR.

1716: COMPLEX APPROACH TO HIGHLY MALIGNANT LONG QT SYNDROME IN A 2-YEAR-OLD GIRL

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Background and objectives: Clinical presentation and therapy of long QT syndrome (LQTS) depends on the genetic type of the syndrome. We present a case of a girl with clinical manifestation suggesting LQTS type 1 or 2, resistant to betablocker therapy.
Methods: A 2-year-old girl was admitted after syncope. Prolonged QT interval and episode of torsade de pointes (TdP) was found.
Results: Beta-blocker therapy was started with metoprolol. One month later syncope recurred and frequent TdP occurred of a few beats to 20 seconds' duration. Propranolol was initiated and increased to supramaximal dose with no effect on TdP frequency and minimal effect on heart rate. Frequency of TdP increased to a few hundred per day with increased duration and frequent loss of consciousness. TdP were related to sympathetic discharge, completely disappearing during sleep. Lidocaine and magnesium demonstrated no effect, analgosedation had a partial effect. An implantable cardioverter-defibrillator (ICD) was implanted and left cervical sympathetic denervation (LCSD) performed. Mexiletine obtained with the help of an international pharmacy resulted in complete elimination of TdP episodes. One year later, the patient was successfully defibrillated from life-threatening TdP recurrence. Genetic testing revealed SCN5A mutation P1332L.
Conclusion: Beta-blocker therapy of LQTS in a girl with clinical presentation suggesting LQTS type 1 or 2 was not successful. A complex approach including ICD implantation and LCSD was needed, with ICD serving as an emergency back-up treatment. Surprisingly, lidocaine was without effect but mexiletine led to disease control. Availability of mexiletine and genetic testing would allow earlier appropriate treatment.

1718: CARDIAC INVOLVEMENT IN PATIENTS WITH BECKER MUSCULAR DYSTROPHY

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Background: Cardiac involvement (CI) is an invariable feature of Becker muscular dystrophy (BMD), and can be even more pronounced than the skeletal muscle weakness. It may be subclinical and detectable only by instrumental investigation, or symptomatic. Asymptomatic CI occurs in most cases, but up to one-third of patients develop dilated cardiomyopathy (DCM) with concomitant heart failure. The degree of symptomatic CI in BMD varies greatly. The aim of this study was to assess the cardiac findings in patients with BMD from the outpatient clinic of our institution.
Material and methods: We did a retrospective review of clinical, electrocardiographic and echocardiographic assessments of patients with BMD.
Results: The study included 11 patients aged 1–16 years at referral to our paediatric cardiology centre. Long-term follow-up was 6.8 ± 4.4 years. Patients were assessed annually from time of diagnosis or more frequently according to the clinical manifestations. Only one adolescent had complaints of poor exercise tolerance. Prevalence of preclinical and clinically evident cardiac involvement was 55%. Electrocardiogram was abnormal in 9% of patients. Echocardiogram was abnormal in 55% of patients. Mild left ventricle enlargement was present in 6 patients. Three patients were started on ACE inhibitors.
Conclusion: Although most patients were asymptomatic, a high percentage had evidence of cardiac involvement. Clinicians need to be aware of the importance of myocardial involvement in patients with BMD and the therapies available. Until aetiology-specific genetic or cell-based therapies are developed, work in this area should concentrate on (1) implementing the complementary consensus recommendations for clinical care and research developed by neuromuscular and cardiovascular specialists and (2) identifying children with cardiomyopathy so currently available treatments can be used appropriately.

1719: TREATMENT OF PATENT DUCTUS ARTERIOSUS IN PRETERM NEONATES: SURGERY OR INDOMETHACIN?

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Objectives: The haemodynamically relevant patent ductus arteriosus (PDA) impairs pulmonary and cardiac function. This prospective study evaluated the treatment and outcome in preterm newborns.
Methods: Between January 2002 and January 2008, 3 043 newborns were enrolled, of whom 231 (7.6%) presented with PDA and were subdivided into groups depending on the treatment. Spontaneous closure occurred in 39/231 (16.9%) and 17 (7.4%) died without treatment. Indomethacin therapy was successful in 100 (43.3%) patients, surgical closure was the first treatment in 55 (23.8%) and 20 (8.6%) undergoing ductal ligation after indomethacin failure. The proportion of neonatal morbidities (chronic lung disease (CLD), necrotising enterocolitis (NEC), haemorrhage intraventricular (HIV),

pneumothorax) was compared between groups. The data collected was analysed with chi-square, Mann-Whitney and multiple logistic regression tests.
Results: The age of diagnosis and echocardiographic initial findings were significantly different in neonates with spontaneous closure and those with significant PDA ($p = 0.00$). PDA internal diameter > 2.0 mm, reversal of diastolic flow in descending aorta and treatment after 8 days were associated with complications (death, indomethacin failure, CLD, NEC and HIV). Indomethacin and surgery gave similar benefits. There were no differences in deaths, CLD or NEC. Surgery was more effective in closing PDA ($p = 0.00$) but it was associated with increased risk for HIV and pneumothorax ($p = 0.00$).
Conclusions: Early PDA diagnosis and treatment in preterm infants improves their outcome. Echocardiographic parameters predicted risk of complications and should be useful to guide the therapy. Indomethacin and surgical closure were safe and there were no long-term complications as a result of surgery.

1722: TETRALOGY OF FALLOT AND MENINGITIS COMPLICATED BY BRAIN ABSCESS: LATE FATAL PRESENTATION IN A CHILD

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Brain abscess is a known complication of cyanotic congenital heart diseases. We report the case of a 4-year-old girl whose first presentation in the hospital was because of symptoms related to the neurological system. The illness was however fatal as a result of the late presentation, which is not uncommon in this environment because of various reasons.

In a resource-poor environment like ours, this is a reminder that proper history taking, clinical examination and subsequent investigations will aid in early diagnosis and subsequent management of such cases to reduce childhood mortality.

1723: COMPARISON OF THE ECHOCARDIOGRAPHIC RESULTS IN INFANTS WITH VENTRICULAR SEPTAL DEFECT AND DIFFERENT NT-PROBNP AND ENDOTHELIN-1 LEVELS

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Aim: To analyse the haemodynamic disturbances assessed by echocardiography in infants with ventricular septal defect (VSD) and different NT-proBNP and endothelin-1 (ET-1) levels.
Material and methods: The study group consisted of 34 infants (aged 38–338 days, mean 130 ± 81 days) (15 boys, 19 girls) with VSD without pulmonary hypertension. In ECHO-2D the following parameters were analysed: pulmonary to systemic flow ratio (Qp/Qs), peak velocity of the pulmonary artery flow (Vmax PA), left atrial to aortic diameter ratio (LA/Ao) and indexed for body surface area, size of the defect (VSD/BSA), left atrial diameter (LAD/BSA), left ventricular internal diastolic diameter (LVIDd/BSA), right ventricular internal diastolic diameter (RVDd/BSA), main pulmonary artery diameter (MPA/BSA).

After the analysis of NT-proBNP and ET-1 levels, the following subgroups were selected: subgroup I – 24 (70%) children with NT-proBNP < 100 fmol/ml; subgroup II – 10 (30%) children with NT-proBNP ≥ 100 fmol/ml; subgroup III – 25 (73%) children with ET-1 < 0.4 fmol/ml; subgroup IV – 9 (27%) children with ET-1 ≥0.4 fmol/ml. Echocardiographic parameters were compared, subgroups I vs II and III vs IV. Statistical analysis was obtained using Student's t test.
Results: Results for subgroups will be presented. No significant differences of the mean values of all echocardiographic parameters in the subgroup III compared to those obtained in the subgroup IV were found.
Conclusions: Higher levels of NT-proBNP indicate more severe haemodynamic disturbances in infants with VSD. The determination of ET-1 level seems to be useless in infants with VSD.

1726: EVOLUTION OF RIGHT VENTRICLE-TO-PULMONARY ARTERY CONDUIT CONCEPT DURING THE NORWOOD PROCEDURE, 10 YEARS' SINGLE-INSTITUTION EXPERIENCE

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Background: The introduction of right ventricle to pulmonary artery (RV-PA) conduit in the Norwood procedure for hypoplastic left heart syndrome resulted in a higher survival rate in many centres. However, RV-PA conduit may result in more frequent unintended interventions becaused of stenosis at many levels.
Methods: We describe the evolution of RV-PA conduit concept for Norwood procedure in single institution, where between 2001 and 2012 more than 320 Norwood procedures with RV-PA conduit were performed. The technical issue of RV-PA placement will be described including the way of proximal and distal implantation, choosing the right or left position according to 'neoaorta', the diameter of shunt and applying of reinforced grafts.
Results: The current technique includes the reinforced, usually 5 mm in diameter shunt localised to the right according to the 'neoarta' with introduction of the proximal part of the conduit into the right ventricle wall.
Conclusion: The evolution of technical issue of RV-PA conduit for Norwood procedure results in smaller number of unintended interventions and facilitates the stage II performance.

1735: A RARE FORM OF CONGENITAL HEART DISEASE IN PREGNANCY

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Background: Univentricle is a rare form of congenital heart disease. It is often associated with other congenital heart defects.
Objective: To describe the clinical presentation and echocardiographic features of a patient presenting with a single atrium and single ventricle in pregnancy.
History and progress: A 19-year-old woman presented with a history of univentricle with left ventricular morphology, univentricular arteriovenous connection, absent right LV valve, pulmonary and tricuspid atresia and dextrocardia. In addition she had situs inversus and a single kidney. She had a classic BT shunt in 1991 at 6 weeks of age. She subsequently developed adhesions around the shunt and stenosis at the insertion site and on either side of the left pulmonary artery. Six years later she had total cavopulmonary connection. Thirteen years later she presented with an unplanned pregnancy, complaining of dizziness and dyspnoea. She subsequently had episodes of syncope, cyanotic spells and worsening dyspnoea. She was admitted for close monitoring and 3 weeks later had an elective caesarean section with no perioperative complications. She was discharged a few days later and during her follow-up period she remained haemodynamically stable. Her baby girl remained in a good condition.
Discussion and conclusion: There is lack of literature on the prevalence and success of pregnancies in this form of a complicated congenital heart disease. The terms complex single ventricle and univentricular heart are used to describe this congenital heart disease. These terms are also used to describe a group of rare heart defects, which have in common, a large single pumping chamber or ventricle. It is a serious problem because without surgery, most children would not be able to survive the first year of life. These patients may present

with congestive heart failure or cyanosis. Complex single ventricle can be diagnosed before birth by fetal echocardiography and as early as 18 weeks into pregnancy. This should be suspected especially if there is a family history of congenital heart disease.

1740: PROGNOSTIC IMPLICATIONS OF VENTRICUALR SEPTAL DEFECTS IN PREGNANCY

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Background: Ventricular septal defects (VSDs) are among the commonest congenital heart disease conditions. These can occur in isolation or as part of a more complex congenital heart disease. In adults they are usually small and restrictive; however if large they are commonly associated with Eisenmenger syndrome or part of complex congenital heart disease. In pregnancy isolated small or moderate-sized VSDs or surgically closed with normal ejection fraction and without pulmonary hypertension are usually well tolerated.
Objective/aim: To evaluate the prognostic outcome of VSD in pregnancy.
Methods and design: Thirty-two patients who presented with VSDs during pregnancy were reviewed. Their clinical profiles and echocardiography were retrieved and re-evaluated. The patients were divided into three groups based on the size of the defect, i.e. small, moderate or large. More than 70% of these VSDs were discovered incidentally during pregnancy. The New York Heart Association functional classification of dyspnoea was used to classify their functional capacity. All these patients were admitted and closely monitored during their peripartum period. They were subsequently followed up 6–8 weeks post-delivery for full cardiac evaluation.
Results: Of these patients 26 had small, 4 had moderate and 2 had large VSDs. Two patients had associated dextrocardia, 1 had associated congenitally corrected transposition of great vessels and 12 had pulmonary hypertension (8 patients had mild pulmonary hypertension and 4 had moderate pulmonary hypertension). The majority of these patients presented with at least class II symptoms (18) and 14 presented with class I symptoms. None of these patients experienced any peripartum complications related to either cardiac lesion or pregnancy. All patients received infective endocarditis prophylaxis during labour. At 6–8 weeks' follow-up, only patients with a large VSD reported at least class II symptoms and needed an earlier intervention.
Conclusion: In this study we predominantly had patients with a small VSD which is associated with favourable overall outcome. One patient had complex congenital heart disease as part of the syndrome. No associated maternal or foetal complications occurred perioperatively or post delivery.

1741: THE USE OF IMMUNOGLOBULIN FOR MYOCARDITIS IN CHILDREN - VARIATION IN PRACTICE ACROSS THE UNITED KINGDOM

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Background: The UK Department of Health has produced updated guidelines for the use of immunoglobulin (Ig) in 2011 which do not include myocarditis. A Cochrane review in 2009 found no evidence to support this indication but recent publications disagree. In the current financial climate British National Health Service funding of Ig is under review.
Objective: To investigate variations in current UK practice for the administration of Ig in children with acute myocarditis.
Method: *Design and setting:* A questionnaire was emailed to paediatric cardiology consultants in all 13 UK paediatric cardiac tertiary units. Paediatric cardiac pharmacists were also contacted. *Main outcome measures:* Use of Ig and status of recipients at preferred administration time (early in illness vs later when ventilated vs only proven viral infection cases).
Results: Eleven centres responded (85% response rate) with considerable variation in practice. Four units do not use Ig in myocarditis. Two units give only if a proven viral cause is known. One unit reported use only in desperately sick ventilated children while the remaining 4 units were using it more liberally early in the illness in non-ventilated patients.
Conclusion: Because of a paucity of evidence-based practice there is wide variation in management in whether Ig is given or not. Furthermore in those units continuing to use Ig for myocarditis there is no agreement on timing of administration (early or later in illness). There is a need for consensus guidelines, particularly because if a shortage occurs Ig will be withdrawn for this indication.

1746: LIVE REAL-TIME 3D ECHOCARDIOGRAPHY IN PAEDIATRIC CARDIOLOGY - FULFILLING THE PROMISE?

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Three-dimensional (3D) imaging is one of the most significant developments in ultrasound technology, providing realistic and unique, comprehensive views of cardiac structures. When initially introduced, it held a great promise in imaging of congenital abnormalities. However, acquisition of full-volume 3D data requires good patient cooperation, and lack of it in the paediatric population results in technically limited, often incomplete studies, causing in turn underuse of 3D. Labour-intensive and time-consuming off-line reconstruction further limits use of this technology.

Live real-time three-dimensional echocardiography (LRT3DE) is performed by acquiring still images and short clips using the narrow-angle 3D option, and applying appropriate views and angles. This readily available technique is practical in children, requires short added examination time, and enables incorporating 3D imaging into the daily clinical setting in a busy hospital clinic. Using LRT3DE routinely, and making it an integral part of the echocardiographic evaluation provides familiarity with its unique advantages as well as its limitations. We find it enhances understanding of anatomy and morphology of cardiac lesions, improves cardiologist-surgeon communication, and aids teaching.

We present examples of our experience in LRT3DE in both simple and complex congenital lesions and valvular heart disease, demonstrating its use and added value compared to 2D imaging.

1749: PATTERNS OF PRENATAL CEREBRAL GROWTH AMONG INFANTS WITH CONGENITAL HEART DISEASE

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Background: Microcephaly is more common in children with congenital heart disease (CHD), and the relation to neurodevelopmental abnormalities has been demonstrated. However confounders such as genetic syndromes and low placental weight were not sufficiently accounted for in existing studies. Additionally the focus has been on complex CHDs, and the impact of more common CHDs has yet to be evaluated.
Materials and methods: A register-based study was performed comprising a validated cohort of 2 947 Danish children born from 2000 to 2008 with CHD. Genetic, newborn and maternal parameters were identified in national registries, and each child was randomly matched to three non-syndromic children without CHD (n = 8841) according to gender and gestational age. By means of linear and logistic regression analysis, newborn head circumference will be adjusted to birth weight, maternal pre-pregnancy weight, smoking status, medical diseases, genetic abnormalities and placental weight

Results: The cohort represents high numbers of diagnostic subgroups: e.g. transposition of the great arteries (n = 189), hypoplastic left heart syndrome (n = 98), tetralogy of Fallot (n = 210), pulmonary stenosis (n = 178), coarctation of the aorta (n = 207), aortic valve stenosis (n = 88), atrioventricular septal defect (n = 184), VSD (n = 904) and ASD (n = 535). Trisomy 21 (n = 180), 22q11.2-deletion/duplication (n = 46) and other genetic abnormalities have been determined. Main outcome measure is newborn head circumference, non-adjusted and adjusted to newborn, maternal and genetic parameters comparing diagnostic subgroups of CHD to healthy controls. Preliminary comparison of unadjusted means revealed significantly smaller newborn head circumference in non-syndromic (-0.233 cm, $p < 0.0001$) and syndromic CHD cases (-1.299 cm, $p < 0.0001$) compared to controls. Analyses are undergoing and the results will be presented at the meeting.
Conclusion: The strength of this study, more than tripling the numbers of the largest study in the field, lies within the possibility to adjust head circumference to confounders such as genetic abnormalities and placental weight through data from unique national registries.

1764: ETHICAL ANALYSIS OF HLHS

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In a limited-resources Middle Eastern country, a foetus was diagnosed intrauterine with hypoplastic left heart syndrome (HLHS) after 120 days' gestation. No paediatric cardiac surgery programme or heart transplant programme is available. It is also not feasible to send patients abroad. The baby was born and required PGE1, and mechanical ventilation for severe cyanosis. Saturation improved with these actions but the baby developed NEC (started on TPN). The baby is obviously PGE1 and ventilator-dependant. Parents agreed to 'no code' status. ICU beds are full and there are 2 cases in ER of severe asthma that require urgent PICU admission with expected excellent outcome.

The presentation will include the following 5 steps:

- Collecting clinical data. The starting point in the ethical analysis of a clinical case consists in gathering information related to: medical aspects (diagnosis, prognosis, potential treatments); personal and relational aspects; and cultural aspects (Islamic perspective).
- Assessing responsibilities. What are the specific responsibilities of health care professionals in the given case? Has the patient (or his/her legal guardian) been adequately informed? What is the role of the family? What are the responsibilities of social bodies (social services, etc.)?
- Identifying ethical problems. What ethical problems are involved in the evolution of the given case?
- Proposing alternative courses of actions. What are the possible courses of action for this case, e.g. abortion vs no abortion; asthma vs HLHS priority? What are the motivating reasons?
- Formulating and justifying ethical judgement.

1767: CHANGING PATTERN OF RHEUMATIC HEART DISEASE IN KANO: DATA FROM THE AMINU KANO TEACHING HOSPITAL ECHOCARDIOGRAPHY REGISTRY

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Background: Rheumatic heart disease (RHD) remains a major public health problem in developing countries. Anecdotal reports across Africa show that the disease is becoming less prevalent in cities and patients are surviving longer, although with high morbidity. Between 2002 and 2006 from our echo register we found 9.8% of 1 312 patients to have RHD, with a mean age of 24 years. We set out to review our current data for RHD to see if there are any changes in patterns of presentation in our centre.
Material and methods: This is a retrospective analysis of prospectively collected echocardiography data between August 2010 and July 2012. The study was conducted at the Aminu Kano Teaching Hospital, Kano Nigeria. The procedure was performed with Aloka SSD 4000. The standard techniques for depicting the anatomical structures of the heart were employed. All the procedures were performed by a cardiologist. Information obtained from the records included the age, gender, clinical diagnoses and echocardiographic diagnoses. Prevalence and patterns were compared with previous findings.
Results: During this period, a total of 1 496 echocardiographic examinations were done. One hundred and four (7.0 %) had RHD. There were 69 females (66.3%) and 35 males (33.7%) aged 30.71 + 13.99 years (range 12–70 years). Forty (37.7 %) were aged 15–24 years. The commonest lesions were mixed mitral valve disease and aortic regurgitation (26.9%) followed by combination of mitral and aortic regurgitation (25%) and pure mitral regurgitation in 17.3 %. Complications of RHD observed included secondary pulmonary hypertension, left ventricular dysfunction, atrial fibrillation and infective endocarditis.
Conclusion: Although there is an improvement in prevalence compared to previous finding, RHD is still an important cause of cardiac morbidity. While the patients are a bit older, they had more severe disease and still had complications at diagnosis.

1769: PERIOPERATIVE VASOPRESSIN RESULTS IN REDUCED LENGTH OF HOSPITAL STAY AFTER THE FONTAN OPERATION

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Background: The Fontan operation is the final palliative surgery in patients with single ventricle physiology. Although early postoperative outcomes have improved over time, the length of hospital stay is still prolonged as a result of persistent chest tube output in the postoperative period. We hypothesised that the use of vasopressin (VP) in the perioperative period would reduce chest tube output by maintaining vascular tone, thereby limiting third spacing and the need for volume replacement.
Materials and method: We retrospectively analysed 31 consecutive patients undergoing Fontan operation between 2008 and 2012. In 2010 VP was introduced as part of the standard management of patients undergoing Fontan operation. The patients were grouped according to VP use (n = 24) or non-use (no-VP, n = 7) in the perioperative period; dosage 0.3–0.5 mU/kg/min. The endpoints analysed were hospital mortality, length of hospital stay (LOS), and chest tube output.
Results: The VP and no-VP subgroups were well matched for age and weight (14.9 kg vs 15.5 kg, p = NS). There was no hospital mortality. The LOS in the VP group was 11.2 ± 2.3 days compared to 18.4 ± 3.6 days in the no-VP group (p = 0.01). Daily chest tube output decreased significantly in the VP subgroup but not in the no-VP subgroup (p = 0.01).
Conclusions: Use of vasopressin in the early postoperative period is associated with reduced chest tube output and length of hospital stay after the Fontan operation.

1771: ISOLATED LEFT VENTRICULAR NONCOMPACTION

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Aim: To analyse the clinical course and echocardiographic features of isolated left ventricular non-compaction (LVNC) in children.
Methods: The study group consisted of 16 patients. Clinical evaluation, chest X-ray, ECG, Holter ECG and echocardiography were performed. For statistical analysis Student's t test was used.
Results: The age at diagnosis was 2 weeks to 18 years (average 6.44 years), follow-up 2 months to 9 years (mean 3.15 years). In two brothers Barth syndrome was recognised. Signs of acute heart failure occurred in 4 children. Chest X-ray revealed cardiomegaly in 7 with features of pulmonary congestion in 2. The ECG was within normal limits in 8, and detected signs of LV hypertrophy in 3, 1st degree atrio-ventricular block in 1, QTc prolongation in 1, ST-T abnormalities in the form of fluttering and/or biphasic T in 3. In 5 patients arrhythmia was diagnosed. In all the evidence of 2-layered myocardium with prominent trabeculations was diagnosed by ECHO, the non-compacted to compacted ratio ranged from 1.6 to 3:1 (mean 2:1). Dilated left ventricle was found in 8 patients, reduced systolic function in 7 (EF 22% to 57%). The cardiac function progressively improved during follow-up in all patients. The mean age of patients with impaired left ventricular function was 5.9 months and the non-compacted to compacted ratio was 2.09:1; in those without left ventricular dysfunction the mean age was 60.8 months ($p < 0,05$) and the ratio 1.93 ($p > 0,05$). Pharmacological treatment with ACEI and spironolacton was used in 8 patients and digoxin in 4 of them.
Conclusions: 1. The clinical course of severe heart failure and significant systolic dysfunction is observed in infants with LVNC. 2. Progressive improvement of the left ventricular function with age is characteristic for LVNC. 3. The non-compacted to compacted ratio has no impact on the left ventricular dysfunction.

1773: PAEDIATRIC CARDIOVERTER-DEFIBRILLATOR IMPLANTATION: STILL A CHALLENGE BUT SAFE AND FEASIBLE

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Background: *Implantable cardioverter defibrillator* (ICD) implantation in children remains a challenge because of their limited body surface area (BSA), making implantation of percutaneous leads difficult, and because of adhesions generated by epicardial patches, which may interfere with normal development of the heart muscle. A new technique of subcutaneous AICD implantation in children with defibrillation coil is reported.
Methods: With the patient supine decubitus, a left anterolateral thoracotomy is performed. The pericardium is opened and an epicardial bipolar electrode for sensing is placed on the left ventricle. The sensing wire is tunnelled subcutaneously and connected to the generator, placed in an abdominal box pocket. A stimulating lead linked to the generator is then tunnelled subcutaneously from the pocket to the back, with the distal end of the coil placed medially to the left scapula.
Results: The technique was successfully applied in 2 children (age 14 and 12 years at implantation) affected by LQTS1 and hypertrophic cardiomyopathy respectively, with hisotry of resuscitation for cardiac arrest. Early after procedure, good values of sensing, pacing impedance, and excellent pacing threshold were found in both cases. Furthermore, the defibrillation threshold was optimal in both cases (14 J). The AICD-PM setting parameters were: brady therapy VVI 40 bpm and tachy therapy with a single VF-window from 194 bpm. At follow-up of 9 and 23 months respectively, electrical parameters and position of subcutaneous lead remained stable. No events of inappropriate shocks were recorded during follow-up.
Conclusions: Placement of AICD with a surgical approach, epicardial pacing and sensing electrode and subcutaneous defibrillation coil, is a feasible, safe and effective procedure in paediatrics. This approach ensures better functioning of the device over time and does not interfere with child growth. Key to success is regular patient follow-up to ensure constant adjustment of sensing parameters for their growth in order to avoid inadequate shocks.

1777: PHARMACOLOGICAL TREATMENT AND LONG-TERM OUTCOME IN PATIENTS WITH LONG QT SYNDROME

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Methods: We observed 17 children at the age from 6 months to 17.5yrs (mean age 10 yrs), 10 girls and 7 boys, with diagnosed long QT syndrome. Patients (pts) were divided into 2 groups: group I – 11 pts (7 girls, 4 boys) with symptomatic long QT syndrome (syncope, dyspnoea); group II – 6 pts (3 girls and 3 boys) with prolongation of QTc segment on ECG but without any clinical symptoms. All pts were in long-term follow-up from 19 months to 7 years (mean observation time 3.5 yrs). Standard 12-lead ECG with assessment of QT interval, corrected QT interval according to Bazett formula and QTc dispersion were done at the beginning and the end of observation period; 24-hour Holter ECG monitoring with QT evaluation was also performed in all pts. All pts from group I were administered beta-blockers (propranolol or metoprolol) with a mean dose of 1.2 mg/kg. Mean standard ECG QTC duration in pts in group I was significantly higher than in group II both in the beginning and at the end of the observation period (471.82 vs 435 ms and 460.91 vs 423.33 ms). A similar result was observed in Qtc duration in Holter monitoring (535 vs 455 ms; 515.91 vs 452.5 ms).There were no significant differences between QTc duration before and after beta-blocker treatment in group I (471.82 vs 460.91 ms) although frequency and intensity of symptoms in this group of pts was expressively diminished. Three pts (27.3%) from group I were referred for ICD implantation.
Conclusions: Beta-blockers diminished clinical symptoms in long QT syndrome. Syncope can be an evident risk factor of sudden cardiac death (SCD) in children. Pharmacological treatment could decrease the SCD risk in children with long QT syndrome.

1778: PARTIAL ANOMALOUS CONNECTION OF PULMONARY VEINS: ANATOMICAL VARIANTS, SURGICAL TREATMENT AND DEVELOPMENT

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Introduction (objective): To spread the institute's experience in the surgical treatment of partial anomalous connection of pulmonary veins (PACPV) and its mid-term results in terms of morbidity and mortality.
Method: A retrospective, longitudinal, observational, descriptive study was undertaken of all patients with PACPV undergoing surgical correction, in the period between January 2000 and December 2010.
Results: Of all 86 patients, 62.5% were male, the average age was 19 years, and the vast majority (91%) were in functional class I, II of NYHA (New York Heart Association). The most common variant of PACPV was two vessels (64.28%), the surgical technique used the most was redirecting the flow through pericardium (66%), followed by the Warden technique (19.6%), and finally direct reimplantation of the anomalous vein (14.2%). There were 3 deaths in the early post-operative period; there were no deaths in late follow-up.
Conclusions: Surgical results of PACPV at our institution were satisfactory in terms of morbidity and mortality, since rates are low and are within the worldwide average. The mid-term prognosis is satisfactory among the remaining patients in NYHA functional class I.

1781: COMPARISON OF EXERCISE CAPACITY IN YOUNG PATIENTS AFTER CORRECTION OF TETRALOGY OF FALLOT AND AFTER ATRIAL SWITCH (SENNING OPERATION) OF TRANSPOSITION OF GREAT ARTERIES IN LONG TERM FOLLOW-UP

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Methods: We observed 49 patients (pts) who were divided into two groups. Group I consisted of 35 patients after correction of tetralogy of Fallot (TOF) (6 females, 29 males), age range from 8.7 to 21.7 yrs (mean 14.6 yrs). Group II consisted of 14 pts after operation of transposition of great arteries (TGA) by Senning method (6 girls, 8 boys) age range from 7.3 to 17.8 yrs (mean 13.1 yrs). In all pts 24-hour Holter ECG monitoring was performed with assessment of heart rhythm and presence of arrhythmias. Treadmill exercise testing (TT) was done in all pts and modified Bruce protocol was used. We estimated total metabolic equivalent (MET), total exercise time, presence of arrhythmias and changes in ST segment. In 9 pts (25.6%) from group I ventricular arrhythmias were noticed, but in 3 subjects only was complex arrhythmia present. In group II dysfunction of sinus node was observed in 5 pts (35.7%). Significant changes of ST segment and dysfunction of right ventricle during TT was present in 5 patients after TOF (14.3%); such changes with additional chest pain could be seen in 6 patients (42.9%) after Senning operation. During TT mean MET parameters achieved by pts after TOF were significantly higher than in pts after Senning operation (14.12 vs 12.76) and TT duration was significantly longer in pts after TOF operation (13:12 vs 10:39). In 3 pts (8.6%) after TOF, reoperation and/or ICD implantation was considered, while another 3 pts (21.4%) needed reintervention.
Conclusions: Patients after correction of TOF have much better long-term prognosis in comparison to patients after Senning operation for transposition of great arteries. Systemic right ventricle dysfunction is frequently observed in patients after Senninng operation of transposition of great arteries. TT could be very useful in determination of high-risk patients after complex congenital heart defect correction.

1787: THE EVIDENCE-BASED 'DECISION ANALYSIS' AND PATIENT/FAMILY SHARED DECISION MAKING IN MANAGEMENT OF CHD

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Decisions in a critical care setting can frequently be difficult and involve several trade-offs and multiple risk-benefit ratios. In the face of this dilemma, the currently most commonly used methods to make a decision in these circumstances depend on the doctor's personal knowledge or experience, nihilism, deferring to other experts, deferring to patients, dogmatism or just blindly following a policy. These methods are not scientifically sound. They not only lack objectivity but more importantly lack evidence supporting the decision, and even sometimes minimise adequate and appropriate involvement of the patient/family. This situation is frequently encountered in making decisions of management of congenital heart diseases.

In the recent era of evidence-based medicine, full arrays of decision support aids have been developed, among which an important method, which is unfortunately infrequently used in PCICU, is the evidence-based Decision Analysis (DA). In this presentation, we will explain this important tool for aiding decision and how it can help in making difficult decisions by utilising the best available scientific evidence together with incorporating the patient/family's own preferences and values with a scientifically based methodology that is not only objective but also is expressed in numerical values of disutility and probability. The presentation will also explain the various tools that can be utilised to achieve proper patient/family shared medical decision making.

1790: CHANCES OF NEWBORNS AND INFANTS TO SURVIVE TO HEART TRANSPLANT IN A LOW DENSITY POPULATION AREA: IMPACT OF MECHANICAL SUPPORT AND ABO INCOMPATIBLE TRANSPLANT

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Background: Prolonged time on a waitlist affects survival pre and post heart transplantation (HTx). We analysed outcomes of patients listed for HTx aged below 3 months in the low population density region of western Canada.
Materials and methods: We reviewed patients listed below age of 90 days from 2006 to 2011, to determine waitlist mortality, outcomes after HTx and factors that could impact outcomes.
Results: Twenty-seven patients were listed during the study period. Congenital heart disease was the predominant diagnosis in 20 patients (74%), and cardiomyopathy, myocarditis and others in the remaining 7. Twelve patients (44%) died or were delisted as a result of clinical deterioration after a median of 32 days (from 7 to 127 days); one was removed after 112 days because of clinical improvement. HTx was performed in 14 (52%) patients after a median waitlist of 51 days (from 2 to 215 days). Mortality in 14 patients who required extra-corporeal life support (ECLS) pre-Tx (9 bridged to transplant, 5 died on waitlist) was not different from patients without ECLS ($p = 0.61$) but time to death on the waitlist trended shorter ($p = 0.09$). In the transplanted group, 7 patients (50%) received an ABO-compatible (ABOc) heart, with 3 post-Tx deaths. The remaining 7 cases received an ABO-incompatible (ABOi) graft and are alive. ABOc patients waited a median of 51 days compared to 49 days for ABOi Tx ($p = 0.53$). Patient death was not associated with prematurity ($p = 0.61$) or birth weight below 2.5 kg ($p = 0.71$). Cumulative survival post-listing was 44%.
Conclusion: HTx outcomes in early childhood are promising; however high waitlist mortality has a negative impact on overall results. Despite having strategies such as ABOi HTx, waitlist mortality in western Canada exceeds rates reported from higher population density areas and is higher than other reported age groups. Further strategies to improve organ availability/allocation are required.

1796: VENTRICULAR SEPTAL DEFECT – AN UNUSUAL SEQUEL OF BLUNT CHEST TRAUMA IN A 7-YEAR-OLD BOY

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Background/hypothesis: Ventricular septal defect (VSD) is the commonest congenital cardiac lesion encountered worldwide. Only very rarely is it acquired, and causation through blunt injury is even rarer.
Materials and methods: A 7-year-old boy suffered blunt trauma to his chest while at play with his peers at school. He had been quite well before then, with no symptoms related to the cardiovascular system and with no growth or developmental delay. He presented 2 days later at our centre with features of acute congestive cardiac failure. Two-dimensional and Doppler echocardiographic examination revealed a rent in the peri-membranous portion of the outlet ventricular septum with the avulsed flap still attached to the septum.
Results: Cardiac failure was refractory to anti-failure therapy and other stabilisation measures. His clinical condition took a rapid turn

for the worse and he succumbed to the ensuing illness 5 days after the trauma, despite stabilisation measures and before any surgical intervention could be undertaken.
Conclusions: Traumatic VSD, though rare, should be considered in cases of acute congestive cardiac failure in otherwise well, active children with a history of blunt trauma to the chest and all such patients should undergo careful echocardiographic evaluation. Prompt surgical intervention has been reported to be life-saving in similar cases.

1799: COMPLETE REPAIR OF TETRALOGY OF FALLOT IN LATE DIAGNOSED PATIENTS

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Aim: In most centres worldwide, correction of tetralogy of Fallot is performed before 9 months of age. However, in developing countries, early repair may be difficult because of many factors, including facilities for the diagnosis. The purpose of this study was to evaluate the early results of surgical repair of tetralogy of Fallot in patients older than a year, in one hospital performing congenital heart surgery in Angola.
Methods: A retrospective analysis was performed of early results of surgical repair in paediatric patients older than 1 year, between June 2011 and June 2012 in one single hospital. Data were gathered from patients' records, preoperative cardiac catheterisation or TC studies, operative intervention and pre- and post-operative two-dimensional echocardiographic scans. Patients submitted to systemic-to-pulmonary shunt were excluded.
Results: In our hospital, 55 patients were treated (male-to-female ratio 1.03:1) with a mean age of 6.4 years (range 13 months to 19 years). Early extubation occurred in the majority of patients (mean 9 hours, range 3–35 hours). Most patients did not present with significant post-operative lesions, except for residual interventricular shunt in 4 cases, mild or moderate pulmonary regurgitation in 6 patients, and mild obstruction of right ventricle outflow in 14 cases. Pleural effusion occurred in 5 patients, with good response to medical treatment. No deaths occurred.
Conclusion: We demonstrated good early results of complete repair of tetralogy of Fallot in paediatric patients with late diagnosis in Angola. The late results follow-up must be evaluated, but this preliminary study reinforces the value of the establishment of local assessment to early diagnosis and treatment of congenital heart diseases in developing countries.

1801: INTERRUPTED AORTIC ARCH: 10 YEARS OF EXPERIENCE IN THE SURGICAL TREATMENT

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Introduction: The interruption of the aortic arch is a rare malformation, representing less than 1% of cases of congenital heart disease, and is associated with 90% mortality if not treated before 1 year of age; death occurs as a combination of the increase of a short circuit form the left to right, ventricular failure and closure of the ductus arteriosus, resulting in hypoperfusion, renal failure and metabolic acidosis. Initial treatment is to maintain a patent ductus arteriosus with prostaglandin administration. Surgical correction is the definitive treatment and must be performed to confirm the diagnosis.
Method: A retrospective, longitudinal, observational, descriptive study of all patients with interrupted aortic arch was performed who underwent surgery for correction of this pathology in the NIC, in the period between January 2000 and December 2010.
Results: Of 20 patients, 16 (80%) presented with type B interruption, 3 (15%) with interruption type A and 1 patient (5%) with interruption type C. The average age at which surgery was performed was 2.9 months (range 3 days to 7 months). The surgical technique used was end-to-end anastomosis. One patient died (5%). At follow-up, the rest of the patients are asymptomatic and without reintervention.
Conclusions: Despite late referral of many patients with interrupted aortic arch, surgical results and developments, assessed by monitoring, are similar to those reported in the world literature.

1803: PERSISTENT LEFT SUPERIOR VENA CAVA DRAINING INTO LEFT ATRIUM WITH NORMAL CORONARY SINUS

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The most common variation in the thoracic systemic venous system is a persistent left superior vena cava draining into a coronary sinus. A rare anomaly is a persistent left superior vena cava connecting directly to the left atrium. In this situation it is believed that the coronary sinus must be absent.

We report an unusual case of a left superior vena caval drainage to the left atrium with normal coronary sinus, which was a preoperative finding during surgical closure of an atrial septal defect in an 11-year-old patient. We rerouted left superior vena caval flow into the right atrium using intra-atrial baffle. The postoperative course was uneventful.

In this case report, we discuss embryological development, clinical profile and surgical techniques to treat this condition.

1806: NORMAL CARDIOVASCULAR RESPONSES TO TREADMILL EXERCISE TEST IN HEALTHY BRITISH CHILDREN

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Background: Normal cardiovascular responses to exercise in childhood are not well defined. Maximum normal blood pressure response to exercise in childhood is overestimated which makes assessment of hypertensive response in disease situations rather difficult.
Aim: To assess normal cardiovascular responses to exercise in healthy British children.
Method: A retrospective review was carried out on all children who underwent exercise testing (Bruce test using a treadmill protocol) at a tertiary institution between 2003 and 2010. One hundred and thirty-seven healthy children (80 males, 57 females) aged 9–16 years were included in the study.
Results: Minimum exercise duration was 12 minutes across the spectrum regardless of age and gender. Although lower VO2max values were attained in females compared to male subjects, there was little change throughout adolescence years. All subjects achieved over 85% of maximum predicted heart rate for age. Younger subjects showed quicker heart rate recovery compared to older individuals. Maximum blood pressure did not exceed 155 mmHg in any age group. Rate pressure product was lower in males compared to females in most age groups but similar in 13–14-year-old group. All values were summarised in a table.
Conclusions: Exercise duration in healthy children is minimum 12 minutes. 85% of maximum heart rate response is more achievable in children than maximum predicted heart rate hence it may be more preferred in clinical decision making. Maximum blood pressure

response is much lower than historically quoted which may necessitate redefining hypertensive response to exercise in childhood.

1823: PROOF OF CONCEPT OF A SMART PHONE-BASED PHONOCARDIOGRAPHIC SYSTEM

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Introduction: Clinical auscultation remains essential for the diagnosis of congenital heart disease in the developing world. In screening programs of Lanzhou First University Hospital, Children's HeartLink, First Affiliated Hospital of Kunming Medical University and China-California Heart Watch in People's Republic of China, children were screened with phonocardiography (iAusc, $n = 52$), standard auscultation (sAusc, $n = 19$) and echocardiography (echo). Our hypothesis is that iAusc detection of murmurs is sensitive to facilitate referral to a cardiac centre.
Methods: Children were examined with an electronic stethoscope connected to an iPhone® that stored, retrieved and transmitted the iAusc data, and then with a commercial stethoscope. Endpoints include pathologic vs innocent murmur, location and intensity for iAusc and their overall concurrence with echo diagnosis. An echocardiogram was performed using standard techniques. Initial difficulty with high-pitched vibratory innocent noises required different earphones.
Results: Organic murmur was differentiated from innocent murmur in each case with both iAusc (48/52, LAP) and sAusc (19/19). iAusc was recorded from the four primary cardiac exam areas on the chest, URSB, ULSB, LLSB, apex. Principal diagnoses included: ventricular septal defect (23), pulmonary stenosis (3), atrial septal defect (5), aortic stenosis (2), normal (10). Sensitivity for murmur type was 48/52 or 92%. Specificity for innocent murmur was 10/14 (71%). Pulmonary hypertension was present for 3 of 4 murmurs in heart disease misdiagnosed as innocent.
Conclusion: Phonocardiography can be used to detect congenital heart disease. We report a proof-of-concept of iAusc to detect heart disease in underserved areas. The iAusc tracings can be emailed to remote experts from a cloud-based server, providing cost-effect access to care with reduced travel for disadvantaged children and a link to the cardiac referral centres. The methodology can provide a venue for international volunteerism for cardiologists based in advanced countries accounting for unfamiliar challenges such as Eisenmenger pulmonary hypertension.

1831: REDUCTION OF RADIATION DOSE FOR CORONARY COMPUTED TOMOGRAPHY USING A 128-SLICE DUAL-SOURCE MACHINE IN CHILDREN AND YOUNG ADULTS WITH CORONARY LESIONS AFTER KAWASAKI DISEASE

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Background: Coronary artery lesions (CAL) of coronary aneurysm followed Kawasaki disease (KD) have progressed into stenosis, occlusion and calcification. Coronary computed tomography (CCTA) has provided much information on the diagnosis of CAL after KD. CCTA using a conventional 64-slice single-source machine provided high radiation exposure. A 128-slice dual-source CT (DSCT) can take coronary artery images very fast and can reduce of the radiation dose.
Aim: The aim of this study is to evaluate the reduction of radiation dose and recognition of the CAL by DSCT.
Patients and methods: A total of 35 patients (median age 11 y 6 m, range 2 y 5 m–45 y) were examined. All patients were administered beta-blockade to reduce the heart rate. Six children were sedated and the other 29 patients could breath-hold. Scan parameters were as follows; mAs adapted body weight (100–310 mAs) at 70 Kv, 80 Kv, 100 Kv and 120 Kv, with prospective ECG gated. To assess radiation dose, we recorded the dose-length product (DLP) in mGy cm and the effective dose in mSy estimated from the DLP. We used two modes to take CCTA, named flash spiral mode (FSM) and adaptive flash cardio mode (AFCM).The quality of images for proximal and middle segments of the right and left coronary arteries was evaluated.
Results: A total of 29 patients (median age; 11 y 6 m) were examined by FSM, and 6 patients (median age; 11 y 0 m) were examined by AFCM. Total DLP was extremely low (median 45 mGy cm) and effective radiation dose was 0.74 mSy (median) by FSM. Total DLP was very low (median 128.5 mGy cm) and effective radiation dose was 1.93 mSy (median) by AFCM. The image of 1 patient (7 y) was not taken by FSM because of movement.
Conclusion; Prospective ECG-gated DSCT can provide adequate CAL images in children and young adults. These methods are associated with extremely lower radiation doses.

1832: ANALYTICAL COMPARISON OF CARIOVASCULAR RISK FACTORS FOR ADULT CONGENTIAL HEART DISEASE AND NORMAL CONTROLS: A CASE-CONTROL STUDY

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Background: Objectives of this study are to identify cardiovascular risk factors in adults with congenital heart disease (CHD) and to provide basic materials for developing media to lower the risk factor levels.
Methods: This study conducted a survey of 240 people in total, including 120 adult patients with CHD and 120 patients in the control group whose selection was based on age, sex, and body mass index (BMI). The survey was conducted regarding fasting blood glucose (FBS), lipid profile, apoprotein A-1/B, occurrence of carotid stenosis, and environmental influences.
Results: Compared to the control group, the CHD group had significantly high FBS, HDL, and apoprotein A-1, but significantly low total cholesterol, LDL, and apoprotein B. The two groups had significant difference in occurrence of carotid stenosis, exercise, and smoking. Regarding differences by sex between the two groups, males of the CHD group had lower total cholesterol and LDH than males of the control group; females of the CHD group had higher FBS and lower total cholesterol than females of the control group. However, there was no difference according to sex in the CHD group. Comparison of acyanotic and cyanotic CHD patients in the CHD group showed that acyanotic CHD patients had higher FBS, total cholesterol and LDH, and lower apoprotein than cyanotic CHD patients. When age and sex were adjusted, cyanotic CHD patients without surgery, cyanotic CHD patients with surgery, acyanotic CHD patients with surgery, and acyanotic CHD patients without surgery in this order had a high risk of developing metabolic syndrome ($p < 0.001$). BMI and smoking were identified as variables influencing metabolic syndrome.
Conclusions: It is believed that regular follow-up of risk factors, BMI control, and education about non-smoking may reduce the risk of developing metabolic syndrome in adult patients with CHD.

1836: POST-RHEUMATIC CARDIAC VALVULAR LESIONS MANAGEMENT IN A TERTIAIRY SUB-SAHARAN CENTRE: THE EXPERIENCE OF THE CARDIAC CENTRE, SHISONG

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Abstract: The aim of the study was to investigate the early post-surgical mortality, and challenges in the care of operated children in St. Elizabeth Catholic General Hospital Cardiac Centre.
Patients and methods: This retrospective analysis included 56 patients aged between 6 and 17 years old who underwent mitral repair or replacement and/or aortic valve replacement from November 2009 through June 2012. Data from patients' records, operative intervention, and preoperative and postoperative two-dimensional echocardiographic studies were reviewed.
Results: A total of 56 patients aged between 6 and 17 years old with a mean age of 10 ± 3.4 years underwent surgical correction of mitral and/or aortic valvulopathy. Mitral regurgitation was the commonest echocardiographic diagnosis present in 51.7% patients; 13.3% patients had mixed mitral valve disease, 35% had pure mitral stenosis. Before surgery, 8 patients were in class IV, 12 in class III, 25 in class II and 11 in class I according to the New York Heart Association classification. Patients were extubated from 5 to 10 hours after surgery with low doses of inotropes. The mean stay in intensive care unit was 1.5 ± 0.5 days. In the early post-surgical period, the ejection fraction (EF) changed from 45.3 ± 1.5 % to 56.1 ± 1.4% ($p < 0.005$) in 3 months and stayed almost the same after 6 months 57,2 ± 2,7% ($p > 0.05$); at 9 months it was 55.1 ± 1.8% ($p > 0.05$), and at 12 months 58.4 ± 1.7% ($p > 0.05$); however the basal part of the interventricular septum was hypokinetic.
Conclusion: Post-rheumatic mitral valve regurgitation is the pathology most encountered. Post-surgical echocardiogram is characterised by motion abnormalities of the basal part of the interventricular septum. Because of financial limitations, poverty and illiteracy of parents, the post-surgical follow-up of patients is challenging.

1840: ANOMALOUS ORIGIN OF THE PULMONARY ARTERY FROM THE ASCENDING AORTA: TWO INSTITUTIONAL REVIEWS OF CASES FROM 1991 TO 2012

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Introduction: Anomalous origin of one pulmonary artery from the ascending aorta is a rare congenital anomaly associated with early onset of pulmonary hypertension and irreversible pulmonary vascular disease.
Methods: A retrospective clinical review ws done of 18 cases presenting to the Divisions of Paediatric Cardiology at the Chris Hani Baragwanath Academic Hospital and Inkosi Albert Luthuli Hospital, which are both tertiary care institutions in South Africa. Data collected included clinical features, diagnosis, operative procedures, pre-operative procedures and post-operative follow-up.
Results: Sixteen infants, 1 child and 1 adult (11 males, 6 females) were diagnosed. The most common presenting features were respiratory distress, a cardiac murmur, congestive cardiac failure and failure to thrive. Median age at presentation was 67 days. Diagnosis was made with echocardiography and confirmed with CT angiogram in 5 patients and angiography in 5 patients. There were 16 patients with anomalous origin of right pulmonary artery arising from the aorta (AORPA) and 2 cases of anomalous origin of left pulmonary artery (AOLPA). Patients were divided into three categories: isolated lesions (7); simple lesions with patent ductus arteriosus (8) and complex lesions (3). One patient with AOLPA had CATCH 22 and a second patient with AOLPA also had Mckusick-Kaufman syndrome. Five patients had successful direct re-implantation of the right pulmonary artery. One patient died the day after surgery following a pulmonary hypertensive crisis, one patient died 20 days after surgery from sepsis, and three patients remain well on follow-up. Three patients were deemed inoperable. The remaining 10 patients died before surgery could be undertaken.
Conclusion: There is a high mortality associated with anomalous pulmonary artery arising from the aorta without surgery. A good outcome can be expected with early surgery before pulmonary vascular disease.

1841: INFECTIVE ENDOCARDITIS IN CHILDREN: A CROSS-SECTIONAL STUDY OF 32 CASES IN OUAGADOUGOU, BURKINA FASO

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Introduction: Infective endocarditis is a transplant of a micro-organism on an often injured endocardium. It is rare in children. This study aimed to determine the frequency of endocarditis in children, and to describe the clinical presentation, data from echocardiography, microbiological profile and clinical course.
Patients and methods: From 1 May 2010 to 30 April 2012, we consecutively included children admitted for infective endocarditis at two medical centres in the city of Ouagadougou: Saint Camille Medical Centre and the teaching hospital Yalgado Ouedraogo.
We investigated the functional and general signs and treatment already received. Physical examination included investigation of an infectious syndrome, pneumonia, heart failure and portals of entry. Blood cultures, blood count, creatinine, blood chemistry, HIV status, electrocardiogram, chest radiography and cardiac Doppler ultrasound were systematically done. The diagnosis of the disease was based on Duke criteria.
Results: Thirty-two cases of endocarditis in children were reported to be 1.5 % of admissions. The average age was 4.6 ± 2.5 years (1–14). The female:male sex ratio was 1.5. Clinical presentation was a common infectious syndrome. Impaired general condition and congestive heart failure were present on admission in 14 cases, respectively. The portal of entry was dental in 14 cases (43.7%), and skin and ENT in 6 cases (18.7%) respectively. A peripheral vein was implicated in 2 cases. In four other cases no portal of entry was found. HIV serology was positive in 6 cases. Blood cultures were positive in 26 cases, for streptococcus in 20 and staphylococcus in 6. Echocardiography revealed vegetation in all cases, localised to the mitral valve in 14 cases. Multiple locations were found in 8 cases. Underlying heart disease was dominated by rheumatic valve disease (19 cases. Treatment consisted of antibiotics, antipyretic treatment and that for heart failure as appropriate. Surgery was indicated in 3 patients as an emergency but they were unable to benefit because of inadequate technical support. Four deaths (25%) occurred as a result of septic shock.
Conclusion: Infective endocarditis in children is common in our practice. The common clinical syndrome is infectious. Streptococcus and staphylococcus are the two organisms found. Surgical forms are immediately lethal. The global prognosis remains poor.

1843: IMPACT OF SOUTH AFRICAN GUIDELINES ON THE USE OF HIGH-SENSITIVITY TROPONINS: A PATHOLOGY LABORATORY PERSPECTIVE

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Introduction: Guidelines on the use of high-sensitivity cardiac troponin (hs-cT) for the diagnosis of myocardial infarction (MI) in South Africa have recently been published. The aim of the study was to evaluate the effect of implementation of the guidelines on hs-cT laboratory measurements.

Method: The study was conducted as a retrospective, comparative study. Results were anonymised by using codes to protect patient privacy. Clinical presentation was not recorded. The fourth-generation hs-cT assay (Roche Diagnostics, South Africa) was employed for hs-cT measurements. Clinical significance of effect was evaluated on data excluding children aged < 12 years.

Results: The total number of requests did not change significantly but a significant shift in distribution of values within hs-cT categories (< 15 ng/l; 15–52 ng/l; 53–100 ng/l; ≥100 ng/l) was observed (+8.6 and –22% for categories 2 and 3 respectively; $\chi^2 = 21.4$; $p < 0.0001$). Fourteen per cent of values exceeded 100 ng/l for both groups. Significantly higher repeat rates were observed following implementation of the guidelines (33 vs 42%; $\chi^2 = 5.9$; $p < 0.015$) as well as lower numbers of patients within hs-cT category 2 with a > 50% increase in hs-cT (18 vs 9%: $\chi^2 = 4.9$; $p < 0.026$). Thirty-five per cent of patients within hs-cT category 3 currently present with a > 20% increase in hs-cT. Serial daily measurements (> 3 days) were requested in 3% of all patients.

Conclusion: Although this study does not specifically inform on the use of hs-cT to diagnose MI, a change in laboratory observations is evident. Implementation of the guidelines appears to be beneficial, but repeat rates are still unacceptably low, irrespective of the indication for the test.

NURSING SCIENCE

400: PARENT EDUCATION DISCHARGE INSTRUCTIONS: A PILOT STUDY, HYDERABAD, INDIA

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Background: Every day nurses worldwide assist patients and families in the transition from hospital to home. Currently, in economically developing countries, children with complex cardiac defects are receiving treatment for their chronic illnesses and are requiring more expanded parental education programmes to manage care after discharge. The aim of this study was to test the feasibility of training Indian nurses to conduct discharge teaching for parents. The study team developed a sustainable computer-based nurse training, the PEDI programme, to increase nurses' knowledge and support role expansion. Our secondary goal was to validate culturally relevant training materials for parents and nurses.
Methods: Institutional review board and ethics committee approvals were obtained. This investigation used a pre-/post-study design to evaluate user satisfaction, nurses' knowledge of discharge teaching content, and documentation of discharge teaching conducted with parents. After nurse training throughout a tertiary Indian paediatric heart hospital, individualised and group parent teaching post-operative training sessions were implemented on the ward. Convenience samples of 40 nurses, 25 parents, and 25 patient charts were obtained pre- and six months post-implementation of the PEDI programme. Focus testing of parents and nurses and retrospective patient chart audits were performed.
Results: There was a 15-point increase in nurses' discharge knowledge, from a mean of 80.6 to 95.7% ($p = 0.0005$) after participation in the training. Nurses' and parents' reported high levels of satisfaction with the parent education material (3.75–4 on a four-point scale). Evidence of discharge teaching in patient medical records improved from 60% (15/25 charts) to 96% (24/25 charts) after implementation of the PEDI programme.
Conclusions: Nurses can play an integral role in educating parents about postoperative care at home for children with complex cardiac defects. Future studies are needed to examine nurse, child, parent and organisational outcomes related to this expanded nursing role in developing countries.

420: EFFECT ANALYSIS AND NURSING METHODS STUDY OF THE MODIFIED BLALOCK-TAUSSIG SHUNTS OPERATION IN CHILDREN WITH CONGENITAL HEART DISEASE

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Objective: Through summarising the therapeutic efficacy of the modified Blalock-Taussig shunts (MBTs) used on children with congenital heart disease (CHD), we aimed to study the methods of peri-operative nursing care and the reasons for complications, and to use the appropriate care counter-measures to increase children's survival rate and improve their quality of life.
Methods: A retrospective study was performed to summarise the peri-operative treatment and nursing process and the subsequent complications of the 94 children undergoing MBTs operation. The peri-operative monitoring and nursing methods included: (1) All the children underwent pre-operative examinations, and low-flow oxygen inhalation and prostaglandin E_1 was administered to them to prevent the closure of the ductus arteriosus. (2) We continuously monitored the children's postoperative cardiac function. (3) We kept a balance of intake and output, electrolytes and acid–base ratios. (4) We used peritoneal dialysis early on. (5) We implemented effective respiratory management. (6) We strengthened the anticoagulant drug application and venous access management to prevent postoperative catheter embolisation.
Results: Ninety-four children's detailed postoperative information was collected, 76 children survived and 18 died. In-hospital mortality was 19.1%. The postoperative S_pO_2 of the survivors increased, which was significantly improved when compared with pre-operative S_pO_2 ($p = 0.000$). Fifteen children have already had the radical (definitive) operation and their postoperative condition is good.
Conclusion: MBTs is a good palliative treatment for children with CHD, as it can improve their hypoxic situation and increase pulmonary blood flow so as to improve the success rate of surgery. Rational use of oxygen, early use of prostaglandin E_1, improved monitoring of postoperative cardiac function, implementing effective respiratory management, early use of peritoneal dialysis, accurate application of anticoagulant drugs, and comprehensive training of parents could significantly increase the surgical success and survival rate and improve the quality of life of the patients.

863: PAEDIATRIC CARDIOTHORACIC SURGICAL SITE INFECTION PREVENTION: A MULTIDISCIPLINARY APPROACH

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Background: Surgical site infection prevention (SSIP) is a vital part of ensuring that all patients experience a complication-free journey throughout their healthcare experience. A great deal of research has been done in the adult healthcare setting on how to prevent surgical site infection but there remains little evidence of SSIP within paediatrics.
Method: Great Ormond Street Children's Hospital initiated an independent SSI team in 2010. The aim was to independently review in-patient's wounds on alternate days and follow them for 30 days post procedure. The cardiothoracic unit multidisciplinary team worked together with the department of infection prevention to establish a care bundle that would be suitable for cardiothoracic patients, from pre-admission to discharge. All patients now follow a specific care bundle that includes the following elements:

- improved MRSA screening compliance
- patient information on SSI and surveillance
- pre-operative washing
- patient skin preparation
- information post discharge
- unit protocol development on visitors, chest closure procedures on CICU, and wound care
- timely investigations into all surgical site infections.

Essential to our work is the ability to follow our patient's journey through his/her recovery up to 30 days post-surgery. We believe we have extended our wound-care program into the community.
Results: The cardiothoracic unit has been able to follow up 80% of all surgical patients. Our results show we have had a 10% reduction in SSI and we currently have a 1% infection rate for deep and organ-space infections. We feel this is due to the multidisciplinary approach to the SSIP.
Conclusions: Since the monitoring process has been in place, we can demonstrate a reduction in infection rates, which ultimately improves our patient experience in their healthcare journey. We're now looking at extending and developing our methods further to continue our rate reduction.

930: RESEARCHING THE OUTCOMES OF AN EDUCATIONAL INTERVENTION WITHIN THE WORKPLACE: A FLEXIBLE WORKFORCE FIT FOR PRACTICE?

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Background: High-dependency nursing of seriously ill cardiac children is fast paced and requires a high level of attention to detail,

outstanding assessment skills and deep critical thinking. These skills are not imbued in the nurse on completion of basic registration training. During a time of change, one of the matrons identified a need for development of the knowledge and skills of practitioners and made contact with the university.
Aim: The matron working in collaboration with UNN helped develop a simulation-based training module but ultimately wanted to know if following this intervention would make the workforce fit for practice, and whether the intervention made the nurses competent paediatric HDU practitioners.
Methods: The educational module uses theory and practice through simulation and reflection on real practice, which is grounded in real case studies, to allow staff to develop the requisite knowledge and skills to recognise and provide emergency care for critically ill children. A collaborative journey began between the matron and the senior clinical academic from the University, which resulted in nurse competence assurance in the recognition of the sick and critically ill child for nurses working in cardiac care.
Results: A research study carried out to assess the cardiac nurse's perception of the impact of the module concluded that this had enhanced the underpinning of knowledge and improved clinical skills in the recognition and safe care of critically ill children.
Conclusion: This has allowed continuing partnership between academic and practice organisations to ensure that provision of educational modules meets identified local and wider workforce development needs. Further research has taken place following on from this initial study.

963: THE IMPORTANCE OF A STRUCTURED NURSING ASSESSMENT OF A PAEDIATRIC CARDIOTHORACIC PATIENT: THE POSITIVE RESPONSE TO CHANGE

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Background: With the changing nursing role and the dependency of their cardiothoracic patients, the importance of patient assessment and excellent structured communication has been considered essential in maintaining patient safety.
Methods: Great Ormond Street Children's Hospital cardiothoracic unit initiated a radical change in January–March 2011 to the way their high- and low-dependency wards assessed their patients. The new structure was devised to recognise the in-depth assessment that is now required in the simple and complex cardiothoracic patient. The implementation of the new structure was supported by senior nurses experienced in the advanced assessment of patients. As part of the development, the University affiliated with the Trust was also consulted, to ensure that the tool met academic standards.

The nursing team concluded that to be succinct and effective, known structures of assessment would need to be adapted to create a new tool. The tools that were used in establishing the new structure were the ABC approach to patient assessment previously established by the Resuscitation Council UK and the 'SBAR' approach originally devised by the naval military. The new tool was implemented over a four-week period with small group training at the patient bedside.
Results: Following implementation, our results showed 100% compliance with the tool. At one year we still maintain an average 95% compliance with the tool. Our figures are only affected by the employment of agency staff, which we are striving to train, to ensure we will again achieve 100% compliance.
Conclusion: We feel that we are achieving high compliance in the use of our tool, as it has been designed by nurses for nurses, and the simplicity of the tool is paramount to our successful implementation. We would now like to share our experience internationally.

999: 'THE IMPACT OF MODERNISING THE WAY WE NURSE': A NURSE'S EXPERIENCE IN MOVING THE CARDIOTHORACIC WARD TO A NEWLY DESIGNED AND BUILT ENVIRONMENT

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Background: The increase in patient numbers and the dependency of patients means the cardiothoracic unit needs to evolve. The availability of a new building to expand into is one that needs precise planning and communication.
Methods: Great Ormond Street Children's Hospital cardiothoracic unit expanded into a new building, increasing capacity to eight high-dependency beds, with 16 ward beds. The unit was designed to provide maximum privacy and well-being for all patients. The nursing process required change as patients were to be nursed in single cubicles not open bays. The facilities of the unit were also improved for patients and visitors, with break-out rooms, play rooms and adolescent facilities.

The change in nursing methods meant preparing the nursing staff was vital to the move. A system of training days was established to provide support in learning how to manoeuvre patients around the ward, carry out emergency procedures, and become familiar with modern technology that would be available to the patients, as well as newly designed patient safety equipment and technology. Nursing staff were given the opportunity to develop working strategies on team days, which were facilitated by senior staff. All strategies were implemented.
Results: The patients were all moved safely and effectively within two hours. All nursing staff completed a follow-up questionnaire to identify lessons to be learned for a further move in four years' time. All staff stated they felt supported throughout their training and the effective communication they had regarding the move gave them confidence in caring for their patients.
Conclusions: We believe that by sharing our experience of transitioning to a new unit we can demonstrate that the key to a happy, effective workforce and patient satisfaction is effective communication and empowering nurses to become involved in the decision-making processes of establishing new working practice.

1002: EVALUATION OF A NUTRITIONAL STATUS INTERVENTION IN CHILDREN WITH CONGENITAL HEART DISEASE

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Objective: To investigate the feeding of children with catch-up growth after nursing intervention.
Methods: From October 2010 to January 2011, children younger than six months with non-cyanotic congenital heart disease (CHD) who had undergone surgery at the Shanghai Children's Medical Centre were selected by convenience sampling. Random allocation was taken to divide them into two groups (n = 56 and n = 58). The control group followed the current clinical feeding regime and education, while the experimental group was given feeding guidance from hospitalisation to post-discharge follow up. During hospitalisation, feeding guidance, including primary feeding information, feeding information especially for children with CHD, feeding behaviour information, and high-calorie feeding principles were given to the children's parents. In addition, the handbook *Feeding Guidance for Children with Congenital Heart Disease* was distributed to them at discharge. The nurses followed them up for three months and continued to guide the parents on how to feed according to the handbook.

In this study, information was collected according to the Baseline Questionnaire for Children with CHD, Feeding Knowledge Questionnaire for Parents, Feeding Index Questionnaire, and State Anxiety Inventory. The effect of the feeding guidance follow-up plan, which aimed at improving parents' feeding knowledge, regulating feeding behaviour, and easing feeding anxiety was evaluated by feeding knowledge scores, feeding index, and level of feeding anxiety scores.
Results: After following them up for three months, the feeding knowledge of the caregivers improved ($p < 0.05$), the feeding index

was higher ($p < 0.05$), and the levels of feeding anxiety significantly decreased ($p < 0.05$) in the experimental group compared with the control group. There was no statistical difference on HAZ, WAZ, and WHZ between the two groups ($p > 0.05$).
Conclusion: Taking the feeding intervention programme could significantly enhance parents' feeding knowledge, relieve their feeding anxiety, and ameliorate feeding behaviour in the short term, however, it failed to promote children's postoperative catch-up growth in the short term.

1011: TETRALOGY OF FALLOT POSTOPERATIVE COMPLICATIONS AND NURSING

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Objective: To provide evidence of clinical nursing care through exploring and analysing the postoperative complications of tetralogy of Fallot (TOF).
Methods: This retrospective analysis was performed to select patients under one year old who underwent TOF operations in our hospital from January 2009 to December 2011, and to describe postoperative complications of these patients.
Results: Patients (556) with different postoperative complications were selected into this study. The following complications: low cardiac output syndrome, cardiac arrhythmia, ventilation-associated pneumonia, atelectasis, hyoxaemia and pleural effusion ranked among the top six, with prevalences of 3.7, 3.6, 3.1, 2.5, 2.0 and 1.8%, respectively.
Discussion: This study highlights the main complications of infant patients after TOF operation, provides evidence of the nursing care, and guides nurses to take some preventive measures with these patients.

1031: STORIES FROM THE HEART: A PROTOCOL FOR INTERVIEWING CHILDREN WITH COMPLEX CONGENITAL HEART DISEASE FROM EDMONTON, CANADA

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Background: Research to date on families and children living with complex congenital heart disease (CHD) has largely involved perspectives from caregivers and healthcare professionals, not the children themselves. Also missing is the perspective of their siblings. Children exposed to repeated invasive medical and surgical interventions from an early age face significant morbidity and considerable stress. The perspectives of these children are necessary to give direct voice to their experiences and effectively plan for intervention.
Objective: The purpose of this project was to identify evidence-based strategies for interviewing children for the development of an interview protocol to gain children's perspectives on living with complex CHD. The research question was: How can we best facilitate children and their siblings to tell their stories about growing up with CHD?
Findings: A protocol was developed based upon literature reviews regarding involving children in research. Key aspects of interviewing children incorporated into the protocol included creating an atmosphere of trust and respect; obtaining consent, assent and maintaining confidentiality; engagement through the participatory process; the use of props and guiding questions to facilitate storytelling; and strategies to reduce power inequities. The protocol was piloted with children with complex CHD ranging in age from five to 15 years. The final protocol consists of a diverse repertoire of interview activities to elicit the stories of children as well as contact with parents before and after the interview with the child.
Conclusion: Hearing from children as they live with complex CHD provides a voice for their experiences and needs. Their perspective also helps clinicians and researchers in planning for family-based interventions. This study provides valuable data regarding the feasibility of obtaining data through stories with this population, and further contributes to the methodological literature concerning involving children with CHD in research.

1032: THE RELATIONSHIP OF CHILD AND FAMILY FUNCTIONING IN FAMILIES OF CHILDREN WITH COMPLEX CONGENITAL HEART DISEASE: A PILOT STUDY

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Background: Parents play a critical role in optimal child development and healthy family functioning. Children who survive their complex congenital heart disease (CHD) live with chronic health issues and can have motor and cognitive disabilities that present parenting challenges. Understanding a range of parent perceptions and family functioning is foundational to developing interventions that optimise child, parent and family well-being.
Aim: The purpose of this pilot study was to: (1) describe a cohort of parents of children with CHD as per their parent and parenting characteristics (i.e. family management style, resilience and parenting stress), and their perceptions of their child with CHD [i.e. child's development, adaptive behaviours, and health-related quality of life (HRQOL)], (2) identify trends through the analysis of individual family profiles, and (3) determine if the questionnaires were effective in identifying the potential intervention needs of parents.
Findings: Questionnaires were sent to 24 families, with a total of 16 families responding. The mean age of the children with CHD was 7.1 years. Most family management styles were child/family focused. A significant difference on the resilience score was found between mothers and fathers. There was a large range in parenting stress, from within a normal range for total stress to a clinically significant range. The mean scores on all indexes of child development and adaptive behaviour were within the normal range. Regarding HRQOL, all children scored poorly on the dimensions of heart problems and treatment, treatment anxiety, and communication. Family profiles indicated higher parenting stress, lower resiliency scores, and a condition-focused family management style in families with a child with more co-morbidities, greater developmental needs, and sub-optimal HRQOL.
Conclusion: Results indicate a range of parent and parenting characteristics. The questionnaires were effective in delineating the needs of parents at different stages of a child's CHD trajectory.

1034: WHAT ARE THE TIMING, SETTING, AND NATURE OF PARENT INTERVENTIONS FOR MOTHERS AND FATHERS OF CHILDREN WITH CONGENITAL HEART DISEASE: A SCOPING REVIEW

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Background: Evidence of the plight of parents of children with congenital heart disease (CHD) has been published since soon after the first successful cardiac surgery programmes were established in the 1950s. Mothers have been the predominant source of parental data. Maternal anxiety, fear, stress and uncertainty have been shown to influence child outcomes, although not in proportion to disease severity. Within this growing body of descriptive research there are repeated recommendations for parental interventions. The research question guiding this scoping review was: What are the timing, setting, and nature of parental interventions for mothers and fathers of children with CHD?
Methods: A scoping review was conducted with 707 articles identified from six bibliographic databases. Two reviewers independently screened articles by title and abstract for inclusion in the review.
Results: Thirteen articles representing 12 intervention studies from

10 centres published between 1975 and 2010 constituted the evidence base for this review. Most interventions occurred within the first few months of the child's birth and/or diagnosis. Most were conducted in the hospital or clinic. One study involved an interactive website and another evaluated a video-conferencing intervention. Seven interventions involved a combination of parent education and psychological support. Four studies concerned home-based management of anticoagulation therapy. Regarding study design, seven studies had a comparison or control group, and two studies involved randomisation. Five studies included mothers only, three included mothers and fathers, and four studies did not specify numbers of mothers and fathers.
Conclusion: Interventions beyond the time of diagnosis are needed as parents face further transitions with their child, such as additional surgeries and entering preschool. Timely parental intervention is required to optimise child and family outcomes. Settings other than hospitals and clinics are needed. Innovative technology to provide interventions at home and interventions for both mothers and fathers are needed.

1073: THE EVALUATION OF A SHORT-TERM EDUCATIONAL STRATEGY: CAN THIS IMPROVE KNOWLEDGE AND CONFIDENCE AMONG PARTICIPANTS IN IRAQ?

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Background: Congenital heart disease in the developing world creates a significant burden of disease and to date the development of sustainable health systems that will treat and manage these conditions has not been a global priority. The International Children's Heart Foundation (ICHF) is a charitable organisation that collaborates with local teams in the developing world to build paediatric cardiac surgical programmes. This project aims to evaluate the effectiveness of a two-week educational intervention in a paediatric cardiac intensive care unit in Iraq during a collaborative surgical visit. The research question is: Does a two-week, mixed-method educational programme have an effect on the knowledge and self-reported confidence level of participants in a paediatric cardiac intensive care unit in Iraq? There is little consensus on competence when applied to clinical practice, however, confidence and knowledge are attributes that have been closely linked with the ability to perform and have been shown to guide future behaviours.
Methods: Participants on the course will be asked to complete a pre- and post-course questionnaire, the ICU Education Evaluation Questionnaire (IEEQ), which has been designed and tested to measure knowledge and self-reported confidence levels before and after the educational intervention. The project will seek to establish correlation links between the identified variables, namely, the education programme, knowledge and self-reported confidence levels of participants.
Results: Data were collected from 8 to 22 September 2012. It is anticipated that there will be a difference in both confidence and knowledge among the group.
Conclusions: The ability of ICHF to tailor future educational interventions toward the requirements of the local team will improve the uptake of education and improve the competence of the whole team.

1150: PAEDIATRIC NURSE PRACTITIONER-MANAGED CARDIOLOGY CLINICS: PATIENT SATISFACTION AND APPOINTMENT ACCESS

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Background: Appointment waiting times exceeded 40 days in the out-patient cardiology department at a children's hospital. To address the gap in available appointments, paediatric nurse practitioner-managed cardiology clinics were implemented.
Methods: A sample of 128 patients who presented concurrently in physician or paediatric nurse practitioner-managed cardiology clinics was recruited for participation. The hospital's ambulatory patient satisfaction survey was used to measure level of patient satisfaction with healthcare. Survey responses were evaluated using Fisher's exact test. Appointment waiting times were compared pre- and post-implementation of paediatric nurse practitioner-managed clinics.
Results: Sixty-five physician and 63 paediatric nurse practitioner families completed the satisfaction survey. There was no statistically significant difference in patient satisfaction between clinic types. Appointment waiting time decreased from 46 to 43 days, which was not statistically significant, but was clinically important. Paediatric nurse practitioner clinics evaluated a statistically higher percentage of urgent appointments compared to physician clinics. With the addition of several weekly nurse practitioner-managed cardiology clinics, current waiting times in the out-patient cardiology department are now at less than one week.
Conclusions: Paediatric nurse practitioner-managed cardiology clinics are a strategic solution for improving patient access and facilitating high-quality patient care while earning high levels of patient satisfaction. This healthcare delivery model illustrates successful expanded utilisation of advanced practice nurses.

1180: PERCUTANEOUS PULMONARY VALVE IMPLANTATION: EXPERIENCES OF THE PATIENT AND THEIR CLOSE RELATIVES

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Background: Percutaneous pulmonary valve implantation (PPVI) reduces the number of open-chest procedures during childhood and adolescence. As operative survival has improved, the focus of PPVI has turned to quality-of-life aspects and patient-assessed experience of treatment. This study evaluated the physical and psychosocial aspects of daily life of patients and relatives undergoing PPVI.
Methods: Ten consecutive patients were included in the prospective, qualitative study from April 2007 to June 2011. Patients and close relatives participated in individual in-depth interviews one day before and three months after PPVI. All patients and close relatives had previously experienced cardiac surgery and subsequent paediatric cardiac intensive care.
Results: This less-invasive procedure resulted in an earlier return to daily life and activities compared with previous experiences (median 2.4 days in hospital), with patients resuming their social role and function. Close relatives stated that both the short hospital stay and improved function of their child was of benefit to the child, the family and society.
Conclusion: This study shows that striving for normality of life is a main goal for both patients and their relatives. In facilitating patients to achieve optimal social function in school, at home and with peers, PPVI appears to offer a favourable approach, due to the minimal interference in daily life. Furthermore, this study allows for the evaluation of our health system from both a patient and a family perspective.

1268: INCREASED KNOWLEDGE REQUIRED IN ADULTS WITH RHEUMATIC HEART DISEASE: THE CAPE TOWN EXPERIENCE

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Background: The REMEDY study is a comprehensive, contemporary data collection on patients with rheumatic heart disease (RHD). It is a hospital-based international registry in progress, involving 20 sites in Africa and India. This abstract focuses on the patients enrolled at Groote Schuur Hospital (GSH) in Cape Town.
Methods: Over a two-year period, 420 patients with RHD were interviewed and enrolled into the REMEDY study from the cardiac clinic out-patients department at GSH. At enrolment, demographic data, clinical findings, treatment practises, ECG and echocardiographic details were recorded onto structured record forms.
Results: Among a study population of 420 patients, 52% of the patients were in the age group 30 to 50 years. Eight per cent were severely debilitated in their daily functioning (NYHA III and IV). According to the patients' past medical history, 6% have had major bleeding, 19% stroke, 19% valvuloplasty and 66% valve surgery. A total of 44.7% had been prescribed oral anticoagulation therapy. Of concern was the finding that 40% of patients had two or fewer INR measures in the previous six months, while only 4% had monthly INR checks. In addition, 62% did not know what their goal INR should be. Only 14% of patients had been prescribed secondary prophylaxis, although 66% have had surgery.
Conclusion: These preliminary results highlight the need to improve the knowledge of both health practitioners and patients with regard to treatment practices.

1312: A PILOT STUDY OF A SKIN-TO-SKIN CARE INTERVENTION IN INFANTS WITH CONGENITAL HEART DEFECTS

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Aim: Infants born with complex congenital heart defects (CCHD) demonstrate impaired parasympathetic nervous system function. Early separation impairs infant parasympathetic function, and newborn infants with CCHD and their mothers are often physically separated due to the infants' need for intensive care. Skin-to-skin contact (SSC) is effective in improving infant parasympathetic function in other high-risk infant populations. This study examined the feasibility, acceptability and safety of an intervention designed to enhance infant parasympathetic function through daily SSC between mothers and newborn infants with CCHD.
Methods: Ten infants with CCHD and their mothers were recruited. Mothers provided at least one hour of SSC for 14 consecutive days post-operatively, shortly after initiation of nipple feedings. Feasibility and acceptability were measured with a survey and mothers' written records of duration and frequency of holding. Safety was measured by infants' cardiorespiratory stability during SSC. Measures of parasympathetic function, as indexed by high-frequency heart rate variability (HF-HRV), were collected at feedings before and after the intervention and bi-weekly for four weeks.
Results: Mothers provided a daily mean of 66.58 (SD = 4.85) minutes in SSC over the 14-day intervention. Mothers were positive in their evaluation of SSC, and no adverse events occurred. Improvements in parasympathetic function over time were demonstrated with (1) developmentally appropriate increases in the magnitude of baseline (pre-feeding) HF-HRV ($t = 40.01$, $p < 0.001$), (2) consistent improvements in time to post-feeding HF-HRV recovery, as calculated using event history analysis, and (3) gradual permanent improvements in adaptive parasympathetic responses to feeding in six of 10 infants, as calculated using ARIMA time-series analysis.
Conclusions: SSC is a safe, feasible and acceptable intervention for infants treated for CCHD as neonates. Improved parasympathetic responses to feeding were demonstrated. A randomised clinical trial is needed to more closely examine the effects of SSC on parasympathetic function.

1439: FREQUENT ADVERSE EVENTS IN INFANTS WITH SINGLE-VENTRICLE ANATOMY DURING THE INTER-STAGE PERIOD

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Background: Infants with single-ventricle anatomy s/p post stage 1 repair or those with shunt-dependent lesions are at continued risk for morbidity and mortality during the inter-stage period. Home monitoring programmes (HMP) were developed to detect worsening oxygen saturations, dehydration and growth failure. This study reviews inter-stage adverse events in infants followed in a HMP.
Methods: This was a single-centre retrospective review of 53 infants with single-ventricle anatomy who were in a HMP during the inter-stage period (time from initial discharge to Glenn procedure) over a two-year period, 2009 to 2011. Inter-stage adverse events (changes in oxygen saturation, poor feeding or poor weight gain, inter-current illness and other health problems) were evaluated.
Results: There were 32 infants with HLHS (60%) and 21 infants with other types of single-ventricle anatomy; inter-stage mortality was 5.5%. The average inter-stage period was 119 days. There were 145 adverse events; 50 for changes in saturation, 46 for poor feeding or inadequate weight gain, 29 for inter-current illnesses (bacterial and viral infections) and 19 for numerous other health problems. While not specifically targeted on HMP, infections and other medical problems comprised 33% of events. Management involved ER evaluation for 17 events, and 73 hospital admissions. There were 33 unexpected catheterisations or surgical procedures. Eight patients (15%) had no adverse events; only two patients with HLHS had no events. Eleven patients (21%) had more than five events. Of the nine patients with naso-gastric (NG) or gastric (G)-tube feedings, all had one or more adverse events. Nine patients (17%) needed prolonged hospital care.
Conclusion: HMP detected many adverse inter-stage events of varying aetiologies, which were frequent and often required hospitalisation. A third of the adverse events involved inter-current illnesses or other medical problems. Patients with NG or G-tube feeds and those with HLHS were more likely to have adverse events.

1466: CHARACTERISTICS OF POST-OPERATIVE CARDIAC SURGICAL PATIENTS READMITTED TO THE PAEDIATRIC INTENSIVE CARE UNIT

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Background: The paediatric intensive care unit (PICU) is a 20-bed integrated general and surgical unit within a university-affiliated tertiary children's hospital. The hospital performs approximately 700 cardiac surgical procedures per year, providing comprehensive care for the treatment of neonates, infants, children and young adults with complex congenital cardiac lesions. The PICU Liaison Program was developed in 2005 to facilitate discharge, and support patients and families on the wards. In an effort to reduce readmission rates to PICU, the Liaison Nurse team developed a risk-management tool known as the Clinical Marker Score (CMS). It is used to identify predictive risk factors for readmission during the discharge and follow-up process. The tool is a combination of both objective and subjective score components derived from a case–control regression analysis of PICU discharge and readmission data. It is a generic tool used on PICU patient populations both prior to discharge and on every follow-up visit by the liaison nurse.
Aim: To test the hypothesis that predictors of readmission will vary across the general and cardiac surgical patient population in the PICU. The purpose of this study was to describe the characteristics of cardiac surgical patients readmitted to the PICU.
Methods: This was a retrospective review of all postoperative cardiac surgical patients readmitted to the PICU within 72 hours of discharge. The data period was between January 2005 and June 2012. No interventions were performed in this study.

Results: There were 4 368 cardiac surgical patients discharged from PICU during this six-and-a-half-year period. Of these, 197 (4.5%) were readmitted. The most common cause for readmission was respiratory dysfunction, primarily resulting from effusions, infection or upper airway obstruction. The second highest cause was cardiovascular dysfunction, predominantly sepsis, cardiac failure or arrhythmias.
Conclusions: Respiratory complications were the most common cause for early PICU readmission among paediatric cardiac patients. Further development of the subjective/objective score components specific to this patient group will improve discharge planning and avoid readmission to the PICU.

1659: ACADEMY OF CARDIOVASCULAR EXCELLENCE: DEVELOPMENT OF A CARDIAC CURRICULUM FOR PROFESSIONAL DEVELOPMENT OF STAFF IN A PAEDIATRIC ACUTE-CARE HOSPITAL SETTING

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Background: Caring for patients with congenital heart disease poses many challenges that may impact on clinical performance, productivity, patient safety and outcomes. These challenges are constantly changing due to many factors, such as staff turnover and skills mix, continued technological advances in care, and increasing complexity of children and adults presenting with paediatric cardiovascular disease. We determined the need for a standardised cardiac educational programme that could be offered to all staff on an on-going basis, providing an opportunity for continued development of the knowledge and critical thinking skills necessary to meet these challenges. The Academy of Cardiovascular Excellence (ACE) was developed.
Methods: In 2008, a multidisciplinary group with representatives from the cardiovascular ICU, cardiac catheterisation laboratory, cardiac operating room and non-invasive cardiology in a paediatric acute-care hospital met frequently to discuss the development of this programme. Discussion centred around the core knowledge-based objectives that were felt to be essential in providing care to this patient population along the continuum of care, encompassing all of these clinical areas.
Results: A 48-hour core curriculum was developed and offered over an eight-week period, six hours each week, at a minimum of three times per year. Didactic sessions, group discussion and case-based scenarios promoting critical thinking allowed for the application of content to the participant's specific practice area. An 18-hour advanced curriculum, offered in three sessions of six hours each, enhanced the core knowledge provided in the curriculum.
Conclusions: Course evaluations have indicated a high level of satisfaction with the content and delivery of the ACE programme as well as providing feedback related to opportunities for improvement in the course content since its inception. The programme continues and is evolving further to meet the changing needs of the patient care environment.

1714: NEONATAL SCREENING WITH PULSE OXIMETRY IN 10 858 NEONATES: MULTICENTRE PROJECT IN NORTH-EAST BRAZIL

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Introduction: Congenital heart disease accounts for approximately nine in every 1 000 neonates. Early diagnosis and treatment is needed in about one-third of the cases. When optimised, the arterial pulse oximetry test (POT) adds to physical examination as a low-cost and efficient screening tool.
Objective: To report the results from arterial pulse oximetry and physical examination with a telemedicine network established between two states in north-east Brazil.
Methods: This was a multicentre, descriptive, prospective study from 12 centres between January and July 2012. Nursing teams from all centres were trained to collect data and input in a database. POT was considered normal when two examinations from the right arm and one foot had SpO_2 > 95% and the difference between them was below 2%. Physical examination focused on the presence of murmurs and examination of peripheral pulses. Neonates with abnormal physical examinations or POT were referred to a neonatal screening echocardiogram.
Results: A total of 10 858 POT were performed; 554 were abnormal and 214 babies were referred to a screening echo performed by a neonatologist under cardiological supervision. There were 138 abnormalities detected. The relationship between abnormal POT/collected POT decreased from 14.8% in January to 3% in July. The number of echoes performed was below the number of abnormal POTs except in July.
Conclusion: Neonatal screening for CHD in the state of Paraiba started this year and has already evaluated over 10 000 neonates. The fall in abnormal POTs in recent months reflects a learning curve with the test. Isolated, POT accounted for the detection of 15.5% of CHD, the remaining cases being screened by physical examination alone (81.2%) or physical examination and POT (3.3%). The POT test is easy to perform but requires training and the establishment of routines to be reliable. Physical examination remains the most important tool for screening for CHD.

1780: FACTORS INFLENCING MEANING OF LIFE IN ADOLESCENTS WITH CONGENITAL HEART DISEASE

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Purpose: This study was designed to identify variables associated with meaning of life in adolescents who underwent cardiac surgery for congenital heart disease (CHD).
Methods: Data were collected from 120 adolescents with CHD, aged 15 to 18 years, during out-patient clinic follow up after open-heart surgery in one major cardiac centre in Korea. Adolescents completed measures of meaning of life, self-esteem, self-control, parental attitude, family satisfaction, school adjustment and career maturity. Their New York Heart Association functional class and non-invasive saturation of arterial oxygen were also measured.
Result: The mean total score for the meaning of life scales was 2.05 points, which was slightly low. There was a significant relationship between meaning of life and self-esteem, self-control, parental attitude, family satisfaction, school adjustment, career maturity and functional class. The multiple regression analysis also showed that 53% of the variance in meaning of life in adolescents with CHD could be explained by self-esteem, family satisfaction, school adjustment, parental attitude and functional class.
Conclusion: These results indicate a need to develop nursing interventions to increase self-esteem, family satisfaction, school adjustment, parental attitude and functional class in order to improve meaning of life for adolescents with CHD.

1797: SEVERE LEFT VENTRICULAR COMPACTION IN EARLY CHILDHOOD

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Background: Left ventricular non-compaction is characterised by hyper-trabeculated myocardium. There are deep recesses communicating with the left ventricular chamber, where blood penetrates, with the risk of clots developing. Adult prevalence is estimated at 50 per

100 000 but only 2% will develop symptoms within the first decade of life. We present such a case in an infant.

History: A four-week-old male infant developed poor feeding and increasing breathlessness. He presented collapsed and in shock at six weeks of age, requiring intubation and inotropic support. His worst gas had a pH of 6.96 with a lactate of 16. Echocardiogram revealed a poorly functioning hypertrophic dilated left ventricle with non-compaction. After one week he was extubated.

Receiving regular anticoagulation, diuretics and beta-blocker treatment, he remained clinically asymptomatic for the next 12 months other than having frequent respiratory infections. He grew and developed normally despite dramatically poor echocardiographic left ventricular function (rocking left ventricle with fractional shortening < 10% and abnormal speckle tracking). He was negative for Barth syndrome testing (a known association). At one year of age he rapidly decompensated, requiring ventilation and inotropes. There was no improvement and he was urgently listed for cardiac transplantation. Unfortunately he died from an intracranial haemorrhage while on a Berlin heart ventricular assist device awaiting transplantation.

This case was particularly challenging as the prognosis was so uncertain, with poor outcome reported in 25% of children, while others remain asymptomatic. This causes the family great difficulty with coping. From the onset we were open in sharing the uncertain future with the family, giving psychological as well as medical and nursing support. This child was relatively well until the final deterioration.

Conclusion: Left ventricular non-compaction in childhood is relatively rare and has a variable severity and an undulating course. This case demonstrates the severe end of the spectrum.

1813: PARENTING/CAREGIVING WORK OF MOTHER-AND-FATHER COUPLES WHO HAVE AN INFANT WITH A COMPLEX CONGENITAL HEART DEFECT

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Background: Parents of infants with a complex congenital heart defect (CCHD) often describe caregiving demands and challenges. Little is known about how parents manage with caregiving as a couple through the infant's first year. Although parents have identified the other parent as the most likely source of support, they have also described providing this support as the most challenging aspect of parenting.

Objective: To learn what parents were working on together to parent and give care to an infant with a CCHD through the first year of life, how they were working on caregiving, and agreement or differences in perspectives on the infant or about parenting/caregiving.

Methods: Parents (mothers and/or fathers) of an infant with a CCHD were interviewed in their homes at one, four or six, and 12 months after the infant's birth. Parents responded to semi-structured interview questions concerning parenting experience, including what parents were working on, a question to learn about motivations, goals and intentions concerning the infant, self, and the parent couple.

Results: Among 21 caregiving parents, including three fathers, approximately half were doing parenting work as a couple, 30% were attempting to parent as a couple, and the remainder had no prospect of couple parenting. Disagreement on the infant's condition or what the baby needed for care or parenting was higher when couples were not parenting together, either cooperatively, collaboratively, or in tandem.

Conclusions: Differences in perspective on infant needs may put couples at risk for parenting/caregiving difficulties. Motivations for parenting/caregiving changed for some couples through the infant's first year. Two motivations likely to have continuity were to maintain the couple's relationship with the qualities it had before the infant's birth, and to keep attuned to each other, i.e. to 'stay on the same page'. Studies of support to couples' accomplishment of parenting/caregiving goals are needed.

831: THE IDENTIFICATION AND VERIFICATION OF PUTATIVE KCNE2-INTERACTING PROTEINS WITH RELEVANCE TO LONG-QT SYNDROME

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Background: Long-QT syndrome (LQTS) is a cardiac repolarisation disorder characterised by a prolonged QT interval on an echocardiogram (ECG). The symptoms of LQTS range from minor symptoms such as dizziness and syncope to more severe symptoms such as seizures and sudden cardiac death (SCD). Clinical features of LQTS are a result of the precipitations of torsades de pointes, which is a form of polymorphic ventricular tachycardia. A number of genetic forms of LQTS have been identified with more than 900 mutations in 10 different genes leading to disease pathogenesis. However a large percentage of LQT-affected patients have no mutations within the known LQT genes. Of these known mutated genes, *KCNE2* is one that is associated with LQT6. This is a beta-subunit of potassium ion channels of which the mutations are mainly located in the C-terminal domain. We hypothesised that genes encoding for proteins that interact with *KCNE2* might be identified as disease-modifying genes and this study aimed to use a yeast two-hybrid (Y2H) analysis in order to identify and verify putative interactors of the C-terminal domain of *KCNE2*.

Methods: The C-terminal domain of *KCNE2* was used as bait to screen a pre-transformed cardiac cDNA library using Y2H analysis. Putative interactors will be verified using 3D-colocalisation and co-immunoprecipitation experiments.

Results: A number of putative *KCNE2* interactors were identified by Y2H and are currently being verified. These include filamin C (FLNC), protein tyrosine phosphatase (PTPRK), crystallin alpha B (CRYAB), voltage-dependent anion-selective channel protein 1 (VDAC1), titin (TTN) and cardiac actin (ACTC1).

Conclusion: The genes encoding verified interactors will be screened in our South African panel of LQT patients, to potentially identify novel LQT causative genes. Furthermore, the interactions verified in this study may shed some light on the mechanism of pathogenesis of LQT causative mutations in *KCNE2*.

BASIC SCIENCES

83: FOETAL AND NEONATAL DUCTUS ARTERIOSUS IS REGULATED WITH ATP-SENSITIVE POTASSIUM CHANNEL

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Background: The foetal patency and neonatal closure of the ductus arteriosus (DA) is regulated with oxygen and prostaglandins. The proposed oxygen sensors of foetal and neonatal DA include P450-endothelin and the Kv channel. We hypothesised that the ATP-sensitive potassium channel (K_{ATP} channel) is another oxygen sensor.
Methods: Foetal and neonatal DA were studied in Wistar rats using sulfonylurea drugs, including tolbutamide, chlorpropamide, gliclazide, glimepiride and glibenclamide (K_{ATP} channel inhibitors), diazoxide and pinacidil (K_{ATP} channel openers: KCOs), and rapid whole-body freezing.
Results: Tolbutamide, chlorpropamide and gliclazide easily passed across the placenta, and dose-dependently constricted foetal DA following orogastric administration to near-term pregnant rats. The foetal DA constricted 30% with clinical doses of sulfonylurea drugs, and closed completely with larger doses. Glimepiride and glibenclamide passed across the placenta minimally, and only mildly constricted the foetal DA after maternal administration, but constricted and closed the foetal DA dose-dependently with direct foetal injection. Foetal DA closure was associated with hydrops and foetal death. Diazoxide and pinacidil delayed DA closure following neonatal injection immediately post-natally, and dilated the closed the DA with injection 30 minutes post-natally.
Conclusions: All tested sulfonylurea drugs constricted foetal DA dose-dependently and with complete closure at large doses. KCOs dilated the neonatal DA. These results indicate physiological regulation of foetal and neonatal DA with K_{ATP} channels. This study has several clinical implications. Sulfonylurea-associated foetal death was first reported in South Africa 50 years ago. The mechanism of death remained unclear prior to this study. Sulfonylureas may be useful for closing patent DA in premature neonates. The recently reported reopening of neonatal DA associated with the use of diazoxide for hyperinsulinaemic hypoglycaemia has been proved experimentally. The DA-dilating effect of KCO drugs may be useful as a bridge to surgery in neonatal DA-dependent congenital heart diseases.

163: COMPARISON OF CONTRACTILE PERFORMANCE OF PATIENTS WITH AND WITHOUT MUCOPOLYSACCHARIDOSIS UNDERGOING CARDIAC VALVE REPLACEMENT

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Objective: Progressive valve pathology is the most prominent cardiac manifestation in patients with mucopolysaccharidosis (MPS). Valve and papillary muscle thickening and short chordae lead to valve regurgitation and are associated with systolic and diastolic dysfunction. There is some evidence that differences in venous return might cause decreased preload and reduced end-diastolic volume and ventricular pressure, but the underlying mechanism remains unclear.
Methods: Tissue from six patients (three MPS, three non-MPS) undergoing aortic and/or mitral valve replacement was obtained from the right auricle, transported in an oxygenated Krebs-Henseleit solution and skinned with Triton-X. We performed three experiments on each patient ($n = 18$). The fibres were exposed to a gradual increase in calcium concentration and the corresponding force was measured and recorded.
Results: (1) Calcium-induced contraction was statistically significantly different between patients with and without MPS ($p = 0.03$). (2) The non-MPS male fibres showed significantly higher force values compared to males with MPS ($p = 0.0002$). (3) Female fibres did not show significant differences in contractile behaviour between MPS and non-MPS fibres.
Conclusion: Our data showed a statistically significantly different contractile behaviour of male and female fibres with and without MPS. A lowered calcium sensitivity, which leads to a deficiency in the Frank-Starling mechanism might be the physiological correlation for this observation and may be a clinical indication for optimised pre-operative treatment of these patients.

164: IMPACT OF GENDER ON CONTRACTILE BEHAVIOUR OF SKINNED PAPILLARY MUSCLE OF FEMALE AND MALE PATIENTS WITH MUCOPOLYSACCHARIDOSIS

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Background: Mucopolysaccharidoses (MPS) are a group of metabolic disorders caused by the absence of lysosomal enzymes, which cause dermatan and heparan sulfate to accumulate in the tissue. There is a lack of information regarding the impact of gender on these lysosomal storage diseases.
Methods: We examined right atrial tissue of three patients with MPS I or II ($n = 9$, three preparations per patient) and significant stenosis of the mitral (Δp mean = 17 ± 11.5 mmHg) and aortic valves (Δp mean = 40 ± 21 mmHg) and undergoing valve replacement. Human tissue was obtained from the right atrium before implementation of ECC, and it was prepared and stimulated with a stepwise increase in calcium-containing solution. The results were recorded.
Results: All fibres developed the greatest force (mean: 1.71 ± 0.75 mN) at the highest calcium concentration (pCa 4.5). Male MPS fibres developed statistically significantly less force then female fibres ($p = 0.016$). The contractile behaviour of the female fibres differed significant from the male fibres ($p = 0.02$ for MPS male 1, and $p = 0.03$ for MPS male 2). Calcium sensitivity, i.e. pCa^{2++}_{50} was also statistical significant different between male and female fibres (pCa 5.5, female vs male fibres, $p = 0.01$).
Conclusion: The data showed differences in contractile performance in these fibres and imply a different sensitivity in male and female MPS patients. We feel it is justified to report these limited results because this is an extremely rare kind of disease. It may help to consider specific peri-operative treatment for patients with MPS.

198: MESENCHYMAL STEM CELL-MEDIATED REVERSAL OF BRONCHOPULMONARY DYSPLASIA AND ASSOCIATED PULMONARY HYPERTENSION

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Clinical trials have failed to demonstrate an effective preventative or therapeutic strategy for bronchopulmonary dysplasia (BPD), a multifactorial chronic lung disease in preterm infants, which is frequently complicated by pulmonary hypertension (PH). Mesenchymal stem cells (MSCs) and their secreted components have been shown to prevent BPD and pulmonary fibrosis in rodent models. We hypothesised that treatment with conditioned media (CM) from cultured mouse bone marrow-derived MSCs could reverse hyperoxia-induced BPD and PH.
Methods: Newborn mice were exposed to hyperoxia (FiO2 = 0.75) for two weeks, then treated with one intravenous dose of CM from

either MSCs or primary mouse lung fibroblasts (MLFs), and placed in room air for two to four weeks. Histological analysis of the lungs harvested at four weeks of age was performed to determine the degree of alveolar injury, blood vessel number and vascular remodelling. At six weeks of age, pulmonary artery pressure (PA acceleration time) and right ventricular hypertrophy (RVH; RV wall thickness) were assessed by echocardiography, and pulmonary function tests were conducted.

Results: When compared to MLF-CM, a single dose of MSC-CM-treatment (1) reversed the hyperoxia-induced parenchymal fibrosis and peripheral PA devascularisation (pruning), (2) partially reversed alveolar injury, (3) normalised lung function (airway resistance, dynamic lung compliance), (4) fully reversed the moderate PH and RVH, and (5) attenuated peripheral PA muscularisation associated with hyperoxia-induced BPD. A microRNA expression analysis on the hyperoxia-induced BPD lungs ± MSC-CM intervention is underway.

Conclusion: To the best of our knowledge this is the first evidence that reversal of key features of hyperoxia-induced BPD and its long-term adverse effects on lung function can be achieved by a single intravenous dose of MSC-CM *in vivo*, thereby pointing to a new therapeutic intervention for chronic lung diseases, including pulmonary hypertensive vascular disease.

210: REMOTE ISCHAEMIC PRECONDITIONING WITH, BUT NOT WITHOUT METABOLIC SUPPORT PROTECTS AGAINST ISCHAEMIA–REPERFUSION INJURY IN THE NEWBORN PIGLET IN VIVO

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Introduction: While remote ischaemic preconditioning (rIPC) protects the mature heart against ischemia–reperfusion (IR) injury, the effect of rIPC on the neonatal heart is controversial. The neonatal heart relies almost solely on carbohydrate metabolism, which is modified by rIPC in the mature heart. Glucose–insulin (GI) infusion provides myocardial substrate supplementation, which may compensate for the adverse metabolic effects induced by rIPC in the immature heart.

Hypothesis: rIPC combined with GI improves cardiac function and reduces infarct size compared to control or rIPC alone after IR injury in neonatal piglets *in vivo*.

Methods: Thirty-two newborn (one to four days old) piglets were randomised into four groups: control, rIPC, GI and GI+rIPC. The GI and GI+rIPC groups received insulin in 20% glucose at a rate corresponding to 100 mU/kg/h continuously from 40 minutes prior to ischaemia. The rIPC and GI+rIPC groups underwent four cycles of five-minute limb ischaemia followed by five minutes of reperfusion. Myocardial IR was induced by 40 minutes of occlusion of the left anterior descending artery followed by two hours of reperfusion. Left ventricular pressure was measured using 3-F Millar microtip catheters. Interstitial lactate was measured using microdialysis, and infarct size was measured using triphenyltetrazolium chloride staining.

Results: Systolic recovery (dP/dt_{max} as % of baseline) after two hours' reperfusion was improved in GI+rIPC (84.7 ± 5.3%) compared to the controls (71.2 ± 4.9%, $p < 0.05$) and rIPC (33.9 ± 12.9%, $p < 0.01$) but was not different from the GI group (82.9 ± 8.1%, ns). Lactate levels (% of baseline) were lower in the GI+rIPC (85.5 ± 4.9%) compared to controls (125.1 ± 9.1%, $p < 0.01$) and rIPC group (233.9 ± 31.1%, $p < 0.01$). Infarct size relative to area at risk was 12.7 ± 1.1% in GI+rIPC compared to 16.4 ± 1.5% controls ($p = 0.06$), 18.1 ± 0.8% in rIPC ($p < 0.01$) and 24.1 ± 2.1% in the GI group.

Conclusion: rIPC+GI, but neither rIPC nor GI alone protected the neonatal porcine heart against IR injury *in vivo*. rIPC alone appears to have had detrimental metabolic and functional effects that were compensated for by simultaneous metabolic support with GI infusion.

251: IRON HOMEOSTASIS PLAYED A CRITICAL ROLE IN THE PROCESS OF CARDIOMYOCYTE HYPERTROPHY DURING LEFT VENTRICULAR RETRAINING FOR CHILDREN WITH TRANSPOSITION OF THE GREAT ARTERIES

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Background: The left ventricle (LV) regresses after the neonatal period in patients with transposition of the great arteries (TGA) with an intact ventricular septum (IVS) or restrictive ventricular septal defect (VSD). Pulmonary artery banding (PAB) can induce cardiomyocyte hypertrophy for the subsequent arterial switch operation. We screened the altered plasma proteins after PAB and explored the implications for LV retraining.

Methods: Eight late-referral children with TGA/IVS or small VSD were enrolled in part I of the study. The plasma was collected 30 minutes before and 48 hours after PAB. Differential gel electrophoresis (DIGE) proteomics was used to identify the altered proteins. The significantly changed proteins were then confirmed by ELISA. In part II of the study, children with TGA ($n = 16$) undergoing PAB and matched children undergoing open-chest surgery without cardiopulmonary bypass ($n = 12$) were enrolled and the changed level of the chosen proteins was measured. In part III of the study, cell size and mRNA expression of the biomarkers for cardiac hypertrophy were evaluated in a neonatal cardiomyocyte culture model with the associated protein.

Results: Proteomic analysis revealed significant change in 25 proteins. Furthermore, ELISA analysis showed three differential proteins, including ceruloplasmin (CP), transferrin (TF) related to iron ion homeostasis, and parvalbumin (PVALB) related to heart development, were regulated 1.37-, 1.33-, 1.38-fold, respectively. These changes were confirmed in part II of the study to exclude the involved inflammatory response during open-chest surgery. The *in vitro* study showed that after 48 hours' incubation with TF, the size of cardiomyocytes increased 1.94 times. Meanwhile, the expression of natriuretic peptide precursor A and B and PVALB was significantly enhanced.

Conclusions: Augmented levels of CP and TF indicated iron homeostasis played a critical role in the process of immature cardiomyocyte hypertrophy during LV retraining. TF could directly promote cardiomyocyte hypertrophy and hold therapeutic/prognostic potential in clinical practice.

272: ENDOGENOUS SULPHUR DIOXIDE REGULATES MONOCROTALINE-INDUCED PULMONARY VASCULAR COLLAGEN REMODELLING IN RATS

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Background: Mechanisms for pulmonary hypertension remain unclear. Sulphur dioxide was recently discovered to be generated endogenously in vascular tissue and to have important vascular functions; however, the role of the endogenous sulphur dioxide pathway in the pathogenesis of pulmonary vascular collagen remodelling has not been defined. The objective of the present study was to explore the role of sulphur dioxide, a new gaseous signal, in the regulation of pulmonary vascular collagen remodelling induced by monocrotaline and its regulatory mechanisms.
Methods: A rat model of monocrotaline-induced pulmonary hypertension was developed and administered with an L-aspartate-β-hydroxamate and sulphur dioxide donor to evaluate the effects of sulphur dioxide on pulmonary vascular collagen remodelling. The endogenous sulphur dioxide pathway and collagen metabolism were examined. Transforming growth factor-β_1-stimulated cultured pulmonary arterial fibroblasts were used to further the study.
Results: The results showed significant pulmonary hypertension and pulmonary vascular collagen remodelling in association with the augmented sulphur dioxide pathway. L-aspartate-β-hydroxamate further increased pulmonary artery pressure and aggravated pulmonary vascular collagen remodelling, accompanied by decreased sulphur dioxide production. However, sulphur dioxide donor treatment decreased pulmonary artery pressure, attenuated pulmonary vascular collagen remodelling with inhibited collagen synthesis and augmented collagen degradation, and decreased transforming growth factor-β_1 of the pulmonary arteries. Furthermore, sulphur dioxide could prevent activation of the p38 signalling pathway as well as abnormal collagen synthesis in the fibroblasts.
Conclusions: Up-regulation of the endogenous sulphur dioxide pathway played a protective role in pulmonary artery collagen remodelling induced by monocrotaline. The mechanisms may involve inhibition of the transforming growth factor-β_1 expression and activation of the p38 signalling pathway.

278: EFFECT OF SULPHUR DIOXIDE ON PULMONARY ARTERY PRESSURE IN RATS WITH EXPERIMENTAL ABDOMINAL AORTA AND INFERIOR VENA CAVA SHUNTING

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Objective: To explore the effect of sulphur dioxide on the pulmonary artery pressure of rats with abdominal aorta and inferior vena cava shunting.
Methods: Twenty-five male Sprague Dawley rats were randomly divided into a shunting group, a shunting with sulphur dioxide group, and a control group. Abdominal aorta and inferior vena cava shunting was produced in rats in the shunting group and in the shunting with sulphur dioxide group. After an eight-week shunting, the pulmonary artery pressure of each rat was evaluated using the right cardiac catheterisation procedure.
Results: Compared with the control group, the mean, systolic and diastolic pulmonary artery pressures were raised in the shunt rats (72.88, 58.33 and 43.35%, respectively). Compared with the shunting group, the systolic pulmonary artery pressure of rats in the shunting with sulphur dioxide group decreased by 22.12% ($p < 0.05$), the mean pulmonary artery pressure decreased by 20.63% ($p < 0.05$) but the diastolic pulmonary artery pressure did not change ($p < 0.05$).
Conclusion: Sulphur dioxide decreased the mean and systolic pulmonary artery pressures of rats with abdominal aorta and inferior vena cava shunting.

291: ISCHAEMIA AND ISCHAEMIC PRECONDITIONING IN THE HYPERTROPHIC AND FAILING RIGHT HEART

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Aim: To investigate the response to ischaemia and ischaemic preconditioning in the hypertrophic and failing right heart.
Methods: Male Wistar rats ($n = 74$) were subjected to a sham operation ($n = 24$), moderate pulmonary trunk banding (mPTB, $n = 24$) or severe pulmonary trunk banding (sPTB, $n = 26$). Four weeks after the operation, the right ventricle (RV) weight and function was evaluated. Hearts were quick-frozen ($n = 28$) or isolated and perfused in Langendorff apparatus ($n = 46$) with Krebs-Henseleit buffer. The perfused hearts were randomised to IPC (2×5 min of global ischaemia) or no preceding ischaemia (CON), followed by 40 minutes of global ischaemia and 120 minutes of reperfusion. Measurement of the infarct size/area-at-risk (IS/AAR) ratio and post-ischaemic RV function was used to evaluate the effect of IPC on the right ventricle. The quick-frozen hearts were used to evaluate key components of the GPCR/NPR-AKT-eNOS-PKG and RISK pathways.
Results: The mPTB procedure caused compensated RV hypertrophy and the sPTB procedure caused RV hypertrophy with failure. Hypertrophy of the RV caused an increase in infarct size in hearts from mPTB and sPTB animals compared to sham rats (IS/AAR: 66.5 ± 3.4; 59.3 ± 2.4 vs 35.6 ± 2.9% respectively, $p < 0.0001$). Cardioprotection by IPC was possible in sham and mPTB hearts, measured by a decrease in IS/AAR and improved haemodynamic recovery of RV contractile function. The mPTB procedure did not cause an increase in RV cGMP. In sPTB hearts with hypertrophy and failure, IPC did not improve IS/AAR or haemodynamic recovery and an increase in RV cGMP was observed.
Conclusion: Right ventricular hypertrophy increased infarct size, and when failure was present, abolished cardioprotection by ischaemic preconditioning in the right ventricular myocardium of the rat. The abolition of cardioprotecion was followed by an increase in myocyte cGMP. It will be investigated whether there was a causal connection.

337: MUTATIONS IN CALMODULIN AND VENTRICULAR TACHYCARDIA, SYNCOPE AND SUDDEN CARDIAC DEATH

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Primary arrhythmias such as long QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia (CPVT) may cause syncope and sudden cardiac death. CPVT might be suspected with a history of exercise/emotion-related syncope, sudden cardiac death, normal QT_c on ECG and normal structure of the heart.
Methods: A genome-wide linkage analysis of a large four-generation Swedish family with CPVT-like disease but without mutations in the cardiac ryanodine receptor gene (RYR2) was performed. A candidate gene involving calcium homeostasis was analysed for mutations using sequencing. The functional consequences of the identified mutation were determined using a calcium-binding assay.
Results: A novel locus for a severe dominantly inherited CPVT-like disease was identified on chromosome 14q31-32. Sequencing

revealed a heterozygous mutation (Asn5311e) in CALM1 gene encoding for calmodulin, the intracellular calcium sensor and signalling protein. When screening a collection of 61 arrhythmia samples negative for RYR2 mutations, a second *de novo* missense mutation (Asn97Ser) in an Iraqi patient with CPVT-like disease was found. Both mutations demonstrated compromised calcium binding and the Asn97Ser mutation elicited a defective interaction with RYR2, exclusively occurring at a low calcium concentration.
Conclusion: Missense mutations in calmodulin have been demonstrated. The molecular mechanism of a defective calmodulin-RYR2 interaction may lead to arrhythmias. Calmodulin mutations can be tolerated but might cause CPVT-like disease with syncope and sudden cardiac death.

470: CONTRACTILE PROPERTIES OF HUMAN LYMPHATIC VESSELS *IN VITRO*: A NEW PERSPECTIVE ON PROTEIN-LOSING ENTEROPATHY FOLLOWING FONTAN OPERATION

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Background: Protein-losing enteropathy (PLE) is a devastating late complication after the Fontan operation for univentricular hearts. Although the pathophysiology is not fully understood, high venous pressure is often present and may impair lymph flow, leading to intestinal oedema and PLE. Lymphatic vessels are divided into segments by unidirectional valves and the muscular vessel wall generates phasic contractions pumping lymph centrally. In our study we investigated the contractile properties of human lymphatic vessels to provide insight into the complex pathophysiology of PLE, with a view to future pharmacological treatment.
Methods: Thoracic ducts were harvested with informed consent from 26 patients undergoing oesophageal cancer surgery. Isolated vessel segments were mounted in a wire myograph for isometric force measurements. The diameter–tension relationship was established and the following pharmacological protocols tested: dose–response curves for norepinephrine, endothelin-1 and thromboxane analogue U46619, and phasic contractile activity in the presence of L-NAME and indomethacin.
Results The diameter–tension relationship revealed the vessels generating maximal contractions at a transmural pressure of 21 mmHg. The active curve was flat, indicating the vessels can contract at a wide range of diameters, with peak pressures of approximately 60 mmHg. Phasic contraction frequency was $1.39 \pm 0.35 min^{-1}$. Blocking the NO production with L-NAME, and subsequently prostaglandins with indomethacin, increased the frequency two- and five-fold, respectively. Norepinephrine, endothelin-1 and U46619 induced contractions in a dose-dependent manner with maximal contractions of 40–60 mmHg. High doses of all substances turned the phasic contractions into small oscillations, almost resembling a ventricular fibrillation.
Conclusion: We have shown that the thoracic duct from humans generates phasic contractions and that this activity can be modulated in several ways. Furthermore, the human thoracic duct has the capacity to overcome high venous pressures. Based on our results, we propose a novel approach to treating PLE by pharmacologically increasing lymphatic pumping.

502: AN *IN VIVO* CARDIAC ASSAY TO DETERMINE THE FUNCTIONAL CONSEQUENCES OF PUTATIVE LONG QT SYNDROME MUTATIONS

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Background: Genetic testing for long QT syndrome (LQTS) is now a standard and integral component of clinical cardiology. A major obstacle to the interpretation of genetic findings is the lack of robust functional assays to determine the pathogenicity of identified gene variants in a high-throughput manner. Since zebra fish have cardiac electrophysiology similar to that of humans, the goal of this study was to design and test an *in vivo* high-throughput cardiac assay to distinguish between disease-causing and normal *KCNH2* (*hERG1)* variants using the zebra fish as a model organism.
Methods: We tested the ability of previously characterised LQTS *hERG1* mutations and polymorphisms to restore normal repolarisation in the *kcnh2* knockdown embryonic zebra fish. Fertilised zebra fish eggs were injected with *kcnh2*-morpholino with or without *hERG1* mutant cRNAs. The cardiac phenotypes of embryos (48-hour post-fertilisation) were visually assessed under light microscopy to determine the degree of repolarisation. Results of the zebra fish assay were compared with the current benchmark *in vitro* assay.
Results: The cardiac assay correctly identified a non-disease-causing variant in nine of 10 cases (negative predictive value 90%) while correctly identifying a disease-causing variant in 40/40 cases (positive predictive value 100%).
Conclusions: The *in vivo* zebra fish cardiac assay is as precise as the current benchmark *in vitro* assay for the detection of disease-causing mutations and far superior in terms of throughput rate. Together with emerging algorithms for interpreting a positive LQTS genetic test, the zebra fish cardiac assay provides an additional tool for the final determination of pathogenicity of gene variants identified in LQTS genetic screening.

522: HYDROGEN SULPHIDE UPREGULATED HAEME OXYGENASE 1 EXPRESSION IN RATS WITH VOLUME OVERLOAD-INDUCED HEART FAILURE

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Background: Chronic heart failure (CHF) is a common complication of left-to-right shunt congenital heart disease. The mechanism responsible for CHF is not fully understood. Hydrogen sulphide (H_2S), a newly found gasotransmitter, has been reported to play an important pathophysiological role in the cardiovascular system. The present study was designed to determine the role of H_2S in CHF induced by left-to-right shunt, leading to volume overload.
Methods: Thirty male Sprague-Dawley rats were randomly divided into four groups: shunt group ($n = 8$), sham group ($n = 8$), shunt + sodium hydrosulphide (NaHS) group ($n = 8$), and sham + NaHS group ($n = 6$). Chronic heart failure was induced in the rats by abdominal aorta–inferior vena cava puncture. Rats in the shunt + NaHS and sham + NaHS groups were injected intra-peritoneally with NaHS (H_2S donor) at 56 $\mu mol kg^{-1} d^{-1}$, and at the same time, rats in the shunt and sham groups were injected with the same volume of physiological saline. Eight weeks after surgery, left ventricular HO-1 mRNA expression was measured by real-time PCR. Protein expression of HO-1 was evaluated by Western blots.
Results: Eight weeks after surgery, protein expression of HO-1 was significantly decreased in the shunt group compared with that in the sham group (0.54 ± 0.11 vs 1.04 ± 0.20, $p < 0.05$). Protein expression of HO-1 was significantly increased in the shunt + NaHS group compared with that in the shunt group (1.06 ± 0.10 vs 0.54 ± 0.11, $p < 0.05$). HO-1 mRNA expression was significantly increased in the shunt + NaHS group compared with that in the shunt group (5.86 ± 0.61 vs 1.86 ± 0.29, $p < 0.01$).
Conclusions: H_2S may play a protective role in volume overload-induced heart failure by up-regulating protein and mRNA expression of HO-1.

541: HYDROGEN SULPHIDE AMELIORATES VOLUME OVERLOAD-INDUCED VENTRICULAR REMODELLING BY MATRIX METALLOPROTEINASES (MMP-8, MMP-13) AND THEIR TISSUE INHIBITOR (TIMP-1) IN RATS

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Background: Chronic heart failure is a common complication of left-to-right shunt congenital heart disease, and ventricular remodelling is an important pathophysiological basis of volume overload-induced chronic heart failure. Hydrogen sulphide (H_2S) is a newly found gasotransmitter in the cardiovascular system. But the effect of H_2S on ventricular remodelling induced by volume overload is unknown.
Methods: This study used 30 male Sprague-Dawley rats (120–140 g), which were randomly divided into four groups: shunt group (n = 8), sham group (n = 8), shunt + sodium hydrosulphide (NaHS) group (n = 8), and sham + NaHS group (n = 6). An animal model of volume overload was induced by abdominal aorta–inferior vena cava puncture in the rats. Eight weeks after surgery, left ventricular matrix metalloproteinase-8 (MMP-8), MMP-13 and tissue inhibitor of metalloproteinase-1 (TIMP-1) expressions were measured by real-time PCR, Western blots and immunohistochemistry, respectively.
Results: Eight weeks after surgery, in the shunt group, MMP-8, MMP-13 and TIMP-1 mRNA expression and the ratio of MMP-13/TIMP-1 were significantly increased compared with those in the sham group (all $p < 0.05$). MMP-13 and TIMP-1 mRNA expression and the ratio of MMP-13/TIMP-1 were significantly decreased in the shunt + NaHS group compared with those in the shunt group (all $p < 0.05$). Protein expression of MMP-8, MMP-13, TIMP-1, and the ratios of MMP-8/TIMP-1 and MMP-13/TIMP-1 were significantly increased in the shunt group compared with those in the sham group (all $p < 0.05$). Protein expression of MMP-8, MMP-13, TIMP-1 and the ratio of MMP-8/TIMP-1 were significantly decreased in the shunt + NaHS group compared with those in the shunt group (all $p < 0.05$).
Conclusions: H_2S might play a protective role in volume overload-induced ventricular remodelling by regulating protein and mRNA expression of MMP-8, MMP-13 and TIMP-1.

588: INOTROPIC EFFECTS OF ILOPROST IN THE HYPERTROPHIC AND FAILING RIGHT HEART

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Background: To investigate whether iloprost has a direct inotropic effect in the pressure-overloaded hypertrophic and failing right ventricle (RV).
Methods: Rats were subjected to pulmonary trunk banding (PTB) (n = 8) or sham operations (n = 8). PTB enables evaluation of the direct effect of iloprost in the right heart, excluding the influence from pulmonary vasodilation. After the development of RV hypertrophy and failure, measurements were made at baseline and after intravenous administration of placebo, iloprost 10 ng/kg/min (Ilo10) and iloprost 100 ng/kg/min (Ilo100). Cardiac output, systemic blood pressure and RV function were evaluated by MRI, echocardiography and invasive pressure measurements.
Results: Animals subjected to PTB developed significant RV hypertrophy and failure. RV weight/tibia length ratio was elevated and tricuspid annular plane systolic excursion was markedly decreased compared to the sham animals. Iloprost caused a decrease in mean arterial blood pressure (MAP). In both groups of animals, infusion of Ilo100 induced an increase in stroke volume (placebo vs Ilo100 ± SEM: PTB 0.19 ± 0.008 vs 0.21 ± 0.01 ml, $p < 0.01$, sham 0.25 ± 0.01 vs 0.28 ± 0.01 ml, $p < 0.05$) as well as in dP/dt_{max} (placebo vs Ilo100 ± SEM: PTB 4730 ± 451 vs 5338 ± 605 mmHg/sec, $p < 0.05$, sham 2218 ± 218 vs 2521 ± 386 mmHg/sec, $p < 0.05$). Additionally, an elevation in cardiac output (placebo vs Ilo100 ± SEM: 63.0 ± 5 vs 71.4 ± 5 ml/min, $p < 0.001$) and RV systolic pressure (placebo vs Ilo100 ± SEM: PTB 110 ± 6 vs 121 ± 6 mmHg, $p < 0.01$) were observed in the PTB group. Infusion of nitroprusside, titrated to cause the same level of decrease in MAP as iloprost, did not increase any of the measured parameters.
Conclusion: Our results suggest that the prostacyclin analogue iloprost has inotropic properties, directly improving ventricular function in the hypertrophic and failing as well as in the healthy right heart.

613: VASCULAR HISTOPATHOLOGICAL REACTION TO GORE-TEX STRIPS USED FOR PULMONARY ARTERY BANDING IN AN *IN VIVO* PORCINE EXPERIMENTAL MODEL

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Objective: Pulmonary artery banding (PAB) is used as a surgical palliation to reduce excessive pulmonary blood flow caused by congenital heart defects. Due to the lack of histological studies dealing with tissue remodelling caused by contemporary PAB materials, our aim was to analyse tissue reaction to the presence of Gore-Tex strips fixed around the porcine pulmonary artery.
Methods: Gore-Tex strips were used for PAB in a growing porcine model. After five weeks, histological samples with PAB (n = 5) were compared to healthy pulmonary arterial segments distal to the PAB or from sham-operated animal (n = 1). Using stereology, we quantified the density of vasa vasorum and the area fraction of elastin, smooth muscle actin, macrophages and nervi vasorum within the pulmonary arterial wall.
Results: PAB samples had higher amounts of macrophages, lower amount of nervi vasorum, and a trend towards decreased smooth muscle content when compared with samples without the PAB strips. There was no destruction of elastic membranes, no medionecrosis, no pronounced inflammatory infiltration or foreign body reaction, and no vasa vasorum deficiency following the PAB. All the histopathological changes were limited to the banded vascular segment and did not affect distal parts of the pulmonary artery.
Conclusion: Our results suggest that Gore-Tex strips used contemporarily for PAB in a clinical setting do not cause as severe histological damage to the pulmonary arterial wall after five weeks in a growing porcine PAB model compared with previously published series using different PAB material.

615: REMOTE ISCHAEMIC PRECONDITIONING MODULATES METABOLISM AND IMPAIRS RECOVERY IN THE NEONATAL HEART SUBJECTED TO ISCHAEMIA–REPERFUSION INJURY

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Background: Remote ischaemic preconditioning (rIPC) reduces cardiac injury in older children and adults after cardiac surgery.

Dialysed plasma from mature rabbits undergoing rIPC protects isolated mature rabbit hearts in a Langendorff model, but the effect on the neonatal heart is unknown. The neonatal heart relies almost solely on carbohydrate metabolism, known to be modified by rIPC in the mature heart. We used microdialysis combined with targeted metabolomics to profile metabolism in the immature rabbit heart.
Hypothesis: Treatment of neonatal rabbit hearts prior to ischaemia–reperfusion injury with either *in vivo* rIPC or perfusion with dialysate from adult rIPC-treated rabbits alters myocardial function and metabolism and may be detrimental.
Methods: Fifteen newborn (1–4 days old, 49–72 g) rabbits were randomised into three groups ($n = 5$ in each group): control, *in vivo* rIPC and adult rIPC dialysate group. Plasma from the rIPC-treated adult rabbits was dialysed, added to the buffer and administered to the dialysate group. Hearts were mounted in a Langendorff model and perfused for 55 min stabilisation, 40 min global ischaemia and 120 min reperfusion. A microdialysis probe was inserted intramurally in the LV and samples (10 µl) were analysed using UPLC-MS/MS, targeting lactate and purine metabolites.
Results: During the final 10 min of ischaemia, interstitial lactate concentration almost doubled in the rIPC (7.8 ± 1.0 mM/l) and dialysate group (9.3 ± 0.6 mM/l) compared with the controls (4.6 ± 0.1 mM/l) ($p < 0.05$). Simultaneously, a marked increase in inosine concentrations was measured for the intervention group: rIPC (49.2 ± 4.0 µM/l) and dialysate group (52.5 ± 9.6 µM/l) compared to the controls (17.2 ± 6.0 µM/l) ($p < 0.05$). Functional recovery (dP/dt min) during 30 min reperfusion was impaired in the rIPC (–329.4 ± 34 mmHg) and the rIPC dialysate (–270.3 ± 65 mmHg) groups compared to the controls (–683.8 ± 56 mmHg) ($p < 0.05$).
Conclusions: *In vivo* rIPC and rIPC dialysate increased cytosolic energy expenditure during ischaemia and attenuated functional recovery in the neonatal isolated rabbit heart.

731: PLATELET FUNCTION AND MORPHOLOGY IN IDIOPATHIC PULMONARY HYPERTENSION

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Background: Thrombosis and proliferation are two important pathological features of idiopathic pulmonary hypertension (IPAH). Theoretically, both these processes may be initiated by platelets. A barrage of activated platelets in the pulmonary circulation, similar to what happens in the nail bed to cause clubbing, may lead to all the known pathological changes seen in iPAH.
Methods: Nine consecutive patients of iPAH [median age 24 years (14–47 years)] were included in the study. Nine patients with rheumatic heart disease with pulmonary hypertension [median age 42 years (22–66 years)] were recruited as controls. Blood samples were taken from the superior vena cava, pulmonary artery, left ventricle and femoral artery. Collagen and adenosine-di-phosphate were utilised for the assessment of platelet reactivity, and platelet morphology was analysed under electron microscopy.
Results: There was no difference in the proportion of active platelets between the iPAH group and the control group at various sites. In the iPAH group, the number of active platelets was significantly higher in pulmonary artery compared to that of the femoral artery ($p = 0.01$). By contrast, there was no difference in the number of active platelets between various sites in patients with RHD with iPAH. Platelet reactivity at the various sites did not differ significantly between iPAH and RHD in the PAH groups. There was no significant difference in platelet reactivity measured biochemically by collagen and ADP between the pulmonary artery and the rest of the sites.
Conclusions: We found an increased number of active platelets in the pulmonary circulation compared to the systemic circulation on electron microscopy in iPAH patients. We found no difference biochemically in the level of platelet reactivity between the pulmonary and systemic circulation. Patients with iPAH did not show significantly higher platelet reactivity compared to patients with RHD with moderate to severe PAH.

770: REVERSIBLE PULMONARY TRUNK BANDING. X: WALL STRESS-ASSOCIATED ACTIVATION OF MYOCARDIUM GLUCOSE-6-PHOSPHATE DEHYDROGENASE IS NORMALISED BY INTERMITTENT SYSTOLIC OVERLOAD IN YOUNG GOATS

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Background: Increased myocardial glucose-6-Phosphate dehydrogenase (G6PD) activity has been demonstrated in young goats submitted to traditional pulmonary artery banding (PAB). This biochemical alteration drives superoxide anion generation and elevates oxidative stress by elevated G6PD-derived NADPH in the failing heart. This study sought to assess the myocardial mechanics and kinetics of G6PD activity during intermittent systolic overload of the sub-pulmonary ventricle in a young animal model.
Methods: Thirty young goats with comparable weights were separated into five groups according to the study period duration (0, 24, 48, 72, and 96 hours). A 12-hour systolic overload of the right ventricle (RV) was alternated with a 12-hour resting period with an adjustable PAB. Systolic overload was adjusted to achieve a 0.7 RV/aortic pressure ratio. Echocardiographic and haemodynamic evaluations were performed every day postoperatively. After completing the training programme of each group, the animals were humanely killed for morphological and G6PD activity assessment.
Results: A 130.8% increase occurred in the RV mass of the 96-hour group, compared with the 0-hour group ($p < 0.0001$). Increased RV volume/mass ratio and wall stress observed in the 24-, 48-, and 72-hour groups were associated with increased RV G6PD tissue activity (Pearson correlation, 0.77 and 0.87; $p = 0.05$ and 0.03, respectively). A full recovery of these parameters was observed in the 96-hour group, compared to baseline values. No significant changes were observed in the G6PD activity of the ventricular septum and left ventricle.
Conclusions: G6PD tissue activity is associated with changes in ventricular volume and RV wall stress. This study suggests that intermittent systolic overload for sub-pulmonary ventricle retraining in young goats may improve the altered cardiac energy substrate metabolism and decrease the formation of reactive oxygen species, thus preventing cardiac deterioration post-PAB.

861: RIGHT VENTRICULAR MYOCARDIAL PERFORMANCE INDEX IS PARADOXICALLY DECREASED WITH SEVERE PRESSURE OVERLOAD CARDIAC HYPERTROPHY IN YOUNG RATS

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Background: Although the myocardial performance index (MPI) is usually increased in the presence of RV dysfunction, debate continues over the correlation between right ventricular (RV) MPI and functional derangement in patients with congenital heart disease (CHD). To address this controversy, we took serial measurements of the RV MPI during the development of RV dysfunction induced by

pressure overload.
Methods: RV pressure overload was induced by partial pulmonary arterial banding (PAB) in three-week-old rats. The rats were divided into two groups: mild pulmonary stenosis (PS) group (20–40% stenosis, $n = 20$) and severe PS group (40–70% stenosis, $n = 28$). Sham-operated animals (sham group, $n = 30$) underwent the same surgical procedure without PAB. Pressure-overloaded RV hypertrophy, which was documented by weighing the heart, evaluation of echocardiograms, and cardiac hypertrophy-associated gene expression. The RV MPI was checked one, two, three, five, and eight weeks after PAB.
Results: The RV MPI of the mild PS group did not differ significantly from that of the sham group. The RV MPI of the severe PS group, however, was paradoxically lower than that of the sham group ($p < 0.05$).
Conclusions: The RV MPI was paradoxically deceased in severe RV pressure overload hypertrophy induced by PAB.

898: GLYCOGEN KINASE-3 INHIBITION: GOOD OR BAD FOR THE HEART?

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Background: Glycogen synthase kinase-3 (GSK-3) is a serine-threonine kinase that was discovered as a regulator of glycogen synthase and known as a role player in cardioprotection. Myocardial GSK-3 (1) may regulate expression of SERCA-2a, affecting contractility; (2) may phosphorylate and inhibit IRS-1, disrupting insulin signalling, (3) may regulate growth via interaction with Wnt and hypertrophic signalling pathways. GSK-3 inhibitors are being developed for clinical use.
Aim: To determine whether myocardial GSK-3 and its substrate proteins are dysregulated in obesity and pre-diabetes, and to study the effects of GSK-3 inhibition on the hearts of obese, pre-diabetic rats.
Methods: Pre-diabetic Wistar rats [induced by a diet causing hyperphagia (DIO) for 16 weeks] were compared to age-matched controls. Half of each group was treated with the GSK-3 inhibitor (CHIR118637 – 30 mg/kg/day) for four weeks (weeks 12 to 16 of the diet period). After 16 weeks, echocardiography was performed and glucose tolerance was established, biometric and biochemical parameters were determined, myocardial performance was verified by *ex vivo* perfusion, and protein expression was ascertained in snap-frozen hearts by Western blotting and specific antibodies. Ca^2+ATPase activity was determined spectrophotometrically and cardiomyocytes were used to determine cell size and localisation of NFATc3 and GATA4.
Results: Treated and untreated DIO gained more body weight and intra-peritoneal fat. GSK-3 inhibition improved glucose tolerance and echo parameters in DIO. CHIR had no effect on GSK-3 expression but increased phosphorylation in CHIR. CHIR was associated with increased NFATc3 and GATA4 nuclear translocation. CHIR elevated IRS-2 expression but had no effect on IRS-1 and SERCA-2a.CHIR increased PKB/Akt and phospholamban phosphorylation in DIO rats.
Conclusion: GSK-3 protein may play a role in glucose homeostasis and regulation of IRS-2 expression but its inhibition did not enhance IRS-1 or SERCA-2a expression. CHIR reversed cardiac hypertrophy in DIO rats but it caused hypertrophy in the controls.

907: GENETIC POLYMORPHISMS ASSOCIATED WITH ALLOGENEIC RED BLOOD CELL TRANSFUSION REQUIREMENTS IN PAEDIATRIC PATIENTS UNDERGOING CARDIAC SURGERY

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Background: Patients undergoing repair of congenital heart disease require allogeneic blood transfusions, mainly to allow cardiopulmonary bypass and to offset blood loss from bleeding. Clinical factors associated with increased bleeding risk, including coagulation system activity, platelet reactivity, use of deep hypothermic circulatory arrest, and surgery duration and complexity. We sought to determine whether genetic polymorphisms known to be associated with the coagulation/fibrinolytic system or with platelet function are associated with red blood cell transfusion requirements associated with paediatric cardiac surgery.
Methods: A total of 625 cardiac surgeries in 383 patients were reviewed. Ninety-six SNPs on 53 genes involved in the coagulation/fibrinolysis pathways were assayed using the the Illumina GoldenGate® custom SNP panel; genotyping was successful for > 99% of SNPs. Associations between SNPs and red blood cell transfusions within 48 hours of surgery (adjusted for age at surgery, surgical complexity and pre-operative oxygen saturation) were assessed in regression models adjusted for repeated measures. Bootstrap resampling (1 000 samples) was used to offset multiple comparison bias and exclude SNPs with very low minor allele frequencies.
Results: Median red blood cell requirement was 114 ml/kg (interquartile range: 73–174 ml/kg). Coagulation factor polymorphisms associated with increased red blood cell requirements included factor VIII rs100873005 CC/CG SNPs [+ 40 (14) ml/kg, $p = 0.004$] and factor XI rs2036914 CC SNPs [+ 51 (16) ml/kg, $p = 0.001$]. Additional SNPs associated with a higher volume of red blood cell requirements included alpha-2-macroglobulin precursor rs669 GG SNPs [+ 91 (23) ml/kg, $p < 0.001$], guanine nucleotide-binding protein β3 rs5443 CC SNPs [+35 (18) ml/kg, $p = 0.05$] and chemokine receptor 2 rs1799864 AA/AG SNPs [+42 (16) ml/kg, $p = 0.008$]. Both alpha-2-macroglobulin precursor rs669 and factor XI rs2036914 were also found to be associated with low pre-operative antithrombin activity, a key marker of heparin resistance and increased bleeding volumes.
Conclusions: Patients with congenital heart disease have substantial transfusion requirements during and immediately after paediatric cardiac surgery. While clinical factors are critical in determining the required amount of transfusions, genetic polymorphisms also have a key role in this process.

908: ASSESSING THE INTERLEUKIN-6 -174 G/C SINGLE NUCLEOTIDE POLYMORPHISM AND CORONARY ARTERY DISEASE

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Background: Interleukin 6 (IL6) is a pro-inflammatory cytokine involved in the pathogenesis of chronic inflammatory diseases such as coronary artery disease (CAD). The -174 IL6 G/C promoter polymorphism influences mRNA and protein levels and is implicated in CAD. This polymorphism has been investigated but limited data are available on South African Indian (SAI) and black (SAB) population groups, despite high disease prevalence. This study aimed to assess the -174 IL6 G/C polymorphism in SAI subjects with CAD.
Methods: Polymorphic variants were assessed by polymerase chain reaction–restriction fragment length polymorphism, and IL6 levels were measured by ELISA.
Results: The -174 IL6 C allele was found at a higher frequency in the total SAI subjects (23%) compared to SABs (2%), irrespective of disease status ($p < 0.0001$, OR = 0.0503, 95% CI: 0.0183–0.1388) and in healthy SAI (29%) and SAB (2%) controls ($p < 0.0001$, OR = 0.0507, 95% CI: 0.0152–0.1699). A significant association between

the -174 IL6 G allele and CAD in SAI was found (84 vs 71% – SAI controls; $p = 0.0431$, OR = 0.468, 95% CI: 0.23–0.953). Circulating levels of IL6 were elevated in total (6.58 ± 0.56 pg/ml) and healthy (6.62 ± 0.63 pg/ml) SABs compared to total (1.80 ± 0.22pg/ml) and healthy (2.51 ± 0.57pg/ml) SAI groups, as well as CAD patients (1.46 ± 0.36pg/ml, $p < 0.0001$). Levels of IL6 were elevated in all groups with homozygous -174 IL6 C alleles but only significant in the healthy SAI group (GG: 3.73 ± 0.94 pg/ml vs GC/CC: 0.89 ± 0.5 pg/ml, $p = 0.0001$).
Conclusion: The presence of the IL-6 -174 G allele influences levels of IL-6 and increases the risk of CAD in South African Indians.

922: AUTOPHAGY UPREGULATION IN CARDIOTOXICITY: PHARMACOLOGICAL VS GENETIC MANIPULATION

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Background: Cardiotoxicity is a well-known side effect of anthracyclines such as doxorubicin (DXR), resulting in substantial morbidity. The most widely accepted hypothesis for their mechanism of action is oxidative stress, which leads to the induction of cell death as a direct consequence of DNA damage and/or interference with DNA repair. Autophagy, a major catabolic process, has been shown to play a vital role in cardiac homeostasis. This process is often elevated following various forms of cardiovascular stress. However, whether autophagy participates as a pro-survival or pro-death pathway remains to be determined. This study therefore aimed to determine whether pharmacological or genetic manipulation of autophagy alleviates DXR-induced toxicity.
Methods: H9C2 rat cardiac myoblasts were treated with rapamycin (50 nM - CR) or siRNA (mTOR - CM) for 24 hours to up-regulate autophagy. This was followed by treatment with DXR alone (3 µM - CD) or in combination with rapamcin (RD) or siRNA (MD) for a further 24 hours, where after cell viability, apoptosis, mitochondrial morphology and DXR localisation was assessed.
Results: Assessment of cell viability indicated that groups CM and CD significantly reduced viability [75.48 ± 1.81% ($p < 0.001$) and 65.58 ± 2.25% ($p < 0.01$)] versus the control. Group RD significantly improved viability [78.93 ± 10.85% ($p < 0.05$)] versus CD. Caspase activity was also significantly elevated in group CD [444.60 ± 29.33% ($p < 0.001$)] versus the control, whereas group RD significantly reduced (78.86 ± 7.14%) caspase activity. Normal mitochondrial morphology was not adversely affected in groups CM, CR and RD. However groups CD and MD displayed abnormal mitochondria that were shorter, fragmented and discontinuous.
Conclusions: These results indicate a prospective role for rapamycin against DXR-induced cardiotoxicity and highlight rapamycin as a plausible adjuvant therapy to counteract and improve the life-threatening impediment of DXR's actions in clinical practice.

928: GRAPE SEED PROANTHOCYANIDIN EXTRACT LIMITS CARDIAC DAMAGE

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Background: Heart failure is a leading cause of mortality worldwide. To date, many pharmaceutical agents have been used to treat cardiac hypertrophy and early stages of heart failure. These therapies have proven reasonably effective; however, there is a need for alternative therapeutic strategies, more specifically, natural therapies that are cost effective and safe, to prevent or reverse hypertrophy before it develops into heart failure. Proanthocyanidolic oligomer supplementation (PCO), a grape seed extract, was shown to quicken muscle recovery and reduce inflammation in a skeletal muscle model of injury. Considering the positive effects on muscle recovery, as well as results suggesting that resveratrol, another member of the polyphenol family, could limit the occurrence of cardiac hypertrophy, the possibility exists that PCO might also be beneficial.
Methods: An osmotic mini-pump containing isoproterenol (2 mg/kg/day), a dual β_1–β_2-adrenergic receptor agonist, was used to induce hypertrophy. Male Wistar rats (280–320 g) were orally gavaged with either PCO (20 mg/kg/day) or distilled water for two weeks prior to the subscapular implantation of the pump containing isoproterenol or ascorbate (vehicle). After seven days, the rats were killed and the heart isolated. Paraffin wax-embedded hearts were sectioned (2 µm) and stained with Picro Sirius for fibrosis. A macrophage marker, ED1, was used to determine the infiltration of immune cells.
Results: Results indicated that the isoproterenol groups lost significant amounts of weight one day ($p < 0.05$) after implantation. PCO treatment reduced hypertrophy, as measured by a lower heart weight to tibia length and heart weight to body weight ratio. Histological analysis showed more damage, inflammation and fibrosis in the isoproterenol group receiving placebo treatment (I-PLA) compared to the group receiving PCO supplementation (I-PCO). Both groups displayed significantly more fibrosis than their respective controls.
Conclusion: The cost-effective, over-the-counter PCO supplement resulted in better recovery after isoproterenol infusion.

932: GLUCOSE AND INSULIN IMPROVE FUNCTIONAL RECOVERY AFTER *DE NOVO* ACUTE HEART FAILURE BY STIMULATING THE SINUS NODE

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Background: Glucose–insulin–potassium infusions improved recovery from sinus node dysfunction in patients. GIK infusions given in the ambulance to patients with acute coronary syndrome reduced hard end-points in the IMMEDIATE study, but effects on acute heart failure (AHF) are still unknown.
Methods: We therefore tested GI therapy on isolated rat hearts perfused retrogradely by a modified Krebs-Henseleit solution in the Langendorff system subject to *de novo* AHF. In this model the initial stabilisation phase hearts were perfused at 100 cm H_2O with glucose (11.1 mM) as sole substrate Thereafter AHF was induced by under-perfusion at 20 cm H_2O. We added adrenaline 10^{-8} M to induce a hyperadrenergic stimulus while reducing perfusate glucose to 2.5 mM and adding high free fatty acids (FFA) to the buffer (1.3 mM, 0.1 mM BSA). In the recovery phase the hearts recovered incompletely although the perfusion pressure was restored to 100 cm H_2O with the continued presence of high FFA, and with increased glucose (11.1 mM). Only half of the hearts in this phase received insulin (0.3 mU).
Results: Glucose coupled with insulin in the recovery phase increased the heart rate (168.5 ± 34.5 vs 36.7 ± 25.0 beats/min, $p < 0.01$). LV developed pressure was unchanged in both groups (31.9 ± 7.2 vs 38.9 ± 16.9 mmHg). We attribute this cardioprotective increase of heart rate to the electrophysiological effects of glucose and insulin on sinus node function.
Conclusions: Our data suggest that glucose and insulin improve the heart rate of the acutely failing isolated heart by improving sinus node recovery after *de novo* AHF.

960: ANTI-SENESCENT EFFECT OF STATIN THERAPY IN CORONARY ARTERY DISEASE

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Introduction: Atherosclerosis is a premature-ageing syndrome associated with senescence of vascular endothelial and smooth muscle cells. Premature cellular senescence is induced by extrinsic risk

factors that damage the structure of DNA. Experimental evidence and emerging clinical data suggest that the HMG CoA reductase inhibitors may exert their beneficial effects in patients with coronary artery disease (CAD) by reducing cellular senescence and preventing apoptosis.
Aim: We investigated the effect of prior statin therapy on leucocyte telomere length and the interaction with markers of inflammation, oxidative stress and cellular apoptosis.
Methods: Consecutive patients below the age of 45 years admitted to a tertiary cardiac unit were recruited following confirmation of obstructive CAD on angiography. A population-based, randomly selected, matched, healthy control group who were statin naïve were also studied. Apoptotic activity in peripheral blood mononuclear cells was determined using the Caspase-Glo® 3/7, 8 and 9 assays. Oxidative stress was assessed with athiobarbituric acid assay, which measures the lipid peroxidation end-product malondialdehyde. Leukocyte telomere length was measured with a quantitative PCR-based technique and calculated as the ratio of telomere repeats to single-copy gene copies (T/S ratio).
Results: The mean duration of statin therapy in patients was 31 months. Lipid peroxidation was significantly elevated in patients compared to controls [median/interquartile range 0.0060 (0.0030–0.0140) cases, 0.0035 (0.0025–0055) controls, $p < 0.009$]. Caspase 8, an initiator of apoptosis activated by the extrinsic pathway, was significantly reduced in the cases. Telomere length was significantly longer in the cases [cases 0.71 (0.69–0.73), controls 0.67 (0.63–0.70), $p < 0.001$]. There was no significant difference in LDL cholesterol and hsCRP levels between the groups.
Conclusion: Unrelated to the lipid-lowering, anti-inflammatory and anti-oxidant effects, chronic statin therapy was associated with longer telomere length, a marker of vascular ageing. This anti-senescent effect of statin therapy may emerge as a new strategy in the treatment of atherosclerosis.

980: REGULATION OF CARDIAC HORMONES DURING ASPHYXIA IN NEONATES: STUDIES IN A PIGLET MODEL

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Background: The natriuretic peptides (ANP and BNP) are expressed in the myocardium and related to cardiac dysfunction. In adults they are up-regulated in response to cardiac ischaemia and increased wall stress, as seen in acute coronary syndromes and cardiac failure. Also in neonates, the peptides are pursued as cardiac biomarkers. However, the role of a global hypoxic–ischaemic insult in the newborn period on cardiac natriuretic peptide expression has not been investigated. We investigated this in a 72-hour piglet model of hypoxic ischaemic encephalopathy.
Methods: Eighteen-hour-old piglets were randomly divided into a control and intervention group. The piglets in the intervention group were anesthetised and exposed to global hypoxia (45 minutes) verified by EEG-depression and arterial pH < 7.0, whereas control piglets were only anesthetised. The piglets were awakened and intensively cared for during a total of 72 hours. Blood samples were drawn after intubation but prior to any hypoxia and after 72 hours, and natriuretic peptide concentration was analysed. The piglets were euthanised and regional myocardial biopsies were obtained. These biopsies were analysed for cardiac expression of natriuretic peptides and natriuretic peptide receptors by real-time PCR and Western blot.
Results: The biochemical analyses are being performed and results are pending.
Conclusion: In order to validate whether natriuretic peptides can be used as specific biomarkers of cardiac compromise in neonates, it will be of value to conclude whether the natriuretic peptide expression occurs as a result of global hypoxia *per se* or not. Furthermore, it is of great interest to convey whether the natriuretic peptide expression is regional and corresponds to plasma values in the same manner as seen in adults to verify whether parallels can be drawn from the vast knowledge of natriuretic peptides in adult cardiac disease.

985: DOES CARDIAC MUSCLE RESTORATION OF THE LEFT VENTRICLE DIFFER FROM THE RIGHT VENTRICLE IN THE NORMAL CANINE HEART? A PASSIVE STRESS–STRAIN RELATIONSHIP *IN VITRO* STUDY

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Background: Early deterioration of diastolic ventricular function usually precedes systolic ventricular abnormalities. Normal cardiac muscle restoration is important for adequate relaxation and may depend on muscle fibre orientation. The passive stress–strain relationship as an indicator for cardiac muscle restoration has been established.
Objective: The goal of the study was to compare and characterise the *in vitro* passive stress–strain relationship of the left and right ventricular muscle in the normal canine heart.
Methods: Cardiac muscle tissue from four normal canine hearts was studied. Samples were taken from the ventricular free wall, ventricular septum and papillary muscles of both left and right ventricles. Each sample was divided into three blocks and was studied in three different orientations: longitudinal, radial and circumferential. Each sample underwent compression under four different strain rate conditions, ranging from 10 to 40/sec. Relaxation of the cardiac muscles was recorded by multiple sensors for each sample, and stress–strain loops were calculated from force and displacement data.
Results: Stress–strain relationships of the left and right ventricles were significantly different. Stress of the left ventricular free wall and septum, when measured in the radial and circumferential direction, was significantly higher than for the same amount of strain applied on right ventricular muscle. LV stress = 40.5–60.2 Pascals (mean = 48.2); RV stress = 20.1–38.9 Pascals (mean = 31.2) ($p < 0.001$). When the longitudinal direction was compared, right ventricular stress was significantly higher than left ventricular stress under the same strain conditions, RV stress = 40.7–80.6 Pascals (mean = 49.1); LV stress = 20.4–40.3 (mean = 31.2) ($p < 0.001$). Papillary muscle stress–strain relationship was similar for both ventricles in the longitudinal direction.
Conclusions: Cardiac muscle restoration is different between the left and right ventricles in the normal canine heart. LV muscle is more efficient in the longitudinal direction, while RV muscle is superior in the radial and circumferential orientations. Measurements of cardiac restoration may provide new insight on diastolic function.

1193: CHANGES OF CASPASE 3, BCL2, VASCULAR ENDOTHELIAL GROWTH FACTOR GENE EXPRESSIONS AFTER HUMAN UMBILICAL CORD BLOOD-DERIVED MESENCHYMAL STEM CELL TRANSFUSION IN PULMONARY HYPERTENSION RAT MODELS

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Objective: Pulmonary arterial hypertension (PAH) is difficult to treat and is characterised by increased pulmonary arterial pressure, right heart failure and death. PAH has been shown to be refractory to most of the conventional pharmacological therapies. Human umbilical cord blood-derived mesenchymal stem cells (hUCB-MSCs) are

regarded as an alternative source of bone marrow-derived mesenchymal stem cells. hUCB-MSCs have recently been studied for evaluation of their potential as a source of cell therapy. The purposes of this study were to investigate changes of pulmonary pathology, haemodynamics and gene expression of caspase 3, Bcl2, vascular endothelial growth factor (VEGF), interleukine (IL)-6, and tumour necrosis factor (TNF)-α in monocrotaline (MCT)-induced PAH rat models after hUCB-MSCs transfusion.
Methods The rats were grouped as follows: control group (C group), subcutaneous injection of saline; M group, subcutaneous injection of MCT (60 mg/kg); and hUCB-MSCs transfusion (U group). hUCB-MSCs (3×10^6/ml/cm^2) were transfused by intraperitoneal injection one week after MCT injection.
Results: The mean right ventricular pressure (RVP) significantly decreased in the U group compared with the M group in weeks two and four. RV weight and the ratio of RH/LH + septum significantly decreased in the U group compared to the M group. The number of muscular pulmonary arterioles significantly decreased in the U group compared with the M group in weeks two and four. Medial wall thickness of the pulmonary arteriole significantly decreased in the U group compared to the M group in week two. Gene expressions of caspase-6, Bcl-2, VEGF, IL-6 and TNF-α significantly decreased in week four in the U group compared with the M group.
Conclusion: After hUCB-MSCs transfusion there was improvement of RVH, mean RV pressure and survival rate. Decreases in several gene expressions were observed. Additional research on the dose and frequency of hUCB-MSCs infusion is needed to determine the optimal parameters for PAH treatment**.**

1194: CHANGES OF CASPASE 3, BCL2, AND VASCULAR ENDOTHELIAL GROWTH FACTOR AFTER BONE MARROW CELL TRANSFUSION IN RATS WITH MONOCROTALINE-INDUCED PULMONARY HYPERTENSION

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Background: Pulmonary arterial hypertension (PAH) is difficult to treat and is characterised by increased pulmonary arterial pressure, right heart dysfunction, lung vascular remodelling and death. Bone marrow-derived mesenchymal stem cell therapy has provided an alternative treatment for ailments of various organs by promoting regeneration at the site of pathology. The purposes of this study were to investigate changes of pulmonary pathology, haemodynamics and gene expressions of Caspase 3, Bcl2, and vascular endothelial growth factor (VEGF) in monocrotaline (MCT)-induced PAH rat models after bone marrow cell (BMC) transfusion. The rats were grouped as follows: control group; M group, subcutaneous injection of monocrotaline (MCT); BMC transfusion (B group). BMC were transfused by intravenous injection in the tail one week after MCT injection.
Results: The mean right ventricular pressure (RVP) significantly decreased in the B group compared with the M group in weeks two and four. RV weight significantly decreased in the B group compared to the M group in weeks two and four. The ratio of medial thickening of the pulmonary artery was significantly decreased in the B group compared with the M group in week two. The number of muscular pulmonary arterioles significantly decreased in the B group compared with the M group in week four. The number of muscular pulmonary arterioles increased in the M group compared with the C group in weeks two and four. Gene expressions of Caspase 3, Bcl2, and VEGF significantly increased in the M group compared with the C group and significantly decreased from week two in the B group compared with the M group.
Conclusion: After BMC transfusion, there was improvement of RVH and mean RV pressure. Decreases in several genes were observed. Additional research on the dose and frequency of BMC infusion is needed to better determine the optimal parameters for PAH treatment.

1214: EXOME SEQUENCING IN SYNDROMIC PATIENTS WITH CONGENITAL HEART DISEASE: PERFORMING A TRIO ANALYSIS

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Background: Congenital heart defects (CHD) are a major cause of infant morbidity and mortality. Reaching an aetiological diagnosis in patients with a syndromic heart defect is important, not only to gain insight into their pathogenesis and for genetic counselling on recurrent risks, but especially with regard to providing information on the future prognosis, based on knowledge of the natural course of the disorder. In syndromic cases, an exact aetiological diagnosis can be reached in an estimated 50 to 60% of patients, following careful clinical evaluation, complemented by various genetic tests, including array-CGH. With the advent of exome sequencing, it is now feasible to perform a trio analysis, i.e. sequencing of the coding parts of the genes in both parents and the child, where only the child is affected, in order to identify a candidate gene. For syndromic cases, we hypothesised that these patients have a thus far unrecognised monogenic condition responsible for both the intelligence deficit and the heart defect. Since the majority of syndromes featuring CHD are dominant, it is likely that at least in a subset of these, a *de novo* dominant mutation is present.
Methods: In-solution capture (Nimblegen target-enrichment system) and sequencing will be done on the Illumina HiSeq2000 platform (Genomics Core KULeuven/UZLeuven). Possible causal mutations will be confirmed by traditional Sanger sequencing.
Results and Conclusion: Preliminary results on exome sequencing in trios on the child presenting with congenital heart disease, dysmorphic features and mental retardation will be discussed.

1218: COMPLEMENTARY ROLES OF THE NOVEL BIOMARKER ST2 AND NT-proBNP IN AFRICAN HYPERTENSIVE SUBJECTS

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Background: Although NT- pro brain natriuretic peptide (NT-proBNP) levels have been shown to differentiate hypertensive left ventricular hypertrophy (LVH) without heart failure (HF) from hypertensive HF due to systolic and/or diastolic dysfunction, it has not been very helpful in differentiating hypertensive subjects with LVH from those without. We therefore decided to see the complementary role of soluble ST2, a novel biomarker of biomechanical strain.
Methods: This was a prospective cohort study. Echocardiography was performed on all subjects. LVH was considered present when left ventricular mass, indexed for height$^{2.7}$, was greater than 46.2 g/m$^{2.7}$ in women and 49.2 g/m$^{2.7}$ in men. Plasma ST2 and NT-proBNP was measured using electrochemiluminescence-type immunoassay.
Results: Of 210 subjects studied, 42.9% were female, and the mean age of the study cohort was 50.3 ± 11.3 years. Hypertensive subjects with LVH had higher concentrations of ST2 compared to those without LVH (23.0 ± 8.3 vs 14.5 ± 4.9 ng/ml, $p = 0.001$) and those with

hypertensive HF had higher levels compared with those with hypertensive LVH (134.7 ± 57.3 vs 23.0 ± 8.3 ng/ml, $p = 0.000$). There was however no significant difference between NT-proBNP levels when hypertensive subjects with LVH were compared with those without LVH ($p = 0.68$) but those with heart failure had significantly higher NT-proBNP levels compared with hypertensives with LVH ($p < 0.000$). ST2 has a stronger correlation with clinical and echocardiograhic parameters compared to NT-proBNP. Serum ST2 also correlated well with NT-proBNP ($r = 0.41$, $p < 0.000$). In the assessment of the hypertensive heart disease spectrum, ST2 correlates well with NT-proBNP and has proven to be a better marker.
Conclusions: Plasma ST2 levels appear to be a very useful marker in differentiating the different spectra of hypertension–hypertensive heart disease and may hold a future role in this regard.

1226: GENE EXPRESSION PROFILES IN ENGINEERED CARDIAC TISSUES RESPOND TO MECHANICAL LOADING AND INHIBITION OF TYROSINE KINASES

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Background: Several formulations for engineered cardiac tissue (ECT) have emerged that incorporate stem cells or immature cardiomyocytes into three-dimensional (3D) constructs. These ECTs mature *in vitro*, acquire the features of mature cardiac muscle, appear to involve the p38MAP kinase (p38MapK) pathway, and respond to mechanical load with increased proliferation and maturation. We hypothesised that global ECT gene expression patterns are sensitive to mechanical loading conditions and tyrosine kinase inhibitors.
Methods: We generated 3D ECTs from immature rat embryo heart cells, as previously published, and then treated constructs after five days in culture for 48 hours with mechanical stretch (5%, 0.5 Hz) and/or the following selective inhibitors (birb796 for p38MapK, CI10404 for ERK1/2, or SP60025 for JNK). RNA was isolated from three sets of experiments and assayed using a standard Agilent rat 4x44k V3 micro-assay. The Ingenuity Systems Pathway analyser was used to analyse data from individual experiments, pooled within groups and between groups.
Results: Changes in gene expression in response to mechanical stretch and/or inhibitors were recorded. As anticipated, top pathways altered in response to these stimuli included cellular development, cellular growth and proliferation; tissue development; cell death, cell signalling and small-molecule biochemistry, as well as numerous other pathways.
Conclusion: ECTs display a broad spectrum of altered gene expression in response to mechanical load and/or tyrosine kinase inhibition, reflecting the complex regulation of proliferation, differentiation and architectural alignment during ECT maturation.

1381: CARDIOPROTECTVIE EFFECT OF ACE2 ACTIVATOR ON LEFT VENTRICULAR DYSFUNCTION SECONDARY TO PRESSURE OVERLOAD IN THE RAT

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Objectives: The RAS (renin–angiotensin system) is activated after myocardial infarction, and RAS blockade with angiotensin converting enzyme inhibitors or angiotensin receptor blockers slows but does not completely prevent progression to heart failure. In contrast, angiotensin converting enzyme 2 (ACE2)/angiotensin-(1-7) [Ang-(1-7)]/Mas is recognised as a counter-regulatory axis. Little is known about the role of ACE2 in cardiac dysfunction secondary to pressure-overload.
Hypothesis: In pressure overload-induced cardiac dysfunction, we hypothesised that cardiac expressions of ACE 2 and Ang-(1-7) are down-regulated, and an ACE2 activator can attenuate the development of left ventricular dysfunction through ACE2/Ang-(1-7)/receptor mas axis.
Methods and Results: In the Wistar rats subjected to ascending aortic banding (AOB), starting 29 days after banding, banded rats were treated with DIZE (ACE2 activator) at a dose of 15 mg/kg/day intra-peritoneally or vehicle for 14 days. Subsequently, there was down-regulated cardiac expression of Ang-(1-7) in AOB for 42 days compared to sham-operated rats. DIZE could significantly decrease the mean pulmonary arterial pressure and mean left atrial pressure, and attenuate left ventricular remodelling, respectively, when compared with the vehicle controls. In addition, DIZE caused up-regulated expression of ACE2, receptor mas and endothelial nitric oxide synthase in 42-day banded rats.
Conclusions: These results indicate that activation of ACE2 may provide preventive potential for attenuating the development of left ventricular dysfunction secondary to pressure overload. Further translational study, including oxidative stress in humans, is needed to substantiate the findings.

1422: ARG72 VARIANT OF p53 CODON 72 FUNCTIONAL POLYMORPHISM AND RISK OF CORONARY ARTERY DISEASE IN A SOUTH AFRICAN POPULATION

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Introduction: Atherogenic stimuli induce DNA damage through increased oxidative stress. DNA damage leads to increased expression of p53, a pro-apoptotic gene whose principal function is to protect cells from malignant transformation. p53 is thought to contribute to vascular disease by increasing apoptosis of macrophages and smooth muscle cells in advanced atherosclerotic lesions, rendering them vulnerable to rupture. More recently, p53 has been shown to regulate genes involved in lipid and carbohydrate metabolism. Polymorphisms of the p53 gene have been associated with increased susceptibility to coronary artery disease (CAD).
Methods: A common polymorphism in the p53 gene, Pro72Arg (rs1042522), results in the substitution of arginine (Arg) for proline (Pro) at codon 72 in the amino acid sequence of the protein. The Arg72- has been reported as a more potent inducer of apoptosis than the Pro72 variant. One hundred young (mean age 37.5 years, range 24–45) male Asian Indian patients with CAD confirmed at angiography were compared with 100 healthy control subjects matched for age, gender and ethnicity. Polymorphic variants were assessed by polymerase chain reaction–restriction fragment length polymorphism.
Results: The frequency of p53 codon 72 genotypes were 28% Arg/Arg, 48% Arg/Pro and 24% Pro/Pro in CAD patients compared to 30, 61 and 9%, respectively, in the control group. A significantly higher frequency of the p53 Arg72 allele was found in CAD patients compared to the p53 Pro72 allele (52 vs 40%, $p < 0.0121$, OR = 1.659, 95% CI: 1.116–2.467). Lipid and glycaemic indices were not significantly influenced by the p53 genotypic variants.
Conclusion: The p53 Arg72 allele is associated with increased risk of CAD in this cohort of Asian Indian male patients with premature CAD.

1458: EFFECT OF HUMAN UMBILICAL CORD BLOOD-DERIVED MESENCHYMAL STEM CELL TRANSFUSION IN MONOCROTALINE-INDUCED PULMONARY HYPERTENSION IN A RAT MODEL

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Background: Pulmonary arterial hypertension (PAH) causes right ventricular failure and possibly even death due to progressive increase in pulmonary vascular resistance. Human umbilical cord blood-derived mesenchymal stem cells (hUCB-MSCs) are regarded as an alternative source of bone marrow-derived mesenchymal stem cells because collection of cord blood is less invasive than that of bone marrow. hUCB-MSCs have recently been studied for evaluation of their potential as a source of cell therapy. However, it is rare to investigate use of hUCB-MSCs in PAH. The purposes of this study were to investigate changes in haemodynamics, pulmonary pathology and gene expressions of endothelin (ET)-1, ET receptor A (ERA), endothelial nitric oxide synthase (NOS) 3, matrix metalloproteinase (MMP) 2, tissue inhibitor of matrix metalloproteinase (TIMP), interleukin (IL) 6, and tumour necrosis factor (TNF)-α in monocrotaline (MCT)-induced PAH rat models after hUCB-MSC transfusion.
Methods: The rats were grouped as follows: control (C) group, subcutaneous (sc) injection of saline (0.1 ml/kg); M group, sc injection of monocrotalline (MCT); hUCB-MSC transfusion (U group). hUCB-MSCs (3×10^6/ml/cm^2) were transfused by intraperitoneal injection one week after MCT injection.
Results: The mean right ventricular pressure (RVP) significantly decreased in the U group compared with the M group in weeks two and four. Right ventricle (RV) weight and the ratio of RV/left ventricle (LV) + septum significantly decreased in the U group compared to the M group in weeks two and four. Gene expressions of ET-1, ERA, NOS 3, MMP 2, TIMP, IL-6 and TNF-α significantly decreased from week two in the U group compared with the M group.
Conclusion: After hUCB-MSC transfusion, there was an improvement in RVH and mean RVP. Decreases in several gene expressions were observed. Additional research on the dose and frequency of hUCB-MSC infusions is needed to better determine the optimal parameters for PAH treatment.

1459: REGULATORY B CELLS IN HUMANS: IDENTIFYING THE REGULATORY CAPACITY AND INTERLEUKIN-10 PRODUCTION OF REGULATORY B CELL PHENOTYPES

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Background: In mice, CD1d+CD5+ B cells have regulatory properties associated with interleukin-10 (IL-10) production. In humans, this phenotype is up to 10 times more frequent in infants than in adults. Infants show better heart transplant outcomes than older recipients, including acceptance of ABO-incompatible grafts. However, they also show increased severity of infection with polysaccharide-encapsulated bacteria. We hypothesised that CD1d+CD5+ B cells contribute to the altered immune response during infancy, particularly towards polysaccharides, including ABO-antigens and bacteria capsules.
Methods: CD1d+CD5+ B cells were FACS-sorted from paediatric splenocytes and cultured parallel to non-CD1d+CD5+ B cells using T-dependent (TD; α-IgM+CD40L) and T-independent (TI; CpG or α-IgM+crosslinker) B cell stimuli to measure IL-10 in supernatants by ELISA. The regulatory impact of CD1d+CD5+ B cells on other cells was assessed through proliferation of CFSE-stained (1) peripheral blood mononuclear cells ($PBMC^{original}$), (2) PBMC to which CD1d+CD5+ B cells were added to double the original proportion ($PBMC^{double}$), and (3) PBMC from which CD1d+CD5+ B cells were depleted ($PBMC^{depleted}$) after stimulation with B-cell stimuli or T-cell stimuli (α-CD3+CD28).
Results: IL-10 levels were higher with TI than with TD stimulation, however, little difference was observed between CD1d+CD5+ B cells and non-CD1d+CD5+ B cells. The mean percentage of dividing B cells stimulated with CpG was 21.9 lower in $PBMC^{double}$ than in $PBMC^{original}$ ($p = 0.018$). When stimulated with α-IgM+CD40L, it was 31.9 higher in $PBMC^{depleted}$ than in $PBMC^{original}$ ($p = 0.042$). The percentage of dividing T cells was 26.2 lower in $PBMC^{double}$ than in $PBMC^{original}$ ($p = 0.088$).
Conclusions: These results indicate that CD1d+CD5+ B cells in humans inhibit the proliferation of B and T cells. Since IL-10 production was also found in non-CD1d+CD5+ B cells, these markers do not uniquely identify regulatory B cells in humans. Further analyses to determine the phenotype of IL-10-producing B cells are underway.

1546: EFFECTS OF MELATONIN TREATMENT ON CARDIAC FUNCTION IN A MODEL OF PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary arterial hypertension (PAH) is a disorder characterised by elevated pulmonary arterial pressure, which leads to cardiac hypertrophy and ventricular dysfunction. Current treatments are only marginally effective and additional therapies are required. Melatonin is a natural product that has been shown to be cardioprotective against hypertension and myocardial infarction. We therefore propose that a chronic melatonin treatment may be cardioprotective in a rat model of monocrotaline (MCT)-induced PAH.
Methods: Male Long Evans rats (150–175g) received a single subcutaneous injection of MCT (80 mg/kg), which induced PAH after 28 days. Melatonin was given in the drinking water (4 mg/kg/day) for the 28-day period. Cardiac hypertrophy was confirmed with a ratio of the right ventricle weight over heart weight (RVW/HW). Cardiac functional parameters were assessed at zero and 28 days using isolated heart perfusion and/or echocardiography. These parameters included right ventricular systolic (SP) and diastolic pressure (DP), ejection fraction (EF) and fractional shortening (FS).
Results: MCT increased RVW/HW, reduced EF (92.84 ± 1.33% vs 60.53 ± 4.23%, $p < 0.0003$), FS (28.23 ± 2.68% vs 61.03 ± 2.89%, $p < 0.0002$) and increased SP and DP. Chronic administration of melatonin in MCT-treated rats improved EF (60.5 ± 4.2% vs 84.1 ± 1.7%, $p < 0.0008$), FS (28.2 ± 2.7% vs 48.7 ± 2.1%, $p < 0.0005$), SP and DP.
Conclusions: Our data demonstrate that chronic melatonin improved cardiac function in MCT-induced PAH and suggest a cardioprotective role of melatonin in PAH.

1561: THE ROLE OF NOVEL PROTEIN–PROTEIN INTERACTIONS IN THE FUNCTION AND MECHANISM OF THE SARCOMERIC PROTEIN, MYOSIN-BINDING PROTEIN H (MYBPH)

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Introduction: Left ventricular hypertrophy is a major risk factor for cardiovascular morbidity and mortality and is a feature of common diseases, such as hypertension and diabetes. It is therefore, important to understand the underlying mechanisms influencing its development. Hypertrophic cardiomyopathy (HCM) has been viewed as a model disease in which to study the causal molecular factors underlying isolated cardiac hypertrophy. HCM is described as a disease of the sarcomere, and one of the regions of the sarcomere that has been identified as playing a key role in the regulation of contractility is the C-zone. Identifying binding partners of one of the C-zone proteins, myosin-binding protein C (MyBPC), has led to insights

into the function of this protein. However, myosin-binding protein H (MyBPH) is another member of the myosin-binding protein family, located within this region, of which very little is known. Given the sequence homology and similarity in structure between MyBPC and MyBPH, we proposed that MyBPH may play critical roles in the cardiac sarcomere and possibly in HCM pathogenesis.
Methods: The present study reports the identification and verification of interacting partners of MyBPH with the aim of identifying the role of this protein in the sarcomere using yeast two-hybrid (Y2H) analysis.
Results: Twelve interacting partners were identified, of which three [SUMO-conjugating enzyme UBC9, alpha cardiac actin (ACTC), and myosin 7 (Myh7)] were considered putative physiological interactors based on the plausibility of the interactions as assessed *in silico*. Putative interactors UBC9, ACTC and Myh7 proved to co-localise with MYBPH in differentiated rat cardiomyocyte cells. Furthermore, co-immunoprecipitation confirmed the interaction between MYBPH and UBC9, ACTC1 and MYH7.
Conclusion: The results of this study provide important clues to the function of MyBPH and, in so doing, improve our knowledge and understanding of this protein's role in the cardiac sarcomere.

1627: GLUCOCORTICOID-INDUCED CARDIOPROTECTION: A NOVEL ROLE FOR AUTOPHAGY?

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Ischaemic heart disease is a leading cause of death worldwide, therefore better treatment or prevention of ischaemia–reperfusion (I/R)-induced stress in the heart necessitates a better understanding of the molecular pathways and mechanisms of cell death. The well-established anti-inflammatory and immunosuppressive properties of glucocorticoids have led to their investigation as possible therapeutic agents to reduce ischaemia–reperfusion-induced stress in the heart. However, influences of glucocorticoids on cardiovascular disease and cell death are complex and often contradictory. I/R-induced stress leads to three types of cell death, which include apoptosis, autophagy and necrosis. Although autophagy is foremost a survival mechanism activated during cellular stress, it can also lead to cell death under certain conditions. Many signalling pathways interlink with the autophagic machinery and are activated during I/R-induced stress in the heart, such as the mitogen-activated protein kinase family, which include p38-MAPK. These kinases are subsequently dephosphorylated by appropriate phosphatases. MAPK phosphatase-1 (MKP-1), a dual specificity phosphatase, inactivates the MAPKs by dephosphorylating specific Thr/Tyr residues. Up-regulation of MKP-1 during I/R-induced stress in the heart has been shown to be cardioprotective, however, little information exists regarding the role of autophagy in GC-induced protection in the heart. Therefore, the aim of this study is to describe some of the major signalling pathways activated during I/R-induced stress and the potential role of autophagy in GC-induced cardioprotection. By dissecting out the roles of autophagy and glucocorticoids with regard to shared metabolic effects and signalling pathways in cardiac injury, it is hoped to provide a framework for improved treatment of cardiovascular disease.

1704: DIFFERENTIATING TRANSMURAL FROM TRANS-ANASTOMOTIC GRAFT ENDOTHELIALISATION THROUGH AN ISOLATION LOOP-GRAFT MODEL

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Background: The absence of a physiological intima is the primary reason for low patency in small to medium-sized synthetic vascular conduits. Despite incomplete endothelial surface coverage by trans-anastomotic outgrowth, vascular graft models have yet to distinguish this form of healing from trans-mural capillary sprouting. We have developed an isolation loop-graft model that clearly separates these distinctly different events.
Methods: Trans-anastomotic outgrowth was measured by implanting expanded polytetrafluoroethylene (ePTFE; ID 1.7 mm, IND 15–25 μm) for 2.4 and six weeks (n = 6 per time point) in the abdominal aorta of Wistar rats. High-porosity polyurethane (PU; ID 1.7 mm, 150-μm pore) grafts were then interposed between the ePTFE for two, four, six and eight weeks (n = 6 per time point). Looping the interposition grafts increased their length to 8 cm and they were implanted for six, eight, 12 and 24 weeks (n = 8 per time point). Grafts were analysed by light, immunefluorescence (CD31) and scanning electron microscopy. Endothelialisation was expressed as maximal outgrowth (I_{max}) and segment graft coverage (GSE).
Results: Six-week proximal and distal trans-anastomotic growth rate did not differ (I_{max} = 0.3 ± 0.3 vs 0.3 ± 0.2 mm/week, NS). The composite straight-graft ePTFE zones were too short to isolate trans-mural ingrowth; only 8% of the grafts had mid-graft endothelial coverage without trans-anastomotic breach. All six- and eight-week straight composite grafts had trans-anastomotic encroachment. This outgrowth edge never traversed the endothelium-free isolation zone in the loop grafts (23.6 ± 10.1 mm at 6 weeks and 10.5 ± 45.7 mm at 24 weeks), which separated it from trans-mural mid-graft endothelium. Trans-mural mid-graft endothelialisation reached pre-confluence by six weeks (GSE = 55 ± 45%) and confluence between week 12 and 24 (GSE = 95.0 ± 10.0% and 84.0 ± 30.13%). The sub-intimal thickness stayed constant with a non-significant trend towards regression (91.8 ± 93.9 mm vs 71.4 ± 59.4 mm at six and 24 weeks, respectively; NS).
Conclusion: Trans-mural endothelialisation can be clearly distinguished from trans-anastomotic outgrowth in a high-throughput rat model. A looped interposition-graft model provides sufficient isolation length to separate the two events for up to half a year, and does not result in an increase in intimal hyperplasia.

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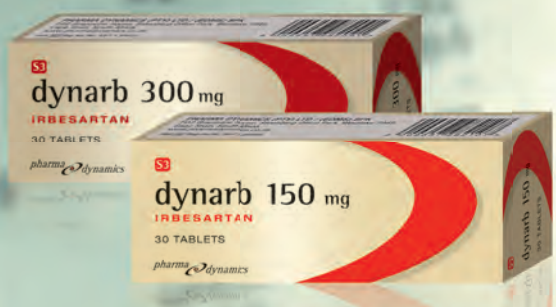

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2. Simon TA et. al. Safety of irbesartan in the treatment of mild to moderate systemic hypertension. American Journal of Cardiology. 1998;82:179-182.
3. Department of Health website: http//www.doh.gov.za – accessed 01/11/2012.

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