Abstract no: 944

Outcomes of patients undergoing open heart surgery in Uganda Heart Institute

Twalib Aliku*[,#], Sulaiman Lubega[#], Michael Oketcho[#], Peter Lwabi[#], John Omagino[#] and Tom Mwambu[#]

*Gulu University, Gulu, Uganda
#Uganda Heart Institute, Mulago Hospital, Kampala, Uganda

Background: Heart disease is a disabling condition and needed surgical service is often lacking in many developing countries. A novel approach in which open heart surgeries are conducted locally by visiting teams alongside skills transfer to build local capacity has been adopted at the Uganda Heart Institute (UHI).
Objectives: We evaluated the progress of open heart surgery at the UHI, to describe the post-operative outcomes and challenges faced in conducting these surgeries.
Methods: Medical records of patients undergoing open heart surgery at the UHI from October 2007 - June 2012 were reviewed.
Results: A total of 124 patients underwent open heart surgery during the study period. The commonest conditions were VSDs in 35.5% (44/124), ASDs in 34.7% (43/124) and ToF in 10.5% (13/124). Foreign charities funded 82.3% (102/124) of the operations, and only 4 patients (3.2%) paid for their surgeries. A local NGO funded the rest. There was increasing complexity in cases operated from predominantly ASDs and VSDs at the beginning to more complex cases like ToFs and TAPVR later on. The local team independently operated 19 patients (15.3%). Post-operative morbidity was low with left ventricular dysfunction and reoperations occurring in 6 (4.8%) patients each were the commonest. Sepsis occurred in only 2 cases (1.6%). The 30-day mortality was 3.2%.
Conclusion: Open heart surgery though expensive is feasible in a developing country. Governments and local charities should direct funding to support treatment of more children with heart disease locally as opposed to referral abroad to increase access to the service.

Abstract no: 945

Pulmonary arterial hypertension associated to congenital heart disease in paediatric and adult Spanish population: Data from REHIPED and REHAP

Maria Jesus del Cerro*, J.M. Oliver*, A. Mendoza[#], Laura Dos[†], A. Rodriguez[‡], M. Quero[§], Pilar Escribano[◊] and REHAP/REHIPED investigators

*Hospital La Paz, Madrid, Spain
#Hospital 12 de Octubre, Madrid, Spain
†Congenital Heart Disease Unit for Adolescents and Adults, De la Santa Creu Hospital, Barcelona, Spain
‡Gregorio Marañón University Hospital, Madrid, Spain
§Hospital Ramon y Cajal, Madrid, Spain
◊Hospital 12 de Octubre, Madrid, Spain

Background: Pulmonary arterial hypertension is a serious complication of congenital heart disease (PAH/CHD), but there isn't enough data about clinical profiles and survival of these patients in the adult and paediatric age. Our objective was to analyse clinical and survival data in adult and paediatric Spanish patients with PAH/CHD.
Methods: Voluntary reporting of 338 adults diagnosed of PAH/CHD from January 1998 - April 2012 from the REHAP (Spanish Registry for Pulmonary Arterial Hypertension in Adults) and of 105 children diagnosed from January 2009 - June 2012 from the REHIPED (Spanish Registry for Paediatric Pulmonary Hypertension). Clinical classification for PAH/CHD was used: (1) Eisenmenger; (2) Non-restrictive shunt with high PVR; (3) Small, shunt with high PVR; and (4) Corrected CHD without residual shunt. Kaplan-Meier curves were analysed, censoring patients at death/transplantation.
Results: 443 patients were included. Mean age was 31+17 years in the adult registry and 5.5+5 years in the paediatric one. Paediatric patientss had worse Functional Class FC ($p=0.003$), and more co-morbidities ($p<0.05$), but they showed higher cardiac index ($p=0.015$), and lower mean pulmonary arterial pressure ($p<0.001$). The most frequent forms in adults were Eisenmenger (68%) and operated shunts (20%), while in paediatrics non-restrictive shunts with high PVR (33%) or operated shunt (33%) were the most common forms, ($p<0.001$). Nevertheless, survival was similar in paediatric and adult patients : 91%, 85%, and 80% at 1, 3 and 5 years from diagnosis. For the whole population, we found significant differences in 5-year survival according to FC at diagnosis (90% in FC I/II, 74% in FC III, 50% in FC IV, $p<0.001$), and in Eisenmenger versus operated CHD (94% vs. 72%, respectively, $p=0.038$).
Conclusions: (1) Clinical profiles in CHD-PAH differ significantly between paediatric and adult patients, but survival was similar in both groups; and (2) In the whole population, predictors for worse survival were FC III/IV at diagnosis and operated CHD.

Abstract no: 951

The epidemiological and clinical profile of Kawasaki disease in Western Australia: A 30-year population-based study

Jelena Saundankar*, Deane Yim*, Benedicta Itotoh*, Ruth Payne*, Gayatri Jape*, James Ramsay*, Darshan Kothari*, Allen Cheng[#] and David Burgner[†,‡]

*Children's Cardiac Centre, Princess Margaret Hospital for Children, Perth, Australia
#Monash University, Melbourne, Australia
†Murdoch Childrens Research Institute, The Royal Children's Hospital, Melbourne, Australia
‡School of Paediatrics and Child Health, University of Western Australia, Perth, Australia

Background/hypothesis: The current epidemiology of Kawasaki disease (KD) in Australia is poorly understood. Previous enhanced national surveillance (1993 - 5), gave an estimated incidence of 3.7/100 000 children aged 0 - 4. In non-Asian countries, the incidence varies between 3.6 in Denmark, 8.1 in the UK, 17.1 in the US and 26.2 in Canada, although many data are regional rather than national.
Materials and methods: We identified all patients hospitalised in Western Australia (population ~2.2 million, of whom 3.4% self-identify as indigenous) from 1979 - 2009 with an ICD discharge diagnosis of KD. We retrieved demographic, clinical, laboratory and echocardiographic data from individual patient files. Age-specific population estimates were from national census data. 95% confidence intervals calculated assuming a Poisson distribution.

Results: 353 KD cases were identified. Male to female ratio was 1.7:1 and the median age was 3 years (IQR 12, 60 months).The mean annual incidence increased from 2.82 per 100 000 children aged 0 - 4 years (95% CI: 1.93, 3.99) in 1980 - 1989; to 8.04 (6.55, 9.76) in 1990 - 1999; to 9.66 (8.01, 11.55) in 2000 - 2009. The highest incidence in the 0 - 4 age group was 15.7 in 2005. Incomplete KD was diagnosed in 40/353 (11.4%) patients. Of the 288 children who had complete echocardiographic studies, 42/288 (14.6%) had coronary artery (CA) ectasia/dilatation and 26/288 (9%) had coronary aneurysms. The only significant risk factor for CA involvement was Asian ethnicity. There were no KD cases in Indigenous children (5.8% of WA population).
Conclusions: KD epidemiology in WA mirrors that of other industrialised, predominantly Caucasian populations. The rising incidence reflects both improved ascertainment and a real increase in disease burden. The current Australian incidence of KD is 2 - 3 fold higher than previously reported and comparable to that in the UK and US. The CA outcomes, which include the pre-IVIG era, are comparable to those reported elsewhere.

Abstract no: 958
Mid-term results of mitral valve repair for rheumatic heart disease in children

Sivasubramanian Muthukumar*, Ranjith Karthekeyan#, Ramya Shri†, Charles Jebaraj†, Dheeraj Reddy* and Thangavelu Periyasamy*

*Department of Cardiothoracic Surgery, Sri Ramachandra University, Chennai, India
#Department of Cardiac Anaesthesia, Sri Ramachandra University, Chennai, India
†Department of Cardiology, Sri Ramachandra University, Chennai, India

Objective: To analyse the techniques and mid-term results of mitral valve repair in children with rheumatic mitral valve disease.
Patients and methods: The study population includes 67 patients who underwent mitral valve repair for rheumatic mitral valve disease in the paediatric age group, <18 years in our institution. The group includes 36 female and 31 male patients. The age varied from 6 years - 18 years with a mean age of 14 years. Forty three patients had only mitral regurgitation, 18 patients had mixed lesions, and 5 patients had pure mitral stenosis. 71% of patients were in NYHA Class III or IV.
Results: All patients underwent mitral valve repair under CPB with aortic and bicaval cannulation. Core cooled to 28°C. The approach to mitral valve included both left atrial and superior septal. We performed anuloplasty using rings, teflon strip and auture only in 63 patients. Neo chordae was created in 22 patients using PTFE suture. Chordal transfer was done in 11 patients. Open mitral valvotomy was done in 13 patients. Intra -peratively, valve was checked by saline injection and TEE. Two years follow-up study was done for those patients. Six patients were lost to follow-up. Post-op ECHO revealed severe MR in 3 patients, moderate MR in 9 patients, mild MR in 31 patients and trivial MR in 18 patients. Four patients were reoperated.
Conclusion: Mitral valve repair in rheumatic patients can be performed with acceptable mortality and good mid-term results. We believe valve repair should be preferred over valve replacement for rheumatic patients in the paediatric age group to avoid problems related to anticoagulation and prosthetic valve function.

Abstract no: 975
Long-term survival and Fontan completion rates after surgery for univentricular heart defects

Jan Sunnegårdh, Mats Synnergren and Håkan Berggren

The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Sweden

Background: To analyse long term outcome after surgery for univentricular heart defects.
Material and methods: All 320 patients (133 girls/187 boys) operated due to univentricular heart defects in our institution before <18 years from January 1994 - January 2010 were included. Patient files were studied and cross-checked as of 1 January 2012 against the National Population Registry in Sweden allowing for reliable and complete data on survival. Two patients (0.6%) emigrated and were lost to follow-up 3 and 6 years after Fontan completion.
Results: Median age and weight at 1st surgery was 11 days (0 - 15.1 years) and 3.6kg (1.2 - 30). Median age of survivors at follow-up was 10.8 years (2.1 - 30.7). 87 deaths (27.1%) occurred with a median age at death of 69 days (3 days - 22.7 years). Median survival time in deceased patients was 29 days (0 - 11.3 years) after the last major surgery. In all 276 patients with their 1st surgery in 1994 - 2009 mortality was 53/124 (42.7%) in patients with classic hypoplastic left heart syndrome, unbalanced atrial septal defect or critical aortic stenosis with left ventricular hypoplasia (A), 12/94 (12.7%) in patients with tricuspid atresia, double inlet left ventricle or pulmonary atresia with intact septum (B), ($p<0.001$). Mortality in children with other complex heart defects (C) was 19/68 (27.9%). In these subgroups the number of patients alive with completed Fontan circulation was 59/124 (47.6%) (A), 73/94 (77.7%) (B), 38/68 (58.9%) (C). Nineteen patients had a heart transplantation with 4 late deaths.
Conclusions: Overall survival was 72.9%. Patients with a morphological left ventricle as systemic ventricle had a high survival (87.3%) and Fontan completion rate (77.8%). The corresponding figures were less favourable in patients with a systemic right ventricle (57.3% and 47.6%) and in those with other complex univentricular heart defects (72.1% and 58.9%).

Abstract no: 982
Anatomy of coronary arteries and aortic arch and its relation to truncal valve dysplasia

Gabriel Liguori*, Yen Ho#, Marcelo Jatene* and Vera Aiello*

*Heart Institute (InCor), University of Sao Paulo Medical School, Brazil
#Royal Brompton Hospital, Imperial College London, United Kingdom

Background: Surgical correction of common arterial trunk (CAT) requires knowledge of the particularities of the malformation, such as the anatomy of the truncal valve, coronary arteries and aortic arch. The truncal valve may show dysplastic leaflets. Anomalies of the coronary arteries and of the aortic arch are also described as frequent findings in CAT. In this study, we sought to check for possible associations of the truncal valve dysplasia with the coronary and aortic arch anatomy.
Materials and methods: Forty three heart specimens with CAT were analysed. Coronary orifices were expressed regarding their number, location and shape. Anatomy of the aortic arch was described as normal, interrupted or coarcted, double, right-sided and presenting aberrant subclavian artery. The truncal valve was described regarding the number of leaflets and, according to its thickness and symmetry, classified as mildly dysplastic, moderately dysplastic, severely dysplastic or without dysplasia.

Results: Thirty one (72%) specimens presented at least a mild degree of dysplasia, 3 (6%) presented abnormal number of coronary orifices (2 single and 1 triple ostium) and 17 (40%) presented at least one type of aortic arch anomaly. Truncal valve dysplasia was associated with a number of leaflets different than 3 (p=0.022). Anomalous shape of the coronary orifices was predominantly described as slit-like for the right coronary orifices and as funnel for left coronary orifices. Anomalous shape of the right coronary orifice was associated with both dysplasia and abnormal number of leaflets (p=0.038 and p=0.025, respectively). Specimens with abnormal anatomy of the aortic arch also showed a trend to present abnormal number of leaflets (p=0.072).
Conclusion: Anomalies of the coronary ostia and aortic arch seems to be related to the dysplasia of the truncal valve. This correspondence should alert both the echocardiographist and the cardiac surgeon to the diagnostic and management of associated lesions.

Abstract no: 988

Infant with mitral valve endocarditis caused by Saccharomyces sp.: Unusual case and successful medical treatment

Adriana Castillo#, Ana Erazo#, Martha Alvarez-Olmos*, Claudia Stapper*, Nestor Sandoval*, Alberto Guerra* and Karina Tovar*

*Fundacion Cardioinfantil, Bogota, Columbia
#Our Lady of the Rosary University, Bogota, Colombia

Background: Invasive fungal infection (IFI) is an emerging complication in paediatric population. Its incidence has recently increased especially in patients admitted to neonatal and paediatric Intensive Care Units (NICU/ PICU). Fungal endocarditis is an IFI with high risk of complications and death. Several risk factors such as prolonged hospital stay, use of central venous catheters and broad-spectrum antibiotics, and the particular vulnerability of the immunological immaturity of the infants, especially preterm newborns, have been identified. In these cases, even non-pathogenic strains might be identified with related risk factors.
Materials and methods: Paediatric case report with unusual fungal mitral valve endocarditis treated medically in Colombia.
Results: We present a 2-month-old male infant with history of complications associated to 28-weeks of prematurity, prolonged hospitalisation in the NICU/ PICU, episodes early and late neonatal sepsis, necrotising enterocolitis complicated with short bowel syndrome transiently treated with probiotics, managed with broad spectrum antibiotics, and carrying a central venous catheter who developed a mitral valve endocarditis caused by Saccharomyces sp., which was isolated from several blood cultures. He was treated initially with fluconazole and changed to deoxicolato of amphotericin B based on laboratory susceptibility, with complete clinical and microbiological recovery. Surgical management was not performed and differed due to the basal patient's condition.
Conclusions: Isolation of this non-pathogenic yeast widely used as probiotic as a cause of endocarditis in a patient with multiple risk factors like our patient's case has been infrequently reported. Nevertheless, it suggests us that there are certain circumstances in which these probiotics should be used with caution.

Abstract no: 989

Long term survival after surgery in patients with congenital heart defects and Down syndrome

Eva Stramvall Larsson, Haykan Berggren and Jan Sunnegayrdh

The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Sweden

Background: To collect reliable and complete data for evaluation of long term survival after paediatric cardiac surgery in patients with Down syndrome.
Materials and methods: All 288 patients (131 girls/157 boys), operated due to congenital heart defects in our institution <18 years from 1994 - 2009 were included. During the study period nearly 50% of all paediatric cardiac surgery in Sweden was performed at our institution. Patient files were cross-checked as of 1 January 2012, against the National Population Registry in Sweden, allowing for reliable and complete data on long-term survival. Two patients emigrated and were lost to follow-up.
Results: Median age at 1st surgery was 0.44 years (0.01 - 15.69 years). 142 (49%) had surgery for atrioventricular septal defects (AVSD), 72 (25%) for ventricular septal defects (VSD), 13 (5%) for isolated Tetralogy of Fallot and 7 for AVSD and Tetralogy. Six patients had univentricular heart palliations. Eleven of the patients with AVSD/VSD had a pulmonary artery banding as the 1st operation. Median age of survivors at follow-up was 10.6 years (3.3 - 32.6 years). Eighteen deaths (6.3%) occurred with a median age at death of 1.6 years (0.06 - 21.36 years). The cause of death was pulmonary hypertension in 4, septicaemias in 5. One patient operated for a VSD at the age of 4 months also had biliary atresia, and was liver transplanted. He died at the age of 3 years. One patient died in an accident. There were 7 cardiac deaths. Fifteen patients developed complete AV-block post-operative, needing a pacemaker. Eleven patients developed significant mitral valve regurgitation and had a mechanical valve prosthesis inserted.
Conclusions: With 93.7% of the patients alive at follow-up long term survival was favourable. The majority of deaths (78%) occurred >30 days post-operatively, reflecting the need for long term follow-up.

Abstract no: 997

Heterotaxy syndrome: Is a prophylactic Ladd procedure necessary in asymptomatic patients?

Charissa R Pockett*, Bryan Dicken#, Jennifer Rutledge*, Ivan M Rebeyka#, David B Ross#, Andrew S. Mackie* and Lindsay M Ryerson*

*Department of Paediatrics, University of Alberta, Edmonton, Alberta, Canada
#Department of Surgery, University of Alberta, Edmonton, Alberta, Canada

Background/hypothesis: Complex congenital heart disease and abnormalities of intestinal rotation are commonly associated with heterotaxy syndrome (HS). Malrotation is the most worrisome intestinal rotation abnormality (IRA) due to the risk of bowel ischaemia and infarction. There is controversy whether asymptomatic infants with HS require screening for IRA and if present, whether a prophylactic Ladd procedure is indicated. As this population grows due to continuing advances in cardiovascular care, it is vital to better understand the natural history of IRA and devise an evidence based treatment model.
The first objective of this study is to determine institutional practice in the management of asymptomatic infants with HS and IRA. The second objective is to prospectively observe a cohort with HS and evaluate their long term outcomes.

Materials and methods: We have begun a prospective, multi-center, observational study using a web-based database to follow infants with HS to 5 years of age. Data collection includes screening methods used for the detection of IRA, management of IRA, cardiac diagnosis, cardiac interventions, long term complications and outcomes. Patient management will not be dictated by the study protocol, given the observational design.
Results: Ethics and scientific approval has been obtained at 2 centers in Canada and 6 patients have been enrolled to date. Fourteen other centers across North America and the United Kingdom are currently obtaining ethics approval. Additional centers are being actively recruited.
Conclusions: Results from this study may change general surgical practice in the management of this complex group of patients. Establishment of this cohort will facilitate future studies of the HS population with respect to gastrointestinal and cardiovascular outcomes.

Abstract no: 1000

Mid-term functional outcome after repair of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)

Lennart Duebener, Claudia Arenz, Peter Murin, Ehrenfried Schindler, Martin Schneider, Peter Zartner, Nicole Toussaint- Gâtz, Nicodeme Sinzobahamvya, Boulos Asfour and Victor Hraska

German Paediatric Heart Centre, Sankt Augustin, Germany

Background: Treatment options for ALCAPA repair include direct implantation of the origin of the left coronary artery into the aorta and the Takeuchi procedure, which involves creation of an aortopulmonary window and a tunnel that directs blood from the aorta to the anomalous left coronary artery. The patient outcome and the incidence of residual and new lesions after the 2 types of ALCAPA repair were analysed.
Methods and patients: From 1995 - 2012 a total of 21 patients (76.2% female) underwent ALCAPA repair at our institution. Four (19 %) had a Takeuchi procedure and 17 (81%) direct implantation of the LCA. Concomitant procedures included mitral valve repair (n=2) and ASD closure (n=1). The mean patient age at time of surgery was 8.3 months (range: 1st day of life - 41.4 months).
Results: Surgical and late mortality was zero. The mean length of follow-up after surgery was 6.2 years (range: 0.4 - 10.9 years). Mean post-operative LV fractional shortening at last follow-up was 38.3±5.6% and mean LVEDD was within normal limits. Reasons for reoperations included residual mitral regurgitation (MR) (n=1) and baffle leaks (n=2). The freedom from reoperation was 81% at 5 years. Mild main pulmonary artery stenosis was documented in 3 patients after Takeuchi repair. Three patients had a moderate degree of residual mitral regurgitation, the remaining patients were free of MR or only had a mild degree.
Conclusions: ALCAPA repair can be performed with low surgical mortality. In the majority of patients after ALCAPA correction, both ventricular function and mitral valve regurgitation normalise over time. Therefore concomitant mitral reconstruction may not be required upon initial repair. Coronary transfer is our preferred technique today because the Takeuchi procedure is associated with a higher rate of reoperations and residual lesions.

Abstract no: 1006

Anomalous origin of coronary arteries from right pulmonary artery

Nawal AlAbdulkarim, Tarek Momenah and Saleh AlGhamdi

Prince Sultan Cardiac Centre, Riyadh, Saudi Arabia

Background: Anomalous origin of coronary arteries from right pulmonary artery AOCARPA is a rare variant of anomalous left coronary artery from the pulmonary artery AlCAPA. It is essential to recognise this anomaly since it causes higher morbidity/mortality of the associated congenital cardiac lesions; also correction of the associated congenital cardiac defect could reduce left coronary artery flow.
Methods: We describe the cases of AOCARPA that were diagnosed at a tertiary care referral cardiac centre during 1990 - 2012.
Results: Forty five patients had ALCAPA, 3 patients (7%) were diagnosed with AOCARPA as described in the Table. All underwent surgical reimplantation of AOCARPA; patient number 2 had uneventful convalescence in spite of being diagnosed late post coarctation repair, patient number 3 died due to sepsis post-operatively, and patient number 1 with single coronary from RPA died during surgery.

Patients	Age month	Sex	Associated Cardiac Defects	Diagnosis delay	Echo	Angios	Anomalous coronary
1	2	Male	ASD	-	DCM/ALCAPA	Yes	RCA/LCA (single)
2	27	Male	Coarctation VSD	Yes	DCM/ALCAPA	Yes	LCA
3	7	Male	VSD	-	DCM/ALCAPA	Yes	LCA

Conclusions: AOCARPA is a rare subset of ALCAPA which diagnosis is suspected by clinical/echocardiographic findings and confirmation requires cardiac angiography, it is usually associated with other congenital heart disease and carries higher mortality.

Abstract no: 1016

Exercise training in children and young adults with corrected Tetralogy of Fallot: Preliminary results of the TOFFIT study

Nienke Duppen*, Michiel Schokking#, Irene Kuipers†, Christiaan Blank‡, Nico Blom§, Arie van Dijk#, Jolien Roos-hesselink*, Tim Takken◊, Maria Hopman# and Wim Helbing*

*Erasmus Medical Centre, Sophia, Rotterdam, The Netherlands
#Radboud University, Nijmegen Medical Centre, Nijmegen, The Netherlands
†Academic Medical Centre, Amsterdam, The Netherlands
‡Wilhelmina Children's Hospital, Utrecht, The Netherlands
§Leiden University Medical Centre, Leiden, The Netherlands
◊Child Development & Exercise Centre, Wilhelmina Children's Hospital, Utrecht, The Netherlands

Objective: To study whether exercise training in children and young adults with corrected Tetralogy of Fallot can improve exercise capacity, physical activity and ECG markers.
Methods and results: The study is a multicentre randomised controlled trial. After randomisation the participants were assigned to an interventional or control group. In total, 92 participants were recruited from 5 participating university medical centres. All participants underwent a cardiopulmonary exercise test, an electrocardiogram and wore a 5-day activity monitor device. The intervention group followed 12 consecutive weeks of supervised aerobic exercise training 3 times a week for an hour at the level of 60 - 70% of heart rate reserve. The results of the 1st 26 participants were included in an interim analysis. Baseline characteristics of the intervention group consisting of 14 participants: age 16.1±2.4 years; BMI 21.3±3; 10 males; baseline characteristics of the control group consisting of 12 participants: age 17.6±3.0; BMI 20.6±3.2; 9 males. VO2peak/kg of the intervention group improved significantly (34±6ml/kg/min vs. 38±8ml/kg/min, p<0.03) whereas the control group did not show any change (35±7ml/kg/min vs. 34±5ml/kg/min). Activity levels did not change in either group. No significant ECG changes were seen (QRS duration intervention group 128±27ms vs. 127±26; control group 128±24ms vs. 125±27ms, QTc duration intervention group 432 ±29ms vs. 427±27, control group 431±19 vs. 425±22ms). Moderate to vigorous activity levels in percentage of total recorded time did not change in either groups (intervention group: 15±6 vs.15±5; control group 14±6 vs. 14±6).
Conclusion: Exercise training did improve VO2peak/kg of children and young adults with a corrected Tetralogy of Fallot without changing ECG markers and physical activity.

Abstract no: 1021

Normal values of arterial valve diameters in neonates

Krzysztof Michalak, Jolanta Binikowska, Katarzyna Ostrowska, Barbara Stanek and Jadwiga Moll
Department of Cardioogy, Polish Mother's Memorial Hospital, Research Institute, Poland

Background: Normal values of various cardiovascular diameters are needed to facilitate proper diagnosis and decision about treatment of children with congenital heart defects. The larger the analysed group, the more accurate and helpful the conclusions and received norms are.
Aim: The aim of this study was to assess the normal values of aortic and pulmonary valves in healthy, term neonates and to correlate this with age (1 - 30 days), weight and body surface area.
Material and methods: 771 transthoracic echocardiographic examinations performed between years 2002 - 2008 were reviewed and data were collected retrospectively. All examinations were performed in healthy, term neonates without any structural heart defect. Measurements were taken from short (pulmonary valve) and long (aortic valve) parasternal axis in diastole. Statistical analysis was performed using Statistica 10 software.
Results: The average age at examination was 10.6 days (SD-7.8), weight 2.6kg (SD-0.7). Aortic valve diameter – average 7.68mm (SD-0.95) – was statistically significantly correlated with age (p<0.001,R=0.29); BSA (p<0.001,R=0.47) and weight (p<0.001,R=0.54). In the case of the pulmonary valve the average diameter was 9.05mm (SD-1.24). It was also correlated significantly with age (p<0.001,R-0.4), BSA (p<0.001,R-0.58) and weight (p<0.001,R-0.62). In both of the valve diameters the strongest correlation and clinical significance was observed with weight. Because all observed correlations was medium or weak we calculated the normal values of aortic (6.3 - 9.7mm) and pulmonary (7 - 11.4mm) valves for whole study group, containing values between 5 and 95 percentile.
Conclusion: The diameters of aortic and pulmonary valves in the neonatal period does not have a strong correlation with age, weight or BSA, despite the fact, that there is statistically significant correlation in all of the cases. It allows use of the common normal values range for the neonatal period. In case of borderline values, the weight should be taken under consideration, because it has the strongest correlation with arterial valve diameters.

Abstract no: 1028

Pulmonary haemosiderosis secondary to severe mitral stenosis in patients undergoing valvuloplasty at Tygerberg Hospital

Jacques Doubell*, Philip Herbst*, Alfonso Pecoraro*, Coenraad Koeglenberg# and Anton Doubell*
*Division of Cardiology, Department of Medicine, Stellenbosch University and Tygerberg Academic Hospital, Bellville, South Africa
#Division of Pulmonology, Department of Medicine, Stellenbosch University and Tygerberg Academic Hospital, Bellville, South Africa

Background: Pulmonary haemosiderosis (PH) secondary to mitral stenosis (MS) is considered rare. This study aimed to determine if PH is more common than currently thought as well as determining factors contributing to its development, impact on lung function and reversibility.
Methods: Chest X-rays (CXRs) of patients who underwent mitral valvuloplasty (17 January 1997 - 10 February 2012) were reviewed for PH. Data collected included date of birth, date of valvuloplasty and pre-procedural echocardiography reports. Patients with PH on CXR were invited to participate in a prospective trial. Patients without PH were selected as controls. In the prospective trial a clinical evaluation, ECG, echocardiography, CXR and lung functions were performed.
Results: Sixty (24.8%) of 242 patients who underwent valvuloplasty at Tygerberg Hospital had CXRs available. The incidence of PH in this cohort of patients with severe MS was 19%. Patients with PH had higher pulmonary pressures (71.1mmHg vs. 55.7mmHg; p=0.04) and had significantly smaller mitral valve areas

(0.85cm^2 vs. 0.99cm2; p=0.09). Patients with PH also had higher peak transmitral pressure gradients (30.1mmHg vs. 24.37mmHg; p=0.10). Although the mean transmitral pressure gradients were higher in the patients with PH this difference was not significant (17.43mmHg vs. 15.02mmHg; p=0.23). Patients with PH were younger at the time of valvuloplasty (29 vs. 38; p=0.02). No difference was seen when comparing the lung functions in terms of ventilatory and diffusion impairment. The data regarding radiological reversibility was inconclusive.
Conclusion: PH occurs more commonly in severe MS than currently thought. Compared to patients without PH, they have more severe MS and underwent valvuloplasty at a younger age. The data suggests that PH is an irreversible process with little impact on lung function but the numbers studied prospectively were too small to provide conclusive evidence regarding the impact on lung function and reversibility.

Abstract no: 1030

Score for quantification of the morphological and functional echocardiographic features of mitral valve in patients with rheumatic heart disease

Cleonice Mota, Carolina Capuruão, Alan Conde, Fãtima Rocha and Zilda Meira
Division of Paediatric Cardiology, Federal University of Minas Gerais, Brazil

Background: Rheumatic heart disease (RHD) still challenges all those involved in its diagnosis and in this context, a more accurate assessment by echocardiography has added valuable information about the functional and anatomic subtract of valvar and subvalvar mitral apparatus (MV).
Objective: To present an echocardiographic score to support the evaluation of MV lesions.
Methods: Out of 298 patients seen during the period of 2009 - 2010, 93 patients (duration of the disease: = 8.0±5.8 years; age at first episode: = mean: 9.2±2.9 years) with definitive diagnosis of RHD, followed since the 1st episode and without surgical intervention were selected and underwent ECHO screening. The ECHO tapes were reviewed by 2 other echocardiographists and the Kappa statistic was used for the assessment of inter-observer variability [Kappa: 0.875 (0.775 - 0.974; CI 95%)].
Results: The patients' data was scored on a rating scale (1 - 15), to quantify the severity of the mitral valve involvement (Table 1). The quantification included 5 groups of morphological and functional variables and 3 degrees of severity. In the comparative analysis, the degrees established for the classification of the scores of morphological/functional features were associated with the correspondent degrees of severity of the haemodynamic findings for mitral regurgitation (p:0.00) and stenosis (p:0.02).

TABLE 1: Quantification of the morphological/functional features of MV according to the degree of commitment

ECHO findings/ score	1	2	3
AMVL thickening	3 - 5mm (mild)	6 - 7mm (moderate)	>8mm (important)
Restrictive or excessive motion	Mild	Moderate	Important
Cordal/commissural abnormalities	Cordal thickening	Commissural and/or cordal fusion	Rupture/flail
Deformity of leaflets	Focal thickening	Dog leg orbprolapse	Coaptation defect
Functional abnormalities (↑chambers - PH)	Mild	Moderate-severe	Pulmonary hypertension

Score:1 - 5→mild, 6 - 10 →moderate,10 - 15 →severe.

Conclusion: Considering the characteristic aspects of the morphological features of the mitral valve in patients with RHD, the stratification by degree of commitment could represent additional support for the echocardiographic evaluation.

Abstract no: 1035

Improving the diagnostic yield of the ECG in tachyarrhythmias via routine Lewis lead recordings

Everhardus Smith, Hellmuth Weich Anton Doubell
Division of Cardiology, Department of Medicine, Stellenbosch University and Tygerberg Academic Hospital, Bellville, South Africa

Background: Determining the origin of tachyarrhythmias is a skill required by all clinicians. Identifying the p-wave is integral to making a diagnosis in tachyarrhythmias. A diagnostic tool that is proposed as an aid in this regard is doing Lewis leads, but there is little evidence for this in the literature. As with any diagnostic tool one would expect that its value is influenced by the users' knowledge and experience.
Methodology: We prospectively studied patients presenting with a tachycardia to the division of cardiology of Tygerberg Hospital over a 6-month period. Both standard ECGs and Lewis leads were performed in all cases. These ECGs were then analysed by 3 sets of participants namely cardiologists, registrars and students. An initial diagnosis was made on the standard ECG only. A revised diagnosis was then given based on the added information provided by the Lewis lead in addition to the standard ECG. These answers were compared to the definitive diagnoses and classified as correct, incorrect and unsure (where a differential diagnosis was given).
Results: More correct diagnoses were obtained in all groups with the addition of Lewis leads. Cardiologists displayed a trend towards improvement with Lewis leads (87% - 96%; p=0.07). The registrar group had the most marked improvement (64% - 82%; p=0.0026). Students did not improve significantly (52% - 54%; p=0.8). Lewis leads was of assistance to students in patients with regular rhythms but not in irregular rhythms.
Conclusion: Lewis leads is a valuable diagnostic tool to enhance ECG interpretation. It aids clinicians in making accurate diagnoses in tachyarrhythmias. The amount of benefit is dependent on the user's knowledge and expertise. Cardiologists and registrars may benefit from routinely registering Lewis leads in tachyarrhythmias. Inexperienced individuals should be wary of using Lewis leads in irregular tachyarrhythmias. Students would benefit from specific training in assessing Lewis leads.

Abstract no: 1040

Impact of age and gender on cardiac pathology in children and adolescents with Marfan syndrome

Goetz Mueller, Jeske Wild, Veronika Stark, Kristoffer Steiner, Florian Arndt, Jochen Weil and Thomas Mir

Clinic for Paediatric Cardiology, University Heart Centre, Hamburg, Germany

Background: Cardiac pathologies are the major aspect in the treatment-strategies of Marfan syndrome (MFS). In this progressive disease less is known about manifestations and progression of cardiovascular symptoms in children. To define a certain decision regarding therapeutic options, knowledge about onset of cardiovascular findings in children is essential.
Materials and methods: From 1998 - 2011 patients with clinical features of MFS were subject to a standardised diagnostic programme. Cardiovascular findings were analysed concerning age at 1st clinical manifestation, prevalence and sex differences, morbidity, mortality and treatment during follow-up.
Results: MFS was diagnosed in 82 patients (46 male, mean age at diagnosis 9.0±5.7 years). At 1st presentation aortic root dilation and mitral valve prolapse was found in 56.1 and 31.7% whereas pulmonary artery dilation and tricuspid valve prolapse were detected in 22.0 and 17.1%. Aortic valve (2.4%) and mitral valve regurgitation (22.0%) are significantly correlated with aortic root dilation ($p<0.01$) and mitral valve prolapse ($p<0.05$) but without relevant progression during childhood and adolescents. Because of progressive aortic root dilation medication was initiated in 41.5% of patients (mean age 8.0±4.5 years). Aortic dissection did not appear. Aortic root surgery in case of severe progression of aortic root dilation was needed in 3.7% of patients (mean age 9.7±2.3 years). Gender specific differences concerning cardiovascular findings, progression of disease or treatment did not appear.
Conclusion: Comparable to adults aortic root dilation is the most frequent cardiovascular finding and associated with relevant morbidity and aim of early prophylaxis. While aortic and mitral valve regurgitation is of minor clinical relevance. Manifestation at early age and slow progression of cardiovascular findings in childhood underline the necessity of repeated echocardiographic examinations in case of suspected MFS for early diagnosis and of course an early start of prophylactic treatment.

Abstract no: 1041

Cardiomyopathy in patients with the Amish & Mennonite variant of propionic acidemia

Devyani Chowdhury[*], Ron Jacob[*], Alison Greidinger[#], Kevin Strauss[#,†] and D. Holmes Morton[#,†]

[*]Lancaster General Hospital, Lancaster, Pennsylvania, United States of America
[#]Clinic for Special Children, Strasburg, Pennsylvania, United States of America
[†]Franklin & Marshall College, Lancaster, Pennsylvania, United States of America

Background: Propionyl-CoA carboxylase deficiency (propionic acidemia, PA) caused by a PCCB c.1606A>G variant is prevalent among Amish and Mennonite (Plain) people. Systemic metabolic crises are infrequent, but they remain at risk for life-threatening cardiomyopathy, which may result from loss of anaplerotic propionyl-CoA flux into the myocardial tricarboxylic acid (TCA) pool.
Methods: Thirty three patients (10.2±5.6 years) homozygous for PCCB c.1606A>G were followed longitudinally at a single centre for 250 aggregate patient-years. We studied cardiac morphology, function, and outcome using ultrasound and cardiac magnetic resonance (CMR) equipped with quantitative T1 mapping software (Siemens Healthcare). Two brothers with symptomatic cardiomyopathy were treated with a dietary anaplerotic mixture designed to supply 2-, 4-, and 6-carbon substrates to the TCA cycle.
Results: Cardiomyopathy (Left ventricular ejection fraction, LVEF<3SD below age-matched controls) developed in 12 (36%) patients. (EF 58% to 7%), and was lethal in 3 children. In PA patients, LV EF (64±11%) was lower than controls (73±5%; $p<0.0001$). 2 brothers with symptomatic cardiomyopathy when treated with the TCA anaplerotic mixture had resolution of symptoms over 4-6 months. LV EF increased from 18% - 56% and 44% - 63%. CMR during the acute phase showed biventricular dysfunction and increased end diastolic volumes but no evidence of tissue fibrosis or oedema (myocardial interstitial volume 26%). After anaplerotic treatment the biventricular function and cardiac volumes normalised and there was no tissue scarring (myocardial interstitial volume 29%).
Conclusion: Cardiomyopathy is common in patients with PA who are otherwise metabolically stable and is the major cause of untimely death resulting from the PCCB c.1606A>G variant. PA should be considered in patients of any age who present with "idiopathic" cardiomyopathy, even when standard metabolic screening tests are normal. The cardiomyopathy of PA may be fully reversible by supporting the TCA anaplerotic function normally served by propionyl-CoA.

Abstract no: 1042

Changes in clinical signs of heart failure after carvedilol therapy in children with left-to-right shunt congenital heart disease

Mahrus Rahman, Teddy Ontoseno, Alit Utamayasa, Hendarti Eddy and Budiyono Budiyono

Department of Child Health, Medical School, Airlangga University, Soetomo Hospital, Surabaya, Indonesia

Background: Heart failure (HF) in children with left-to-right shunt congenital heart disease (L-R shunt CHD) caused heart remodelling, make the symptoms worsen. Tachycardia, tachypnea and hepatomegaly are the cardinal sign of HF. Carvedilol, non-selective beta blocker has good results for HF treatment in adult with tolerable side effects, but there is scarce evidence in children. The objective of this study is to determine the changes in heart rate, respiratory rate, and hepatomegaly after 3 months carvedilol additional therapy on HF standard therapy in children with L-R shunt CHD.
Materials and methods: A randomised controlled trial designed for 30 subjects with Paediatric Heart Failure Score (PHFS)> due to L-R shunts CHD. Subjects divided into carvedilol group (n=15) and placebo group (n=15). 2nd group received standard therapy for HF ACE-I and diuretic. The study last for 3 months. The changes in heart rate, respiratory rate, and hepatomegaly were observed. Statistical analysis using paired t-test and independent sample t-test with confidence interval of 95%. McNemar test and Chi square test, CI 95%.
Results: Nineteen (63.3%) were boys 11 (36.7%) were girls. Mean age 57.6 (SD 43.57) months: 21 VSD (70%), 9 PDA (30%). Heart rate and respiratory rate were significantly decreased in carvedilol group compared with placebo group ($p <0.0001$). No hepatomegaly found. No adverse effect during the study.
Conclusion: Additional therapy with carvedilol on standard therapy HF due to L-R shunt CHD significantly improved clinical signs of HF. Carvedilol is effective and safe as additional therapy on HF standard therapy.

Abstract no: 1047

Erythrocyte deformability mechanism in Tetralogy of Fallot patients with iron deficiency

Teddy Ontoseno

Cardiology Division, Department of Child Health Medical Faculty, Airlangga University, Surabaya, Indonesia

Background: Iron deficiency in patients with Tetralogy of Fallot (TF) is a highly adverse condition. Iron deficiency in TF patients is related to erythrocyte deformability and the incidence of fatal complications of the disease. However, the mechanism of erythrocyte deformability in TF patients with iron deficiency so far remains unexplainable.
Objective: To disclose the mechanism of erythrocyte deformability in TF patients with iron deficiency. This is an observational study on human beings. This study involved 4 groups: TF patients with iron deficiency (Group I), TF patients without iron deficiency (Group II), non-TF patients with iron deficiency (Group III), and non-TF patients without iron deficiency (Group IV).The variables of erythrocyte deformability used in this study were SaO_2, transferrin saturation, H_2O_2 molecule level in erythrocyte, and spectrin-denaturated as well as the number of erythrocyte passing through a device membrane.
Result and conclusion: The reduction of SaO_2 resulted in the decrease of transferrin saturation followed by the increase of H_2O_2 molecule level, leading to the rise of the number of spectrin-denaturated erythrocytes and to reduced erythrocyte deformability and the reduction of the number of erythrocyte passing through a device membrane. Iron therapy may increase transferrin saturation, reducing thereby the erythrocyte H_2O_2 molecular level, and lead further to the reduction of the number of spectrin-denaturated erythrocytes count and improved deformability of erythrocyte and increase the number of erythrocyte passing through a device membrane. This study also produced a model describing the contribution of each discriminator. The discriminator model of the occurrence of erythrocyte deformability in TF patients with and without iron deficiency can be applied as a novel basic concept to underscore and optimise the management of TF patients who still have no opportunity to have cardiac corrective surgery.

Abstract no: 1050

Usefulness of N-Terminal pro-B-type natriuretic peptide as biomarkers for congestive heart failure treatment of congenital heart diseases

Sevcan Erdem, Pånär Kocak, Alev Kiziltas and Nazan Ozbarlas

Department of Paediatric Cardiology, Faculty of Medicine, Cukurova University, Adana, Turkey

Background: The aim of this study is to determine plasma N-Terminal pro-B-type natriuretic peptide (NT pro-BNP) levels in infants with congenital heart disease (CHD) and congestive heart failure (CHF) before and after decongestive treatment, and correlate these with the heart failure score.
Methods and results: The study comprised 46 infants with CHD aged from 21 days - 26 months who were categorised into CHF mild, moderate and severe according to the modified Ross scoring system. The patients were evaluated before and 10 and 30 days after decongestive treatment. Before treatment; 15 patients had mild CHF, 27 moderate CHF and 4 severe CHF. Mean NT-proBNP level was 4983.4+6326pg/ml. 10th day after the treatment; 27 of them (59%) mild, 18 (39%) moderate, and 1 (2%) had severe CHF. Mean NT-proBNP level was 2177.1+2629.8pg/ml. 30th day after the treatment; 41 of them (89%) had mild CHF, 5 11%) had moderate CHF. None of them had severe CHF. Mean NT-proBNP level was 1701.8+2126.4pg/ml. NT-proBNP levels were decreased with decongestive therapy ($p<0.05$). NT-proBNP level was lower on the 10th day of therapy than before therapy and NT-proBNP level was lower on the 30th day of therapy than the 10th day of therapy. There was no significant correlation between NT-proBNP and Ross scoring at 10th day of the therapy. There was a significant correlation between NT-proBNP and Ross scoring at 30th day of therapy.
Conclusion: Plasma NT-proBNP levels are elevated in infants with CHD with left-to-right shunt before treatment, and decreased with decongestive therapy, nevertheless clinical evaluation is more important to determine the severity of CHF.

Abstract no: 1051

Profile of the presentation and evolution of rheumatic fever in children and adolescents

Cleonice Mota, Rosangela Graciano, Fatima Rocha and Zilda Meira

Division of Paediatric Cardiology, Federal University of Minas Gerais, Brazil

Background: Rheumatic fever (RF) and rheumatic heart disease (RHD) represent a health burden worldwide. As a condition carried throughout life, it has repercussions at all ages and accounts for an important number of repeated hospitalisations and deaths.
Objectives: To analyse the profile of the clinical and epidemiological presentation and its relation with the severity and evolution of cardiac involvement.
Methods: This cohort study was carried out on 823 consecutive patients aged from 2.7 - 18.9 years and with a mean follow-up of 7.6±2.8 years (from 1984 - 2004). The following variables were analysed: age at first attack, gender, clinical manifestations and recurrences; family antecedents, previous pharyngo tonsillitis and pattern of severity.
Results: The 1st episode was most frequent in the age group of 6 - 15 years (:9.2±3.1 year), without gender predisposition, except for chorea (F/M:1.7/1.0; p:0.0013). Previous pharyngo tonsillitis was reported by 54.9%. The prevalence of RF in patients' families (14.2%) was higher than among relatives of children without the disease (p:0.0000). At the first attack, 96.4% patients presented MR, isolated (44.2%) or associated to AR. Out of those with RHD, 97.6% showed mitral and/or aortic involvement (isolated AR:2.4%) with regurgitation in 78.8% and mixed lesions in 21.2%, without patients with AS. Severe carditis was more prevalent in children with 2 or more recurrences, and valvar sequels more significant in those with severe carditis (p:0.0001); 34.8% of patients showed complete resolution of cardiac findings, mostly without recurrences, and presented mild regurgitant lesions but none with severe valvar involvement. Significant decrease was seen in the occurrence of severe carditis, surgery and death after the control of recurrences (p:0.0000).
Conclusions: The valvar sequels were influenced by the severity of the carditis and by the number of recurrences. Considering the difficulties in the approach of primary prevention, the authors reinforce the need for effective strategies of secondary prophylaxis to reduce morbidity and mortality.

Abstract no: 1055

Innsbruck experience with Kawasaki disease

Martin Schwienbacher and Ulrich Schweigmann

Department of Paediatrics, Division of Cardiology, Pulmonology, Innsbruck Medical University, Austria

Background: We describe long term follow-up of 3/89 patients with Kawasaki disease seen in our institution over a 33-year period, with different clinical course of cardiac or coronary artery involvement.
Materials and methods: A retrospective description including clinical investigation, echocardiography and angiography was performed.
Results: Patient 1 had a myocardial infarction in the acute phase of Kawasaki disease 33 years ago. At that time aneurysms in both carotids, subclavian, renal and mesenterial arteries were detected by sonography and angiography. Last heart catheterisation 6 years ago revealed 2 aneurysms of the left ventricle (one at the apex and one at the basis, 3cm diameter each one) and a hypokinetic left ventricle. He is in stable clinical condition and denied any therapy. **Patient 2** developed a 17 x 11mm aneurysm of the left coronary artery. Twelve years later, a 90% stenosis of the left coronary artery and a 70% stenosis of the left anterior descending artery, as well as the ramus circumflex, were detected by angiography. He underwent aortocoronary bypass with a bilateral arteria thoracica interna bypass. He is in excellent clinical condition under treatment with a platelet aggregation inhibitor. **Patient 3** was diagnosed with a 9 x 6mm aneurysm of the right coronary artery during the acute phase of Kawasaki disease. Spontaneous regression was observed within 12 years. The right coronary artery shows no signs of stenosis or thrombosis in angiography, ventricle function is normal and the patient is in excellent clinical condition, receiving clopidogrel.
Conclusions: In a 33-year period 2/89 patients evolved giant coronary artery aneurysms, 1 patient therefore underwent aortocoronary bypass surgery. One patient developed 2 aneurysms of the left ventricle and multiple aneurysms in the great arteries affecting multiple central arteries.

Abstract no: 1056

Myocardial response to exercise after paediatric heart transplant: A bicycle exercise study

Barbara Cifra, Henrik Brun, Cedric Manlhiot, Brian McCrindle, Anne Dipchand and Luc Mertens

Division of Cardiology, Department of Pediatrics, The Hospital for Sick Children, University of Toronto, Canada

Background: Data on myocardial systolic and diastolic response to exercise of the transplant heart is limited. We used semi-supine cycle ergometry (SSCE) stress echocardiography to evaluate left ventricular (LV) systolic and diastolic reserve in pediatric heart transplant (P-HTx) recipients and we compared the exercise response to healthy controls.
Materials and methods: 43 P-HTx and 24 age and gender matched controls were included. A stepwise SSCE protocol was used. Peak systolic and early diastolic tissue Doppler velocities were measured in the lateral and septal basal segments and values were expressed versus heart rate. LV myocardial acceleration during isovolumic contraction (IVA) was measured in all the subjects at incremental heart rates to evaluate the force-frequency relationship (FFR). LV longitudinal strain was also quantified at rest and during exercise.
Results: At rest early diastolic tissue Doppler velocities (E') were reduced in P-HTx group in the lateral LV wall (11.1vs. 13.7cm/s p=0.001) and the basal septum (8.1 vs. 11.1cm/s p<0.001). Lateral and septal S' values did not differ significantly between the groups. At peak, all S' (8.1 vs. 11.1cm/s p<0.001) and E' (8.1 vs. 11.1cm/s p<0.001) velocities were lower in P-HTx. The change in E' and S' values from baseline to peak was lower P-THx compared to controls. Also the E/E' ratio was higher in P-HTx in the lateral wall and in the septum. The contractile response as studied by the FFR, is blunted in P-HTx. LV longitudinal peak systolic strain values increase during exercise in both groups, but the P-HTx have lower strain value than controls.
Conclusions: P-HTx showed a reduced systolic contractile response as well as a reduced diastolic response to exercise compared to controls. This is not related to the heart rate response. The clinical and prognostic implications of these findings require further study.

Abstract no: 1058

Assessment of pulmonary vascular volume and lung perfusion in patients with hypoplastic left heart syndrome in Fontan circulation using CMR imaging

Eileen Pardun[*], Michael Jerosch-Herold[#], Christopher Hart[*], Inga Voges[*], Dominik Gabbert[*], Hans-Heiner Kramer[*] and Carsten Rickers[*]

[*]Department of Congential Heart Disease and Paediatric Cardiology, Kiel, Germany
[#]Brigham and Womens Hospital Boston, Boston, Massachusetts, United States of America

Introduction: Angiographic data indicate that children with Hypoplastic Left Heart Syndrome (HLHS) in Fontan-circulation have a smaller anatomical size of central pulmonary arteries. We utilised novel MRI-techniques to assess pulmonary vascular volume and lung perfusion to investigate whether HLHS patients have lower values than lung healthy controls.
Materials and methods: 31 children with HLHS (4.9±2.3 years) and 8 lung healthy controls (9.8±6.4 years) were studied. A modified contrast-enhanced dynamic MR-angiography sequence was used to assess pulmonary vascular and total volume of the right and left side of the lung. With use of dedicated software we measured the lung volume, relative pulmonary vascular volume, the rate of parenchymal contrast enchancement ("up-slope"), as well as the parenchymal mean transit time (MTT) in order to assess pulmonary vascular status and perfusion. Standardised cardiopulmonary exercise testing on a treadmill was performed in all patients and compared to a cohort of paediatric volunteers.
Results: The indexed pulmonary total lung volume and the relative vascular volume in HLHS were significantly reduced compared to the control group (634±105 vs. 945±274ml/m^2, p<0.05; 6.5±3.7 vs. 9.5±2.4%/m^2, p<0.05). Lung perfusion in children with HLHS was also impaired compared to healthy children (MTT: 10.8±2 vs. 6.7±3s, p<0.01; Up-slope: 0.32±0.24 vs 1.12±0.305s-1m-^{2}; p<0.01s-1m-^{2}; p<0.01). Relative vascular volume showed no difference between the right and left lung and correlated significantly with oxygen-uptake measured by standard spirometry (Spearman's r=0.78; p<0.01).
Conclusions: Advanced MRI methods enable the assessment of pulmonary perfusion in HLHS patients in Fontan-circulation. HLHS patients have a reduced total lung volume, pulmonary vascular volume, and an impaired lung perfusion, compared to controls. The reduced vascular volume in HLHS patients appears to be an important limiting factor for pulmonary function. Long term follow-up investigations are important to detect a potential further decline of values during growth and which implication may occur for the function of the Fontan-circulation.

Abstract no: 1062
Congenital long QT syndrome: A single-centre experience

Sevcan Erdem, Alev Kiziltas, Osman Kucukosmanoglu and Nazan Ozbarlas
Department of Paediatric Cardiology, Faculty of Medicine, Cukurova University, Adana, Turkey

Background: Congenital long QT syndrome (LQTS) is characterised by prolonged ventricular repolarisation (QT interval prolongation) and a propensity for syncope and sudden death secondary to Torsades de Pointes.
Method and materials: In this report we identified 16 congenital LQTS patient that followed by Cukurova University Department of Paediatric Cardiology from 2000 - 2012. The mean age of the patients was 104 months, the mean follow-up period was 24 months, mean corrected QT interval was 0.52msn. At the pre-diagnosis period 9 patients (56%) have syncope, convulsion or cardiac arrest history, and 3 patients were treated with anti-epileptic drugs that misdiagnosed as epilepsy.
Results: During the follow-up period, 2 patients received implantable cardioverter defibrillator (ICD). One of them was a boy with symptomatic bradycardia in Holter ECG record and cardiac arrest history of 1st degree family member, and the other was a girl with recurrent sustained ventricular tachycardia with syncope even though medical therapy. All patients treated with beta adrenergic blockade as soon as diagnosed LQTS, and potentiated through maximal heart rate on treadmill exercise test. During the follow-up period none of the patients had cardiac arrest. We diagnosed LQTS to 2 patient's asymptomatic 1st degree family member just with ECG screening.
Conclusions: Being the survivor of sudden cardiac arrest, history of recurrent syncope, history of sudden cardiac arrest in family members, and diagnosis of LQTS in family members and genetic determination must take into consideration during clinical follow-up of a congenitally LQTS. Beta Adrenergic blockade, avoidance of triggering factors, ICD or permanent pacemaker implantation for high risk patient, sympathetic ganglion blockade are treatment choose in LQTS. Physician must screen family members with ECG, and also must overemphasise the history of recurrent syncope attacks, convulsion or sudden cardiac arrest at patients with suspicion of LQTS diagnosis.

Abstract no: 1063
Value of radiofrequency ablation vs. cryo-ablation for atrioventricular nodal re-entry tachycardia in children

Matthew Oster[*,#,†], Zhou Yang[#], Kay Steward-Huey[*], Michelle Glanville[*], Arlene Porter[*], Robert Campbell[*,†], Brad Webb[*] and Margaret Strieper[*,†]
[*]Childrens Healthcare of Atlanta, Atlanta, Georgia, United States of America
[#]Emory University Rollins School of Public Health, Atlanta, Georgia, United States of America
[†]Emory University School of Medicine, Atlanta, Georgia, United States of America

Background: Radiofrequency ablation (RFA) and cryo-ablation are 2 strategies for treating atrioventricular nodal re-entry tachycardia (AVNRT) in children, but it is unclear which strategy may offer better value.
Materials and methods: We performed a retrospective cohort study of all AVNRT ablation cases for children (age≤18 years) from 1 July 2009 - 30 June 2011 at a single institution. Cost data included fixed cost, miscellaneous hospital costs, and labour costs. Outcomes data regarding the acute long term success (6 months) of the ablations were collected through chart review. T-test and regression analysis were conducted to investigate any association between ablation procedure type (RFA, cryoablation, or combination of both), and the cost and long term success rate of the ablation procedures, adjusting for patient (age, BSA) and provider (experience) characteristics.
Results: Of 96 unique cases, 48 were cryo-ablation only, 42 RFA only, and 6 were combination (4 starting with RFA, 2 starting with cryo-ablation). Acute success was 100% for the cryo-ablation-only and RFA-only cases and 83% for the combination cases. There were no notable adverse events. The average total cost was US$9 636 for cryo-ablation cases, US$9 708 for RFA cases, and US$10 967 for combination cases. The difference in the cost between cryo-ablation only cases and RFA only cases was not statistically significant. The unadjusted long-term success rate was 79.1% for cryo-ablation only cases, 92.8% for RFA only cases, and 66.7% for combination cases ($p<0.05$ for cryo-ablation only vs. RFA only). After adjusting for patient and provider characteristics, the difference between cryo-ablation only and RFA only was not considered significant.
Conclusions: There was no difference in costs or short term success rate for cryo-ablation vs. RFA for treatment of AVNRT in children. Differences in long term success rates may be attributed to multiple factors other than the choice of procedure and may lead to differences in long term costs if children are referred for repeat ablation.

Abstract no: 1064
Impact of BT shunt size on tricuspid regurgitation in hypoplastic left heart syndrome

Hiroko Asakai[*], Bryn Jones[*], Igor Konstantinov[#], Yves D'Udekem[#], Christian Brizard[#] and Michael Cheung[*]
[*]Royal Children's Hospital, Parkville, Victoria, Australia
[#]Royal Children's Hospital, Parkville, Victoria, Australia

Background: The prognosis for hypoplastic left heart syndrome (HLHS) has greatly improved over the past years, but there is still a significant risk of inter-stage mortality particularly between stages 1 and 2 of surgical palliation. Tricuspid regurgitation (TR) is known to be a risk factor for post-operative and inter-stage mortality. We hypothesised that a smaller BT shunt would lead to a reduction in ventricular volume loading and also a reduction in TR. This study aimed to evaluate the effect of BT shunt size on the severity of TR.
Methods: A retrospective review of all patients with HLHS who had a Norwood procedure in our institution from January 2006 - December 2011 was performed. Patient demographic, echocardiograms, MRI, operative, and post-operative data were reviewed. The impact of BT shunt size on severity of TR at the time of BCPS was analysed.
Results: In this time period, 64 neonates with HLHS underwent a Norwood procedure with a modified BT shunt at our institution. 32 (32/64) had a 3.5mm BT shunt (Group 1) and 32/64 had a 3.0mm BT shunt (Group 2). Survival to BCPS was 53/64 (83%) overall, with 25/32 (78%) in Group 1 and 28/32 (87%) in Group 2. The prevalence of significant TR (≥moderate) on pre-operative assessment in preparation for BCPS was 9/25 (36%) in Group 1 and 2/28 (7%) in Group 2 ($p=0.01$). Tricuspid valve repair was performed 7/25(28%) in Group 1 and 4/28 (14%) in Group 2 ($p=0.21$). The rate of non-fatal serious adverse events was similar in the 2 groups.
Conclusion: A smaller sized modified BT shunt at the initial Norwood procedure reduces the prevalence of significant tricuspid regurgitation. Further investigation and long term follow-up is required to determine other potential complications of this surgical strategy.

Abstract no: 1065

Effect of beraprost on pulmonary artery pressure in pulmonary hypertension due to left-to-right shunt congenital heart disease

Mahrus Rahman*, Teddy Ontoseno*, Alit Utamayasa*, Ery Oliviato* and Windhu Purnomo#

*Department of Child Health Medical School, Airlangga University, Soetomo Hospital, Surabaya, Indonesia
#Faculty of Public Health Airlangga University, Surabaya, Indonesia

Background: Late diagnosis and surgical management of left-to-right shunt congenital heart disease (L-R shunt CHD) caused pulmonary hypertension. To date there is no effective drug to decrease pulmonary artery systolic pressure. Beraprost an oral prostacyclin analogue which acts as a pulmonary artery vasodilator was expected to decrease PASPS in children with L-R shunt CHD.
Materials and methods: A pre-test/post-test study was conducted with 17 subjects aged 2 months-16 years old, with VSD, ASD, PDA or combination, who develop pulmonary hypertension. Patients with other structural cardiac defects or with pulmonary infection were excluded. Echocardiography examinations were performed before and after administration of beraprost 1 µg/kgBB/day for 3 months. Vmax TR, Vmax L-R, PASP were measured and M-mode of pulmonary valve including A-wave, EF slope and mid-systolic notch were examined. Adverse reactions were noted, monitoring was performed every 2 weeks.
Results: Vmax TR decreased 0.7±0.79m/second (p=0.004), Vmax L-R increased 0.6±0.64m/second (p=0.018) and PASP decreased 18.3±21.90mmHg (p=0.003). There were no significant changes in wave, EF slope and mid-systolic notch.
Conclusions: There was decrease of pulmonary artery systolic pressure after administration of beraprost in children with L-R shunt CHD who develop pulmonary hypertension.

Abstract no: 1067

Delayed uptake and washout of contrast in infarcted myocardium shown with 4-D CT in pigs

Sebastian Udholm*, Sofie Laugesen*, Peter Agger*, Jesper Honge*, Morten Smerup*, Morten Battcher# and Jens Michael Hasenkam*

*Department of Cardiothoracic & Vascular Surgery, Aarhus University Hospital, Skejby, Denmark
#Cardiac Imaging Centre, Aarhus University Hospital, Skejby, Denmark

Background: Assessment of ischaemic and potentially viable myocardium plays an important role in the planning of revascularisation of patients with critical coronary atherosclerosis. Cardiac computed tomography (CT) is being used increasingly in the functional- and morphological analysis of cardiovascular diseases and specifically in the screening and evaluation of atherosclerotic lesions in coronary artery disease. The aim of the present study was to investigate the pattern of contrast distribution over time in pigs with experimentally induced anterior myocardial infarctions to evaluate the feasibility of using simple CT enhancement as a viability marker.
Methods: This study was conducted as an animal experimental follow-up study. Twelve female pigs weighing 50kg were subjected to ischaemic myocardial lesions induced by 60 minutes of balloon occlusion of the left anterior descending artery immediately distal to the first diagonal branch followed by removal of the balloon and reperfusion. After 6 weeks, CT was performed using intravascular contrast agent. Measurements of radio density in Hounsfield units in the infarct zone and the non-ischaemic lateral wall were performed at 20 seconds, 1, 3, 5, 8 and 12 minutes after contrast injection.
Results: We found highly significant differences in radio density between the 2 zones at all time points except t=1 minute. The healthy myocardium showed a more rapid uptake and washout of contrast compared to the infarct zone. In particular the ratio between early and late uptake is a strong marker of viable myocardium.
Conclusion: The delayed uptake and washout of contrast agent in the infarct zone compared to the zone of healthy myocardium corresponds well with existing knowledge from MRI and other methods and most likely reflects the impairment of the vascular bed in the infracted myocardium. The present study shows promise for future clinical application of CT in a combined assessment of coronary anatomy and myocardial viability.

Abstract no: 1069

3-dimensional rotational angiography in patients with single ventricle after total cavo-pulmonary connection

Sebastian Goreczny*, Pawel Dryzek*, Tomasz Moszura*, Jacek J. Moll#, Andrzej Sysa* and Jadwiga A. Moll*

*Department of Cardiology, Polish Mother's Memorial Hospital, Research Institute, Rzgowska Lodz, Poland
#Department of Cardiac Surgery, Polish Mother's Memorial Hospital, Research Institute, Rzgowska Lodz, Poland

Background: Three-dimensional rotational angiography (3-DRA) is an emerging technology that could be a beneficial adjunct to fixed plane angiography and could enhance diagnostic capabilities in patients with single ventricle after various stages of palliation. We report our total experience using 3-DRA in patients after total cavo-pulmonary connection (TCPC).
Methods: A retrospective analysis of all patients after TCPC who underwent 3DRA was performed. Philips Allura system was used to acquire non-gated, breath-held images. During a 240 degrees, 4.1 seconds isocentric rotation, 122 angiographic images were acquired and automatically reconstructed in real time.
Results: Between September 2010 and June 2012, we performed 41 3-DRA's in 21 patients after TCPC. All but 1 patient underwent total of 32 interventions. Twenty four 3-DRA's were performed prior to the intervention, remaining 17 to asses results of percutanous treatment. Median age and weight was 9.13 years (4.1 - 24.1) and 24.8kg (13 - 56), respectively. Median contrast dose for 3-DRA and for total study was 1.45ml/kg (0.9 - 2.4) and 4ml/kg (1.5 - 9.8), respectively. Median area dose for the whole study, time of fluoroscopy and total time of study was 330.2 cGycm2 (144.8 - 1325.2), 15.2 minutes (9.3 - 54.4) and 70 minutes (55 - 165), respectively. Rotational angiography provided median of 96 projections (78 - 105) and the fallowing 3D reconstruction was successful in all studies. Both allowed for precise visualisation of the external tunnel, superior vena cava and pulmonary arteries. Overall quality of 3-DRA images was graded as good in 32 (78%) studies and satisfactory in 9 (22%). None of the 3-DRA's was graded as poor, however 8 additional angiographies (3-DRA = 3, fixed plane = 5) were performed to better visualise the left pulmonary artery.
Conclusions: After undergoing TCPC 3-DRA provided a large number of projections with relatively small amounts of contrast and allowed for perfect visualisation of the Fontan circuit.

Abstract no: 1083

Assessment of tricuspid annular plane systolic excursion in patients with hypoplastic left heart syndrome in Fontan-circulation

Eileen Pardun*, Dominik Gabbert*, Michael Jerosch-Herold#, Christopher Hart*, Inga Voges*, Miriam Michel*, Ana Andrade*, Minh Pham*, Hans-Heiner Kramer* and Carsten Rickers*

*Deparment of Congential Heart Disease and Paediatric Cardiology, Kiel, Germany
#Brigham and Women's Hospital Boston, Boston, Philadelphia, United States of America

Background: The tricuspid annular plane systolic excursion (TAPSE) is an established echocardiographic parameter to assess right ventricular (RV) systolic long axis function in biventricular hearts.
Aim: To investigate long and short axis function in patients with Hypoplastic Left Heart Syndrome (HLHS) utilising MRI.
Material and methods: Forty children (5.8±2.5 years) with HLHS in Fontan-circulation and 10 healthy controls (10.6±5.2 years) underwent a comprehensive cardiac MRI study to evaluate global and regional RV function. We used CMR cine-imaging (TR/TE/α = 1.1/1.6/60, FOV: 240 x 260) for analysis of global (EF, and cardiac index-CI) and regional ventricular function. TAPSE was analysed with in-house developed software that allowed 3-D reconstruction of ventricular geometry.
Results: HLHS patients had a lower TAPSE and cardiac index compared with healthy subjects. In contrast to healthy subjects TAPSE and RV-EF did not show significant correlation in HLHS patients with a rudimentary LV. In HLHS patients without a visible LV cavum we found a significant correlation between RV-EF and TAPSE (r=0.62; p=0.01). Furthermore, HLHS patients with a rudimentary LV had a significantly reduced septal wall motion and a cardiac index as compared with HLHS patients without a rudimentary LV (4.6±1.5 vs. 6.9±2.1mm; p=0.001; 3.4±0.8 vs. 2.8±0.9ml/m^2/min; p=0.01).
Conclusion: Patients with HLHS have impaired long-axis RV function compared to healthy controls. A rudimentary LV impairs in particular contraction in the septal segment, resulting in a reduced long-axis and global RV function. This may be of prognostic significance for the long term outcome in HLHS patients.

Abstract no: 1090

Can minor congenital mitral valve variants be differentiated from rheumatic mitral valve disease?

Nigel Wilson, Tom Gentles, Paul Tan, Anthony Concannon and John Stirling

Starship Children's Hospital, Auckland, New Zealand

Background: The echocardiographic features of congenital mitral valve prolapse and severe mitral rheumatic heart disease (RHD) are well established. Mild forms of these entities are harder to differentiate. A frequent clinical scenario in regions with high prevalence RHD is whether mild mitral regurgitation is of congenital or rheumatic aetiology.
Methods: Fifteen echocardiograms with mild mitral regurgitation (MR) were reported by 2 experienced observers and 2 trainee cardiologists without knowledge of patient demographics. The aetiology of the MR was judged congenital if there was classical mitral valve bi-leaflet prolapse, accessory mitral valve leaflet or scallop, diastolic excessive movement, or mitral valve cleft. 5 echocardiograms had minor congenital mitral valve features previously defined by a panel of 3 cardiologists.
Results: The range of congenital mitral anomalies diagnosed was 1/4 of 5 positive studies (Kappa 0.25 - 0.46).
Conclusions: Minor congenital mitral valve variations may cause mitral valve regurgitation but they are not easily differentiated from MR due to RHD. Minor congenital mitral valve anomalies can cause a false positive diagnosis of RHD.

Abstract no: 1102

Long QT molecular autopsy: Results of 6 years of a population-based clinical service for autopsy negative sudden death in 1 - 24 year olds

Jonathan Skinner*#, Jackie Crawford*, Warren Smith*, Paul Morrow*†, Ian Hayes*‡ and Donald Love*†

*Cardiac Inherited Disease Group New Zealand
#Starship Childrens Hospital , Auckland, New Zealand
†Lab Plus, Auckland District Health Board, New Zealand
‡National Clinical Genetic Service, New Zealand

Background/hypothesis: Families and forensic pathologists seek a diagnosis with urgency following the tragedy of a sudden, unexpected and unexplained death in children and youth. We report the results from 6 years of a national molecular autopsy service in New Zealand (population 4.3 million) focusing on this age group.
Methods: Following a national protocol, pathologist save DNA at all sudden unexpected natural deaths in young people, and refer for cardiac genetic opinion and molecular autopsy when the standard autopsy and ancillary tests are uninformative.
Results: Between April 2006 and February 2012, DNA was preserved on 132, 1 - 24 year olds who had died suddenly and unexpectedly without an overt cause at initial autopsy. In 67 cases, ancillary tests (histology/toxicology/microbiology etc.) revealed a cause of death (mostly infection and poisoning). The remaining 65 DNA samples underwent genetic sequencing of 6 genes linked to long QT syndrome or Brugada syndrome. 39/65 (68%) were of European ancestry and 17/65 (26%) Maori. Rare variants were identified in 13 of 65 cases (20%), 7 in SCN5A, 2 in KCNQ1 and one each in KCNH2, KCNE1, KCNE2 and KCNJ2. Nine of these 13 deaths (69%) were nocturnal. Uncertainty remains over the pathogenicity of 5 of these variants. Familial evidence of cardiac ion channel disease on cardiological testing is present in 5 families thus far.
Conclusions: Sudden unexpected natural death in 1 - 24 year olds occurs with a minimal incidence of 5 per million general population per year, half of which remain unexplained after standard autopsy. Abnormalities in genes linked to long QT and Brugada syndromes occur in 20% of these, with SCN5A being the commonest involved, and with most deaths occurring during sleep. Interpretation of the molecular genetic results remains a significant challenge and involves a multidisciplinary approach, including engaging with the family early in the process.

Abstract no: 1103

Detection of coronary artery lesions late after the arterial switch operation for transposition of the great arteries in children and adolescents: Use of low dose multi-detector computed tomography (MDCT)

Hopewell Ntsinjana*#, Oliver Tann*#, Graham Derrick#, Catherine Owens#, Silvia Schievano*# and Andrew Taylor*#

*UCL Institute of Cardiovascular Science, Centre for Cardiovascular Imaging, Los Angeles, California, United States of America
#Great Ormond Street Hospital for Children, London United Kingdom

Background: One of the most catastrophic late sequelae of the arterial switch operation (ASO) for transposition of the great arteries (TGA) is the development of coronary artery stenosis in approximately 7.2% of cases. The "gold standard" imaging modality to detect these complications is conventional invasive X-ray angiography. Non-invasive retrospective triggered 64-MDCT has been shown to be safe and accurate in some centres, albeit higher effective radiation dose than conventional angiography (4.5 vs. 3.1mSv).(1)
Aim: To assess the feasibility of a low dose MDCT in detection of coronary artery lesions with good image quality late after ASO for repair of TGA.
Materials and methods: Prospective study of 14 patients operated on for transposition of the great arteries with the arterial switch operation underwent MDCT angiography. Ethical approval to carry out this study was obtained from our institutional ethics committee, and all patients signed informed consent. Examinations were performed using prospective ECG-triggered study at a dual-source 64 row MDCT. No beta-blockers were used for these patients. Two independent investigators analysed MDCT angiography images for quality and the ability to detect coronary artery stenosis, anatomy and course. The effective dose was derived from the product of dose-length product (DLP) and conversion coefficient for the chest based on patient age using Shrimpton and Wall method.(2)
Results: The median age was 15 years (IQR 13.8 - 16.4 years) with 78.6% males and 21.6% females. Proximal coronary arteries were visualised in all patients despite step artifact in 2/14. Left coronary artery origin stenosis was identified in 2/14 patients. The mean dose-length product (DLP) was 40±9.4mGycm and mean effective radiation dose was 0.55±0.13mSv. The average heart rate was 74/minute.
Conclusion: Prospective ECG-triggered MDCT angiography provides good quality and interpretable images with an added advantage of lower radiation dose in coronary artery imaging in adolescents.
References: 1. P. Ou, et al. JACC: Cardiovascular Imaging 2008;1:331-339. 2. P.C. Shrimpton, et al. Radiation Protection Dosimetry 2000;90:249-52.

Abstract no: 1113

Management of haemostasis in paediatric patients after placement of ventricular assist device: Report from the Berlin EXCOR™ Paediatric Ventricular Assist Device IDE study

Patti Massicotte*, Marie Steiner# and Lisa Bomgaars†

*Stollery Childrens Hospital, University of Alberta, Alberta, Canada
#University of Minnesota, Minnesota, United States of America
†Baylor College of Medicine, Houston, Texas, United States of America

Background: Efficacious ventricular assist device (VAD) support in paediatric patients depends on successful haemostasis management, balancing the risk of thromboembolic complications with potential life-threatening bleeding. The experience with the haemostasis management algorithm developed for the EXCOR™ Paediatric IDE study is described.
Materials and methods: All patients enrolled in the study are included. Coagulation parameters and thromboembolic and haemorrhagic events were reviewed for relatedness to anti-thrombotic therapy. All events were centrally adjudicated. Evaluation of adherence to the haemostasis management algorithm was based on mean results of primary (PTT, INR, anti-factor Xa) and secondary (thromboelastogram, platelet mapping) laboratory tests at pre-specified times.
Results: 109 patients in primary study cohorts 1&2 (n=48), continued access patients (n=20) and compassionate use cohort (n=41) at IDE sites are included. Adherence to antithrombotic guidelines was acceptable. Pump change occurred in 52% of all patients; half were hypercoagulable based on lab or prior thrombotic history. Major bleeding occurred in 46% of all patients. Mean haemostatic parameters in patients with major bleeding trended to higher anti-factor Xa, PTT, and arachidonic acid platelet inhibition. Bleeding events were probably related to haemostasis management in 18% of total IDE group events. Neurologic events occurred in <30% of all patients; mean haemostatic parameters at the time of event trended toward lower anti-factor Xa and PTT on UFH, and higher net ADPG. Neurologic dysfunction was probably related in 9% and possibly in 42% events to antithrombotic therapy intensity.

TABLE 1: Targeted events

	All cohorts (n=109)	
	No. events	No. patients (%)
Pump change	114	57 (52%)
Major bleed	77	50 (46%)
Neurologic event	31	28 (26%)

Score: 1 - 5→mild, 6 - 10 →moderate, 10 - 15 →severe.

Conclusions: Implementation of a haemostatic algorithm is crucial to minimise VAD-related adverse events. Adherence to an algorithm was possible in the setting of a multicentre study. Incidence of significant bleeding and thromboembolic events is acceptable when balanced against potential life-saving benefits of the VAD. Additional algorithm refinements and a standardised approach may further improve safety and efficacy of the haemostatic management.

Abstract no: 1115

Reliability of echocadiographic parameters in predicting the grade of pulmonary regurgitation in patients with Tetralogy of Fallot: Echo compared to cMRI

Franziska Degener*, Nikolaus A. Haas*, Kai Thorsten Laser*, Axel Moysich*, Eugen Sandica#, Hermann Korperich† and Deniz Kececioglu*

*Department of Congenital Heart Disease, Bad Oeynhausen, Germany
#Department for Surgery of Congenital Heart Disease, Bad Oeynhausen, Germany
†Institute for Radiology, Nuclear Medicine and Molecular Imaging, Bad Oeynhausen, Germany

Introduction: In Tetralogy of Fallot the extent of pulmonary regurgitation (PR) and right ventricular (RV) volume load are the main criteria for long term assessment. Today the gold standard for evaluation is cardiac magnetic resonance imaging (cMRI). Reliable echocardiographic parameters for assessing PR are missing.
Methods: (1) Analysis of 219 sequential cMRIs and echocardiographic examinations in 118 patients over 1 year (53 female, 65 male, mean age 15.7±5.1 years, mean BSA 1.6±0.3m²). Data were obtained within the German competence network for congenital heart disease. (2) cMRI: RV and left ventricular volumes and ejection fractions were measured, PR was defined as mild (≤25%), moderate (>25 ≤40%) and severe (>40%). (3) Echocardiography: RV diameters in M-Mode, tricuspid valve annulus diameter, tricuspid annular plane systolic excursion (TAPSE), tricuspid regurgitation, right ventricular pressure (RVP), pressure half time (PHT), ratio of the PR jet width to the pulmonary valve annulus (PR/PVA) and the ratio of the regurgitation time divided by the diastolic time interval (PRi) were assessed.
Results: PR severity correlated with RV end diastolic volumes ($p<0.001$). TAPSE showed a significant difference between mild and moderate ($p=0.033$) as well as mild and severe PR ($p=0.004$). PHT showed similar results comparing the PR group mild to severe ($p=0.001$) and moderate to severe ($p=0.018$). Analogical results were obtained by the PRi with $p=0.028$ (mild/severe) and $p=0.022$ (moderate/severe) and the PR/PVA ratio between mild and severe PR ($p=0.049$).
Conclusion: With increasing severity of PR there is a decline of RV function measurable by TAPSE. A short PHT and PRi indicate a more rapid equilibration of RV and pulmonary artery pressure and consequently a shorter PR time and PR/PVA ratio. TAPSE, PHT, PRi and PR/PVA ratio are reliable and reproducible echocardiographic tools to assess the severity of PR.

Abstract no: 1117

Electrocardiographic changes after carvedilol therapy of congestive heart failure in children with left-to-right shunt congenital heart disease

Teddy Ontoseno

Dr Soetomo Hospital, Surabaya, Indonesia

Background: Heart failure (HF) is the most common complication of left-to-right shunt congenital heart disease (L-R shunt CHD). Sympathetic activity and stimulation of rennin angiotensin aldosterone system are the compensation. Excessive and prolonged compensations lead to heart remodelling. Medical treatment is necessary in optimising the heart function. Carvedilol is a non-selective beta-blocker, used as additional drug in HF. The purpose of this study is to determine effects of carvedilol on changes in heart rate, R-wave amplitude in V6, S-wave in V1 and R/S ratio in V1.
Materials and methods: Randomised controlled trial study, 30 HF L-R CHD, divided into 2 groups: Treatment group 16 children with carvedilol + standard therapy (ACE-I and diuretic); and control group 14 children with standard therapy. ECG records performed before and after therapy. The changes in heart rate, R-wave amplitude in V6, S-wave amplitude in V1 and R/S ratio in V1 were measured. Adverse effects were monitored. Study duration lasted for 3 months.
Results: Heart rate decreased significantly in the treatment group compared to the control group, 17.7% vs. 4.1% ($p=0.002$). R-wave amplitude in V6 decreased significantly in the treatment group compared to the control group, 23.6% vs. 7.5% ($p=0.03$). S-wave amplitude in V1 decreased significantly in the treatment group compared to the control group, 27.9% vs. 10.1% ($p=0.008$). R/S ratio in V1 increased significantly in the treatment group compared to the control group, 51.6% vs.15% ($p=0.31$). No adverse effects were found.
Conclusions: Carvedilol is effective in improving ECG parameters and safe as therapy additional to the standard therapy for CHF in children with L-R shunt CHD.

Abstract no: 1120

Management of haemostasis in patients after placement of a ventricular assist device: Use of point-of-care INR monitoring to manage Vitamin K antagonists

Patti Massicotte*, Mary Bauman*, Aisha Bruce*, Holger Buchholz† and Stefan Kuhle#

*Stollery Childrens Hospital, University of Alberta, Edmonton, Canada
#Obstetrics and Paediatrics, Halifax, Canada
†Mazankowski Heart Institute, University Hospital, Edmonton, Canada

Background: Vitamin K antagonists (VKA), required for haemostasis management in patients with ventricular assist devices (VADs) must be regularly monitored to avoid both thrombotic and bleeding complications. The gold standard monitoring method is the prothrombin time (PT) testing of plasma collected via venipuncture and expressed as an INR. The point of care POC-INR monitor is convenient for patients, uses a capillary whole blood sample and produces an INR within minutes. The POC INR monitor is demonstrated to be accurate in adults and children requiring VKA therapy facilitating more frequent INR testing; a requirement for VAD patients.
Methods: Prospective cohort study including all patients with a VAD treated with VKA to determine the accuracy of the CoaguChek XS® by comparing whole blood INR results from the CoaguChek XS® to plasma laboratory INR results. Meters were loaned to patients for use. Prior to patient use, comparisons between POC-INR and lab-INR were performed. 158 meter validations were performed from 2 time points of 16 patients receiving warfarin by drawing a venous blood sample for laboratory PT-INR measurements and simultaneous INR using the POC-INR meter. Agreement between CoaguChek XS® and laboratory INR was assessed using the concordance correlation coefficient (CCC).

Results: VKA therapy for VAD thromboprophylaxis was received by 6 children, median age 4.1 years (range 3.1 - 15.5 years) and 10 adults, median age 51.2 years (range 18.6 - 63.0 years) who had n=5 EXCOR, n=1 Heartware, n= 1 EXCOR, and n=9 Heartmate VADs, respectively. There were a mean of 13 (range 2 - 27), 6 (range 4 - 14), and 9 INR tests per patient for the Berlin Heart, Heartmate and Heartware, respectively. The CCC was 0.65 and 0.72 in children and adults, respectively.
Conclusions: The CoaguChek XS® meter readings showed a moderate to good agreement when compared to laboratory INR test results.

Abstract no: 1125
Natriuretic Peptide (proBNP) as a marker of cardiovascular disease and connatal infection in pre-term infants

Juan Pablo Miranda[*,#], Mauricio Pinto[*], Rodrigo Diaz[*], Ronald Leiva[*,#], Gabriel Penaloza[*], Freddie Rybertt[*], Rodrigo Salas[*,#], Jose Antonio Salinas[*], Yalda Lucero[*] and Magdalena Castro[*]

[*]Clinica Las Condes, Santiago, Chile
[#]Hospital Barros Luco, Santiago, Chile

Introduction: Natriuretic peptide (proBNP) is a molecule secreted by the myocardium in response to pressure or volume overload.
Objective: Determine the relationship between proBNP levels and the presence of cardiovascular disease and/or peri-natal infection in pre-term neonates.
Patients and methods: A prospective study including pre-term neonates (<32 weeks and/or <1 500g) was conducted at the Neonatal Unit of Las Condes Clinic from June 2011 - June 2012. ProBNP levels were determined between 48 - 72 hours after birth by electrochemiluminescence assay. Echocardiogram, CBC, CRP and proBNP tests were performed. Patients were divided into 4 groups according to the presence of cardiovascular disease (PDA >1mm, CHD, cardiomyopathy and/or pulmonary hypertension) and/or connatal infection. The study was approved by the local IRB. Logistic regression and ROC analysis were performed to determine the predictive value and a cut-off point for proBNP.
Results: Fifty three patients were recruited with a median gestational age of 29.6 weeks (range 24 - 33 weeks) and a mean birth weight of 1 316.4g (750 - 1890). The median proBNP levels were 5 215pg/ml (662 - 100 080) for the entire study group; 17 691pg/ml (3 761 - 100 080) for patients with cardiovascular disease and 2 855pg/ml (662 - 6 750) ($p<0.005$) for patients without cardiovascular disease. ProBNP levels of 17 730pg/ml corresponded to patients with associated infection and 3 184pg/ml ($p<0.005$) to those without infection. To detect cardiovascular disease, proBNP levels with 80% sensitivity, 79% specificity, a positive predictive value of 60% and a negative predictive value of 91% were obtained based on the ROC curve analysis and a cut-off value of 9 970pg/ml. By regarding proBNP to detect peri-natal infection, 92% sensitivity and 73% specificity were obtained with a cut-off value of 7 522pg/ml.
Conclusions: The results in this study suggest the need for proBNP measurement to accurately predict cardiovascular disease in premature newborns. Possible peri-natal infection of the patient should be considered.

Abstract no: 1131
Systemic venous anomalies in the Middle East

Antonio Corno, Sami Alahdal and Karunamoy Das
King Fahad Medical City, Riyadh, Saudi Arabia

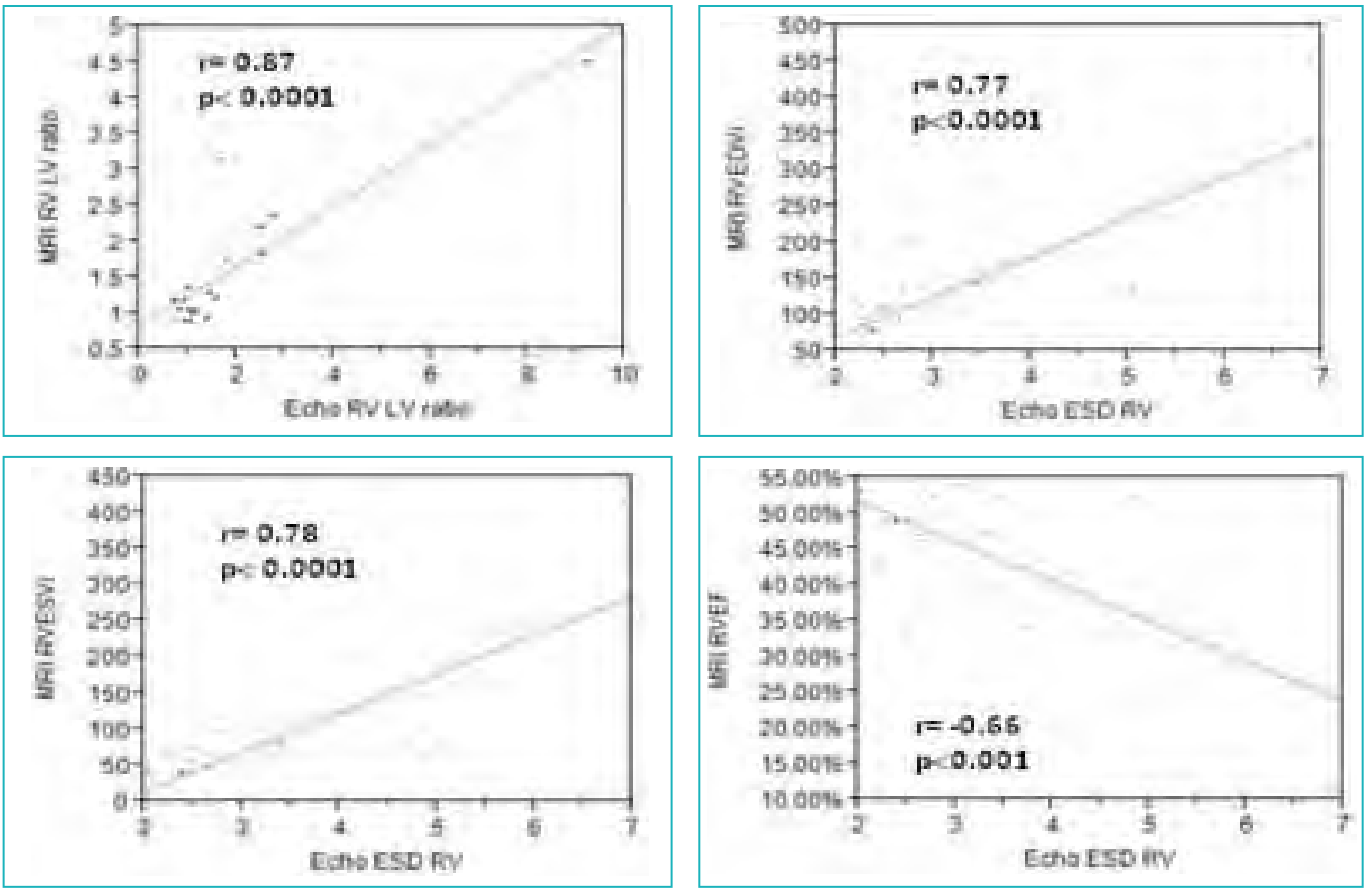
Introduction: The purpose of this study was to evaluate the incidence of systemic venous anomalies in patients requiring congenital heart surgery in the Middle East.
Materials and methods: From September 2010 - March 2012, 155 consecutive patients who had surgery for congenital heart defects were reviewed for the presence of the following systemic venous anomalies: (1) persistent left superior vena cava (PLSVC); (2) inferior vena cava (IVC) interruption; and (3) retro-aortic innominate vein. The modalities utilised for the diagnosis were also reviewed: echocardiography, computed tomography (CT) scan or intra-operative.
Results: Systemic venous anomalies were identified in 28/155 patients (18.1%). PLSVC was present in 21 patients (13.5%), median age 4 months (range 3 days - 18 years), median body weight 6kg (range 2.6 - 34.0kg). IVC interruption was identified in 5 patients (3.2%), median age 2 months (range 30 days - 26 years), median body weight 3.7kg (range 2.3 - 68.0kg). Retro-aortic innominate vein was diagnosed in 3 patients (1.9%), mean age 5 years (range 10 months - 5 years), mean body weight 12kg (range 4.5 - 14kg). The diagnosis was established pre-operatively in 22/28 (78.6%) patients: in 14/28 (50%) with echocardiography and in 8/28 (28.6%) with CT scan. In 6/28 (21.4%) patients the diagnosis was made intra-operatively. The incidence of the systemic venous anomalies in this study was higher than previously reported in the literature. Compared to literature a total incidence of systemic venous anomalies of 18.1% (vs. 4 - 14%), with PLSVC of 13.5% (vs. 4 - 11%), IVC interruption of 3.2% (vs. 0.6 - 2%), and retro-aortic innominate vein of 1.9% (vs. 0.2 - 1%).
Conclusions: This study showed a higher incidence of systemic venous anomalies in the Middle Eastern population with congenital heart defects than in previous literature reports. In a substantial percentage of patients (21.4%) the diagnosis was intra-operative. A better pre-operative screening should be performed in all patients with congenital heart defects to identify all systemic venous anomalies for a more precise planning of the surgical approach.

Abstract no: 1134
Echocardiographic right-to-left ventricular ratio in systole correlates with cMRI measurements in children with pulmonary hypertension

Pei-Ni Jone, Brian Fonseca, Dunbar D. Ivy and Adel Younoszai
Paediatric Cardiology, Children's Hospital Colorado, University of Colorado, Colorado, United States of America

Background: Pulmonary hypertension (PH) increases right ventricular (RV) pressure resulting in RV dilation and ventricular septal shift towards the left ventricle (LV). A ratio of systolic RV/LV diameter seeks to combine these effects into a single measure and has been shown to correlate well with pulmonary vascular resistance in children with PH.
Aim: In this study we seek to validate the measure by comparing this ratio to the identical ratio by cardiac MRI (cMRI) as well as cMRI indices of biventricular volume and function.
Methods: Seventeen children with PH, median age 12 years, (4 - 23 years), had 18 echocardiograms and CMRs within 72 hours. Echocardiographic data included: RV end-systolic diameter (RVESD), LV end-systolic diameter (LVESD) (parasternal short axis) and RV/LV ratio. CMR data included: RVESD, RV end-diastolic

volume, RV end-systolic volume, RV ejection fraction (RVEF), LVESD, LV end-diastolic volume, LV end-systolic volume, LV ejection fraction (LVEF), and RV/LV ratio. Echocardiographic measures were correlated with CMR.
Results: Echocardiographic RV/LV ratio correlates significantly with CMR RV/LV ratio in the Figure. However, systolic RV/LV ratio does not correlate with CMR indices of RV or LV size or systolic function. Echocardiographic RVESD correlates significantly with CMR RV volume and function in the Figure. Although echocardiographic LVESD had a negative correlation with CMR LVEF (r=-0.62, p<0.001), there were no significant correlations between echocardiographic LVESD and CMR LV volumes.



Conclusion: There is good agreement between the systolic RV/LV ratio obtained by echocardiography and CMR. Interestingly, echocardiographic RVESD in isolation appears to be an easily obtainable and accurate descriptor of RV size and function in children with PH.

Abstract no: 1137

Clinical performance of small calibre high voltage implantable cardioverter defibrillation (ICD) leads in children and young adults

Maully Shah, Karen Smoots, Akash Patel and Christopher Jansen
The Children's Hospital of Philadelphia, University of Pennsylvania, Philadelphia, Pennsylvania, United States of America

Background: Recent reports have called attention to ICD lead-related adverse events with small calibre leads.
Aim: To define the incidence of lead-related adverse events of small calibre ICD leads at a single site paediatric centre.
Methods: Clinical and lead performance information was collected retrospectively of patients ≤30 years of age with small calibre right ventricular (RV) ICD lead implantation from 1995 - 2011. Small calibre ICD leads were defined as lead diameter ≤7.6 French, and standard ICD leads were of diameter ≥8.6 French.
Results: Out of 142 patients with RV ICD lead implantation, 37 (26%) had small calibre ICD leads, and 85% of these were single chamber ICDs. The most frequent small calibre ICD lead implanted was Medtronic Sprint Fidelis 6931 (Medtronic, Minneapolis, MN) in 43% (n=16), followed by Medtronic Sprint Fidelis 6949 in 24% (n=9), Riata 1582 (St. Jude Medical, St. Paul, MN) in 19% (n=7), and Riata ST 7002 in 14% (n=5). In addition, 105 patients had standard calibre ICD leads. The mean age at ICD implantation was 15.8±5.7 years, with a mean follow-up time of 3.4±1.6 years. All implantations were associated with acceptable lead performance at initial implant. Lead fractures occurred in 13 (35%) patients at an average of 3.1±1.2 years after ICD implantation. Medtronic Sprint Fidelis 6931 and 6949 leads were most frequently affected (92% of all conductor fractures). When compared to standard-sized leads (5 lead failures), small calibre ICD leads have a significantly increased complication rate: 35% vs. 5% (p<0.05).
Conclusion: ICD lead-related complication rates were higher with small diameter ICD leads than standard calibre ICD leads. The lead-related adverse events were more frequent in the Sprint Fidelis leads when compared to the Riata leads, emphasizing that lead design in addition to diameter size may play a significant role in lead performance.

Abstract no: 1139

Cardiovascular magnetic resonance in patients with repaired Tetralogy of Fallot: The goal standard in assessment of injectable pulmonary valve implantation and follow-up

Stefano Marianeschi[*], Aurelio Secinaro[#], Benedetta Leonardi[†], Giacomo Pongiglione[†], Antonio Amodeo[‡], Simone Ghiselli[*], Nicola Uricchio[*], Stefano Pedretti[§], Giuseppe Annoni[◊] and Alberto Roghi[Δ]

[*]Congenital Heart Surgery, Niguarda Hospital, Milan, Italy
[#]Radiology Department, Bambino Gesã¹ Hospital, Rome, Italy
[†]Paediatric Cardiology Department, Bambino Gesã¹ Hospital, Rome, Italy
[‡]Paediatric Cardiac Surgery, Bambino Gesã¹ Hospital, Rome, Italy
[§]Cardiology Department, Niguarda Hospital, Milan, Italy
[◊]Paediatric Cardiology Department, Niguarda Hospital, Milan, Italy
[Δ]Imaging Cardiology Department, Niguarda Hospital, Milan, Italy

Background: Severe pulmonary regurgitation, progressive dilatation and dysfunction of the right ventricle are the most frequent late morbidity post Tetralogy of Fallot repair. Pulmonary valve replacement is often indicated in these patients. Bio-integral injectable pulmonary valve (IPV) is an innovative less invasive technique, often done off cardiopulmonary bypass (CPB). Cardiovascular Magnetic Resonance (cMRI) is fundamental to assess patient suitability for IPV insertion and to control the follow-up.
Material and methods: From January 2006 - June 2012 we performed 10 pre-operative CMRs. Five of these patients also underwent a CMR 3 months - 6 years post-IPV insertion. We measured the diameters of: right ventricular patch, pulmonary valve and pulmonary bifurcation and the length of the pulmonary trunk.
Results: Ten patients with implanted IPV 3 months - 6 years post-IPV insertion CMR showed an improvement of right ventricle end diastolic volume. The IPV was continent and the mean transvalvular gradient was below the gradient of traditional pulmonary valve prosthesis.
Conclusions: cMRI is a safe and effective method. It is necessary before IPV insertion to exclude contra-indications and to determine the need for CPB. In the follow-up cMRI should measure the pulmonary valve efficiency, the transvalvular gradients and right ventricle function. IPV is also better detected by cMRI than traditional prosthesis valve that present focal artefacts that can obscure small jets.

Abstract no: 1143

Double-chambered right ventricle: Clinical and echocardiographic characteristics of a series of 11 cases

Emilia Josefina Patio Bahena, Nilda Espinola- Zavaleta, Mirna Yabur, Maria Elena Soto, Juan Calderón and Alfonso Buendia

Instituto Nacional de Cardiología, Mexico

Introduction: The double-chambered right ventricle (DCRV) is an uncommon congenital heart disease and represents 0.5% of these congenital cardiopathies.
Objective: A series of 11 cases with diagnosis of double-chambered right ventricle is presented frompaediatric age to adulthood (2 - 64 years).
Material and methods: The median age of studied patients was 10 years (range: 2 - 64). 36.45% of cases were asymptomatic and the diagnosis was made because of an incidental finding of cardiac murmur. In other cases patients did not thrivein the lactating stage and in older children a worsening of NYHA functional class was observed. The echocardiogram showed double-chambered right ventricle by a muscular band in all studied patients. The most frequent associated anomalies were: ventricular septal defect, patent foramen ovale, subvalvular aortic stenosis, and Tetralogy of Fallot. Seven patients went to surgical treatment and now 6 of them are in NYHA functional class I.
Conclusions: This abstract is of special importance as it represents the biggest series of patients with double-chambered right ventricle studied in Mexico. Results are also comparable to the studies reported in the literature. Worsening in the NYHA functional class was the predominate symptom. The clinical manifestations can be presented from lactating babies to adulthood and the degree of obstruction could have a progressive character. Echocardiography is the method of choice in the diagnosis of these patients. 91% of patients with DCRV who went for surgical treatment had excellent haemodynamic and functional results at mid-term follow-up.

Abstract no: 1144

Mortality and morbity in young pacemaker and intracardiac defibrillator carriers. A focus on epicardial vs. endocardial strategy in long term follow-up

Luca Tomasi[*], Alessandra Cristofaletti[*], Luca Barozzi[#], Paolo Moggio[*], Maria Antonia Prioli[*], Lucia Rossetti[*], Fabrizio De Rita[#], Giovanni Battista Luciani[#], Giovanni Morani[*] and Corrado Vassanelli[*]

[*]Department of Cardiology University Hospital Civile Maggiore, Verona, Italy
[#]Department of Cardiosurgery University Hospital Civile Maggiore, Verona, Italy

Background: The choice of the most appropriate procedural technique for paediatric cardiac pacing is still debated.
Aim of the study: To evaluate the outcome, in terms of mortality and morbidity, of young PM or ICD carriers.
Patients and methods: All PM or ICD carriers in our centre, aging less than 18 years were included. Indications for device therapy, peri-procedural complications and long term follow-up were revised. An echocardiogram was performed in patients with isolated congenital atrio-ventricular block (iAVB) focused on the presence of aortic dilation.
Results: Thirty four patients, 28 with PM and 6 with ICD, were followed for a median time of 7.8 and 2.4 years respectively. The overall survival in patients carrying an ICD was 100%, 93% ± 5% in the PM group. All 4 deaths were complex congenital heart disease patients (CHD). The major complications were lead failures (19%) and infections (12%). The complication-free survival was 57±12% at 10 years in all patients. Infections were responsible for a worse outcome in CHD. Time to first complication was significantly shorter for epicardial pacing systems (3.35 years vs. 5.63 years, p=0.016). The difference between epicardial and endocardial systems was not significant in terms of complications and electrical parameters. iAVB was associated with significant dilation of the ascending aorta (median z-score 2.32 Q1-Q3:0.55-3.73, p=0.047).
Conclusions: PM/ICD carriers experience a significant number of complications. Systemic infections are responsible for the majority of deaths, especially in patients with CHD and endocardial leads. For this reason epicardial leads used as long as possible in CHD patient might be a reasonable choice. The challenge for the future is to increase longevity of pacing systems and to reduce number of re-intervention. The positive correlation between dilation of the ascending aorta and iAVB in stimulated hearts needs further investigations.

Abstract no: 1145

Using remote monitoring of implantable cardiac defibrillators and pacemakers in a paediatric tertiary care centre

Maully Shah*, Karen Smoots* and Jason Imundo#

*The Children's Hospital of Philadelphia, University of Pennsylvania, Pennsylvania, United States of America
#Hershey Children's Hospital, Pennsylvania State University, Pennsylvania, United States of America

Background: The number of paediatric and adult congenital heart disease patients with implantable cardiac defibrillators and pacemakers continues to grow annually. Cardiovascular implantable electronic devices (CIED) require regular follow-up to ascertain technical integrity. There is a paucity of published literature in the paediatric population regarding remote monitoring in patients with CIED.
Aim: The primary objective was to describe the use of remote monitoring in paediatric patients with CIED at a large tertiary care children's hospital. The secondary objective was to determine the potential clinical benefit to patient safety and care, and determine if remote monitoring decreases the number of hospital visits due to CIED-related issues.
Methods: A single centre, retrospective, observational chart review was performed. Data was obtained and reviewed for patients who have CIED and are followed at The Children's Hospital of Philadelphia from January 1991 - November 2011.
Results: Fifty patients were included: 15 with pacemakers, 35 with implantable cardiac defibrillators. The mean age was 19 years. The data sent by the remote monitoring system was monitored for 12 months to determine clinically actionable events (CAE). Of 633 total transmissions 41 resulted in a CAE in 19 patients. The CAE were comprised of the following: 29 arrhythmias, 5 lead malfunctions and 7 other issues. In the pacemaker group, the alerts that were most frequently activated were those concerning atrial tachyarrhythmia, battery exhaustion, lead impedance as well as sensing and threshold measurements. In the ICD therapy group, the alerts nearly always activated were those concerning a detection setoff, battery exhaustion or critical values of impedance.
Conclusions: Remote monitoring is a reliable method for following-up patients with CIEDs. Accurate alert settings personalised to the patient's clinical features are essential for effective management and avoidance of excessive flow of data.

Abstract no: 1146

Unusual atrio-fascicular accesory pathway in an adolescent with pre-excitation and long term tachycardia

M. Cecilia Gonzalez*, Thierry Sluysmans*, Jean-Benoit le Polain de Waroux# and Christophe Scavee#

*Paediatric Cardiology, Cliniques Universitaires St Luc, Brussels, Belgium.
#Cardiology Department, Cliniques Universitaires St Luc, Brussels, Belgium

Background: Mahaim fibers are rare decremental and antegrade only fibers connecting the right atrium to the right ventricle. We present an extremely rare case of an atrio-fascicular fiber with proximal insertion in a para-Hissian position.
Case presentation: A 17-year-old patient with pre-excitation and history of tachycardia leading to heart failure at age 2 was referred with recurrence of the arrhythmias despite therapy.
The Electrophysiology study showed at baseline a negative HV interval in sinus rhythm. Decremental atrial pacing lead to progressive AH prolongation and loss of pre-excitation with normalisation of the HV interval. Dual AV nodal physiology was excluded. Atrial stimulation induced a usual wide complex tachycardia with persistence of a retrograde His. Late atrial extrastinuli during tachycardia advanced the QRS complex proving pre-excitation of the ventricle via the AP. Atrial mapping on sinus rhythm indentified the earliest signals in the right antero-septal position. On the ventricular site, the earliest potentials were identified on the moderator band. Ventricular pacing excluded retrograde accessory pathway conduction. Cryo and RF ablation of the atrial insertion were unsuccessful. We finally cryoablated successfully the ventricular insertion over the right bundle potential, leaving the patient with a right bundle branch block on the surface ECG but otherwise normal His and infraHis conduction. Unsurprisingly given the chosen technique, pre-excitation recurred overnight. The patient requested a 6 month period before attempting a distal insertion RF ablation.
Conclusion: The case of para-Hissian atrio-fascicular connection represents an extremely rare finding and a further challenge for the electrophysiologist in terms of correct diagnosis as well as therapy. In our patient, a precise diagnosis was done by pacing maneuvers. Recurrence after cryoablation of the moderator band confirms once again that this approach is of little long-term success in the setting of accessory pathways.

Abstract no: 1147

Arrhythmias in patients with Tetralogy of Fallot: A national database study

Mei-Hwan Wu*, Hui-Chi Chen#, Jou-Kou Wang*, Chun-Wei Lu* and San-Kuei Huang†

*National Taiwan University Hospital, Taipei, China
#Genomics Research Centre, Academia Sinica, Taipei, China
†Bureau of National Health Insurance, Taiwan

Objective: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease and the incidence was 0.6/1 000 live births in Taiwan. Though the surgical outcome is generally good, the long term mobility and mortality are increasingly recognised. A population study from Taiwan, a country with national health insurance, easily accessible medical care and child health index similar to that of the U.S.A. would adequately reflect the long term risk.
Methods: We retrieved the data of ToF patients from our national health insurance 2000 - 2010 database.
Results: From an average 23 million population, there were 6 073 ToF patients: a prevalence rate of 44.2/100 000 (95%CI, 42.4 - 45.9) for paediatric population (aged <18 years) and 9.5/100 000 (95%CI, 9.1 - 10.0) for adults. Among them, 224 (3.8%) patients had arrhythmias during the study period spanning over 11 years, including 165 (74%) patients with tachycardia and 59 (26%) patients with bradycardia. Male dominance was noted for all types of arrhythmias, particularly

for ventricular tachycardia and sudden cardiac death. The mean age of each type of arrhythmia was highest in patients with atrial fibrillation (incidence= 0.7%, n=45, 44.4±15.6 years), followed by bradycardia-tachycardia syndrome (0.07%, n=4, 28.9±10.9 years), atrial flutter (0.18%, n=11, 29.7±17.8 years), sick sinus syndrome (0.13%, n=8, 27.9 ±16.6 years), ventricular tachycardia (0.46%, n=28, 24.4±15.3 years), supraventricular tachycardia (1.19%, n=72, 16.4±15.8), and atrioventricular block (0.77%, n=47, 15.8±13.9 years). Arrhythmia intervention was performed in 17 patients during the study period (RFCA in 12, ICD in 3 and pacemaker in 26), given an annual risk of arrhythmia intervention of 0.028%.
Conclusion: In this Asian national cohort spanning over 11 years, arrhythmias occurred in 3.8% of ToF patients, particularly in males. Tachycardia account for almost 3/4 of the arrhythmias and was a growing problem during long term follow-up.

Abstract no: 1148
Dextrocardia and associated cardiac malformations: Experience from a tertiary centre in Saudi Arabia

Muhmmad Arif Khan, Yahya Almasham, AbdurRehman Almoukirish, Omar Galal and Tarek Momenah
Prince Salman Heart Centre, King Fahad Medical City, Riyadh, Saudi Arabia

Introduction: Dextrocardia is an abnormal congenital positioning of the heart in the chest with the apex pointing towards right side. Isolated dextrocardia without other cardiac lesion does not require treatment. Dextrocardia with situs solitus or situs ambiguous is usually associated with complex congenital heart disease and require attention.
Objective: To determine the frequency of dextrocardia and associated cardiac defects in children presented at our institution.
Materials and methods: Retrospective study. All patients diagnosed with dextrocardia from January 2008 - July 2012 and referred to our tertiary cardiac centre were included in the study and their charts and echocardiograms were reviewed.
Results: A total of 8 648 patients were seen during this period: total number of new patients 2 657 (30.72%) of total. Dextrocardia was found in 30 (0.35%) of total cohort. Of dextrocardia cases 14 (46.7%) had situs solitus, 14 (46.7%) situs inversus and 2 (6.6%) situs ambiguous. The dominant were male with male to female ratio 60:40. Majority 25 (83.4%) of patients had associated congenital heart disease while 5 (16.7%) had no congenital heart. Of those with structural heart defects: 13 (52%) had situs solitus, 10 (40%) situs inversus and 2 (8%) patients fell in the situs ambiguous group.
Conclusion: The frequency of dextrocardia is 0.35% in our study which is in accordance with rest of world (0.2 - 0.8%). In addition congenital heart disease was more common in dextrocardia with situs solitus or situs ambiguous.

Abstract no: 1152
Risk of adverse cardiac events in children and adolescents with severe rheumatic left-sided valvar lesions

Maria Concepcion Sison, Edison Ty, Cecilienne Acosta, Jonas Del Rosario, Vincent Aluquin and Olympia Malanyaon
University of the Philippines, Philippine General Hospital, Manila, The Phillipines

Background/hypothesis: Rheumatic heart disease (RHD) and rheumatic fever (RF) continue to be a scourge in developing nations. A number of patients who need valvar surgery cannot afford the intervention. With limited resources, government hospitals need to prioritise patients needing immediate surgery.
Aim: To determine demographic and echocardiographic variables which are risk factors for the development of adverse cardiac events (ACE) in children with severe rheumatic left-sided valvar lesions.
Method: 376 echocardiograms of paediatric (<19-years-old) patients with RHD done at the Philippine General Hospital from January 2002 - December 2003 were reviewed. Based on the latest echocardiograms of 296 patients, they were grouped according to the left-sided valve most severely affected. Demographic and echocardiographic parameters of patients with ACE (death, AF, thrombus formation, and ≥2 admission/2-year period) were compared with those without ACE among patients with the same left-sided valvar lesion. A p-value <0.05 was considered significant.
Result: The following factors were found to increase the risk of ACE: (1) Severe mitral stenosis (p<0.0001); (2) Severe MR with concomitant moderate MS, severe TR, LVESD ≥3.5cm (RR=2.44; 1.27 - 4.68; p= 0.006), or LVEDD ≥5.5cm (RR=2.19; 1.15 - 4.14; p= 0.01); (3) Severe AR with an EF <60% (RR=5; 1.15 - 21.78; p=0.03), or LVESD ≥4.5cm (RR=4.17; 1.17 - 14.8; p = 0.04); and (5) moderate TR or PR in patients with combined severe MR and AR. Patients with severe MR and ACE were also found to be older (14.35±2.42 vs.12.65±3.3) and had larger LA size (5.18±1.05 vs. 3.99±1.14). The absence of PR in patients with combined severe MR and AR conferred significant freedom (p<0.05) from ACE. Based on these findings, amongst the cohort of 178 paediatric RHD patients with severe left-sided valvar lesions and who were still alive at the time of study at least 88 patients (49.4%) were in immediate need of surgery/ intervention.
Conclusion: The risk of adverse cardiac events in children and adolescents with RHD appear to be associated with the severity and nature of valvar involvement and, depending on the valvar lesion, on certain demographic and echocardiographic variables which could guide the clinician regarding timing of surgery.

Abstract no: 1155
Dilated cardiomyopathy with severe left ventricular systolic dysfunction in a patient on highly active antiretroviral therapy

Twalib Aliku*,#, Sulaiman Lubega# and Peter Lwabi#
*Department of Paediatrics, Gulu University, Uganda
#Uganda Heart Institute, Kampala, Uganda

Background: Cardiovascular dysfunction is a recogniwed complication of HIV infection in children. Cardiac complications of HIV usually occur late in the course of the disease or may be associated with drug therapy and hence become more common as therapy and survival improve. Left Ventricular (LV) dysfunction at baseline is a risk factor for death independent of the CD4 cell count, HIV viral load, and neurological disease.
Clinical case: We present the case of a 15-year-old girl diagnosed with HIV and subsequently started on highly active antiretroviral therapy (HAART) who presented with a 2-month history of cough, orthopnea and haemoptysis. She had been on zidovudine, lamivudine and Nevirapine for the previous 5 years.

Clinically she had pedal oedema, a tachycardia of 118 beats per minute, BP=106/79mmHg, a laterally displaced apex, a 3rd heart sound, basal crepitations and a tender hepatomegaly. She had a total lymphocyte count of 1 624cells/ml, ESR of 103mm/hr and the CRP was 132mg/dl. Her CD4 count was 51cells/ml. The viral load was 36 869 copies/ml. ECG showed a sinus tachycardia with diffuse ST-T segment changes and LVH with strain. Echo revealed dilated left heart chambers with severe LV systolic dysfunction and a small pericardial effusion. She was started on heart failure medicines. Two weeks later her symptoms had markedly improved but she was getting transient weakness of her left limbs. A repeat echo showed an intracardiac thrombus in the LV apex. Anticoagulation was started. Her HAART regimen was switched to abacavir, lamivudine and lopinavir. Six months after diagnosis, she was asymptomatic with normal LV size and normal systolic function on echo. Her CD4 count had risen to 360.
Conclusion: Early cardiology referral of patients on HAART with suspected cardiac dysfunction could lead to better treatment outcomes.

Abstract no: 1157

A wide QRS complex tachycardia with group beating in a paediatric patient with heart failure

M. Cecilia Gonzalez*, Jean-Benoit le Polain de Waroux#, Francoise Mascart* and Christophe Scavee#
*Paediatric Cardiology, Cliniques Universitaires St Luc, Brussels, Belgium
#Cardiology Department, Cliniques Universitaires St Luc, Brussels, Belgium

Background: Idiopathic sustained ventricular arrhythmias are a rare cause of cardiomyopathy in the pediatric age. We present a peculiar case of primary sustained ventricular tachycardia with an unusual group beating pattern and heart failure at diagnosis.
Case presentation: A previously healthy 11-year-old girl was referred following a pre-syncopal episode while playing sports. On arrival, 12 lead ECG showed a sustained wide complex tachycardia with a left inferior axis and VA concordance. A prolonged strip showed an incessant arrhythmia, regularly interrupted by narrow QRS complexes. A transthoracic echocardiography demonstrated a left ventricular ejection fraction of 35%. A dose of adenosine confirmed loss of retrograde concordance for one beat without interruption of the arrhythmia. The patient was urgently taken to the EP lab. The intra-cardiac recordings revealed a repetitive pattern of 3 wide beats followed by a narrow complex. Interestingly, this pattern was accompanied by progressive prolongation of the retrograde VA activation. We discovered that the ventricular activity was conducted retrogradely in an alternative pattern by a fast pathway and a decremental slow pathway. The third retrograde P-wave was reconducted anterogradely by the fast pathway and depolarised the ventricular septum, creating a fusion beat. This last ventricular event blocked retrogradely and gave place for the phenomenon to starts again. The ventricular tachycardia was successfully ablated at the lateral free wall of the right ventricular outflow tract. After ablation, the EP study confirmed a dual AV node physiology. Incremental ventricular pacing at the right ventricular apex reproduced the observed phenomenon of a retrograde Wenckebach in the slow pathway without induction of supraventricular tachycardia despite aggressive protocol. No accessory pathway was found. One month after the procedure, the left ventricular ejection fraction normalised.

Abstract no: 1165

Congenitally corrected transposition of the great arteries: Outcomes of 46 survivors subjected to different surgical approaches

Marisa Di Santo, Erica Stelmaszewski, Alejandra Villa, Alejandra Mori, Analia Martin, Mariela Mouratian, Victoria Lafuente, Jose Suarez and Horacio Capelli
Garrahan Children's Hospital, Buenos Aires, Argentina

Background: Outcome of congenitally corrected transposition of the great arteries (CCTGA) depends on the associated intracardiac defects. Different surgical repairs have been proposed. Late outcomes are still poorly understood.
Objectives: To compare the outcomes of different surgical techniques in 46 patients (pts) with CCTGA.
Methods: From 1998 - 2012, 46 patients (age 47.5 months) underwent surgery. Group I (9) Single-ventricle repair (Glenn + extracardiac conduit); Group II (11) Anatomical correction (4 double switch + VSD closure, 4 Senning + Rastelli and 3 Mustard + Rastelli) and Group III (26) Conventional surgery (5 VSD closure, 3 Tricuspid valve replacement and 18 conventional Rastelli). Mean follow-up (months) was: Group I, 53.3; Group II, 36; Group III, 105. Tricuspid regurgitation (TR), aortic regurgitation (AR), systemic ventricle dysfunction, and AV block were assessed.
Results: Group I: (19.5%) AV-valve regurgitation was mild and non-progressive in 18%. All patients had normal ventricular function and NYHA class I and there was 2nd degree AV block in 1 (11.1%). Survival was 83% at 120 months. **Group II:** (24%) TR was moderate-severe in 4 (36%). After surgery it improved in 3. Six developed transient ventricular dysfunction (54.5%). Four had mild-moderate AR. Neither AV block nor residual lesions were found in the atrial switch repair. Survival was 89% (at 120 months). Four patients needed reoperation. **Group III:** (56.5%) The main complications were progressive TR in 16 (p=0.001), associated with mild ventricular dysfunction in 3 and AV block in 10 (p=0.002). Seven patients required a pacemaker implantation post-operatively and 3 during follow-up. Six required reoperation (26%). Survival was 87% at 120 months.
Conclusions: Conventional repair were satisfactory, despite TR progression during follow-up. AV block and pacemaker implantation were relevant in this group. Anatomical correction showed decreasing TR, without AV block complication, but increased risk for AR at mid-term follow-up. Survival rates were similar among the 3 groups.

Abstract no: 1173

Initial experience with a novel echo-based magnetic tracking system for reconstruction of right ventricular volumes and function in adults with congenital heart disease

Helen Dormand, Heiko Schneider, Paul Dunne, Vaikom Mahadevan, Bernard Clarke, Andreas Hoschtitzky and Jaspal Dua
Manchester Heart Centre, Manchester Royal Infirmary, Oxford Road, Manchester, United Kingdom

Background: Long-term monitoring of RV function in ACHD patients is important and continuous. It is critical for timing the intervention. CMR is the gold standard for this but remains time-consuming, costly resource and impossible in some patients. Technology has developed which combines magnet-based tracking with bedside echocardiography, which can be performed in much less time and cost than CMR. We hypothesised that this could be used in majority of ACHD patients with average echo windows.

Materials: We utilised industry-developed tracking-system and patented software installed on a standard computer, linked-up with a standard 2D-Echocardiography machine. Key anatomical points acquired in expiration were then processed through the software and sent securely via the internet to the industry server, where the system generated a 3-Dimensional model within seconds, providing volumetric measurements and ejection-fraction of the RV, to the user.
Methods: Twenty one (9m) patients, mean-age 27.9 years (range17 - 64 years), diagnosis: repaired Tetralogy of Fallot-16; Pulmonary stenosis-5, underwent standard echocardiography. Key anatomical points were placed and image reconstruction undertaken. In 8 patients, CMR-data was available for comparison.
Results: Fifteen scans were suitable for reconstruction. Limited acoustic windows made reconstruction difficult, but poor delineation of valve planes and RV apex was even more important. In 6/8 patients the echo and CMR-derived RVEF were within 6%. In 2 patients with limited echo images, the RVEF differed by >10%.
Conclusions: In the majority of ACHD patients with ToF, this novel and rapid technique could be performed in outpatients to assess the RV. Reconstruction can be performed in less than 10 minutes; accuracy depends on visualising right heart valves and the apex. The use can be extended to other CHD conditions like systemic RV. This will greatly reduce costs and waiting times. Further experience and data is required.

Abstract no: 1175

Nurse-led echocardiographic screening for rheumatic heart disease in Fiji: Results from a pilot study

Daniel Engelman*,#, Joseph Kado†, Bo Remenyi‡, Samantha Colquhoun*,‡, Jonathan Carapetis‡, William May†, Nigel Wilson§ and Andrew Steer*,#

*Centre for International Child Health, University of Melbourne, Australia
#Royal Children's Hospital, Melbourne, Australia
†Colonial War Memorial Hospital, Suva, Fiji
‡Menzies School of Health Research, Darwin, Australia
§Starship Children's Hospital, Auckland, New Zealand

Background: Rheumatic heart disease causes significant morbidity and mortality in Fiji, where the prevalence of RHD in children is approximately 1%. Antibiotic prophylaxis is most effective for mild cases, many of whom are asymptomatic. Screening with echocardiography is the most sensitive screening approach, but is limited by lack of resources, particularly experienced cardiologists and technicians. Training nurses in basic echocardiography for screening is a potential solution.
Hypothesis: We hypothesised that with simple training and supervised field experience in echocardiographic screening for RHD, nurses will have high sensitivity in the identification of children requiring referral for RHD, when compared with diagnosis by a cardiologist.
Methods: We designed an 8-week training syllabus for primary health workers in basic echocardiography for RHD, using a simplified screening protocol and referral criteria based on significant mitral or aortic regurgitation. Seven Fijian nurses participated, and were assessed in a pilot study at the end of the training programme. Nurses screened 16 children, including selected cases with known RHD. A technician performed an extended echocardiogram on all children. All echocardiograms were reported by a paediatric cardiologist.
Results: Four subjects met echocardiographic criteria for definite or borderline RHD, and all were correctly identified for referral by all nurses (sensitivity 100%). The overall specificity was 45% (range 0 - 67%). Fifty five of 56 (98%) nurse screening echocardiograms were of adequate quality for diagnosis.
Conclusions: The high sensitivity observed in this pilot supports the feasibility of training nurses in echocardiographic screening. The study has been expanded to a larger fieldwork phase involving screening 2 000 Fijian school-children, and is powered to formally assess the sensitivity of nurse-led echocardiographic screening. These results will assist resource-poor countries to evaluate the utility of this novel screening strategy.

Abstract no: 1177

Computer-based surgery planning and the Y-graft: The next innovations of Fontan's procedure

Christopher M. Haggerty*, Kirk R. Kanter#, Maria Restrepo*, Timothy C. Slesnick†, Jarek Rossignac‡, Thomas L. Spray§, Mark A. Fogel◊ and Ajit P. Yoganathan*

*Wallace H. Coulter Department of Biomedical Engineering, Georgia Institute of Technology and Emory University, Atlanta, Georgia, United States of America
#Division of Cardiothoracic Surgery, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, Georgia, United States of America
†Division of Paediatric Cardiology; Department of Paediatrics, Emory University School of Medicine and Children's Healthcare of Atlanta, Atlanta, Georgia, United States of America
‡College of Computing, Georgia Institute of Technology, Atlanta, Georgia, United States of America
§Division of Cardiothoracic Surgery, Children's Hospital of Philadelphia, Philadelphia, United States of America
◊Division of Cardiology; Children's Hospital of Philadelphia, Philadelphia, United States of America

Background: The total cavopulmonary connection (TCPC) for single ventricle lesions creates adverse hemodynamics that are hypothesised to negatively impact long-term outcomes. Patient-specific computational modeling may provide novel means to improve blood flow characteristics in these patients. This study reviews our experience with two novel advances: pre-operative surgical modeling and a bifurcated Fontan baffle (Y-graft).
Materials and methods: Cardiac magnetic resonance (CMR) images are segmented to create patient-specific TCPC models and provide flow information. A specially designed anatomy editing tool can be used to accurately mimic Fontan surgery by simulating baffle placement. Blood flow simulations using computational fluid dynamics characterize hemodynamic metrics (i.e., power loss and hepatic flow distribution) and compare connection performances.
Results: Computer-based surgical planning has been prospectively used for 16 patients. The primary indication has been pulmonary arteriovenous malformations, which are believed to form in the absence of hepatic nutrients in the blood reaching pulmonary arterial segments. Thus, the modeling objective was to optimally distribute hepatic blood flow to the lungs. In 5 of 16 patients, a Y-graft Fontan baffle was the modeling recommendation based on beneficial flow distribution characteristics, and was surgically implemented. Follow-up in select patients using arterial oxygen saturations (Table 1), 4-dimensional phase velocity CMR, and post-operative simulations have shown favorable clinical outcomes and consistency between model predictions and operative results. Based on hypothetical energetic improvements, the Y-graft was used for 11 separate patients (Table 2). It is too early to conclude on energetic characteristics compared to traditional extracardiac connections, however operative outcomes and simulated flow distribution results are both positive.

TABLE 1: Arterial oxygen saturation improvements in 8 surgical planning patients

Pre-operative arterial O_2 Saturation (%)	74±8.5
Post-operative arterial O_2 Saturation (%)	89±8.9
Average improvement	15±5.0
Follow-up duration (months)	9.6±8.6

Mean±St. Dev.

TABLE 2: Haemodynamic results from 13 Y-graft patients

Age (years)	6.2±5.6
BSA (m^2)	0.82±0.40
Cardiac index (L/min/m^2)	3.6±1.0
Pulmonary flow distribution (%LPA)	47±23
Hepatic flow distibution (%LPA)	43±21
Power loss (mW)	1.6 (0.7 - 3.5)
TCPC energy dissipation index	0.035 (0.026 - 0.068)

Mean±St. Dev. Median: Interquartile range. BSA: body surface area.

Conclusions: Computer-based surgery planning is an exciting new paradigm for patients with single ventricle lesions with the potential to deliver patient-specific benefit. Findings have motivated the use of the Y-graft Fontan, which may provide both improved energy efficiency and flow distribution for the TCPC.

Abstract no: 1178

Clinical clues to identify anomalous origin of left coronary artery from the pulmonary artery in patients diagnosed with congenital mitral regurgitation or dilated cardiomyopathy

Orhan Uzun*, Sunitha Sampath#, Giridhar Dhandayuthapani†, Yusuf I. Ayhan*, Alison M. Hayes†, Amos Wong*, Dirk G. Wilson*, Victor D. Ofoe* and John L. Gibbs#

*Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, Wales, United Kingdom
#Department of Paediatric Cardiology, Leeds General Infirmary, Leeds, United Kingdom
†Department of Paediatric Cardiology, Bristol Royal Children's Hospital, Bristol, United Kingdom

Background: Children presenting with isolated mitral regurgitation (MR) or dilated cardiomyopathy (DCM) may have anomalous origin of left coronary artery from the pulmonary artery (ALCAPA).
Methods: Twenty-year review of ALCAPA from 3 centres was undertaken.
Results: There were 41 patients (27 females and 14 males). Age ranged from 1 month - 17 years (median 5 months). It took from 1 week to 8 years to diagnose ALCAPA in 11 patients. The commonest reasons for referral were cardiac failure in 23 (56%), respiratory distress or asthma in 21 (51%), failure to thrive in 15 (37%), and incidental murmur in 11 (27%) patients. The initial echocardiographic diagnosis was DCM in 29 (70%), congenital MR in 10 (24%) and PDA in 2 (5%). MR was considered as the primary pathology in 8 (18%) hence its association with ALCAPA was overlooked for as long as 5 years. Eighteen patients (43%) exhibited dilated right CA, and in 7 (17%) the origin of left CA could not be demonstrated. In 15 (37%) patients abnormal retrograde flow in the pulmonary artery was noted. The commonest ECG findings were nonspecific T-wave or ST segment changes in 24 (59%) and 18 (43%) patients respectively. Twenty two (54%) patients had pathological Q-waves in lead aVL. Nineteen (46%) patients required cardiac catheterisation to confirm ALCAPA. In 2 patients ALCAPA was suspected after PDA ligation or during mitral valve replacement. 39 (95%) patients had surgery immediately after diagnosis, but 2 were not operated. Post-surgical echocardiograms showed resolution of MR and left ventricular dysfunction in 29 (70%). 3 (16%) patients died after surgery.
Conclusion: Isolated MR and left ventricular dilatation are important and frequent findings in ALCAPA syndrome. Any child with echocardiographic diagnosis of congenital MR or DCM should prompt a careful search for ALCAPA.

Abstract no: 1179

Fetal and post-natal outcomes of 22q11 deletion and associated congenital heart defects

Orhan Uzun*, Kadir Babaoglu*, Yusuf I. Ayhan*, David Tucker#, Amos Wong*, Dirk G. Wilson* and Victor D. Ofoe*

*Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, Wales, United Kingdom
#Congenital Anomaly Register and Information Service for Wales (CARIS), Wales, United Kingdom

Background: 22q11.2 deletion is the most common chromosomal microdeletion syndrome. It has a strong association with conotruncal heart defects and may exert negative influence on the clinical outcomes.
Methods: Review of patients with 22q11.2 deletion who either presented to the University Hospital of Wales with congenital heart disease or reported to the Congenital Anomaly Register and Information Service for Wales (CARIS) between 1990 and 2011.
Results: 102 cases of 22q11.2 deletion were reported during the study period, giving a gross 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 congenital heart defect (CHD) (93%). Seven cases had a normal heart. In 18 cases diagnosis was established in fetal life: pregnancy was terminated in 4 cases, still birth was found in 1 and in 13 cases pregnancy resulted in a live birth (72%). Eight four patients were diagnosed post-natally. Mean follow-up duration was 16.2+11.4 years (range 1 - 52 years, median 14.5 years) 8 patients died after birth, 7 in the first year of life, and 1 at age 3.8 years.

TABLE 1: Frequency of congenital heart disease associated with 22q.11.2 deletion

Heart defect	(n)	(%)
IAA	18	17.6
VSD	17	16.6
TOF	15	14.7
TA	14	13.7
VSD-PA	11	10.7
DORV	5	4.9
TGA-DILV	1	0.9
AVSD	1	0.9
ASD	3	2.9
PS	3	2.9
ALSCA	2	1.8
VSD	1	0.9
PDA	1	0.9
RAA	1	0.9
BCAV	1	0.9
ARSCA	1	0.9
Total	**95**	**93**

Conclusions: 93% of patients with 22q11.2 deletion exhibited CHD. In spite of the ante-natal detection rate being low, survival was similar for all cases irrespective of the presence of CHD. Most deaths occurred within the 1st year of life but beyond infancy survival was favourable.

Abstract no: 1191

A new method for manufacturing flexible replicas of congenital heart disease using stereolithography and vacuum casting technique: Rehearsal of surgical operation

Isao Shiraishi*, Ken-ichi Kurosaki*, Suzu Kanzaki#, Masatoshi Takeda† and Hajime Ichikawa‡

*Department of Paediatric Cardiology, National Cerebral and Cardiovascular Centre, Japan
#Department of Radiology, National Cerebral and Cardiovascular Centre, Japan
†CrossEffect, Inc, Kyoto, Japan
‡Department of Paediatric Cardiac Surgery, National Cerebral and Cardiovascular Centre, Japan

Background/hypothesis: Stereolithography is a rapid prototype technology whereby an ultraviolet laser beam selectively polymerizes and solidifies photosensitive and polymeric liquid plastic. By using this technique, 3D volumetric image data of multi-slice (MS)CT can be converted into plastic models that enhance our spatial perception of real-life anatomy and pathology. However, the materials of the stereolithography are limited to several photosensitive plastic or urethane, which are not satisfactory for simulation surgery. Recently, a vacuum casting method has been developed, where more delicate models with different stiff materials can be manufactured.
Materials and methods: Three-dimensional volumetric data sets of MSCT angiography of congenital heart disease (CHD) were converted into standard triangulated language files to make stereolithographic biomodels representing the both outer and inner surface of the heart. Then, urethane with appropriate stiffness was injected by using the vacuum casing method. After solidification of the urethane, the casts were carefully removed.
Results: We have made replicas of CHD including VSD, ASD, TOF, DORV, SV, crisscross heart, and ccTGA. The vacuum casting in association with stereolithography enabled us to manufacture replicas with similar texture and structure of the real heart. This technique also allowed the surgeon to cut and suture, facilitating the simulation of the surgical operation.
Conclusion: The vacuum casting in association with stereolithography is a promising technique for the preoperative practice and simulation of individual surgery, and planning of novel and innovative surgical procedures of CHD. This technique could be helpful for rescuing children with CHD.

Abstract no: 1192

Damus-Kaye-Stansel anastomosis mitigates the risk of systemic outflow tract obstruction in children with single ventricle cardiac anomalies

Bahaaldin Alsoufi, Mubashir Khan, Mamdouh Al-Ahmadi, Avedis Kalloghlian, Abdullah Al-Wadai and Zohair Al-Halees

Heart Centre, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

Background: Children with various single ventricle (SV) pathologies are at risk of developing systemic ventricular outflow tract obstruction (SVOTO) following volume unloading with cavo-pulmonary connection (CPC). We aim to evaluate the efficacy of Damus-Kaye-Stansel anastomosis (DKS) at time of CPC in decreasing late SVOTO risk.
Methods: Retrospective review of SV patients who underwent DKS concurrent with CPC between 1997 and 2012 was performed. Clinical, echocardiographic and angiographic outcomes were analysed.
Results: Thirty six children with SV underwent DKS at time of Glenn bidirectional CPC (n=29) or Fontan total CPC (n=7). Underlying anatomy was double inlet left ventricle (n=19), double outlet right ventricle (n=7), unbalanced atrioventricular septal defect (n=4) and other (n=6). Prior palliation included pulmonary artery band (n=35), atrial septostomy (n=9), coarctation/arch repair (n=10). Median age at time of DKS was 8.7 months (range 3.5 months - 8.9 years) and median weight was 6.3kg (range 5 - 27kg). Overall survival was 87% at 5 years. Deaths were unrelated to DKS or SVOT complications. Pressure difference between the systemic ventricle and the aorta decreased from 14±15mmHg pre-operatively to 0 after DKS (p=0.005). At last follow-up, none of the patients developed recurrent SVOT gradient; 73% of them had zero or trivial aortic/neoaortic valve regurgitation while 20% and 7% had mild or moderate regurgitation, respectively. None of the patients had evidence of compression of the left pulmonary artery or bronchus. None of the patients had heart block requiring permanent pacemaker insertion. 86% of patients have reached or are suitable candidates awaiting final palliation stage.
Conclusions: DKS can be safely performed in conjunction with CPC without added mortality hazard. It's very effective mitigating SVOTO risk with durable, good semilunar valves function. Our data support an aggressive approach of performing concurrent DKS with CPC in children with SV pathologies at risk of developing SVOTO.

Abstract no: 1197

Development of the Vanguard demonstration site for RHD among school-aged learners in the South Africa

S. Nkepu, M.E. Engel, C. Lemmer and L Zuhlke

Department of Medicine, Health Sciences Faculty, University of Cape Town, Observatory, South Africa

Background: In South Africa anecdotal information suggests that rheumatic heart disease (RHD) is still the leading acquired heart disease in young adults. Recent data on the prevalence of RHD in school-aged learners remain scanty, with 2 earlier studies suggesting an estimate around 7:1 000. However, this figure is thought to be an underestimation given that the screening was performed using auscultation, which is less sensitive than echocardiography.
Objective: We have developed as part of ASAP, an echocardiography-based RHD screening programme for learners within the Vanguard community of the Western Cape. Outcomes include determining the prevalence of RHD, monitoring the disease progression in screen-positive participants referred for appropriate follow-up, and evaluating the cost-effectiveness of such a programme.
Method: We present the issues involved in the establishment of the demonstration site, the lessons learned, and the ongoing challenges facing the research team, having screened in excess of 2 500 participants. In brief, we detailed our experience in five areas:

- Engaging the community on all levels;
- Completing a situational analysis and gathering background data, including sampling frame and the random sampling procedure;
- Consent process;
- On-site data collection and management; and
- Post-screening responsibility and continued community involvement.

Conclusion: We concluded that screening for RHD is feasible in the community setting using schools as the sample frame. We firmly believe that our experience will serve as a meaningful resource for other similar research programmes.

Abstract no: 1200

Effects of adrenergic receptor blocker treatment on right heart failure

Stine Andersen*, Asger Andersen*, Jacob Gammelgaard Schultz*, Jan Mäller Nielsen*, Sarah Holmboe*, Steffen Ringgaard# and Jens Erik Nielsen-Kudsk*

*Department of Cardiology, Institute of Clinical Medicine, Aarhus University Hospital, Denmark
#MR centre, Aarhus University Hospital, Denmark

Background: Late diagnosis and surgical management of left-to-right shunt congenital heart disease (L-R shunt CHD) can cause pulmonary hypertension. To date there is no effective drug to decrease pulmonary artery systolic pressure. Beraprost an oral prostacyclin analogue acting as a pulmonary artery vasodilator was expected to decrease PASPS in children with L-R shunt CHD.
Materials and methods: A pre-test/post-test study was conducted with 17 subjects aged 2 months - 16 years old, with VSD, ASD, PDA or a combination thereof, who develop pulmonary hypertension. Patients with other structural cardiac defects or with pulmonary infection were excluded. Echocardiography examinations were performed before and after administration of beraprost 1 µg/kgBB/day for 3 months. Vmax TR, Vmax L-R, PASP were measured and M-mode of pulmonary valve including A-wave, EF slope and mid systolic notch were examined. Adverse reactions were noted, monitoring was performed every 2 weeks.
Results: Vmax TR decreased 0.7±0.79m/second (p=0.004), Vmax L-R increased 0.6±0.64m/second (p=0.018) and PASP decreased 18.3±21.90mmHg (p=0.003) were observed. There was no significant changes of wave, EF slope and midsystolic notch.
Conclusions: There was decrease of pulmonary artery systolic pressure after administration of beraprost in children with L-R shunt CHD who develop pulmonary hypertension.

Abstract no: 1202

Bicycle stress echocardiography in children: Feasibility, safety and determination of interobserver variability

Paolo Ciliberti*, McLeod Ian*, Cairello Francesca*, Giardini Alessandro*,# and Jan Marek*,#

*Department of Cardiology, Great Ormond Street Hospital, London, United Kingdom
#Institute of Cardiovascular Science, University College London, United Kingdom

Background: In adult population exercise stress echocardiography (ESE) is a well validated technique providing a dynamic evaluation of myocardial structure and function. In children its use is rather scarce due mainly to precipitous rapid drop in peak heart rate after exercise. The aim of this study hence was to assess the feasibility, the safety and the reproducibility of ESE using on-line scanning in semi-supine cycloergometer protocol in a wide spectrum of children.
Materials and methods: Between July 2008 and April 2012, 42 patients (mean age 14.1±2.9) underwent a bicycle ESE. Two independent observers without knowledge of any patient data interpreted all stress studies, grading quality of each acquired image and presence of regional wall motion abnormalities (RWMA).
Results: ESE was successfully performed and well tolerated by all patients. Rest HR was 82±13.6, peak HR was 153.4±19.7 (161.5±18.3 excluding heart transplant patients and patients on beta-blockers). Among 464 views acquired, the visualisation was optimal in 403 (87%), suboptimal in 32 (7%) and inadequate in 29 (6%). Among 10 patients with hypertrophic cardiomyopathy we were able to assess a significant left ventricular outflow tract gradient increase (>25mmHg) during exercise in 3 patients (33%). ESE was performed in 29 patients with congenital or acquired coronary abnormality (Kawasaki disease, heart transplant recipients, congenital coronary abnormalities, transposition of the great arteries after arterial switch operation). In this group, the RWMA were revealed in 8 patients (28%). The agreement between observers showed a K index of 0.7276 (95% CI = 0.6497 - 0.8055) for the image quality and a K index of 0.5125 (95%CI = 0.4782 - 0.5468) for the RWMA analysis.
Conclusions: Bicycle stress echocardiography performed by on-line scanning during exercise is feasible, safe and reproducible modality in children. Further data to assess its diagnostic accuracy are however needed.

Abstract no: 1204

Associated pulmonary atresia is a significant risk factor for mortality in symptomatic neonates with Ebstein's anomaly

Christopher Knott-Craig, Steven Goldberg, TK Susheel Kumar, Jean Ballweg and Thomas Chin

Le Bonheur Children's Hospital, University of Tennessee, Memphis, Tennessee, United States of America

Background: Ebstein's Anomaly (EA) presenting in the severely symptomatic neonate remains a complex entity with high mortality. There is no accepted standard approach to surgical repair, and opinions vary between complete biventricular repair and single ventricle (Fontan) pathways. More than half of neonates with EA have associated anatomical pulmonary atresia (PA). The outcome in this subset is not well defined. We report on our experience with the early and intermediate outcome of EA/PA.
Methods: From 1994 - 2011, 35 consecutive patients (26 neonates, 9 young infants) with EA were operated on by a single surgeon (CKC). Of the 26 neonates, 16 had associated anatomical pulmonary atresia (61%, 16/26) and 7 had functional PA. For the entire cohort (n=35), follow-up is complete in 93% of patients, over a period of 18 years (mean 6.4 ± 4.6).
Results: Thirty two of the 35 patients had a complete 2-ventricle repair, and 3 patients had a single ventricle palliation (BT shunt, n=2, Starnes palliation, n=1). Hospital mortality for the group was 20% (7/35). For patients with associated PA, mortality was 37.5% (6/16) compared to 5.2% (1/19) for patients without PA ($p<0.05$). The hospital mortality for patients with EA/PA undergoing biventricular repair was 46% (6/13), and the late mortality was 22% (2/9). In our more recent experience (2005 - 2011) we altered our management of patients with EA/PA to include either a RV-PA valved conduit or a single ventricle palliation (n=7) with no early mortality. Kaplan-Meier 15-year survival estimates were 79%±15% for neonates without pulmonary atresia, and 40%±15% for those with pulmonary atresia ($p=0.03$).
Conclusions: The surgical management of EA with pulmonary atresia in neonates continues to evolve. The early survival of neonates with EA/PA is improved when a RV-PA conduit is included in the repair or the patient is stratified to a single ventricle palliation.

Abstract no: 1206

Rare complications of congenital bicuspid aortic valve disease are possibly due to coronary arteriopathy

Ian Ternouth*, Sanjeevan Pasupati#, Rajesh Nair# and Adam El Gamel#

*Taranaki District Health Board, New Zealand
#Waikato District Health Board, New Zealand

Case report: We present a case of a patient with unusual complications post-Bentall repair and aortic valve replacement, possibly due to arteriopathy involving the proximal coronary trees. A 46-year-old seemingly well woman was referred for assessment of palpitations. She was found to have a bicuspid aortic valve with mild aortic stenosis, mild-to-moderate aortic regurgitation, but significantly dilated aortic root, left ventricular dilatation, all of which increased in size under surveillance. CTCA showed normal coronary arteries. She was referred for Cardiac Surgery, which was complicated by emergency Vein Graft to LAD as the patient did not come off pump easily. She was initially well, then presented with acute chest pain 4 months later. A small 4mm defect in the anterior wall of the prosthesis with a large pseudo-aneurysm was found on Echo confirmed on angiography and CTA, and subsequently closed with an Amplatz device. Angiography then showed mild RCA (FFR0.91) and L main stenosis. The vein graft was occluded. Patient subsequently presented 2 months later with crescendo angina, and was found to have critical 95% + left main coronary artery stenosis, stented successfully. The RCA stenosis was confirmed but did not merit PCI.
Results: This patient seems to have developed rapid severe intimal hyperplasia possibly triggered by instrumentation on the substrate of Bicuspid aortopathy/ arteriopathy. Bicuspid aortic valves are known to be associated with cystic medial necrosis and premature coronary artery disease, abnormal vascular reactivity, coarctation, cranial aneurysms; but this patient developed severe coronary disease within a few months of having normal coronary anatomy, suggesting possible rapid intimal hyperplasia. We have not found any similar reports in the literature. Echo and 3-D CT images will be displayed.

Abstract no: 1208

Pulmonary veno-occlusive disease in children

***Cornelia Woerner*[*], *Ernest Cutz*[†], *Hartmut Grasemann*[‡], *Shi-Joon Yoo*[#] *and Tilman Humpl*[*]**

[*]Labatt Heart Centre, Hospital for Sick Children, University of Toronto, Canada
[#]Division of Cardiac Imaging, Hospital for Sick Children, Toronto, Canada
[†]Department of Pathology, Hospital for Sick Children, Toronto, Canada
[‡]Department of Respirology, Hospital for Sick Children, Toronto, Canada

Background: Pulmonary veno-occlusive disease (PVOD) is a rare lung disease, and accounts for 5 - 10% of all cases with pulmonary hypertension (PH) of unknown etiology. Incidence, prevalence and etiology of PVOD in children are not well defined. The mortality remains high, which is partly related to limited treatment options.
Methods and results: This retrospective analysis (1985 - 2011) summarises symptoms, associated factors, treatment and outcomes of 9 paediatric patients (5 female, 4 male) with histological confirmation of PVOD. PH was diagnosed at a mean age of 13.5 years (range 8 - 16 years), followed by the definitive diagnosis of PVOD at a mean age of 14.3 years (range 10 - 16 years). Symptoms, such as decreased exercise tolerance (n=6) and/or shortness of breath (n=9) preceded the diagnosis by 2 years on average; the mean survival time after diagnosis was 14 months (range 0 - 47 months). Computed tomography (CT) of the lungs showed typical radiological features. Treatment included home-oxygen (n=5), diuretics (n=9), warfarin (n=4) and pulmonary vasodilators (n=4). Four children were listed and 3 had successful lung transplants. 8 patients died, including 2 after lung transplant. One transplanted patient survived with good quality of life.
Conclusions: PVOD is an important differential diagnosis for patients with PH. CT is a valuable tool to image lung abnormalities; the definitive diagnosis can be only made by lung biopsy, which subjects the patient to an additional risk. Early listing for lung transplantation is essential, as the mean survival time is only 14 months.

Abstract no: 1212

Anomalous origin of a coronary artery from the aorta: Outcomes of treatment strategies

***Francesca Romana Pluchinotta*[*,#], *Massimiliano Carrozzini*[#], *Gabriele Egidy Assenza*[*], *Elisabeth Beran*[*], *Stephen P. Sanders*[*] *and Francis Fynn-Thompson*[*]**

[*]Boston Children's Hospital, Boston, Massachusetts, United States of America
[#]University of Padua, Padua, Italy

Background: Anomalous origin of a coronary artery from the aorta (AOCA) is often an incidental finding but can be associated with myocardial ischaemia and sudden death, especially in athletes. Because AOCA is rare and the natural history unclear, there are no established management guidelines. We sought to investigate outcomes of various treatment strategies.
Material and methods: We reviewed records of patients with the diagnosis of AOCA seen in our hospital from 1995 - 2012. We included all patients with coronary origin from an inappropriate sinus or single coronary ostium and excluded patients with other congenital or acquired coronary abnormalities or a CHD. Diagnosis, presentation, cardiac testing results, treatment, and outcome were recorded.
Results: We identified 114 patients, 72 (63%) male, ages 0 - 49 years (mean 9.7±9.7 years). The LCA arose from the right sinus in 20 (18%), the RCA from the left sinus in 73 (64%), and the LCA from the non-coronary sinus in 3 (3%). There was a single coronary ostium in 18 (16%). 53 (46%) were symptomatic at presentation. Diagnosis was made by echocardiography in all and confirmed by TEE (53%), cardiac MRI (37%), catheterisation (19%), or angio-CT (17%). Surgery was performed in 48 (42%) patients, including unroofing (41), reimplantation (4), CABG (2), and excision of a prominent sinotubular ridge (1). There were no surgical deaths but 13% of patients had complications. The mean CICU stay was 2 days (range 1 - 16) and the mean hospital stay was 5 days (range 3 - 20). There were no deaths or significant morbidities during follow-up (mean 33 months). Ten patients reported continued symptoms, 5 after surgical repair. Fifty two patients were physically active.
Conclusions: Death or other significant clinical events are rare in young patients with AOCA. Surgical correction can be performed with very low risk but symptoms can persist regardless of treatment strategy.

Abstract no: 1216

Diffusion tensor magnetic resonance imaging tractography detects myofiber architecture in developing human fetal hearts

***Prashob Porayette*[*], *William Kostis*[#], *Guangping Dai*[#], *Van Wedeen*[#], *Stephen Sanders*[*] *and David Sosnovik*[#]**

[*]Boston Children's Hospital, Boston, United States of America
[#]Massachusetts General Hospital, Massachusetts, United States of America

Background: The timing and mechanism of development of the helical myoarchitecture of the human heart is largely unknown. Understanding normal development is essential for detection of alterations in myofiber architecture associated with congenital and acquired heart disease. Diffusion tensor MRI (DTI) tractography exploits the preferential diffusion of water along the direction of muscle fibers to define myofiber structure and orientation nondestructively.
Hypothesis: We hypothesised that DTI tractography could delineate myofiber architecture during heart development.
Materials and methods: Normal human fetal hearts ranging from 10 - 19 weeks gestational age (GA), stored in 10% formalin, were studied. DTI data were obtained using a 4.7 Tesla magnet and 24 diffusion-encoding directions. The primary eigenvectors of the diffusion tensor field were integrated into streamlines and color-coded by their helix angle (HA). Hearts were then sectioned for histology parallel to the LV free wall and stained with H&E. Fiber orientation (DTI tractogram) by MRI at multiple levels through the LV wall was compared with the registered histology sections.
Results: Fiber orientation by DTI correlated well with histology. At 10 weeks the myocardium resembled an isotropic tissue without distinct myofiber patterns. However, by 19 weeks of gestation, LV myoarchitecture closely resembled that seen in adult hearts. The helix angle varied smoothly from a left-handed helix (0° to -90°) in the subepicardium (epi) to a right-handed helix (0° to 90°) in the subendocardium (endo). Fibers in the midwall (mid) were circumferential (± 0°). The characteristic crossing helical pattern was present, but less well developed, at 14 weeks.
Conclusions: DTI tractography shows that developing human myocardium is relatively isotropic at the end of embryogenesis (10 weeks), develops microstructural anisotropy by 14 weeks, and essentially resembles adult myocardium by mid-gestation (19 weeks).

Abstract no: 1222

Rheumatic fever, a South African perspective: A pilot study assessing adherence, notification and secondary prophylaxis

Jacques Liebenberg, Phillip Herbst and Anton Doubell

Division of Cardiology, Department of Medicine, Stellenbosch University and Tygerberg Academic Hospital, Bellville, South Africa

Hypothesis: (1) The notification system for acute rheumatic fever (ARF) is underutilised and inadequate to establish a central data base of rheumatic fever patients; (2) Adherence to secondary prophylaxis is poor; and (3) Factors underlying adherence is multifactorial and can be addressed by the health care system.
Methods and material: Thirty four patients with previous ARF (modified Jones criteria) or established rheumatic heart disease (RHD) (echocardiography) were subjected to semi-structured interviews. Adherence was gauged against provincial guidelines as a gold standard. Echocardiograms were assessed for morphological and functional abnormality. All patient names were cross-correlated with provincial notification data to evaluate the effectiveness of the notification system.
Results: The notification system for ARF at Tygerberg Hospital (TBH) is ineffective with utilisation figures during the past 9 years of only 12.67%. Adherence figures were found to be inadequate: 33.8% in the high-risk period (<25 years) and 11.06% in the total recommended period. Patients' knowledge of their diagnosis, information transfer at the time of diagnosis and the presence of a reminder system were some of the strongest predictors of adherence. Xhosa speaking patients living far from health care facilities had a poor adherence. Lack of social support, the absence of indoor toilet facilities and long waiting times for treatment were significant predictors of poor adherence.
Conclusion: Gross under-reporting of ARF occurs in TBH. Adherence to secondary prophylaxis is poor. Factors which are modifiable by the health care system which predicted adherence included patients' knowledge of their condition, information transfer at the time of diagnosis, patient perception of the need for prophylaxis and the presence of a reminder system.

Abstract no: 1223

Prognostic value of submaximal cardiopulmonary exercise parameters for cardiac morbidity in Fontan patients

Wei-Hsuan Chang[*], Chun-An Chen[*,#], Ssu-Yuan Chen[†], Hsin-Hui Chiu[*], Jou-Kou Wang[*,‡], Yu-CHuan Hua[‡], Chung-I Chang[§], Ing-Sh Chiu[§], Yih-Sharng Chen[§] and Mei-Hwan Wu[*,‡]

[*]Department of Paediatrics, National Taiwan University Hospital, Taipei, Taiwan
[#]Graduate Institute of Clinical Medicine, College of Medicine, National Taiwan University, Taiwan
[†]Department of Physical Medicine and Rehabilitation, National Taiwan University, Taiwan
[‡]Cardiac Childrens Foundation, Taiwan
[§]Department of Surgery, National Taiwan University Hospital, Taipei, Taiwan

Background: Submaximal exercise parameters are associated with an increased risk of hospitalisation in patients with heart failure, but the implication in patients with Fontan circulation remains unknown. Therefore, we investigated the prognostic value of submaximal exercise parameters in Fontan patients, in whom maximal exercise effort is often limited.
Methods and results: Fifty two patients who received Fontan completion underwent cardiopulmonary exercise tests at our institute at least 12 months afterwards. We evaluated 2 maximal parameters (peak oxygen consumption (VO_2) and heart rate reserve (HRR)) and 2 submaximal parameters (oxygen uptake efficiency slope (OUES) and minute ventilation (VE) to carbon dioxide elimination (VCO_2) slope). Peak VO_2 and the OUES were expressed as the percentage of predicted values. During a median follow-up of 22.7 months, 11 patients (21%) exhibited cardiac morbidity, defined as cardiac-related hospitalisation. Time-dependent receiver operating characteristic curve analysis demonstrated that submaximal parameters were related to 2-year cardiac morbidity (area under the curve for the OUES .781, p=.018; for the VE/VCO_2 slope .714, p=.04), whereas peak VO_2 and HRR were not. The optimal threshold value for the percentage of the predicted OUES was 45%, and for the VE/VCO_2 slope 37%. Furthermore, the OUES conveyed independent prognostic information beyond resting oxygen saturation and a history of heart failure or protein-losing enteropathy.
Conclusion: Submaximal exercise parameters provide superior prognostic information to maximal exercise data for predicting cardiac morbidity in Fontan patients. Moreover, the association between the OUES and cardiac morbidity is independent of relevant baseline clinical information.

Abstract no: 1230

Long term management of co-morbidities associated obesity in children with Preader-Willi syndrome

Renny Suwarniaty, Anik Puryatni, Haryudi Aji Cahyono and Januar Wijaya

Department of Child Health, University of Brawijaya, Saiful Anwar Hospital, Malang, Indonesia

Background: Preader-Willi Syndrome (PWS) is the most common syndromic form of obesity, caused by deletion in the 15q11 - 13. Management of obesity and its complications is a critical part of care.
Objective: To present long term management of co-morbidities associated obesity in children with PWS.
Case: A 7-year old girl admitted to Saiful Anwar General Hospital due to dyspnoea on effort. She was born to non-consanguineous and healthy biological parents. Her developmental progress was delayed. She had mild mental retardation, speech articulation defect and global developmental delay. She developed feeding difficulty at infancy but started to become preoccupied with food accompanied by compulsion to eat at 3 years old. At 5 years of age she had morbid obesity. Physical examination showed dysmorphic features. Genetic testing resulted in deletion in the 15q11 - 13. We found cardiomyopathy dilatation and type 2 diabetes mellitus as a complication of obesity. Echocardiography showed decreased ejection fraction and mild MI/MR. Long term management includes monitoring and management of comorbid conditions such as medication of cardiomyopathy, a well-balanced low calorie diet, regular exercise, environment modification and anti-diabetic agent. Early intervention and special education, followed by supportive employment, are appropriate to address the developmental disabilities. Physical, occupational, and speech therapy was started since the establishment of diagnosis. Those resulted in a decrease of BMI, improvement of cardiomyopathy, improvement of developmental and behavioural problems including food-seeking behaviour, thus improving the quality of life.
Conclusion: Comprehensive management of Preader-Willi Syndrome resulted improvement of the quality of life.

Abstract no: 1234

The use of computed tomography angiography (CTA) in the evaluation of congenital heart disease

Darshan Reddy
Department of Cardiothoracic Surgery, Inkosi Albert Luthuli Central Hospital, Durban, South Africa

Background: Whilst echocardiography is the commonest non-invasive imaging modality using in the diagnosis of congenital heart disease, CT angiography may be useful to further elucidate anatomical detail that is crucial to accurate surgical planning.
Methods: Between 2009 to present at our institute, CT angiography has been used selectively to evaluate pulmonary artery size, pulmonary venous drainage, aortic arch abnormalities and coronary artery abnormalies in patients with congenital heart disease. Furthermore, select patients who underwent corrective or palliative surgery were evaluated by CTA.
Results: The use of CTA as an adjunct to echocardiography has enabled accurate pre-operative assessment and facilitated surgical decision-making.
Conclusions: The frequent use of CTA has enabled cardiologist, congenital cardiac surgeons and radiologist alike to familiarise themselves with the anatomical details evident on CT angiography, and correlation with the intraoperative findings enhances the interpretation of the study.

Abstract no: 1236

Intervention for re-coarctation in the Single Ventricle Reconstruction trial: Incidence, risk and outcomes

Kevin Hill
Duke University Medical Center, Durham, North Carolina, United States of America

Objectives: To determine the incidence of re-coarctation (re-CoA), risk factors and outcomes in patients with single right ventricle lesions after Norwood.
Methods: The cohort included subjects randomized to right ventricle-pulmonary artery shunt (RVPAS) or modified Blalock Taussig shunt (MBTS) in the Single Ventricle Reconstruction (SVR) Trial. Re-CoA was defined by intervention, either catheter-based or surgical. Univariate analysis and multivariable Cox proportional hazard models were performed adjusting for center.
Results: Of the 549 SVR subjects, 97 (18%) underwent 131 interventions (92 balloon aortoplasty; 39 surgical) for re-CoA. Intervention typically occurred at pre-stage II catheterisation (n=71, 54%) or at stage II surgery (n=38, 29%). Median (range) age and catheterisation gradient at first intervention were 4.9 months (1.1 - 10.5) and 17mmHg (0 - 60). Center intervention rates varied from 0 - 50%. In multivariable analysis, re-CoA was not associated with assigned shunt type, but was associated with actual shunt type received (HR 2.0 for RVPAS vs. MBTS, p=0.02), and Norwood discharge peak echo-Doppler arch gradient (HR 1.07 per 1mmHg, p<0.01). No other demographic, anatomic or surgical variables predicted intervention. Subjects with re-CoA demonstrated comorbidities at pre-stage II evaluation including higher pulmonary arterial pressures (15.4±3.0 vs. 14.5±3.5mmHg; p=0.05), higher PVR (2.6±1.6 vs. 2.0±1.0 WUxm^2; p=0.04) and increased echocardiographic volumes (end-diastolic volume: 126±39 vs. 112±33ml/BSA1.3; p=0.02). There was no difference in 12-month post-randomisation transplant-free survival for those with and without re-CoA (91% vs. 93%; p=0.7).
Conclusions: Intervention for re-CoA was common and varied by center. Intra-operative crossover due to complex arch anatomy may explain the association with receipt of an RVPAS. Although those undergoing intervention demonstrated comorbidities prior to stage II, there was no difference in 1-year transplant/mortality. Further evaluation is warranted to evaluate effects of morbidity of re-CoA.

Abstract no: 1239

Factors associated with serum B-type natriuretic peptide levels in infants with single ventricle

Ryan Butts[*], Daphne Hsu[#], Victor Zak[†], David Hehir[‡], Paul Kantor[§], Jami Levine[◊], Renee Margossian[◊], Derek Williams[Δ], Andrew Atz[*] and Paediatric Heart Network Investigators

[*] Medical University of South Carolina Children's Hospital, Greenville, South Carolina, United States of America
[#]The Children's Hospital at Montefiore, The Bronx, New York, United States of America
[†]New England Research Institutes (NERI), Watertown, Massachusetts, United States of America
[‡]Children's Hospital of Wisconsin, Wisconsin, Milwaukee, United States of America
[§]The Hospital for Sick Children, Toronto, Canada
[◊]Children's Hospital of Boston, Boston, Massachusetts, United States of America
[Δ]Brenner Children's Hospital, Winston-Salem, North Carolina, United States of America

Background: Data regarding the value of B-type natriuretic peptide (BNP) in infants with single ventricle (SV) physiology are lacking. The objective of this analysis was to describe the changes in BNP in infants with SV physiology before and after superior cavopulmonary connection (SCPC) surgery.
Methods: BNP levels were measured by a core laboratory pre-SCPC (5.0±1.6 months) and at age 14 months during a multicentre trial of ACE inhibition therapy in infants with SV. Associations between BNP, clinical, and laboratory variables were assessed using log-transformed BNP. To identify the predictors of BNP, longitudinal analysis was performed for each set of grouped variables (Echo, catheterisation, growth), and multivariable analysis was performed using patient characteristics at both visits and neurodevelopment predictors at the 14-month visit.
Results: BNP was significantly higher (p<0.01) at pre-SCPC (n=173) compared to 14 months (n=134); median (IQR) 80.8pg/ml (35 - 187) vs. 34.5pg/ml (17 - 67). BNP>100pg/ml was present in 72 (42%) of subjects pre-SCPC and 21 (16%) at 14 months. Patients with BNP at both visits (n=117) had a median (IQR) decrease of 32 (1 - 79, p<0.01). In longitudinal analysis, higher levels of BNP were associated with increased echocardiographic end-systolic volume Z-score (p=0.01), greater degree of AV valve regurgitation (p<0.01), lower weight Z-score (p<0.01), and lower length Z-score (p=0.02). In multivariable analyses at 14 months, higher BNP was associated with presence of arrhythmia post-SCPC surgery (p<0.01), prior Norwood procedure (p<0.01), and longer length of hospital stay post-SCPC surgery (p=0.04), as well as lower Bayley Psychomotor Developmental Index (p=0.015).
Conclusion: BNP decreases in infants with SV after SCPC surgery. Elevations in BNP are associated with increased ventricular dilation, increased atrioventricular valve regurgitation, poorer growth and neurodevelopment outcomes. Therefore, BNP may be a useful seromarker in identifying infants with SV at risk for worse outcomes.

Abstract no: 1240

Magnetic resonance assessment of myocardial scarring and ventricular function before and after repair of anomalous left coronary artery from the pulmonary artery

Heiner Latus*, Kerstin Gummel*, Stefan Rupp*, Klaus Valeske#, Hakan Akintuerk#, Christian Jux*, Juergen Bauer*, Dietmar Schranz* and Christian Apitz*

*Paediatric Heart Centre, Children's Hospital Giessen, Germany
#Division of Cardiovascular Surgery, University Clinic Giessen, Germany

Background: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare cardiovascular anomaly. Left ventricular (LV) dilatation and dysfunction (DCM) evolves due to diminished myocardial perfusion caused by coronary steal phenomenon. Myocardial scarring has been shown in ALCAPA patients late after repair however the incidence of myocardial scarring before surgery and its impact on post-operative course is still unknown. Accordingly, the aim of the study was to assess myocardial function and viability using cardiac magnetic resonance imaging (cMRI) before and in the short term follow-up after ALCAPA repair.
Methods: Eight patients (mean age 10.0±5.8 months) with diagnosed ALCAPA underwent cMRI before and after (mean time interval 4.9±2.5 months) coronary re-implantation procedures (6 direct re-implantation, 2 Takeuchi method). cMRI included functional analysis and late gadolinium enhancement (LGE) for detection of myocardial scars.
Results: Severe LV dilatation (mean indexed LVEDV 171±94ml/m^2) and dysfunction (mean LV-EF 22±10%) was present in all patients and improved significantly after surgery in 7 of 8 patients (mean LVEDV 68±42ml/m^2, p=0.02; mean LV-EF 58±19%, p<0.01). Myocardial scarring (1 x apical subendocardial, 1 x midventricular transmural) was present in 2 of the 8 patients (25%) and did not predict post-operative course or functional recovery. Early follow-up CMR showed an unchanged degree of infarcted myocardium in these patients. Although functional recovery was sufficient, transmural scarring not evident before reimplantation was found at follow-up cMRI in 2 patients.
Conclusions: Despite often severely compromised LV function pre-operatively, myocardial scarring was only present in the minority of our patients. According to our experience, myocardial dimension and function recovered quickly after coronary reimplantation, independently of myocardial scarring. Further studies are needed to elucidate the endogenous repair mechanisms that are responsible for the re-remodelling of the LV in patients with ALCAPA.

Abstract no: 1244

Inter-parametric correlation between echocardiographic markers in pre-term infants with patent ductus arteriosus

Arvind Seghal*,# and Samuel Menahem†

*Monash Newborn Unit, Monash Medical Centre, Victoria, Australia
#Department of Paediatrics, Monash University, Melbourne, Australia
†Monash Heart Centre, Melbourne, Australia

Background: Various echocardiographic parameters are studied in the assessment and the evaluation of the patent ductus arteriosus. In isolation, many of these parameters have low sensitivity and specificity for ductal significance when compared to ductal size.
Aim: To correlate various echocardiographic parameters with ductal size (transductal diameter) in infants with symptomatic patent ductus arteriosus and ascertain the sensitivity, specificity, positive and negative predictive values and likelihood ratios of various parameters to predict a duct ≥3mm in size.
Methods: Pre-term infants less than 32 weeks gestation who were evaluated for the presence of a patent ductus arteriosus were assessed. The following echocardiographic parameters were measured: transductal diameter (TDD), ductal velocity, end diastolic left pulmonary artery (LPA) flow, ductal diameter to left pulmonary artery ratio (TDD:LPA), left atrial to aortic root ratio (LA:Ao), left ventricular output to superior vena cava flow ratio (LVO:SVC), transmitral E/A ratio and isovolumic relaxation time (IVRT).
Results: Fifty two infants were enrolled with a mean gestational age 26±2 weeks and mean birth weight of 837±240g. The mean transductal diameter was 2.8±0.5mm. Transductal diameter correlated significantly with ductal velocity, end diastolic LPA flow, TDD:LPA, LA:Ao and LVO:SVC ratio. No significant correlation with transmitral indices was noted. Amongst parameters, LVO:SVC ratio had the highest specificity (0.83) and sensitivity (0.95) to detect a duct of ≥3mm. The area under the curve was 0.95 (95% CI: 0.85-0.99), indicating a 95% probability that a randomly selected patient with LVO:SVC ratio ≥4 will have a ductal size ≥3mm.
Conclusions: Significant correlations between ductal size and surrogate markers of pulmonary over circulation were noted. A combination of echocardiographic markers may help assess the magnitude of the haemodynamic impact of a patent ductus arteriosus.

Abstract no: 1245

Marfan syndrome in an 8-year-old boy

Renny Suwarniaty, Haryudi Aji Cahyono and Nur Ramadhan

Department of Child Health, Medical School, University of Brawijaya, Saiful Anwar, Malang, Indonesia

Background: Marfan syndrome is a systemic disorder of the connective tissue associated with mutations. Early diagnosis and advances in medical technology have improved the quality of life for people with this disease. It can be determined by multisystem disorder with manifestations typically involving the cardiovascular, skeletal, and ocular systems. Rupture of the aorta poses a serious complication of Marfan syndrome.
Objective: An 8-year-old boy weighing 27kg, height 138cm, came to the hospital on 20 March 2009, for endocrinology counselling. The boy looked healthy. He was taller than expected from his genetic background. A craniofacial found a small chin and malar flattening. There was no chest deformity and from extremities was found wrist sign and thumb sign. An ophthalmologist diagnosed simple ectopia lentis. Echocardiography revealed elongation of the aorta. He is the 2nd child, and his father became blind at about 3 years of age. This patient fulfils the Ghent Criteria (3/5 criteria). Beta blocker was prescribed to prevent the progressivity of the elongation of aorta, and followed by serial echocardiography regularly every 6 months.
Conclusions: The diagnosis of Marfan syndrome use clinical manifestation (Ghent Criteria) without genetic testing. Medication with beta blocker and serial echocardiography is necessary to early detection the elongation of the aorta may cause spontaneous rupture of the aorta.

Abstract no: 1250

Clinical course and prognosis of hypertrophic cardiomyopathy in Egyptian children

Sonia Elsaiedi, Zeinab Salah and Reem Ibrahim
Cairo University Children's Hospital, Cairo, Egypt

Background: Hypertrophic cardiomyopathy (HCM) is an important cause of disability and death in patients of all ages. Egyptian children may differ from Western and Asian patients in the pattern of hypertrophy distribution, clinical manifestations and risk factors.
Objectives: The present work aimed to register the clinical characteristics and outcomes of our children with HCM over a period of 7 years, and to determine whether reported adult risk factors for SCD are predictive of outcome in these affected children.
Study design and methods: A retrospective study that reviewed the clinical data of 128 HCM paediatric patients. These data included personal and family history, physical examination, baseline laboratory measurements, ECG, Holter and echocardiographic results. Logistic regression analysis was done for detection of risk factors for death.
Results: 51/128 patients died during the period of the study. Extreme LVH (interventricular septal wall (IVS) thickness or posterior wall (PW) thickness Z score >6, presence of sinus tachycardia and supraventricular tachycardia (SVT) were independent risk factors for prediction of death in patients of HCM.
Conclusions: In our Egyptian tertiary care centre, HCM has a relatively bad prognosis. Infants have a worse outcome than children presenting after the age of one year. A poorer prognosis in HCM is predicted by extreme LVH, presence of sinus tachycardia and SVT.

Abstract no: 1251

Initial experience with cardiac resynchronisation therapy in paediatric patients

Himal Dama[*], E.G.M. Hoosen[*], A. Nzimela[*], J. Ragadu[*] and B. Vezi[#]
[*]Department of Paediatric Cardiology, Inkosi Albert Luthuli Central Hospital, University of KwaZulu-Natal, South Africa
[#]Department of Cardiology, Inkosi Albert Luthuli Central Hospital, University of KwaZulu-Natal, South Africa

Introduction: Cardiac resynchronisation therapy (CRT) is an effective intervention in adults with heart failure who do not improve after optimisation of medical therapy. Experience in paediatrics is limited in terms of patient numbers and long term follow-up. We present our experience with 2 paediatric patients on CRT at our institution.
Case reports and discussion: Patient 1 was diagnosed with an ASD and a large VSD that was surgically corrected at one year of age. The surgery was complicated by complete heart block necessitating the insertion of an epicardial pacemaker. Over the next 3 years he developed dilated cardiomyopathy (DCMO) which did not improve on optimal medical therapy. A CRT pacemaker system was inserted epicardially at age 4. He now displays improved exercise tolerance on the 6 minute walk test with improved LV function. **Patient 2** presented as a neonate with complete heart block. An epicardial pacemaker was inserted. Over the next 5 years she developed DCMO despite optimal medical therapy. A CRT pacemaker system was inserted epicardially at the age 5. She now has improved symptoms and LV function. CRT has emerged as an effective treatment strategy for pacemaker-induced DCMO as chronic RV stimulation can adversely influence LV function over time. The indications, procedures and outcomes for CRT in paediatrics is evolving. CRT has shown to improve symptoms, exercise capacity and quality of life.
Conclusion: CRT was associated with improved outcomes in our 2 patients with pacemaker-induced DCMO.

Abstract no: 1257

Syndromic phenotypes of Ugandan children with congenital heart disease

Andrea Beaton[*], Laura Harris[*], Twalib Aliku[#], Sulaiman Lubega[#], Peter Lwabi[#], Brendan Lanpher[*] and Craig Sable[*]
[*]Children's National Medical Centre, Washington DC, United States of America
[#]Uganda Heart Institute, Kampala, Uganda

Background: This study determined the occurrence and pattern of recognisable genetic syndromes in Ugandan children with congenital heart disease.
Methods: A medical geneticist, a genetic counsellor, and cardiologists from the Uganda Heart Institute (UHI) at Mulago Hospital, and Children's National Medical Centre in Washington, DC, evaluated children with congenital heart disease. Cardiac diagnoses were confirmed by echocardiography. Phenotypic assessment was performed and family history obtained.
Results: 124 children with congenital heart disease were evaluated over a 1-week period in February 2012. Mean age was 5.5±5.5 years. The most common diagnoses were ventricular septal defect (n=40), Tetralogy of Fallot (n=28), pulmonary stenosis (n=12), atrial septal defect (n=11), double outlet right ventricle (n=8), patient ductus arteriosus (n=7), and truncus arteriosus (n=6). By phenotypic evaluation, 26 (20%) of these children had a suspected genetic syndrome: 22q11 deletion (n=10), Noonan (n=5), Down (n=4), 3 Turner (n=3), Kabuki (n=2), Holt-Oram (n=1) and CHARGE (=1). A majority of patients phenotypically positive for 22q11 deletion syndrome (9/10) had a conotruncal abnormality: either tetralogy of Fallot (7), D-transposition of the great arteries (1), or truncus arteriosus (1). A majority with a Noonan's syndrome phenotype (4/5) had pulmonary stenosis. An additional 15 patients (12%) had a pattern of dysmorphic features suggesting an unrecognisable syndrome. 5 children (4%) had a family history of 1 or more 1st - or 2nd-degree relatives with congenital heart disease.
Conclusions: This is the 1st phenotypic survey of CHD in an East African population. Our data suggest a substantial number of children with known congenital heart disease have underlying genetic abnormalities. Truncus arteriosus constitutes a larger than expected proportion of children with congenital heart disease. Coarctation of the aorta is underrepresented. Genotypic studies are needed to evaluate the full spectrum of genetic variation seen in this population.

Abstract no: 1259

Genetic susceptibility to endomyocardial fibrosis

Andrea Beaton*, Joshua Hoffman#, Charles Mondo†, Nezih Cereb‡, Michael Mongomo†, Juergen Freers† and Craig Sable*

*Children's National Medical Centre, Washington DC, United States of America
#Vanderbilt University, Nashville, Tennessee, United States of America
†Uganda Heart Institute, Mulago Hospital, Kampala, Uganda
‡Histogenetics, Westchester, New York, United States of America

Background/hypothesis: Endomyocardial fibrosis (EMF) is the most common form of restrictive cardiomyopathy. Cases have been reported from tropical and subtropical regions around the equator and have repeatedly been shown to cluster both within families and within select ethnic groups. Eosinophilia is an independent risk factor for EMF, suggesting that both genetic and environmental factors play a role in disease development. This study was designed to investigate the genetic susceptibility to EMF, and possible links with eosinophilia. It sought associations between EMF and inherited alleles in the human leukocyte antigen (HLA) system, including antigens HLA-A, -B, -C, -DR, and –DQ. Alleles in the HLA-A1 region have previously been associated with predisposition to allergic and hypersensitivity reactions
Materials & methods: Fifty patients with EMF and 50 controls were recruited from the Uganda Heart Institute and the cardiology ward at Mulago Hospital. EMF was confirmed by echocardiography. Blood was obtained from peripheral veni-puncture and sequence-based typing was used to identify HLA class I (HLA-A, -B, -C) and class II (DRB1 and DQB1) genes. Chi square analysis was used to identify any difference in class I and class II HLA alleles between cases and controls.
Results: HLA-A*02:02 appeared more frequently at site A1 in cases than in controls (29% in cases compared to 4% in controls). In light of this difference, we performed a secondary analysis distinguishing between those with the allele and those without. In this new analysis, there was a significant association (p=0.005) between EMF and presence of the HLA-A*02:02 allele.
Conclusions: HLA-A*02:02 at site A1 was more commonly found in patients with EMF than in controls. Further investigations are needed to more fully understand the role of the HLA system in EMF development and the link between genetics and eosinophilia.

Abstract no: 1260

Tricuspid valve regurgitation

Timothy Colen*, Shelby Kutty#, Richard Thompson*, Chodchanok Vijarnsorn*, Jeffrey F Smallhorn* and Nee Scze Khoo*

*University of Alberta, Edmonton, Canada
#University of Nebraska College of Medicine, Omaha, Nebraska, United States of America

Background: Tricuspid regurgitation (TR) is a risk factor for morbidity and mortality in hypoplastic left heart syndrome (HLHS). Our group previously described association between HLHS medium-term TR and laterally-displaced anterior papillary muscle (APM). This study aims to determine if congenital position of the APM is an early risk factor for TR in HLHS.
Methods: Neonates with HLHS were prospectively assessed prior to stage-1-palliation (S1P) with two-dimensional (2-DE) and three-dimensional echocardiograms (3-DE). 2-DE was used to assess TR, RV fractional area change, sphericity index and end-diastolic area. Neonates with moderate or greater TR prior to S1P were excluded. 3-DE datasets were analysed off-line to extract spatial coordinates of TV annulus, leaflets and PM (TomTec Inc, Germany), with annulus size, leaflet areas, prolapse and tethering volumes calculated using MatLab-based software (MathWorks Inc, Mass.).
Results: Forty neonates were assessed pre-S1P and at mean follow-up of 23±19 months. Patients were divided by degree of TR at follow-up; 15 patients had moderate or greater TR (Group A) whilst 25 had mild or less TR (Group B). Group A had an increased risk of death or cardiac transplant (60% vs. 28%, p<0.05) and rate of TV surgery (33% vs. 0%, p<0.01). Group A and B had similar RV size, shape and function. No difference was found using 3-DE assessment of the APM position. However, group A patients had increased TV tethered volume (0.69 vs. 0.47ml/m^2, p<0.05), increased 3-D tenting height p<0.05), with a trend for greater leaflet area (8.5 vs. 7.2cm^2/m^2; p=0.06). No differences in annular area and TV prolapse were present.

Abstract no: 1267

Establishing left ventricular function in children with aortic insufficiency via exercise stress-test

Igor Lebid*, Oleksandr Romaniuk*, Nadia Rudenko*, Illya Yemets*, Ievgeniia Liebied#, Tetyana Kobets# and Tetyana Kuzmenko#

*Ukrainian Children's Cardiac Centre, Ukraine
#Crimea State Medical University, Simferopol, Ukraine

Method: LV function was evaluated in 56 children (31 boys and 25 girls) with congenital AI, NYHA 1- 2. Median age: 14.9 years (10 - 18). Group 1 (36) had mild AI (P1/2t 485.04±16.85ms) and Group 2 (20) moderate AI (P1/2t 324.74±39.41ms). Echo and stress-echo were performed (Phillips HD11XE) to evaluate the LV in all patients. Stress agent was step-test (power calculation by formula W=p•h•n•1.33, W-power (kg/min); p-weight (kg); h-height of step (m); n-number of raises per min, 1.33-coefficient. 6kg/min/kg on the step for 3 minutes). Thirty normal children were evaluated as control. An echo-Doppler study recorded LV end-diastolic (EDD, EDV) and end-systolic (ESD, ESV) dimensions, volumes; ejection fraction (EF) pressure gradient on aortic valve.
Results: Patients with mild congenital AI had no significant differences in LV parameters after stress-test compared to healthy children (p>0.05): EDD (4.06±0.13cm), ESD (2.23±0.06cm), EDV (79.05±3.95ml), ESV (17.84±1.19ml) that in control group (4.15±0.06cm, 2.16±0.04cm, 77.22±2.55ml, 15.90±0.78ml). EF in Group1 (77.56±1.09%) did not differ from that of healthy children (79.56±0.63%) (p>0.05). In children with moderate AI there were significant higher LV parameters after stress-test: ESD (2.41±0.11cm), EDV (90.03±4.13ml), ESV (21.97±1.96ml) compared with the same indexes in healthy children (p<0.05). EF in this group (73.84±2.18%) was lower than in healthy children group (p<0.05).
Conclusion: Estimation of LV function during stress echo in patients with AI detects early signs of dysadaptation for developing the heart failure. This is very important for children with moderate AI as an objective indication for surgery correction.

Abstract no: 1271

Valve surgery in Brazilian children and adolescents with rheumatic heart disease: Time to operation and associated factors

Regina Elizabeth Mäller*, Maria Virginia Marques Peixoto#, Maria Cristina Caetano Kuschnir†, Dayse Pereira Campos‡ and Ana Claudia Almeida#

*National Institute of Cardiology, Ministry of Health, Rio de Janeiro, Brazil
#Fernandes Figueira Federal Institute, Fiocruz, Rio de Janeiro, Brazil
†State University of Rio de Janeiro (UERJ), Rio de Janeiro, Brazil
‡Evandro Chagas Clinical Research Institute, Fiocruz, Rio de Janeiro, Brazil

Background: Rheumatic heart disease (RHD) remains the most common acquired heart disease in children and adolescents worldwide, being responsible for high morbidity and mortality, often demanding valve surgery in acute or chronic phase of disease.
Hypothesis: Valve surgery is associated with variables/factors from 3 distinct dimensions: socio-demographic; clinic; and echocardiographic in children and adolescents with RHD.
Materials and methods: Longitudinal observational study of hospital based population, using Kaplan-Meier method for time estimation and Cox regression model for hazard risk (HR) evaluation of associated variables. Cohort: 3 - 20 year old patients, registered 1986 - 2006, and followed-up until 2011. RHD confirmed through Doppler echocardiography. Variables evaluated at patient's first visit, separated in 3 dimensions: socio-demographic (gender, age group, skin colour, residence region, decade of diagnosis); clinic (disease status, NYHA functional class, number of previous rheumatic episodes, secondary prophylaxis, infectious endocarditis, atrial fibrillation); echocardiographic (valve lesion and severity, left atrium diameter, systolic left ventricle diameter, left ventricle function, pulmonary hypertension, rupture of mitral chordae). For database: ACCESS 2000. For statistical analysis: R Programme. Significance by $p<0.05$.
Results: 348 patients, 58% female, 39% underwent valve operation. Median age at register: 12.5 years. Median follow-up time: 9.0 years (2 - 21 years). Median time until surgery: 22.3 years. Univariate analysis: all variables except residence region ($p>0.5$) presented significance (HR>1). Multivariate analysis: final model included: decade of diagnosis (HR 1.36), NYHA functional class II-IV (HR 1.97), number of anterior episodes (HR 1.72 - 2.15), endocarditis (HR 2.01), valve lesion and severity (HR 2.15 - 2.89), left atrium diameter ≥40mm (HR 2.67), systolic left ventricle diameter ≥45mm (HR 2.67) and rupture of mitral chordae (HR 2.68).
Conclusions: Valve surgery is associated with socio-demographic, clinic and echocardiographic factors in Brazilian children and adolescents. Although most patients underwent surgery in 1st 2 years, long-term surveillance of this population is mandatory.

Abstract no: 1273

Assessment of systemic endothelial function in children and adolescents with pulmonary arterial hypertension

Christian Apitz, Anna Werz, Ines Kock, Heiner Latus, Jochen Kreuder and Dietmar Schranz

University Children's Hospital, Giessen, Germany

Background: Pulmonary endothelial function is known to be affected in patients with pulmonary arterial hypertension (PAH). We hypothesised that PAH is a progressive panvasculopathy, affecting both pulmonary and systemic vascular beds, and that systemic endothelial dysfunction correlates with pulmonary vascular changes.
Methods: Systemic endothelial function was assessed by the vascular response to reactive hyperaemia and was recorded non-invasively by peripheral arterial tonometry (EndoPAT) under standardised conditions. Digital reactive hyperaemic index (RHI) was examined in 25 children and adolescents (mean age 21.1±10.1 years; 14 females) with pulmonary arterial hypertension (IPAH n=13, PAH-CHD n=12; 8 of them with Eisenmenger syndrome). Measurements were compared with functional data assessed by echocardiography and cardiac catheterisation.
Results: Mean tricuspid annular plane systolic excursion (TAPSE) was 20.9±4.6 mm. Mean pulmonary artery pressure (MPAP) of all patients was 68±27.4mmHg, and ratio of pulmonary vascular resistance to systemic vascular resistance 1.0±0.6. Vasodilator response to acetylcholine (mean pulmonary flow ratio) was 2.1±2.2. Mean RHI was lower in IPAH (1.6±0.6) and Eisenmenger patients (1.5±0.4) compared to PAH-CHD without Eisenmenger patients (2.1±0.7) ($p=0.04$). RHI correlated with TAPSE ($p=0.001$). There was no correlation between RHI and pulmonary endothelial function assessed by vasodilator response to acetylcholine.
Conclusions: Systemic endothelial function may be impaired in children and adolescents with IPAH and Eisenmenger syndrome and correlates with RV systolic function. However, according to our data there was no relation between systemic vascular changes and pulmonary endothelial function therefore different mechanisms may contribute to their pathogenesis and progression.

Abstract no: 1279

Contribution of atrioventricular plane displacement to left and right ventricular stroke volume in healthy subjects and patients with pulmonary regurgitation

Sigurdur Sverrir Stephensen*,#, Katarina Steding-Ehrenborg*, Hakan Arheden*, Einar Heiberg* and Marcus Carlsson*

*Department of Clinical Physiology, Skane University Hospital, Lund University, Denmark
#Department of Paediatric Cardiology, Skane University Hospital, Lund University, Denmark

Background: Atrioventricular plane displacement (AVPD) contributes to 60% of left ventricular (LV) stroke volume (SV) and 80% of right ventricular (RV) SV in healthy subjects. However, there is no data in patients with RV volume overload. The aim was to quantify the physiological effect of RV volume overload on the contribution of AVPD to ventricular pumping
Materials and methods: MRI was performed in 29 healthy subjects and 22 patients with moderate to severe pulmonary regurgitation (PR) due to surgically corrected Tetralogy of Fallot or pulmonary stenosis (mean age 23.5±13.1). LVSV and RVSV were calculated by delineation of the endocardium of both ventricles in diastole and systole. The longitudinal contribution to the SV was calculated for both ventricles using a previously described method using AVPD and the epicardial area of the basal part of the ventricle.

Results: In the patient group regurgitant fraction was 44±10% and RV end-diastolic volumes (EDV) were increased (278±84ml) compared to LVEDV (154±56ml, p<0.0001). The contribution of the AVPD to the LVSV did not differ between healthy subjects (59±2%) and patients with PR (56±13%, p=0.244). However, the contribution of AVPD to the RVSV was significantly lower in patients with PR (46±8%) compared to healthy subjects (78±2%, p<0.0001).
Conclusion: Patients with PR have normal longitudinal contribution to LVSV, but decreased longitudinal contribution to RV pumping compared to healthy subjects. This is caused by decreased AVPD in volume overloaded RVs which only in part is compensated by an increased RV area. Instead, increased septal movement towards the RV and radial contraction of the free wall contribute to the major part of the RVSV in patients with volume overload due to PR.

Abstract no: 1280
Cardiac pacing in patients with the old Fontan-Kreutzer atriopulmonary connection

Ricardo Gomez*, Tomasa Centella*, Maria Jesus Lamas*, David Cabestrero*, Ana Coca#, Elena Alvarez#, Elvira G. Lestache† and Inmaculada Sanchez†

*Congenital Heart Surgery Unit. Ramon y Cajal University Hospital, Madrid, Spain
#Paediatric Intensive Care Unit, Ramon y Cajal University Hospital, Madrid, Spain
†Paediatric Cardiology Unit, Ramon y Cajal University Hospital, Madrid, Spain

Background: Atrial arrhythmias occur frequently and represent an important complication in the long term follow-up of patients with atriopulmonary connection. The loose of A-V synchrony is clearly associated with a worsening of the functional status and the failure of the univentricular circulation.
Material and methods: From 1975 - 2011, 75 patients with aortopulmonary connection are being followed-up at our institution. Forty eight patients (64%) have developed arrhythmias. Eleven patients (age ranging from 5 - 41 years) needed a pacemaker because of Sinus Node Dysfunction (5 patients), A-V Block (3) and Atrial Tachyarrhythmias (3).
Results: A DDDR pacemaker was implanted in 5 patients, VVIR pacemakers were implanted in 4 cases and an AAI pacemaker was implanted in 2 patients. Seven devices were endovenously placed and 4 devices were epicardial. The measured mean parameters at the implantation in the atrium and the ventricle respectively were: threshold 1.18 and 0.925mv. at 0.5msec; p-wave 1.64 and R-wave 11.18; impedance: 615 Ohms and 725 Ohms. For ventricular stimulation, 4 intravenous leads were placed in the coronary sinus, 3 bipolar contact epicardial leads and 2 unipolar contact epicardial leads. Regarding the atrial leads, 5 were endocavitary active fixation leads and 2 bipolar contact epicardial leads. Mean follow-up is 7.7 years (9 months - 23.7 years). There has been one late death at a conversion procedure to TCPC. Functional status has improved in all the remaining cases.
Conclusions: The use of the appropriate devices to control the frequent atrial arrhythmias in these patients improves the functional class, thus delaying the need for a conversion procedure or a heart transplant. It is mandatory to use the adequate techniques to achieve the goal of maintaining the A-V synchrony in these patients.

Abstract no: 1284
Mitral valve repair in children with rheumatic heart disease

Yogesh Sathe, Mayekesavan Prabhu, D. Muthukumarvel and Prashant Shah

Paediatric Cardiac Surgery, India

Introduction: Rheumatic Heart Disease (RHD) is common in rural India. Mitral valve is most commonly seen in RHD. Children with rheumatic mitral valve disease present either mitral regurgitation or stenosis. The option of mitral valve repair option and subsequent results has been studied at our centre.
Material and methods: This is a retrograde single centre study from March 2009 - June 2012. Total number of patients = 90: male 41, female 49; aged from 5 - 18 years, mean age 10.6 years: 79 had mitral regurgitation and 11 mitral stenosis. 76 patients had mitral valve repair, 14 required mitral replacement. Mitral repair: chordal shortening (11); neo-chordal formation (17) and thinning of leaflet (25); suture anuloplasty (32) and ring anuloplasty (35). All patients underwent epicardial or transoesophageal Echo post-operatively. Mean CPB time was 130 minutes. While mean aortic cross-clamp time was 78 minutes. Five patients had Grade II MR on TEE which was accepted as the left atrial pressure was low. No early or late mortality. Mean hospital stay 7.4 days.
Results: On discharge 8 patients had mild MR while 5 patients had Grade II MR. We have completed follow-up for 2 years. Three patients required mitral valve replacement after 2 years, while 3 patients developed new Grade III MR.
Conclusion: Mitral valve repair is a good option for children with rheumatic mitral valve disease. By modifying surgical techniques most mitral valves can be repaired with good surgical results.

Abstract no: 1291
Retrospective evaluation of patients with Kawasaki disease

Pelin Kosger, Birsen Ucar, Ali Yildirim, Gokmen Ozdemir, Tevfik Demir and Zubeyir Kilic

Department of Paediatric Cardiology, Medical Faculty, Eskisehir Osmangazi University, Eskisehir, Turkey

Background: The diagnosis of Kawasaki disease requires the presence of 5 days of fever and at least 4 of the 5 principal clinical features, including bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy. The most serious complication is coronary artery aneurysm.
Methods and results: In this study, 18 patients diagnosed with Kawasaki disease in Eskişehir Osmangazi University Hospital Department of Paediatrics between 1996 and 2012 were evaluated. During admission, the mean age of the patients was 40.6±22.5 months (8 - 90 months) and the duration of fever was 8.8±4.4 days (3 - 20 days). All patients had high fever and second commonly seen finding was changes in lips and oral mucosa (94.4%). The other findings were changes in the extremities (83.3%), rash (72.2%), non-exudative conjunctivitis (55.5%) and cervical lymphadenopathy (27.7%). 12 (66.7%) of the cases were diagnosed with complete and 6 (33.3%) with incomplete Kawasaki disease. Coronary artery dilatation was observed in 6 patients (33.3%), mitral insufficiency in 4 (22.2%), pericardial effusion in 1 (5.6%), increased end-diastolic diameter of the left ventricle in 1 (5.6%) patient. Sixteen patients were given intravenous immunoglobulin (IVIG) and acetylsalicylic acid (ASA). Two patients, 1 complete and 1 incomplete, could be given only ASA. Coronary artery pathology was not shown in these 2 patients. One patient was given a 2nd dose of IVIG because of the persistence of fever. During follow-up, coronary artery aneurysm was observed in 4 (25%) and coronary artery stenosis in 1 (6%) of the patients given IVIG.
Conclusion: Early diagnosis and treatment is important to prevent coronary artery complications. It is essential to suspect Kawasaki disease and to perform echocardiographic evaluation for cardiac involvement in patients with fever persisting longer than 5 days.

Abstract no: 1293

Evaluation of ventricular septal defect with real-time 3-D echocardiography

Jia-Kan Chang*, Yen-Ying Yang#, Ta-Cheng Huang#, Chu-Chuan Lin#, Jun-Yen Pan† and Kai-Sheng Hsieh#

*Department of Paediatrics, Cheng-Hsin General Hospital, Taipei
#Department of Paediatrics, Veterans General Hospital, Kaohsiung, Taiwan
†Division of Cardiovascular Surgery, Veterans General Hospital, Kaohsiung, Taiwan

Background: As a dynamic three-dimensional structure, the heart can be hard to understand in conventional two-dimensional (2D) plane imaging. Hence, real time 3-D (RT-3-D) imaging can be used for assessing cardiac structures and intracardiac lesions. This study assessed a RT-3-D platform for delineating ventricular septal defect (VSD) geometric characteristics.
Materials and methods: Nine patients with VSD (4 female, 5 male) enrolled in this study. The VSD types included 3 perimembranous cases, 3 inlet cases, and 3 outlet cases. The Philips IE 33 system was used to acquire both the 2-D and the RT-3-D echocardiography images. These patient lesions were subsequently diagnosed and analysed for clinical comparison.
Results: The 2-D VSD diameter was 6.78±1.55mm (range: 4.2 - 10.1mm). The RT-3-D mean maximum and minimum VSD diameters were 7.27±1.74mm and 6.37±1.66mm, respectively. All patients underwent VSD surgical repair; the respective intra-operative maximum and minimum VSD diameters were 7.29±1.83mm (range: 5.0 - 11.5mm) and 6.17±1.98mm (range: 3.0 - 10mm). The correlation coefficient between 2-D and RT-3-D mean maximum diameters was r=0.966, between surgical diameters it was r=0.967, while it was r=0.945 between the RT-3-D mean maximum and intra-operative diameters.
Conclusion: "Real-time" 3-D echocardiography can be a good diagnostic tool to clearly delineate the size and position of a VSD. In future, it may pre-operatively assess VSD and device closures.

Abstract no: 1301

Cost-effectiveness of echo-based rheumatic heart disease screening

Justin Zachariah*,#, Rosemary Wyber† and Mihail Samnaliev*

*Boston Children's Hospital, Boston, Massachusetts, United States of America
#Harvard Medical School, Boston, Massachusetts, United States of America
†Harvard School of Public Health, Boston, Massachusetts, United States of America

Background: Rheumatic heart disease (RHD) is a leading cause of mortality and morbidity in young citizens of low- and middle-per capita income settings. Echocardiography-based screening approaches dramatically expand the number of children identified at risk of progressive RHD. We developed a Markov model to project the cost-effectiveness of this nascent screening approach.
Methods: A Markov model was constructed comparing a No-Screen to Echo-Screen approach. Both scenarios commit staff to provide: secondary prophylaxis; prophylaxis transportation; consumables; heart failure medications; anticoagulation and monitoring; general practitioner and/or cardiology follow-up appropriate to the severity of RHD; valve replacement in a fraction of compromised patients including:operative costs, consumables, valve, and post-operative stay; and severity-appropriate lost wages to patient/parent. The screen scenario posits technician-driven limited screening Echo followed by detailed cardiology evaluation in screen positive children. The screen scenario entailed one-time costs for staff, transportation, echo machine, and a single day lost wages. RHD-related states were categorised as well (utility weight 0.9), dead (utility 0), silent RHD defined as visible on echocardiography but silent by auscultation (utility 0.75), auscultation-audible RHD (utility 0.75), previously audible but now resolved RHD (utility 0.75), RHD with functional compromise in activities of daily living (utility 0.58), and RHD post valve replacement (utility 0.58). Sensitivity analyses varied Echo accuracy, surgical availability, disease prevalence, and screening associated costs. Results are denominated in Australian dollars and future utilities and costs were discounted by 3.5% per year.
Results: The Echo-Screen strategy may be cost-effective, and under certain circumstances, dominates the No-Screen strategy. This result appears insensitive to screening costs, surgical availability, echo accuracy, but does appear to be affected by RHD prevalence.
Conclusions: Contrary to our expectation, a 2 -tage Echo screening approach in a health system committed to providing secondary prophylaxis may be robustly superior under a variety of circumstances.

Abstract no: 1302

New Ghent criteria for Marfan syndrome: Clinical implications

Silvia Alvares, Vasco Lavrador, Esmeralda Martins and Marilia Loureiro

Central Hospital of Porto, Porto, Portugal

Background: Marfan syndrome (MFS) is a connective tissue disorder with major features in cardiovascular, ocular and skeletal systems. The diagnostic criteria were revised in 2010 and more weight was given to the aortic root dilatation, ectopia lentis and genetic study.
Materials and methods: Retrospective study of every patient evaluated for MFS in our centre over the last 15 years. We aimed to analyse the practical clinical implications of the revised nosology in a paediatric population.
Results: The study included 18 patients (9 girls), with the 1st evaluation in Paediatric Cardiology at the mean age of 6.7 years (6 days - 16.8 years). There was a positive family history in 4 cases. The diagnosis of MFS by the classical Ghent nosology was established in 12 patients; 8 patients at the first consultation and in the remaining 2.8 years later, confirming that the MFS has an evolving phenotype. With the revised criteria, the diagnosis of MFS remained in 11/12 patients and 2 new patients were diagnosed. The diagnosis of the 5 rejected patients was: 2 as MASS phenotype (in the absence of aortic dilatation), 2 as mitral valve prolapse syndrome and 1 as potential MFS in patients <18 years. The diagnosis was not confirmed because the Z-score of the aortic root was <2 or 3.
Conclusions: The revised Marfan nosology led to a different diagnosis in 3 cases; 1 of MFS patients was reclassified as MASS; conversely, 2 were reclassified as MFS in the presence of aortic dilatation. The diagnosis of MFS was rejected mostly because of the absence of aortic root dilatation defined as Z-score ≥2 or 3. All patients suspected of MFS maintain follow-up in Paediatric Cardiology, considering the possibility that they may develop classic MFS with time.

Abstract no: 1308

Relationship of a left atrial myxoma and the mitral valve is best evaluated by 3D echocardiography

Marie Jose Raboisson*, Ahmed Abdul Salam#, Hatem Hosny† and Nancy Poirier‡

*Ste Justine University Hospital Centre, Montreal, Canada
#Aswan Heart Centre, Aswan, Egypt
†Aswan Heart Centre, Aswan, Egypt
‡Ste Justine University Hospital Centre, Montreal, Canada

Background: 75% of myxomas are located in the left atrium, the majority originating from the inter-atrial septum. They can come in contact with vital structures. We used intra operative 3-D TEE to reassess a myxoma of the left atrium just before surgery.
Methods and results: A 51-year-old woman consulted the Aswan Heart Centre for progressive mild dyspnoea. Her physical findings were compatible with bronchial reactivity without signs of cardiac pathology. Her chest X-ray and ECG were normal. The TTE showed a sessile mass of 13 x 18mm compatible with a myxoma attached to the left side of the inter-atrial septum without other anomalies. She was brought to the operating room for elective resection of the myxoma. A real time 3-D TEE was performed pre-operatively followed by a full volume acquisition with a systematic cropping of the 3-D TEE dataset in multiple plans. Reconstructions showed a tumour measuring 14 x 19mm attached to the lower part of the inter-atrial septum in close contact with the anterior leaflet of the mitral valve as well as the mitral annulus. The space between the mitral annulus and the myxoma was estimated at 5mm. Trans-mitral blood flow was normal. There were echo lucent areas in the tumour compatible with necrosis. After cardiopulmonary bypass and cardiac arrest with blood cardioplegia, a right atriotomy was performed and the myxoma completely resected without traumatising the mitral valve. The septum was then reconstructed with an autologous pericardial patch. The post-operative course was simple and the patient discharged at day 6.
Conclusion: 3-D echocardiography facilitates description of left atrial myxoma better than 2-D. It provides a better understanding of the relationships with adjacent structures especially regarding the space between the tumour and vital structures and should be considered as a 1st line exam in order to help plan surgery and avoid complications.

Abstract no: 1314

Persistent retinal vascular changes in Kawasaki syndrome: Potential role in coronary risk stratification

C. Lim*, I. Wong#, A. Pang‡, X. He*, C. Hia*, C. Cheung†, T. Wong# and S.Quek*

*Department of Paediatrics, National University Health System, Singapore
#Department of Ophthalmology, National University Health System, Singapore
†Singapore Eye Research Institute, Singapore
‡Department of Ophthalmology, Tan Tock Seng Hospital, Singapore

Background/hypothesis: We hypothesise that the inflammatory effects of Kawasaki Syndrome (KS) causes longstanding changes in the retinal vasculature akin to traditional coronary risk factors like diabetes mellitus and hypertension.
Materials and methods: We compared the retinal vascular dimensions of a cohort with a history of KS to matched controls. All subjects underwent high-resolution digital retinal photography in which the diameters of all arterioles and venules coursing through a specified area 1/ to 1 disc diameter from the optic disc were measured with a computer programme (IVAN), according to a published standardised protocol. Central retinal arteriolar equivalent (CRAE), central retinal venular equivalent (CRVE), arteriole-to-venule ratio (AVR) was calculated for each retina photo.
Results: Thirty two subjects with a history of KS and 138 controls were examined. KS subjects had a mean CRAE of 149.77µm, a mean CRVE of 211.61 while controls had mean CRAE of 147.25µm (p:0.300), and a mean CRVE of 221.11 (p:<0.001). Cases were matched for age, gender, ethnicity, body surface area and also controlled for calibre of neighbouring vessels.
Conclusions: KS results in independent, persistent and substantial narrowing of the retinal venules. Such an effect may be related to endothelial dysfunction and serve as a potential marker for incipient coronary vasculopathy.

Abstract no: 1320

Two cases of post-Fontan warfarin-induced tracheobronchial cartilage calcification

Luke Eckersley, Nigel Wilson, Chris Occleshaw and John Stirling

Green Lane Paediatric and Congenital Cardiac Service, Starship Children's Health, Auckland, New Zealand

Background/hypothesis: This study identifies tracheobronchial cartilage calcification in children with congenital heart disease. Calcification of the tracheobronchial airways has been previously found to be more common in adults taking warfarin, and has been described in children who are receiving warfarin following mitral valve replacement.
Materials and methods: A 9-year-old female who had undergone a Fontan repair 6 years previously underwent a Cardiac CT to further image her pulmonary arteries. An incidental finding of extensive tracheobronchial cartilage calcification was noted. We then conducted a retrospective review of all the paediatric Fontan patients who had undergone cardiac CT to look for calcification of the tracheobronchial cartilage.
Results: Ten paediatric Fontan patients had cardiac CT scan. Two patients with extensive calcification of the tracheobronchial airways were identified. A 9-year-old female with a cardiac diagnosis of hypoplastic left heart syndrome had undergone a staged repair with Fontan completion at age 3. A 16-year-old male with a cardiac diagnosis of tricuspid atresia underwent staged repair and Fontan completion at age 3.5 years. They had received continuous warfarin for 6 and 13 years respectively. Other common causes of airway calcification were excluded.
Conclusions: We describe warfarin-induced tracheobronchial calcification in patients following the Fontan procedure. This finding has potential implications for airway growth and vascular calcification.

Abstract no: 1322

HIV+ Malawian children with decreased exercise performance and normal cardiac strain

***Amy Sims*[*,#], Mina Hosseinipour*[*,†], Irving Hoffman*[†], Charles van der Horst*[†], Peer Kazembe*[‡], Madeline McCrary*[†], Robert McCarter*[#] and Craig Sable*[#]**

[*]Fogarty International Clinical Research Fellows Programme, Bethesda, Maryland, United States of America
[#]Children's National Medical Centre, Washington DC, United States of America
[†]University of North Carolina, Chapel Hill, North Carolina, United States of America
[‡]Baylor International Centre of Excellence, Lilongwe, Malawi

Background/hypothesis: Traditional measures of cardiac function are often normal in HIV+ children on antiretroviral therapy (ART). Previously, strain analysis has shown subtle cardiac dysfunction in HIV+ adolescents and young adults. We hypothesised that, using speckle tracking, we would detect subtle cardiac dysfunction in this larger cohort of Malawian children.
Materials and methods: In this prospective observational study of children aged 4 - 18, we recruited 241 HIV+ youth, and 95 HIV- controls. An echocardiogram and 6 minute walk test (6MWT) were performed. CD4 count and HIV viral load were performed on HIV+ subjects. Ejection fraction, strain, and strain rate were measured. Within the HIV+ group, analysis of covariance was implemented to compare means of cardiac function and performance on the 6MWT in HIV+ children on and off ART as well as HIV- children controlling for age, sex, and BMI. Multiple linear regression models evaluated whether cardiac function was related to CD4 count or log viral load. T-tests based on linear contrasts were used to compare function measures between those with detectable and undetectable viral loads.
Results: The HIV- subjects performed better on the 6MWT (p=0.001). HIV+ on ART averaged 470m, HIV+ off ART averaged 460m, and HIV- averaged 500m. Cardiac function and strain were normal for all groups. EF and strain were not related to CD4 count or log viral load. Among HIV+ participants, children with undetectable viral loads had more negative (better) global circumferential strain (GCS) as compared to those with detectable viral loads (p = 0.02).
Conclusions: HIV+ children had decreased exercise performance but did not exhibit decreased strain compared to controls. Detectable viral loads were associated with worsened GCS, suggesting that viral suppression may delay the onset of cardiac dysfunction. Although these children do not currently exhibit clinical cardiac dysfunction, long term evaluation is warranted.

Abstract no: 1324

Hand-held echocardiography: Initial experience in use for assessment of rheumatic heart disease in indigenous children in Western Australia

Jim Ramsay and Joan Sharpe

Princess Margaret Hospital for Children, Perth, WA, Australia

Background: ARF and RHD remain a major problem in indigenous children in Western Australia (WA) particularly in the remote communities in the Kimberley region. It is difficult to triage the care of children presenting with signs of ARF without some echocardiographic assessment. This is difficult in WA as the geography and population distribution means there is very limited access to any paediatric Echo skills in remote areas. With the recent introduction of hand-held Echocardiographic equipment (HHE) it was decided to trial the use of such technology in the Kimberley.
Methods: A HHE was taken to outreach clinics in the Kimberley over a 6-week period and >30 patients with RHD and a number with signs suggestive of ARF had an Echo study performed using HHE. This included 2-D echo images in long-axis, short-axis and 4-chamber views and colour Doppler images from same views. This was followed by a more formal complete study using standard portable Echo equipment (SPE). The images between the studies were compared to assess whether there was good correlation between the degrees of mitral and/or aortic regurgitation.
Results: There was very good correlation between HHE studies and the SPE studies. It was possible using HHE to detect even trivial to mild valvar regurgitation. Chamber sizes and significant valvar morphological changes could be reasonably defined, although this was more limited than the SPE studies.
Conclusions: This initial experience using HHE has been encouraging and warrants further more scientific exploration. HHE could be a useful tool to train non-cardiology staff in remote WA to triage management of patients with possible ARF in consultation with paediatric cardiologists. HHE may be useful for quick screens of patients with known RHD to select patients for more complete Echo studies.

Abstract no: 1325

Epidemiology of acute rheumatic fever in Manitoba, Canada: A 10-year retrospective study

***Maria Elena Lautatzis*[*], Lillie Catherine Cholakis*[#], Ilan Buffo-Sequeira*[†], Reeni Soni*[†] and Dionysios Pepelassis*[†]**

[*]University of Manitoba, Manitoba, Canada
[#]University of Winnipeg, Manitoba, Canada
[†]Department of Paediatrics, Variety Children's Heart Centre University of Manitoba, Canada

Objective: The aims and objectives of this study are to determine the incidence of Acute Rheumatic Fever (ARF) between First Nation (FN) and non-First Nation (n-FN) children in the Province of Manitoba, Canada.
Methods: This is a retrospective study conducted at the Children's Hospital of Winnipeg and Variety Heart Centre, Winnipeg, MB, Canada. Children with discharged diagnosis of ARF were identified through the Children's Hospital of Winnipeg medical records and the Electronic Echocardiography data based at the Variety Heart Centre. The diagnosis of ARF was based on the revised Jones criteria.
Results: Over a 10-year study period (2000 - 2010), a total of 61 patients meeting the Jones criteria for the diagnosis of ARF are included in this study. The overall annual incidence rate of ARF in the province of Manitoba was 1.613/100 000 with 4.748/100 000 for FN and 0.618/100 000 for the n-FN children (p-value <0.001). The overall incidence of ARF among girls was 1.562/100 000 (FN girls 4.984/100 000 n-FN girls 0.475/100 000, p<0.001). Overall incidence of ARF among boys was 1.661/100 000 (FN boys: 4.522/100 000, n-FN: 0.754, p<0.001).

Abstract no: 1327

Acute effect of inhaled iloprost in children with pulmonary arterial hypertension associated with congenital heart disease

Gu Hong, Li Qiangqiang, Zhang Chen, Liu Haiju, Zhuo Ling, Li Aijie, Wang Xiaofeng, Wu Bangjun and Guo Baojing

Beijing Anzhen Hospital affiliated to Capital Medical University, Beijing, China

Objective: To evaluate the safety and efficacy of inhaled iloprost in children with pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) using measurements of haemodynamic parameters.
Methods: The study selected children hospitalised with severe PAH associated with systemic-to-pulmonary shunt type CHD in the Department of Paediatric Cardiology of Beijing Anzhen Hospital from June 2007 - October 2011. All of the children underwent left- and right-sided cardiac catheterisation, during which iloprost was administered by atomisation inhalation. Changes in haemodynamic parameters before and after Iloprost administration were recorded. Adverse events were also recorded.
Results: In total, 89 children with severe PAH associated with systemic-to-pulmonary shunt type CHD (mean age 10.8±4.7 years) were enrolled. Following inhalation of iloprost, the mean pulmonary arterial pressure decreased from 79±11mmHg to 74±12mmHg ($p<0.01$), while the pulmonary to systemic blood flow ratio (Qp/Qs) increased from 1.15±0.45 to 1.71±1.36 ($p<0.01$). Pulmonary vascular resistance index (PVRI) decreased from 19.73±9.31 Wood Units per m^2 prior to inhalation to 15.49±8.87 Wood Units perm^2 following inhalation ($p<0.01$), but there was no significant change in systemic blood pressure (prior to inhalation 85±13mmHg, following inhalation 83±11mmHg, $p>0.05$).
Conclusion: Inhalation of iloprost during cardiac catheterisation in children with severe PAH associated with systemic-to-pulmonary shunt type CHD can lower pulmonary vascular resistance, while causing no significant change in systemic blood pressure and without producing any obvious adverse reactions.

Abstract no: 1328

Outcome of bosentan treatment in paediatric patients with pulmonary arterial hypertension associated with congenital heart disease

Hong Gu, Aijie Li, Chen Zhang, Xiaofeng Wang, Qiangqiang Li, Bangjun Wu, Baojing Guo and Yinglong Liu

Beijing Anzhen Hospital affiliated to Capital Medical University, Beijing, China

Objective: To describe the outcome of paediatric patients with pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) treated by bosentan, which is known to be safe and effective in patients with idiopathic pulmonary hypertension (IPAH).
Methods: In this single centre, open-label, uncontrolled, observational study, 23 patients with PAH associated with CHD were treated by bosentan: The mean age was 9.12±3.6 years (range 2.1 - 14.7 years); they were treated for a mean of 13.3±7.5 months (range 6 - 31 months). The 6-minute walking test, percutaneous blood oxygen saturation (SpO_2%), New York Heart Association functional class (NYHA) and cardiac catheterisation data were compared before and after bosentan treatment.
Results: After treatment, 6MWT (n=17) has improved from 458±16m - 496±69m ($p=0.035$). SpO_2% has increased from 89%±5% - 91%±5% ($p=0.009$). In addition, the NYHA improved. One in NYHA class IV increased to class III, and one in class III increased to class II , other class II kept stable ($p=.001$). Twelve patients have had cardiac catheterisation before and after bosentan treatment, Qp/Qs increased from 0.97±0.40 - 1.16±0.40, PVRI decreased from 20.8±8.8 - 18.1±7.7 Wood unit *m^2 changes had no statistical significance. Bosentan was well tolerated by all patients, except 1 patient who had temporary lower gastro-intestinal bleeding.
Conclusion: Bosentan was safe and effective. Bosentan caused significant improvements in 6MWT, SpO_2 and NYHA function class and improvements in Qp/Qs and PVRI in children with PAH associated with CHD.

Abstract no: 1331

Subvalvar aneurysms in children at a Southern African tertiary care centre

Antoinette Cilliers, Paul Adams, Gcina Dumani and Barend Fourie

Division of Paediatric Cardiology, Chris Hani Baragwanath Hospital, University of the Witwatersrand, Johannesburg, South Africa

Introduction: Subvalvar aneurysms which may be subaortic or submitral were described originally in young Africans in the 1960s, but later reports have included other race groups. Proposed aetiologies include infections particularly tuberculosis, post traumatic, post-surgery, and ischaemia. An underlying congenital or developmental weakness between the muscular ventricular wall and the fibrous valve annulus is speculated. Complications such as rupture of the aneurysm, coronary artery compression, thromboembolism, and sudden death support the need for surgical intervention.
Method: Children with subvalvar aneurysms were sourced from a paediatric cardiology computerised database compiled over a 20-year period at a Southern African tertiary care centre.
Results: Nine children with an average age of 8.8 years (range, 1 - 16) were diagnosed with submitral aneurysms using echocardiography. The male to female ratio was 1:2 and all were black. Five presented in heart failure and with varying degrees of mitral valve incompetence; 1 with an incidental murmur, 1 with syncope, 1 with a pneumonia and 1 with abdominal TB and Human Immunodeficiency Virus HIV) infection. Two patients were found to have additional left ventricular cavity aneurysms and, at surgery, 2 aneurysms were found to have ruptured into the left atrium 4 patients had positive Mantoux tests and 1 was confirmed to have myocarditis and non-compaction of the myocardium on MRI scan. Surgery was undertaken in 6 patients. Histological examination of operative specimens showed peri-cardiac tuberculosis in 2, features suggestive of rheumatic fever in 1, a false aneurysm in 1 and, non-specific changes in the remaining 2. Two deaths were recorded; patient died suddenly pre-operatively and immediately post-operatively.
Conclusion: Subvalvar aneurysms are a rare cause of mitral regurgitation and heart failure in children. Diagnosis is readily confirmed on echocardiography. Tuberculosis is a frequent association, but the aetiology remains inconclusive. There may be an underlying congenital predisposition.

Abstract no: 1340

Congenital central hypoventilation syndrome (Undine syndrome) with recurrent hypercapnia and hypoxemia is likely to act as endothelial preconditioning

Damian Hutter*, Alain Wimmersberger#, Roman Brenner†, Stefano Rimoldi†, Emrush Rexhaj†, Jean-Pierre Pfammatter*, Mladen Pavlovic*, Urs Scherrer† and Yves Allemann†

*Centre of Congenital Heart Disease, University Hospital, Bern, Switzerland
#Santa Maria Hospital, Visp, Switzerland
†University Hospital, Bern, Switzerland

Introduction: Undine syndrome (US) is a rare disease with severely impaired central autonomic control of breathing and dysfunction of the autonomous nervous system. The incidence is estimated to be 1:200 000 live births. We hypothesised that these patients have a higher risk for pulmonary arterial hypertension (PHT) and long term systemic vascular dysfunction due to recurrent hypercapnia and hypoxia. We examined 7 patients with US at baseline and high altitude in regards to pulmonary artery pressure and systemic vascular disease and compared them with 6 age and sex matched subjects.
Methods: Seven patients with US (5 female, 2 male, mean age 19±3 years) and 6 age and sex matched healthy controls (3 female, 3 male, mean age 20±2 years) were examined at 550m and at 3883m above sea level with echocardiography (to measure pulmonary artery pressure). Vascular function was measured at 550m by flow mediated dilatation with and without oxygen.
Results: All US patients had mild to moderate PHT at 550m above sea level. PHT was more pronounced in males than in females (41±5mmHg vs. 27.4±3mmHg) while no PHT was found in the controls. All US patients had high normal systemic vascular function while controls had normal systemic vascular function. In 3 883m above sea level all US subjects showed only mild increase in PHT in regards to baseline while controls developed moderate to severe pulmonary hypertension (38±7mmHg vs. 55±17mmHg).
Conclusion: (1) Despite recurrent hypercapnia, US patients show high normal vascular function. We speculate endothelial preconditioning (recurrent hypercapnia as stimulus). To date recurrent hypercapnia was believed to be most harmful for endothelium function properties. (2) The presence of PHT at 550m above sea level is not significantly aggravated by high altitude in comparison with healthy controls. This underlines the hypothesis of endothelial preconditioning and identifies environmental hypoxia as key trigger for PHT in these patients.

Abstract no: 1341

Rheumatic heart disease in a tertiary hospital in Malaysia

Hung Liang-Choo, Jasminder Kaur Amarjit Singh and Lee Phaik-Ngan

Institute of Paediatrics, Kuala Lumpur Hospital, Malaysia

Background: Acute rheumatic fever (ARF) and chronic rheumatic heart disease (CRHD) remain one of the leading causes of acquired heart disease in children, especially in developing countries. Malaysia, being a developing country should share the same disease burden. Literature search revealed that there were several articles on rheumatic heart disease from Malaysia.[1,2,3,4] Two retrospective reviews in a single centre reported 42 patients over 4 years[1] and 313 patients over 30 years.[2]
Materials and methods: We conducted a retrospective review of the records of all patients with a diagnosis of ARF or CRHD in our hospital from July 2011 - July 2012.
Results: There were a total of 20 patients. The male to female ratio was 1.5:1. The racial breakdown was 18 Malays, 1 Chinese and 1 Indian. A total of 12 patients had ARF, 1 patient had Sydenham chorea with mild mitral regurgitation on echocardiographic examination, and 7 patients had CRHD. Of the 12 patients with ARF, 2 were <5 years old, 7 were 5 - 9 years old and 3 were >10 years old. The patient with Sydenham chorea was a 9.5-year-old Malay boy. Of the 12 patients with ARF, 6 had their first attack of ARF with carditis, 5 had acute-on-chronic rheumatic heart disease, and 1 patient had ARF with no carditis. Of the 11 patients who presented with acute rheumatic carditis, 7 (63.3%) were in NYHA functional class I or II, 1 (9.1%) in class III and 3 (27.3%) in class IV. One patient required intubation and ventilation. All 11 patients had mitral valve involvement, 5 with severe mitral regurgitation, 8 patients also had aortic valve involvement, 2 with severe aortic regurgitation on echocardiographic examination.
Conclusions: Rheumatic heart disease is not uncommon in Malaysia. A quarter of patients with acute rheumatic carditis presented with severe congestive cardiac failure.
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Abstract no: 1342

Poor efficacy of drug therapy for PDA closure in Indian preterm babies

Sanju Yadav*, Dinesh Yadav*, Sheetal Agarwal*, Ajay Dudeja#, Mukesh Beniwal*, N.K. Dubey*, Arti Maria* and Pankaj Gupta*

*Division of Paediatric Cardiology and Division of Neonatology, Kalawati Saran Children Hospital, New Delhi, India
#Department of Paediatrics and Neonatology, Kalawati Saran Children Hospital, New Delhi, India

Background: Ibuprofen is being used as an alternative to indomethacin in medical management of PDA. Due to paucity of data on efficacy of these drugs for PDA closure in India, a study was designed to assess and compare their efficacy for PDA closure in preterm Indian neonates.
Methods: A prospective randomised controlled study on clinically diagnosed and echocardiographically confirmed hemodynamically significant PDA in preterm neonates was designed. Patients were randomly assigned to receive either oral ibuprofen at a dosage of 10, 5, 5mg/kg every 24 hours or 3 doses of oral indomethacin (0.20 - 0.25mg per kg 24 hourly) starting on third day of the life or when diagnosed. A second course of ibuprofen/indomethacin was given, if PDA failed to close within 48 hours after first course. Surgical ligation was considered if PDA did not close after two courses of treatment. Patients were monitored for complications like oliguria, bleeding, NEC, IVH, oxygen dependency and gastrointestinal side effects.

Results: The baseline characteristics were comparable in both groups. Of the 83 children with PDA, 57.8% received oral ibuprofen and 42.1% received oral indomethacin. The overall closure rate of PDA was 60% and 65.7% in the ibuprofen and indomethacin group respectively, however in babies with postnatal age 3 - 5 days; the closure rate was 69.2% and 100% respectively. Complications were similar in both the groups.
Conclusion: The efficacy of both the drugs was similar. However, the overall closure as well as closure in subset of patients with post natal age 3 - 5 days in ibuprofen group was much lower than reported in international literature. Cytochrome P450 enzyme causes increase plasma clearance of the drug, which is negligible at birth and increases with post natal age. Poor closure in our study could be because of genetic difference in pharmacokinetics of drugs in Indian population.

Abstract no: 1343

Longterm follow-up (40 years) of patients post neonatal intervention for congenital critical aortic stenosis at a single-centre

Michelle Carr[*], Xavier Iriat[*], Paolo Ciliberti[*], Ian Sullivan[*], Graham Derrick[*], Martin Kostolny[*], Victor Tsang[*,#], Catherine Bull[*,#] and Jan Marek[*,#]

[*]Department of Cardiology, Great Ormond Street Hospital, London, United Kingdom
[#]Institute of Cardiovascular Science, University College London, United Kingdom

Background: Survival with congenital critical aortic valve stenosis (CAS) can be successfully achieved by surgical or catheter interventions. Longterm outcome and follow up remains uncertain.
Methods and results: A 40 year (1970 - 2010) review of 96 patients, requiring intervention for neonatal (<30 days) CAS. Surgery (SX) was undertaken in 61 patients and balloon dilation (BD) in 35 patients. Before 2000, SX was more common (56/63 cases) while BD was favored after 2000 (28/33 cases). Follow up data was available for 88 patients. There were 29 (32.9%) reported deaths, early death (<90 days) accounted for 22 (25%). Early death accounted for 4 of the 5 deaths since 2000. Twenty (22.7%) patients died without any further intervention, one patient had a heart transplant and another patient had a single ventricle palliation. Overall free survival rate at 5, 10 and 20 years was 73%, 73% and 61%. Of the 57 remaining survivors, 52 (87%) had at least one reintervention, with further reinterventions required in 21(36.8%). There were 33 aortic valve replacements (AVR) in 32 patients, 31(35.2%) patients had a Ross or Ross-Konno procedure. Freedom from AVR at 5, 10 and 20 years was 78%, 63%, and 44%. The median time interval between initial procedure and AVR for the BD group was 1.32 years (mean 3.9±4.7, range 1 day - 14.4 years) this was shorter than for the SX patient group, 8.9years (mean 7.24±5.1, range 1 day - 16.5 years). For patients presenting for initial procedure since 2000, AVR was undertaken in 14/34, with a median time to AVR of 1.3 years.
Conclusions: This study highlights the acute and long term mortality associated with neonatal CAS. Catheter based intervention has become more common but is still associated with early mortality. Reintervention is highly likely in survivors, over 50% of survivors requiring surgical AVR, and a trend towards earlier surgical AVR at our institution in recent years.

Abstract no: 1344

Association of modified Blalock-Taussig shunt and scoliosis in the children with congenital heart disease

Jing-Ming Wu[*], Min-Ling Hsieh[*], Jieh-Neng Wang[*], Hok-Keong Chang[#], Wen-Lan Yen[†], Yu-Jen Yang[*] and I-Ming Jou[*]

[*]National Cheng Kung University Hospital, Taiwan
[#]Specailist Hospital, Johor, Malaysia
[†]Cheng Ching Hospital, Taichung, Taiwan

Background: Previous literatures showed congenital heart disease highly related to scoliosis. The operative methods such as median sternotomy, lateral thoracotomy were predisposed to the development of scoliosis. Nowadays, long term follow-up for thess patients is possible with the improved survival rate of congenital heart disease patients. The role of Blalock-Taussig shunt in scoliosis has not yet been reported as a risk factor in those patients.
Patients and methods: A retrospective review of spinal deformity in congenital heart disease patients who received Blalock-Taussig shunt, open heart surgery or lateral thoracotomy was performed in the National Cheng Kung University Hospital from 1989 - 2005. Serial chest roentgenograms were taken before and after operation in 221 patients to assess for the patients of spinal deformity. The curvature of the spinal deformity was defined by Cobb's angle. Comparison of scoliosis was made among the operative methods, and type of congenital heart disease. Follow-up was done up to 5 years and above.
Results: Total 45 patients had scoliosis, with curvature greater than 10 degree. Most of them (93.3%) had cyanotic heart disease. The incidence in patients who received Blalock-Taussig shunt was relatively high compared to median sternotomy and lateral thoracotomy. There was clinically significant with p-value<0.05.
Conclusions: Blalock-Taussig shunt may be one of the risk factors for scoliosis in patients with congenital heart disease. The prevalence of scoliosis increases in patients with cyanotic heart disease and treated with modified Blalock-Taussig shunt.

Abstract no: 1353

Peri-operative risk factors for in hospital death or retransplantation in paediatric heart transplant recipients

Rachel Vanderlaan, Cedric Manhiot, Jennifer Conway, Brian McCrindle and Anne Dipchand

Hospital for Sick Children, Toronto, Ontario, Canada

Introduction: While advances in surgical techniques and immunosuppression regimens have contributed substantially to the success of paediatric heart transplantation (HTx), increased understanding of peri-operative risk factors associated with death or retransplantation (ReTx) could potentially improve survival to hospital discharge.
Methods: Peri-operative risk factors were explored in 226 paediatric HTx recipients between 1995 and 2010.
Results: Mortality prior to hospital discharge occurred in 20 patients (9%), a further 5 patients (2%) underwent ReTx for early primary graft failure of whom 1 died peri-operatively. Death or ReTx in 9 patients (36%) occurred <48 hours of HTx secondary to primary graft failure (n=4), operative complications (n=3)

or multisystem organ failure (n=2), including 4/5 ReTx. Death or ReTx >48 hours was secondary to primary graft failure (n=6), infections (n=4), multisystem organ failure (n=3), rejection (n=2) and post-operative complications (n=1). In a multivariable regression model, factors associated with increased hazard of peri-operative death or ReTx were earlier year of HTx (HR:1.2, p=0.001), peri-operative use of factor VIIa (HR:32.9, p=0.001), post-operative chest reopening (HR:11.0, p=0.001), post-operative use of extracorporeal life support (HR:7.7, p<0.001), rejection prior to discharge (HR:6.0, p=0.03), donor negative rhesus factor (HR:6.2, p=0.001), and higher donor BMI (HR:1.033/kg/m^2, p=0.001). UNOS Status 1 (vs. Status 2) at the time of HTx was associated with increased hazard of death or ReTx from causes other than primary graft failure (71% vs. 27%, p=0.05), implying that patients on life support are at risk of death from non-cardiac causes despite receiving a HTx.

Conclusions: Important peri-operative risk factors for early death or ReTx post-paediatric HTx include surrogates of poor graft function or bleeding, and early rejection. Understanding risk factors that impact peri-operative outcomes may help with difficult decisions around timing of listing for primary HTx and appropriateness of relisting for ReTx.

Abstract no: 1354

Smartphone app help optimise inter-stage home monitoring for infants with single ventricle

Girish Shirali, Dawn Tucker, Richard Stroup and Brent Kevern

Childrens Mercy Hospital, Kansas City, Missouri, United States of America

Background: While the surgical results of the Norwood procedure have improved dramatically over the past 2 decades, post-discharge interstage morbidity and mortality remains a significant problem. While inter-stage home monitoring is widely employed to try to minimise adverse interstage events, the needs for data entry and interpretation by the family lead to a system that is imperfect at many levels. Advances in technology may facilitate improvements in automating home monitoring. We explored the use of an iPad and wireless technologies to help build an optimal solution for interstage monitoring.

Materials and methods: We started with standard devices that are in current usage for inter-stage monitoring (pulse oximeters and weight scales). We constructed an iPad app that uses Bluetooth and other wireless technologies to automate the upload of data from these devices to the iPad. The iPad camera is used to obtain a 10-second video of the patient. Family members are responsible for entering intake and output manually. All data is automatically transmitted to the hospital servers and integrated with the electronic medical record (EMR). Machine learning algorithms that have been developed within the EMR helps trend measurements and to triage clinical scenarios. The ability to view videos of the patient in a non-linear manner promises to yield important and previously unavailable insights as well as educational value for both providers and families.

Results: The prototype app has been developed and is functional. Its integration with an EMR system (Cerner) has been validated; validation with other EMR systems is in progress.

Conclusions: The ability to use a consumer device for automated interstage home monitoring could be an important advance in the care of these fragile and high-resource infants. Such technology could also be used for other high-risk children with congenital or acquired heart disease.

Abstract no: 1357

Australian and New Zealand population-based study of paediatric idiopathic pulmonary arterial hypertension

Michelle Rose[*], Clare O'Donnell[#], Ingrid King[*], Suzanna Vidmar[*], Robert Justo[†], Andrew Bullock[◊], Gary Sholler[§], Malcolm Richardson[Δ], Megan Sherwood[§] and Robert Weintraub[*]

[*]The Royal Children's Hospital, Melbourne, Australia
[#]Starship Hospital, Auckland, New Zealand
[†]The Mater Hospital for Children, Brisbane, Australia
[‡]Princess Margaret Hospital, Perth, Australia
[§]The Children's Hospital at Westmead, Sydney, Australia
[◊]The Women's and Children's Hospital, Adelaide, Australia

Background: Idiopathic pulmonary arterial hypertension (IPAH) in children is a rare and progressive condition with a poor natural history. Currently available therapies have not been trialled in children and little information exists about long-term survival in treated children.

Methods: A multicentre, web-based Australian and New Zealand registry for children with PAH was established to better understand the profile of children with IPAH, aiming to describe demographics, presenting features, clinical course and outcomes of children aged 3m - 18 years at time of PAH diagnosis, from 1 January 2002. A group of 9 historic controls with IPAH diagnosed from 1970 - 1990 were used for comparison.

Results: Of the 24 patients enrolled with IPAH, 54% were female and 75% Caucasian. The mean (SD) age at diagnosis was 9.3 (4.90) years with 50% in WHO functional class III at presentation. During follow-up, 20 (83%) were treated with an ERA (exclusively Bosentan monohydrate), 17 (71%) with PDE5 inhibitor (sildenafil) and 10 (42%) with either epoprostenol or iloprost; 19 (79%) received warfarin. The mean duration of follow-up was 3.45 years, with an annualised mortality of 6%. Study end-points of death/transplant at 9-years post PAH diagnosis was 25% for patients in the current era compared to 60% for historic controls (p<.007). Freedom from death, transplant or epoprostenol initiation for current era patients was 30% at 7 years after diagnosis. The severity of PAH from cardiac catheterisation was similar between patients and controls, except for a lower mean CI of 2.57+0.93 in current era group vs. 3.85+1.85 (p<.05) for the historic controls.

Conclusions: Multiple medical therapies, in particular, a high utilisation of epoprostenol have resulted in improved survival for children with IPAH. A better understanding of childhood IPAH will facilitate medical care, permit standardisation of therapeutic guidelines, and allow better representation in the healthcare arena.

Abstract no: 1358

Echocardiographic longitudinal study in children with acquired immunodeficiency syndrome

Maria Suely Bezerra Diógenes, Regina Calia Menezes Succi, Valdir Ambrosio Moisas, Solange Bernardes Tatani, Celia Maria Camelo Silva and Antonio Carlos Camargo Carvalho

Federal University of Sao Paulo, Brazil

Background: Cardiovascular abnormalities in children with acquired immunodeficiency syndrome (AIDS) is slow and progressive in occurrence. The most frequent echocardiographic abnormalities are left ventricular dilation and dysfunction.
Purpose: To study prospectively the cardiovascular profile of children with acquired immunodeficiency syndrome (AIDS) by Echocardiography.
Material and methods: Forty six HIV-1 positive children born to HIV-infected mothers that progressed to AIDS, 25 males and 21 females, age range from 4 months - 11 years, average 6 years, classified according to CDC's 1994 revised classification system, were longitudinally studied by two-dimensional and Doppler Echocardiography from 1995 - 2012. The mean follow-up period was 6 years for each child.
Results: Cardiac abnormalities were encountered in 12 children (26%): mild tricuspid regurgitation (16.6%); mild mitral regurgitation (8.3%); mild aortic regurgitation (8.3%); severe dilation of ascending aorta (8.3%); mild dilation of left ventricle (8.3%); pericardial effusion with cardiac tamponade (16.6%); mild–to-severe left ventricular dilation and dysfunction (33.3%); severe pulmonary hypertension with right ventricular dysfunction (8.3%). Eight cases belonged to advanced clinical-immunologic stage (C3) and the others had moderate signs and symptoms of AIDS. There was one case (C3) of reversibility of severe left ventricular dilation with dysfunction which is doing fine at the age of 17 years.
Conclusions: Cardiac abnormalities initially appeared in children with moderate symptoms (B1) of AIDS and were more frequent in the ones with severe symptoms (C3). The most frequent abnormality was dilated cardiomyopathy and it was reversible in one case.

Abstract no: 1361

Ten years experience in implantation of Contegra graft in young patients: Single centre study on outcomes

Phan-Kiet Tran, Edin Omerbasic, Jens Johansson-Ramgren, Sune Johansson and Torsten Malm

Children's Hospital, University Hospital, Lund, Sweden

Background: It is shown that the use of bovine jugular vein graft (Contegra) to reconstruct the RVOT is currently, in the mid-term perspective, the best alternative to homografts. Recent studies also report on a sustained freedom from re-intervention in the long term perspective for graft-sizes 16mm>. Here, we investigate the outcome in a group that consist of predominantly young patients, and consequently also a higher number of smaller sized grafts. We also investigated the value of post-implantation right ventricular and pulmonary arterial pressure to predict future stenosis.
Material and methods: A total of 94 grafts were implanted from 2002 - 2012. Mean patient age was 3.3 years (3 days - 18 years, median 1.25 years). Total follow-up time was 226 patient years (25 days - 10 years, median 1.84 years). Diagnoses included: 37 Pulmonary Atresia, VSD with or without MAPCA (39%), 15 Tetralogy of Fallot (16%), 14 Truncus (15%), 10 Transposition of Great Arteries (11%) and 18 other diagnosis (19%).
Results: Implanted graft sizes ranged from 12 - 22mm, with the predominance of 12mm (9%), 14mm (27%) and 16mm (38%) grafts. There were 3 mortalities, all post-operatively in-hospital, none related to the graft. Freedom of re-intervention was 67% for graft sizes 12 - 14mm and 93% for graft sizes 16mm>. Post-implantation RV pressure could not predict future risk of stenosis development, 39/4mmHg vs. 36/2mmHg, p>0.05 stenosis vs. normal respectively.
Conclusion: Compared to previous reports, our study group is made up by younger patients and a higher proportion of smaller grafts. Nevertheless, our data supports previous studies that have shown that smaller graft sizes, 12mm and 14mm, is the main independent risk factor for re-intervention. The Contegra grafts continue to a reliable and readily accessible alternative to homografts.

Abstract no: 1364

Role of reflux oesophagitis in precipitating ischaemic symptoms

Datshanna Naidoo and Sunil George

University of KwaZulu-Natal, Durban, South Africa

Background: A link has been postulated between ischaemic heart disease (IHD) and gastric-oesophageal reflux disease (GORD). To what extent GORD may precipitate angina is not clear. In 1985 Davies suggested that reflux may precipitate angina events. We designed a study to investigate the role of GORD in precipitating angina.
Hypothesis: It is possible that reflux symptoms may precipitate angina events.
Aim: To determine whether the presence of GORD may serve as a trigger for symptoms of ischaemia in subjects with coronary artery disease (CAD); to document the electrocardiographic changes (ECG) changes that occur during reflux.
Method: Patients with endoscopically confirmed GORD, as well as recent MI admitted to CCU constituted the study group. Patients with GORD underwent endoscopy and acid installation to determine whether GORD could induce ischaemia. Hydrochloric acid (0.1N) was instilled during endoscopy and the ECG simultaneously recorded. Ischaemia was detected by ST changes during ECG monitoring. Nuclear imaging with Methoxyisobutylisonitrile (MIBI) scanning was performed to look for objective evidence of ischaemia. Twenty normal volunteers served as control subjects.
Results: To date 23 subjects with GORD have been studied. ST segment changes were noted in 12 out of 23 and 5 showed ischaemia on MIBI scanning. In the IHD group 21/35 showed ST changes with 33/35 showing changes consistent with ischaemia on MIBI scanning. Other ECG changes were frequent during acid installation. Only 3 subjects showed no changes on the ECG monitoring.
Conclusion: Acid reflux produces ischaemic changes on the ECG recording and may lower the threshold for angina and coronary events. We present evidence to suggest that coronary vascular changes may arise from oesophageal stimulation and may lead to the development of myocardial ischaemia.

Abstract no: 1377

Successful surgical correction of Type II truncus arteriosus at 3 years of age

Amitabha Chattopadhyay, Debasree Gangopadhyay, Biswajit Bandyopadhyay, Hemant Nayak, Navdeep Singh, Prashant Thakur, Mahua Roy, Rhitajyoti Sengupta, Mrinalendu Das and Debasis Das

Department of Paediatric Cardiology and Paediatric Cardiac Surgery, Rabindranath Tagore International Institute of Cardiac Sciences, Mukundapur, Kolkata, India

Background: Truncus arteriosus is an uncommon congenital cardiac malformation. The condition presents in early infancy with features of pulmonary over-circulation as the neonatal pulmonary hypertension regresses. We report the clinical case of a girl from rural Bengal whose 1st presentation to a paediatric cardiac facility took her 3 long years after birth.
Materials and method: Initial clinical examination and investigations showed cardiomegaly, with biventricular enlargement and pulmonary plethora. On echocardiography she was diagnosed to have type II truncus arteriosus with mild truncal valve regurgitation. Cardiac catheterisation was done for operability which showed pre-oxygen pressures of the aorta and pulmonary artery to be 100/56/75mmHg and 84/49/69mmHg. Post-oxygen aortic and pulmonary artery pressures were 105/60/80mmHg and 72/40/50mmHg respectively, which showed a drop of 20mmHg in the mean PA pressures post oxygen, depicting operability. She underwent corrective surgery with closure of VSD with a flap technique and reconstruction of right ventricular outflow tract with 16mm valved conduit.
Results: Post-operatively she was managed with inhaled nitric oxide, sildenafil and bosentan. She required prolonged ventilation and inotropic support. Her recovery was good and echocardiography on 27th post-operative day documented good bi-ventricular contractility, estimated pulmonary arterial systolic pressure of 34mmHg and mild truncal valve regurgitation.
Conclusion: Most of the world literature on surgical correction of truncus arteriosus report surgery at a very young age which is explained by the early irreversible change of pulmonary vasculature these patients develop if remain uncorrected. Our patient along with few other published reports testify to the fact that meaningful surgical outcome is still possible in a select subset of patients with this condition who present late, if chosen carefully on clinical grounds.

Abstract no: 1378

Vascular mechanics at rest and during exercise after arterial switch operation for complete transposition of the great arteries

Robin Hay-Son Chen, Wilfred Hing-sang Wong and Yiu-fai Cheung

Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Hong Kong University, Hong Kong

Background: Progressive neoaortic root dilatation and regurgitation after arterial switch operation (ASO) for complete transposition of the great arteries (TGA) are well documented. We tested the hypothesis that neo-aortic stiffness is increased and is associated with neo-aortic dilatation in patients after ASO. We further explored the changes in mechanics of the neo-aorta during exercise stress.
Methods: Thirty patients (22 males) aged 16.2±2.1 years and 22 healthy controls (15 males) were studied. Central and peripheral arterial pulse wave velocity (PWV), carotid (c-AI) and radial (r-AI) augmentation indices and central systolic blood pressure (CsBP) were assessed by applanation tonometry. Dimensions of the aortic annulus, sinus, sinotubular junction, ascending aorta, and right carotid artery dimensions were determined at rest and during exercise by 2-dimensional echocardiography. Aortic strain, distensibility, aortic and carotid stiffness indices were calculated.
Results: At rest, patients compared with controls had higher c-AI, heart-carotid PWV, CsBP, and r-AI (all $p<0.05$), while brachial-ankle arterial PWV were similar. During at rest and exercise, patients had significantly lower aortic strain and distensibility, and greater systolic blood pressure, and aortic and carotid stiffness were significantly lower ($p<0.05$). Aortic root dimensions at all levels were significantly greater in patients compared with controls (all $p<0.05$). Patients with aortic dilatation had higher CsBP and aortic stiffness at rest, and lower aortic strain and distensibility at rest and at submaximal exercise (all $p<0.05$). Linear regression model identified resting aortic distensibility ($\beta=-0.57$, $p=0.005$) and age at operation ($\beta=0.40$, $p=0.005$) as significant determinants of aortic sinus Z-score. Significant aortic regurgitation was identified in 18.8% (6/32) of patients, in whom significant higher Z-scores for aortic annulus and sinotubular junction were found (both $p<0.05$).
Conclusions: In adolescents late after ASO for TGA, aortic root dilatation and regurgitation is prevalent and is associated with stiffening of central arteries at rest and during exercise.

Abstract no: 1379

Presentation and treatment outcome of TAPVC in older population: A single centre experience from India

Amitabha Chattopadhyay, Debasree Gangopadhyay, Biswajit Bandyopadhyay, Hemant Nayak, Navdeep Singh, Prashant Thakur, Mahua Roy, Rhitajyoti Sengupta, Mrinalendu Das and Debasis Das

Department of Paediatric Cardiology and Paediatric Cardiac Surgery, Rabindranath Tagore International Institute of Cardiac Sciences, Mukundapur, Kolkata, India

Background: TAPVC is a rare congenital heart disease which is diagnosed in the neonatal period or in infancy. This study investigates the spectrum of presentation and treatment outcome of an Indian cohort who presented >age 5 with TAPVC.
Methods: Isolated TAPVC cases were identified from our institutional database from 2003 - 2012. We reviewed the medical records of 12 patients that belonged to the age group specified and analysed the data.
Results: Total 98 patients were diagnosed with TAPVC. Of them12 patients (12.24%) presented after 5 years. Median presenting age was 13.25 years (range 5 - 30 years) with 8 males (66.66%) and 4 females (33.33%). Unobstructed supracardiac TAPVC was the most common diagnosis and shortness of breath on exertion the most common presenting complaint. All patients were investigated with ECG, chest X-ray, echocardiography pre- and post-operatively. All were in sinus rhythm pre- and post-operatively except 1 who had transient atrial fibrillation in the immediate post-operative period. TAPVC with moderate pulmonary arterial hypertension was the most common echocardiographic diagnosis pre-operatively. All the patients underwent rerouting of pulmonary veins and closure

of ASD with flap technique. One patient died 40 days after operation from sepsis and multi-organ failure. Mean duration of ventilation was 20 hours. Median duration of hospital stay after surgery was 10.5 days. The post-operative period was uneventful for all except 1 who required longer duration of inotropic support and ITU stay. Only 1 patient required oral sildenafil for PAH post-operatively. The median duration of follow-up was 25.5 months (range 1-83 months). Three patients were lost to follow-up. All patients were asymptomatic during follow-up and were in sinus rhythm.
Conclusion: Patients diagnosed late with TAPVC are those who are naturally selected for better survival. Operative outcome is good with complete reversal of PAH in majority.

Abstract no: 1387

Adenosine improves the efficacy and cost-efficiency of transoesophageal electrophysiology studies to risk stratify patients with asymptomatic Wolff-Parkinson-White syndrome

Christopher Snyder[†], Walter Hoyt[*] and Joni Steinberg[#]
[*]Department of Paediatric Cardiology, University of Virginia, Charlottesville, Virginia, United States of America
[#]Department of Global Health Systems and Development, Tulane University, New Orleans, Louisiana, United States of America
[†]Rainbow Babies and Children's Hospital, Case Western University School of Medicine, Cleveland, Ohio, United States of America

Introduction: All patients with Wolff-Parkinson-White (WPW) Syndrome, even if asymptomatic, are at risk of sudden death during atrial fibrillation (AFib). This risk can be determined by measuring the shortest pre-excited R-R interval during AFib. This study evaluated adenosine's potential to induce AFib during transoesophageal electrophysiologic studies (TEEPS) when atrial-pacing modalities alone failed to do so and analysed its cost-savings potential during the workup of asymptomatic WPW.
Methods: A retrospective review of patients with WPW who had a TEEPS performed.
Inclusion criteria: Evidence of WPW on electrocardiogram, no history of arrhythmias or associated symptoms, age <18 years, and failure to induce AFib by initial TEEPS protocol. When TEEPS protocol was unsuccessful, adenosine 0.2 milligrams/kilogram was administered via rapid intravenous push during atrial-burst pacing. AFib was considered successfully induced if persisting >20s. If induced, patients were classified as at RISK of sudden death if the shortest pre-excited R-R interval during AFib was <250ms or NO RISK if ≥250ms. Using 2011 and 2012 Medicaid reimbursement data the cost of adenosine during TEEPS was compared to proceeding directly to a transvenous electrophysiologic study (TVEPS).
Results: Inclusion criteria was met by 7 patients. Adenosine and atrial-burst pacing induced AFib in 4 of these patients (57%). Of those induced, 3 (75%) had NO RISK and 1 (25%) had RISK. No complications occurred. The average cost of TEEPS was $999, TVEPS $4524, and adenosine $84.21. The average cost of the adenosine decision-arm was $2669.35, resulting in an average cost-saving of $1854.65.
Conclusions: To improve the efficacy and cost-efficiency of TEEPS to risk stratify patients with asymptomatic WPW, we recommend rapid intravenous adenosine administration during atrial-burst pacing when prior modalities have failed to induce AFib.

Abstract no: 1389

Reference echocardiographic measurements in low birth weight infants of a developing country

Stephen Brown, Daniel Buys, Salomi Jacobs and Carri-Lee Greig
Department of Paediatric Cardiology, University of the Free State, Bloemfontein, South Africa

Background: To obtain a set of reference echocardiographic values in a group of low birth weight infants of central South Africa as none exist for sub-Saharan Africa.
Methods: Over a 12-month period, 290 infants were included. Standardised M-mode, two-dimensional and systolic functional assessments (Shortening fraction[SF], myocardial performance index [MPI]) were carried out according to the guidelines of the American Society of Echocardiography. Studies were performed by 3 experienced echocardiographers and reviewed by a paediatric cardiologist. A longitudinal study was also included to examine changes in these indices from days 1 - 28 of life.
Results: Median weight was 1.36kg (range: 0.69 - 2.50) with a median gestational age of 31 weeks (range: 26 - 38). 87 (29%) infants were small for gestational age (SGA). Body surface area (BSA) and weight had a near perfect correlation (r=0.98). Inter-observer variation was less than 6%. Cardiac dimensions increased with increase in body weight. SF and MPI for left and right ventricles were - 34.6±6%, 0.29±0.14 and 0.23±0.15 respectively. Dimensions of SGA infants did not differ from other infants of comparable weight. Longitudinal data showed that all dimensions from birth to 28 days remained within the ranges determined by the study. Comparison with international reference ranges showed that local interventricular septal and posterior wall thicknesses as well as left atrial dimensions were significantly larger ($p<0.01$).
Conclusion: Cardiac dimensions increased parallel to increase in body weight. BSA and weight correlate excellently and either can be used. Longitudinal data indicate that reference values obtained are applicable to infants from 0 - 28 days of age. The differences in certain cardiac dimensions from international reference values demonstrate that regional differences exist and emphasise the need for development of local reference ranges.

Abstract no: 1395

Congenital complete heart block: Review of cases seen at Chris Hani Baragwanath academic hospital from 1986 - 2012

Gcina Dumani, Barend Fourie, Paul Adams and Antoinette Cilliers
Division of Paediatric Cardiology, Chris Hani Baragwanath Academic Hospital, University of Witwatersrand, Johannesburg, South Africa

Introduction: Congenital complete heart block is a rare conduction anomaly which may either be isolated or associated with congenital structural heart disease. Pre-natal diagnosis, identification of high risk neonates and early planned management is associated with improved outcomes.
Methods: Retrospective clinical review of 30 cases from 1986 - 2012 presenting to the Division of Paediatric Cardiology at the Chris Hani Baragwanath Academic Hospital which is a Southern African Tertiary care institution. Data related to clinical features, diagnosis, pacemaker interventional procedures and follow-up was collected.
Results: Thirty four patients (18 males, 16 females) were diagnosed: 33 (97,1%) post-natally, and 1 ante-natally. Eight patients were delivered by Caesarean section for fetal distress including 6 premature babies. Presenting features included bradycardia, congestive cardiac failure, respiratory distress, cardiac murmur, cardiomyopathy with poor left ventricular function and metabolic acidosis. Median age at diagnosis was 2 days. Antibodies Ro/Lo were positive in 21 patients (61.8%).The major associated cardiac lesion was patent ductus arteriosus in 15 patients, 4 of whom had left ventricular non-compaction. Four patients had

heterotaxia associated with atrioventricular septal defects. Other cardiac defects found in 2 patients were valvar pulmonary stenosis and a secundum atrial septal defect. The median ventricular rate was 50 beats per minute. The median atrial rate was 150 beats per minute. Four patients (11.8%) had QTc interval >460 milliseconds. Two patients had a wide QRS duration greater than 120 milliseconds. Thirteen patients (38.2%) including 6 neonates had permanent pacemaker insertion at median age of 5 weeks. Thirteen patients (38.2%) died prior to permanent pacemaker placement including 3 patients with heterotaxia. Three patients died after permanent pacemaker insertion.
Conclusion: Congenital heart block in our setting has a high mortality and is linked with premature delivery. The majority of patients are associated with autoimmune antibodies.

Abstract no: 1397

Investigating syncope in children: Where to draw the line

Ranya Hegazy, Sonia ElSaeidy, Wael Lotfy and Osama AbdelAziz
Department of Paediatrics, Faculty of Medicine, Cairo University, Egypt

Background: Syncope is the temporary loss of consciousness and postural tone resulting from an abrupt transient decrease in cerebral blood flow. Its death-like quality makes it an alarming presentation for parents and investigating it is troublesome, expensive and often fruitless. The present work aimed at determining the yield of diagnostic tests in paediatric syncope at a tertiary paediatric referral centre and to report on the prevalence of cardiac diagnoses among these patients.
Materials and methods: The current work is a retrospective study conducted at a tertiary referral arrhythmolology service. The clinical charts of 1896 paediatric patients presenting with a primary complaint of syncope with an average age of 8.3±2.1 (3.4 - 17) years were reviewed by the investigators. Statistical Package of Social Science (SPSS) version 14 was used for the analysis of data. A complete history, physical examination, 12-lead ECG, EEG, echocardiography and Holter monitoring were done for the whole study group.
Results: The commonest trigger for syncope in the study population was early following exercise (n=526) and the commonest prodrome was palpitation, noted in 190 patients. A murmur was present in 147 of our patients (7.7%) while 107 (5.6%) had abnormal ECGs. Echocardiography revealed a cardiac cause in 109 (5.7%) of them, 2 of them were missed by ECG. The most common cardiac cause was HOCM. All patients were offered ambulatory 24-hour ECG. One patient with sick sinus syndrome was diagnosed only with Holter. EEG diagnosed a neurological cause for syncope in 88 (4.6%) patients, all suspected through history taking.
Conclusions: 12-lead ECG and history taking are the most cost beneficial investigations in diagnosing syncope in children. Transthoracic echocardiography, Holter monitoring and EEG have low yield in paediatric syncope and should be reserved to when an abnormality is suspected from history or 12-lead ECG.

Abstract no: 1398

The yield of ambulatory 24-hour Holter monitoring in paediatric patients

Ranya Hegazy, Wael Lotfy and Osama AbdelAziz
Department of Paediatrics, Faculty of Medicine, Cairo University, Egypt

Background: The diagnosis of arrhythmias may be a tedious process due to the episodic and often transient nature of the disease, hence the need for longer periods of ECG recording. Holter monitoring (HM) has been established as one of the most effective non-invasive clinical tools in the diagnosis, assessment and risk stratification of cardiac patients. However studies with regards to its value in children remain debatable. The present work aims at determining the value of HM in the diagnosis and management of children.
Materials and methods: The present work is a retrospective study conducted at a tertiary referral arrhythmolology service. Holter records of 4 015 paediatric patients (52% males and 48% females) with an average age of 4.8±2.9 years (2 days - 16 years) were reviewed. The indications for which Holter monitoring was done were analysed as well as all the abnormalities diagnosed and factors that may increase Holter yield.
Results: The most common indications for were palpitations (23%), syncope (16%), cardiomyopathy (14%), chest pain (8%), evaluation of anti-arrhythmic therapy (8%), post-operative assessment (3.7%) and complete AV Block (4.5%). A sum of 602 Holter recordings was found abnormal with a total diagnostic yield of 15%. The highest contribution to diagnosis was in post-operative assessment (37%), cardiomyopathy (28%) and palpitations (14%) where the most common abnormalities were frequent supraventricular/ventricular premature beats, supraventricular tachycardia, ventricular tachycardia and AV block. Diagnostic yield was lowest in patients' syncope (0.4%) and chest pain (0%). An abnormal ECG was significantly associated with a higher diagnostic yield (p=0.0001).
Conclusions: HM has an extremely valuable role in the assessment of high risk patients (post-operative and cardiomyopathy) and in palpitations. However, in children with syncope and chest pain HM has a low yield. In this group of patients an abnormal ECG is more likely to be associated with abnormal Holter recordings.

Abstract no: 1401

Tele-cardiology partnership between Washington, DC and Marrakech, Morocco: Supporting a growing paediatric cardiovascular service in the developing world

Yassine Boukaidi[*], Drissi Boumzebra[*], Saloua Karimi[*], Mary Fuska[#], Molly Reyna[#], Philip Hopkins[#], Ron Dixon[#], Svetlana Sinykin[#], Ouahbi Karim[*] and Craig Sable[#]
[*]Hospital Ibn Tofail, Marrakech, Morocco
[#]Children's National Medical Centre, Washington DC, United States of America

Background: Only 7% of the world's population has access to modern paediatric cardiac care, resulting in nearly 6 million children with treatable conditions who are denied care. Telemedicine has the potential to help bridge this gap by providing remote consultation and distance education.
Methods: Children's National Medical Centre developed a tele-medicine partnership with Marrakech, Morocco (4 - 5 hour time difference) to augment the skills of the paediatric cardiovascular team. Video-conferencing units and satellite dishes were installed in 2009 with subsequent training in 2010.

Results: Live monthly video-conferences were started in 2009, increasing to weekly in 2011 between cardiovascular teams in Washington (CS, SS) and Marrakech (DB, YB). Patient data and echocardiograms were reviewed in real time. The Children's technical team managed conferences remotely. Thirty eight conferences occurred in the last 12 months. Fourteen were cancelled due to scheduling conflicts (no technical difficulties). Ninety five cases/73 patients were presented; 22 were discussed more than once. Most common diagnoses were Tetrology of Fallot (n=14), transposition of the great arteries (n=10), double outlet right venticle (n=9), atrioventricular canal (n=8), and ventricular septal defect (n=6). Mean age was 4.8 years (3 days - 30 years). 44 cases <2 years. Mean oxygen saturation was 83%; 22≤80%. Additional imaging was recommended in 22 patients; improvement in echocardiography skills was observed. Cardiac surgery was performed in 25% of patients, half had a difference in approach as a result of the teleconference. Three operations (Tetralogy of Fallot, atrioventricular canal, D-Transposition) were performed sucessfully in infants for the 1st time. Meetings with US and Moroccan government officials contributed to ongoing support. Focus on barriers including technology, satellite availability, language, funding and time difference has contributed to the sustainability of the project.
Conclusions: Tele-medicine is an innovative and practical means to augment the skills of paediatric cardiovascular surgery teams in the developing world.

Abstract no: 1402

Intermediate term results following post-cardiotomy extracorporeal membrane oxygenation (ECMO) support in congenital heart surgery (CHS) patients

Syed Murfad Peer*, Dominic A. Emerson*, Michael Philippe-Auguste*, David Zurakowski#, Pranava Sinha*, Richard A. Jonas*, John T. Berger* and Dilip S. Nath*

*Children's National Medical Center, Washington DC, United States of America
#Children's Hospital, Boston, United States of America

Background: There is considerable data regarding in-hospital results of CHS patients (pts) requiring post-cardiotomy ECMO; however there is limited information on mid-term outcomes.
Material and methods: A retrospective single institutional review of 25 consecutive CHS pts who survived to hospital discharge following post-cardiotomy ECMO between January 2003 and June 2008 was completed. Primary endpoint was survival at last follow-up (f/u). Secondary endpoints included evidence of (1) neurological deficits (2) renal injury (3) respiratory failure and unplanned (4) cardiac re-interventions (5) hospitalisations.
Results: Median age at ECMO support was 4 months. Primary indications for ECMO included cardiac arrest (n=12.48%), low cardiac output (n=7.28%), failure to wean from CPB (n=5.20%), and hypoxia (n=1.4%). There were 24 survivors; 1 death occurred within 48 hours of hospital discharge. Median f/u was 3.4 years (IQR:1.7 - 6.0). Kaplan-Meier patient survival is 95% at 3 years (95% CI: 88% - 99%). Neurological deficits were present in 7 pts (cognitive deficits in 5 pts; motor deficits in 3 pts). No pts had evidence of renal dysfunction. At hospital discharge, 3 pts had respiratory failure requiring tracheostomy and at latest f/u. 1pt remained dependent on mechanical ventilation. Echocardiogram in 23 pts demonstrated normal systemic ventricular (SV) function in 19 pts and mild-moderate SV dysfunction in 4 pts. Unplanned cardiac re-interventions were required in 13/19 pts (68%): both percutaneous and operative in 4 pts (21%), solely percutaneous in 6 pts (32%) and solely operative in 3 pts (16%). Unplanned hospital re-admissions were documented in 9 pts.
Conclusions: Post-hospital discharge outcomes of CHS pts requiring post-cardiotomy ECMO are encouraging with respect to survival, respiratory and renal assessments. While SV function is largely preserved in the mid-term, rate of unplanned cardiac re-interventions is high. Neurological impairment remains a concern with evidence of both cognitive and motor deficits.

Abstract no: 1405

Repair of anomalous left coronary artery from the pulmonary artery: Outcomes and follow-up

Gladys Salgado*, Rodrigo Oribe*, Solana Villafañe Molina*, Marta Lopez*, Jose Alonso*, Jose Suarez# and Horacio Capelli*

*Department of Cardiology, Hospital de Pediatria J.P. Garrahan, Buenos Aires, Argentina
#Deparment of Cardiovascular Surgery, Hospital de Pediatria J.P. Garrahan, Buenos Aires, Argentina

Backround: We reviewed surgical early and late results of children with anomalous left coronary artery from pulmonary artery (ALCAPA).
Material/Methods: Between July 1994 and June 2012, 40 patients with ALCAPA were repaired in our hospital. The median age was 4.5 months (range 1 - 156), mean weight 6kg (range 3 - 63). Patients were divided in 2 groups: (1) Direct coronary transfer (n=34) and (2) Coronary ligation (n=6). Three simultaneous annuloplasties were performed. The mean follow-up was 4.12 years (range 0.25 - 16.1).
Results: There were 2 hospital deaths: group I: 1/34 (2.9%), group II: 1/6 (16.6%) and one late sudden death in each group. Both techniques were equally effective regarding left ventricular shortening fraction (LVSF) (long term follow-up: 38% vs. preoperative: 23%) and mitral valve competence (mitral regurgitation (MR) was absent or mild in 76% of patients in long term follow-up). One patient required mitral valve replacement two months after coronary transfer. Four patients underwent a redo: second mitral annuloplasty, coronary artery bypass grafting, mitral valve re-replacement and pulmonary supravalvular stenosis repair (1, 10, 11 and 13 years after surgery). All survivors remain asymptomatic. Sixteen patients were catheterised during follow-up. In 4 patients left coronary artery was occluded, everyone with normal LVSF and 2 of them, with normal excersice test. It was performed in 15 patients: 5 were abnormal, 2 of them however had a normal coronary angiography.
Conclusions: A dual-coronary system could be stablished safetly in children with ALCAPA. Early mortality was related to severity of preoperative left ventricular dysfunction. Late results are satisfactory, with marked improvement of left ventricular LVSF and mitral valve competence. The echocadiographic assesement and exercise test are usefull tools to evaluate a potential myocardial injury, however those 2 methods didn't predicted patency of the coronary arteries. Therefore, a coronary angiography should be included in the follow-up evaluation.

Abstract no: 1410

Cardiac resynchronisation therapy (CRT) in paediatric cardiology: A new tendency or an exception?

W. Tetsuji Tamaki, Christiane Zambolin, Elizabeth Crevelari, Roberto Costa, Silvana Nishioka, Martino Martinelli, Anisio Pedrosa and Fabio Jatene

InCor, HCFMUSP, Brazil

Background: Biventricular pacing was introduced in the middle 90's as new modality of treatment for patients with drug-refractory dilated heart failure (F.E.<0,3) and ECG evidence of dyssynchronous ventricular contraction. Initially the 3rd lead was exclusively placed by thoracotomy in the left ventricle wall (epimyocardial). The 1st system to be placed via left ventricular leads through the coronary sinus was introduced in 2000. This study reports our clinical experience with Cardiac Resynchronisation Therapy, in patients <18 years annually since 1997.

Methods and results: We review our cases in CRT, since the 1st surgical procedure in 1997 - 2012. Since 1997 we implanted 1 135 CRT, but only 16 patients were <18 years old (1.41%). In both groups there were a major prevalence of male 68% and 75% in adulthood (Group I) and <18 years (Group II) respectively (Table 1). Group II is described in Table 2. Five patients had Complete Congenital Heart Block with pacemaker VVI. They developed Dilated Myocardial Insufficiency (from 2 months - 8 years) and elected to have CRT.

TABLE 1: Resynchronisation cases since 1997 - 2012

Year	Patients(>18y)	Patients(≤18y)	Total	% ≤18y
1997	3		3	0.00%
1998	6		6	0.00%
1999	9		9	0.00%
2000	24		24	0.00%
2001	88	1	89	1.12%
2002	86	4	90	4.44%
2003	131	1	132	0.76%
2004	145	1	146	0.68%
2005	113		113	0.00%
2006	89	2	91	2.20%
2007	109	2	111	1.80%
2008	62	2	64	3.13%
2009	56	1	57	1.75%
2010	63	1	64	1.56%
2011	92		92	0.00%
2012	43	1	44	2.27%
Total	**1119**	**16**	**1135**	**1.41%**
Age (mean. sd)	**61.6 (13.0)**	**12.1 (4.8)**		
Female	354(32%)	4(25%)		
Male	765(68%)	12(75%)		

TABLE 2: Etiology of CRT patients under 18 years old

RG	2001	2002	2003	2004	2006	2007	2008	2009	2010	2012	Diagnostic
5096962G					1						CCHBupgrade
5106557B						1					CTGA Ebstein Hipopl. RV EP
5134176E				1							T4F
5144514C								1			CCHBupgrade
5151092J		1									Rastelli operation
5173119A						1					T4F
5234028D		1									CCHBupgrade
5257587F							1				RCM FA Ablation 6 year upgrade
5268877H					1						DORV Hipopl. LV CIA CIV
5282046B			1								CCHBupgrade
55310866I		1									CTGA CIA CIV EP
55311008H							1				TGA
55370106H		1									T4F
554549/01										1	CCHBupgrade
55505498C									1		CTGA 8 years from first op.
6044994F	1										Sub Aortic Stenosis
Total	**1**	**4**	**1**	**1**	**2**	**2**	**2**	**1**	**1**	**1**	

CCHBupgrade: Complete Congenital Heart Block/upgrade VVI to CRT. T4F: Tetralogia Fallot. TGA: Transposition of Great Arteries. CTGA: Correct Transposition of Great Arteries. CIA: Intra-Arial communication. CIV: Intra-Ventricular Communication. EP: Pulmonary Stenosis. Hipopl. (R/L)V: Hipoplaisa of (Right/Left) Ventricule. RCM: Restrict Miocardiopathy. FA: Atrial Fibrilation. DORV: Double Outlet of the Right Ventricle.

Discussion: CRT implantation in children is still limited by generator size, lead diameter incompatibility to vascular dimensions and, in adults, the epicardial leads are exceptions whereas they are more commonly found in children. CRT is indicated in very specific cardiological condition. Even in those cases the most optimistic expectancies show three quarters of responders and low impact in survival. Besides the growing indication and feasibility in infants and neonates we still have a lack of evidence, a consistence rationality for them.

Conclusion: Besides our experience with adults and their trials with CRT in the paediatric cardiology there isn't enough evidence to answer Why? For whom? When? or How?

Abstract no: 1411

Lateral tunnel growth: Magnitude and impact on the total cavopulmonary resistance

Maria Restrepo*, Lucia Mirabella*, Elaine Tang#, Christopher M. Haggerty*, Malavika S. Mundkur*, Mark A. Fogel†, Anne Marie Valente‡, Doff B. McElhinney‡ and Ajit P. Yoganathan*

*Georgia Tech and Emory University, Atlanta, Georgia, United States of America
#Georgia Institute of Technology, Atlanta, Georgia, United States of America
†Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America
‡Childrens Hospital Boston, Boston, Massachusetts, United States of America

Background: The total cavopulmonary connection (TCPC) is usually performed with a lateral tunnel (LT) pathway or an extra-cardiac (EC) conduit. Growth potential of the LT pathway has been hypothesised as one of the main benefits of this strategy compared to an EC conduit. However, the nature, magnitude, and haemodynamic implications of LT growth have not been characterised.

Materials and methods: LT (n=7) and EC (n=3) patient-specific TCPC anatomies and flows were reconstructed from retrospective serial cardiac magnetic resonance images (cMRI). Vessel centre lines were extracted using the Vascular Modelling Toolkit for the Fontan pathway (FP), superior vena cava (SVC), descending aorta (dAo) and pulmonary arteries (PA). Parameters computed included: minimum, mean, and maximum diameters, FP volume, Nakata index and McGoon ratio. To understand the impact of growth on TCPC resistance (RTCPC), computational fluid dynamic simulations were performed for the LT group by simulating both serial changes (S1, S2) and absence of growth (S3).

Results: Results are summarised in the Table. Absolute and normalised diameters increased in all vessels in the LT patients; the EC patients' vessels growth was less than expected. Changes in the FP volume were one order of magnitude larger in the LT than in the EC. Haemodynamically, average RTCPC was seen to increase serially by 20%, while in growth absence it increased by 57%, indicating that TCPC growth helps limit the RTCPC increase while flow rate changes in time.