TRACK 5: CARDIOLOGY AND THE IMAGING REVOLUTION

Abstract no: 1

Real time 3-D 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 3-dimensional echocardiography (RT3-DE) is reported to show better correlation with magnetic resonance imaging (MRI) in estimating LV and LA volume than conventional 2-dimensional echocardiography (2-DE). On the other hand, the volume measurement in RT3-DE is also reported to be significantly smaller than those in MRI, necessitating the establishment of normal values of RT3-DE itself.
Aim: To identify the normal values of LV and LA volume measured by RT3-DE in Japanese children.
Methods: Sixty four normal school students (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, 2-D biplane method, and RT3-DE. 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 RT3-DE data sets and were analysed.

- RT3-DE: LV end-diastolic volume index (LVEDVI) = 51.4±5.7ml/m^2, LV end-systolic volume index (LVESVI) = 21.0±4.2ml/m^2, max LA volume index (LAVI) = 21.4± 5.0ml/m^2, min LAVI = 7.7± 2.7ml/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±10.8ml/m^2, LVESVI = 23.5± 5.5ml/m^2, LVEF = 66.8±6.1%.
- 2-DE biplane: max LAVI = 22.8± 5.6ml/m^2 LV end-diastolic volume on RT3-DE 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 RT3-DE images. The discrepancy of LVEDV between RT3-DE and M-mode was significant and the measurements of RT3-DE were constantly smaller than those of M-mode.

Abstract no: 4

Reduced aortic elasticity and ventricular dysfunction late after paediatric meningococcal septic shock: A precursor of atherosclerosis?

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Objectives: To prospectively assess aortic elasticity and biventricular systolic and diastolic function in pediatric patients after meningococcal septic shock (MSS) by using magnetic resonance imaging (MRI).
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.
Methods: Eighteen pediatric MSS survivors (8 male; age 14.5 years ± 3.9; MRI 8.2 years ± 2.4 after MSS) treated with at least 2 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: 4.1m/s±0.3 vs. 3.3m/s±0.5, p<0.01; PWV descending aorta: 3.9m/s±0.9 vs. 3.2m/s±0.4, 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 52.1gram/m^2±8.4 vs. 36.0gram/m^2±9.9, p<0.01; RV 26.8gram/m^2±6.5 vs. 10.4gram/m^2±5.0, p<0.01). Also, delayed biventricular relaxation was found after MSS: E -wave deceleration time was significantly prolonged across the mitral valve (MV) (184msec±61 vs. 116msec±28, p<0.01) and tricuspid valve (TV) (192msec±67 vs. 126msec±40, p<0.01) with loss of diastasis time (MV: 22msec±35 vs. 159msec±92, p<0.01; TV: [13msec±24 vs. 113msec±70, p<0.01]). Also, peak filling rates corrected for end-diastolic-volume (PFREDV) across the MV and TV were significantly reduced (MV: PFREDV of E-wave 2.54±0.56 vs. 3.08±0.63, p=0.01; PFREDV of A-wave 1.10±0.26 vs. 1.31±0.30, p=0.03; TV: PFREDV of E-wave 1.81±0.44 vs. 2.09±0.29, p=0.04; PFREDV of A-wave 1.11±0.22 vs. 1.42±0.39, p<0.01). Increased PWV in 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, p0.01, 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 pediatric 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.

Abstract no: 6

Predicting sub-endocardial ischaemia in humans

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Background: In 1972[1] we demonstrated that a ratio $\frac{DPTI}{SPTI}$ <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/100ml blood);[2] the critical ratio is ~10. This ratio probably applies to normal human hearts,[3] 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,[4] so that MVO_2 ∝ 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.[5] These variables can be quantitated easily by echocardiography and applied to patients.

References: 1. G.D. Buckberg, et al. Experimental subendocardial ischaemia in dogs with normal coronary arteries. Circ Res 1972;30:67-81. 2. J. Brazier, N. Cooper, G.D. Buckberg, The adequacy of subendocardial oxygen delivery: The interaction of determinants of flow, arterial oxygen content and myocardial oxygen need. Circulation1974;49:968-977. 3. R.J. Barnard, et al. Ischaemic response to sudden strenuous exercise in healthy men. Circulation 1973;48:936-42. 4. W. Grossman, D. Jones, L.P. McLaurin. Wall stress and patterns of hypertrophy in the human left ventricle. J Clin Invest; 1975;56:56-64. 5. B.-E. Strauer Myocardial oxygen consumption in chronic heart disease: Role of wall stress, hypertrophy and coronary reserve. Am J Cardiol 1979;44:730-740.

Abstract no: 7

Abnormal ventricular torsion: Key to diastolic dysfunction

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Background: 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 counter clockwise; the angular difference in these rotations (torsion) is measured by magnetic resonance imaging (MRI) 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 - 90msec 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 - 90m/sec hiatus is essential for normal function.

Results: In disease, excessive prolongation of torsion by persistence of contraction of 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.

Conclusion: 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.

Abstract no: 17

Low recurrence rate in treating atrioventricular nodal reentrant tachycardia with triple freeze-thaw cycles

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Background: Cryoablation is an alternative to radiofrequency ablation in treating atrioventricular nodal reentrant 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.

Materials and methods: We performed retrospective single center chart review of consecutive patients ≤18 years of age with AVNRT who underwent cryoablation between January 2007 and August 2009. During cryoablation, a 6mm tip cryocatheter was used with temperature set to -80°C. Test lesions were performed at the presumed slow pathway location based on combined anatomic and electrophysiologic approach. If successful, ablation was then continued with triple freeze-thaw cycles (FTC) of 4 minutes each.

Results: A total of 53 patients (age range: 6.1 - 18.4 years, mean: 13.6 years, median: 13.2 years) underwent slow pathway modification with cryoablation. Acute success was achieved in 51 (96.2%) cases. Transient atrioventricular block was seen in 19 (35.9%) cases, but no patient had permanent heart block. Number of FTC was 3 in 47 (92.2%) patients. Less than 3 FTC were given in 2 patients due to transient heart block and 4 FTC were given in 2 patients with suspected catheter movement. Procedure duration was 177±56 minutes; fluoroscopic time was 14±11 minutes. Mean follow-up was 30.7±10 (range 12 - 52, median 31) months. Recurrence of supraventricular tachycardia was seen in only 1 (1.96%) patient.

Conclusion: Triple FTC cryoablation lesions resulted in a comparable low recurrence rate as RF ablation in treating AVNRT without increased complications.

Abstract no: 25

Outcome of 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 asplenia. Cardiac defects included dextrocardia 44%, single ventricle 66% and common atrioventricular valve in 100% of

cases. Transposition or double outlet ventricle was seen in 56% and 44%, respectively. Total anomalous pulmonary venous drainage appeared 75%. Pulmonary outflow obstruction was identified in 91%. Arrhythmias were evident in 28%. 2 (6%) had atrioventricular block and pacemaker treatment. Surgery was performed on 78% of patients, 7 patients (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 <4 weeks of age had mortality of 60% at 5-year period and those requiring surgery at 4 weeks or later had mortality 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.

Abstract no: 33
Cardiovascular disorders in adolescents with chest pain

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Background: Chest pain is one of the chief complaints that make parents 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 are more often due to musculoskeletal problems, gastrointestinal, lung, idiopathic, and psychogenic.
Purpose: To acknowledge the involvement of the 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 from January 2008 - January 2011. The presence of established cardiovascular disorders were based on history, physical examination, electrocardiography and echocardiography.
Results: Found 13/25 adolescents with chest pain who have cardiovascular abnormalities. Of the 25 teens that came with chest pain, most of which showed normal electrocardiographic results, only 9/25 who have dysrhythmias are sinus tachycardia and 8 having a first degree AV block. Echocardiography examination showed only 4 patients with abnormal cardiac anatomy. There is no correlation of nutritional status and adolescents with cardiovascular abnormalities adolescents with chest pain (p=0.206 and p=0.632). There is a positive correlation of sex 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 have no correlation with cardiovascular abnormalities.

Abstract no: 36
Therapeutic role of mobilised bone marrow cells in children with non-ischaemic dilated cardiomyopathy

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Background: 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.
Subjects and methods: This case control prospective study was conducted on 20 children with idiopathic dilated cardiomyopathy following up at the Cardiology Clinic Children's Hospital, Ain Shams University who were compared to 10 age and sex 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-cells assessment in peripheral blood before and one week after granulocyte colony stimulating factor intake for 5 consecutive days.
Results: A significant improvement in echocardiographic data and increase of the cluster of differentiation 34 T-cells was found in patients post granulocyte colony stimulating factor intake. The percentage of change of the cluster of differentiation 34-T cells showed no significant correlation with the percentage of change of the 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.

Abstract no: 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 crescent-shaped 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 echocardiography 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: Multi-parametric transthoracic echocardiographic analysis including global function parameters for RV ejection fraction (RVEF), fractional shortening (RVFS), Tei index and dP/dtmax; longitudinal function parameters (tricuspid annular plane systolic excursion (TAPSE), lateral tricuspid annulus TDI peak systolic velocity (S'), tricuspid regurgitation (TR) evaluation and asynchrony assesssment, 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/dtmax, TAPSE and 2-D 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. Though, 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.

Abstract no: 43

Multi-parametric 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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Purpose: 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 multi-parametric echographic approach including 2-D strain and 3-D for RV volumes and function assessment, in comparison with MRI.
Methods and results: We performed a complete echocardiographic study including 2-D parameters (TAPSE, S' TDI, Tei indices, fractional area change (FAC)), 2-D strain and 3-D and a non-sedated cardiac MRI in 26 consecutive patients with repaired ToF (rToF) before pulmonary valve replacement and one year after surgery. TAPSE, S' TDI and 2-D 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<0.01and r=0.68, p<0.01, respectively). Despite RV volumes underestimation, 3-D 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: Global approach of RV function using 2D (FAC) or (3D) parameters seems reliable in patients with repaired ToF. The commonly used TAPSE and S'TDI focused on segmental analysis of RV inflow are less sensitive probably because RV inflow is less affected by RV remodelling related to initial surgical repair.

Abstract no: 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 anomalies and of the subvalvular apparatus. Its presentation in adults is not frequent.
Objective: In this work, we assessed the clinical and echocardiographic aspects as well as the treatment of 5 adult patients with CAMV.
Methods: A complete clinical history was made for each patient and also an electrocardiogram, chest X-ray, and transthoracic echocardiogram.
Results: Two patients were in functional class (FC) I, 1 in FC II, and 2 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;
- Tri-leaflet mitral valve with subaortic obstruction;
- Double mitral orifice associated with bicuspid aorta and aortic coarctation, and
- Tunnel-forming mitral valve associated with ostium primum inter-atrial communication and pulmonary arterial hypertension and pulmonary artery hypertension.

One patient was subjected to mitral valve change, 1 to dilation of the aortic coarctation, and another is under sitaxentan treatment, with improvement in the FC. The 2 remaining patients are awaiting surgery.
Conclusions: Review of these cases with CAMV reveals the relevance of the echocardiogram in the evaluation of the mitral valve and of the subvalvular apparatus, because it allows for the identification of different types of malformations and their haemodynamic repercussion, to be able to propose the precise and timely treatment for these patients.

Abstract no: 46

Anatomo-echocardiographic correlation in complex congenital heart disease

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Background: Echocardiography is a valuable noninvasive 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 a correct interpretation of diagnostic images and clinical decisions.
Objective: The objective of this study was to establish the anatomo-echocardiographic correlation in complex congenital heart disease from heart samples with equivalent findings to those of echocardiographic images.
Material and methods: Thirty hearts and 50 patients with Ebstein's anomaly, 60 hearts and 24 patients with atrioventricular septal defect, 15 hearts and 24 patients with absence of right atrioventricular connection were studied. The samples correspond to the Collection of the Embryology Department.
Results and conclusions: The anantomo-echocardiographic correlation clearly showed that the anatomical findings of the hearts correspond with the echocardiographic images of complex congenital heart disease and provide an adequate understanding of the echocardiographic images in terms of an accurate diagnosis, treatment, therapeutic decisions and prognosis.

Abstract no: 48

Ventricular non compaction associated with complex congenital heart disease

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Objective: To describe the clinical behaviour and the echocardiographic features of 6 patients with ventricular non-compaction associated to complex congenital heart disease.
Methods: A complete clinical history and the echocardiographic study were performed in 6 patients with complex congenital heart disease. Jenni's criteria for the echocardiograph assessment of ventricular non-compaction were used.
Results: In all cases prominent trabeculae and deep inter-trabecular recesses in the ventricular wall filled by direct blood flow from the ventricular chamber were visualised by colour Doppler imaging. The ratio of non-compacted to compacted layer was of 2.5±0.19. The associated congenital heart diseases were: Uhl's anomaly, the absence of right atrioventricular connection, single ventricle, cleft of mitral valve, transposition of the great arteries, double inlet in left ventricle. Two of these patients went to surgery: one for Blalock-Taussig shunt and the other one for pulmonary banding (Table 1).

TABLE 1: Characteristics of studied patients

Patient	Age	Sex	Ventricular non- compaction	Congenital heart disease	Surgery	NYHA
1	27	Male	Left ventricle	Uhl's anomaly	No	II
2	27	Male	Left ventricle	Tricuspid atresia and pulmonary atresia.	Blalock-Taussig shunt.	II
3	21	Male	Single ventricle	Single ventricle and total anomalous drainage of pulmonary veins	No	II
4	30	Male	Left ventricle	Cleft of mitral valve	No	II
5	3	Male	Biventricular	Corrected transposition of the great arteries	Pulmonary Banding	II
6	19	Male	Left ventricle	Double inlet left ventricle. Corrected transposition of the great arteries	No	II

Conclusions: The clinical manifestations were presented in the childhood and they were associated with congenital heart defects. The echocardiography is a diagnostic method, which allows the assessment of the ventricular non-compaction.

Abstract no: 50

Contribution of MRI and CT scan in diagnosis and management of congenital heart disease

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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.
Patients and methods: This is a descriptive study from January 2008 - March 2011 including patients with CHD who underwent Trans-Thoracic Echocardiography (TTE) and in whom MRI or CT was performed
Results: We identified 10 cases who underwent MRI and 8 cases CT scan. Patients's average age was 10 years.The indications of MRI were always preoperative assessment of:

- Fallot's syndrome;
- 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 anatomic diagnosis of:

- Hypoplastic left pulmonary artery (LPA);
- Tight coarctation of the aorta;
- A bi-carotid trunk;
- Rupture of a LV aneurysm within the pericardium; and
- Distal migration of a pulmonary banding.

CT was indicated for:

- Stenosis at the origin of the pulmonary branches after ligation of the ductus arteriosus; and
- Fallot tetralogy with pulmonary hypoplasia.

CT scan provided a better description of the aorta, but also the pulmonary arteries and their abnormalities.

Conclusion: MRI and CT scan can be highly contributive in preoperative assessment of congenital heart defects. They can be indicated for more accurate anatomic diagnosis prior to surgery and may prevent the use of invasive cardiac catheterisation.

Abstract no: 54

Intra-atrial rhabdomyoma in a newborn producing pseudo pre-excitation

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Background: 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 (Movie 1).

Methods and subjects: 1st presentation to our clinic was for further diagnostic and therapy. Except from the cardiac tumours other physical findings were unremarkable for age and gender. Initial ECG showed atrial fibrillation with ventricular rate of 230/minute, which was terminated by a single direct current shock of 6 Joule. (Figure 1) After cardioversion ECG showed slow atrial rhythm with frequent atrial premature contractions (PAC) and deformation of the PR interval imposing as pseudo pre-excitation, followed by a normal QRS complex with seemingly abnormal ventricular repolarisation. We suspected the origin of these pseudo pre-excitation as being caused by isolated atrial depolarisation disturbances due to tumour caused heterogeneous endocardial activation. Also the seemingly abnormal ventricular repolarisation rather represents excessive delay in atrial repolarisation superimposed on the ventricular repolarisation (Figure 2). Due to development 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 (Figure 3).

Conclusions: Recognisable, the QRS complex before and after surgical resection of the rhabdomyoma is identical, underlining the atrial origin of the repolarisation abnormalities before surgery.

Abstract no: 57

Congenital junctional ectopic tachycardia: Clinical presentation, responses 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.

Materials and methods: Twenty patients were seen in our practice in arrhythmias with congenital JET from 2001 - 2010, with a mean follow-up of 5.9 years (±5.3). All of them 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. They were subdivided according to the age of presentation into Group 1: (younger than 1 year old):16 patients with a mean age at diagnosis of 38 days old (1 - 90 days old) and Group 2: older than 1 hospitalisation: 4 patients with a mean age of 8.7 hospitalisation (7 - 11 hospitalisation).

Results: Group 1: Median HR at diagnosis was of 182 (±32) bpm, it was incessant in all of our patients (p0.002), 87.5% (p0.006) showed signs of heart failure and ventricular dysfunction, and 81% (p0.02) needed to be admitted. In all cases HR (mean of 122 between 90 - 157bpm) was controlled by 45 days (15 - 120 days). Only one patient had a positive family history. **Group 2:** Median HR of the JET was127 (±38) bpm and in 2 pts, paroxistic (p0.09). All of them referred palpitations (p0.0002) and 1 patient required admission. In all cases the HR (mean of 81 bpm, between 69 - 98bpm) was controlled by 35 days (5 - 120 days). In all cases there was a 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 combina-tion with other drugs (p0.02). During follow-up, all patients persist with intermittent JET, with HR under control and heart rate within normal ranges for age.

Conclusions: Children younger than 1-year-old presented generally with incessant JET, ventricular dysfunction and heart failure requiring admission and a combination of drugs. In children older than 1-year-old, 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 remains present in all patients during the follow-up, without spontaneous cure in serial Holter studies.

Abstract no: 59

Scaling cardiac structures in children

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Background/hypothesis: Body composition influence 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 by many investigators been suggested to be the best scaling factor. Data on LBM are, however, not available in clinical practice. We assessed if cardiac dimensions normalised by LBM compares with cardiac dimensions normalised by various anthropometric methods in younger children.

Materials and methods: 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 2-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) in diastole 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 normalised with various anthropometric methods. Summary of partial correlation coefficients, with adjustment for gender, are displayed in Table.

Cardiac structures normalised by LBM	BSA (m^2)	Height $(m)^{2.7}$	Body mass (kg)	BMI (kg/m^2)
LVM	0.93	0.93	0.79	0.75
LVDD	0.79	0.81	0.73	0.41
LVSD	0.86	0.85	0.80	0.60
Sep	0.89	0.89	0.80	0.66
Post	0.91	0.91	0.83	0.71
LA	0.83	0.85	0.65	0.39

Conclusions: This investigation shows close relations between cardiac dimensions normalised by LBM and cardiac dimensions normalised by BSA or height$^{2.7}$. These results support the use of these as appropriate denominators in paediatric echocardiography to normalise cardiac dimensions for differences in body size.

Abstract no: 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.
Materials and 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 cine- and phase-contrast imaging was 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±1.3m/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 10-3mmHg-1, $p>0.05$).
Conclusions: About half of our HLHS patients show 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.

Abstract no: 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-Vol_{min}) and the LA volume before atrial contraction (LA-Vol_{bac}) were higher in patients (LA-Vol_{min}: 25.3±7.6 vs. 20.8±5.4 ml/m^2, LA-Vol_{bac}: 33.2±9.8 vs. 26.8±6.2ml/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 during long-term follow-up.

Abstract no: 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 7 females), aged from 1 month - 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, 2-dimensional mode, colour Doppler, pulse-wave Doppler and continuous-wave Doppler were performed.
Results: (1) Combination of atrioventricular 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 4 cases. (2) Coexistence of atrioventricular valvular abnormalities was seen in 3 cases, including complete atrioventricular septal defect (2 cases) and straddling tricuspid valve (1 case). In addition, functional tricuspid regurgitation was detected in 19 cases (severe in 4 cases, moderate in 8 cases and mild in 7 cases), and functional mitral regurgitation in 10 cases (severe in 1 case, moderate in 1 case and mild in 8 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 coexisted abnormalities was 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 coexisted abnormalities. Additionally, attention should be paid to the functional tricuspid regurgitation in patients with ccTGA.

Abstract no: 73
Lung biopsy diagnosis of operability associated with congenital heart disease and pulmonary hypertension in 358 patients

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Background: One performed lung biopsy diagnosis in order to determine the operability in a total of 358 patients in these 5 years as requested by the department 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), operative indication was determined by the degree of hypoplasia of small pulmonary arteries. Operability of 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 patients. In simple atrial septal defect (ASD), ASD closure was indicated in 21 patients but not in 6. Radical surgery was indicated in 50 patients with AVSD but not in 12. The latter 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 jn 49. Among 7 patients with Tetralogy of Fallot, 1 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 is a prospective study in the patients with congenital heart disease and pulmonary hypertension, results have the confidence of cardiac surgeon, paediatric cardiologist and cardiologist who asked for lung biopsy diagnosis for pulmonary vascular disease.

Abstract no: 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: Retrospective review of children with CHD hospitalised and diagnosed with HRV from January - May 2012. Outpatient, pre-operative, operative and post-operative variables were reviewed with interest in management strategies and clinical impact.

Results: Nine patients tested positive for HRV. Median age was 5 months (range 1 - 9); single ventricle defects (n=3); trisomy 21 (n=2); community acquired HRV (n=5); nosocomial HRV (n=4); post-operative patients (n=3); and surgical management was altered with elective palliative procedure instead of complete repair in 2 patients. Median length of mechanical ventilation associated with HRV was 11 days (range 0-70); median length of non-invasive ventilation was 9 days (range 0 - 45). Post-operative hospital stay increased significantly in patients with single ventricle physiology and in patients with trisomy 21 compared to 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; atrioventricular septal defect repair median length of stay 6 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 preoperative evaluation of respiratory viral bronchiolitis in high risk children prior to cardiac surgery.

Abstract no: 80
Cardiac entrapment and injury due to epicardial pacemaker wires

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Background: 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 7 patients (incidence of <0.02%).
Methods and results: 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 7-year-old patient, unidentified pacemaker lead compression is felt to have caused sudden unexpected death. Post mortem angiography confirmed compression of the circumflex coronary artery. Two other patients, aged 2 and 5 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 repositioned 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.

Abstract no: 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 9 patients with TGA and status post atrial redirection surgery (SenningMustardTGA), endocardial ventricular stimulation (overdrive DDD-mode, 80 - 90bpm) 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. 90bpm; 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 6 out of 7 patients systemic dP/dtmax was even higher during BiVP and systVP than during AAI pacing.
Conclusions: In a population of patients with SenningMustardTGA, acute hemodynamic 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.

Abstract no: 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 of late.
Aim: To assess the left ventricle systolic function in children with primary arterial hypertension with the use of tissue Doppler method.
Material and 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 ventricle myocardial performance index were significantly > in children with arterial hypertension (0.46±0.08 vs. 0.36±0.03). Significantly < 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±2.84cm/s vs. 10.9±2.19 and 16.16±3.30cm/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 is observed. Left ventricle systolic function estimated with the use of standard echocardiographic indices was normal, except myocardial performance index, the value of which was significantly higher compared to the control group.

Abstract no: 91

Syncope unit in a paediatric population: A single-centre experience

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Background: Syncopes are frequent in the paediatric population. The majority is 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 developed in adult population have demonstrated major improvement in diagnostic process, hospitalisation reduction time, with favourable long term outcome. We report our experience of syncope management in children and adolescents through a dedicated syncope unit.
Methods: In this ongoing study prospectively enrolled 45 consecutive patients (13±3 hospitalisation, 65% male) from January 2011 - June 2012, referred for loss of consciousness (LOC) in a dedicated paediatric syncope unit involving a paediatric cardiologist, a nurse, a physiotherapist and a psychologist. All patients underwent initial evaluation including medical history assessment, physical examination, 12-lead ECG and echocardiography to exclude non-cardiogenic syncopes. If initial assessment was abnormal, they underwent targeted tests that differed according to suspected aetiology. Patients with neuro-cardiogenic syncope underwent specific physiotherapy training and a consultation with a psychologist.
Results: The most common causes of LOC were neuro-cardiogenic 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 seizure and was transferred to neurologic department. Mean hospitalisation duration was 0.9±0.5 days. Head-up tilt testing was positive in 62% neuro-cardiogenic syncope. Echocardiograms and exercise tests were not contributive for diagnosis. After a mean follow-up of 9±4 months, including physiotherapist and or psychologist specific care, syncope recurred in 5 patients (12%).
Conclusion: Syncope unit in paediatric population with dedicated team improves diagnostic process, reduces hospitalisation and decreases syncope recurrence when adapted follow-up is proposed.

Abstract no: 94

Impact of the 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 etiology of acquired heart disease in children and adolescents world wide. The major outcomes are heart failure (HF) and death. Nevertheless survival of these patients after the onset of HF is unknown. In addition, the impact on prognosis of social conditions by individual community's human development index (HDI) remains opened.
Purpose: To assess children and adolescents survival rate with RHD and HF, using probabilistic databases linkage methods. To study the influence of residence geographic regions and HDI on the prognosis.
Methods: We performed probabilistic databases linkage from Brazilian hospital admission and death certificates (2001 - 2007). We used the Chi-square, analysis of variance, Kaplan-Meier method for survival curve, and compared groups by log rank test. We estimated hazard ratios (HR) with confidence intervals 95%, followed by Cox proportional hazards model. The significance was achieved by $p<0.05$.
Results: 780 patients were hospitalised by the first time for RHD and HF, with 421 (54.0%) deaths along 7 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 1, 54.9% at 2 and 37.2% at 6 years. There was no difference in survival between gender ($p=0.107$), but by residential geographic regions ($p=0.0001$), with the lower in North and Northeast regions. HDI was lower in North (0.727±0.727) and Northeast (0.682±0.078) compared with Midwest (0.796±0.049), Southeast (0.784±0.051), and South (0.800±0.038) regions ($p<0.0001$). In Cox analysis, the increase in the overall HDI of 0.01 point 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 education (0.968; 0.958-0.977; $p<0.001$) dimensions.
Conclusion: Patients with RHD hospitalised for HF are at increased risk of death. The increase of 0.01 point in HDI reduces that risk.

Abstract no: 97

Atrial septal defect in infancy: A single-center 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 management of 5 patients who were operated in infancy.
Methods: Between January 2009 and June 2012, 5 patients were operated with isolated diagnosis of ASD , the mean age was 6 months , weight 4.5kg, 3 were operated for failure to thrive, 1 for repeated respiratory tract infections and 1 was operated in emergency 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 of them had autologous pericardial closure. Two patients had 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 is 18 months, 6 months - 3 years), 3 children have their pulmonary pressures normalised, with good catch up growth, 2 children still have elevated PAP, with mild FTT, they are being followed up closely and are on sildenafil and Bosentan.
Conclusion: ASD can present in infancy, and would benefit by surgical closure. A proportion of them would have pulmonary hypertensive issues. Whether a concomitant lung biopsy or a fenestrated closure would prognosticate and hasten their post-operative course needs to be evaluated.

Abstract no: 102

New insights into aspects of pulmonary diffusing capacity in Fontan patients

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Background: Functional univentricular heart patients, palliated a.m. Fontan lack a sub-pulmonary ventricle and consequently have non-pulsatile pulmonary bloodflow. Fontan patients are known to have reduced pulmonary diffusing capacity, however, the cause of this reduction remains unclear.
Aim: To assess possible determinants in the aetiology of reduced diffusing capacity and to ascertain if these could be increased. Furthermore, we aimed to search for predictors of reduced diffusing capacity.
Material and methods: Eighty seven Fontan patients (mean age 16.3±7.6 years) performed sitting pulmonary function tests using the single breath method and 2 different mixtures of gas. Furthermore, 72 of the 87 patients performed maximal cycle ergometer tests using the Innocor™ re-breathing technique thereby estimating cardiac output and stroke volume. Ten Fontan patients and 9 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 haemoglobin 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 the 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.

Abstract no: 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 by analysing mortality from the current burden related to age.
Materials and methods: All Danish citizens with Fontan completion from 1981 - 2009 were identified (n=235). Surviving patients performed an exercise test, Holter monitoring, echocardiography, pulmonary function test, and blood sampling. Their medical histories 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 was 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 4 years and survival of the peri-operative period, the probability of being alive by age 40 would be 72%, the probability of clinically relevant arrhythmia 45% and the probability of severe exercise intolerance 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.

Abstract no: 104

Predictive value of response to acute vaso-reactivity testing in children with idiopathic pulmonary arterial hypertension

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Background: Acute testing of vaso-reactivity during catheterization is an important factor in the evaluation of idiopathic pulmonary arterial hypertension (IPAH). 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 a decrease in mean pulmonary artery pressure (mPAP) of ≥20%. The purpose of our study was to assess the predictive value of vaso-reactivity 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 vaso-reactivity 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 3 different response criteria: reduction of mPAP ≥20%, mPAP ≥30%, and mPAP ≤40mmHg.

Results: Baseline mPAP was 65.2±18.3mmHg, mean change in mPAP during vaso-reactivity testing was 25.8±19.1%. 24 of the 42 patients showed more than 20% reduction of the mPAP, 18 of them to a mPAP below 40mmHg. Mean follow-up after catheterization was 55.3 (±40.5) months. Freedom from serious cardiovascular events (lung transplantation or death) was 86% after 2 years, 76% after 3 years, and 57% after 5 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). Predictive value of mPAP ≥30% and ≤40mmHg was superior to reduction of mPAP ≥20% (log rank (Mantel Cox) Chi square 9.98; p=0.002).

Conclusions: Acute vaso-reactivity testing has an impact on outcomes in paediatric IPAH patients although a stricter definition of the response criteria seems to be more reliable to predict serious cardiovascular events.

Abstract no: 109

Rheumatic fever in Estonian children over two decades (1992 - 2011) based on data from children's clinic, Tartu University Hospital

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Background: There is almost no data published about rheumatic fever (RF) among Estonian children. We report our data from the South-Estonian region with a population of approximately 100 000 people.

Methods: Retrospective 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 all patients. The first attack occured at age of 3 - 16 years. Most cases occured from November to April. Cardiac involvement was present in 26 cases (70%): endocarditis in 24 with mitral, in 8with aortic, in 5 with tricuspid and in 11 with multiple valve involvement, peri-carditis in 3, and myocarditis in 1 case. Huntington's chorea was diagnosed in 15 cases (41%); only chorea in 7 (19%). Joint involvements where present in 22 cases (59%). All children were treated with conventional anti-rheumatic and symptomatic therapy. Prophylaxis with benzathine benzylpenicillin was administrated to all children. After, children with cardiac involvement's 1st attack, 6 recovered without sequalae. The others had only mild mitral or aortic valve regurgitations and in 2 cases mild dilatation of LV persisted, and required administration of Enalapril. In 5 cases there are no further data available after the 1st attack. A 2nd attack occured in 7 cases: 5 with cardiac involvement and 2 with chorea. Even after a 2nd attack only mild mitral insufficiencies persisted in only 2 cases. None of our children needed to be operated on during childhood.

Conclusions: RF is a rare disease in Estonia nowadays. In 1995 a small outbreak 8.5:100 000 was evident. Since 2000, 0 - 2 cases per year were diagnosed in our hospital. However, some new cases may still occur in years to follow and it is important to take the possibility of RF into consideration and not being late with well-timed appropriate treatment to prevent its subsequent complications.

Abstract no: 110

No difference in cardiac performance between critical neonatal and non-neonatal patients 1 year post coarctectomy

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Background: Nowadays, the repair of a critical neonatal coarctation has low surgical mortality. It is unknown if subsequent post-operative cardiac performance in this subgroup of coarctation patients differs from those in 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 1st year after coarctectomy.

Materials and methods: Children (0 - 17 years) undergoing an aortic coarctectomy were included and 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, 1 day post-operatively and 1-year post-operatively (11.4±8.3 months post-operatively). 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 1st year following repair. One year post-operatively LV systolic performance had normalised, while LV diastolic performance was still impaired as compared to controls in both neonatal (LV E' 8.7±3.1 vs. 13.3±3.8cm/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±2.4cm/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 1st year following repair. Subsequently, 1-year post-operatively no differences were observed in RV systolic or diastolic performance between neonatal or non-neonatal coarctation patients and controls.
Conclusions: One year post-operatively LV diastolic performance was still impaired in both neonatal and non-neonatal coarctation patients, while RV systolic and diastolic performance was 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.

Abstract no: 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; f: 38; m: 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. 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. Reinterventions in 56 patients included ICD change because 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 respect to the occurrence of appropriate shocks no differences could be found between the primary and secondary prevention group. Patients younger than 6 years were more at risk of an appropriate shock than older children. Methods to prevent further shocks 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, stay at risk for further shocks. Lead-related problems are an important reason for reinterventions.

Abstract no: 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 venacava. This can present either in infancy a sick child or as a relatively asymptomatic older child. There can be other associated conditions which determine the timing of presentation.
Case report 1: Forty five day 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 left thoracotomy with partial improvement in clinical condition. Catheterisation showed multiple collaterals from 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 4 months old and is now planned for pneumonectomy.
Case Report 2: A 7-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 reimplantation of the inferior pulmonary vein into the left atrium without cardiopulmonary bypass through right thoracotomy, with uneventful postoperative course.
Conclusion: Scimitar syndrome can present as extremes, with 1 child having persistent severe pulmonary hypertension and respiratory issues, presenting as neonate, ending up requiring a pneumonectomy, and the other as relatively asymptomatic child where off pump reimplantation 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 determine the timing of presentation.

Abstract no: 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: 14/64 patients (21%) after DSO and 7/29 (24%) after CR showed significant tachycardia. Of these, 12 after DSO and 5 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 ($p< 0.02$). 2 cases (3%) after DSO and 2 cases (7%) after CR developed ventricular tachycardia, 1 with DSO was treated medically and the other was given an implantable cardioverter defibrillator. Thirteen of the 64 patients (20%) after DSO and 5 of the 29 (17%) after CR had bradycardia. Of the 13 cases with bradycardia after DSO (complete atrioventricular block (CAVB) in 7, advanced AVB in 2, and sick sinus syndrome (SSS) in 4), 10 underwent permanent pacemaker implantation (PMI). Among the 4 cases with SSS, 2 underwent PMI 4 - 10 years after DSO. Among the 9 cases with AV block, 4 underwent PMI soon after DSO. CAVB progressed in these 4 patients and PMI was performed 1 - 13 years after DSO. 1 case already had congenital CAVB, and for this case, PMI was performed at the DSO. 5 cases had CAVB soon after CR, and all 5 cases 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.

Abstract no: 123

Just a headache? Brain abscesses in right-to-left shunt lesions

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Background: Although congenital heart disease is a major risk factor for the development of brain abscesses in the paediatric population, the overall incidence is low, becoming even lower with early correction of right-to-left shunt lesions nowadays.
Case reports: We encountered 2 adolescents with brain abscess in recent years. The 1st 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 complaint was headache 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 4 weeks of antibiotics she could return home without any residuals. The 2nd patient was a 21-year-old man with functional univentricular heart and Eisenmenger syndrome. Due to a new onset of headaches he presented at our clinic on the weekend, was admitted and about to be discharged on the following day. As with the young girl, he did not have any fever, neurologic deficits or elevated inflammatory markers. Bearing the previous case in mind, he got a MRI showing 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. A repeat MRI after 10 days showed progression, thus Clindamycin was added for a total of 6 weeks resulting in regression of the abscess.
Discussion: 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.

Abstract no: 125

Reference values for QT and Qtc measurements after brisk standing in healthy (7 - 13 years of age) pre-puberal schoolchildren

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Background/hypothesis: 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 shown to discriminate between normal individuals and genetically proven LQTS patients.
Aim: This research was undertaken to establish reference values for pre-puberal children (7 - 13 years of age) for QT duration and QTc prolongation after brisk standing

Methods: Fifty seven pre-puberal (Tanner score ≤3) healthy schoolchildren underwent a BST during continuous ECG registration. The subjects rested in supine position for 2 minutes, stood up briskly and stayed in vertical position for 1 minute upon which they laid down quickly and rested 2 minutes in supine position. QTc maximal, minimal and mean values where 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 the 57 subjects 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 en girls (Table 1).

TABLE 1: Baseline values and response to brisk standing girls (n=28) vs. boys (n=29)

	Girls	Boys	p-value	All subjects
Age, years	9.9±1.1 (7.0 - 12)	10.2±1.1 (8.1 - 12)	0.429	10.1±1.1 (8.0 - 12.0)
Baseline HR pre standing, beats/minute	76.6±9.2	74.2±9.2	0.330	75.4±9.2 (58 - 100)
Baseline HR post standing	77.5±8.3	73.8±8.8	0.108	75.6±8.7 (60 - 100)
Baseline QT pre standing, ms	355.1±21.0	360.8±21.2	0.320	358±21 (317 - 414)
Baseline QT post standing, ms	349.3±20.2	350.6±21.7	0.821	350±21 (315 - 398)
QT during max tachycardia, ms	356.5±23.1	356.9±25.3	0.688	358±21 (322 - 413)
QT during max QT stretching, ms	355.6±25.9	358.6±24.8	0.656	357±25 (317 - 410)
Baseline QTc pre standing, ms	407.3±17.0	405.6±27.3	0.780	407±23 (367 - 508)
Baseline QTc post standing, ms	404.5±20.0	396.3±27.3	0.203	400±23 (364 - 495)
QTc during max tachycardia, ms	486.6±34.9	483.5±2.6	0.715	485±32 (414 - 562)
QTc during max QT stretching, ms	485.0±36.4	481.9±30.5	0.728	483±33 (423 - 569)
Response to standing				
Time to maximal HR, s	8.6±2.6	8.7±2.9	0.904	8.6±2.7 (2.2 - 15)
Increment in HR, beats/minute	35±9.9	35.5±7.0	0.850	35±9 (20 - 62)
Maximal HR, beats/minute	112.1±11.2	109.3±11.1	0.348	111±11 (87 - 129)
ΔQT during max tachycardia, ms	1.75±17.9	-1.27±16.3	0.507	0.2±17 (-34 - 40)
ΔQTc during max tachycardia, ms	79.3±28.8	77.9±22.6	0.840	-0.9±17 (-32 - 44)
ΔQT during max QT stretching, ms	0.46±15.9	-2.14±17.4	0.557	79±26 (19 - 144)
ΔQTc during max QT stretching, ms	77.7±29.3	76.3±27.8	0.854	77±28 (14 - 158)

Data are mean ± SD (range). HR: Heart Rate, ΔQT: QT interval change from baseline, ΔQTc: corrected QT interval change from baseline, QTc: corrected QT using Bazett's formula.

Whole group QTc prolongation at maximal tachycardia at BST was significantly longer in children compared to adults (79±26ms vs. 50±30ms) (Viskin, et al, JACC 2010 55;18:1955-61)
Conclusions: QTc prolongation after brisk standing in children is more pronounced than in adults. Using adult values for children would thus yield false positive results with the risk of over-diagnostics and over-treatment. Comparison of these BST reference values with DNA positive children from 7 - 13 years is therefore warranted.

Abstract no: 129

Management and outcome of isolated partial anomalous pulmonary venous connection from the left upper lobe

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Background: An anatomic subset of partial anomalous pulmonary venous connection (PAPVC) is venous drainage of the left upper lobe (LUL) of the lung to the in nominate vein via a vertical vein. The operative approach, outcomes, and follow-up strategy for this lesion are not well-reported.
Aim: 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, post-operative 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 6 patients were female (50%). The operative procedure involved median sternotomy, cardiopulmonary bypass, ligation of the vertical vein at its insertion to the in nominate vein, and anastomosis of the vertical vein to the left atrium. There were no peri-operative or late deaths. Post-operative 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 post-operative 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 reintervention. Four patients (33%) had post-operative complications, including 2 with atrial fibrillation, 1 with post-pericardiotomy syndrome, and 1 with left phrenic nerve dysfunction. Median length of hospital stay was 4 (3.5) days.

TABLE 1: Patient characteristics and outcomes

Variable	n=12
Age (median, [25th, 75th percentile])	14 (6.37)
Weight (kg)	71 (22.84)
Median follow-up (months)	26 (5.81)
Median pre-operative pulmonary artery pressure (mmHg)	22 (21.25)
Cardiopulmonary bypass time (minutes)	91 (55.318)
Cross clamp time (minutes)	41 (0.142)
Patients with post-operative echocardiography	11 (92%)
Patients with post-operative MRI	3 (25%)
Post-operative complications	4 (33%)
Median post-operative tricuspid regurgitant gradient (mmHg)	27 (20.47)

Conclusions: PAPVC from the LUL to the in nominate vein via a vertical vein can be repaired using this technique with low morbidity and mortality, and without post-operative pulmonary venous obstruction. Pulmonary hypertension was not observed during follow-up. Post-operative imaging of LUL-PAPVC repair can be difficult, and MRI should be considered as an alternative to echocardiography in imaging these patients.

Abstract no: 131
The Taranaki regional area plan to save kids 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 <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 Cardioversion (DCC). Time to DCC shock is proven to be critical to outcome.
Methods: We report on a project in New Zealand's Taranaki Region to place Ambulatory Automatic Defibrillators (AEDs) throughout the community. Taranaki is on the west coast of New Zealand's North Island. Population 109 000, area 7 257 km², population 84% European ancestry, 15% Maori. Since the Sudden Unexpected Death 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. Aim is to get as good 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 also aware of all AED placements and will either tell the emergency caller or ask an equipped business to deliver if nearby.
Results: Several devices have been used appropriately since being placed, though not successfully yet.
Conclusion: It is feasible to get wide geographical coverage of AEDs and use modern technology to improve usage.

Abstract no: 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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Background: Taranaki is a province on the West Coast of New Zealand, population 109 000, 85% White, 14% Maori.
Aim: 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.
Methods and material: We are meant to have all sudden unexpected/unexplained deaths in persons 2 - 40 years reported to us. Referrals for syncope are also investigated once/if referred. Investigations include autopsy, DNA studies in index cases; clinical examinations, ECG, ETT, echocardiography and, where appropriate, cardiac MRI and 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 is not an easy service to implement and is very time and resource consuming.

Abstract no: 142

Born with huge mediastinal mass: 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. Intra-pericardial teratomas are significantly rare and generally seen in infants and young children. Interestingly, malignant teratomas are extremely rare with a prevalence of 4-6 cases in 10 000. It presents itself mainly in female newborns weighing <3kg. 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 intra-pericardial 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 then 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 malignant immature teratoma. Chemotherapy was started on day 4 post extubation. Child remained in stable condition to date.
Conclusion: Immediate post birth detection and surgical management were essentially life saving. Yet prognosis of this child is considered to be poor in light of the incomplete resection of the tumour and the course of chemotherapy required.

Abstract no: 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 and high incidence of heart transplantation (HTX). Pulmonary artery banding (PAB) has been proposed in patients with ccTGA to retrain the sub-pulmonic 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-center observational study was performed to evaluate dilatable surgical PAB in infants and young children with LV-DCM.
Results: Since April 2006, 10 infants and 2 toddlers with LV-DCM referred for HTX received a PAB. Additional, 4 patients underwent repair of the left-sided total TAPVR, re-implantation of an ALCAPA, mitral valve repair or replacement. There was no hospital mortality, but clinical improvement in all patients. Median age at operation was 126 (1 - 756) days of the 12 patients without additional operation. The pressure gradient across the PAB increased significantly. The LV ejection fraction increased from median 15% pre-PAB to 43% at discharge home, and 47%, 3 - 6 months later. The median LVEDD and z-score decreased from 45 - 30mm ($p>0.001$) and +6.1 - +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 functional class 1. Two patients, both with non-compaction DCM, deteriorated 5 and 6 months after PAB-de-banding and finally died. One listed for HTX.
Conclusion: In young children with LV-DCM and still preserved right ventricular function, PAB led to an improvement of LV and mitral valve function by ventricular interaction and still unknown factors.

Abstract no: 147

Global and regional circumferential and radial myocardial deformation and torsion in elite high school athletes: Physiologic 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.
Aim: The purpose of this study was to assess the physiologic impact of long term training and the type of sport participation on parameters of LV radial and circumferential deformation as well as LV torsion in this same cohort.
Methods: Standard two-dimensional (2-D), spectral Dopplerand tissue Doppler were prospectively performed utilising 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, & basal short-axis segments) and global LV strain.

Results: Traditional measures of LV systolic & diastolic function as well as tissue Doppler, global circumferential 2-D strain and averaged radial 2-D 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 vs. 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 physiologic basis of augmented regional contractile reserve seen with high intensity training and may also assist in distinguishing athletic from myopathic increases in LV mass.

Abstract no: 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.
Objective: The purpose of this study was to identify common associated symptoms and their clinical significances in children with KD.
Subjects and methods: We retrospectively reviewed the medical records of 121 children who were treated for KD at Presbyterian Medical Centre from January 2005 - June 2010. We collected and analysed all clinical symptoms, laboratory data and echocardiographic findings.
Results: We identified 9 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. As compared with complete KD, the incidence of seizure was significantly higher in patients with incomplete KD. Vomiting was strongly associated with 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.

Abstract no: 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 post surgurey the patients are considered healthy with normal physical capacity. If this is actually true, it still has not been clarified thus making it 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 EKG. End points were: maximal oxygen uptake, maximal workload and anaerobic ventilatory threshold. For the latter both absolute and relative thresholds were measured using V-slope. Before each test a spirometry was made to measure FVC, FEV1 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 38.0ml $O^2kg^{-1}min^{-1}$ (31.6 - 40.8ml $O^2kg^{-1}min^{-1}$) vs. 45.8ml $O^2kg^{-1}min^{-1}$ (41.1 - 49.9ml $O^2kg^{-1}min^{-1}$) in control subjects, $p<0.01$. Furthermore, absolute and relative anaerobic thresholds were reduced in VSD patients, median 22.1ml $O^2kg^{-1}min^{-1}$ (17.5 - 25.9ml $O^2kg^{-1}min^{-1}$) and 60.0% (54.0 - 72.7%), respectively, vs. 33.5ml $O^2kg^{-1}min^{-1}$ (25.1 - 41.6ml $O^2kg^{-1}min^{-1}$) 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.

Abstract no: 157
Shades of bradycardia: An evaluation of the fetal heart rate across gestational age in long QT syndrome

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Introduction: Long QT syndrome (LQTS) is rarely recognised in utero although sinus bradycardia is a common manifestation. Normally, fetal heart rate (FHR) decreases as gestation progresses. The effects of gestational age on FHR in fetal 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 fetal bradycardia is gene-specific.
Methods: We reviewed FHR throughout gestation from obstetrical records of 42 paediatric patients with LQTS at 3 paediatric cardiac centres from 2002 - 2011. For comparison, we determined 3rd - 95th percentile range of FHR at 10 - 40 weeks (wks) from 547 normal fetuses. Bradycardia was defined as FHR ≤3rd

percentile for gestational age or ≤110bpm (peri-natal definition). We assessed best FHR predictor of LQTS. LQTS was confirmed by post-natal 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 (4), SCN5A (6), KCNE1 (2) and multiple (1); 3 had uncharacterised (de novo) mutations; and 3 were not tested. Bradycardia incidence depended on the definition: 15% of FHR readings were ≤110 bpm and 66% ≤3rd percentile. Confirmation of fetal LQTS resulted in diagnosing unsuspected LQTS in 3 families.
Conclusions: FHR varies widely in LQTS. A FHR ≤3rd percentile for gestational age increases the sensitivity to ascertain fetal LQTS compared to the FHR ≤110bpm 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 fetal bradycardia. These findings may improve detection of fetal and neonatal LQTS.

Abstract no: 160

Predictors of disease progression in paediatric dilated cardiomyopathy

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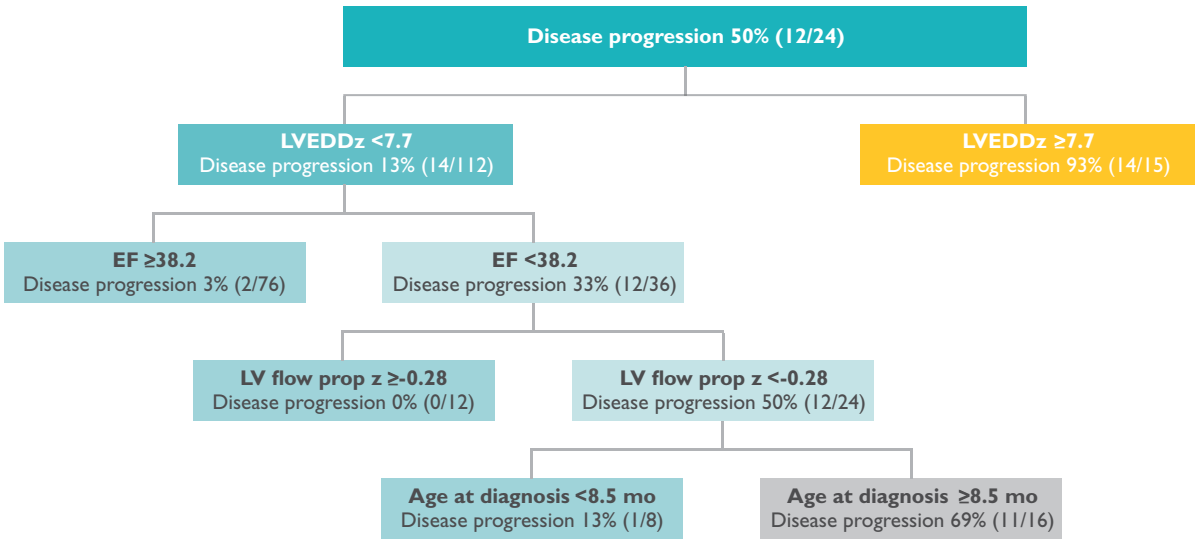
Background/hypothesis: Dilated cardiomyopathy (DCM) is the leading indication for heart transplantation in children. Identifying patients at risk for disease progression/transplantation remains elusive.
Materials and methods: The Paediatric Heart Network Ventricular Volume Variability Study evaluated 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 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 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 LVEDDz ≥7.7, LV ejection fraction <38.2%, LV inflow propagation velocity (colour m-mode) Z-score <-0.28, and age at diagnosis ≥8.5 months. (Figure 1.)

FIGURE 1: CART results



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.

Abstract no: 161

Physical activity recommendations in congenital and electrophysiological heart disease: A survey of Canadian health care 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. from July 2011 - December 2011, approximately 350 health care providers working with this group of patients were recruited by email 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, amongst 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.

Abstract no: 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 compare long term results of balloon valvuloplasty with new surgical methods such as valve shaving and plasty that provide promising short term results in selected newborns.

Material and methods: This is a retrospective follow-up study of all the 126 patients initially treated with 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 - 28 days (median 2 days) and the follow-up period reached 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 years 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 the 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 the 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 hazard of death.

Abstract no: 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 encouragements may be needed. Little is known of 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 1 year, centred on interactive use of mobile phone and Internet technology.

Hypothesis: We hypothesise that e-Health encouragements and interactive monitoring of intensive exercise over 1 year can improve physical fitness, physical activity and HRQoL.

Methods: 216 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. The 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 with new knowledge regarding how physical activity in adolescents with CHD can be increased and possibly co-morbidity reduced.

Abstract no: 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: From September 2003 - March 2012 53 consecutive newborns with PA and IVS were reviewed: 10 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 in 3 groups: no to mild RH hypoplasia (n=35), moderate hypoplasia (n=10) and severe in 8 patients. Thirty two neonates with PA-IVS underwent attempts of percutaneous balloon pulmonary valvoplasty as primary procedure that was successful in 28 patients. Mortality rate was 10.7%, there was no procedure-related death or major complications. Freedom from reintervention was 32%. Nineteen patients required 23 additional surgical procedures after BPVP, 10 of them in 10 days period after BPVP. Primary surgical procedure was performed in 21 patients (including 3 patients after unsuccessful BPVP). There was 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 (4/6), severe dysplasia of TV (2/2) ($p<0,001$). Five patients were refused treatment because of RVDCC (n=3) and severe dysplasia of the right ventricle (n=2). Out of 44 patients that survived 23 already had biventricular circulation; 4 are scheduled for biventricular repair, 4 patients are scheduled for 1½ ventricle circulation, and 8 are waiting for 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.

Abstract no: 188
Quantification of quality in congenital heart surgery

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Background: Estimation of quality in congenital heart surgery should reflect procedures complexity, achieved survival and observed post-operative unfavourable events. It should be quantified to facilitate bench-marking.
Methods: Procedures complexity was determined by Aristotle basic complexity score. Hospital and 30-days survival was applied. Surgical performance was estimated as a product of complexity score multiplied by achieved survival. Observed morbidity was calculated as score according to the methodology of Sata and co-authors (Eur J Cardiothorac Surg 2012; 41: 898-904). 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: 542 primary procedures, including 46 (8.5%) without cardio-pulmonary bypass, were evaluated. Total cavo-pulmonary 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 therefore 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 in congenital heart surgery for year 2011 was quantified at 7.64 - 2.26 = 5.38 points.
Conclusions: Such quality quantification adequately reflects the 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 congenital heart surgery programmes.

Abstract no: 200
Factors affecting growth from birth to Norwood discharge: Results from the single ventricle reconstruction trial

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Background/hypothesis: Growth failure after the Norwood procedure is a potentially modifiable risk factor for medical morbidity and neuro-developmental outcome. We sought to characterise growth patterns and to determine risk factors for poor growth between birth and Norwood discharge.
Materials and 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 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 2 shunt groups (p=.32). Growth faltering was present in 91%, with FTT in 37% at discharge. Independent risk factors associated with a decline in WAZ (R2=0.33) were male gender (p<.001), higher birth weight (p<.001), pre-Norwood enteral feeds (p=.01), longer cardiopulmonary support time (p=.03), and more ventilator (p=.04), and hospitalisation days (p<.001).
Conclusions: Nearly all infants discharged after the Norwood procedure in the SVR trial had growth faltering and >1/3 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. Sub-optimal growth was associated with longer intra-operative support and medical morbidity. Strategies to improve growth during the Norwood hospitalisation warrant further attention.

Abstract no: 203

Outcomes of congenital heart defects associated with 22q11 deletion in chromosomal microdeletion syndrome

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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: 102 cases of 22q11.2 deletion were reported during the study period, suggesting a prevalence of 1.2 per 10 000 total births (1 in 8 335 total births). Ninety five cases had both 22q11.2 deletion and at least 1 associated congenital heart defect (CHD) (93%). Interrupted aortic arch, ventricular septal defect, Tetralogy of Fallot, 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 fetal life; 13 babies were delivered live (72%), pregnancies were terminated in 4, and still birth occurred in 1. In 84 patients 22q11.2 deletions were confirmed postnatally. Eighty eight 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 8 died after birth, 7 in the 1st year of life, and one at age 3.8 years.
Conclusions: 93% of patients with 22q11.2 deletion exhibited a CHD. In spite of the ante-natal detection rate being low, survival rates were similar for all cases irrespective of the presence of CHD. Conotruncal abnormalities were the most prevalent heart defect. Most deaths occurred within the 1st year of life but beyond infancy survival was favourable.

Abstract no: 204

Mis-diagnosis of Bland-White-Garland syndrome: Report of 2 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. The syndrome may cause sudden death in infants and young people but in this case report we present 2 different types of presentation.
Case report: 1st case: A 3-year-old girl was diagnosed with dilated cardiomyopathy since infancy. Her electrocardiography showed prominent Q-wave in lateral leads. Dilated right coronary artery was shown by echocardiography. The **2nd case** was a girl with prolapsed mitral valve and chest pain but similar to the first case she had prominent Q-wave in lateral leads at her electrocardiography and dilated right coronary artery but without heart failure.
Conclusion: ALCAPA in children may present with ambiguous presentation from dilated cardiomyopathy and full-blown heart failure to an atypical chest pain attributed to prolapsed mitral valve.

Abstract no: 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. Liver stiffness 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 anaesthesia (Fi02=21%) using an Optitorque 5 French catheter. The patients underwent 10 LS measurements (median value taken as representative) by transient elastography (Fibroscan®, Echosens, France) 24 hours before catheterisation. The results of LS are expressed in kilo Pascals (kPa).

Results: Fourteen children (mean age=9±6 years old, 64% male) and 14 adults (mean age=34±17 years old, 66% male) have been included so far. Catheterism 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 assessment of a complex congenital heart defect (n=16). Mean right atrial pressure was 8.2±3.3mmHg and mean LS was 8.1±4.4kPa. 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.

Abstract no: 206
Is QRS axis pattern associated with the type of surgical repair in adults operated for 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 old). 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±17mL/m2. Mean QRS duration was 156±16ms 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°). 9 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 is correlated to the type of surgery.

Abstract no: 208
Incidence and natural history of innocent heart murmur in newborn babies

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Introduction: This study was aimed to determine the incidence and also define the origin and natural history of innocent heart murmur in the newborn.
Methods: In a 21-month prospective study, 2 849 newborn babies underwent routine examination by paediatricians. Infants with murmur were re-examined by paediatric cardiologists. Term babies with clinical diagnosis of an innocent heart murmur were studied. Each baby had a complete echocardiographic study and was followed up at 2 and 6 months until the murmur had disappeared or the heart was totally normal.
Results: Clinical 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 6 cases (20%), small patent ductus arteriosus (PDA) in 5 cases (17%), small ventricular septal defect (VSD) in 3 cases (10%), mild pulmonary valve stenosis in 1 case (3%) and isolated mild tricuspid regurgitation in 5 cases (17%). Only VSD and pulmonary valve stenosis were considered to be pathologic. Follow-up was performed at the age of 2 months: heart murmur had disappeared in 23 cases (82%), PDA and mild pulmonary stenosis had resolved in all patients but PPS was still present in 1 of 6 cases. At 6 months: murmur had disappeared in 26 cases (93%), 2 of the 3 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 6 months.

Abstract no: 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 poor resource countries remains poor when compared to the developed counterparts. There are still paucity of data on the severity and determinants of outcome in childhood cardiac failure in Nigeria.
Objectives: The aim of the study is 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: 220 consecutive cases of CCF admitted to paediatric wards and the children's emergency rooms of LUTH with diagnosis of heart failure over 1-year period was studied prospectively. Diagnosis of heart failure was based on the presence of at least 3 or 4 cardiac signs of heart failure. The severity of CCF was determined by using heart failure severity index score 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 till a definite outcome was determined.

Results: Predominate 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 which 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 readmission for heart failure within the study period (p<0.01).
Conclusion: Heart failure in Nigerian children is associated with an unacceptably high mortality. 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.

Abstract no: 222

Familial recurrence of congenital heart disease caused by mutation in NKX2-5

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Background: Familial recurrence presents in 3 - 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.
Materials and 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 are listed in Table 1.

TABLE 1: Classification of study population

Group	Title	# individuals
1.	Conotruncal	16
2.	Atrioventricular septal	7
3.	Anomalous pulmonary venous return	2
4.	Left ventricular outflow tract obstruction	20
5.	Right ventricular outflow tract obstruction	6
6.	Septal	49
7.	Heterotaxy	0
8.	Single ventricle/complex	3
9.	Septal + LVOTO	4
10.	Septal + RVOTO	1
11.	Other association	0
12.	Other related defects*	22
13.	Vascular ring	1
14.	CHD**	5
Total		**136**

*The patients were classified using the method published by Botto, et al.[5] Group 12-14 is added by the authors to complete the description of the study population. The classification has been done in a hierachial manner. *Other related defects: bicuspid aortic valve, valve insufficiencies, patent ductus arteriosus and arrhythmia/block. **CHD: confirmation of the diagnosis is pending.*

Results/discussion: One family had a single nucleotide deletion in exon 1 of NKX2-5 causing a frame shift. The family had 5 living (3 with ASD2, 1 with ASD2 and first-degree AV-block, 1 with muscular VSD and ASD) and 1 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 deaths have 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.

Abstract no: 223

Global strain rates are 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 (2-DST) 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) 2-DST and conductance catheter studies were performed 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) (rs=-0.4, p<0.01), but not with global strain (S) (rs=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 is not affected by preload and correlates 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.

Abstract no: 231

Current outcomes of post-operative extracorporeal membrane oxygenation support in children with functional single ventricle pathologies

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Background: Improved survival with post-operative 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 post-operative ECMO and compare results between children with SV versus bi-ventricle (BV) pathologies.
Methods: Demographics, anatomic, surgical and support details of children who received post-operative ECMO (2007-2012) were entered into multi-variable regression analysis to determine factors affecting survival.
Results: There were 95 patients, (3 days - 16 years), that were divided into SV (n=28) and BV (n=67) groups. ECMO was initiated in 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 reoperation while on ECMO. Forty patients (42%) survived >24 hour after ECMO discontinuation and 37 (39%) were discharged alive. Mean ECMO duration was 4.9±3.1 days (3.8 & 5.5 days in survivors vs. non-survivors, p=0.003). Survival for ECMO initiated in OR vs. ICU was 43%&40% (p=0.66), and was 38% & 39% for ECPR and non-ECPR patients (p=1.0). Survival of SV and BV patients was 32% & 42% (p=0.18). In SV group, outcomes diverged with best survival after BT shunt (60%) or Norwood (50%). On the other hand, survival was dismal following PA band, Glenn, Fontan, TAPVC in Heterotaxy patients (0%). On multi-variable analysis, cardiac reoperation 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 post-operative support including SV patients. Results in SV vary with Norwood and BT shunt having better prognosis. Timely ECMO prior to emergence of complications and surgical correction of residual lesions might improve survival.

Abstract no: 240

Incidence of isolated aortic dilation in patients with Turner's syndrome

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Background: 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: Retrospective study with data collected from medical records and echocardiography studies. Patients with TS seen at our centre from 1992 - 2010, free of structural cardiac malformations were eligible when they had ≥2 echocardiography. Patients with previous cardiac surgery were excluded. Age ranged from infancy to adulthood. Of 120 patients, 33 (27.5%) had 1 or > cardiac anomaly; 18 (15%) presented with bicuspid aortic valve, 14 (11.6%) with coarctation of the aorta, 7 (5.9%) with aortic valve stenosis and 3 (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 for TS patients. The impact of confounding factors such as genetic variants (mosaicism), growth hormone therapy and vasoactive medication is yet to be determined.

Abstract no: 242

Usefulness of pro-BNP as a marker for myopericarditis in the early diagnosis of Kawasaki disease

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Background: The early diagnosis of Kawasaki disease (KD) is sometimes difficult and it is critical to achieve the optimal treatment result, especially in incomplete or a-typical KD. We investigated the correlation of serum levels 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.
Materials and 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 2 categories and defined as follows; (1) Significant mitral valve regurgitations; and (2) Significant pericardial effusion.
Results: In the KD patients, significant mitral valve regurgitations were present in 20 patients (20.8%), significant pericardial effusion in 8 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 927.3pg/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 could be helpful in the diagnosis.

Abstract no: 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 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 hypothesise that early post-operative echocardiography will predict which patients are at higher risk of re-intervention for significant SVPS following ASO.
Methods: Single institution retrospective review of Manitoba newborns (n=63) who had ASO for D-TGA from 1991-2010 was undertaken (independent of when, where and who performed their surgery). First post-operative- and most recent trans-thoracic echocardiograms (TTE) of all surviving patients (n=59) were reviewed for SVPS. Patients were categorised as requiring re-intervention for SVPS ("re-intervention"-group) vs. those who did not ("no re-intervention"-group). Univariate analysis using Fisher's Exact Test was used to analyse parameters summarised in Table 1. Significant TTE parameters (gradients >40mmHg plus 2 or more levels of stenosis) were analysed using the Kaplan-Meier method to calculate probability of freedom from re-intervention.

TABLE 1: Summary of univariate analysis

Univariate variable	Re-intervention: SVPS (n=4)	No re-intervention (n=55)	P-values
Atrial Septostomy pre-ASO	4	52	0.8
d-TGA + VSD	4	17	0.013
Complex d-TGA	2	9	0.3
Pre-ASO diagnosis of RVOTO/PA hypoplasia	2	7	0.1
Time to ASO (mean days)	13.5	12.35	n/s
Post-operative TTE >2 locations of stenosis	4	6	0.0007
Post-operative TTE >40mmHg gradient (CW)	4	4	<0.0001
Both (>2 locations and >40mmHg)	4	2	<0.0001

Results: Mean follow-up period was 9.3 years. First post-operative TTE demonstrating 2 or more levels of stenosis, stenosis gradients >40mmHg, both last mentioned parameters combined, as well as D-TGA plus VSD, were all significantly more prevalent in the "re-intervention"-group (Table 1). Patients that had gradients >40mmHg and stenosis at 2 or more levels, the 5-year probability of freedom from re-intervention for SVPS was 40% compared to 100% for those without the 2 aforementioned parameters (log rank p=0.0001).
Conclusion: SVPS with multiple levels of stenosis causing a gradient >40mmHg at initial TTE post-ASO, allows 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.

Abstract no: 245

Randomised controlled trial of intravenous immune globulin in acute myocarditis in the 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. The paediatric literature is scant, and there are no randomized controlled studies of immunosuppressive therapy in childhood myocarditis. Hence, this study was designed to assess whether intravenous immune globulin (IVIG) improves left ventricular function and survival in children with acute myocarditis.
Materials and 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 (2g/kg) plus prednisolone (2mg/kg/day) or prednisolone (2mg/kg/day) alone for 6 weeks duration. Left ventricular function was assessed at 7 days, 1 month, 3 months, 6 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 9 (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 6 months and 12 months (p=0.01 and p=0.009 respectively). Left ventricular ejection fraction (LVEF) was also higher in IVIG group at 6 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 the groups.
Conclusion: Compared to steroids, IVIG is associated with significant improvement of left ventricular function in acute myocarditis at the end of 12 months but without significant difference in survival.

Abstract no: 246

Repair of total anomalous pulmonary venous connection in infancy: A single-centre experience in 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 at our institute from July 2006 - March 2012. There were 48 males and 21 females. Median age was 89 days and median weight 3.77kg. 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. 42% deaths were caused by pre-operative infections.

Type	Number (%)	Mortality (%)	Obstructed (%)
Supracardiac	34 (50%)	5 (14%)	13 (38%)
Intracardiac	18 (26%)	0	6
Mixed	6 (8%)	3 (50%)	3 (50%)
Infracardiac	11(16%)	6 (54%)	8 (72%)
Total	69	14 (20%)	30 (43%)

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 >2 days significantly reduced the mortality (p=0.49). 40% had delayed sternal closure.

Operative data	Overall	Supracardiac	Intracardiac	Mixed	Infracardiac
Bypass time	84.6 minutes	91.8	68.7	92.8	89.6
Cross-clamp time	28 minutes	30.5	22.3	38.8	28.2
DHCA (75%)		18.4	7.3	23.1	12.2

Mean ventilation was 109 hours. 98.5% 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 post-operative infections. 42% deaths were due to delay in diagnosis and pre-operative infections, and 14% to pulmonary hypertensive crisis. There was 1 late death. In a follow-up of 3 months to 6 years (n=53), no patient has residual pulmonary hypertension.
Conclusion: Delay in diagnosis and surgery and rampant use of broad spectrum antibiotics still contribute to the high mortality of patients with treatable CHD and a good long term prognosis

Abstract no: 250

Changes in plasma hydrogen sulfide and its 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 following coronary artery lesions.
Method: A prospective unmatched case-control study was designed. 48 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 sulfide (H2S) was examined by sulfide-sensitive electrode method.
Results: Plasma H2S levels in KD patients during acute period were significantly lower than those during convalescent period (34.37±8.11 vs. 38.17±8.63, $p<0.05$). Plasma H2S levels in KD patients during 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 H2S and C-reactive protein, and between the plasma level of H2S 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: 44.705μmol/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: 43.78μmol/L).
Conclusion: Plasma H2S level in acute period might be a potentially useful biomarker for assisting the diagnosis of KD and predicting coronary lesions.

Abstract no: 252

Children suffering from postural orthostatic tachycardia syndrome with a marked increase in erythrocytic hydrogen sulfide 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 sulfide plays an important role in the pathogenesis of POTS. The present study was to explore the predictive value of erythrocytic hydrogen sulfide in predicting the therapeutic efficacy of midodrine hydrochloride for children with POTS.
Methods: Sixty eight children were included in the study, of whom 28 children suffered from POTS (POTS group) and 40 healthy children served as control group. Children in POTS group received midodrine hydrochloride treatment. The erythrocyte hydrogen sulfide production was measured by sensitive sulphur electrode and a receiver operating characteristic (ROC) curve was used to test if erythrocyte hydrogen sulfide could predict the therapeutic response to midodrine hydrochloride for children with POTS.
Results: Hydrogen sulfide production from erythrocyte was significantly higher in children with POTS than control subjects ($p<0.001$). Erythrocyte hydrogen sulfide production in responders to midodrine hydrochloride was significantly higher than that in non-responders ($p<0.05$). ROC curve revealed that the area under curve was 0.857 with a 95% confidence interval (CI) of 0.715 - 0.999. Erythrocytic hydrogen sulfide 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 sulfide could serve as a useful predictor of therapeutic response to midodrine hydrochloride in POTS of children.

Abstract no: 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 unfavorable, is also still unclear. In this study, we characterised 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.
Material and 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: Variable correlates with CI are shown in Table 1. 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/dtmax and normalized heart rate, and was strongly predictive of CI (R2=0.877). Table 2 shows variable correlates with TCPC-EDI. Ventricular volumes were all negatively correlated with TCPC-EDI; EDV had the strongest relationship. Q_s did not significantly correlate with any investigated variables.

TABLE 1: Correlates with cardiac index

Parameter	Correlation (p)	p-value	Multivariate oefficient	Multivariate p-value
Age (years)	-0.25	0.09		
BSA (m^2)	-0.22	0.17		
TCPC-EDI	-0.17	NS		
ESV ($mL/BSA^{1.3}$)	0.12	NS		
EF (%)	0.30	0.061		
ln(EDV) ($mL/BSA^{1.3}$)	0.43*	0.006	2.7	<0.001
ln(SV) ($mL/ BSA^{1.3}$)	0.58*	<0.001		
Sys. dV/dt_{max} (EDV/s)	0.40*	0.011	0.71	<0.001
Dias. dV/dtmax (EDV/s)	0.43*	0.006		
HR ($bpm/BSA^{0.4}$)	0.25	0.12	0.027	<0.001

*SV: stroke volume. HR: heart rate. NS: not significant. *: p<0.05.*

TABLE 2: Correlates with TCPC power loss (TCPC-EDI)

Parameter	Correlation (p)	p-value
Age (years)	0.15	NS
BSA (m^2)	0.22	0.18
Q_s ($L/min/m^2$)	-0.24	0.14
ESV ($mL/BSA^{1.3}$)	-0.36*	0.021
EF (%)	0.06	NS
EDV ($mL/BSA^{1.3}$)	-0.52*	0.001
SV ($mL/BSA^{1.3}$)	-0.42*	0.008
Sys. dV/dt_{max} (EDV/s)	-0.07	NS
Dias. dV/dt_{max} (EDV/s)	-0.09	NS
HR ($bpm/BSA^{0.4}$)	0.28	0.09

**: p<0.05.*

Conclusions: EDV (preload), BSA-normalised HR, and systolic dV/dt_{max} are independent predictors of CI in single ventricles. TCPC power loss is inversely related with ventricular volumes, particularly preload.

Abstract no: 256

Cardiovascular profiles 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 being 8 years (4½ months - 16 years) (n=43). The mean GFR was 23.22ml/1.73m^2/min. Malnutrition was universal in all stages with reflux nephropathy as the commonest etiology. 9/41 patients with LVH (LV Mass Index >38g/$m^{2.7}$) and 8/39 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 value 0.03). Patients with ESRD had higher mean CIMT.Values (0.14cm, p value=0.03) than other stages. 57% of patients with uncontrolled systolic hypertension and 43% with uncontrolled diastolic hypertension had LV dysfunction (p value 0.04). Significantly higher CIMT was noted in those with uncontrolled systolic and diastolic hypertension (0.15cm), which persisted even after control. Children with LVH had significantly higher mean CIMT (0.124cm±0.006) than those without (p value <0.0001). Vitamin D intake was associated with better LV functions (p value 0.03).
Conclusion: Cardiovascular disease is mainly subclinical and noted in all stages of CKD with large vessel disease worsening exponentially with ESRD. Eccentric LV hypertrophy with hypertension is an important risk factor contributing to LV dysfunction and persistent large vessel disease. Vitamin D therapy has a positive impact on global LV functions.

Abstract no: 261

Senning's 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 little 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%) surgical surviving patients, submitted to Senning's operation, with a mean follow-up time of 14.7±3.1 years. Electrocardiogram, echocardiogram and chest radiograph series were performed every 6 months. Thirty six patients of this series underwent Holter study and ergometric testing to evaluate the physical capacity. Three patients living overseas were excluded.
Results: There was only 1 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 39 surviving patients was 77.1% (30) or normal right ventricular function was 76.5% (29), 10 - 20 years after operation. The incidence of sinus node and right ventricular dysfunction increased gradually over time. No reoperations and pacemaker implantations were performed. Functional Class I=30 (85.7%) cases and Functional Class II=5 (14.3%) cases.
Conclusions: In our experience, patients with simple TGA submitted to Senning's procedure, presented with the following during late follow-up:

- Low incidence of right ventricular dysfunction and active arrhythmias;
- Low mortality and no sudden deaths were recorded;
- Good quality of life; and
- Satisfactory surgical results (free of reoperation or definitive pacemaker implantation).

Abstract no: 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 etiology and management have yet to be determined. We hypothesise that aortic dilation occurs in children with CHB and that it will regress following the implantation of a permanent pacemaker. We will also attempt to identify an association between the presence of maternal antibodies and the degree of aortic dilation.
Method: Retrospective review of charts and echocardiograms of patients with CHB, followed at our institution over the past 27 years. Comparison of the target structures diameter is based on Z-scores derived from normal populations and dilation is defined as Z-score ≥ 2.
Results: Seventeen patients met inclusion criteria in our preliminary review. Mean age at diagnosis was 5.2 months and 53% were males. Positive maternal antibodies were found in 5 cases, 2 were negative and 10 were unknown. All the patients underwent pacemaker implantation (mean age of 17 months). Aortic dilatation was found in 7 (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, 4 patients amongst the 7 had positive maternal antibodies (the 3 others having an unknown serologic status).
Conclusion: Although those are preliminary data, ascending aortic dilatation seems to be present in a large proportion of patients with congenital heart block. This associated aortic dilatation seems to be regressing after implantation of a pacemaker. The mechanism involved in this phenomenon might be related to the regularisation of stroke volume. Positive serologic 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.

Abstract no: 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 1 in 3500 boys. Cardiomyopathy is a major source of morbidity and mortality. While steroid therapy slows musculoskeletal impairment, 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, 7 of 63 (11%) patients on steroids died compared to 10 of 23

(43%) without steroids (p=0.0010). Survival rates at 5, 10, and 15 years of follow-up were 100%, 98% and 79% for patients with vs. 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. +0.92mm per year, p=0.0105).

Conclusion: In patients with Duchenne muscular dystrophy, steroid therapy is associated with a substantial reduction in all-cause mortality and new-onset and progressive cardiomyopathy.

Abstract no: 266

Assessment of atrial septal defects with real time 3-D transoesophageal echocardiography: A new insight into dynamic changes in the 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 3-dimensional transoesophageal echocardiography (R-T3-D TEE) may provide superior imaging assessment of ASD compared to conventional 2-dimensional TEE. The objectives of this study were to compare measurements of ASD size obtained by R-T3-D and 2-D TEE and to study the dynamic changes of ASD during the cardiac cycle.

Methods: R-T3-D and 2-D 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 axis were measured offline using Xcelera QLab software and compared with values obtained by 2-D imaging. Defect areas by 3-D planimetry were evaluated for changes during cardiac cycles.

Results: Out of 34 ASDs; 28 were oval, 4 were multiple and 2 were complex. R-T3-D TEE en face view enabled better appreciation of ASD shape and orientation in those with multiple and complex ASDs. There were high agreement between ASD diameters measured by R-T3-D and 2-D TEE along both major axis (19.6±5.5mm vs. 19.0±5.3mm, r=0.92, p<0.001) and minor axis (14.8±4.4mm vs. 14.8±4.1mm; r=0.93, p<0.001). There was significant change in ASD size during 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±3.1mm, 4.1±2.5mm and 0.69±1.8cm^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: R-T3-D TEE is highly accurate in assessing anatomy and size of ASD. It also provides new insight on the dynamic changes of ASDs' size and shape during the cardiac cycle.

Abstract no: 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 - 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 - 2007. Patients younger than 6years at latest follow-up and post-partum women were excluded. Baseline characteristics and most recent investigations were obtained. Consenting patients were prospectively reviewed with standardized 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); 4 of 15 patients were male. All patients were treated with corticosteroids and cyclosporine, 2 of 15 patients also received intravenous immunoglobulin post-diagnosis. Long-term follow-up occurred at median 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% have mild left ventricular dilatation 10 years post-illness. There was no evidence of diastolic dysfunction by routine echocardiographic assessment in our paediatric population.

Abstract no: 273

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 of children and adolescents.

Methods: Fifty seven consecutive children and adolescents hospitalised for syncope were collected from Peking University First Hospital from January 2008 - 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 radio-immuno-assay method. Logistic regression analysis was used to find out 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 orthostatic tachycardia syndrome and 2 micturition syncope), 10 (17.5%) had cardiac arrhythmias, 11 (19.3%) had structural cardiac/cardiopulmonary disease, and 2 (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 (1955.95±3322.24 vs. 31.01±23.59 and 36.83±25.63pg/ml, p=0.000). Whereas, 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±25.63pg/ml, p=0.991). At cut-off of 71.24pg/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 differential diagnosis of arrhythmic-cardiac syncope and non-cardiac syncope in children and adolescents.

Abstract no: 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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Background/Hypothesis: The International Children's Heart Foundation provides surgical care to children in developing countries and since 2010 has used the SIS-ECM (Cormatrix) for reconstruction and or repair of congenital heart defects in 174 patients. We report our experience with this material and describe its versatility.

Materials/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, 6 Tetralogy with absent pulmonary valve, 4 Truncus arteriosus, 4 Rastelli, 3 DORV with RV to PA conduit, 2 TOF redo RV to PA conduit), 39 primary tetralogy repairs, 34 valve leaflet repairs (included with primary repair), 65 septal patches, 7 arterial switch repairs, 5 AVSD repairs and 5 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 early required 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 reoperations.

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 term and mid term follow up. Complete freedom from calcification is an added benefit.

Abstract no: 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 score 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 2 - 19 years (mean 11.8±2.9 years). 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.

Abstract no: 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 post-operative 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 (Sata S, et al. Eur J Cardiothorac Surg 2012;41:898-904). Two groups of patients with similar age who underwent similar types of procedures and with the same Aristotle complexity score (8 points) from January 2006 - 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 the so-called unfavourable period (Full Moon and Moon in Leo). Patients in Group 2 (n=79) underwent surgery during presumed favourable moon phase (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, vs. 4.21±4.96 and 14.53±8.51 days for Group 2: p values=0.62 and 0.30, respectively. 57.3% (43/75) patients in Group 1 and 57.0% (45/79) in Group 2 had an uneventful post-operative 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: Results quality did not vary with moon phases. One should not worry at all about lunar phases when planning and performing congenital heart surgery.

Abstract no: 295
Permanent pacemaker in pregnancy: Analysis of 6 cases

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Background and aim: Outcome of pregnancy in patients with a pacemaker is not fully understood because of limited number of cases. We present 6 cases and discuss their management issues.
Methods: We conducted the retrospective analysis of 7 pregnancies in 6 patients with pacemaker implantation during 2008 - 2012 in our institution. The course of pregnancy, peri-natal outcome, maternal baseline disease and pacing mode were studied.
Results: All 6 patients had atrioventricular block AVB. Of these, 3 patients had structural cardiac lesion (single ventricle in 2 and atrio-ventricular septal defect in 1), and other patients had normal structure heart (cardiomyopathy in 1 and congenital AVB in 2). In 5 patients, implantation of the pacemaker was performed before pregnancy. The mode of pacemaker was DDD in 4 (transvenous lead), and VVI in 1 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 fetal death. Three patients had uneventful 4 vaginal deliveries. One patient had delivered a baby by Caesarean section for intrauterine growth retardation and non-reassuring fetal status. One patient who had single right ventricle with a pacing due to surgical AVB developed cardiac decompensation in the 3rd trimester, and was induced at 36-week gestation. One patient with VVI pacing system required to change the ventricular pacing rate due to the failure of the cardio-acceleration during the intra partum period.
Conclusion: There were no maternal or neonatal crucial complications. Outcome of pregnancy in patients with a pacemaker was uneventful due to multidisciplinary medical care.

Abstract no: 309
Longitudinal comparison of outcomes between pre- vs. post-natally diagnosed infants with transposition of the great arteries

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Background: Pre-natal diagnosis of transposition of the great arteries (TGA) may decrease peri-operative morbidity but little data exists regarding its long term impact or costs. Linking the Utah Birth Defects Network (UBDN, a state-wide surveillance system) and the Utah Population Database (UPDB, state-wide in-patient discharge abstracts and charges) one 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 pre-natally (predx) and post-natally (postdx) diagnosed TGA.

Methods: TGA cases identified at the UBDN (born from 1997 - 2009) were reviewed and pre-term gestation excluded. Linkages to the UPDB identified in-patient encounters and charges for cases (through December 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 generalised linear regression with gamma distribution and log link function, and in-patient encounters, using multivariate modelling, adjusting for demographic and clinical risk factors.
Results: Of 108 cases identified, 12 (11%) were pre-natally diagnosed. Predx were similar to postdx in gestational age (38.6 vs. 38.8 weeks), birth weight <2.5kg (0% vs. 4%, p=.62) and additional congenital defects (8% vs. 10%, p=.39). There were 7 deaths (6%) with no difference in survival between groups (mean follow-up 7.4 years). Predx cases had similar hospitalisations (median 2 in both groups), but higher total LOS (23 vs. 19 days, p<.004) as well as higher costs for both mothers ($8200 vs. $5200) and index cases ($290,000 vs. $151,000) after adjusting for gestational age, birth weight and additional congenital defects (p=.001).
Conclusions: Predx TGA cases had longer hospitalisations after adjusting for available risk factors. Both mothers and infants in the predx group had higher longitudinal inpatient medical costs. Ongoing investigation should explore specific cost components responsible including additional testing, surgical delays and other risk factors.

Abstract no: 311

Myocardial performance index (Tei index) in children and adolescents in 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 held at Hospital Infantil Joana de Gusmão, in Florianopolis-SC, Brazil. The left ventricular MPI (LVMPI) and right ventricular MPI (RVMPI) was measured in 121 healthy children and adolescents and in 7 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, in a minimum 30 day intervals between the measurements.
Results: Among the healthy individuals, the age ranged from 3 - 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 to LVMPI and moderate to RVMPI and inter-observer reproducibility was considered high to LVMPI and low to RVMPI.

Abstract no: 316

Arrhythmia phenotype during fetal life predicts LQTS mutation: Risk stratification of peri-natal Long QT syndrome

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Background: Torsades de Pointes (TdP) and/or 2° atrioventricular block (2° AVB) are signature rhythms for peri-natal LQTS known for their high morbidity and mortality. We hypothesised that the clinical profile of patients with these complex fetal arrhythmias might be genotype specific.
Methods: Peri-natal records of LQTS subjects exhibiting complex fetal arrhythmias were reviewed. Fetal echocardiograms, neo-natal ECG, and genetic testing were evaluated.
Results: We studied 11 LQTS subjects exhibiting complex fetal arrhythmias. Mutations in one of the 3 major LQTS-susceptibility genes were identified in 9: SCN5A (5), KCNH2 (2), and KCNQ1 (2). Most mutations were de novo including 4 with SCN5A-R1623Q. TdP occurred in 7 fetuses (mean gestational age=30.5 weeks) and 8 neonates; 2° AVB occurred in 4 fetuses/neonates. TdP exhibited 2 patterns: incessant and fast (>270bpm) or intermittent and slow (<250bpm). 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. Fetuses 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). Pre-natal treatment was administered in 6 cases without maternal complications; 4 fetuses improved and TdP ceased in 2. Despite medical and pacemaker therapy, cardiac arrest (n=6) resulting in sudden death (n=1) was common.
Conclusion: Complex rhythm phenotypes of fetal LQTS have genotype-suggestive features that, along with QTc duration, may risk stratify the peri-natal management of LQTS.

Abstract no: 321

Left axis deviation in the paediatric population: A modern study

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Background: Few recent and few large studies have investigated the associations of left axis deviation on electrocardiograms in children. Using the electronic medical record and the computer database of electrocardiograms within an integrated healthcare system, we documented these associations.
Methods: During a 5-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° - -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 atrioventricular septal defect (59), partial atrioventricular septal defect (22), tricuspid atresia (10), single ventricle arrangement (24), atrial septal defect (19), pulmonary stenosis (18), coarctation of the aorta (10), left ventricular hypertrophy/ cardiomyopathy (15), aortic stenosis (12), Tetralogy of Fallot (11) (majority post-operative), patent ductus arteriosus (5), transposition of the great arteries (5) and isolated anomalous pulmonary venous return (4). There were 57 with conduction abnormalities; Wolff-Parkinson-White syndrome (37), paced rhythm (17), Long QT syndrome (2) and complete heart block without a pacemaker (1). There were 10 patients with Noonan syndrome, 32 with short stature or other genetic abnormalities. 18 had scoliosis, pectus excavatum or dextrocardia.
Conclusions: This study documents the associations of left axis deviation in children and demonstrates the utility of using the electronic medical record within an integrated medical system to gather such data.

Abstract no: 326

Surgical and interventional management of patent ductus arteriosus in premature infants in Western Australia: A 9-year experience

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Background: There is significant controversy regarding optimum PDA management in preterm infants. KEMH is the sole tertiary peri-natal hospital caring for all infants born <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 <30 weeks in WA from January 2003 - 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 grams) received surgical or interventional closure at PMH during the study period. 11 infants had surgical closure during their initial neonatal hospital admission and 20 after discharge; 17 interventional and 3 surgical. All surgical and interventional procedures produced successful closure. Post surgery 3 had a pneumo thorax and one vocal cord palsy. One death occurred late and was 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.

Abstract no: 332

Nurse-led echocardiographic screening for rheumatic heart disease in Fiji: design of a training syllabus

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Background/hypothesis: 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 the training of 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 8-week syllabus, including extensive supervised field experience, would provide appropriate and adequate training for echocardiographic screening for RHD.

Methods: We designed an 8-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 7 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.

Abstract no: 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 7 of these patients later-on 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 control group over 6.6 months was 1.59±1.22 (-0.5; 4.0) mm. DDAR was significantly lower in patients under medication with BB ($p=0.056$) and AT ($p= 0.001$) compared to control group.
Conclusion: In this small and 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 seems to be more effective. These findings require confirmation in further studies with a prospective and randomised study design.

Abstract no: 342

The 6-minute walk 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 (VO2max) has been used as a prognostic marker to stratify adults with (severe) heart failure. In children VO2max may also have prognostic value, but it may be challenging to measure, especially at young age. Therefore, we determined whether the 6-minute walk test (6MWT) can 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 6 minutes, is easy to perform and provides a good reflection of daily life activity.
Methods: In a prospective longitudinal multi-centre 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 they 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 that reached one of the predefined primary endpoints, the distance covered during the 6MWT was significantly lower (Z-score -5.7 (±2.9)) in the 3 months before the endpoint was reached, as compared to those not reaching a primary endpoint (-2.2 (±2.1)) ($p < 0.05$).
Conclusion: The 6-minute walk 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 6-minute walk test may have prognostic value in children with DCM.

Abstract no: 344

Reduced health-related quality of life in children 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 significant 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 ($r2 = 0.5$-0.8; $p<0.01$). To a lesser degree we observed agreement between parents' and children's report on the psychosocial scales as "mental health" and "self esteem" ($r2 = 0.2$, $p<0.05$).
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.

Abstract no: 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 - 2010 with IE at 1 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 1033 IE cases, 663 had HD and 370 had NHD. Table 1 shows the differences between HD and NHD cohort. Factors associated with poor outcome in 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 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).

TABLE 1: The differences between HD and NHD cohort

	HD (n=663; 53%)	NHD (n=370; 30%)	p-value
Age (months)	86±76	116±72	0.0001
Staphylococcus	24%	38%	0.0001
Streptococcus	20%	13%	0.0029
Stroke	8%	15%	0.0009
Cardiac surgery performed	18%	10%	0.0004
Mortality	6.8%	3.5%	0.02
Hospitalisation days	31±44	27±27	0.76
Hospitalisation cost (median)	$140 655	$131 893	0.48

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; in addition, renal failure in the NHD and vasoactive medication use in the HD cohort were associated with poor outcome.

Abstract no: 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 amongst school aged children and to determine the burden of rheumatic heart disease in this Pacific Island nation.
Materials and 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 3 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 6 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 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.

Abstract no: 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.
Materials and methods: School-based screening of grade 3, 6 and 9 students at all 4 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, to support and expand the local RHD programme.
Results: Of the total of 2 374 school-aged children of Nauru, 462 pupils aged 6 to 15 underwent echocardiographic screening for RHD. 53% were female. Seven cases of definite RHD were identified including 2 previously known cases; a prevalence of 15.1/1,000. Five had mild disease, 1 moderate and 1 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.

Abstract no: 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 frequently have been detected before birth, and they are the main group of the population of neonatal Ebstein's anomaly. We performed this study to find the outcome variables in current population of neonatal Ebstein's anomaly.
Materials and Methods: The records of 59 patients with neonatal Ebstein's anomaly who managed in Asan Medical Center 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 were died during the neonatal period. One year and 5 year survival rates were 78.6% and 76.3%, respectively. Variables found to be related to the time to death on univariate analysis were fetal 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 fetal distress (p=0.004) and pulmonary stenosis / atresia (p<0.001) remained significant.
Conclusion: Fetuses in whom Ebstein's anomaly is diagnosed should be closely monitored throughout pregnancy, especially those with pulmonary obstruction. A strict cooperation between obstetrician, neonatologist, pediatric cardiologist, and pediatric cardiac surgeon is essential for their survival.

Abstract no: 368

Secondary QT prolongation due to external force to the precordial region may induce commotio cordis

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Background: Commotio cordis (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 a thoraco abdominal region. We studied the clinical, electrocardiographical and laboratory features to clarify whether there are any relationship between CC and secondary LQT.
Patients and methods: We reviewed the charts and ECGs of 10 patients (two girls and 8 boys, 6 - 15 years) who showed LQT in the first ECG following blunt trauma against chest and abdomen, and analysed their clinical backgrounds, laboratory data and ECGs.
Results: (1) Clinical features: The causes for strong shock were as following; A traffic accident (4 patients), a collision in sports (3 patients), and a fall (3 patients). 5 patients suffered from liver, spleen and intestinal tract damages, 3 patients showed a skull fracture, and 1 exhibited haemo-pneumothorax, and another showed severe dyspnoea. (2) ECG findings: The first ECG on arrival disclosed long QTc (442 - 503) and 2 peaks at 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. (3) Laboratory data: Laboratory studies on arrival showed increased blood glucose concentrations (132 - 222) and decreased potassium levels (2.8 - 3.9), both which subsequently normalised.
Conclusions: CC usually results from VF following the mechanical stimulus added to the precordial region. The excited sympathetic nervous system at a severe shock induces over adrenaline secretion. This condition could enlarge an intra and extra cellular gradient of potassium ion, which results in LQT. Children following blunt trauma against chest might incur sudden cardiac death from TdP due to secondary LQT.

Abstract no: 374

Prognostic implications of 2-D, 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 pts at baseline and follow-up. iPAH patients alive (Group 1) were compared with those dead/transplanted (Group 2) at time of study. Cut points predictive of survival were generated (ROC) and Kaplan Meier survival 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 Group1 and Group 2 at presentation. However, at follow-up, despite similar pulmonary artery pressures and PVRi, RV function as shown in Table 1 were significantly worse in Group 2. A small pericardial effusion was seen in 3 patients in Group 2 vs. 0 in Group 1. RV function was significantly decreased in iPAH vs. cPAH patients during follow-up (Table 2). In iPAH survival was significantly different based on RV function cut points (RV fractional area changes <15.5%, RV end-systolic area >18.4cm^2/m^2, RV end diastolic diameter cm Z-score >4.8 and TAPSE Z-score <-4.3).

TABLE 1

	1st Echo (at diagnosis)				Last follow-up Echo			
	All patients	Group 1 (n=12)	Group 2 (n=6)	p-value	All patients	Group 1 (n=11)	Group 2 (n=6)	p-value
Heart rate (bpm)	95±23	92±21	103±29	0.568	95±22.3	79±20.5	103±20.4	0.062
RV systolic pressure (mmHg)	94±30	99.6±32	82.8±30	0.522	82±27.8	92.1±26.8	92.8±20.6	0.950
RV subjective function	Good	13				14		
	Mildly reduced	1	4				1	
	Mod reduced		1				2	
	Severe reduced		1				3	
RA volume (ml/m^2)	37±13	32.8±8	48±19	0.082	52±36	33.7±10.8	86±46	0.005
RV end-diastolic area (cm^2/m^2)	24.5±8	23.1±3	27.5±13	0.815	26.5±13	20.9±2.6	36.6±19	0.036
RV fractional area change (%)	19±6	19±5.4	17±7.6	0.756	21±0.09	24.5±8	14.2±6	0.009
TAPSE (cm)	1.31±0.4	1.4±0.37	1.12±0.45	0.258	1.5±0.59	1.81±0.5	0.97±0.37	0.008
TAPSE	-4.69±2.48	-4.29±2	-5.5±3	0.482	-	-	-6.7±0.89	0.002
Z-score					3.75±3.2	2.13±2.4		
LV end systolic eccentric index	2.3±0.67	2.1±0.64	2.7±0.68	0.129	1.55±0.42	2.04±1.12	2.6±0.86	0.093
LV end diastolic eccentric index	1.6±0.33	1.64±0.33	1.6±0.33	0.859	2.26±1.02	1.42±0.34	1.78±0.51	0.190
Pericardial effusion		None	None				None	n = 3

TABLE 2

Variable	1st Echo (at diagnosis)			Last follow-up Echo		
	iPAH (n=18)	cPAH (n=36)	P*	iPAH (n=17)	cPAH (n=31)	P*
RV systolic pressure mmHg	85±17	71±3	0.024	83±16	60±24	0.025
RV end-diastolic area cm^2/m^2	26±10.6	21±5	0.074	27±13	17.6±8	0.01
RV end-systolic area cm^2/m^2	21±7.4	14.8±4.4	0.001	24±15	13.3±5.3	0.002
RVED Z-score	4.2±1.9	2.7±1.6	0.027	5±2.4	2±2.1	0.001
RV fractional area change (%)	19±6	30±7	0.0001	21.1±9	30±10	0.003
RA volume index	39±15	35±20	0.184	60±42	35±20	0.016
TAPSE Z-score	-4.7±2.4	-3.4±2.5	0.044	-3.6±3.4	-4.2±2.1	0.442
LV end-systolic eccentric index	2.3±0.7	1.9±1.8	0.002	2.4±1.1	1.4±0.33	0.003
LV end-diastolic eccentric index	1.7±0.37	1.3±0.26	0.01	1.6±0.5	1.3±0.19	0.012

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.

Abstract no: 375

Arterial switch operation: Timing of surgery based on neuro-imaging

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Objectives: We examined relation between duration of pre-operative hypoxemia and prevalence of post-operative brain injury in neonates with TGA which were managed with 2 different strategies.

Materials and methods: From September 2009 - March 2012 93 newborns with TGA underwent brain MRI. Patients were divided into 2 groups according to ASO time. Group A includes 35 patients that underwent surgical repair in first hours of life (4±2 hours). MRI was performed pre-operatively in patients aged 3.6±1.9 hours, 30 of them have got post-operative MRI in 10.8±3.2 days of age. Group B includes 58 patients with conventional surgical approach that

underwent ASO at age of 8.1±4.0 days in 43 BAS was performed prior surgery. All of them had pre-operative MRI in age 6.6±4.0 days. Correlation between MRI findings and systemic arterial oxygen saturation was analysed.
Results: Parenchymal brain damage was diagnosed in 40% of patients (n=12) from Group A in comparison to 57% (n=33) from Group B. Post-operative 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 correlation in risk of brain injury with level and duration of systemic hypoxemia. In patients with WMI the average value of SaO2 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, without stroke 2.5±1.5 days.
Conclusion: Prolonged systemic hypoxemia is associated with higher risk of brain injury. Our brain MRI-based research shows that early TGA repair allows prevention of brain injury associated with prolonged systemic hypoxemia.

Abstract no: 378

Correlation between QRS duration, pulmonary artery regurgitation and right ventricle performance in totally corrected Tetralogy of Fallot patients

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Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart diseases with a good prognosis when totally corrected. Despite this progress, pulmonary artery regurgitation and progressive disorders of right ventricle impress its long term prognosis after corrective surgery by 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 recoursed to paediatric cardiology clinic in children's medical centre of Tehran. Thirteen leads ECG was used to calculate QRS duration and continuous Doppler echocardiography used to calculate pulmonary artery regurgitation (total time of pulmonary artery insufficiency/diastolic time) while the Tei index was used to measure performance of right ventricle. QRS duration was measured in milliseconds (ms) units in D2 lead and categorised into 3 groups: 1st group: patients with normal QRS and QRS duration <120ms; 2nd group: patients with long QRS and 120≤ duration <160. 3rd group: patients with QRS duration ≥160. Pulmonary artery regurgitation was measured in percentage and its index (PRi) divided in 2 groups: Mild regurgitation as PRi ≥70% and severe regurgitation as PRi <70%.
Results: Twenty one of 57 patients were in 1st group (36.8%). Amongst 36 patients with long QRS, 24 patients (42.1%) were in 2nd group and 12 patients (21.1%) were in 3rd group with a QRS duration >160. Only 1 patient had a QRS duration >180. PRi in 1st group was 80±12% and PRi in 2nd and 3rd group was 75±8.8 and 63±15 respectively making the difference (p<0.001) significant. Right ventricular function (RV MPI) in 1st group was 0.27±0.13; in 2nd and 3rd group 0.32±0.12 and 0.34±0.15 respectively and the increment amongst the 3 groups was not significant (p=0.143).
Conclusion: Increment in QRS duration more than 120ms in totally corrected ToF patients can be relevant to indicate increment of pulmonary artery regurgitation.

Abstract no: 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.
Materials and methods: Retrospective case note review.
Results: A 7-month-old Nigerian male infant presented at the Children outpatient clinic with complaints of recurrent cough and bluish discoloration 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 7.6kg. 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 lady. There was no history of exanthomatous febrile illness, ingestion of herbal concoctions or exposure to irradiation in pregnancy. No history of hypertension or heart disease in the family. His birth weight was 3.2kg. 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.

Abstract no: 386

Patients with unbalanced atrioventricular septal defect and a smaller left ventricle: What is the outcome?

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Background: Borderline unbalanced atrioventricular canal defect and borderline small left ventricle (uCAVC-smallLV) is characterised by atrioventricular valve index (AVVI) 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 vs. univentricular palliation.

Patients: From 1992 - 2012, 18 patients (pts) (8M/10F) with uCAVC-smallLV underwent surgery; 7pts (Group 1) had univentricular palliation due to associated atrioventricular valve (AV) malformations and/or big ventricular septal defect. In 11pts (Group 2) 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 vs. univentricular palliation were evaluated.

Results: Group 1/Group 2 at time of surgery (SURG): median age 8.9/2.7 months (p=0.01). 2 early deaths were present in: Group 1 on post-operative 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 prospering according to patients' weight: all were under 3 percentile at SURG, not changing during FU in Group 1 (p=0.2), but reaching normal weight 5 years after operation in Group 2 (p=0.042). More than mild AV/mitral valve regurgitation in Group 1/Group 2 were present: in 2 (28.6%)/4 (36.4%) patients (p=0.07). Late re-operations for severe regurgitation was performed in 2 patients, with no difference between Group 1/Group 2 (p=0.94): AV valvuloplasty in 1 (14.3%) vs. mitral valvuloplasty in 1 (9.3%), 2.8/4.6 years post-operatively. 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-smallLV biventricular repair should be used. During follow-up these patient are generally doing better; and mitral valve is reaching normal values. On the other hand, mitral as well as AV valve regurgitation may represent a severe problem after both types of correction.

Abstract no: 388

Left ventricular twist in children: Impact of age on torsion in children

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Background: The recently introduced method, speckle tracking echocardiography, represents simplified, objective and angle-independent modality for quantification of regional myocardial deformation. As published, there was no significant change in LV torsion with ageing and there might be some difference in LV rotation at base and apex.

Objective: The purpose of this study was to assess the relationship of LV rotation for torsion twist with ageing in children.

Methods: Forty healthy children were recruited and divided into 2 groups of 20 preschool age (2 - 6 years of age) and 20 school age children (7 - 12 years of age). After obtaining conventional echocardiographic data, apical and basal short axis rotations were assessed with speckle tracking echocardiography. LV rotations in basal and apical short axis planes were determined of 6 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 certain trend between 2 age groups in each basal and apical rotation. In basal and apical rotation, the values of preschool age children are greater than those of school age children at antero septal, anterior, lateral, posterior, inferior and septal in all 6 segments.

Conclusion: There was some trend of higher rotation value in preschool age children rather than school age children, with decrease rotation and torsion value in ageing during childhood from 2 - 14 years old. Although there was no statistically significant age-related change in LV torsion from rotation data, the lower trend with aging for rotation and torsion twist during childhood should be necessary for further investigation.

Abstract no: 393

A feasibility study of left ventricular rotation and torsion by 2-D 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: 14 CON (median age 11.1 years) and 5 PT (median age 14.8 years) exercised to volitional fatigue. 2-D echo basal and apical short axis views were obtained at rest, at each stage of SSCE, immediately - and 3 minutes post-exercise. R and T were obtained by standard techniques. Each variable was measured 3 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 CVs were as high as 50%.

	Basal		Apical		Torsion	
	CON	PT	CON	PT	CON	PT
Rest	-6.2	-5.3	10.6	10.4	15.0	12.2
Early exercise	-7.1	-7.2	11.3	7.3	16.5	10.8
Immediately post	-7.0	-5.6	9.9	10.4	16.0	11.6

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.

Abstract no: 394

Non-invasive assessments 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 (ELVI) to arterial elastance (EAI), 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 EAI:ELVI 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 6 trans-positions 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.

	CON (n=20)	Marfan (n=17)	TOF (n=17)	CoA (n=14)	TGA (n=6)
E_{AI}	2.38±0.49	2.44±0.83	2.59±0.64	1.91±0.36	2.16±0.76
E_{LVI}	5.07±0.11	4.07±1.45	5.97±2.56	4.91±2.43	3.35±0.47
E_{AI}/E_{LVI}	0.49±0.11	0.62±0.11	0.60±0.22	0.37±0.14	0.64±0.20
HE	0.82±0.04	0.84±0.03	0.85±0.03	0.83±0.03	0.81±0.03

E_{AI} was lower in CoA vs. ToF ($p=0.027$). E_{LVI} was higher in CoA vs. TGA ($p=0.048$). E_{AI}:E_{LVI} was lower in CoA vs. Marfan ($p<0.001$), TOF ($p=0.001$) and TGA ($p=0.005$). 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. 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.

Abstract no: 395

Lead removal in young patients with a congenital heart disease in view of lifelong pacing

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Background and 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 is evaluated retrospectively.
Methods and results: Over the last 6 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 3 leads the lead tips were retained, while 3 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 can 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.

Abstract no: 396
Tele-monitoring in children with a congenital heart disease and electronic devices

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Background and aim: In children and young adults self-perception and self-responsibility is not fully developed. An automated cardiac monitoring system can assist to diagnose clinical problems early or to anticipate device failure in the seriously affected patients with congenital heart diseases (CHD).
Method and population: Out of 200 patients 65 patients received a pacemaker (PM) or defibrillator (ICD) with the Home Monitoring (HM) option. Patients' age at implantation ranged from 5 weeks to 37.6 years [median 12.4 years]. The individual follow-up time from the daily monitoring data is 7 days up 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-physiologic studies, reprogramming of parameters, modifications in medication and sports or required further observation. 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 7 patients. 5 patients had 19 episodes with anti-tachycardiac 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 for 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.

Abstract no: 398
Norwood's procedure: A success story for the child or surgeon - and for how long?

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Introduction: The Norwood procedure is a series of 3 open-heart surgeries that gradually improve certain life-threatening forms of congenital heart disease. The 1st 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 3 surgeries is done at a different age, beginning from infancy and spanning through the toddler years. The 1st 2 surgeries (Stage I and Stage II) are used to temporarily relieve blood flow problems to and from the lungs. The 3rd surgery (Stage III) is used to further improve circulation. The Norwood procedure cannot cure the underlying heart defects.
Methods: A retrospective review of our unit's experience with more than 20 patients underwent the Norwood procedure between 1 January 2010 and 30 June 2012. We also reviewed a significant number of cases done abroad but had regular follow-ups done locally.
Results: Although the results of the modified Norwood procedure as palliation for hypoplastic left heart syndrome have improved considerably, in-hospital mortality remains high (up to 46%) while 1-year survival is only 1%.
Conclusion: Our study and supporting data from literature had showed that Norwood Procedure despite being life-saving for infants <1 week old with HLHS but has yet to give strong evidence for 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 the light of the overall results.

Abstract no: 399
The impact of congenital heart disease on outcomes in 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 in children with EA/TEF in the current era.
Materials and methods: A retrospective review of patients with EA/TEF treated at 2 academic institutions from 1995 - 2011 was performed. The presence of cardiac defects, other anomalies, surgical intervention, and outcomes were recorded.
Results: Among 231 babies with EA, 117 (51%) had CHD. Neonates with EA/TEF and CHD had a 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 systemic/pulmonary veins, Tetralogy of Fallot, aortic arch anomalies, atrioventricular 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, though CHD

neonates had 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, though mortality was higher in the presence of CHD. Our data indicate that CHD was not directly responsible for death and mortality may therefore be multi-factorial. 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 good outcome in the majority.

Abstract no: 403
Silver dressings for sternotomy incision care in paediatric cardiac patients to decrease surgical site infections

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Background/hypothesis: 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.
Materials and methods: Institutional review board approval was obtained to examine the infection prevention 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 - 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 5 days post-operatively or until discharge. The ASEPSIS wound score is a validated wound assessment tool; it was utilised to assess for wound infections. The study team supervised all dressing changes during the study period. 30-day follow-up for SSIs was performed. This randomised controlled trial evaluated silver-impregnated dressing vs. our standard dressings on SSI rates.
Results: There were no SSIs in either study group. We found that there were no differences in type of dressing utilized 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.

Abstract no: 404
Myocarditis in children: A 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: 44.7% were <1 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 >5 days in 85%. Tachycardia, gallop, bradycardia, hypotension, hyper dynamic precordium and cardiogenic shock were the presenting signs. Fulminant myocarditis was diagnosed in 5 (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. 2-D 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 8 (21%) requiring immune-modulators. Hospitalisation ranged from <5days in 7.8% to >15 days in 2%. CCF was difficult to treat in 5 (13.1%). Two patients died during the acute phase and 4 in sub-acute phases of illness. On 3 month follow-up, those with fulminant myocarditis had earlier normalisation of LV functions than those with acute myocarditis (p= 0.048). Twenty six children (68.4%) had complete recovery with 6 (15.7%) recovering partially.
Conclusion: Acute myocarditis often presents with non-cardiac symptoms, thus delaying clinical diagnosis. Fulminant myocarditis has better long term outcome than acute myocarditis.

Abstract no: 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 2-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 was assessed via a separate questionnaire.
Results: Male:Female ratio was 2.1:1. Mean age was 9.5 years with 2/69, being <3 years. Over-crowding was noted in 62.3% and 80% belonged to the lower socio-economic class. Carditis (78.2%), poly-arthritis (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 to 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 2-D 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 6 patients requiring valve surgery. Compliance to 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 does hamper scholastic performance, increase economic burden and lower the self-esteem of the child.

Abstract no: 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. 2-D 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 4-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 3 stages were analysed using 1-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.

Abstract no: 416

Protein-losing enteropathy after Fontan operation: Gastro-intestinal 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. Sulphated glycosaminoglycans (s-GAG) play an important role in maintaining the integrity of the intestinal mucosa. Congenital deficiency of intestinal s-GAG, and acquired deficiency in disorders of glycosylation, leads 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, immuno-histochemistry s-GAG staining was performed on tissue specimens from duodenum and terminal ileum.
Results: Age at study was 15.8+5.6 years; albumin was 2.8+0.9gm/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 4 (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 amongst 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 sulphate).

Abstract no: 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 the ventricles and inter ventricular septum (IVS), is not reported much in literature.
Aim: To analyse and evolve the echocardiographic (TTE) diagnostic criteria for "apical non-compaction".
Material and results: From January 2011 - July 2012, 60 consecutive cases that fulfilled standard echocardiographic criteria for LVNC and the additional criteria (1) Swiss cheese appearance of IVS, looking like a delta of the river; and (2) Non-compaction of right ventricle (RV) on TTE formed the study material. The diagnosis of ANC was totally missed by the echo-cardiographer and either only specific lesions or Swiss cheese VSD were diagnosed. The diagnosis was made after review. Age ranged from 3 days - 35 years, with 36 males and 24 females. The associated lesions were present in all 60 cases. Fifty two patients had acyanotic heart disease (86.7%) and 8 patients had cyanotic heart disease (13.3%). 16 of 60 cases had pump failure (26.7%). 7 cases had LV dysfunction, 7 had RV dysfunction and 2 had biventricular dysfunction, 28 patients (46.7%) had pulmonary hypertension; 2 (3.3%) had thrombus. 3 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 human beings.
Conclusion: For the first time in the world we are presenting the largest series of ANC. ANC is probably being ignored. ANC is invariably associated with other serious congenital cardiac malformations, which worsen pump failure.

Abstract no: 424

Impact of the peri-natal 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 the fetal to post-natal 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 fetus. Little data exists regarding the peri-natal transition in congenital heart disease. We sought to explore the impact of the peri-natal transition on myocardial function and cerebral blood flow in hypoplastic left heart syndrome (HLHS).
Method: Pregnancies with fetal HLHS were prospectively enrolled (n=8). Echocardiography was longitudinally performed at pre-natal (38±0.5 weeks) and post-natal (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 fetal HLHS was 416±86ml/kg/min and did not change at 4 - 12hours after birth (414±49ml/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±111ml/kg/min, respectively) due to increases in both SV and more so HR ($p<0.05$). CO correlated positively with PaO2 ($r=0.39$, $p<0.05$). During the same period, MCA-PI progressively increased from the fetal stage through all time points ($p<0.05$).
Conclusion: In HLHS, in contrast to the normal peri-natal transition, CO progressively increases from levels comparable to the fetus at 4 - 12 hours, to >150% by 3 - 5 days. This is likely a secondary to decreasing pulmonary vascular resistance in the presence of patent ductus arteriosus. Changes in pulmonary vascular resistance may also contribute to increasing MCA-PI. Ongoing investigations into the peri-natal transition in HLHS in comparison to normal neonates will provide further insight into how the myocardium adapts in HLHS to post-natal demands and its influence on the peripheral circulation.

Abstract no: 426

Brain abscess in cyanotic congenital heart disease: A 5-year review

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Background: Brain abscess in cyanotic congenital heart disease (CCHD) accounted for 5 - 10% of all kinds of brain abscesses in children. The prolonged uncorrected lesions, chronic hypoxemia and polycythemia were the most common of risk factors for brain abscess in CCHD.
Objective: To report the cases of brain abscess with CCHD in our institution.
Material and methods: Retrospective study of all patients admitted to our institution with the diagnosis of brain abscess and cyanotic congenital heart disease from July 2006 - July 2010.
Results: There were 9 patients ranging from 3 - 11 years of age, of which 6 males and 3 female. The clinical presentations were fever, vomiting, headache and seizure. Two patients developed hemi-paresis and 5 presented with seizures. The duration of fever before admission ranged from 11 days to 1 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 6 cases and solitary lesions in 3. The type of CCHD were Tetralogy of Fallot, complex cyanotic CHD with right isomerism, TGA and TAPVD. Burr-hole aspiration was performed in 5 cases: 1 case revealed Acinetobacter calco-acetricus, and 4 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: When cyanotic CCHD in children is not corrected or the repair thereof delayed after 2 years of age, these children can potentially develop brain abscess with poor outcome and prognosis.

Abstract no: 435

3-D speckle tracking echocardiography in the assessment of left ventricular volume and function in normal children: A comparison with cardiac MRI

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Background: 3-D speckle tracking echocardiography (3-DSTE) 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. 3-DSTE 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 9 years of age were investigated using CMRI and trans-thoracic echocardiography to assess LV function and volumes. 3-D images were acquired using a Phillips IE33 echocardiographic machine and a X7-2 probe, and these were stored for off-line analysis using Tomtec 4-D LV analysis software. Parameters derived from speckle tracking analysis included global longitudinal strain (GLS), torsion, EF, EDV and ESV which was correlated with CMRI derived indices.
Results: LVEDV derived from 3-DSTE correlated significantly with that derived from CMRI (63.4±11.6 vs. 72.2±11.7, $r=0.43$ $p<0.001$). These volumes differed significantly from 3-DSTE tending to underestimate LV volume. The LVESV derived from the 2 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. 46.67.8, $r=0.215$ $p=0.12$). EF derived from 3-DSTE 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 3-DSTE EF ($r=-0.38$ $p<0.005$). Torsion showed no correlation with CMRI derived parameters of function or volume.
Conclusion: 3-DSTE shows significant correlation with CMRI derived parameters of cardiac function and volume. 3-DSTE 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.

Abstract no: 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 33.9mm±7 (n=60) in the group with infant repair vs. 35.8mm±5 in the group with later repair (n=33) (p=0.17). Measurements were also compared in patients that initially required palliative procedures vs. those who did not (34.mm±7 (n=20) vs. 34.6mm±6 (n=71) p=0.77). Patients who had primary repair at <6 months did not show a significant difference to those performed later (34.8mm±6 (n=69) vs. 33.8mm±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 34.6mm±6 vs. 30.9mm±2.9 (p<0.001.)

Conclusion: This study confirms progressive aortic root dilatation 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 preventative therapy need further evaluation.

Abstract no: 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 68 children aged 9 years old had assessments of longitudinal myocardial deformation in the basal septal segment of the left ventricle using 2-D 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: Higher child's 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, whilst 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 ongoing follow-up.

Abstract no: 439

Impaired right ventricular contractile reserve late after surgical closure of isolated atrial septal defect

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Background and hypothesis: 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.

Materials and 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 160bpm. M-mode images and colour-coded tissue Doppler recordings from apical 4-chamber view were analysed offline.

Results: Patients had lower tricuspid annular peak systolic excursion (TAPSE) (14.2±3.1mm) at rest as 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±1.8cm/s, controls 9.7±1.6cm/s, p<0.001) and, as the highest measured S' during exercise (11.7±2.8cm/s vs. 15.3±2.7cm/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±0.6cm/sec^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±1.2cm/sec^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.

Abstract no: 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 6 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 >200µmol/l were excluded. Survival was determined telephonically at 6 months.
Results: Twenty eight per cent of the total population had LQTc. During follow-up 15 patients (4.7%) died suddenly, 73% of whom had 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±9.0mmHg vs. NQTc: 15.95±7.5mmHg; 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 6 months. QTc also strongly associates with left ventricular systolic and diastolic dysfunction, hypercholesterolaemia and a negative family history of IHD.

Abstract no: 441

Exercise capacity and quality of life in adolescents after surgical closure of atrial or ventricular septal defects during childhood

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Background and hypothesis: Patients with congenital heart disease generally report good quality of life but they also show reduced exercise capacity of varying degree 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.
Materials and 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 (VO2peak). 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 scale ranged from 0 - 100.
Results: The VO2peak 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 are shown in Table 1. By linear regression analysis we found no statistically significant relationship between the group wise Z-score of VO2peak and any of the subscale results or total score results from the quality of life assessment.

TABLE 1: Quality of life assessment and comparison results (NS = difference non significant)

Age < 18 years					
	Patients N=16		Controls N=51		p-values
Reported by	Self	Guardian	Self	Guardian	
Physical subscale	93.8±9.1	87.3±15.9	95.6±5.8	88.9±18.6	NS (0.831/0.365)**
	p=0.028*		NS (p=0.063)*		
Psychosocial subscale	93.1±8.0	84.9±14.6	89.3±9.7	87.5±16.8	NS (0.141/0.413)**
	p=0.027*		NS (p=0.551)*		
Total Score (PedsQL™)	93.3±7.4	85.7±14.1	91.1±7.3	88.0±16.6	NS (0.207/0.397)**
	p=0.017*		NS (p=0.756)*		

Adult			
	Patients N=16	Controls N=52	p-values
Physical subscale	73.9±22.4	80.0±13.9	NS (0.418)**
Mental subscale	75.5±16.4	77.7±15.0	NS (0.470)**
Total score (SF-36™)	77.7±18.2	82.0±13.6	NS (0.347)**

*Mann-Whitney U test. **Wilcoxon test.*

Conclusions: Adolescents operated for isolated heart septal defects during childhood reported normal quality of life compared to the healthy population. Patients <18 years of age reported better quality of life than estimated by their guardians. Our patients had lower aerobic exercise capacity compared to normal but we found no relationship between VO2peak and any of the reported subscale scores or total scores of quality of life for either the patient group or control group.

Abstract no: 452
Do clinically important racial differences in atrial septal defect morphology exist?

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Background: Sinus venosus defects are considered uncommon, representing only 5-10% of inter-atrial communications, with the inferior subtype being particularly rare and more difficult to diagnose. Suspecting and correctly identifying this unusual anatomy facilitates appropriate repair.
Material and methods: A retrospective review was undertaken of patients who underwent repair of inter-atrial communications at our Central American centre from October 2006 - 2012 to determine the anatomic subtype. Partial atrioventricular canal defects were excluded.
Results: Fifty four patients (53 children and 1 young adult, age range 14 months - 23 years) underwent surgery for inter-atrial communication. Sinus venosus defects with partial anomalous pulmonary venous connection were encountered in 10 patients (19%, p=0.025), with half of these (5 patients, 9.2% of the total operated) being inferior subtype. The remaining 44 patients (81%) had secundum atrial septal defects, 6 who (11%) had an additional diagnosis of pulmonary stenosis requiring valvotomy, and 1 who had a small patent ductus arteriosus. 1 patient (1.9%) died as a result of a ruptured oxygenator during bypass; the remaining patients had an uneventful post-operative course, spending 1 night in intensive care and being discharged at a median of 2 days post-operatively. All surviving patients had post-operative 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.

Abstract no: 455
Pre-natal diagnosis improved the post-natal cardiac function in a 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 peri-natal care and is known to be associated with more stabile pre-operative haemodynamics. The aim was to determine whether pre-natal diagnosis of HLHS has an impact on post-natal myocardial function.
Methods: We reviewed a consecutively encountered cohort of 66 HLHS infants born in Finland between 2003 and 2010. Post-natal global and segmental right ventricular fractional area change (FAC), strain rate (SR) and myocardial velocity (V) were analysed from apical 4-chamber view using Velocity-Vector- Imaging technique (Syngo USWP 3.0, Siemens). Intra- and interobserver 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 Norwood procedure.
Results: 25 infants (38%) had a pre-natal diagnosis. Pre-natally 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 pre-natally had less acidosis (pH 7.30 vs. 7.25, p=0.005) and end-organ dysfunction (alanine aminotransferase 33±38 vs. 139±174U/l, p=0.0001; creatinine 78±18 vs. 81±44mmol/l, p=0.05). No deaths occurred among the pre-natally diagnosed infants but 4 deaths were recorded among post-natally diagnosed infants (p=0.15).
Conclusions: A pre-natal diagnosis of HLHS is associated with improved post-natal right ventricular function, reduced metabolic acidosis and end-organ dysfunction. Pre-natal diagnosis is important for optimal prognosis of these infants.

Abstract no: 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 is to determine whether children who underwent congenital cardiac repair with open-heart surgery at <1 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 7-15 years and underwent open-heart surgery at <1 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 <1 year of age are more likely than healthy controls to have a positive screening score for ADHD.

Abstract no: 466
Recurrent giant left ventricular aneurysm of tuberculous etiology in a child

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Case report: A 10-year-old girl was admitted with a 6-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. 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 beating heart. The patient's intra and post-operative 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 poly-serositis condition were inconclusive. The etiology of the aneurysm was not clearly elucidated. Eight months after surgery the girl was brought back with dyspnoea. Chest radiograph revealed right-sided pleural effusion. Cytologic examination of pleural fluid revealed lymphocyte predominance. The Mantoux test was strongly positive. The patient was started on anti-tuberculous therapy. Fourteen months after surgery the girl was readmitted with atelectasis of the left upper lobe. Bronchoscopic examination showed 50% narrowing of the left main bronchus due to extrinsic compression. Left ventriculography disclosed antero-apical giant aneurysm. Reoperation 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 and 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: rare diagnosis of cardiac aneurysm caused by tuberculosis, especially in a child; its recurrence, which is even more rare; and the successful surgical treatment thereof.

Abstract no: 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 etiology. 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.
Material and 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. 89.633.5), 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 did found higher levels in the lipid profile and in the cIMT in children with regression of coronary aneurysms compared with children without coronary aneurysms.
Conclusions: There is no clear evidence of increased atherosclerosis in Mexican children with KD, but there is evidence of an altered lipid profile and in the cIMT in patients with KD with coronary lesions compared with children with KD without coronary lesions which warrant further study.

Abstract no: 477
Familial arrhythmogenic right ventricular dysplasia: MRI 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. To correlate MRI data with other 2010 ARVD task force criteria.
Materials and methods: Retrospective study (January 2001 - June 2012), 70 MRI in 36 children (1 month to 20-year-old, mean 10.7 years) from 22 families with proven 1st degree relative with ARVD. Mean number of MRI by patient was 1.9; 21 patients had 2 or more MRI for a mean follow-up of 2.3 years.

Results: MRI examinations were: normal in 50, non-diagnostic in 2 and abnormal in 18 (11 patients). Anomalies consisted of abnormal cardiac contractility: dyskinesia (n=5), akinesia (n=7) and hypokinesia (n=6), with only 1 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, no LV anomaly. Mean age of the positive cases was 13.6 years. Only 3 patients were symptomatic (cardiac arrest, syncope, ventricular tachycardia). Of the other 2010 ARVD task force criteria, 2 patients also had epsilon wave in the right precordial leads on ECG. 6 patients underwent implantable defibrillator, no appropriate shock, and 1 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 precede 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.

Abstract no: 478
Prevalence of pulmonary hypertension in children with adenoid or adeno-tonsillar hypertrophy in Kenya

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Background/hypothesis: Adeno-tonsillar hypertrophy is a common condition in childhood, but local prevalence is unknown. The condition has serious complications of pulmonary hypertension and cor-pulmonale are common and devastating.
Materials and methods: This was a cross-sectional descriptive study in children aged 0 - 12 years attending ENT clinic 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 ≥25mmHg estimated by the Chemla equation (0.61sPAP+2mmHg).
Results: The prevalence of pulmonary hypertension in children with adenoid or adeno-tonsillar hypertrophy at KNH was 21.1% (95% CI 14.3% to 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 adeno-tonsillar hypertrophy for early identification of pulmonary hypertension.

Abstract no: 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.
Design: A retrospective series of children with acute viral myocarditis were reviewed to confirm the diagnosis.
Methods: CA diameters of the proximal right and the left CA were measured at onset and during the 1st 2 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 >2StDev 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 3 boys who met the selection criteria from 2000 - 2006. 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 cases (78.5%), whereas the history of familial/personal acute infectious illness was present in the remainder. Cardiac enzymes were elevated in 9 patients, normal in 2 and not available in 3. CA involvement was detectable in 9/14 (64.3%) cases; dilatation in 3 (21%) and occult dilatation in 6 (42.9%). Peak CA Z-score was at 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.

Abstract no: 480
Natriuretic peptide release in acute Kawasaki disease predicts gamma globulins 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 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 hypo-natremia is associated with severe KD suggesting an inappropriate secretion of the anti-diuretic hormone (ISADH).
Objectives: We hypothesised that increased serum NT-proBNP correlates with the severity of KD in terms of resistance to gamma globulins (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 was inversely proportional to serum sodium (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 concentration, 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 hypo-natremia during acute KD. It is associated with higher resistance to IVIG, 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.

Abstract no: 481
Surgical congenital heart disease categories and gender: Different incidence, but similar long term survival

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Background: Sex 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, Rigshospitalet (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: 3 256 patients (48.1%) were females. Median age at 1st 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 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.

Abstract no: 483
Surgically treated pulmonary stenosis: 50 years of follow-up

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Background: The natural history of the surgically relieved pulmonary artery stenosis is only limitedly described, since balloon valvuloplasty has replaced surgery. We hypothesise that the incidence of morbidity and mortality increase with increasing time and that more than 30% have been re-operated due to pulmonary regurgitation before the age of 40 years.
Materials and methods: A retrospective follow-up study of the population treated surgically for pulmonary stenosis at Aarhus University Hospital 1 January 1957 - 31 December 2000. Patients were identified by their personal identification number; their hospital records were reviewed and present status noted.
Results: Of 87 patients, 6 were excluded due to missing information. The age at primary surgery was <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 remaining had an infundibular stenosis. 3 patients died (age 0, 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 reintervention. Pulmonary valve replacement due to pulmonary regurgitation was the most common reintervention (60%). Out of 19 patients >40 years and 15 patients between 30 and 40 years of age 47% and 28% respectively, have had a reoperation.
Conclusion: With a mean long term follow-up of 25 years (max 54 years), reintervention was necessary in 24% and almost half of the patients reaching the age of 40 years had a reoperation. This retrospective study shows that there is a call for prolonged follow-up and reintervention in patients operated for simple pulmonary stenosis.

Abstract no: 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 imperforate anus (IA), the outcome of anorectal surgical reconstruction in our centre, and to determine the need for performing echocardiography in all patients with IA.
Material and 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 - December 2010), were reviewed. The average annual delivery rate of this hospital is 5 500.
Results: During the study period, 61 patients of IA were diagnosed that showed an incidence of 1 per 992 live births, and rate of CCA among IA subjects was 15(24.6%). In 12 patients (19.6%), the associated CCA were of mild form and reconstruction of the anorectum went smoothly. Three patients (4.9%) had significant CCA and died.
Conclusion: The incidence of imperforate anus in our hospital is 1 per 992 Live births with the association with CCA is 24.6%. The Majority of associated CCA with IA were of the mild types.

Abstract no: 486

Sinus node dysfunction due to myocarditis

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Introduction: The most common ECG findings associated with myocarditis are nonspecific 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) resolved after few days with Intravenous Immunoglobulin (IVIG) treatment.
Case report: A 17-year-old caucasian male was hospitalised after four day history of progressively worsening chest pain and dyspnea. On physical examination, his vital signs were normal. Cardiovascular system examination revealed no murmur or pericardial rub. 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 was consistent with myocarditis. His elevated creatinine kinase (1,113 U/L), CK MB 84.9ng/ml, troponin-T 4.900ng/ml and myoglobin 211ng/ml. Cardiac MRI revealed diffuse myocarditis, global hypokinesia, low ejection fraction. Adenoviral and coxsackie B1 antibody titers were mildly elevated. IVIG was started in the first day of hospitalisation. On day 3, telemetry showed badycardia with rate of 28 and sinus pauses 6.7 sec. 24-hour Holter revealed bradycardia of lowest rate of 27 with frequent sinus pauses up to 6.1 sec. On day 10 resolved sinus pauses. Subsequently patient showed improvement of cardiac function. Telemetry normal sinus rhythm without sinus pause, for 5 consecutive days prior to discharge. A repeat 24 hour holter monitor after 3 weeks revealed maximum sinus pause of 1.68 sec.
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 may resolve once the inflammation improves.

Abstract no: 488

Echo-Doppler assessment of arterial stiffness in paediatric patients with Kawasaki disease

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Background: There is growing evidence to suggest that patients with a history of Kawasaki disease (KD) experience increased arterial stiffness. 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 (from 2002 - 2012) for any patient who had KD follow-up >1 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 (ß-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 2 groups. The PWV was higher among KD patients compared to controls (458±153 vs. 370±61cm/s, p=0.0008). The Zc, (Ep), and ß-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 2 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 ß-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 indexes.
Conclusions: Arterial stiffness is increased in children after Kawasaki disease. There was no association between coronary artery involvement and PWV.

Abstract no: 495

World Heart Federation echocardiographic criteria for rheumatic heart disease allows for reproducible diagnosis world-wide

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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 is to determine if 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 epidemiologic 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 diseases. Overall agreement in categorising echocardiograms as normal, borderline and definite RHD (primary end-point) 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 end-points, the presence of pathologic 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 utilised by experienced physicians. The ability of less experienced physicians and community health workers to diagnose RHD by echocardiography need 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.

Abstract no: 496

Wrestling manoeuvre as the culprit in acute severe aortic regurgitation

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Background: 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 exam findings revealed a new onset ¾ diastolic murmur and blood pressure of 174/38mmHg 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.
Method: 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.
Results: 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, Ross procedure was performed with excellent results. His aortic valve pathology result showed slightly thickened valve cusps without evidence of vegetation of micro-organisms.
Conclusions: Since the new exam findings and symptoms have developed immediately after wrestling, we conceptualise 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's "choke-hold" manoeuvre.

Abstract no: 512

Adjustable bilateral pulmonary artery banding for hypoplastic left heart and its variants

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Background and objectives: 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 - January 2012, we retrospectively evaluated 17 children who underwent BPAB and received the Norwood Procedure or definitive repair (9 in the adjustable BPAB group and 8 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 O2 saturation; along with non-absorbable sutures (5 - 0 Prolene) a few millimetres apart from the absorbable sutures. In the non-adjustable BPAB, a 2mm 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 4 children in the non-adjustable group.
Conclusion: Adjustable BPAB is effective in preventing stenosis of the pulmonary artery and allows for pulmonary artery growth.

Abstract no: 516

Outcomes of primary repair of atrioventricular septal defects in children: An experience of Red Cross Children's Hospital, 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 1st 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 1 year.
Results: Median age was 8 (3 - 26) months (9 aged <6 months, 10 aged 6 - 12 months and 12 aged >12 months). Mean weight was 6±2.4kg. The majority (23/31) had Down syndrome (D.S). Twelve children required pre-surgical cardiac catheterisation. An acceptable surgical result was achieved in 90% (28/31). There were 3 re-operations; 2 right and 1 left atrioventricular valve anuloplasty. Early (30-day) mortality was 13% (4/31); 3 in-hospital deaths attributable to infection and 1 "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 which reduce survival following successful surgery and discharge from hospital should be addressed.

Abstract no: 523

Study on the diagnosis and treatment of childhood supraventricular tachycardia with intracardiac electrophysiology attached with 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 electrophysiologic study (EPS) and radio-frequency catheter ablation (RFCA) in children.
Methods: Fifty patients with SVT were enrolled in the study from December 2007 - July 2012. The data of ECG and the results of RFCA (ablation success, complications and recurrence) were studied retrospectively.
Results: Among the 50 patients, 29 cases (58%) with atrial ventricular re-entrant tachycardia (AVRT); 10 cases (20%) with atrial ventricular node re-entrant tachycardia (AVNRT); 6 cases (12%) with atrial tachycardia (AT). Two cases with AVRT accompanying with AVNRT; 1 case with AVRT and AT at the same time. The procedure was abandoned in 2 patients because of considering the risk in 1 case (AVRT and accessory pathway (AP) location near his bundle), and 1 case not induced by EPS. No recurrant tachycardia and complication was found in all 50 cases to date.
Conclusions: RFCA is a safe procedure with a high success rate and low complication for tachycardia management, but the indications for RFCA should be carefully considered in very young patients.

Abstract no: 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 programs was reviewed for generalisable implementation experiences.
Results: Twenty-four individuals' components of comprehensive RF/RHD control programs 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 schema.
Conclusions: Existing literature contains considerable lessons on the design and implementation of comprehensive RF/RHD control programs. Fashioning these guidelines and programmatic experiences into a conceptual framework schema benefits clinicians, policy makers and RHD advocates. Extending the framework may yield a model to isolate and evaluate individual programme components.

Abstract no: 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 multicenter studies. This single centre study was undertaken to review our experience with ICD implantation in children and young adults with relatively different etiologies.
Methods: We retrospectively reviewed the records of the paediatric patients who underwent to cardioverter-defibrillator implantation at the Instituto Nacional 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 7 patients and biventricular in 1.6 patients received 30 shocks. 4/15 patients (26%) from the secondary prevention group experienced at least 1 appropriate shock during a median period of 3.4 years (range: 1.2 months - 6.5 years). Eight inappropriate shocks were delivered in 2 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 complication was zero and the incidence of inappropriate shocks was low. Careful programming is mandatory to reduce the inappropriate shocks.

Abstract no: 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 still unclear. This study was undertaken to review our initial experience with CRT in children.
Methods: We initiated the CRT program in children on January 2011; only 2 children have been included.
Results: Case 1: A 5-year-old girl was transferred because of progressive LV dysfunction. At age 1 she underwent a ventricular septal defect closure. A 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 follow-up no clinical improvement was apparent. 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. 3 endocardial leads were placed on the right atrial appendage, the RV apex and on the LV postero-lateral 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 which underlying anatomical and patho-physiological substrate is more effective. In less developed countries there are financial limitations to apply this and other new therapies to all the patients who could possibly benefit.

Abstract no: 548

Extensive myocardial infarction in a 11-year-old girl: Case report

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Background: 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 start with catecholamines blood pressure remained low.
Methods and results: Primary echocardiogram revealed a depressed myocardial function. Initial blood work showed white blood cell count of 27 000, abnormal I/T ratio, CK 7000U/l, CK-MB 688ug/l, Troponin 12 ug/l. The girl's history was unremarkable. Repeated echocardiogram after 4 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 V3 - V5, ST elevation in V1 - V2. Based on these findings we primarily diagnosed an acute myocarditis (DD acute vasculitis with coronary vasospasm, hypoxic ischaemia after near drowning). Within the following 2 days her LVEF recovered (EF 50 - 55%). A sudden episode of nausea and vomiting on day 3 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 5. However, despite regained normal coronary flow the LV function remained depressed (EF 20 - 25%). After 6 months patient underwent heart transplantation.
Conclusion: Presence of segmental myocardial dyskinesia/hypokinesia always implies further diagnostic imaging regardless of the patient's age.

Abstract no: 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 the cases. Regardless the reported low incidence for 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 6 months (2 - 3 times per month). The girl presented with developmental delay. The mother had ablation at the age of 18 years for WPW. Other family history was unremarkable. Detailed neurologic evaluation (2 EEG, psychomotor skills) was unremarkable. At the age of 18 months the frequency of LOC increased after an event-free period of 3 months. Echocardiogram revealed a large atrial septum defect. The standard ECG and the 24 hour Holter exam was unremarkable, but a 7-day ECG recording revealed 2 episodes with sinus pauses of up to 3.4 seconds both during daytime when the toddler was awake. The diagnosis of a sick sinus syndrome was made. A VVI pacemaker (back-up heart rate 80/minute) was put in place and ever since no episodes of sudden LOC have been reported. The closure of the atrial septal is deferred to the age of 4 - 5 years.
Conclusion: This case illustrates the importance of detailed cardiac assessment for sudden LOC in childhood even for toddlers with obvious neurologic 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 were suspicious for any kind of arrhythmia.

Abstract no: 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 2 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 pathologic murmur on examination or an ECG consistent with ASD (QRS duration >100 msec with rSR' in Lead V1) prompted echocardiographic screening (ECHO) by a paediatric cardiologist.
Results: CHD was identified in 47 of 1 615 patients (3.0%) screened over 5 days, 14 (29.8%) of whom had an ASD by ECHO. All patients with ASD had an abnormal exam, 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.

Abstract no: 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 published literature attempting to distinguish the most effective preventative measure. Some abstracts 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 - 16.5 years). Median follow-up was 4.3 years (2 days - 11.9 years). Of these, 120 of 134 patients were started on warfarin, whilst only 10 were started on aspirin. Four patients died in the early post-operative period before establishment on warfarin or aspirin. In the warfarin group, 4 patients converted to aspirin. One patient had significant extradural haematoma following a bicycle accident, which needed drainage. One patient developed post-operative haemorrhagic stroke, whilst 2 patients converted out of choice. Of the remaining 116 patients on warfarin, 7 had minor complications with self-limiting epistaxis (4 patients), superficial bruising (1 patient), bleeding into pleural cavity (1 patient), and clot in left atrium (1 patient due to low INR). One patient developed clot on fenestration plug device and was put on warfarin and aspirin. 7/8 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. 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.

Abstract no: 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 extra-cardiac vasculature in newborns with congenital heart disease is not widely utilised. 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 echo-cardiography. 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 - July 2012 was performed. All underwent an initial reference scan, with subsequent dynamic images acquired within a single breath-hold (Table 1). Indication for referral, diagnosis made from the MR scans and correlation with surgical findings was 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 0.1mmol/kg). Median age: 5 days, range 2 - 23 days. Mean weight 3.1kg, range 2.1 - 4.5Kg. (Table 1). Seven patients proceeded to surgery based on the MR where findings were confirmed; 1 required no further intervention; 1 required diagnostic catheterisation to assess Multiple Aorto-Pulmonary Collateral Arteries (MAPCAs).

TABLE 1: Scan parameters and distribution of diagnoses

Scan parameter	mean	range
Voxel size	1.55mm	1.2 - 1.78mm
Slices	110	55 - 130
TR/TE	5.2/1.5ms	4.7 - 6.2/1.4 - 1.8ms
Reference scan duration	15.8secs	8 - 24secs
Dynamics		9 - 20
Time per dynamic	1.85sec	2 - 2.7secs
Time for dynamic scan	24.4secs	17.2 - 35secs
Total scan duration	41.5secs	31.4 - 59secs
SENSE	x2	
Keyhole	20%	
Flip angle	25 degrees	
Indications for scan		**Number**
Assess arch anatomy		3
Assess pulmonary venous drainage		3
Assess collateral vessels from aorta		3
Diagnosis		**Number**
Pulmonary atresia with VSD and MAPCAS		3
Interrupted aortic arch		2
Partial anomalous pulmonary venous drainage		2
Anomalous right subclavian artery		1
Heterotaxy syndrome with total anomalous pulmonary venous drainage		1

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.

Abstract no: 569

Predictors of outcome in paediatric idiopathic cardiomyopathy

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Objectives: Idiopathic dilated cardiomyopathy (IDCM) is a severe illness with high mortality in the pediatric population, the purpose of this study was to highlight our experience with the clinical course and outcome of IDCM.
Methods and material: Patients' files were reviewed retrospectively for diagnosed cases of IDCM in the pediatric cardiology unit of King Abdulaziz University Hospital, Jeddah, Saudi Arabia from January 2003 - June 2011, data about full history, clinical examination 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. The presenting age was 14 months (median) with a range of 2 months - 12 years with a female predominance (63%). On presentation; cardiomegaly was noted in 72(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 difference in several areas. Group 1 had 40(48.2%), Group 2 had 23(27.7%) while 20(24.1%) were in Group 3 including 9 cases who died. Survival rate over 3 years was 78%. Older age was associated with a worse outcome (spearman's rho=0.3, p=0.04).
Conclusion: The majority of subjects presented during the first year of life. The three year survival rate was 78%. Favorable outcome correlated with younger age at presentation.

Abstract no: 579

Cardiac lesions in neonates with gastro-intestinal malformations

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Introduction: The association between congenital structural cardiac lesions and major gastrointestinal malformations such as tracheo 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 tracheo-oesophageal fistula. It was decided to do an institutional retrospective analysis of this association at an African tertiary care centre.
Method: This study is 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 to determine whether or not gender preponderance exists and document the geographical distribution of these neonates.
Results: An average of 20 000 - 25 000 live births per year occur in the neonatal unit at Chris Hani Baragwanath Hospital. Over the 5-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. The most lesions occurred in neonates with anorectal malformations (27%) and omphalocoele (38%). The most commonly found lesions are 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 catchment area which includes Southern Gauteng and the North West Province.
Conclusion: The findings are similar to that of 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.

Abstract no: 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 para-septal pathways and described paradoxical or hypo kinetic septal motion. Data regarding non-septal pathways is 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±17kg, were assessed. All had WPWs, structurally normal hearts and underwent successful ablation (cryo-energy in 4; radiofrequency in 12). Septal/para-septal pathways were present in 6 (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 pre-ejection times (11.0±3.3ms, 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 4 patients with LVEF<50% prior to ablation, 2 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±7.1ms (p=0.015) vs. 1.4±11.4ms (p=0.655).
Conclusion: LV systolic function and dyssynchrony improve after ablation of antegrade conducting accessory pathways in children, with greater changes for non-septal pathways.

Abstract no: 589

Left ventricular submitral aneurysms

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Objective: Retrospective institutional review 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 compromises 31 patients treated surgically at 2 institutions from 2001 - 2012. Patients were of African ancestry (n=25) and mixed ancestry (n=6). Natural history, clinical presentation, histopathological findings, aetiology, 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 as to the degree of posterior mitral annulus involvement. In Group I, (n=18) a single neck, in Group II, (n=3) multiple necks, 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, extracardiac approach in 2 and combined approach in 9 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.

Abstract no: 600

Biophysical properties of the aorta in adolescent females with anorexia nervosa (AN)

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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. 66 adolescent females with AN and 21 adolescent female controls exercised on a semi-recumbent ergometer in 3 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 3 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 (VO2), minute ventilation (VE), respiratory exchange ratio (RER), and arterial-venous oxygen difference (a-vO2) were determined using open circuit spirometry.
Results: Patients with AN had a significantly lower BMI (16.7 vs. 19.7kg/m^2, p<0.001), total work (1126 vs. 1914 J/kg, p<0.001), total test duration (13.8 vs. 20.8 minutes, p<0.001), peak VE (47.4 vs. 72.0L/min, p<0.001), and VO2 (31.3 vs. 39.7mL/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. controls. HR, FS, MVCFc, and CI showed no difference at pre-exercise, increased with exercise, and were lower at peak exercise in AN vs. controls. WS decreased with exercise and was lower in AN 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-vO2 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 as compared to controls.

Abstract no: 621

Kawasaki disease analyses in a reference paediatric hospital in southern Brazil from 1980 - 2012

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Background: Kawasaki disease is a systemic and acute vasculitis with unknown etiology and actually considered the main cause of acquired heart disease in developed countries' children. The clinical presentation is typically characterized by 5 days or more of fever, associated with at least 4 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 analyze epidemiological, clinical and therapeutic aspects of the disease in patients at the Joana de Gusmão Children's Hospital, Florianópolis - Santa Catarina, from 1980 - 2012.
Method: Using data from medical records, cases were analysed considering age, sex, colour, 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, more frequently since 2001, predominatly children <5 years old, boys, white race and from Florianópolis. Out of patients, 70% fulfilled the diagnostic criteria. Anaemia, leucocytosis with neutrophilia, thrombocytosis, increased ESR and CRP were frequent findings. Cardiac involvement occurred in 53.3%, predominantly with coronary alterations. 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. The cardiac compromise was found to be frequent, especially in the coronary arteries, probably due to the long time lapse to identification.

Abstract no: 624

Lone atrial fibrillation in an adolescent

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Background: We report a 15-year-old adolescent girl with a good past health, who presented with sudden onset of palpitation and dizziness. Examination revealed fast, irregular apical beats (170 beats/minute) with normal blood pressure.
Methods and results: 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 more recurrence of AF.
Conclusions: 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.

Abstract no: 625

Ventricular hypertrophy with outflow tract obstruction vs. dilated cardiomyopathy in neuroblastoma

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Background: Catecholamine-associated hypertension secondary to neuroblastoma can occasionally be demonstrated, but cardiomyopathies caused by neuroblastoma have been rarely reported. We report 2 cases of neuroblastomas with different extremes of severe cardiomyopathic changes and complications.
Methods and results: The 1st case was a 2-month-old girl with a retroperitoneal mass which was incidentally found in an ultrasonic examination during a post-natal evaluation of ante-natal hydronephrosis. Hypertension was detected and echocardiogram showed severe concentric left ventricular hypertrophy with mid-ventricular obstruction, with a peak pressure gradient of 51mmHg, simulating hypertrophic obstructive cardiomyopathy (HCOM). The 2nd case was a 30-month-old girl 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. Choices of anti-hypertensives were different. Beta-blocker was used in the 1st case to promote left ventricular filling, while angiotensin-converting enzyme inhibitor was used in the 2nd case. Anthracycline group of chemotherapy was avoided in the initial phase of treatment. Chemotherapy and subsequent removal of tumour led to successful normalisation of blood pressure and regression of abnormal cardiomyopathic changes.
Conclusions: HOCM-like feature secondary to catecholamine-secreting neuroblastoma is exceedingly rare. The very young onset of HOCM suggested that the remodelling of the heart had already started during her fetal period. Updated management of catecholamine-induced cardiomyopathy associated with neuroblastoma is also discussed.

Abstract no: 627

Routine head ultrasound scans are not indicated in the pre-operative evaluation of infants with congenital heart disease

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Background/hypothesis: Head ultrasound scans (HUS) are a routine part of the pre-operative evaluation of young infants with CHD in many institutions. HUSs has 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.
Materials and methods: 167 infants born at 35 weeks gestation or greater due to undergo surgery for CHD were included in this 3-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 neuro-radiologists 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 5 infants (3%) and MRI in 44 infants (26%) ($p<0.001$). The abnormalities on HUS were: intraventricular haemorrhage in 4 that was not confirmed on MRI performed within a few days after HUS and peri-ventricular leukomalacia in 1. The predominant MRI abnormality was white matter injury (32 infants). Other findings included infarct (14), haemorrhage (5), and lesion or infarct of deep nuclear grey matter (2).
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 is not indicated in asymptomatic term or near-term neonates undergoing surgery for CHD.

Abstract no: 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 neurologic 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.
Materials and 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 physiologic, were excluded from the analysis.

Results: We report 4 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 (4), ToF (3), PS (2) and 1 each of Ebstein anomaly, PA/IVS, ASD, and DORV.
Conclusions: This is the 1st 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 in this area of cardiac development.

Abstract no: 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 of age) with ASO-surgery in the neonatal period were studied. Seventeen 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 NE ([3H]NE) in 8 of these children to examine total body and cardiac sympathetic function at baseline and following 5 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 spill over doubled vs. baseline ($p<0.002$) and the coronary venous-arterial concentration of [3H]dihydroxyphenylglycol increased 4-fold ($p=0.04$).
Conclusions: Arterial switch operation performed neonatally appears to leave cardiac vagal function intact and, although cardiac sympathetic activation in response to adenosine occurs, cardiac neuronal NE re-uptake is impaired. This may be pro-arrhythmic by reducing removal capacity of NE from the cardiac synaptic cleft.

Abstract no: 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 pre-natal risk factors and CHD risk.
Methods: Patients with CHD and healthy controls enrolled in Heart Centre Biobank were studied. Pre-natal exposure data and CHD anatomic subtypes were obtained from questionnaires and medical records. The cardiac phenotype was compared by pre-natal 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 1 risk factor, and 27.5% had ≥2 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 pre-natal 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 are associated with specific CHD phenotypes. Additional studies will evaluate if genetic factors increase the susceptibility to development of fetal CHD in pregnancies with environmental exposures.

Abstract no: 659
Genetic associations with anthracycline cardiotoxicity in paediatric cancer patients

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Background: Anthracycline cardiotoxicity (ACT) is the 3rd 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: 139 paediatric cancer patients were prospectively enrolled through the Heart Centre Biobank. Seventy seven patients were genotyped for 3 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 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% patients had at least one abnormal measure (≤55%) of LVEF at any time point. All 3 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 is associated with LV posterior wall thinning during follow-up. Additional candidate SNPs are being genotyped and will help early identification of patients with a genetic susceptibility to ACT that can guide pre-emptive risk-reduction measures.

Abstract no: 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/hypothesis: 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 6 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 and RV ejection fraction (RVEF), 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 echo-cardiograms was 11.9 years and for MRI 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.

Abstract no: 673

Spectrum of congenital heart disease among children presented at Uganda Heart Institute

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Background: Congenital heart diseases are among the commonest birth defects globally. The Echo diagnosis of children who presented to the Uganda Heart Institute was reviewed over a period of 5 years (January 2007 - December 2011). The Uganda Heart Institute is a super specialty centre located in Mulago Hospital the country's main referral hospital. It is the main centre that offers paediatric cardiac services in the country.
Objectives:

- To describe the common congenital cardiac lesions among children in Uganda.
- To establish a registry for congenital heart disease. (To date no large scale database exists.)

Methodology: An ongoing registry has been compiled since 2007 by generating Echo reports and storing them digitally. These records were retrieved and analysed. Data was 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 there are genetic or environmental factors to account for these variations.

Abstract no: 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 is to determine the incidence of rhythm and conduction disturbances in ccTGA patients with 2 adequate-sized ventricles.
Patients and methods: Retrospective analysis of records of 49 patients from a single centre was 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 3 months (1 day - 34 years) and mean period of follow-up was 4.5±1.8 years (1 month - 22 years). Forty seven of them had associated heart anomalies. The most common associated lesion was VSD (38 patients). Pulmonary valve abnormalities were 2nd most common lesion. Pulmonary stenosis was more common than pulmonary atresia (17 vs. 6 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 (9 patients). Conventional biventricular repair was and double switch procedure were performed equally (5/5 patients). Tricuspid valve replacement was performed in 2 patients. On initial examination, 2 patients had 1st degree AV block, 1 2nd degree AV block and 1 congenitally complete AV block; additionally, 1 patient had atrial ectopic rhythm, 1 left bundle branch block. Supraventricular tachycardia was detected in 3 patients. At follow-up, complete AV block developed in 5 patients after intracardiac surgery. Pacemaker implantation was required for these patients and 1 patient with congenitally complete AV block.
Conclusions: Patients diagnosed as ccTGA should be followed-up 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.

Abstract no: 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 - 8.8kPa. 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 catheterization 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 1 had a persisting fenestration, and 6 had hepatomegaly. All had an extra-cardiac repair. Liver enzyme values were higher in the Fontan group (ALT30U/L vs. 16U/L; p<0.003 and GGT49U/L vs. 11U/L; p<0.0001). Elastography values were significantly higher in Fontan patients compared with controls (17.1kPa (11 - 39) vs. 4.7kPa (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.

Abstract no: 693

Outcomes of paediatric hypertension: Data from Rehiped (Spanish registry for paediatric pulmonary hypertension)

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Objectives: There is lack of data about epidemiology and survival in paediatric pulmonary hypertension.
Methods: From January 2009 - June 2012, 21 referral and non-referral centres collected data of 225 patients, with mean pulmonary artery pressure (mPAP)>25mmHg and pulmonary vascular resistance index (PVRI)>3WU.m^2. We included all Dana Point (DP) ethiologies (groups I to V) and analysed differences in survival according to DP ethiologic group, age at diagnosis (dx), functional class (FC), syncopes, chromosomopathy, right atrial pressure (RAP), mPAP and cardiac index (CI), PVRI, PVRI/SVRI.
Results: Patients were classified as: Group I (PAH n=142, 61%), Group II (left heart disease n= 31, 14%), Group III (respiratory disease n=41,18%, half of them with bronchopulmonary dysplasia), Group IV (thromboembolic PH n=2, 1%), Group V (n=10, 4.5%, mostly inherited metabolic diseases, but 31% had multi-factorial PH). Median age at dx was 4.3±4.9 years (50% <2 years). 21% were premature, and chromosomopathies/syndromes were present in 38%. Functional class at dx was III/IV in 53%, without significant differences in mPAP, RAP, PVRI/SVRI or CI, between the different DP groups. For the whole cohort, 1 and 3 year survival was 80% and 74%. Mortality risk factors (univariate analysis): DP ethiology group (3 years 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 dx (p <0.001), RAP (p=0. 006), and age at dx (3-year survival 60% for <2 years, 92% between 2 - 8 years, and 85% for >8 years, p<0.001). Multivariate analysis: ethiology (p<0.001), age at dx (p<0.001), FC at dx (p<0.001) and RAP (p=0.002).
Conclusions: Age at diagnosis is a significant risk factor: Worse survival in the younger patients (often frequently missed in referral centre-based registries). In severe paediatric PH, prognosis is better in PAH than in the other DP ethiologic groups.

Abstract no: 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 7018 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 until 1 January 2012.
Results: Number of patients increased from 1233 in 1971 - 1980 to 2286 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 - 80, and 25.4% in 2001 - 2010), and a decrease in surgical treatment of anatomically simpler defects. Overall 30-days post-operative 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 - 80 who survived >16 years, 50 (4.5%) had HLHS, AVSD, IAA, PA or UNI. In contrast, among the 1 682 patients operated in 1991 - 2000 surviving >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 is increasing rapidly, since more of these defects are treated surgically, and with substantially improved survival.

Abstract no: 699

Management and clinical features of paediatric pulmonary arterial hypertension in Spain: Data from Rehiped (Spanish registry for paediatric pulmonary hypertension)

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Objectives: There is lack of information about 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 - June 2012, 21 centres (referral and non-referral) included cases, aged 2 months - 18 years at diagnosis (dx), with mean pulmonary arterial pressure mPAP >25mmHg, Pulmonary Vascular Resistance Index PVRI > 3WU.m^2, Pulmonary Wedge Pressure <15mmHg.
Results: 142 patients were included: Congenital heart disease (PAH/CHD, n=105, 74%), idiopathic/familial (IPAH n=32, 21%), porto-pulmonary (n=3, 2.1%), HIV infection (n=1, 0.7%), connective tissue disease (n=1, 0.7%). 42% were prospective cases. Mean age at dx: 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 dx. Haemodynamic data: mPAP 46+18mmHg, PVRI 8.7+7.8WU.m2, PVRI/SVRI 0.7+0.5, Cardiac Index (CI) 4.6+2l/min/m^2. Only 6% of IPAH children were responders in the vaso-reactivity test. Of the whole group, 92% received PH drugs (46% in combination, 23% prostanoids), 7% oral anti-coagulation and 14% anti-agregation. For the whole group, 1, 3 and 5 years survival was 89%, 85% and 79%, respectively, without differences between IPAH and PAH/CHD. Mortality risk factors (univariate analysis): younger age at dx (p=0.013), FC III/IV (p<0,001), CI (p=0.005) and RAP (p=0.014). Multivariate analysis: age at dx (HR0.84 [CI 95% 0.73-0.96], p=0.009), FC (p <0.001), IC (HR 0.46 [CI 95% 0.3-0.7], p<0.001), RAP (HR 1.2 [CI 95% 1.06-1.38], p=0.012).
Conclusions: Rehiped registry patients had younger age at diagnosis, >% of prospective cases, and <% 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 dx was also risk factor for mortality.

Abstract no: 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.
Materials and methods: The study was a retrospective, observational study of all patients <20 years of age who received an ICD over a 4-year period (July 2007 - 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 6 patients received a transvenous ICD. Underlying diagnoses and age at implant were similar for the 2 groups. There were no implant complications with subcutaneous ICDs, but 1 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 2 patients with a subcutaneous ICD and 1 patient with a transvenous ICD. No subcutaneous ICD required revision, but 2 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.

Abstract no: 705

Prevalence of myocardial fibrosis by CMR 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 of myocardial fibrosis (MF). Cardiac magnetic resonance (CMR) imaging with late gadolinium enhancement (LGE) can detect a small and focal MF.
Aim: To assess the occurrence of MF in children with HCM and to evaluate its clinical significance.
Material and methods: Fifty six children with HCM underwent CMR from January 2010 - June 2012. All 56 patients were divided into 2 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%), fatigue ability (80% vs. 65%). Among children with MF LV wall thickness (mean 22mm vs. 14mm; p=0.0001) and LV mass index (mean 105g/m² vs. 86g/m²; p=0.003) were significantly greater compared with those without MF. In 79% patients in Group I and in 36% in Group II LV mass was increased (p=0.003). In Group I nsVT episodes were more frequent than in Group II (10% vs. 4%). Patients in both groups differed significantly regarding the occurrence of major risk factors for SCD (67% vs. 31%; p=0.016). The LV wall thickness≥30mm and cardiac arrest were observed only in Group I (20%; p=0.025 and 7% respectively) and family history of SCD more frequent was 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 2 patients as secondary prevention and in 8 as primary prevention.
Conclusion: (1) The presence of myocardial fibrosis is related to an unfavourable clinical course of HCM in children. (2) Visualising LGE in CMR may contribute to better risk stratification of SCD and may help with making therapeutic decisions in HCM patients.

Abstract no: 707

Pulmonary hypertension in bronchopulmonary dysplasia: Clinical findings, associated cardiovascular abnormalities and outcomes

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Background and objectives: Severe pulmonary hypertension (PH) worsens the prognosis of bronchopulmonary dysplasia (BPD). Clinical characterisation, incidence of cardiovascular abnormalities (CVA) and outcome in this setting haven´t yet been fully established.
Methods: 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 - December 2011. Median gestational age was 27 weeks (IQR 24 - 27.3) and median birth weight 740g (IQR 620g - 830g). 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 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%): 9 systemic to pulmonary collaterals, 7 pulmonary vein stenosis, 4 ASD, 1 restrictive VSD and 9 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 32mmHg (20 - 40.5). At a median follow-up of 35 months (IQR 17 - 84): 7 patients had undergone closure of shunts, 21 received specific drug treatment for PH (PH improvement or resolution in 9), 3 showed spontaneous improvement and 8 (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.

Abstract no: 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.
Aim: To evaluate the effectiveness of pain management for benzathine penicillin injections.
Methods: A BUZZY vibrating cold pack device and 0.25mls of 2% lignocaine, mixed with the BP prior to administration, were offered to patients. A survey was conducted evaluating pain scores at 4 time points (delivery, 2 minutes 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 <16 years compared with older patients. Paired data pre and post intervention was available (n=119). Mean pain score at delivery changed from 5.4/10 pre intervention to 2.4/10 post intervention (p ≤.001). Pain scores were significantly reduced at all 4 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 5 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.

Abstract no: 711

The usefulness of scintigraphy with 99mTc-anti-granulocyte antibody for diagnosis and follow-up in children with myocarditis

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Background: Clinical diagnosis of myocarditis (myo) 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.
Aim: To investigate whether scintigraphy with 99mTc-anti-granulocyte antibody (99mTc-AGA) is useful for diagnosis and follow-up in children with myo and to determine its correlation with EMB and clinical features.
Material and methods: From 2005 - 2010, 11 children, aged 6.6 - 17 years, mean 13±8 years, presenting with symptoms of myo were evaluated at the time of initial presentation and 6, 12 and 24 months after the first study. Patients' demographics and clinical symptoms, as well as results of echocardiography, electrocardiography, EMB and scintigraphy with 99mTc-AGA were analysed. In all patients myocardial scintigraphy was performed with estimation of anti-granulocyte antibody uptake. EMB was done in 10 patients at the time of initial presentation and in 8 after 6 months.
Results: In 11 in 10 (91%) positive antigranulocyte uptake was observed, with EMB confirming myocarditis in 8 (80%) children. In scintigraphy after 6 months positive uptake was found in 9 (82%), with EMB performed in 8 showing persistent myocarditis; after 12 months scintigraphy indicated positive uptake in 7 (64%) and after 24 months only in 4 (36%) of patients. The mean LVEF was 59%±12 at initial presentation and 61%±10, 65%±12, 66%±7 after 6, 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 in follow-up; (3) The control scintigraphy performed in follow-up after 6, 12, and 24 months allowed to evaluate resolved or persistent myocarditis; and (4) Myocardial scintigraphy results indicate that inflammatory process in the myocardium decreases significantly after 12 months from the onset of the disease.

Abstract no: 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 6-day window was screened at Duhok Teaching Hospital (Kurdistan, Iraq) as part of a Ministry of Health-directed demonstration project. All infants were 1st screened with a VScan® handheld device (VS; assessment of 2-D and colour-flow data) and then using a Seimen's Cypress® echocardiogram machine (SC; assessment of 2-D 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 as compared with 24 for SC. VS and SC found 150 and 154 PDAs respectively. Both diagnosed 167 of 178 newborns with PFOs. VS diagnosed 2/3 infants with muscular ventricular septal defects (VSD) identified by SC. Both machines diagnosed 5 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.

Abstract no: 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 - 25 May 2012, a convenience sample of 176 newborns were screened at Azadi Teaching Hospital in Duhok, Iraq, by 2 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 176/276 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 3. There were 9 structural defects (5.1%). Structural defects include 3 cases of tiny muscular ventricular septal defect (VSD) (1.7%), 2 cases of atrial septal defect (ASD) (1.1%), 3 cases with dysplastic aortic valves (1.7%), 1 aneurysmal atrial septum (0.6%), 1 dysplastic tricuspid valve (0.6%) and 1 bicuspid aortic valve (0.6%). None of these defects were considered haemodynamically significant.
Conclusion: This study is the 1st 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.

Abstract no: 717

Epidemiology of pulmonary arterial hypertension in Spanish paediatric population: Data from Rehiped and Rehap

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Background: Nowadays there is 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 2 months - 18-years-old from the Rehiped (Spanish Registry for Paediatric Pulmonary Hypertension) and Rehap (Spanish Registry for Pulmonary Arterial Hypertension in Adults) registries.
Methods: Voluntary reporting (from referral and non-referral centres) of their paediatric patients (aged 2 months - 18 years) diagnosed with PAH. Consecutive cases diagnosed from January 2009 - June 2012, were selected to provide data on average annual incidence from 2009 - 2012, and point prevalence (June 2012). PAH was defined by a mean pulmonary artery pressure (PAP) >25mmHg, pulmonary vascular resistance (PVR) >3 Wood Units (WU) and pulmonary capillary wedge pressure <15mmHg.
Results: A total of 205 PAH patients, (mean age 8+5.7 years) were 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 10.1 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 2 months - 18 years.

Abstract no: 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 4 - 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 were 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 ($\beta=0.45$, $p<0.0001$), LV stiffness ($\beta=0.28$, $p<0.0001$), RV stiffness ($\beta=-0.21$, $p=0.001$) and age ($\beta=0.25$, $p=0.001$) were independent determinants of Qp/Qs ($r=0.65$, $p<0.0001$).
Discussion: The present study confirmed for the 1st time that increased LV stiffness independently increases Qp/Qs in ASD. Because increased Qp/Qs by 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.

Abstract no: 721

Neurocognitive outcomes of infants with single ventricle physiology seen in complex congenital heart disease clinic

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Background: Earlier intervention and exposure to various peri-operative techniques and care have contributed to improved survival rates, but may also adversely impact future developmental processes and outcomes.
Aims: Determine which medical characteristics of the baby are predictive of which neurodevelopmental outcomes. Determine changes of an individual baby's neurodevelopment over time. Describe neurodevelopmental outcomes of an aggregate population of infants with congenital cardiac disease and early surgical repair followed with 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 influence of peri-operative procedures, including time on ECHMO post-operative, 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 comorbid 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 are 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.

Abstract no: 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 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. Similar data for pulse oximetry is given in the table considering a value of <92% as abnormal.

	Age <24 hours		Age >24 hours	
	Sensitivity	Specificity	Sensitivity	Specificity
Cyanotic CHD	25.00%	90.57%	52.00%	91.96%
PDA dependent CHD	25.00%	90.56%	68.42%	92.00%

Conclusion: The prevalence and spectrum of significant CHD in a community level hospital in India is not very different to that reported in 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.

Abstract no: 729

Löffler myocardiopathie in a 9-year-old boy

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Background: Löffler Myocardiopathie is a restrictive cardiomyopathy associated with eosinophilia. Eosinophilic states that may occur in association with Löffler myocardiopathie include hypereosinophilic syndrome, eosinophilic leukaemia, carcinoma, lymphoma, drug reactions or parasites.
Methods and material: The authors present a 9-year-old male patient that was admitted to the Brazilian Medical Institute (IMIP) in June 2005 with pallor for a month, lower backache for 1 week and fever for 5 days before admission. His vital signs included heart rate = 140bpm, respiratory rate = 28bpm, blood pressure = 90 x 60mmHg. On physical examination was found general state compromised, tachypnoea (+/4+), pallor (3+/4+).

Results: There was no sign of cyanosis and pulses were present with normal amplitude. Heart rhythm was irregular due to extrasystoles (20bpm), 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 6cm from the right costal margin. Joint examination was normal. Laboratory findings included microcytic and hypochromic anaemia with haemoglobin of 5gm/dl, thrombocytopenia with platelet count of 47.000/mm^3 and leucocytosis with white blood cells = 74.000/mm^2 and 83% of eosinophils.
Conclusions: Myelography revealed hyperplasia of the eosinophil lineage and 15% of linfoblastis infiltration. Chest radiograph showed cardiomegaly with normal pulmonary flow and the 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 thrombus. Magnetic resonance revealed imaging consistent with Löffler myocardiopathie in its biventricular fibro-thrombotic. He was submitted to a new myelography that 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.

Abstract no: 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 3-dimensional echocardiography, to assess the structural and functional changes in mitral valve apparatus during an episode of acute rheumatic fever.
Methods: Twenty two consecutive cases of 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 4 weeks of steroid therapy. Age-matched chronic rheumatic mitral regurgitation patients were included as controls.
Results: Mitral regurgitation was severe in 8 (36%), moderate in 12 (55%), and mild in 2 (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 was no significant changes in echocardiographic parameters except the LV end diastolic dimensions, which decreased from 54.17±9.75mm at baseline to 52.5±10.18 mm on follow up (p=0.02). The thickness of leaflets at tip, mid part and base of anterior/posterior leaflets during acute activity was higher than the control. The thickness of the mid-part of anterior mitral leaflet was 5.1±0.6mm in the study group initially and 4.9±0.96mm on follow up, while the control group had a thickness of 3.87±0.8mm (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 favorable changes on left ventricular dimensions measurable by echocardiography.

Abstract no: 734

Left atrial myxoma mimicking rheumatic 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.
Methods and material: The authors present an 11-year-old female admitted to the Brazilian Heart Institute with intermittent fever associated with dyspnoea on moderate exertion for 6 months. Progressive symmetric arthralgia in the knees, followed by the ankles impaired her walking ambulation. She had asthenia and diffused abdominal pain. She didn't have any history of repeated episodes of tonsilitis. The patient had a diagnosis of rheumatic fever in another city and used benzathine penicillin for 6 months before referral.
Results: On admission she had fever, pallor, finger clubbing and watch-glass nails. Heart rhythm was regular with a murmur of mitral insufficiency and a murmur of mitral stenosis. The abdomen exam showed splenomegaly. She had oedema and pain with movement of 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 which histopathology was compatible with myxoma. She was discharged from hospital on medication. Four months after the surgery she was readmitted 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. Post-operative was uneventful and the patient was discharged from hospital 7 days after surgery.

Abstract no: 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 randomized 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 7 months (range = 1 - 32 months). Primary end point was a composite end point of death, hospitalisation and referral for surgery.

Results: Fourteen (35%) patients in the conventional arm and 10 (25%) patients in the betablocker arm had reached primary end point (p=ns). Worsening of heart failure occurred more commonly in the conventional treatment arm compared with propranolol treated arm (27.5% vs. 5% respectively; p=0.015). Two patients in the conventional treatment arm and in 1 patient in 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, hospitalization or need for surgery.

Abstract no: 739

Right ventricular outflow mass in a newborn with pulmonary atresia: Echocardiograph and histopathological aspects

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Background: Typical tumours of the paediatric age group are fibroma, rhabdomyoma and teratoma. Non-neoplastic mass may consist of thrombus and infections, which can be defined by a pathological examination.
Methods and material: The authors describe a 5-month-old female child that had suffered from progressively increasing cyanosis since birth observed by her mother. When she was 5 months of age it was observed a murmur in a paediatric consultation and she was referred for paediatric cardiology at the Brazilian Heart Institute.
Results: At admission her weight was 6.0kg, her height 61cm, heart rate 110bpm, O_2 saturation 73%, normal breath, cyanosis (+++), normal pulses, systolic murmur in left sternal border and no hepatomegaly. Electrocardiogram showed left ventricular overload. Chest radiography with 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 x 12mm) beneath the pulmonary valve with no mobility was observed. The ductus arteriosus was patent and the pulmonary arteries were well developed.
Conclusions: The child underwent surgery with the opening of the pulmonary valve and excision of the mass of the right ventricle outflow. The pathologic findings were a necrohaemorragic material with foci of calcification consistent with thrombus. The child was discharged from the hospital 20 days after surgery in good condition. The association between cyanotic heart disease and thrombus which is related to slow pulmonary blood flow rather than degree of cyanosis or coagulation abnormalities has been shown.

Abstract no: 742

Causes of hempotysis 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 tried to analyze the predictors of haemoptysis in patients with Eisenmenger syndrome and the underlying causes using computerised tomographic pulmonary angiogram (CTPA).
Materials and 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 1 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 6MWT. The only statistically significant finding was the reduced 6 MWD in patients with haemoptysis (323.8±81.7 meters) as compared to patients without haemoptysis (385.2±92.6 meters) (p=0.03). The CTPA was abnormal in 13 patients and normal in 4 patients. The most common extraparenchymal lesion was the presence of collaterals (5 patients). The more described cause of haemoptysis – pulmonary thrombus was seen only in one patient. One patient had a pseudoaneurysm from a branch of left pulmonary artery, which was closed with a coil. One patient was diagnosed to have military tuberculosis. The most common intraparenchymal lesion was the presence of mosaic pattern suggestive of recent pulmonary hemorrhage. Overall 7 patients underwent a therapeutic procedure based on the finding of finding on 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.

Abstract no: 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.
Aim: To analyse the results of mitral valve repair in the correction of mitral regurgitation in children and adolescents with rheumatic heart disease.
Methods and material: Medical records from 29 patients with mitral regurgitation submitted for mitral valve repair from 2002 - 2011 at the Brazilian Heart Institute (IMIP) were reviewed and supplemented by physical examination and echocardiography. Age ranged from 6 - 16 years with a median of 10.9 years.

Post-operative period median was 6.21 years with at least 1 year and no more than 10 years. Four (13.8%) patients still hadsevere mitral regurgitation post-operatively: 2 required mitral valve replacement, and the 2 others are on clinical follow-up. Two other patients required valve replacement, one due to several mitral stenoses and the other due to major haemolysis. With the exception of 4 patients who had to be reoperated, all the others were in functional class I (New York Heart Association). 6 (20,7%) patients had moderate mitral regurgitation and 17 (58.6%) mild mitral regurgitation. In 8 (27.6%) patients mild mitral stenosis was detetected, 8 (27.6%) showed moderate mitral stenosis and 2 moderate to severe. The left ventricular systolic function was decreased in 4 (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 post-operative period.

Abstract no: 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. 66 adolescent females with AN and 21 adolescent female controls exercised on a semi-recumbent ergometer in 3 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 3 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 (VO2), minute ventilation (VE), respiratory exchange ratio (RER), and arterial-venous oxygen difference (a-vO2) were determined using open circuit spirometry.
Results: Patients with AN had a significantly lower BMI (16.7 vs. 19.7kg/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 minutes, $p<0.001$), peak VE (47.4 vs. 72.0 L/min, $p<0.001$), and VO2 (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 anorexia nervosa vs. controls. HR, FS, MVCFc, and CI showed no difference at pre-exercise, increased with exercise, and were lower at peak exercise in anorexia nervosa vs. controls. WS decreased with exercise and was lower in anorexia nervosa 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-vO2 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 as compared to controls.

Abstract no: 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.
Methods and material: The authors describe the clinical aspects of a 112 children and adolescents admitted to cardiology ward of the hospital Instituto de Medicina Integral Professor Fernando Figueira from 2004 to 2010. A survey was conducted of medical records for data collection. Median age was 10.4 years and median length of stay for acute rheumatic fever hospitalisation was 16 days. 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 was 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%).
Results: Because it is a reference hospital for paediatric cardiology there was a high incidence of carditis (96 cases corresponding to 85% of the patients) and heart failure (57 cases corresponding to 51% of the patients) with 13% of the patients requiring the use of amines during hospitalisation. Another cardiac parameter observed was the presence of 29 (26%) patients with mitral regurgitation, 9 (8%) with aortic regurgitation and 45 (40%) with both lesions. The incidence of arthritis 33 (29%) was low and only 7 patients (6.25%) had chorea. Subcutaneous nodules were rare as usual with only 2 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: In this abstract one can observe 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.

Abstract no: 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.
Aim: To determine whether pre-natal diagnosis (PreN) has neuro-developmental benefit in patients with complex congenital cardiac defects compared to post-natal diagnosis (PostN).
Materials and methods: Single centre retrospective review of all patients with complex congenital cardiac defects that had complex surgery before 6 weeks of age, from 2001 - 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 - 24 months of age excluding children with chromosome anomalies or syndromes affecting development (n=13). Two-year mortality was 14.8% (12/88); 1 patient lost to follow-up. Grouped score 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 [2.3mmol/l (SD 1.6) vs. 4.4mmol/l (SD 5.0), p=0.024 (t-test)] and lower median cardio-pulmonary bypass flow rates >10 minutes [100ml/kg/min (IQR=54-126) vs. 125ml/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)], though mean scores were similar. There was no difference in motor delay. No NDO were significantly different after propensity match analysis.
Conclusion: Patients with pre-natal diagnosis of complex congenital cardiac defects had significantly lower pre-operative lactate levels with lower prevalence of cognitive delay. Longer follow-up and larger cohorts might lead to more significant beneficial effects of pre-natal diagnosis on ND.

Abstract no: 765

Pulmonary hypertension in the presence of an aortopulmonary window in patients living at high altitude do not affect morbi-mortality outcomes

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Background/hypothesis: Aortopulmonary window is a rare abnormal congenital communication. Early surgical correction should be offered before the development of irreversible pulmonary hypertension.
Aim: To present the morbidity and mortality in the surgical correction of this pathology in patients with and without pulmonary hypertension who live at high altitude.
Materials and methods: Case review of 17 patients who underwent surgery from January 1990 - March 2012 at 2 640 meter 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, this was diagnostic 41.2% (7/17).
Results: Global median age was 10.5 months (IQR 1.4 - 211), females 58.8% (10/17), pulmonary hypertension 41.2% (7/17), PDA most frequent cardiac associated anomaly 47.1% (8/17). Time of pump mean 64±18.4 minutes, aortotomy in 76.5% (13/17), goretex patch in 35.3% (6/17), low cardiac output and arrhythmias were observed in 52.9% (9/17). Mortality 5.9% (1/17). Survival at 30 days was 100%, follow-up after 10 years 37.5% (6/16). No statistical differences were found in pre-operative or pos-toperative variables in patients with or without pulmonary hypertension.

	Pulmonary hypertension		p-value
	Without n=10	With n=7	
Age in months	7.2 (2.7 - 13.9)	11.3 (6.5 - 21.3)	0.24
Weight in grams	5.2 (4.7 - 7.5)	8 (5.3 - 42.5)	0.1
Pre-operative ejection fraction %	71.7±10.7	69.1±8.9	0.65
Mortality number	0	1	0.7
ICU median days stayed	3 (2 - 7)	2 (2 - 5)	0.8

Conclusions: Pulmonary hypertension in the presence of an aorto pulmonary window lesion in children seems not to behave different from other typea of lesions with left-to-right shunt, mortality is low and apparently not related to these findings.

Abstract no: 768

Infective endocarditis: Improving positivity of modified Duke's criteria in developing countries

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Background: Successful management of infective endocarditis (IE) depends upon timely accurate diagnosis both aspects 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.
Patients and methods: A retrospective 10-year chart review of children younger than 14 years of age, suspected or diagnosed with infective endocarditis was done.
Results: Fifty patients were diagnosed with 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 history of prior antibiotic use. Single blood culture was positive in 42%, 2nd and 3rd blood cultures were positive in 30% and 10% respectively. 84% had vegetations on echocardiography. Thrombocytopenia was seen 28%. Modified Dukes criteria were positive in 62%, probable in 16% and rejected in 22%. By including C-reactive protein and ESR (raised in 80% and 60% respectively) diagnostic sensitivity was improved. We further modified Duke's criteria to include 1 positive blood culture and thrombocytopenia to evaluate if it further improved positivity. Positivity of Dukes criteria improved to 70%, possibility and rejection reduced to 10% and 18% respectively. All 9 rejected by the modified Dukes 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 remains the mainstay of diagnosis. Modified Dukes criteria may have to be further relaxed to improve diagnostic application in such situations and we have found that using 1 positive blood culture with known organism and thrombocytopenia may be helpful.

Abstract no: 782

Higher incidence of endocarditis in bovine jugular vein grafts compared with cryo-preserved homografts

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Background: Both cryo-preserved 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 from 2000 - 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 43.1 months, range 1 day - 50 years), median follow-up 40.1 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 1 with CH (0.75%; p=0.0009) occurring at a median age of 12 years (range 6 -21 years) and median time post conduit implantation 44 months (20 days - 10 years). BC were positive in 17 patients (Staphylococcus aureus (11), Streptococcus viridans (2), Cardiobacterium hominis (2), Staphylococcus epidermididis (1) and Hemophilus parainfluenzae (1)). Conduit replacement was required in 14/22, 3 patients had recurrent EC of the revised conduit. EC(+) patients had significantly reduced freedom from reoperation at 1.5 and 7 years: EC(-): 95.7±1.2%, 80.0±2.7% and 69.2±3.5%; 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 is associated with a significantly higher incidence of bacterial endocarditis. Furthermore, endocarditis of the BJVB is associated with conduit deterioration and more frequent reoperation. This information may be useful in the decision about which conduit to use for RVOT reconstruction in children with congenital heart disease.

Abstract no: 783

Long term cardiac function after anatomical repair and functional repair in corrected TGA and TGA

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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 long term. Furthermore, physiologic mechanism failure of the systemic right ventricle (sRV) in ccTGA is not fully understood.
Objective: To evaluate cardiac function in patients with ccTGA long after anatomic repair and functional repair.
Methods: Retrospective review of the catheterisation data from 2005 - 2011 was carried out in consecutive patients older than 16 years of age with ccTGA and complete TGA. Patients were divided into 4 groups; in TGA, atrial switch group (n=12) group and arterial switch group, in ccTGA, 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 4 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 atrial switch group (anatomical LV) in TGA (60±6 vs. 44±77%, $p<0.01$). In ccTGA, however, EF of the LV in 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 arterial switch group (45±8 vs. 61±11%, p=0.017). CI was also significantly lower in DSO group compared to arterial switch group (2.3±0.4 vs. 3.2± 0.6, $p<0.05$).
Conclusion: LV and RV function after DSO are compromised. Systemic RV function in conventional Rastelli group is also compromised.

Abstract no: 784

The comparative study: Right ventricular assessment in post Tetralogy of Fallot patients by echocardiogram with cardiac MRA

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Background: Post ToF repaired 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 (cMRA) is considered the clinical reference method for RV assessment. Echocardiogram is an alternate tool for evaluating cardiac anatomy and function.
Objective: Assessment of the RV parameters by using Echocardiogram compared to cardiac MRA.
Methods and material: Twenty patients (mean age 14±2 years) after ToF repaired were recruited from June 2011 - March 2012. The RV was evaluated by cardiac MRA and followed by echocardiogram. 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 and the cardiac MRA parameters were Right Ventricular Ejection Fraction (RVEF), Right Ventricular End Diastolic Volume index (RVEDVi) and severity PR. Comparative analysis were assessed by Pearson's sample correlation coefficient, Crosstab Kappa, sensitivity and specificity of area RVEDi from ROC curve analysis.
Results: There is a correlation between RVEDVi to area 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). 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 ≥20.43cm²/m² from echocardiogram was correlated with RVEDVi ≥160ml/m² from cardiac MRA (sensitivity 64% and specificity 83%) from ROC curve analysis.
Conclusions: Echocardiogram is an effective tool for RV evaluation in TOF with PR. Measurement of area RVEDi, FAC, TAPSE and degree of PR correlate well with cardiac MRA parameters.

Abstract no: 787

Residual pulmonary stenosis has the impact to prevent pulmonary regurgitation and right ventricular dilatation in the 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 3 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 repaired ToF group was 24.6±22.8 (0 - 109)mmHg. While there was no significant difference in W1 between the 2 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. In the repaired ToF patients, multivariate regression analysis demonstrated that pulmonary wedge pressure and PR independently increased W2, while pulmonary stenosis reduced W2 (standardised ß: 0.40, 0.39, -0.30, p=0.0024, 0.012, 0.016, respectively).
Conclusions: The results indicated that residual PS serves to reduce PR, while left ventricular diastolic dysfunction, represented by the high pulmonary wedge pressure, increases it. These data raise potential caveat to perform angioplasty to relieve PS, and also suggest importance of preservation of left ventricular diastolic function to improve long-term outcome of repaired ToF with residual PR.

Abstract no: 799

Paediatric 3-D echocardiography facilitates decision-making in management of congenital heart diseases

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Background: The improved image quality and ease of use of current real-time 3-D echocardiographic (3-DE) systems promotes incorporation of study into routine use. The conditions which would benefit most from 3DE are evolving and is being defined. We describe conditions for which 3-DE provides information and that has an impact on decision-making in the management of congenital heart diseases.
Materials and methods: During 2009 - 2012 patients at the University's referral medical centre were assessed via 2-DE and additional 3-DE using Philips iE33 3-DE system with either 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 3-DE was due to inconclusive or questionable information from standard 2-D 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 2-DE studies, 3-DE were performed for more information required in 5% of studies. The patients were male 55% with age ranging from birth to 15 years old. Incremental information obtained by 3-DE included: clarification of septal morphology and the 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 3-DE 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. 3-DE findings were in concordant with surgical findings in 98% of cases. 2-DE would miss the important information in 7% of studies.
Conclusion: Paediatric 3-DE provides incremental information to facilitate decision-making in improved management of congenital heart disease.

Abstract no: 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), left–to-right shunt, leads to the activation of compensatory mechanisms. If these mechanisms excessively activated, cardiac remodelling will occur characterised by changes of dimensions and function of left ventricle. Carvedilol, a novel third-generation non-selective ß-blocker, can postpone this ventricular remodelling.
Objective: To determine changes of dimensions and function of 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 by using echocardiography for 3 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 9 (30%) PDA. There were significant differences between the 2 groups. The mean of changes of 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 on E/A ratio 0.07 (0.32) vs. 0.03 (0.31), p=0.71 between 2 groups.
Conclusion: There were significant changes on LVIDs, LVIDd, LVPWd, IVSd, volume, mass, ejection fraction, and the shortening fraction, without any difference on E/A ratio, in children with CHD-left-to-right shunt and heart failure post carvedilol therapy.

Abstract no: 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 from 1991 - 2011 were identified from the departmental database. Subjects were included if they underwent repeat catheterisation either pre-Fontan 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. 7/16 (44%) had a systemic right ventricle. There were 10 patients pre-Fontan and 6 patients post-Fontan completion who had had repeated assessment. All patients were treated with ACEI; 2 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 13mmHg (IQR 12-15.8mmHg) to 10mmHg (IQR 8.3-12.5mmHg) was demonstrated (p=0.001), as well as a fall in mean atrial pressure from 11.5mmHg (IQR 9.9-13.1mmHg) to 9.2mmHg (IQR 7-10.6mmHg) (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 demonstrates 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.

Abstract no: 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 exist.
Aim: 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.

Materials and methods: Portable echocardiography was performed on 1 087 predominantly Caucasian children aged 5 - 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 1 morphological abnormality of the mitral valve (MV); a thickened anterior MV leaflet (defined as ≥3mm) was most common (19 children;. 214 children (19.5%) had some degree of mitral regurgitation (MR) with the majority reported as trivial; 11 children had MR jets ≥2cm seen in at least 1 view; and only 2 children had MR which met all 4 WHF criteria for significant MR; Aortic regurgitation (AR) was found in 23 (2.1%): 5 children had AR jets ≥1cm, 3 of whom had bicuspid aortic valves; there were no cases of mitral or aortic stenosis; congenital abnormalities were detected in 16 children (1.5%); none of the children met the WHF criteria for Definite RHD; and 5 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.

Abstract no: 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 amongst the highest prevalence rates of rheumatic heart disease in the world, as documented through local registers of clinical cases, but an epidemiologic survey has never been undertaken.
Aim: To establish the prevalence of RHD in high-risk indigenous Australian children using the new 2012 World Heart Federation diagnostic criteria.
Materials and methods: Portable echocardiography was performed on 4 158 predominantly Aboriginal and Torres Strait Islander children aged 5 - 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 14/1 000 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 1/3 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.

Abstract no: 816

Western Australia's recent experience in delineating patent ductus arteriosus morphology by echocardiography prior to device closure

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Background/hypothesis: The Children's Cardiac Centre in Perth, Western Australia uses high quality cardiac ultrasound equipment in its day to day practice. Interventional management of PDA's 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.
Method: Retrospective review of 2 D echocardiograms of all patients with PDA's that 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 PDA's 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 PDA's. Decisions regarding choice of appropriate device can almost always be made prior to proposed interventional procedure. This has important implications with regards to limiting stock of expensive interventional devices required as well as to the process of informed consent when discussing the procedure with patients and their carers.

Abstract no: 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: Though a rare clinical entity, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) 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 inpatient unit with marked restlessness and irritability. He was inconsolable, had marked respiratory distress, cool extremities, central and peripheral cyanosis. The radial and brachial pulses were absent. The mean arterial pressure was 65mmHg, Heart rate of 160 beats per minute with a third heart sound. Laboratory tests showed an elevated CK-MB of 112.5u/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 6 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.

Abstract no: 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.
Method: Review of all cases who had established pulmonary hypertension and coarctation from January 2008 - January 2012.
Results: Three patients were identified: **Case 1:** A 22-year-old lady 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 8-year-old boy was referred for differential cyanosis and it was noted he had large PDA with COA. He underwent stent angioplasty of the COA like the previous patient. His PA pressure was systemic which normalised on follow-up. **Case 3:** A 7-year-old girl 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. Post-operatively 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 medically managed.
Conclusion: The PDA with COA is a protected circulation as the pulmonary blood is shunted in to the descending aorta thus the pulmonary vasculature is not exposed to high blood flow. So it is safe to close the PDA and stent the COA. The VSDs with coarctation have increased pulmonary blood flow and gets irreversible pulmonary hypertension early. However treating the coarctation will help them, but they need careful pre- and post-op management.

Abstract no: 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 recovers completely or does well for years. In this retrospective study we sought to describe outcome and to identify predictors of outcome.
Methods: Children presenting with DCM from 2005 - 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, those: Group 1 reaching a primary endpoint (death, heart transplantation (HTx) or mechanical circulatory support (MCS)); Group 2 recovering (left ventricular end-diastolic dimension(LVEDd) and SF<p95) or Group 3 with ongoing disease.
Results: 108 children were included [median follow-up 1.8 years (range 0 - 5.4)]. Twenty five (23%) children reached a primary endpoint (10 died, 3 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 year (0.03 - 1.1)] than the time to recovery [0.8 year (0.3 - 2.5)] (p<0.05). Fifty four (50%) children had idiopathic DCM, 21(19%) had myocarditis 15 (28%) children with idiopathic disease reached a primary endpoint, as compared to only 1 (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 per year), in contrast to those who recovered (-2.9 per 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 per year) ($p<0.05$).
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.

Abstract no: 841
Oxidative phosphorylation disorders among children with severe cardiomyopathy

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Background/hypothesis: Cardiac involvement occurs in 17 - 40% of children with oxidative phosphorylation (OXPHOS) disorders, but the incidence of OXPHOS disorders among children with primary cardiomyopathy is unknown.
Materials and methods: We analysed data of all children with documented cardiomyopathy who underwent OXPHOS testing from 1984 - 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, 1 (1.6%) was considered probable and 7 (11.3%) were considered possible. These included 13 of 38 (34.2%) of children with dilated cardiomyopathy, 5 of 11 (45.5%) of children with hypertrophic cardiomyopathy, 1 of 5 (20%) of children with left ventricular non-compaction; and 0/8 (0%) children with restrictive cardiomyopathy. Of 51 children without any documented extra-cardiac abnormalities, 8 (15.7%) were considered definite and 6 (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.

Abstract no: 847
Results of cardiac catheterisation and treatment of protein-losing enteropathy in children with HLHS after Fontan procedure

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Objective: Protein-losing enteropathy (PLE) occurs in 3% - 15% of patients with Fontan circulation. All require adequate medications, cardiac catheterisation (CC) and some re-intervention. We reviewed Cardiac Catheterisation Laboratory (cathlab) database at University of Cracow Childre's Hospital, to identify HLHS patients after a Fontan procedure (FP), who underwent CC from January 2001 - July 2012.
Results: At that time, 330 HLHS children were palliated using FP, with only 1 post-operative death. Of 21 patients subjected to CC, in 10 (50%) 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 3 months-7.5 years, x-2.6 years. Only in 1pt (with PLE recognised 3 months post-FP), no structural cardiac changes were found. Two patients revealed narrowed inter-atrial communication and required re-operation, 7 had significant left, and 1 bilateral pulmonary artery branch stenosis demanding balloon pulmonary angioplasty (BPA) (in 5pts with stents). Before BPA, the dimension of the most narrowed pulmonary artery branch ranged 1.5 - 6.2mm and after -5.8 - 12mm, x-9.1mm. Systemic venous pressure ranged 13-22mmHg, x-16mmHg, RVEDP: 4 - 13, x-7.6mmHg, Sa02: 88-99%, x-95.1%. The follow-up ranged 0.5-7.5 years, x-2.2 years. All patients were treated with diuretics (Furosemide, Spironol, Hydrochlorothizide), ACE inhibitors (Enarenal Captopril), and aspirin (1 also Warfarin), and received specific high protein MCT products-enriched diet, 3 additionally received steroids (2) and sildenafil (1). Significant improvement was achieved in 8/10, with protein levels increasing from x-42.4g/l±6.5g/l to 53.5g/l±9.5g/l.
Conclusions: Post-FP, the majority of HLHS and PLE patients may reveal various structural changes, which impede Fontan circulation. Management of PLE requires elimination of such changes and complex, mostly symptomatic treatment.

Abstract no: 849
Computational fluid dynamics in Fontan patients to evaluate energy loss during simulated exercise

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Background/hypothesis: In most patients with a functional single ventricle a total cavopulmonary connection (TCPC) is created. Exercise intolerance is common Fontan-patients. It has been suggested that energy loss (Eloss) inside the TCPC may play an important role in reduced exercise performance.
Aim: To establish whether Eloss inside the TCPC plays a significant role during (simulated) exercise.
Materials and methods: In 15 patients a 3-dimensional reconstruction of the TCPC was created using anatomical data from Cardiac Magnetic Resonance imaging (cMRI). Phase-contrast cMRI was used to obtain through-plane flow rates from inferior vena cava (IVC) and superior vena case (SVC) during rest and during

exercise, simulated with Dobutamine (7.5µg/kg/min) intravenously. Patients underwent cardiopulmonary exercise testing to assess maximal oxygen uptake. CFD-simulations were performed using the flow rates obtained by cMRI. Total energy loss (Eloss) inside the TCPC-structure was calculated for both conditions in each patient. A condition where only IVC flow was increased twofold compared to 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±0.37mW during rest, 1.07±0.64mW during simulated exercise and 2.97±2.49mW with twofold IVC flow. The correlation between cardiac index and Eloss was exponential (Rsq=0.393, p<0.000). The increase in Eloss depended on the specific anatomy of each patient. No correlation was found between the increase of Eloss during dobutamine-infusion and exercise capacity as assessed by maximum oxygen uptake.
Conclusions: Eloss inside the TCPC-structure is limited but increases with (simulated) exercise. This is due to increased blood flow, but also depends on patient-specific anatomy of the TCPC. We did not observe a correlation between Eloss and exercise capacity.

Abstract no: 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 with other congenital heart disease, extracardiac anomalies, and some genetic problems. Up-to-date, there have been a few reported cases. Most of them are case report with complex cardiac abnormality.
Material and methods: We detected 20 cases through the increasing awarenes of this anomaly in the last 3 years. Approximately 9 250 echocardiaograms performed in this period. We describe 20 cases with this anomaly with emphasis on the clinical features with genetic, cardiac and extracardiac abnormalities
Results: The ages of the patients ranged from 1 day - 13 years at the time of initial diagnosis. There were 10 feamale 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. Each of ASD and PS were detected in twelve of 20 patients (60%). Aortic arch abnormality such as right aortic arch and coarctation was seen in 6 patients. One patient had left persistent superior vena cava. 45% of the cases were associated with genetics syndrome (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 was studied in 11 patients. All of them had normal karyotypes and FISH results. Six patients underwent succesfull operation. Three of 20 patients died in the longterm follow-up. The remaining cases were clinically stable and were being followed without surgery.
Conclusions: CPA may be related to different congenital heart disease. Detection of CPA may be an important clue to the presence of structural heart disease and chromosomal abnormalities.

Abstract no: 851

Rare cases of left ventricular aneurysm in children: A single-centre experience

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Background: LV aneurysms in children are very rare and often results from a congenital defect in the posterior portion of the mitral valve annulus. We present 2 unique cases of children with tuberculosis (TB) and acquired sub-mitral aneurysms.
Case report: Case 1: A a-febrile 2-year-old girl with progressive dyspnoea, CHF and systolic murmur at apex for 8 days. CXR showed marked cardiomegaly with pulmonary venous congestion. Echocardiogram revealed a huge LV aneurysm at sub-mitral valve area causing moderate MS, severe MR and TR. EF was 50%. MRI showed a large pseudoaneurysm at posterior wall of 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 posterior leaflet and annulus. LV aneurismorhaphy and tricuspid valve repair were also done. The post-operative course was uneventful. Pathological examination suggests necrotising granulomatous inflammation. AFB from lymph node was negative. **Case 2:** A 2.5 year-old boy, with history of TB contact, presented with prolonged fever and dyspnoea. Physical examination revealed no sign of CHF or murmur. CXR showed mild cardiomegaly. He suddenly collapsed after complaining of severe epigastric pain immediate after administration. CPR was unsuccessful. Autopsy revealed a rupture of huge LV aneurysm at the sub-mitral valve area with massive intrapericardial blood clot, generalised lymphadenopathy, pleural effusion and ascites. Even though AFB from myocardial tissue was negative, pathological finding suggested granulomatous inflammation. TB endocarditis has been reported sporadically. Most cases presented as LV aneurysm at sub-mitral valve area.
Conclusions: Patients with LV aneurysm are at potential risk for life-threatening conditions leading to sudden death. Echocardiography and MRI are crucial modalities to define morphology and extent of the aneurysm and surgical planning. Management should be individualised and mainly directed toward early diagnosis to prevent moribund outcomes.

Abstract no: 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 <5 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 1 - 2 years, and 80% of cases present before the age of 4 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 9-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. A 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 haemiplegia 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 previous undiagnosed myocardial infarction is presented. With early treatment, these complications are rarely seen.

Abstract no: 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 have performed clinical and genetic investigations in 8 Saudi Arabian families with a history of unexplained sudden deaths of children. Additionally, we also investigated the pathology of repeated intrauterine fetal deaths in 2 families.

Methods and results: Clinical symptoms, ECG phenotypes and genetic findings led to the diagnosis of LQT1 in 4 families (recessive) and LQT2 in 4 families (3 recessive, 1 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 intrauterine stages of life in the recessive LQT2 patients. LQT1 causal mutation c.387 -5 T>A in the KCNQ1 gene was detected in 3 families. LQT2 causal mutation c.3208 C>T (p.Q1070X) in the KCNH2 gene were identified in 2 families. In one family with sudden death of 5 siblings, we identified an unclassified variant c.1179 G>T (p.K393N) in the KCNQ1 gene. This variant is present in heterozygous form in about 2% of the healthy Arabs, but not in Caucasian healthy 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 1st line of genetic investigation before proceeding to comprehensive screening for all LQTS causal genes. Further, c.1179 G>T (p.K393N) in KCNQ1, though 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.

Abstract no: 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. Biomarkers for congestive heart failure are becoming more available and 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 levels done. A detailed echocardiogram was done. Modified inotrope score (MIS) was calculated for 1st 5 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 grams. 50% 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.

Test	Abnormality
ProBNP	>1 000pg/ml
Fractional shortening	<29%
Myocardial performance index	>0.38
TDI E/e'(lateral)	>8
Mitral spectral Doppler E/A	<1 or > 1.8

Twenty six newborns with poor systolic and diastolic function had ECG abnormalities and only 2 with normal function had the same (p=0.008). ECG was a poor predictor of isolated diastolic function abnormalities.

TABLE 1: Echocardiography and serum proBNP

	Systolic function	Diastolic function	ProBNP/MIS	Number
Group 1	Good	Abnormal	Abnormal/>1 000	4 (10%)
Group 2	Good	Abnormal	Normal/<1 000	5 (12.5%)
Group 3	Poor	Abnormal	Abnormal />1 000	22 (55%)
Group 4	Poor	Abnormal	Normal/<1 000	4 (10%)
Group 5	Good	Normal	Normal/<1 000	1 (2.5%)
Group 6	Good	Normal	Abnormal/>1 000	4 (10%)

Sensitivity	70%
Specificity	20%
Prevalence of poor function	87.5%
Positive predictive value	87%
Negative predictive value	8.7%

Conclusions: The serum natriuretic peptides are a useful tool to assess cardiac failure in the sick newborn and need to be more widely used. Available but underutilised tests like ECG can also help in early diagnosis of cardiac failure in the newborn.

Abstract no: 862
Evaluation of cardiac autonomic function by 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 1 - 17 years admitted to the emergency department with acute CO poisoning and 40 healthy age- and sex-matched controls. Carboxyhaemoglobin (COHb) and cardiac enzymes were measured at admission. 24-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 discharge.
Results: Time domain indexes (SDNN, SDANN, rMSSD, SDNN index and NN50) are similar between patient and control groups (p>0.05). Among the frequency domain indexes, mean HF level of the CO poisoning group was higher than controls (p=0.018) while LF levels were similar (p=0.636). LF/HF ratio was significantly lower in 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, Pd values were higher in CO poisoning group (p<0.05). On discharge, QTd and QTcd were still longer in CO poisoning group than the controls (p<0.05).
Conclusions: Frequency domain indexes recorded within the first 5 minutes on admission, especially 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.

Abstract no: 870

Does measurement 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 US regarding the benefit of screening oxygenation saturation (O_2Sat) prior to newborn discharge. We sought to determine if assessing O_2Sat would increase the sensitivity of CHD diagnosis during humanitarian, population-based paediatric screening.
Methods: During a humanitarian screening in 2 rural provinces in Mongolia (Mandal Gobi and Dalanzagdad), children from 1 month - 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 pathologic murmur on examination or oxygen saturations <94% prompted echocardiographic screening (ECHO) by a paediatric cardiologist.
Results: Previously unrepaired CHD was identified in 61 of 822 patients (7.0%) screened over 4 days with 133 ECHOs performed (16.1%). Identified cardiac defects included: 19 haemodynamically insignificant ventricular septal defects (VSDs), 9 haemodynamically significant VSDs, 10 patent ductus arteriosii, 8 atrial septal defects and 2 Tetralogy of Fallot (ToF). Remaining lesions were only minor valve abnormalities. Forty nine patients (6%) had O_2Sat <94% on screening. Of these patients, 7 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 1 patient had oxygen saturation <94% with a negative auscultatory exam (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 add little to the diagnostic accuracy of physical examination alone, to warrant its use as a tool for population-based screening of paediatric patients.

Abstract no: 873

Effectiveness of indomethacin in full term infants with symptomatic patent ductus arteriosus

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Background: Common treatment methods of 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 pre-term neonate with significant PDA. However, there is controversy of effectiveness of indomethacin in full term neonate with significant PDA. Therefore, we attempt to evaluate the effect on indomethacin treatment to patients with significant PDA among full-term infants with birth weight (BW) ≥2 500g and a gestational age (GA) ≥37 weeks.
Methods: We retrospectively reviewed 29 infants with significant PDA and a BW of >2 500g and a GA of 37 weeks> who were admitted to Chonnam National University Hospital from 2007 - 2009. During indomethacin therapy, feeding was prohibited and water intake restricted (60 - 80ml/kg/day). Indomethacin (0.25mg/kg/day) was intravenously administrated as a single dose at 12 - 24-hours intervals. Patents 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: Indomethacin treated 13 (44.8%, responder) of 29 patients with PDA were completely closed. Eight (27.6% partial responder) were incomplete close but clinical symptoms such as congestive heart failure were improved without any particular treatment.
Conclusions: Indomethacin therapy might be a useful medical treatment option prior to considering surgery for PDA in full term infants.

Abstract no: 874

Rheumatic heart disease in Namibia: 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.
Aim: To collect data to assist with resource development and distribution with the end objective of reducing the prevalence, morbidity and mortality from a much neglected disease.
Materials and methods: This is a prospective, national, hospital-based registry of patients referred to the RHD Clinic at Windhoek Central Hospital. Questionnaires document patient's presentation, clinical course, investigations, complications, management and demographic at enrolment. The study was initiated in July 2010 in collaboration with the Global Registry for RHD (REMEDY).
Results: 193 patients are enrolled with the distribution of cases reflecting regional population density. 58% are male and 42% female, 81% from 10 - 40 years and 5% <10 years. 32% have severe disease (NYHA III - IV). The mitral valve is most commonly affected (77% with MR, 40% with MS) followed by tricuspid then aortic valve disease. 19% have 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 the preceding 6 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 amongst 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.

Abstract no: 876

Echocardiographic pattern and severity of valve abnormalities in children with rheumatic heart disease in Uganda

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Background: Rheumatic heart disease (RHD) is the commonest acquired heart disease in children worldwide but in Uganda, data is scarce regarding its morbidity and mortality. The disease has a progressive course and patients will usually require valve surgery.
Objectives: (1) To describe frequency of the different heart valves affected; and (2) To describe the relationship of the disease severity with regards to age and sex.
Methodology: 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 - 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. Twenty eight (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 1 (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 higher in females (p value=0.04), while severe AR was significantly more in males (p value=0.007). There was no difference in disease severity between children <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.

Abstract no: 877

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) have 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 was affected by high-dose beta-blocker therapy in children and adolescents with HCM.
Materials and methods: The study included 14 patients (2 females), mean age 9.2 (range 1.8 - 17.7) years, with familial HCM and moderate to severe hypertrophy. They were paired in 7 untreated and treated pairs (treatment: metoprolol or propranolol 5 - 20mg/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 1 year. Ultrasound examinations were performed with Philips IE33 and analysed with 3-D QLab advanced software (version 7). We took 3-D 4-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%> left ventricular end-diastolic volume/m2 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) vs. 27.9 (18.4 - 39.4) which did not reach significance (p=0.08). Resting heart rates were not different, 76 (53 - 95) vs. 76 (56 - 94). Calculated resting cardiac output was non-significantly 32% higher in the beta-blocker group, 2.13 (1.41 - 3.31) vs. 1.61 (1.13 - 2.01)L/m^2 BSA for untreated group (p=0.07).
Conclusions: Beta-blocker therapy does not reduce cardiac output at rest, and the trend for improvement suggests that the increase in resting left ventricular volumes is beneficial.

Abstract no: 878

Outcomes of patients with Tetralogy of Fallot with absent pulmonary valve syndrome: 37-years' 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 tracheo-bronchial 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 from 1975 - 2012 was conducted. The median age and weight at repair were 0.9 years (4 days - 24.2 years) and 6.9kg (1.8 - 56kg) respectively. Pre-operative intubation was required in 15 (30%) patients and 21 (41%) required urgent surgery. 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%); 2 (4%) underwent a Lecompte manoeuvre.
Results: Operative mortality was 14% (7/51): 1975 - 1989:19% (3/16); 1990 - 2000: 19% (4/21); 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% versus 88% and 81% in early and middle eras), with better ante-natal diagnosis (64% vs. 6% and 19% respectively). Overall survival at 5, 10 and 20 years was 81.4±5.6%. On multi-variate 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 5, 10 and 20 years was 79.7±6.9%, 69.4±8.2% and 52.1±9.8%. No difference was found between conduit, monocusp or valveless techniques. Risk factors for late re-operation include prematurity (p=0.001) and neo-natal primary repair (p=0.007). Longer post-operative 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 post-operative ventilation and mortality.

Abstract no: 879
Comprehensive imaging tools: 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 high mortality. RI often presents in association with severe cardiac malformations, isomeric arrangement of the bronchi and abnormalities of 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 post-natal 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 where 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.

Abstract no: 881
Exome sequencing as part of a strategy to identify important variants in congenital heart disease

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Background: Exome sequencing is emerging as a strategy to identify potentially causal variants in families with 2 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. Sporadic forms of congenital heart disease constitute the vast majority of cases.
Methods: Commercial exome sequencing with screening out of common variants was performed using usual bioinformatics 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 modelled in vitro to assess functional impact.
Conclusions: Emerging sequencing technologies need to be paired with powerful bioinformatic 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.

Abstract no: 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 was aimed to investigate serial change of NT-proBNP as a prognostic factor for outcomes of paediatric dilated cardiomyopathy (DCMP) in a single tertiary centre.
Materials and methods: Retrospective review included 69 DCMP patients who were treated at Samsung medical centre from 2004 - 2011. Echocardiographic measurements and NT-proBNP were serially analysed at the time of diagnosis, 3-months, 6-months, and last follow-up. They were classified into 3 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 of 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 level showed statistically significant differences between 3 groups. The levels of NT-proBNP elevated significantly higher in Group III at 3-month, 6-month, last follow-up except at diagnosis. And 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 6-months (p=0.040) was a significant predictor of adverse outcome. Patients with the degree of improvement from diagnosis to 6 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 of 6-months from diagnosis could predict adverse outcomes in paediatric DCMP and could be used a guide for a long term treatment plan.

Abstract no: 886

Chest ultrasound in evaluation of post-operative lung oedema in children with congenital heart disease

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Background: Post-operative 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 hypoxemia. 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 echo dense 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 of the newborn. Accordingly, we explored whether C-u/s can be used to assess post-operative LWC of patients with CHD.
Patients and methods: We studied 6 patients with TGA and 8 patients with acyanotic shunt defects (0.2 - 0.7 and 2.0 - 6.2 months of age, respectively). The measurements were done at <2 and <30 hours post-operatively. Static lung compliance (L-Cst) was measured under anaesthesia by the double occlusion technique (Lab manager 4.52i; Erich Jaeger GmbH; Germany). C-u/s was performed using an 18-MHz linear transducer (Esaote, Italy) at bilateral transverse sections of 3 designated segments. The digital images were blinded, the proportional area of the B-lines in each section was graded from 1 (0%) - 5 (>75%), and a mean chest sonographic score (C-SS) of the 6 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 post-operatively (p=0.01) and on first post-operative day (p=0.01) than patients with shunt defects. L-Cst did not significantly differ between patient groups. (Table).]

	Patient group	N	Median	Range
Post-operative (1-2h) C-SS	TGA	6	2.51	2.13 - 4.00
	Shunt defect	8	1.78	1.00 - 3.78
1st post-operative day C-SS	TGA	6	3.00	2.17 - 3.70
	Shunt defect	4	1.61	1.29 - 2.11
Post-operative (1-2h) L-Cst	TGA	5	6.3	5.6 - 7.3
	Shunt defect	6	5.15	4.7 - 10.2
1st post-operative day L-Cst	TGA	5	5.20	4.10 - 7.44
	Shunt defect	1	7.10	7.10 - 7.10

Conclusions: Measurement of LE by ultrasound is a quick, easy and safe procedure, which may prove to be useful in post-operative evaluation of patients with CHD. Complex open-heart surgery or significant hypoxemia, or both, may increase and prolong post-operative LWC.

Abstract no: 890

Insight into dynamic 3-D 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 young population.
Methods: 40 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. RT3-DE data set was cropped into 15 slices (spaced 24°) 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 3-D graph using our customised software (MathWorks Inc). Measurements included annular area, bending angle and height, and tethered and prolapsed volume of the leaflets, and antero-lateral (APM) / postero-medial PM (PPM) angle.
Results: There was a strong correlation between the annular bending angle and the height throughout all phases of cardiac cycle (range of p-values 0.007 to p<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 of 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.

Abstract no: 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: 169 patients with KD and CAA (1999 - 2012) were reviewed. 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 1/5 years after acute KD the proportion of CAA showing resolution were: 57% - 78% and 90% - 100% depending on coronary branch. For giant CAA (n=51), proportion of CAA showing resolution at 1, 5 and 10 years were 0 - 7%, 15% - 34%, 40% - 73%. Patients with small/large CAA were not at risk for either thrombosis/stenosis. Patients with giant CAA were at substantial risk [20% (2 months), 33% (5 years) and 47% (10 years)] despite 82% on anticoagulation, 43% of thrombosis had important consequences [thrombolytics (7), myocardial infarctions (3), death (1)]. Freedom from stenoses was 96%/77% at 1/5 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, high erythrocyte sedimentation rate, C-reactive protein and neutrophils at 3 months and 1 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 more likely in younger patients. Despite aggressive thromboprophylaxis strategy, patients with giant CAA continue to be at substantial risk of thrombotic and stenotic complications.

Abstract no: 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 if 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 Illumine Golden Gate® 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 7 SNPs known to be associated with increased venous thrombosis risk, including 1 known to affect fibrinogen levels and 3 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 rs 5982 (19% vs. 6%, p=0.04), GG polymorphism in fibrinogen alpha chain rs 2070006 (62% vs. 38%, p=0.02), CC polymorphism in coagulation factor V rs 3753305 (31% vs. 17%, p=0.05), and AG/GG polymorphism in plasminogen rs 13231 (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.

Abstract no: 902
Eisenmenger in infancy: Is it triggered by combined pressure-volume pulmonary blood flow rather than increased pulmonary venous pressure alone? Immediate and midterm normalised pulmonary artery pressures in a 6-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 2 years, failure of

normal regression of the intimal smooth muscles occurs. We report on a 6-year-old child from Togo with severe pulmonary hypertension due to an untreated cor triatriatum.
Material: 6-year-old boy from Togo. At rest slight tachypnea of 40 - 45/minutes, thoracic deformation (cardiac voussure), saturation in room air >96%.
Methods: Echocardiography with diagnosis of a cor triatriatum, severe pulmonary arterial hypertension (TI gradient 150mmHg, BP 100/45mmHg, gradient over membrane of cor triatriatum 55/15mmHg). No atrial or ventricular septal defect. At mild exertion (walking to outpatient clinic) 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 6 weeks improved physical performance with no desaturation while walking. Echocardiography with no evidence for 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 - 48 months a fixed pulmonary hypertension these patients obviously have a great potential to recover from pulmonary hypertension regardless their age.

Abstract no: 903

Increase in use of pooled human anti-thrombin replacement therapy in paediatric patients

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Background: Anti-thrombin is an essential part of the coagulation system. A number of paediatric patients including those with congenital heart disease may have low anti-thrombin activity. For patients with critically low anti-thrombin activity, endogenous human anti-thrombin from pooled donors is available for replacement therapy. We sought to determine current trends in the use of anti-thrombin supplementation in paediatric patients.
Methods: Hospital records of all patients who received anti-thrombin supplements at The Hospital for Sick Children from 2002 - 2011 were reviewed. Indication for anti-thrombin 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 anti-thrombin 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 anti-thrombin 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 anti-thrombin level was 46% (25th - 75th percentiles: 32% - 61%). Number of patients receiving anti-thrombin increased from 33 in 2002 to 81 in 2011 (+5 patients/year, p=0.002). During this period, use of anti-thrombin for cardiac patients not on ECMO remained stable (+0.6 patient/year, p=0.38) as did use for non-cardiac patient (+0.4 patient/year, p=0.16). However, use of anti-thrombin supplementation in the setting of ECMO significantly increased (+3 patients/year, p=0.02) during the study period.
Conclusions: Anti-thrombin 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.

Abstract no: 905

Intra-cardiac 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 2 tertiary centres for all children with DCMO.
Method: Retrospective review of paediatric cardiology databases. All DCMO patients between 0 - 14 years seen from January 1983 - December 2010 at Chris Hani Baragwanath Academic Hospital (CBAH), Johannesburg and from January 2003 - December 2011 at Inkosi Albert Luthuli Central Hospital (IALCH), Durban, both tertiary institution 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 1 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 anti-coagulated 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%.

Abstract no: 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 male preponderance (8 girls, 25 boys). Twenty five patients were of different ethnic background with majority being South Asian or British Asian (11). All patients had carditis of varying degree manifest predominantly with valvulitis. Mitral and or aortic regurgitation were 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 form of mitral valve repair or replacement (4 patients) and 1 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 8 weeks. Twelve patients had been undertreated with steroids before. Secondary prophylaxis with oral Penicillin over a follow-up period of mean 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 being rare.

Abstract no: 909

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/hypothesis: 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.
Materials and methods: The Barwon Infant Study (n=1 250) is a regional non-selected Australian birth cohort. Aortic IMT is measured on 4-week-old infants by 2 trained research staff using trans-abdominal ultrasound (GE vivid ITM with vascular probe). A subset of babies has aIMT performed by both staff to assess inter-observer consistency. Data was 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 (a) sonographers and (b) analysts using Pearson's correlations, Bland-Altman plot and 2xn Chi2 analysis.
Results: Among the first 292 babies, aIMT approximated a normal distribution (mean 0.564mm, sd 0.06mm). The inter-observer correlation of aIMT measurements from infants scanned by both sonographers (n=17 - date) was 0.8449, p <0.001. The inter-operator correlation between measurements of aIMT made by both analysts (n=115 - date) was 0.8783, p<0.001 (mean difference 0.004mm, 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 is reproducible and unaffected by environmental and behavioural factors examined. (These are interim data. Data on 500 subjects will be available by February 2013.)

Abstract no: 915

Comparison of echocardiographic 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.
Materials and methods: From the complete Swedish national material of patients that 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 Götland 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 ≥3cm has a low sensitivity of 33%, and a relative risk of 2.9 (5%CI

1.5 - 5.2). Two paediatric wall thickness criteria are much better: a wall thickness ≥2cm, 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 ≥2cm. 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 x 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.

Abstract no: 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 - 11.3 per 1 000 live births. We report the findings on the spectrum of echocardiographically diagnosed CHDx from 3 different centres across Nigeria.
Methods: Over a period of 42 months children who were referred for echocardiographic evaluation in the centres located in 3 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, Illinois, U.S.A.).
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 1st 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 perimembranous, 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 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.

Abstract no: 924
Successful management of faucial diphtheria 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 2nd 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 5-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 delirium 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 4th day, she had myocarditis as a complication. Methylprednisolone 4mg/kg/times was administered and then continued at 3mg/kg/day for a week. The 1st 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 2nd to 4th routine throat swab culture showed no more C. diphtheriae. The FNAB from the persistent bull neck found a neck abscess. Incision and drainage were performed. After 1 month of treatment, she was given DPT immunisation and discharged.
Conclusion: A complicated case of diphtheria required comprehensive management to achieve an optimum outcome.

Abstract no: 927
Persistent 5th aortic arch: A clinical conundrum

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Background: Persistence of the embryonic 5th aortic arch is a rare and enigmatic condition with variable anatomical forms and physiological consequences. As such, it may be significantly under-diagnosed as a primary pathology.
Methods and results: A search of the surgical database revealed 5 cases of persistent 5th aortic arch (PFAA). Four cases were diagnosed with congenital heart disease antenatally; 1 case presented at 6 weeks of age. In 2 cases there was isolated PFAA with significant left-to-right shunting causing heart failure. In another 2 cases the PFAA provided the only source of pulmonary blood flow in obstructive right heart lesions. In these 4 cases the PFAA mimicked the arterial duct which was absent in all. The PFAA was sensitive to prostaglandin E1 in one case with pulmonary atresia. The 2 cases with isolated PFAA were treated by surgical ligation avoiding the need for cardiopulmonary bypass and the 2 cases with pulmonary atresia have been successfully repaired. The PFAA in the 5th 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 1st 4 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 E1. The differential diagnosis for the 5th 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.