

**TABLE 1:** Geometrical comparison

	LT (n=7)			EC (n=3)			
	T1	T2	Normalised Change	T1	T2	Normalised Change	
Body surface area (m <sup>2</sup> )	0.9±0.2	1.4±0.2		0.8±0.1	1.4±0.3		
Age at CMR (years)	8.7±3.4	13.5±1.9		8.7±2.1	12.7±1.5		
Mean diameter (mm)	FP	14.9±3.8	18.4±4.7	5.4±2.1	15.1±0.3	15.8±0.6	0.8±0.7
	SVC	11.7±3.4	15.0±5.4	5.2±5.3	12.9±0.2	14.5±1.7	2.0±1.3
	dAo	10.3±1.5	13.6±1.8	5.2±2.0	11.7±0.5	11.8±1.0	0.3±0.9
	LPA	10.6±3.0	11.7±3.0	1.6±3.6	10.3±2.1	11.2±5.2	0.3±4.8
	RPA	10.9±3.9	14.4±3.3	5.2±3.0	11.1±2.8	11.3±4.4	-0.3±2.2
Diameter at PA branching (mm)	16.4±5.7	18.9±9.0	3.8±6.2	15.6±1.7	15.1±1.8	-0.6±0.1	
FP volume (cm <sup>3</sup> )	16.1±7.6	29.4±15.7	20.9±14.8	13.0±1.9	16.8±3.0	5.4±1.1	
Mean Nakata index (mm <sup>2</sup> /m <sup>2</sup> )	207.5±52.6	205.2±45.4		237.7±99.4	154.4±60.6		
Mean McGoon ration	2.1±0.4	1.9±0.3		1.8±0.3	1.9±0.7		

**TABLE 2:** Haemodynamic metrics for the LT group

	T1	T2
Cardiac index (L/min/m <sup>2</sup> )	2.8±0.6	2.8±0.4
TCPC resistance (WU)	S1 (T1 geometry,T1 flow)	0.5±0.3
	S2 (T2 geometry,T2 flow)	0.5±0.4
	S3 (T1 geometry,T2 flow)	0.7±0.8
	Serial increase (S2 - S1)	20%
	Growth absence (S3 - S1)	57%

Results are shown as mean ± standard deviation. T1: initial scan, T2: second scan, FP: Fontan pathway, LPA: left pulmonary artery, RPA: right pulmonary artery. All geometrical changes were normalised by the square root of the change in body surface area.

**Conclusions:** Geometric changes over an average span of almost 5 years showed cross-sectional and volumetric growth of the LT pathway. Based on simulations, growth had a positive effect on haemodynamic efficiency by reducing RTCPC even when some growth was accounted for. These findings support the rationale of LT TCPC growth potential, and may have implications for understanding the relative benefits of different approaches to the Fontan procedure.

**Abstract no: 1412**

**Distribution of heart disease in a multicentre paediatric cardiac registry**

**Pouya Hemmati<sup>\*</sup>, James St. Louis<sup>#</sup>, Lazaros Kochilas<sup>#</sup> and Lee Pyles<sup>#</sup>**

<sup>\*</sup>University of Minnesota Medical School, Minneapolis, Minnesota, United States of America

<sup>#</sup>University of Minnesota Amplatz Children’s Hospital, Minneapolis, Minnesota, United States of America

**Background/hypothesis:** The incidence of congenital heart disease is generally 8:1 000 live births. The University of Minnesota’s multicentre Paediatric Cardiac Care Consortium (PCCC) has enrolled children undergoing cardiac catheterisation, operation or autopsy examination since 1982. The study comprises all registered children born from 1991 - 2002 to state residents of Arkansas (AR), Minnesota (MN) and Missouri (MO). We hypothesise that age-stratified incidence of heart disease can be established for these states to investigate paediatric cardiac case-finding and management.

**Materials and methods:** De-identified PCCC records for children born from 1991 - 2002 in AR, MN, and MO were analysed and categorised by diagnosis to calculate birth rate-adjusted incidence. Conditions with abnormal oximetric saturation (see table) that were detected by 3 months of age are “Oximeter-Screenable” anomalies.

**Results:** The rate of registered heart disease for subjects born in 1991 - 2002 for the 3 states is 46.3 per 10 000 live births (1.8 million births). In PCCC registrations within the 1st 3 months of life, 17.1/10 000 are “Oximeter-Screenable” and 13.7/10 000 are potentially cyanotic (e.g. PS, CAVC), compared with the total incidence of 37.0/10 000. MO shows significantly higher registration rates for CAVC (Poisson distribution p=0.012), coarctation (p=0.0015), ToF (p=0.028) and VSD (p=0.0029). AR registers more than half of left-to-right shunts within the 1st 3 months [9.4/10 000 for <3 months vs. 18.5/10 000 all ages (Poisson distribution p=0.00064)].

	AR	MN	MO	Total	n total
<b>All registrants 1992 - 2001</b>					
Cyanotic	9.1	7.9	9.7	8.8	1 579
Left-to-right shunt	18.5	18.0	23.0	20.7	3 586
Overall rate	47.4	41.0	50.6	46.2	8 232
<b>Registration in 1st 3 months of life 1992 - 2001</b>					
Oximeter screenable: TOF,TGA,TAPVC,Truncus, PA, TA, HLHS	17.4	17.0	17.1	17.1	3 040
Left-to-right shunt	9.4	7.8	7.1	7.8	1 387
<3-month overall rate	40.2	36.1	36.1	37.0	6 567

Rates per 10 000 live births, n: number of registered patients.

**Conclusions:** PCCC-registered overall heart disease rates show no significant differences among states. Although other anomalies often present with desaturation, only 46% of heart disease registered within 3 months of life is "Oximeter-Screenable". Therefore, additional screening methods are needed to optimally manage paediatric heart disease. Incidence and timing of presentation of specific anomalies and categories from a large cohort were analysed to allow insights into detection and management of paediatric heart disease and can be used to investigate mechanisms of environmental and genetic causation.

**Abstract no: 1415**

**Diastolic filling impairment during exercise limits exercise capacity in Fontan patients**

**Alexander van de Bruaene\*, Andre la Gerche\*\*#, Guido Claessen\*, Pieter de Meester\*, Sarah Devroe‡, Jan Bogaert‡, Piet Claus§, Hein Heidbachel¶, Werner Budts\* and Marc Gewillig°**

\*Department of Cardiovascular Medicine, University Hospitals Leuven, Belgium

#University of Melbourne Department of Medicine, St Vincent's Hospital, Australia

‡Department of Anaesthesiology, University Hospitals Leuven, Belgium

‡Radiology Department, MIRC, University Hospitals Leuven, Belgium

§Laboratory of Cardiovascular Imaging and Dynamics, University of Leuven, Belgium

°Paediatric Cardiology, University Hospital Leuven, Belgium

**Background:** Better imaging techniques are needed to assess haemodynamic changes that limit exercise performance in Fontan patients. This study aimed at evaluating changes in end-diastolic and end-systolic volumes (EDV and ESV) using a novel cardiac magnetic resonance imaging (cMRI) methodology during mild, moderate and strenuous exercise.

**Methods:** Seven Fontan patients (5 male, age 19±5 years) underwent cMRI at rest and during supine exercise on a programmable cycle ergometer. Systemic ventricular volumes were obtained at rest (heart rate 77±13bpm) and during mild (105±10bpm), moderate (129±15bpm) and strenuous (144±13bpm) exercise. Images were acquired using an ungated, free-breathing real-time cMRI sequence (12 - 18 contiguous 8mm slices). Software was developed to allow for synchronisation of short and long-axis images with compensation for respiratory phase translation. Endocardial borders were delineated using a bi-plane model. Simultaneously, pulmonary artery pressures were measured during exercise using a fluid-filled catheter.

**Results:** Cardiac output (CO) increased continuously during exercise (6.6±1.9 vs. 9.4±1.8 vs. 11.1±3.5 vs. 11.5±3.4l/min; p<0.0001). The increase in CO depended on a 94±40% increase in heart rate. Stroke volume (SV) did not change from rest to mild exercise (87±22 vs. 90±20ml; p=0.458) and decreased during moderate and strenuous exercise (90±20 vs. 85±22 vs. 79±17ml; p<0.0001). EDV increased from rest to mild exercise (162±39 vs. 170±43ml; p=0.040), but decreased during moderate and strenuous exercise (170±43 vs. 164±47 vs. 158±45ml; p=0.004) whereas ESV did not change during exercise (74±28 vs. 80±32 vs. 78±35 vs. 80±38ml; p=NS). Mean pulmonary artery pressures increased during exercise (10±4 vs. 14±4 vs. 18±4 vs. 22±5mmHg; p<0.0001).

**Conclusions:** In Fontan patients, CO augmentation during exercise is predominantly dependent upon increasing heart rate. Impaired ventricular filling causes a decrease in SV already evident at moderate exercise, suggesting that decreased preload reserve is an important determinant limiting exercise capacity.

**Abstract no: 1417**

**Population study of 332 consecutive newborns with hypoplastic left heart syndrome (HLHS): A single centre experience**

**Anna Mazurek-Kula, Jadwiga A. Moll, Jacek J. Moll and Andrzej Sysa**

Polish Mother's Memorial Hospital, Lodz, Poland

**Background:** Our hospital is one of the leading centres for HLHS treatment in Poland.

**Aim:** The aim of the study was the estimation of peri-natal and anatomic data of consecutive newborns with HLHS operated at our institution.

**Material and methods:** Retrospective analysis of peri-natal data and anatomical findings of 332 newborns with HLHS [226 boys (68%) and 106 girls (32%)] treated by staged Norwood operation at our institution from 1992 - 2011.

**Results:** Pre-natal diagnosis was made in 58% of patients (68 - 75% in last 5 years), mean pregnancy duration was  $39.4 \pm 1.8$  (31 - 43) weeks; mean mother's age was  $26 \pm 5$  (17 - 43) years, in 147 cases (44%) child with HLHS came from the 1st pregnancy. Mean Apgar score was  $8.7 \pm 1.4$  (1 - 10), mean birth mass was  $3236 \pm 505.6$ g (1995 - 4430g). Ten patients (3%) came from twin pregnancies. 5 patients (1.5%) had coexistent extracardiac malformations. In 4 cases (1.2%) our patient was the 2nd child with HLHS of the same parents. In 6 families (1.8%) cardiac or extracardiac malformations in HLHS patient's siblings were confirmed (in 2 families TGA in sibling). In 1 patient Turner syndrome (45.X) was diagnosed. Anatomic subtypes of HLHS: MA/AA in 123 (37%); MS/AA in 113 (34%); MS/AS in 93 (28%); and MA/AS in 3 (1%) patients. Right ventricle myocardial performance index (RV-MPI)  $0.521 \pm 0.18$  (0.2-0.968) vs.  $0.3 \pm 0.078$  (0.183-0.445) in control group of 50 healthy newborns. Restrictive atrial communication was confirmed in 33 patients (10%). Mean ascending aorta diameter was 3.8mm (1 - 7.5mm). Severe tricuspid regurgitation was diagnosed in 40 patients (12%).

**Conclusions:** Patients with HLHS is usually male, good developed, full term delivery newborn of a young mother. HLHS rarely coexists with other malformations or genetic disorders. In our material MA/AA, MS/AA, MS/AS subtypes occurred with similar frequency, MA/AS was very rare. RV-MPI for HLHS patients is significantly higher comparing with healthy neonates.

#### Abstract no: 1421

### Association of temporary complete AV block and junctional ectopic tachycardia after surgery for congenital heart disease

**Christian Paech\***, **Patrick Flosdorff\***, **Ingo Dahnert\***, **Martin Kostelka#**, **Meinhardt Mende†** and **Roman Antonin Gebauer\***

\*Department of Paediatric Cardiology, Heart Centre, University of Leipzig, Germany

#Departement of Cardiac Surgery, Heart Centre, University of Leipzig, Germany

†Insitute for Medical Biometry, University of Leipzig, Germany

Junctional ectopic tachycardia (JET) is a postoperative complication with a mortality rate of up to 14% in patients after surgery for congenital heart disease. This study evaluated the association of intra- and / or postoperative temporary third degree AV block and the occurrence of junctional ectopic tachycardia.

**Methods:** Data was collected retrospectively from 1 158 consecutive patients who underwent surgery for congenital heart disease in the period of 2006 - 2010.

**Results:** The overall JET incidence was 2.8%. Temporary third degree AV block occurred in 2.8% of cases. Onlasting third degree AV block requiring pacemaker implantation occurred in 1.2% of cases. Post-operative JET was significantly correlated to temporary AV block ( $p < 0.001$ ) occurring in 56% of patients with temporary AV block. No case of postoperative JET was reported in a patient with onlasting AV block. Furthermore bivariat regression analysis showed a statistically significant correlation between postoperative JET and age at operation ( $p < 0.001$ ), cardiopulmonary bypass time ( $p < 0.001$ ) and aortic cross clamping time ( $p < 0.004$ ).

**Conclusion:** There is an association of temporary complete AV block and postoperative JET. In addition onlasting complete AV block seems to be negatively correlated to postoperative JET.

#### Abstract no: 1426

### Quality of life in families where a child had staged treatment for hypoplastic left heart syndrome (HLHS)

**Anna Mazurek-Kula**, **Barbara Wisniewska**, **Katarzyna Ostrowska**, **Tomasz Moszura**, **Pawel Dryzek**, **Jadwiga A. Moll**, **Andrzej Sysa** and **Jacek J. Moll**

Polish Mother's Memorial Hospital, Lodz, Poland

**Background:** In spite of increasing number of survivors with HLHS after staged Norwood operation, data about quality of life in this group of patients and their families is limited.

**Aim:** To assess the quality of life of children with HLHS and their families.

**Methods:** Mothers of 54 children with HLHS completed the questionnaire to assess subjectively quality of their children's life and the impact of child's illness on the family. All children (age 4 - 16 years) were operated at our institution. In 35% child with HLHS was the only child in family.

**Results:** Problems in physical activity in HLHS patients were reported in 79%, emotional problems in 25%, educational in 9%. Development estimated as normal was reported in 89% of patients. 79% of patients attend normal schools or kindergartens. Good tolerance of frequent hospitalisations was reported in 75% of cases. Child illness is connected with strong parental stress (73%), and negative emotions like sadness (41%), fear and helplessness (42%). Own family support, support groups of parents and religious faith were considered as most helpful. Only 13% of mothers looked for professional psychological care. 94% of responders assessed familial atmosphere as good, in 67% child's illness strengthened parental marriage. Impact of child's illness on family material situation was assessed as significantly negative in 79%. In 59% of families the father is the only working parent.

**Conclusion:** Patients with HLHS are active members of society; they attend normal schools and kindergartens although their physical activity is limited. The family functions well, but the child's illness is a reason for strong parental stress and indicates material problems. An increase in the number of HLHS survivors indicates the need for continued studies concerning neurodevelopmental outcomes, quality of life and family functioning in this group of patients.

**Abstract no: I427****Pediatric cardiac intervention programme in upper Egypt: More benefits and fewer risks****Alaa Roushdy\*<sup>#</sup>, Maiy El sayed\*<sup>#</sup>, Mohamed Gazi\*<sup>#</sup>, Sherien Abdelsalam\* and Magdi Yacoub\*<sup>†</sup>**

\*Aswan Heart Center, Magdi Yacoub Heart Foundation, Aswan, Egypt

<sup>#</sup>Cardiology Department, Ain Shams University, Cairo, Egypt<sup>†</sup>Cariothoracic surgery, Harefield Heart Science Centre, London United Kingdom

**Background/hypothesis:** The high prevalence of congenital heart disease among developing nations due to their high birth rate and the highly qualified medical staff needed to deal with such patients makes the development of a sustainable cardiac centers for the treatment of children and young people with heart diseases, in countries where the facilities for such treatments are unavailable a top priority.

**Patients and methods:** The study included all children suffering from congenital heart disease who were referred for elective percutaneous cardiac intervention in Aswan Heart Center over a period of 6 months. Patients and procedural data were recorded including age, sex, weight, height, underlying heart disease, type of the procedure, procedure duration, outcome as well as any intra or post procedural complications and the severity level of these complications.

**Results:** Eighty four children were included in the study; their mean age was 4.4±4.3 years; the youngest patient was 2 months old while the oldest was 18 years old. Patients underwent a wide range of percutaneous interventional procedures most of which were balloon pulmonary valvuloplasty (n=18), ASD device closure (n=18), PDA device closure (n=2). Five patients underwent 2 interventional procedures in the same setting. The mean procedure duration was 52.7±32.7 minutes and the maximum hospitalisation period was less than 48 hours. The success rate was 97.6% (n=82) and the mortality rate was 0%. Periprocedural complications occurred in 19 patients, none of which were life threatening and the majority were self limiting intra procedural arrhythmias

**Conclusion:** The development of a specialised paediatric cardiac interventional center in remote area that lacks this service is very cost effective. Such a center will not only provide excellent medical service to children in need but will also unload the available surgical theaters allowing these centers to deal with more complex and challenging cases.

**Abstract no: I430****Quality of care and outcomes among children undergoing cardiac catheterisation: A charity based programme in Upper Egypt****Maiy El Sayed\*<sup>#</sup>, Alaa Roushdy\*<sup>#</sup>, Mohamed Gazi\*<sup>#</sup>, Sherien Abdelsalam\*, Baher Matta Hanna\*, Joaquim Miro\*<sup>†</sup>, and Magdi Yacoub\*<sup>‡</sup>**

\*Aswan Hear Centre, Magdi Yacoub Heart Foundation, Aswan, Egypt

<sup>#</sup>Cardiology Department, Ain shams University, Cairo, Egypt<sup>†</sup>Centre Hospitalier Universitaire, Sainte-Justine, Montreal, Quebec, Canada<sup>‡</sup>Cadiothoracic Surgery, Harefield Heart Science Centre, London, United Kingdom

**Background/hypothesis:** The inhabitants of Aswan and neighboring Upper Egypt amount to approximately 24 million people. Currently there are no local specialised cardiac services for this large population. This tremendous need motivated Professor Yacoub and Chain of Hope teams to stimulate the development of a new unit to offer cardiac facilities at the highest level.

**Patients and methods:** All children with congenital heart disease who were referred for elective cardiac catheterisation in Aswan Heart Center over a period of 2 years were subjected to a baseline 2D echocardiogram and a follow up echocardiogram after device closure or valvuloplasty. Percutaneous cardiac interventions were done by a pediatric cardiologist with a minimum of 5 years experience. A dedicated paediatric anesthetist was responsible for the general anesthesia/deep sedation throughout the procedure. The patients were admitted in a specialised post catheterisation care unit for 24 - 48 hours post procedure. All demographic data as well as periprocedural hospital period including the procedural outcome and any complications were recorded and analysed.

**Results:** The study included 214 patients; 146 patients underwent 157 interventional procedures while the remaining 68 underwent diagnostic catheterisation. The interventional procedures included, ASD device closure (n=50), PDA device closure (n=42), valvuloplasty (n=38), VSD device closure (n=4) and coarctation stent (n=8). The mean age was 4.7±4.1 years; the youngest patient was 2 months old while the oldest was 18 years old. The success rate of the interventional procedures was 96.6% (n=141) and the mortality rate was 0%.

**Conclusion:** Establishing a specialised pediatric cardiac catheterisation center in a remote area with results comparable to highly qualified international centers is feasible with the help of experienced local and international teams as long as a continuous funding and development plans can sustain such centers.

**Abstract no: I447****The use of implantable cardioverter defibrillators in paediatric patients****Ana Maria Rocha Pinto E Silva\*, Valquiria P. Campagnucci\*, Wilson L. Pereira\*, Alexey P. Peroni\*, Eduardo G. Chamlian\*, Sylvio M.A. Gandra#, Argemiro Scatolini Neto#, Uelra R. Lourenaso#, Barbara O. Eira# and Luis A. Rivetti\***

\*Heart Surgery Unit, Faculty of Medical Sciences, Santa Casa de São Paulo Hospital, São Paulo, Brazil

<sup>#</sup>Arrhythmia Unit, Faculty of Medical Sciences, Santa Casa de São Paulo Hospital, São Paulo, Brazil

**Background:** The implant of devices in children is always a challenge. Implantable Cardioverter Defibrillator (ICD) therapy has been indicated in 14 children from March 2003 - June 2012 at Santa Casa de Sao Paulo Hospital. Ages ranged between 8 and 16 year old.

**Aim:** To analyse all pathologies, techniques, medical treatment and events related to these children.

**Methods and material:** The following diagnoses were observed: Long QT syndrome (n=2), hypertrophic cardiomyopathy (n=3), Brugada syndrome (n=1), LV non-compaction (n=1), congenital heart disease post-operative (n=3), dilated cardiomyopathy (n=1), catecholaminergic ventricular tachycardia (n=1), rhythmogenic right ventricular tachycardia (n=1) and Idiopathic VT (n=1). Syncope (n=4), ventricular tachycardia (n=6), or recovery from sudden death (n=4) was the indication for ICD. In all cases an endovascular endocardial approach for implanting ICDs was used. The prostheses were located below the left pectoralis major muscle, in 12 patients and below the rectus abdominis muscle in 2 patients. The defibrillation threshold has been distributed as follows: 15J (n=1), 20J (n=11), and 36J (n=2).

**Results:** The children have been followed from 1 month up - 9 years. Each patient received pharmacological treatment for the arrhythmias with specific drugs. Seven patients had no events. Inappropriate shocks occurred in 6 patients. Three of them needed ablation due to atrial tachycardia. One patient had appropriate shocks. Two patients had lead dysfunction and needed replacement.

**Conclusion:** Cardioverter defibrillator implantation was successfully done by endovascular approach in our paediatric patients. The follow-up of this group has showed that ICDs are the solution for those children with tachyarrhythmia whose medical treatment failed.

#### Abstract no: 1449

### Ante-natal diagnosis of congenital heart disease in Nova Scotia: A 20-year retrospective study on survival and surgical outcomes

Paul D'Alessandro\* and Krista Jangaard\*#

\*Dalhousie University, Halifax, Nova Scotia, Canada

#IWK Health Centre, Halifax, Nova Scotia, Canada

**Background/hypothesis:** Effective ante-natal diagnosis of congenital heart disease can improve outcomes and survival in certain conditions. We hypothesised high, moderate, and low detection rates for HLHS, TOF, and TGA respectively.

**Aim:** To examine ante-natal detection rates for hypoplastic left heart syndrome (HLHS), transposition of the great arteries (TGA), and Tetralogy of Fallot (ToF) among live births at our tertiary care centre; and measure outcomes in infants with ante-natal diagnoses.

**Materials and methods:** Charts for live births from July 1989 - December 2010 were identified from the Nova Scotia Atlee Peri-natal Database and reviewed retrospectively. Stillbirths and infants with extra-cardiac abnormalities were excluded.

**Results:** Of the 215 618 live births in Nova Scotia during this time, 23 infants were born with HLHS, 25 with TGA, and 40 with ToF. The rates of ante-natal diagnosis of HLHS, ToF, and TGA among live births were 57.7%, 60.0%, and 68.9%, respectively. The number of HLHS live births declined to zero by 2008. One year survival for all infants born with HLHS was 0, regardless of timing of diagnosis. A pre-natal diagnosis of ToF was not associated with increased survival or decreased morbidity. A pre-natal diagnosis of TGA was associated with significantly shorter time to NICU admission (1.1h vs. 29.5h,  $p < 0.03$ ). There were no significant differences between maternal or neonatal factors between cohorts. The commonest morbidities at most recent follow-up were ADHD and behavioural difficulties.

**Conclusions:** The low incidence of ante-natal HLHS diagnosis with a decline in HLHS live births suggests an increase in terminations. Incidence and benefit of ante-natal diagnosis of ToF remain moderate. Preliminary data suggests that ante-natally diagnosed infants born with TGA receive definitive management faster. Further review of TGA outcomes is pending. The notable incidence of ADHD could reflect latent hypoxic sequelae or chronic disease-related psychosocial issues.

#### Abstract no: 1451

### Donor-recipient size matching in paediatric heart transplantation: Is weight the most appropriate parameter to predict outcomes in all age groups?

Ahmed Bayoumi, Hua Liu and Francis Fynn-Thompson

Department of Cardiac Surgery, Boston Children's Hospital, Massachusetts, United States of America

**Background:** The aim was to retrospectively review our practice of donor-recipient (D/R) size matching by weight criteria alone and simultaneously reassess and compare post-transplant outcomes using the additional variables of height and body-surface area (BSA).

**Methods:** We retrospectively reviewed the medical records of 211 patients who underwent orthotopic heart transplantation at Boston Children's Hospital over the past 20 years. The patients were divided into the following age groups: <1 year ( $n=30$ ), 1 - 12 years ( $n=107$ ) and >12 years ( $n=74$ ). Donor/recipient weight, height and BSA ratios were determined for each age group at the date of transplantation. Outliers were identified for each variable and analysed separately. We compared the average and median of continuous outcomes such as length of stays, need for open chest, post-transplant filling pressures and survival.

**Results:** In our cohort the median D/R weight ratio was 1.276 (0.2 - 5.0), BSA ratio 1.172 (0.2 - 2.0) and height ratio 1.061 (0.3 - 2.0). There was no difference in post-transplant survival when comparing outcomes by weight, BSA or height ratios. Increased D/R size ratio was associated with significantly increased likelihood of delayed chest closure, ICU and total hospital length of stay in all ages. The odds ratio was highest for the youngest age group with disproportionate number of outliers (33.3 %) but this effect was less significant when D/R matching was done by BSA. The younger patients were more likely to receive size-mismatched hearts than older recipients.

**Conclusions:** The current practice of using weight for donor-recipient size matching in paediatric heart transplantation does not seem to impact long-term outcomes. In younger patients, an increase in donor to recipient size ratio increases post-operative recovery. However, the use of BSA is a more reliable tool for matching heart sizes in all age groups.

#### Abstract no: 1453

### Clinical and surgical evolution of tricuspid atresia at a paediatric hospital in Argentina

Ana Maria Susana De Dios\*, Fernanda Biancolini\*, Julio Cesar Biancolini#, Jesus Damsky Barbosa<sup>†</sup>, Miguel Granja<sup>†</sup> and Jose Vargas<sup>‡</sup>

\*Paediatric Cardiology, Pedro de Elizalde Children's Hospital, Argentina

#Paediatrics, Pedro de Elizalde Children's Hospital, Argentina

<sup>†</sup>Cath Lab, Argentina

<sup>‡</sup>Cardiovascular Surgery, Pedro de Elizalde Children's Hospital, Argentina

**Purpose:** Follow-up of patients (p) with tricuspid atresia (TA).

**Methods:** Retrospective review of 45 clinical records of patients with TA that were assessed at Pedro de Elizalde Children Hospital from August 2005 - August 2012. Grouped according to EDWARDS and BURCHEL classification. Analysis based on clinical exam, radiology (Rx), electrocardiography (EKG), echocardiography, catheterisation records, and surgical treatment.

Statistical analysis: multivariable test  $p < 0.05$ .

**Results:** Forty four patients had TA; 44 situs solitus (SS); and 1 was indeterminate (Si).

**Type I:** Forty three patients ventricle-arterial concordance (95.5%).

- IA: 3 patients had decreased pulmonary flow (PQ) (7%). Treatment: 2 systemic pulmonary anastomosis (SPA) and 1 Glenn shunt. Complication: 1p stenosis left pulmonary branch, 1p needed occlusion of inferior vena cava.
- IB (58%): 24 patients normal PQ. Treatment: 2 Park Septectomy, 14 SPA, 5 Glenn shunt, 1p Glenn pulsatile, 3 atrio-pulmonary connection, 3 extracardiac cavopulmonary connection. Complications: 1 needed mitral anuloplasty; 1 endocarditis; 1 AV dissociation, required a pacemaker; 1 W.P. White.
- IC (32%): 12 patients increase PQ. Diagnosis: all 1st month: 4 with pulmonary hypertension. Treatment: 3 SPA; later: 2 Glenn, 1 Glenn pulsatile, 2 Atrio-pulmonary connection. Complications: fibrillo-flutter 4 years later in 1, AV block in 1.

**Type II:** Ventricle-arterial discordance (5%).

- IIA: 2 patients, decrease PQ, Si, RBVF, severe pulmonary stenosis and mitral insufficiency. Treatment: 1 SPA that needed stent in pulmonary artery branch, Glenn dysfunction.

**Type III:** Left sided right ventricle (I-loop).

- No patients. Follow-up only in 23 patients.

**Conclusion:** Type I was the most frequent (95%) [IB (56%) and IC (33%)]. Cyanosis was the most common clinical manifestation (66%). EKG: superior axis was found in 19/45 patients, left ventricle hypertrophy in 82.9% and left anterior hemi-block in 70% of the patients. Regarding surgical procedure: 21 patients needed systemic pulmonary anastomosis; 12 Glenn shunt (9 non-pulsatile, 3 pulsatile); 6 atrio-pulmonary connection; and 4 extracardiac tube connection.

**Abstract no: I461**

**Deteriorating paediatric cardiac ward patients: A 12-month review**

*Kim Trussler and Mary Wilson*

Mater Children's Hospital, Australia

**Background:** To identify deteriorating patients and to describe characteristics of events and mortality.

**Methods:** We retrospectively analysed data of all children from our paediatric cardiac ward requiring secondary emergency response team (SERT) attendance, and/or unplanned Paediatric Intensive Care Unit (PICU) admission from January - December 2010. Data were extracted from medical charts and the ward's clinical information system.

**Results:** Thirty six events were observed in 28 patients. Sixteen (44%) were SERT calls, which included 4 (11%) episodes of CPR, and 20 (56%) separate PICU reviews. Overall 27 (75%) required transfer to PICU. Most common reasons for triggering SERT calls or PICU transfer were cyanotic episodes (28%), low cardiac output state (23%), bradycardia (11%) and arrhythmia (SVT/VT, 11%). There was a statistical significance between cause of deterioration and whether the child was categorised as either medical or surgical  $p < .05$ . Of the children that deteriorated 86% were aged <12 months. Medical children were the most likely to deteriorate (57%) and it was more likely to occur in the close observation unit within the ward (69%) rather than in the general ward area. A slow deterioration as opposed to an acute deterioration occurred in 64% of cases. Timing of deterioration varied with 18% occurring within 24 hours of PICU transfer and 36% within 72 hours. More events (72%) occurred outside of office hours. Of the 28 patients there was a 24% (n=) mortality rate.

**Conclusions:** Deteriorating cardiac patients on the paediatric cardiac ward carry a high mortality long term. The implementation of a clinical early warning tool, specifically designed for the cardiac ward patient, may be more efficient in detecting the deteriorating patient.

**Abstract no: I468**

**Longevity of palliation provided by neonatal ductal stenting before definitive surgical correction for congenital heart diseases with duct-dependent pulmonary circulation**

*Sivakumar Kothandam, Anilkumar Singhi, Sreeja Pavithran, Anpon Bhagyavathy and Robert Coelho*

MIOT Hospital, Chennai, India

**Background:** Ductal stenting (DS) in duct-dependent pulmonary circulation is less morbid than neonatal Blalock-Taussig shunt. We aim to study if DS provided adequately long palliation.

**Methods:** A prospective interventional clinical follow-up study of consecutive infants followed after successful DS was done from 2 tertiary referral centres. Patients were divided into 3 anatomic groups; A: following pulmonary valvotomy for critical pulmonary stenosis and atresia, for continuing duct dependency; B: Tetralogy of Fallot with pulmonary atresia where palliation is needed till conduit replacement; and C: univentricular hearts where ductal stent is needed till bidirectional Glenn shunt surgery. Interstage mortality was studied.

**Results:** Among a total of 24 infants, 4 patients in group A followed for 26 - 54 months had adequate oxygen saturations and no residual gradient. Tricuspid valve, pulmonary annulus and right ventricle grew with age. After discontinuing anti-platelets, 2 stents were patent after 1 year. In 8/11 group B patients, corrective biventricular repair using conduits was done after 5 - 14 months (body weight 5 - 7.5kg). One patient awaits conduit repair. Bidirectional Glenn shunt and confluence repair was done in 7/9 group C patients after 8 - 15 months (weight 6 - 8.5kg). The hilar pulmonary artery growths in B and C groups were adequate for surgical repair. No patient needed stent re-dilatations or additional shunts. Four patients had sudden death at home and no autopsy was done.

**Conclusions:** Duration of palliation by DS is sufficient to allow adequate somatic growth before next surgery. DS is acceptable palliation after pulmonary valvotomy and univentricular hearts where short term patency is sufficient. In patients needing biventricular repair with conduits, a longer term of palliation may be desirable and ductal stent does not provide adequate pulmonary flows for a very long time. There is an interstage mortality of 16% of unknown causes after DS.

**Abstract no: I472****cMRI as a non-invasive definitive diagnostic tool for complex cyanotic congenital heart disease**

**Ramyashri Chandrasekaran\***, **Manokar Panchatcharam\***, **Muthukumar Sivasubramaniam\***, **Periyasamy Thangavel\***, **Jebaraj Rathinasamy\***, **Asha Moorthy\***, **Muralidharan Thoddi Ramamurthy\***, **Murthy Jayanti Sathyanarayana\*** and **Thanikachalam Sadagopan\***

\*Department of Cardiology, Sri Ramachandra Medical College, Chennai, India

#Department of Paediatrics, Sri Ramachandra Medical College, Chennai, India

**Aim:** To study the feasibility of cMRI as a useful advanced diagnostic tool in evaluation of CCHD in our institute from June 2011 - May 2012.

**Material and methods:** Twelve Patients with CCHD in the age group 8 - 50, female: male ratio 1:1, all these patients had through clinical examination along with 2-D Echo evaluation. cMRI was the final decision-making tool for all these patients. None of them had CT/ cardiac cath for delineation of anatomy. After confirmation of diagnosis 9/12 patients underwent surgical correction. Two patients refused surgery; 1-DORV with severe PAH was inoperable.

**Results:** Out of our 12 patients we had: Tetralogy of Fallot (3), DORV in its various forms (4), CCTGA (2), unbalanced AV canal defect with reduced pulmonary flow (1), DILV L-TGA with PS (1), Tricuspid Atresia with failed Glenn (1). Two patients with ToF had total correction, 1 DORV with pulmonary atresia, MAPCAS had unifocalistaion with central shunt, 1 DORV was identified with non-confluent PAS had total correction, 1 DORV with PS with non-routable VSD had a Fontan, DILV & unbalanced AV canal had single-staged Fontan, TA also had a Fontan, 1-CCTGA underwent double switch surgery. All these patients who were admitted for surgery had on table assessment of PA pressures, and all of them were found to be fit for final surgery. The one whom we predicated to be not suitable for surgery was confirmed by cardiac cath.

**Conclusion:** cMRI is a dynamic modality which can give all the necessary information to help us to plan the management strategy in CCHD with decreased pulmonary blood flow, thus avoiding a need for catheterisation studies or CT angiogram in most of the complex congenital heart diseases.

**Abstract no: I475****Functional indexation of RV parameters in repaired TOF patients: A better reflection of clinical status?**

**Heynric Grotenhuis, August Oudshoorn, Cedric Manlhoit and Shi-Joon Yoo**

The Hospital for Sick Children, University of Toronto, Canada

**Background/hypothesis:** Routine cardiac magnetic resonance imaging (MRI) is a well-accepted imaging method for assessment of right ventricular (RV) and left ventricular (LV) function parameters and degree of pulmonary insufficiency after repair of Tetralogy of Fallot (TOF). Poor correlation with clinical status as expressed by New York Heart Association (NYHA) class has however been reported. We hypothesised that a combination of degree of RV dilatation [expressed by RV end-diastolic volume (RV<sub>EDV</sub>) and end-systolic volume (RV<sub>ESV</sub>)], indexed for LV function [expressed by LV ejection fraction (LV<sub>EF</sub>)] may better correlate with clinical status than these parameters individually.

**Materials and methods:** 227 repaired TOF patients (mean age 12.0 years ± standard deviation 4.6 years) were studied with routine cardiac MRI. RV<sub>EDV</sub> and RV<sub>ESV</sub> were indexed for body surface area (1) and LV<sub>EF</sub> (2), respectively. Clinical status as expressed by NYHA class was derived from clinic visit at around the time of MRI. A univariable model was used for statistical analysis.

**Results:** RV volumes (mean ± standard deviation) were: RV<sub>EDV</sub><sup>1</sup>: 144.96±38.49ml/m<sup>2</sup>, RV<sub>EDV</sub><sup>2</sup>: 2.60±0.81, RV<sub>ESV</sub><sup>1</sup>: 76.74±28.25ml/m<sup>2</sup>, RV<sub>ESV</sub><sup>2</sup>: 1.39±0.59. Other parameters were: LV<sub>EF</sub>: 56.81±6.18%, RV<sub>EF</sub>: 48.07±9.24%, LV<sub>stroke volume</sub>: 46.86±9.03ml, RV<sub>stroke volume</sub>: 68.09±17.67ml, LV<sub>EDV</sub><sup>1</sup>: 84.28±19.52ml/m<sup>2</sup>, LV<sub>ESV</sub><sup>1</sup>: 36.35±9.71ml/m<sup>2</sup>, pulmonary regurgitation fraction (PRF): 32.51±13.61%. RV<sub>ESV</sub><sup>2</sup> and RV<sub>EDV</sub><sup>2</sup> showed much higher correlation (0.0002 and 0.007, respectively) with a higher NYHA score than RV<sub>ESV</sub><sup>1</sup> and RV<sub>EDV</sub><sup>1</sup> (0.80 and 0.03, respectively), or LV<sub>EF</sub> (0.43), RV<sub>EF</sub> (0.05) and PRF (0.11).

**Conclusions:** RV volumes indexed for LV<sub>EF</sub> show a much higher correlation to clinical status than all routinely used RV and LV parameters as indexed by BSA. RV volume indexation for LV<sub>EF</sub> that combines degree of RV dilatation and LV (dys-)function may therefore be more useful in clinical follow-up of TOF patients.

**Abstract no: I477****Heart rate corrected pulmonary artery acceleration time correlates with pulmonary capacitance and RV stroke work in children with pulmonary hypertensive vascular disease: A simultaneous echocardiographic and cardiac catheterisation study**

**Prashant Bobhate, Tim Colen, S. Kaneko, Jean Trines, Nee Khoo and Ian Adatia**

Stollery Childrens Hospital, University of Alberta, Edmonton, Alberta, Canada

**Background:** Pulmonary capacitance calculated at cardiac catheterisation is a strong independent predictor of mortality in pulmonary hypertensive vascular disease (PHVD). However cardiac catheterisation is invasive, risky and unsuited for frequent assessments. There is a pressing need for non-invasive correlates of catheter-obtained haemodynamic parameters. We sought to investigate relationships between Doppler-derived indices and invasively obtained haemodynamic measurements.

**Methods:** We performed transthoracic echocardiograms with Doppler interrogation on children undergoing cardiac catheterisation with PHVD after induction of anaesthesia. We measured tricuspid regurgitation velocity (TRV) and heart rate corrected pulmonary artery acceleration time (PAAT). From cardiac catheterisation data we calculated pulmonary capacitance index (PCI) and right ventricular stroke work index (RVSWI).

**Results:** We studied 17 consecutive patients (11 males, median age 6 years, range 0.4 - 15). Mean PA pressures were 40±20mmHg and mean Rp was 9.9±6WU/m<sup>2</sup>. Peak TR velocity correlated with systolic pulmonary artery pressures (PAp) (r=0.79, p<0.01). Heart rate corrected PAAT correlated negatively with PCI (r=-0.58, p=0.03) and RVSWI (r=-0.68, p=0.01).

**Conclusion:** Increased heart rate corrected PAAT was associated with reduced PCI and RVSWI, both measures of the efficiency of the RV performance and pulmonary artery-right ventricular coupling. Heart rate corrected PAAT is measured non-invasively by Doppler. Heart rate corrected PAAT is easy to obtain, reproducible and may prove useful in the assessment of children with PHVD.

**Abstract no: I481**

**Pulmonary vein stenosis in formerly premature infants**

**Linda Mahgoub\*, Ashok Kakadekar#, Frank Dicke†, Jeffrey Fineman‡, Andrew Redington§ and Ian Adatia\***

\*Stollery Children's Hospital, University of Alberta, Edmonton, Canada

#Royal University Hospital, Saskatoon, Canada

†Alberta Children's Hospital, Calgary, Canada

‡San Francisco Children's Hospital, University of California, San Francisco, United States of America

§Hospital for Sick Children, Toronto, Canada

**Background:** Pulmonary vein stenosis (PVS) is a rare disorder that may occur as an isolated lesion or in association with congenital heart defects. It can be acquired following cardiac surgery or interventions around the pulmonary veins. However, a group of ex-premature infants who develop PVS has been identified, although, the etiology and association with prematurity remains poorly understood.

**Methods:** We reviewed all available clinical and laboratory data in patients with a diagnosis of pulmonary vein stenosis. We excluded patients with total anomalous pulmonary venous drainage, atrial isomerism and gestational age  $\leq 35$  weeks.

**Results:** We identified 11 patients with pulmonary vein stenosis, 8 were male, median gestational age was 27 weeks (25 weeks - 34 weeks), median birth weight was 860g (432g - 2100g), and 3 patients were of twin pregnancies whose twin siblings were unaffected. Most patients were diagnosed with chronic lung disease and needed significant respiratory support after birth. In 9/11 initial neonatal echocardiograms did not report abnormal pulmonary vein flow. The median age at diagnosis was 5 months (3 months - 2 years) the diagnosis was most often made by or suspected by echocardiography because of apparent worsening of chronic lung disease. 8/11 patients underwent CT scan or MRI. The left pulmonary vein was the most commonly stenosed (91%) and all patients underwent a suture-less surgical repair. Median survival after pulmonary vein surgery was 6 months (4 months - 10 months). In 7/11 PVS recurred and 4/11 patients died.

**Conclusion:** Pulmonary vein stenosis should be considered if an ex-premature baby has late deterioration of chronic lung disease or evidence of pulmonary hypertension by echocardiogram. Further imaging by CT scan or MRI may be required to complete the diagnosis. Pulmonary vein stenosis appears to develop post-natally but the aetiology remains unknown. The response to surgery and late recurrence appear to be similar to pulmonary vein stenosis in infants born at term.

**Abstract no: I483**

**Serial measures of systemic to pulmonary arterial collateral flow in patients with superior and total cavopulmonary connections**

**Kevin Whitehead\*, Matthew Harris\*, Andrew Glatz\*, Yoav Dori\*, Marc Keller#, Jonathan Rome\* and Mark Fogel\***

\*Division of Cardiology, Children's Hospital of Philadelphia, United States of America

#Department of Radiology, Children's Hospital of Philadelphia, United States of America

**Background:** We have described a method of quantifying systemic to pulmonary collateral (CollF) flow in patients with superior (SCPC) and total (TCPC) cavopulmonary connections using magnetic resonance phase contrast velocity mapping (PC-MRI). Cross-sectional data suggests that CollF decreases with time after TCPC completion, but may take years. We wished to examine more acute changes in physiology and CollF from SCPC to TCPC by comparing serial (paired) PC-MRI data.

**Methods:** We retrospectively reviewed PC-MRI data to identify patients who had CollF quantified at both SCPC and TCPC. CollF, systemic blood flow ( $Q_s$  = total caval flow), and pulmonary blood flow ( $Q_p$  = total pulmonary vein flow) were quantified as previously described. Fenestration flow was quantified in TCPC by the difference in caval and pulmonary artery flow.

**Results:** Of 104 SCPC and 113 TCPC CollF studies, 26 patients had both SCPC and TCPC studies performed on average 5 months before and 7 months after TCPC completion. Indexed CollF was  $1.4 \pm 0.7$  L/min/m<sup>2</sup> at SCPC and  $1.7 \pm 1.1$  L/min/m<sup>2</sup> at TCPC ( $p=0.12$ ), which was a significantly higher fraction of aortic flow in TCPC compared to SCPC ( $37 \pm 18\%$  vs.  $31 \pm 14\%$ ,  $p=0.03$ ). There was a significant decrease in  $Q_s$  from  $3.2 \pm 0.7$  L/min/m<sup>2</sup> at SCPC to  $2.8 \pm 0.8$  L/min/m<sup>2</sup> at TCPC ( $p=0.004$ ).  $Q_p$  increases significantly, primarily as a result of increased cavopulmonary flow. Fenestration flow averaged  $48 \pm 42\%$  of the inferior vena caval (IVC) flow at TCPC, with a significant decrease in right to left shunt from SCPC to TCPC. However, in 4 patients fenestration flow equaled or exceeded IVC flow with flow reversal in the Fontan baffle above the fenestration in 2 patients.

	SCPC	TCPC	p-value
n	26	26	
Age (years)	2.9±1.2	3.9±1.2	
Time from TCPC (years)	-0.40±0.4	0.63±0.3	
$Q_{Ao}$ (L/min/m <sup>2</sup> )	4.6±0.7	4.4±1.0	p=0.2
$Q_{coll}$ (L/min/m <sup>2</sup> )	1.4±0.7	1.7±1.1	p=0.12
$Q_{coll}/Q_{Ao} \times 100$ (%)	31±14	37±18	p=0.03
$Q_{coll}/Q_{PV} \times 100$ (%)	45±18	41±20	p=0.22
$Q_p$ (L/min/m <sup>2</sup> )	3.2±0.7	4.0±1.0	p=0.001
$Q_s$ (L/min/m <sup>2</sup> )	3.2±0.8	2.8±0.8	p=0.004
$Q_p / Q_s$	1.1±0.4	1.5±0.7	p<0.001
Right to Left Shunt (L/min/m <sup>2</sup> )	1.5±0.5	0.5±0.4	p<0.001

**Conclusions:** CollF does not decrease after TCPC completion, and as a fraction of aortic flow initially increases, despite a significant reduction in right to left shunt.  $Q_s$  decreases and  $Q_p$  increases significantly after TCPC completion.

**Abstract no: I486****Coronary artery dysplasia with persistent sinusoids: Rare cause of fatal myocardial infarction in an infant****Pankaj Gupta\*, Ella Sugo# and Owen Jones\***

\*Sydney Children's Hospital Network, NSW, Australia

#Prince of Wales Hospital, Randwick, NSW, Australia

**Introduction:** Myocardial infarction in infancy is a very rare and serious event. Coronary artery dysplasia with persistent sinusoids is extremely rare defect. We report on a previously healthy infant with normal heart, who developed a massive myocardial infarction and died secondary to severe coronary artery dysplasia.

**Case report:** A 2½-month-old boy was transferred to intensive care unit of our hospital with a brief history of feeding intolerance and respiratory distress leading to cardio-respiratory arrest and requiring cardiopulmonary resuscitation with inotropic infusion. Other than congenital abnormalities such as mildly dysplastic left hip, multiple cortical renal cyst, undescended left testis and Talipes left foot, he had asymptomatic post-natal course. Initial arterial blood gas showed severe lactic acidosis. Chest roentogram showed cardiomegaly with pulmonary congestion. ECG revealed persistent pulseless electrical activity. Echocardiogram demonstrated structurally normal heart with severely depressed biventricular function. Prolonged cardiopulmonary resuscitation was ceased after failure to gain any significant cardiac output and he was pronounced deceased. Microorganism investigations including bacterial culture and nucleic acid analysis for viruses were all negative. Post-mortem examination showed significantly enlarged heart, acute and sub-acute ischaemic changes, widespread myocardial fibrosis, normal coronary artery origin and course and, most importantly, persistent myocardial sinusoids. Histology confirmed marked coagulative necrosis with a-cellular fibrosis in subendocardial region of ventricles, dystrophic calcification, and persistent sinusoids. There were no inflammatory infiltrates suggestive of myocarditis. Coronary specimen revealed intimal thickening and myxoid changes, duplication and fragmentation of internal elastic lamina and hypertrophy of media smooth muscle, along with presence of myocardial bridging pattern.

**Conclusion:** Physicians and pathologists should be aware of unusual coronary artery dysplasia in managing infants with severe ventricular dysfunction.

**Abstract no: I487****Aortic disease and recurrence of congenital heart disease in 1st- and 2nd-degree relatives of patients with persistent ductus arteriosus****Regina Bokenkamp, Liesbeth de Bruijn, Janine J. Houwing-Duistermaat, Marco C. DeRuiter, Robert E. Poelmann, C. Gittenberger-de Groot and Nico A. Blom**

Leiden University Medical Centre, Leiden, The Netherlands

**Background:** Persistent ductus arteriosus (PDA) and familial thoracic aneurysm and dissection (TAAD) are genetically heterogeneous. Data on the prevalence of aortic disease including TAAD, the familial recurrence of PDA, and other congenital heart disease (CHD) provide essential information for the investigation of genetic factors of these diseases.

**Methods and results:** Retrospective patient-based study using a questionnaire assessing the prevalence of cardiovascular disease in family members of children with PDA. A thorough family history provided information on 3 generations. The data were analysed by a binominal test. The prevalence in the general population was derived from a Dutch population-based study. Over the last 30 years 320 patients >3 months with isolated PDA have been treated in our institution. Data on 175 patients and their 2 336 family members were available. The prevalence of aortic disease was higher in the study group compared to the general Dutch population. Aortic disease was reported in 4/590 (0.7%) of 1st grade relatives and 29/1 339 (2%) of 2nd grade relatives. In 2nd grade relatives the prevalence of TAAD was increased to 0.7% (9/1 339) compared to 0.016% in the general population ( $p < 0.001$ ). The recurrence rate of CHD was higher (2.7%) in 1st-degree relatives than in 2nd-degree relatives (0.79%).

**Conclusion:** These data provide evidence for a genetic association of PDA and aortic disease. The prevalence of PDA and other CHD is increased in 1st grade relatives of the index patient, suggestive of common genetic factors in the determination of CHD and aortic disease.

**Abstract no: I488****3-D en face echocardiographic reconstruction of simple and complex ventricular septal defects on right ventricular septal surface using simple bedside protocol****Sivakumar Kothandam, Anil Kumar Singhi and Sreeja Pavitran**

Paediatric Cardiology Unit, MIOT Hospitals, Chennai, India

**Background:** Increasing clinical use of three dimensional Echocardiograms (3-DE) has enabled volume rendering of ventricular septal defects (VSD) on the right ventricular septal surface to guide understanding of cardiac morphology. This will help decision-making on surgical or interventional closure. A simple 3-DE protocol to show all VSDs from right ventricular surface in a uniform format will aid understanding among all team members.

**Methods:** We studied 40 patients with simple (single or multiple VSD) and complex VSD (Fallot tetralogy, atrioventricular canal, double outlet right ventricle, transposition of great arteries and Truncus arteriosus) using 3-DE en face reconstruction. We adopted a uniform subxiphoid live 3-DE acquisition of a sector width that covers the entire interventricular septum. This acquisition gives good reproducible images in most patients <10 - 12 years using  $\times 7 - 2$  matrix probe (Philipsmedical, Andover, MA). The probe is positioned in coronal plane in subxiphoid window to display the entire ventricular septum from right ventricular inflow to outflow. Using volume rendering, an enface reconstruction of the right ventricular septal surface is presented to give anatomical location of the ventricular septal defect.

**Results:** Out of 40 patients good 3-DE acquisition was achieved in all except one with optimal images. Twenty two patients had different types of simple VSD, (single in 17 and multiple in 5), 4 patients had atrioventricular canal, 2 had tetralogy, 6 had double outlet right ventricle, 3 had truncus arteriosus and 2 had transposition of great arteries. The 3-DE reconstruction was accurate in showing the VSD location to the interventionist (2 perimembranous VSD and 2 muscular VSD) and surgeons.

**Conclusion:** A simple right ventricular septal surface en face reconstruction of VSD by bedside live 3-DE greatly enhances the understanding location of VSD and aids in planning closure.

**Abstract no: I490**

**Myocardial function following repair of anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) in children**

**Nitha Naqvi\*<sup>#</sup>, Nagarajan Muthialu\*, Sonya Babu-Narayan<sup>#</sup>, Shreesha Maiya<sup>#</sup>, Prathiba Chandershekar\*, Victor Tsang\* and Jan Marek\***

\*Great Ormond Street Hospital, London, United Kingdom

<sup>#</sup>Royal Brompton Hospital, London, United Kingdom

**Background/hypothesis:** We hypothesised that speckle tracking may improve detection of myocardial dysfunction and provide new pathophysiological insights in anomalous coronary artery from the pulmonary artery (ALCAPA).

**Method:** Echocardiography including speckle tracking was performed in 22 children with ALCAPA (8 male, median age at surgery 0.4 years; IQR:0.21-1.05) pre- and post-operatively and in 22 healthy controls. Measurements included global and segmental longitudinal, radial and circumferential peak deformation (strain) and synchronicity index (SI) defined as agreement of time to peak strain measurements between segments per subject summarised using intraclass correlation coefficient.

**Results:** Global strains were lower in unoperated patients than controls (longitudinal: -46 vs. -123;  $p < 0.001$ ; circumferential: -48 vs. -118;  $p < 0.001$ , radial: 110 vs. 357;  $p < 0.001$ ) and improved post-operatively (longitudinal: -46 pre- vs. -82 post;  $p = 0.002$ ; circumferential: -48 vs. -96;  $p = 0.012$ ; radial: 110 vs. 317;  $p = 0.001$ ). Unoperated patients with normal 2-D function ( $n = 8$ ) had significantly impaired strain. Global dyssynchrony significantly improved post-operatively (longitudinal SI 0.93 pre- vs. 0.94 post-; circumferential 0.85 pre- vs. 0.9 post-; radial 0.71 pre- vs. 0.88 post-). Global time to peak shortened (longitudinal 2.236 pre- vs. 1.589 post;  $p < 0.001$ ; circumferential 2.037 vs. 1.447;  $p = 0.005$ ; radial 2.169 vs. 1.602;  $p = 0.01$ , ms). Despite overall global improvement some abnormalities remained. Strain improved in the majority of segments but apical septal and anterolateral segments remained abnormal. Post-systolic index improved in some segments but pre-systolic stretch persisted.

**Conclusions:** Both global contractility (strain) and global synchrony (coordinated contraction) improved after repair of ALCAPA suggesting recovery of hibernating myocardium. Contractility in some segments supplied by the anomalous left coronary artery failed to improve following ALCAPA repair suggesting a degree of irreversible myocardial damage. 2-D speckle tracking identified impairment of function not revealed by standard Echocardiography.

**Abstract no: I494**

**An unusual case of coarctation of the aorta and patent ductus arteriosus**

**Daria Kulikova\*, Igor Polivenok\*, Olga Buchneva\*, Yulia Psiklova\*, Yaroslav Shafer\*, Frank Molloy<sup>#</sup>, Robert Arnold<sup>†</sup>, Christian Gilbert<sup>#</sup>, Valery Bojko\* and William Novick<sup>#†</sup>**

\*AMS of Ukraine, Kharkov, Ukraine

<sup>#</sup>International Children's Heart Foundation, Memphis, Tennessee, United States of America

<sup>†</sup>Royal Liverpool Children's Hospital, Liverpool, United Kingdom

<sup>‡</sup>Health Sciences Centre, University of Tennessee, Memphis, Tennessee, United States of America

**Background:** We describe a highly unusual variant of juxta-ductal coarctation of the aorta (CoAo) and the lessons learnt by our team.

**Case report:** 5-month-old female patient was hospitalised with bilateral pneumonia and referred for cardiac evaluation. Echo showed marked biventricular hypertrophy, CoAo gradient 45mmHg, patent ductus arteriosus (PDA) 3mm, pulmonary hypertension (PH) 75mmHg. ECG suggested biventricular hypertrophy. Cardiac catheterisation: AscAo=142/60mmHg, RV=110/10 mmHg, PA=110/50 mmHg. Thoracotomy revealed CoAo and PDA. During PDA ligation with simultaneous aortic clamping there was severe bradycardia and collapse, without signs of ischaemia on ECG. Inotropes were ineffective. Patient recovered with direct cardiac massage, declamping of the aorta and removal of PDA ligature. Our interpretation was vagal stimulation. After stabilisation, PDA ligation was repeated with resection of coarctation with end-to-end anastomosis. Further temporary haemodynamic instability occurred during chest closure but was stabilised, however 3 hours after surgery the child suddenly developed refractory cardiac arrest.

**Results:** The post mortem study showed: PDA ligated, anastomosis of the aorta end-to-end. A single coronary artery was found on the front of the pulmonary trunk in 0.7cm from the valve, which divided into anterior and posterior descending branches and circumflex branch. No coronaries were present from aorta. Right ventricular wall was 8mm and left measured 11mm.

**Discussion:** We suppose that ischaemic symptoms of anomalous origin of coronary supply were not seen in this case due to CoAo with PDA resulting in high PA pressure maintaining high coronary perfusion pressure. After correction of CoAo and PDA, PA pressure fell and led to an acute fall in coronary perfusion. Following post mortem we reviewed the pre-operative cardiac catheterisation and aortogram, which clearly showed the absence of coronary arteries from the aorta – not seen by our team focusing on the aortic arch and ductus.

**Abstract no: I495**

**Comparative effectiveness among different immunoglobulin for Kawasaki disease**

**Ming-Chih Lin\*<sup>#†</sup>, Mei-Shu Lai<sup>#</sup>, Yun-Ching Fu\*<sup>†</sup> and Sheng-Ling Jan\*<sup>†</sup>**

\*Taichung Veterans General Hospital, Taichung, Taiwan

<sup>#</sup>School of Medicine, National Yang-Ming University, Taipei, Taiwan

<sup>†</sup>Institute of Epidemiology and Preventive Medicine, National Taiwan University, Taiwan

**Background:** Kawasaki disease is the leading cause of acquired heart disease among children in most industrialised countries. Immunoglobulin is used to treat Kawasaki disease, but few studies have ever evaluated the comparative effectiveness of immunoglobulin from different manufacturing processes. Moreover, those studies were limited by small case numbers and lack of longitudinal follow-up.

**Aim:** To evaluate the comparative effectiveness of immunoglobulin from different manufacturing processes.

**Material and methods:** The data come from National Health Insurance Research Database of Taiwan. From 1997 - 2008, patients admitted diagnosed with Kawasaki disease and receiving immunoglobulin therapies for the 1st time were enrolled in the research cohort. The immunoglobulin manufacturing process was

divided into  $\beta$ -propiolactation, acidification and containing IgA. The comparative effectiveness was evaluated by portion of treatment failure, acute aneurysm formation, chronic aneurysm formation and recurrence.

**Results:** In total 3 830 children were enrolled in the cohort. For treatment failure,  $\beta$ -propiolactation has the relative risk of 1.45 (95% CI 1.08 - 1.94). However, acidification and containing IgA were non-significant. For acute aneurysms, acidification has the relative risk of 1.49 (95% CI 1.17 - 1.90). But  $\beta$ -propiolactation and containing IgA were non-significant. For chronic aneurysm,  $\beta$ -propiolactation has the relative risk of 1.44 (95% CI 1.18 - 1.76) and acidification could protect it with the relative risk of 0.82 (95% CI 0.69 - 0.97). Containing IgA was non-significant. For recurrence, all 3 factors were non-significant.

**Conclusions:** This is the 1st longitudinal study in evaluating immunoglobulin effectiveness. This study has also the largest case numbers in this issue. In summary,  $\beta$ -propiolactation immunoglobulin has higher risk for treatment failure and chronic coronary aneurysm. Acidification might increase the risk for acute coronary aneurysm. Further prospective study may be needed to elucidate the real causal relationship between manufacturing process and outcomes.

#### Abstract no: 1496

### Epidemiology of Group A streptococcal pharyngitis in the Vanguard community demonstration site, South Africa

Mark Engel\*, Ronald Gounden\*, Andrew Whitelaw\*<sup>#,</sup>, Dylan Barth\*, Karen Cohen\*, Gary Maartens\*, Jim Dale<sup>†</sup> and Bongani Mayosi\*

\*University of Cape Town, Rondebosch, South Africa

<sup>#</sup>National Health Laboratory Services, Sandringham, Johannesburg, South Africa

<sup>†</sup>University of Tennessee, Memphis, Tennessee, United States of America

**Background:** Incidence rates of Group A streptococcal (GAS) swab-positive pharyngitis from 3.9 - 95/100 child-years have been reported. However, data from South Africa are scant and an understanding of the incidence of GAS pharyngitis among children within a local context is an important component of any Acute Rheumatic Fever and Rheumatic Heart Disease (RHD) control programme. Treating all symptomatic GAS sore throats in susceptible individuals in the community with a course of oral or parenteral penicillin presents the opportunity for primary intervention of RHD. The primary aim of this study was to determine the crude incidence rate of GAS pharyngitis-associated disease burden among children with sore throat.

**Methods:** We conducted a prospective, clinic-based study from 2008 - 2012. Following enrolment, participants with pharyngitis, aged 5 - 15 years were examined physically, before rendering a throat swab specimen for microbiological culture. SA census data were used to estimate crude incidence rates.

**Results:** We enrolled 840 participants with a mean age of 8.17 years, presenting with pharyngitis as a symptom. GAS was positive in 181 patients (21.6%). The crude incidence of pharyngitis and GAS-pharyngitis was estimated to be 837 and 180.4 cases/105 py respectively. A significantly higher incidence of GAS pharyngitis was observed in the younger age group (I.R.R 2.265, 95% CI 1.61 - 3.21), a trend seen throughout each year of the study period.

**Discussion:** This is the first community-based prospective incidence study of GAS-positive pharyngitis in Africa. While it is likely that our prevalence and incidence rates are an underestimate, our data nevertheless confirm that the complaint of GAS pharyngitis is common, particularly amongst younger children. Our data call attention to the need for vigilance as to the correct management of sore throat.

#### Abstract no: 1497

### Isolation of the right innominate artery with retrograde flow into the pulmonary artery

Keng Yean Wong, Ching Kit Chen, Tze Liang Choo, Min Yu Tan and Teng Hong Tan

Cardiology Service, KK Women's and Children's Hospital, Singapore

Isolation of the innominate artery is a rare congenital vascular defect. It is often associated with connection to the pulmonary artery. The majority of patients described have right aortic arch and isolation of the left innominate artery. A literature review by J Manner, et al. in 1997 showed only one of 16 patients had an isolated right innominate artery. We present a patient with a left aortic arch and isolation of the right innominate artery.

**Case report:** The patient is a six-year-old girl, diagnosed at birth with a large ventricular septal defect and pulmonary hypertension and 22q11 deletion. She had surgical repair of the VSD at 3 months of age. Post-operative echocardiogram showed no residual ventricular septal defect but the presence of a small patent ductus arteriosus. A cardiac catheterisation was done with the intention to close the duct. The angiogram showed a left aortic arch with absent right innominate artery. The left carotid, left vertebral and left subclavian arteries were seen. There was no patent ductus arteriosus seen in the usual position. Retrograde flow following the aortogram shows the right innominate artery draining via a duct into the main pulmonary artery. Surgical correction is considered in view of the arterio-venous steal. Manner et al. proposed that the pathogenesis of this condition is related to derangement of the embryonic aortopulmonary septation due to abnormal migration of neural crest cells.

**Conclusion:** Isolated right innominate artery is a very rare condition. This was seen in an asymptomatic patient with chromosome 22q11 deletion, unsuspected at the time of surgical repair of VSD, and can be a challenge diagnose.

#### Abstract no: 1501

### Evaluation of cardiac function using speckle-tracking echocardiography and tissue Doppler imaging in thalassemia major patients

Jacqueline Ellero\*, Sudeepthi Singarayar\*, Owen Jones\* and Christoph Camphausen\*<sup>#</sup>

\*Cardiology Department at Sydney Children's Hospital, Randwick, Sydney, Australia

<sup>#</sup>School of Women's & Children's Health, University of NSW, Sydney, Australia

**Background:** Speckle-tracking echocardiography (STE) and tissue Doppler imaging (TDI) are new techniques available for objectively quantifying the movement of individual cardiac segments. Increasing data suggest that STE and TDI provide reliable estimates of cardiac contractility and provide more detail than conventional echocardiography (CE).

**Objective:** To clarify the value of STE and TDI in the early detection of myocardial dysfunction in chronically transfused thalassemia major patients.

**Methods:** Multi-centre study using STE, TDI and CE to examine 66 thalassemia patients (including 14 children) and 132 age, gender, height and weight matched controls. STE will provide strain and strain rate for each of the 16 heart segments. All data will be compared across iron overload stratifications and between

patients and healthy controls. STE performed before and after the blood transfusion (usually 4 units of packed red blood cells) in all patients will help to quantify the effect of the increase of cardiac pre-load on STE. Data acquisition is performed on Philips IE33 machines and is currently underway. Each echocardiography study is de-identified and will be assessed by 3 different experienced cardiologists utilising the latest release of the Philips QLab Software (version 9) to test for inter-observer variability. The examiners will save their results in an electronic worksheet (Ms Excel), which will be analysed automatically. A custom-made software package will extract relevant data (i.e. global and regional, longitudinal and radial, end-systolic strain and peak systolic strain rate) and prepare for statistical analysis (SPSS). All measurements will be correlated with T2\* MRI data for all adult patients. T2\* MRI measures myocardial iron-load.

**Results/conclusions:** Available in January/February 2013.

**Abstract no: 1507**

**Rheumatic mitral valve disease in children: A big problem in a developing country**

**Euclides Tenorio<sup>\*</sup>, Cleusa Lapa<sup>#</sup>, Cristina Ventura<sup>†</sup>, Fernando Moraes<sup>‡</sup> and Carlos Moraes<sup>§</sup>**

<sup>\*</sup>Cirurgião cardiovascular, The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

<sup>#</sup>Chefe da cardiologia pediátrica, The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

<sup>†</sup>Cardiologista pediátrica, The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

<sup>‡</sup>Coordenador da cirurgia cardiovascular, The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

<sup>§</sup>Chefe da cirurgia cardiovascular, The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

**Background:** Rheumatic valvular heart disease remains a serious problem for public health in developing countries, being a common cause of morbimortality and responsible for half of the cardiovascular internments among children in Brazil.

**Objective:** This study was to assess the long term results of mitral valve repair in children with chronic rheumatic heart disease.

**Method:** From January 2002 - July 2012, 163 patients underwent mitral valve surgery at our institution 70: 42 (94%) underwent mitral valve reconstruction, 62 (38.05%) submitted mitral valve replacement to a bioprosthesis, and 31 (19.21%) underwent a 2nd replacement. The age varied from 3 - 16 years mean aged at surgery was 11.5±3.5 years; 92 (56.44%) children were female and 71 (43.56%) male. Eighty eight children (54%) were in the New York Heart Association functional class III and 49 (30%) in class IV. Reparative procedure included posterior leaflet extension with a pericardial patch in 46 (65.7%), and 24 patients (34.3%) received a Carpentier ring.

**Results:** In the groups of valve repair there was 1 early death by a metabolic disturbance and 2 children needs reoperation. One had a rupture of the plasty 24 hours after surgery and another presented severe haemolysis in the post-operative period. Both of them were submitted to mitral valve replacement. The period of follow-up was from 6 months - 10 years. In the group of valve replacement the durability of the bioprosthesis was 16 - 60 months, with a mean of 33 months.

**Conclusion:** Mitral valve repair in children with rheumatic heart disease is feasible and provides acceptable long-term results. In spite of a rather short bioprosthesis durability in our poor population, the socio-economic level must be considered in the choice of the valve substitute.

**Abstract no: 1511**

**Establishing a rheumatic heart disease control programme in Sudan: Achievements and challenges**

**Sulafa Ali<sup>\*#</sup> and Abdelmoniem Elseed<sup>#</sup>**

<sup>\*</sup>Department of Paediatrics and Child Health, Faculty of Medicine, Sudan Heart Institute and University of Khartoum, Sudan

<sup>#</sup>Department of Paediatrics and Child Health, Faculty of Medicine, University of Khartoum, Sudan

**Introduction:** Rheumatic heart disease (RHD) continues to have a significant burden on the health of young people in the Sudan with a prevalence of 10:1 000. Control programmes initiated by World Heart Organisation based on awareness and secondary prevention seized in 1990.

**Materials and methods:** The authors, inspired by the experience of the Pan African Society of Cardiology (PASCAR) and the RHD Group of the World Heart Federation's last meeting in Dubai in May 2012 initiated a RHD control programme initiative based on the Awareness, Surveillance, Advocacy and Prevention (ASAP) initiative of PASCAR.

**Results:** Primary prevention protocol is based on research done locally that validated a clinical algorithm for diagnosis of bacterial pharyngitis. The programme included protocols for primary and secondary care physicians as well as health assistants, conducting workshops for their training. Initiation of awareness programmes for physicians, medical students, school staff and the public and initiation of a local registry with more emphasis on primary prevention aspect. The project was approved after long discussions with Ministry of Health and the scientific societies (Sudan Heart Society and the Sudanese Association of Paediatricians). It is included in the School and Adolescent Health Programme.

**Conclusion:** The abstract throws light on the achievements and difficulties of this project.

**Abstract no: 1513**

**Under-diagnosed cardiomyopathies in the paediatric age**

**Sulafa Ali**

Department of Paediatrics and Child Health, Faculty of Medicine, Sudan Heart Institute and University of Khartoum, Sudan

**Introduction:** The commonest type of paediatric cardiomyopathy (CMP) is the dilated type (DCMP) which constitutes 70 - 80% of cases.

**Materials and methods:** In this report we reviewed 2 series of patients with other types of CMP which are not commonly reported but we believe that they are under-diagnosed.

**Results:** The 1st type is known as tropical restrictive CMP or endomyocardial fibrosis (EMF). We found that EMF represented 18% of cases of CMP seen at a tertiary centre. We reviewed the clinical and echocardiographic features of patients with EMF and discussed differentiation of this entity from diseases that can mimic it. The 2nd type of CMP is non-compaction CMP (NCCMP) which has recently gained a lot of attention and is also thought to be under-diagnosed. We describe a series of 52 patients in Arab/African patients and it was found that NCCMP represented at least 20% of cases of CMP. Within this disease we reported unique clinical and echocardiographic associations. Female preponderance, a relapsing course and mitral valve regurgitation were distinctive features of NCCMP in our area.

**Conclusion:** We need to increase the awareness of physicians about these types of CMP so that appropriate treatment and counselling can be instituted.

**Abstract no: 1514****The Aristotle comprehensive complexity score predicts mortality after surgical ligation of patent ductus arteriosus in preterm infants***Jae Young Lee, Yun Hee Jang, Eun-Jung Lee, Sena Moon, YoungAh Youn, Ju Young Lee, In Kyung Sung and Jung Eun Kim*

Seoul St. Mary's Hospital, The Catholic University of Korea, Korea

**Background:** The outcome measure after surgical ligation of patent ductus arteriosus (PDA) in preterm infants is often complicated by prematurity associated co-morbidities. The Aristotle comprehensive complexity score (ACCS) has been proposed as a useful tool for complexity adjustment in the analysis of outcome after congenital heart surgery. The aims were to define preoperative risk factors for mortality and to demonstrate the usefulness of ACCS to predict mortality after surgical ligation of PDA in the preterm.

**Material and methods:** Included were 49 preterm babies (<35 weeks of gestation), from May 2009 - July 2012, who had surgical ligation of PDA. Mean (range) gestational age and birth weight were 28 weeks (23 - 35 weeks) and 1 102 g (520 - 2 280g), respectively. Mean (range) age and weight at operation were 16 days (4 - 44 days) and 1 199g (400 - 2 880g), respectively. Initial oral ibuprofen was ineffective in 24 patients and contraindicated in 25. All surgical ligations were done at bedside in the neonatal intensive care unit. Preoperative clinical and laboratory profiles were reviewed and ACCS was derived.

**Results:** Eight out of 49 (16.3%) died after a median of 14 days (range, 2 - 73 days) after PDA ligation. Patients who had contraindications for oral ibuprofen (OR, 8.94; p=0.049), coagulopathy (OR, 12.13; p=0.025), renal dysfunction (OR, 28.88; p=0.003), intraventricular hemorrhage  $\geq$  grade III or seizure within 48 hours of operation (OR, 34.00; p=0.002), and ACCS >15 (OR 415.00; p<0.001) were significantly associated with an increased risk for mortality. At multivariate logistic regression analysis, ACCS >15 was the only independent risk factor (OR, 103; p=0.023) for mortality.

**Conclusions:** The ACCS, especially procedure independent complexity factors, is a useful tool to predict mortality after ligation of PDA in the preterm.

**Abstract no: 1520****Clinical and echocardiographic features of children with rheumatic heart disease and their serum cytokine profile***Sulafa Ali\*#, Inaam Nooreldaim Eldaim\*, Samia Osman† and Sahar Bakhite#*

\*Sudan Heart Institute, Khartoum, Sudan

#University of Khartoum, Khartoum, Sudan

†Jafar Ibn Ouf Hospital, Khartoum and Ministry of Health, Sudan

**Introduction:** Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) constitute important public health problems in developing countries.

**Materials and methods:** Children with ARF and RHD seen at Children's Hospital in Khartoum, Sudan from May 2008 - 2009 were examined clinically and via Echocardiography. Blood cytokines [interleukin 10 (IL10)], tumour necrosis factor alpha (TNF- alpha) and interferon gamma (IFN-gamma) were done.

**Results:** Thirty six children were enrolled; 63% had established RHD, and 37% ARF. Mitral regurgitation (MR) was the most common lesion (94%). 95% of the valve lesions were severe. The serum interleukin IL10 level ranged between 3 - 6pg/ml. TNF alpha levels were 9 - 100pg/ml in 12 patients (40%), 101 - 1000pg/ml in 10 patients (33%) and >1 000 in 8 patients (26%). The level of IFN gamma ranged between 2 - 7pg/ml in all patients except 2 (84 and 135pg/ml respectively).

**Conclusion:** RHD is manifested with severe valvular lesions and a high TNF alpha which indicates and ongoing inflammation.

**Abstract no: 1523****Coarctation of the aorta in infants: Clinical, therapeutic and prognostic characteristics in 52 cases***Euclides Tenorio, Luziene Bonates, Cleusa Lapa, Cristina Ventura, Izabella Brandão, Fernando Moraes and Carlos Moraes*

The Medical Institution, Prof. Fernando Figueira, Recife, Pernambuco, Brazil

**Objective:** To describe the clinical and therapeutic characteristics of 52 infants undergoing surgical repair of coarctation of the aorta.

**Material and methods:** Fifty two infants aged <1 year who underwent surgical correction of coarctation of aorta from July 2000 - January 2012.

**Results:** Thirty nine (75%) children were male. The median age at diagnosis was 30 days, ranging from 20 days - 1 year. The onset of clinical manifestations in 20 (51.9%) cases occurred within the 1st month of life, 13 (25%) cases from the 1st to the 6th month of life, and 19 (36.5%) cases between the 6th month and the 1st year of life. The most significant finding was semiological absence or reduction of arterial pulses in the lower limbs in 52 patients and all clinical signs of congestive heart failure in 46 (88.5%). Cardiomegaly was commonly found in all children. There was a predominance of right ventricular overload in half the cases. Echocardiographic study showed an association of coarctation of the aorta with other intra-cardiac abnormalities in 24 (46.1%) of cases, with the bicuspid aortic valve and VSD more frequently presented. Ventricular systolic dysfunction was observed in all children. Regarding the surgical procedure, angioplasty of the left subclavian artery flap was performed in 46 (90.2%) cases, which is associated with pulmonary artery banding in 4 (7.8%) and angioplasty bovine pericardium was performed in 4 (7.8%) cases. Paradoxical hypertension was observed in 44 (84.6%) cases. The mortality rate was 11.5%.

**Conclusion:** In our series there was a predominance of congestive heart failure as the initial manifestation of the disease. The importance of pulse palpation in the lower limbs as a means of early detection of this anomaly was confirmed, once again.

**Abstract no: I531****The Aristotle comprehensive complexity score predicts mortality after surgical ligation of patent ductus arteriosus in pre-term infants***Jae Young Lee, Yun Hee Jang, Eun-Jung Lee, Sena Moon, YoungAh Youn, Ju Young Lee, In Kyung Sung and Jung Eun Kim*

Seoul St, Mary's Hospital, Khartoum, Seoul, South Korea

**Background:** The outcome measure after surgical ligation of patent ductus arteriosus (PDA) in pre-term infants is often complicated by prematurity associated co-morbidities. The Aristotle comprehensive complexity score (ACCS) has been proposed as a useful tool for complexity adjustment in the analysis of outcome after congenital heart surgery. The aims were firstly to define pre-operative risk factors for mortality and secondly to demonstrate the usefulness of ACCS to predict mortality after surgical ligation of PDA in the pre-term.

**Material and methods:** Included were 49 pre-term babies (<35 weeks of gestation), from May 2009 - July 2012, who had surgical ligation of PDA. Mean (range) gestational age and birth weight were 28 weeks (23 - 35 weeks) and 1 102 gram (520 - 2 280g), respectively. Mean (range) age and weight at operation were 16 days (4 - 44 days) and 1 199 gram (400 - 2 880g), respectively. Initial oral ibuprofen was ineffective in 24 patients and contra-indicated in 25. All surgical ligations were done at bedside in the neonatal intensive care unit. Pre-operative clinical and laboratory profiles were reviewed and ACCS was derived.

**Results:** Eight out of 49 (16.3%) died after a median of 14 days (range, 2 - 73 days) after PDA ligation. Patients who had contra-indications for oral ibuprofen (OR, 8.94; p=0.049), coagulopathy (OR, 12.13; p=0.025), renal dysfunction (OR, 28.88; p=0.003), intraventricular haemorrhage  $\geq$ Grade III or seizure within 48 hours of operation (OR, 34.00; p=0.002), and ACCS >15 (OR 415.00; p <0.001) were significantly associated with an increased risk for mortality. At multivariate logistic regression analysis, ACCS >15 was the only independent risk factor (OR, 103; p=0.023) for mortality.

**Conclusions:** The ACCS, especially procedure independent complexity factors, is a useful tool to predict mortality after ligation of PDA in the pre-term.

**Abstract no: I532****Echocardiographic analysis by 3-D, tissue Doppler, AFI, Strain, SR of the right and left ventricular dysfunction in surgically repair patients with Tetralogy of Fallot***Ana Maria Susana De Dios, Romina Carugati, Fernanda Biancolini, Julio Cesar Biancolini, Victorio Luccini, Ackerman Judith, Manso Paula, Martinez Ines, Florencia Levantini and Damsky Barbosa Jesus*

Department of Paediatric Cardiology, Pedro de Elizalde Children Hospital, Buenos Aires, Argentina

**Objectives:** Our study focuses on echocardiographic assessment of the right and left ventricular systolic function in response to volume overload resulting from pulmonary regurgitation in surgically treated patients with Tetralogy of Fallot (ToF).

**Methods and results:** We included 63 patients with severe pulmonary regurgitation after surgical correction of ToF, in whom AFI automatic function image by Two-Dimensional Speckle Tracking was performed to measure Left Ventricular Longitudinal Strain and Strain Rate in right (RV) and left (LV) ventricle. Age:  $14 \pm 4$  years, surgical correction was done at  $2.7 \pm 1$  year, follow-up  $11.8 \pm 6$  years. Dividing them into 3 groups: (1) RV <100ml/m<sup>2</sup>, (2) RV 100 - 120ml/m<sup>2</sup>, (3) RV:>120ml/m<sup>2</sup>. The 1st group, shows a decrease in RV medial tissue Doppler: velocity systolic (S') and diastolic E-wave and IVAm/s<sup>2</sup>, minimal dysfunction in AFI RV, and depress Strain and SR medial RV; the 2nd group basal and medial dysfunction RV and basal LV; and the 3rd group: RV>120ml/m<sup>2</sup> tissue Doppler S' <0.05m/s, and E-wave and IVA m/s<sup>2</sup> include inversion E/A ratio, IVAm/m<sup>2</sup> RV and LV decrease. AFI RV and LV show severe dysfunction; Strain and SR RV basal and medial was positive and LV basal and medial remained the same.

**Conclusions:** Echocardiographic analysis offers great possibilities for assessment of right and left ventricular dysfunction, identifying in particular, as well as timing the selection of patients for re-intervention.

**Abstract no: I533****Improving cardiac patient education in a multi-lingual society***Lindy Mitchell and Jayneel Joshi*

Steve Biko Academic Hospital, University of Pretoria, Pretoria, South Africa

**Background:** Steve Biko Academic Hospital is the cardiac referral centre for 4 of the 9 provinces in South Africa, including 2 with large rural populations, namely Limpopo and Mpumalanga provinces. As such, our patients speak a wide selection of the 11 official languages. The occasional foreign national also finds their way to our doors and it is not unusual for as many as 8 different languages to be spoken on any outpatient clinic day.

Many of the parents and caregivers we see are also illiterate or only have primary level literacy skills. This provides a challenge to the cardiologist who has to explain often very complex congenital cardiac defects to patients and parents who do not speak his/her home language/languages.

Adequate informed consent for procedures must also be obtained and patient compliance with treatment schedules and dosages, which often change from visit to visit, must be ensured in order to offer the child the best care and management possible. Skilled, informed interpreters are not always readily available, so we have had to find other methods of getting our message across. We present some of the teaching aids and materials that we use in our unit to improve communication with our patients and their caregivers.

**Abstract no: I536**

**Geographical information systems (GIS) as an aid to investigate concordance patterns among rheumatic heart disease (RHD) patients: A pilot project**

Dylan Barth\*, Liesl Zuhlke#, Alexia Joachim\*, Blanche Cupido\*, Bongani Mayosi\* and Mark Engel\*

\*University of Cape Town, Rondebosch, South Africa

#Red Cross War Memorial Children's Hospital, Rondebosch, Cape Town, South Africa

**Background:** Lack of concordance to monthly INR monitoring in RHD patients is considered to be a contributor to stroke and other complications. Various factors for this non-concordance have been suggested including limited access to health care facilities. We hypothesise that the rate of default in patients correlated with the travel distance from the patient's residence to the health care facility to which they have been referred. GIS enables the visual representation of data to establish patterns and trends between patients' follow-up visits for INR monitoring and their geographical context. The aim of this study was, therefore to investigate these associations by developing a spatial display instrument, mapping the residential addresses of patients against their respective designated referral clinic.

**Methods:** Patient data, including residential addresses, referral clinics and concordance patterns were obtained from the REMEDY database. REMEDY is a RHD registry in its pilot phase targeting 3 000 patients. Addresses were converted to geographical coordinates and ArcGIS 9.3.1® was used for mapping and spatial analyses. The geo-coded addresses were further checked in ArcGIS for errors in coordinate data. The travel distance between individual residential addresses and referral clinics were calculated and compared to concordance patterns of patients.

**Results:** RHD patients (n=26) reside between 1.2 - 26.2km from their referred clinics (mean 9.68±8.1 km). The average period between clinic visits was 32 days (range, 18 - 43 days). Preliminary results suggest that concordance to monthly INR monitoring is not associated with patients' travel distance.

**Discussion:** This is the first attempt at the application of GIS within this area of RHD. Distance from referred clinic does not impact upon patients' attendance at INR monitoring. Nevertheless, GIS presents as an ideal tool to visual relationships between RHD patients' follow-up visits for INR monitoring and their geographical context.

**Abstract no: I538**

**Risk factors associated with the development of systemic-pulmonary collateral flow in single ventricle patients with superior cavopulmonary connections**

Andrew Glatz, Jonathan Rome, Neil Harrison, Adam Small, Yoav Dori, Matthew Gillespie, Matthew Harris, Mark Fogel and Kevin Whitehead

Division of Cardiology, Children's Hospital of Philadelphia, Philadelphia, Massachusetts, United States of America

**Background:** Systemic-pulmonary collateral flow (Collf) is common in patients after superior cavo-pulmonary connection (SCPC). Risk factors associated with Collf are unclear. We sought to identify risk factors for Collf in a cross-section of patients with SCPC.

**Methods:** A retrospective chart review of events from birth to the time of study was performed for SCPC patients who had Collf quantified by cardiac magnetic resonance imaging (cMRI). Collf was reported as indexed flow (L/min/m<sup>2</sup>), as a percentage of aortic flow (Collf/Ao), and as a percentage of pulmonary venous flow (Collf/PV).

**Results:** From April 2008 - August 2011, 96 SCPC patients at 2.6±1.1 years of age and 799±400 days from SCPC had Collf measured at 1.6±0.7L/min/m<sup>2</sup>, comprising 33.5±11.1% of aortic flow and 48.3±15.9% of pulmonary venous flow. Two of 3 Collf indices were higher in patients with a prior BT shunt vs. no prior BT shunt (pulmonary artery band, right ventricle-pulmonary artery conduit, or no initial palliation) (Collf: 1.7±0.8 vs. 1.4±0.4L/min/m<sup>2</sup>, p=0.04; Collf/Ao: 35.2±11.6 vs. 28.6±7.6%, p=0.009) and in those with a Hemi-Fontan vs. bidirectional Glenn (Collf/Ao: 38.5±13.5 vs. 32.2±10%, p=0.02; Collf/PV: 57.6±20.1 vs. 45.8±13.8%, p=0.003). By Spearman testing, positive correlations exist between Collf indices and indicators of peri-operative morbidity (Table 1). We did not find associations between Collf and pulse oximetry, haemoglobin, or haemodynamics pre-Stage 2 catheterisation.

**TABLE 1:** Risk factors associated with systemic-pulmonary collateral flow

Risk factor	Total cohort (n=96)		Total Collf (L/min/m <sup>2</sup> )		Collf/Ao (%)		Collf/PV (%)	
	Median	Range	rho	p	rho	p	rho	p-value
Total ICU duration (d)	17.5	2.261	0.23	0.036	0.29	0.008	0.39	0.0003
Stage I hospital duration (d)	23	2.263	0.18	0.1	0.26	0.02	0.3	0.005
Stage II hospital duration (d)	7	2.263	0.2	0.05	0.3	0.004	0.37	0.0002
Total hospital duration (d)	33.5	5.342	0.25	0.02	0.33	0.002	0.39	0.0003
Total # of chest tubes	2	1.7	0.25	0.02	0.29	0.005	0.22	0.04
Stage I chest tube duration (d)	2	0.49	0.2	0.06	0.28	0.01	0.27	0.01
Total chest tube duration (d)	3	0.54	0.21	0.04	0.24	0.02	0.2	0.05
Total chest operations	2	1.9	0.23	0.03	0.24	0.02	0.19	0.06

**Conclusion:** Collf occurs commonly in patients with SCPC and is related to surgical history, chest tube and hospital duration. These data support the hypotheses that peri-operative morbidity and pleural inflammation may play a role in the development of Collf.

**Abstract no: I540**

**Post mortem imaging in paediatric cardiology**

**Francesca Romana Pluchinotta\*<sup>‡</sup>, Prashob Porayette\*, Sanjay P. Prabhu<sup>#</sup>, Lisa Teot<sup>†</sup> and Stephen P. Sanders\***

\*Department of Cardiology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>#</sup>Department of Radiology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>†</sup>Department of Pathology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>‡</sup>Paediatric Cardiology Unit, Department of Paediatrics, University of Padua, Italy

**Background:** Post mortem imaging (PMI), used widely in forensic pathology, could substitute for autopsy in paediatric cardiology patients, increasing effective autopsy rate and facilitating quality improvement.

**Materials and methods:** Paediatric cardiology patients who died during the study period and in whom an autopsy was planned were eligible. PMI included magnetic resonance imaging (MRI) of heart, brain and abdomen (see table), and multi-detector computed tomography (MDCT) angiography, using iodinated contrast diluted 1:8 with PEG, injected in the inferior vena cava (8ml/kg) and aorta (5ml/kg).

**Results:** During a 4-month period 5 patients underwent PMI followed by autopsy; both MDCT and MRI in 4 and MDCT only in 1 because of metal ECMO cannulae. Cardiac anatomy, condition of surgical repair, and abdominal anatomy by PMI correlated perfectly with autopsy. All major intra-cranial findings (intra-cranial haemorrhage, parietal infarct, white matter atrophy) were diagnosed by both PMI and autopsy. Cause of death by PMI and autopsy was congruent in 3 cases: not determined in 2 cases and intra-cranial haemorrhage with herniation in 1 case. PMI (MDCT only) showed severe bilateral pulmonary injury, found to be infarction at autopsy, but missed left ventricle and pulmonary artery thrombosis. PMI missed diffuse left and right ventricular infarction seen by histology in 1 case. MRI and MDCT were complementary, with MRI better for delineation of cardiac and abdominal anatomy, and brain abnormalities, but MDCT was superior for delineation of central vasculature, especially the coronary arteries, and pulmonary disease. Position of lines and tubes, condition of the body wall and skin, and skeletal anatomy were demonstrated by the MDCT 3-D data set.

**TABLE 1:** Post mortem MRI parameter of the chest, the abdomen and the brain in human subjects.

	Planes	Sequence	FoV (mm)	Slice thickness (mm)	TR (msec)	TE (msec)	NEX	Flip Angle
Heart	Axial	T2 SENSE	160	4mm/skip 0 2mm/skip 0	4 000	102	2	90°
	Short axis, 4-chamber; 2-chamber	T2	160	4mm/skip 0	4 000	102	3	90°
	3D morphology CLEAR	T2	140	0.66mm/skip 0	6.76	3.38	3	90°
Brain	Axial, Sagittal	T1	200 - 210	4mm/skip 0	450 500	15 15	2 3	69° 69°
	Axial	T2	200 - 210	2mm/skip 0	4 000	100	3	90°
	Axial	FFE	200 - 210	4mm/skip 0	495	20 - 25	1	18°
Abdomen	Coronal	T2	160	4mm/skip 0	4 000	102	3	90°

**Conclusions:** Our preliminary study in human cadavers shows that PMI is feasible in a paediatric hospital environment. The small sample size precludes analysis of diagnostic accuracy. Nevertheless, we are encouraged by the diagnostic performance of PMI.

**Abstract no: I542**

**Post mortem imaging of ante mortem myocardial ischaemia**

**Francesca Romana Pluchinotta\*<sup>§</sup>, Prashob Porayette\*, Patrick O. Myers<sup>#</sup>, Peter Chen<sup>#</sup>, Eric Feins<sup>#</sup>, Lisa Teot<sup>†</sup>, Sanjay P. Prabhu<sup>‡</sup> and Stephen P. Sanders\***

\*Department of Cardiology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>#</sup>Department of Cardiac Surgery, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>†</sup>Department of Pathology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>‡</sup>Department of Radiology, Boston Children's Hospital, Boston, Massachusetts, United States of America  
<sup>§</sup>Paediatric Cardiology Unit, Department of Paediatrics, University of Padua, Italy

**Background:** Post mortem MRI (PMRI) has been used in forensic pathology to detect ante mortem ischaemia but the sensitivity is unknown. We tested the survival time required for detection of ante mortem ischaemia by PMRI in a pig carcass model.

**Materials and methods:** Nine pigs (7 - 35kg) underwent surgical ligation of the distal LAD (8) and/or a RCA branch (4) and were euthanised 1 - 6 hours after ligation. PMRI (T1, T2, PD, and spin echo 3-D volumetric sequences) was performed 12 - 48 hours after euthanasia. Images were inspected, and signal intensity of 17 myocardial segments was measured serially. Heart sections were submitted for histology.

**Results:** Ligation produced discoloration and dyskinesia of the target segment(s) in all cases. MRI T2-weighted sequences (TE 102, TR 4 000) showed the ischaemic area as hyper-intense in 4/4 LAD ligations with ≥4 hours of ischaemic time and in 0/4 with <4 hours. Histological evidence of ischaemia was present

in 4/4 after 4 hours. Right ventricular ischaemia was visible on MRI after 6 hours of ischaemic time in 1 animal. The right ventricular myocardium is thinner than the left ventricular myocardium, making it difficult to distinguish myocardium from epicardium. Intra-observer variability for MRI signal intensity of the myocardium was 0.93 (95% confidence interval 0.91 - 0.94) for the LV and 0.18 (95% confidence interval -0.03 - 0.37) for the RV. The percent difference in signal intensity between ischaemic and normal myocardium was greatest on T2-weighted images up to 12 hours after death and then decreased with increasing time, approaching that of the non-ischaemic areas by 24 - 48 hours (Table 1).

**TABLE 1:** Percent difference in signal intensity between normal and ischaemic LV myocardium in relation to time after death on MRI T2-weighted images.

Ischaemic time (hours)	Case <sup>#</sup>	Imaging time after death (hours)		
		<12 h	12 - 24	24 - 48
≤2	1, 2, 3 and 4	5.7%	6%*	13.1% <sup>#</sup>
4	7 and 8	24% <sup>§</sup>	10.1%	
6	5 and 6	17.9% <sup>°</sup>	5.7% <sup>†</sup>	5.9% <sup>‡</sup>

\*animals 2,3 and 4. <sup>#</sup>animal 1. <sup>†</sup>animal 5. <sup>‡</sup>animal 6. <sup>§</sup>p=0.0001 on Wilcoxon rank sum test. <sup>°</sup>p=0.0013 on Wilcoxon rank sum test.

**Conclusions:** Ischaemic lesions of the LV, but not the RV at least 4 hours old can be reliably detected as hyper-intense areas on T2 weighted PMRI. By 24 - 48 hours after death, surrounding myocardial density approaches that of the ischaemic area obscuring it.

#### Abstract no: 1544

### Spectrum and outcome of paediatric patients undergoing right ventricular pacing

Ameena Amod<sup>\*</sup>, Annurupa Bhagwandin<sup>\*</sup>, Andiswa Nzimela<sup>\*,†</sup> and Ebrahim GM Hoosen<sup>\*,†</sup>

<sup>\*</sup>Department of Cardiology, Inkosi Albert Luthuli Central Hospital, University of KwaZulu-Natal, Durban, South Africa

<sup>#</sup>Paediatric Cardiology, Inkosi Albert Luthuli Central Hospital, University of KwaZulu-Natal, Durban, South Africa

<sup>†</sup>University of KwaZulu-Natal, Durban, South Africa

**Background:** Pacemaker implantation (PMI) in the paediatric populations is uncommon with implantation performed in selected patients with congenital heart block or following cardiac surgery.

**Aim:** To evaluate the spectrum and outcomes of patients undergoing right ventricular (RV) pacing in the centre's paediatric population.

**Method:** Retrospective chart review of all paediatric patients undergoing PMI since 2003.

**Results:** Seventeen patients were identified. Twelve were congenital complete heart block (CCHB), of which 2 had associated structural abnormalities and 5 were heart block post cardiac surgery. The age ranged from 7 days - 13 years. All implants were single chamber rate responsive devices with 82% epicardially placed and 17.6% endocardial. Epicardial placement was initially on the RV free wall except for the last 3 which were placed on the right ventricular outflow tract. One late mortality and 4 early mortalities in neonates with CCHB were recorded. Associated morbidity in these patients were: (1) Hydrops fetalis in a patient with left isomerism and TAPVC; (2) Long QT syndrome with poor ventricular function post pacemaker implantation; and (3) Gram negative sepsis 9 days after PMI. Sudden unexpected death shortly after PMI occurred in 1 neonate. Late mortality occurred from pneumonia in a 14-month-old child with right isomerism/AVSD who underwent PMI and pulmonary artery banding at 4 months of age. Two patients with progressive heart failure at follow-up attributed to pacemaker-induced cardiomyopathy were upgraded to biventricular pacing with good results.

**Conclusion:** While patient numbers were insufficient to reach any clear conclusion, patients undergoing RV PMI had good outcomes, the exception being those with heterotaxy syndrome and neonates with comorbidities. Alternative pacing techniques to avoid or manage pacemaker induced cardiomyopathy may be indicated and requires further investigation.

#### Abstract no: 1545

### Quantitative gated SPECT valuable to effectiveness of cardiac resynchronisation therapy in functionally single ventricles

Tomomi Nishimura<sup>\*</sup>, Im Sam Park<sup>\*</sup>, Tomomi Ueda<sup>\*</sup>, Tadahiro Yoshikawa<sup>\*</sup>, Naoki Wada<sup>#</sup>, Makoto Ando<sup>#</sup>, Yukihiko Takahashi<sup>#</sup> and Kanki Inoue<sup>†</sup>

<sup>\*</sup>Department of Paediatric Cardiology, Sakakibara Heart Institute, Tokyo, Japan

<sup>#</sup>Department of Paediatric Surgery, Sakakibara Heart Institute, Tokyo, Japan

<sup>†</sup>Department of Cardiology, Sakakibara Heart Institute, Tokyo, Japan

**Background:** Cardiac resynchronisation therapy (CRT) is an effective treatment for severe left heart failure. Recently, CRT is increasingly used in children and congenital heart disease (CHD) patients, but published data about its effectiveness are limited comparison with adults with left heart failure. Furthermore, for CHD patients except cardiomyopathy, it is hard to evaluate the outcome because of a complicated form.

**Methods:** We describe a case of a 35-year-old woman with functionally single ventricle who started CRT 4 years ago because of failing Fontan. At baseline and after CRT, her clinical status by New York Heart Association (NYHA) class, ejection fraction (EF) and ventricular dyssynchrony [using quantitative gated single-photon emission computed tomography (QGS) from Cedars-Sinai QGS software] were evaluated. Improvement in NYHA class and EF was considered responders. Using 99mTc-sestamibi the end-diastolic volume (EDV), end-systolic volume (ESV), ventricular ejection fraction (EF), summed motion score (SMS), and summed thickening score (STS) were calculated with QGS.

**Results:** We found improvements in: EDV: 210ml - 186ml; ESV: 173ml - 120ml; EF: 18% - 35%; SMS: 32 - 7; STS: 27 - 13; and uptake of nuclide to myocardium. Phase analysis derived from QGS, polar map was indicated as heterogeneous distribution in pre-CRT and was improved at follow-up. This case showed an improvement in NYHA class and EF, and, since these are considered responders, the results were positive.

**Conclusions:** Radionuclide study is a useful tool with which to examine and evaluate synchrony, because it is reproducible, simple and visual.

**Abstract no: I562**

**Severe left ventricular inflow obstruction in an infant - an unusual complication of dilated coronary sinus: Case report and review of literature**

**Pankaj Gupta\* and Christoph Camphausen#**

\*Department of Cardiology, Sydney children's hospital network, Sydney, Australia

#Department of Cardiology, Sydney children's hospital, Sydney, Australia

**Introduction:** Dilated coronary sinus secondary to persistent left superior vena cava is a common entity. However, dilated coronary sinus causing severe left ventricular inflow obstruction is an infrequent finding. We report on a 4-month-old infant who presented with congestive cardiac failure, secondary to marked enlargement of coronary sinus with consequent obstruction of left ventricular inflow

**Case report:** A 4-month-old infant was referred to our paediatrics tertiary hospital with history of poor weight gain, breathlessness during feeding and frequent sweating. Cardiovascular examination revealed right parasternal heave with loud diastolic murmur in mitral area. ECG showed right axis deviation with marked right ventricular hypertrophy. Chest roentgenograms showed mild cardiomegaly and passive pulmonary venous congestion. 2-D Echocardiogram suggested diagnosis of Cor triatriatum. However, acquisition of stereoscopic 3-D volume dataset identified the exact nature of the obstruction and revealed persistent left superior vena cava and markedly dilated coronary sinus causing severe narrowing of inflow pathway of left ventricle. Operative finding confirmed the diagnosis with presence of markedly dilated coronary sinus causing narrowing of left ventricular inflow and absence of any membrane in the left atrium. He underwent reduction plasty of coronary sinus which included longitudinal incision of coronary sinus, excision of a segment of its wall and reconstruction of the dilated coronary sinus wall. An excellent result was achieved with no residual left ventricular inflow obstruction in the follow-up.

**Conclusion:** Dilated coronary sinus is a rare but possible etiology for left ventricular inflow obstruction. Reduction plasty of coronary sinus is a safe and effective procedure for treating this entity. In addition, stereoscopic 3-D is effective alternative to conventional catheterisation for diagnosing this entity.

**Abstract no: I565**

**Investigation of idiopathic dilated pulmonary artery: the role of different non-invasive imaging modalities: A case report**

**Shuk Han Lee and Louisa Poon**

Department of Paediatrics, Queen Elizabeth Hospital, Hong Kong, Thailand

**Case description:** A 1-year-old Chinese girl with Noonan syndrome, mild mental retardation and left renal agenesis was followed up for idiopathic pulmonary artery dilatation. At age 7, her echocardiogram and right heart catheterisation showed idiopathic pulmonary artery dilatation. Trans-thoracic echocardiogram was repeated at 14 years of age. It showed dilated main pulmonary artery, measuring 3.38cm in diameter and dilated left coronary artery. The left main coronary artery measured 6.2mm and the left anterior descending artery measured 5.2mm in diameter. There were multiple low velocity turbulent jets coming in the mid-right ventricular free wall. There was no evidence of pulmonary hypertension from Doppler pressure gradient assessment of pulmonary regurgitation.

**Method and results:** Left and right cardiac catheterisation was performed with a suspicion of coronary arteriovenous fistula to the right ventricle. Left coronary angiogram did not show any fistula but retrograde contrast flow to the right coronary artery. Attempt to cannulate the right coronary artery was unsuccessful. Detailed review of the angiogram showed suspicious anomalous origin of right coronary artery from the main pulmonary artery but we could not confirm the diagnosis by manipulating a catheter in it. A CT coronary angiogram with 3-D reconstruction was later performed. It showed beautifully the anomalous origin of the right coronary artery from the main pulmonary artery. MRI perfusion scan with adenosine stress test showed no evidence of myocardial ischaemia. The patient was treated conservatively.

**Conclusions:** One should be careful with the diagnosis of idiopathic pulmonary artery dilatation. In this case, the combined role of non-invasive imaging including echocardiogram, CT coronary angiogram and cMRI perfusion scan can replace traditional invasive cardiac catheterisation to reveal the true underlying cause.

**Abstract no: I571**

**Course of defibrillation thresholds in mid-term follow-up after ICD-implantation using the extracardiac technique**

**Thomas Kriebel\*, Wolfgang Ruschewski#, Heike Schneider\*, Matthias Müller\*, Matthias Pietzka\*, Ulrich Krause\* and Thomas Paul\***

\*Department of Paediatric Cardiology and Intensive Care Medicine, Georg-August-University Göttingen, Germany

#Department of Thoracic and Cardiovascular Surgery, Georg-August-University Göttingen, Germany

**Introduction:** There is limited experience in ICD implantation in infants and small children to date. The previously described extracardiac technique offers an effective and safe ICD-implantation technique in these patients. However, data of mid-term follow-up especially with regards to the course of the defibrillation thresholds (DFT) during the growth of the patients are lacking.

**Patients and methods:** An extracardiac ICD-system was implanted in 19 patients [mean age: 5.6 (range 0.2 - 9.8) years] as previously been described. In brief: Under fluoroscopic guidance a defibrillator lead was tunnelled subpleurally along the course of the 2nd - 6th rib until almost reaching the vertebral column. After a partial inferior sternotomy, bipolar steroid-eluting sensing and pacing leads were sutured to the atrial wall in selected patients and to the anterior wall of the right ventricle in all patients. The ICD device was implanted as "active can" in the upper abdomen. Sensing, pacing, and DFT as well as impedances were verified intra-operatively, 3 months later and every 12 months respectively.

**Results:** In all patients, intra-operative DFT between the extracardiac lead and device was <15 Joule (mean 11.2J). No serious complications were noted. No serious complications were noted. After a mean follow-up of 1.8 years DFT remained stable or below <20 Joule in 15/19 patients. Revision was required in 3 patients due to unacceptable DFT >20 Joule. Using multiple regression analysis a significant correlation between the size of the patient and mean DFT was noted. In 3 patients inadequate ventricular sensing was noted. Revision of the system could be avoided in 2/3 patients by an alternative sensing option between the ventricular leads and subpleural shock electrode.

**Conclusions:** The extracardiac technique offers a safe and effective approach for ICD implantation in infants and small children. However, regular DFT testing is mandatory to recognise failure of the system.

**Abstract no: I573**

**Risk factors and natural history of isolated non-compaction cardiomyopathy in childhood**

*Alejandra Villa, Carlos Rosental, Marisa Di Santo, Natalia Napoli and Alejandra Mori*

Garrahan Children's Hospital, Buenos Aires, Argentina

**Background:** Left ventricular non-compaction (LVNC) is an uncommonly reported genetic disorder of endocardial morphogenesis. Clinical and prognostic heterogeneity is described. The purpose of this study was to identify the clinical characteristics and risk factors of children with LVNC.

**Methods and result:** We retrospectively reviewed 24 children with LVNC evaluated at the Garrahan Children's Hospital from January 1996 - December 2011. The median age at presentation was 2 years (range, 15 days - 13 years). Median follow-up was 4 years (range, 2 years - 15 years). Seventeen patients (70%) had EKG abnormalities, six patients LBBB, two patients WPW. Both ventricles were involved in 8 patients (33.3%) and only the left ventricle in 16 patients (69.1%). Left ventricular systolic function was depressed in 12 patients (50%), with a median ejection fraction of 30% (range, 20% - 66%) at diagnosis. Of 9 patients who presented with depressed left ventricular contractility, 66% had function recovery. During the follow-up, 8 patients (33.3%) suffered any adverse event. Five patients with biventricular and 3 with left ventricular compromise were put on the heart transplantation and 2 required ventricular assist device.

**Conclusion:** In this cohort of patients with LVNC 69.9% remain in functional class I-II. Risk factors for adverse events, heart transplantation or death were biventricular involvement ( $p < 0.05$ ) and conduction disturbances ( $p < 0.001$ ).

**Abstract no: I574**

**Peripheral micro-vascular function is affected in young individuals at risk for HCM and correlates with fine changes in myocardial function and electrophysiology**

*Eva I. Fernlund\*, Todd T. Schlegel#, Pyotr Platonov†, Jonas Carlsson†, Annika Maxedius\* and Petru Liuba\**

\*Paediatric Cardiology Department, Lund University, Lund, Sweden

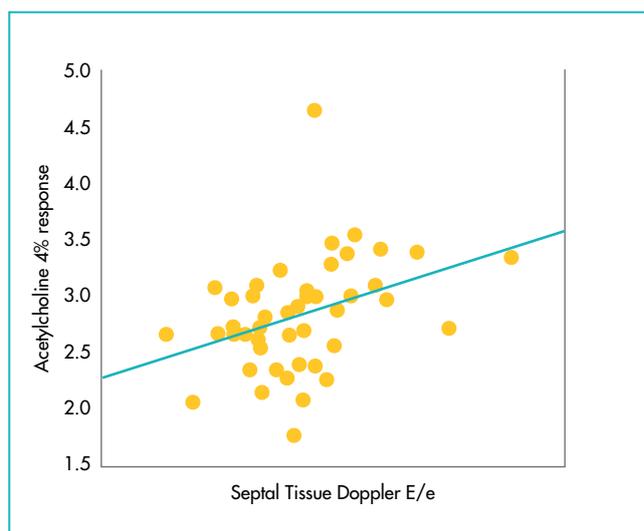
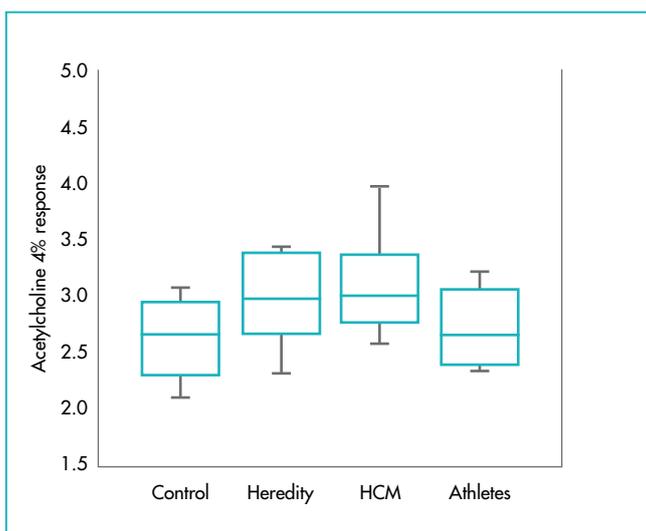
#NASA Johnson Space Centre, Houston, Texas, United States of America

†Cardiology Department, Lund University, Lund, Sweden

**Background:** Recent data from our centre suggested that advanced ECG analysis combined with Tissue Doppler (TD) has increased sensitivity in identifying individuals at risk for developing hypertrophic cardiomyopathy (HCM), and in distinguishing HCM from athlete's heart. Adult patients with HCM appear to have functional abnormalities not only in coronaries but also in peripheral vessels. Whether the latter are present in young individuals, and whether they can be used as additional index for stratifying the risk for HCM have not been yet studied.

**Methods:** The study population, part of a cohort from an ongoing prospective study, consisted of children and adolescents (age 4 - 27, median 13.9 years) with familial HCM heredity, either without left ventricular hypertrophy (HCM-risk: Z-score for IVS and/or PW < 2.5 SD;  $n = 10$ ) and HCM-patients with LV hypertrophy (HCM: Z-score > 2.5 SD;  $n = 10$ ), and compared to age and gender-matched healthy controls ( $n = 20$ ) and athletes ( $n = 10$ , endurance physical exercise > 10 hours/week, Z-score > 2SD). Conventional ECHO, TD, advanced 12-lead ECG (A-ECG) for calculation of spatial mean QRS-T angle by Kors (Cardiax®, IMED Co Ltd, Budapest/Hungary and Houston/USA) were used. Cutaneous micro-vascular responses to acetylcholine (Ach) and sodium nitroprusside were assessed by laser Doppler with iontophoresis.

**Results:** As compared to controls and athletes, the micro-vascular responses to Ach were increased in patients with HCM and LVH, and also in those at risk for HCM (i.e., HCM heredity without LVH;  $p = 0.09$ ). In this latter group, micro-vascular responses to Ach correlated significantly to both spatial mean QRS/T angle by Kors and E/E ratio by TD ( $p < 0.05$ ,  $r > 0.3$ ). With the exception of left ventricular mass (LVM) and myocardial thickness, which were increased in both athletes and HCM patients, there were no differences in any other measured variable between controls and athletes.



**Conclusion:** Independent to the presence of LVH the pre-symptomatic HCM-risk individuals and HCM patients demonstrated enhanced peripheral micro-vascular reactivity, probably reflecting compensatory vasoactive mechanisms in response to HCM-related noxious factors. The enhanced peripheral micro-vascular reactivity are present in both pre-symptomatic HCM-risk individuals and HCM patients, but not in athletes or normal controls, and seem to be associated with fine abnormalities in myocardial electrophysiology and function.

**Abstract no: 1575**

**Complication capture to enhance paediatric cardiac surgery outcomes**

David Maldonado<sup>\*#</sup>, Hayley Burton<sup>#</sup>, Andrew Warren<sup>\*#</sup>, Stacy O'Blenes<sup>\*#</sup> and Camille Hancock Friesen<sup>\*#</sup>

Dalhousie University, Halifax, Nova Scotia, Canada  
 IWK Health Centre, Halifax, Nova Scotia, Canada

**Background:** With current low mortality rates in paediatric cardiac surgery, we can now focus on reducing complications to enhance patient outcomes. In the absence of a risk-adjusted score for morbidity, we have developed a system of tracking programme-wide complications. The system generates a flag at the time of any excess observed:expected (O:E) complications.

**Methods:** Consecutive procedures and 30d complications have been recorded prospectively at the IWK Heart Centre since 2009, using the Multi Institutional Database Committee definitions. Inclusion criterion was a procedure with an assignable Risk Adjusted Congenital Heart Surgery (RACHS) category, which includes 70% of cases. A morbidity score was generated for each procedure factoring in frequency and severity of complications. Observed outcomes were collected (2011 - 2012) and expected outcomes were calculated based on historic complications (2009 - 2011). An overall O:E plot was generated case-by-case. Complications were also stratified by RACHS category and organ-system. Flags were generated to indicate unexpected complication frequency or severity.

**Results:** In 2011 - 12, 116 procedures were performed on 106 patients, 86 of which had an assignable RACHS category. The operative mortality was 5/116 (4.3%). Ninety two complications occurred in 31 procedures. The overall O:E plot was flagged 4 times as a result of 9 complications (arrhythmia, neurologic and pulmonary). The arrhythmia flags were generated by spontaneously resolving arrhythmias in 3/4 cases. The single neurologic flag resulted from a subdural haematoma in a procedure requiring pre-operative ECMO. Pulmonary flags were generated in 4 neonatal procedures (1 reintubation, 4 long term intubation). There was excess morbidity in RACHS category 4 and 6.

**Conclusions:** This complication capture and stratification system allows real-time identification of excess complication occurrence. We anticipate that the ability to detect complications will allow focused changes to improve patient outcomes.

**Abstract no: 1576**

**Dual anti-platelet therapy in paediatric cardiovascular practice**

Srinivas Ananth Narayan<sup>\*</sup>, Elizabeth Orchard<sup>#</sup> and Georgina Stepney<sup>\*</sup>

<sup>\*</sup>Children's Hospital, Oxford, United Kingdom  
<sup>#</sup>Oxford Heart Hospital, Oxford, United Kingdom

**Introduction:** Antiplatelet drugs are increasingly used in paediatric patients to prevent thromboembolic episodes, as an alternative to warfarin. Aspirin remains the most frequently used antiplatelet agent; however there has been an increased interest in the use of newer agents (1). Clopidogrel is an antiplatelet drug that acts by irreversibly modifying the ADP purinergic platelet receptor. Clopidogrel use in children has been reported principally by American centres predominantly in patients with congenital cardiac disease. However, the dosage, duration and indications vary widely. Experience in UK remains very limited.<sup>(1,2)</sup>

**Objective:** The aim of the study was to assess the indication, dosage, duration of therapy and safety of dual antiplatelet therapy with aspirin and clopidogrel in paediatric patients with cardiovascular disease.

**Methods:** The study included consecutive children with cardiovascular disease, presenting over a 2-year period with an indication for anticoagulation (excluding mechanical valve replacement), who were treated with aspirin and clopidogrel. Patient demographics, diagnosis, indication, dose, duration of treatment and adverse events were recorded.

**Results:**

	Median (range) (n=21)
Age	4.08 (0.02 - 13.61) years
Clopidogrel dose	0.3 (0.18 - 1.12) mg/kg
Sex	Male 12 (57%) Female 9 (43%)
Duration of treatment	23.14 (0.57 - 101.3) weeks
Aspirin dose	4.46 (1.6 - 9.44) mg/kg

Indication for use	Number
Systemic-pulmonary artery shunt	7
Fontan	7
Glenn shunt	3
Stent	3
Others	3

There were 2 adverse events:

- 4-year-old boy with gastrointestinal bleed, 7 days after starting clopidogrel at 0.8mg/kg/day. An endoscopy confirmed a gastric ulcer. He was also taking aspirin 75mg (4.17mg/kg) and has now been recommenced on warfarin.
- 6.5-year-old girl with haemorrhagic cerebrovascular accident 5 days after starting clopidogrel at 0.2mg/kg/day. cMRI showed arteriovenous malformations in her brain. Her aspirin was restarted at 50mg (2.83mg/kg) with no further adverse events.

**Discussion:** Combined use of aspirin and clopidogrel is increasing in children. Compared with warfarin, dual antiplatelet therapy has the advantage of ease of administration, with no monitoring required. However there is limited knowledge of the bleeding risks associated with dual antiplatelet therapy in children.<sup>(3)</sup> This study highlights the need for caution and vigilance in treating children with clopidogrel in combination with aspirin, especially at higher clopidogrel doses.

**References:** 1. Y. Finkelstein, L. Nurmohamed, M. Avner, L.N. Benson, G. Koren. Clopidogrel use in children. J Paediatr. 2005 Nov;147(5):657-61.  
 2. C. Gentilomo, Y.S. Huang, L Raffini. Significant increase in clopidogrel use across U.S. children's hospitals. Paediatr. Cardiol. 2010 Feb;32(2):167-75.  
 3. L. Mertens, B. Eyskens, D. Boshoff, M. Gewillig. Safety and efficacy of clopidogrel in children with heart disease. J Paediatr. 2008 Jul;153(1):61-4.

**Abstract no: 1578****Changes in mesenteric flow, heart rate variability and systemic micro-vascular function after total cavopulmonary connection-relationship to pulmonary haemodynamics and post-operative pleural effusion**

Mikaela Dolk\*, Katarina Hanseus\*, Jens Johansson#, Thomas Higgins\*, Annica Maxedius\* and Petru Liuba\*

\*Cardiology, Pediatric Heart Centre, Skåne University Hospital, Lund, Sweden

#Cardiac Surgery, Pediatric Heart Centre, Skåne University Hospital, Lund, Sweden

**Background:** Prolonged pleural effusion (PE) after total cavopulmonary connection (TCPC) remains a serious complication with important clinical and economical impact. Downstream pulmonary action of mediators released in response to abnormal circulation in the gut along with changes in cardiac autonomic nervous activity have been suggested among possible mechanisms. We aimed to assess this hypothesis by studying in a prospective manner the profiles of mesenteric flow, systemic micro-vascular function, and cardiac autonomic function derived from the analysis of heart rate variability (HRV).

**Methods:** Fifteen patients (median age at surgery: 2.1 years) with univentricular heart physiology and previous Glenn surgery were assessed serially before and after TCPC (extracardiac conduit in all patients), until discharge, for superior mesenteric flow velocities, systemic micro vascular function via laser Doppler (baseline and reactive skin perfusion in response to acetylcholine (Ach) and sodium nitroprusside), and for certain HRV indices (standard deviation of the normal-to-normal RR intervals (SDNN), LF and HF; BioMedical Inc, Taiwan).

**Results:** The mesenteric diastolic flow VTI and the micro vascular perfusion and reactivity to Ach decreased post-operatively especially on day 1 after surgery ( $p < 0.05$ ). HRV LF and HF were reduced post-operatively at all time points ( $p \leq 0.05$ ). The mesenteric diastolic flow velocity on day 1 correlated with the pre-operative transpulmonary gradient ( $p = 0.02$ ,  $r = -0.7$ ), and both with the duration of PE ( $p < 0.05$ ,  $r > 0.4$ ). Lower pre-operative values of SDNN were associated with higher pre-operative transpulmonary gradients ( $p = 0.01$ ,  $r = -0.8$ ), and to some extent with PE duration ( $p = 0.06$ ,  $r = -0.6$ ). Neither the demographic variables (age at surgery, time between Glenn and TCPC, weight) nor the heart-lung machine duration were associated with PE duration.

**Conclusion:** TCPC is ensued by abnormalities in mesenteric flow, systemic micro vascular function and cardiac autonomic function. Although several of these variables show some association with PE duration, given the small sample size, further studies are needed to validate these findings and to interrogate the mechanisms.

**Abstract no: 1581****How is the peri-patch myocardium in ventricular septal defect (VSD) patch repair**

Lucy Eun\*, Han Ki Park#, Nam Kyun Kim\* and Jae Young Choi\*

\*Paediatric Cardiology, Yonsei University Severance Cardiovascular Hospital, Seoul, Korea

#Cardiovascular Surgery, Yonsei University Severance Cardiovascular Hospital, Seoul, Korea

**Background:** Strain rate (SR) and strain ( $\epsilon$ ) in tissue Doppler imaging provide new non-invasive measurements of myocardial function, independent of heart motion.

This study assesses the extent of peri-patch regional myocardial function after patch repair of ventricular septal defect (VSD). Myocardial SR and  $\epsilon$  were recorded from the peri-patch myocardium and remote septum from patch area in 20 patients (1 month - 10 years of age, mean 2.6 years). Distance between the patch and the point of returning to remote normal  $\epsilon$  profile was measured. Compared to the remote myocardial region, peri-patch myocardium had decreased peak longitudinal SR ( $-3.3 \pm 1.8 s^{-1}$  vs.  $-4.8 \pm .3 s^{-1}$ ,  $p < 0.05$ ), delayed time to peak longitudinal SR ( $151 \pm 81$  ms vs.  $119 \pm 56$  ms,  $p < 0.05$ ), decreased peak  $\epsilon$  (longitudinal;  $-20 \pm 8\%$  vs.  $-28 \pm 10\%$ , radial;  $20 \pm 14\%$  vs.  $34 \pm 22\%$ ,  $p < 0.01$ ), and delayed time to peak  $\epsilon$  (longitudinal;  $316 \pm 76$  ms vs.  $241 \pm 67$  ms, radial;  $341 \pm 94$  ms vs.  $269 \pm 81$  ms,  $p < 0.0001$ ). The mean distance from the patch to the remote patch  $\epsilon$  curve was  $2.65 \pm 0.77$  mm.

**Conclusion:** Peri-patch myocardium after repair of VSD has delayed and diminished contraction as compared to more remote normal myocardium.

**Abstract no: 1595****Geometric characterisation of 100 patient-specific total cavopulmonary connections and its relation to haemodynamic outcomes**

Elaine Tang\*, Maria Restrepo#, Chris Haggerty#, Lucia Mirabella#, James Bethel#, Kevin Whitehead#, Mark Fogel# and Ajit Yoganathan#

\*School of Chemical and Bio-molecular Engineering, Georgia Tech, Atlanta, Georgia, United States of America

#Department of Biomedical Engineering, Georgia Tech and Emory University, Atlanta, Georgia, United States of America

†Westat, Inc. Rockville, Maryland, United States of America

‡Division of Cardiology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States of America

**Background:** The total cavopulmonary connection (TCPC) anatomies are complex and have great patient-to-patient variability. The geometric characteristics can impact the hemodynamic outcomes, namely: (1) TCPC energy dissipation which can affect ventricular function; and (2) Unbalanced hepatic flow distribution (HFD) which increases the risk of pulmonary arteriovenous malformations (PAVM). In this study, we present a large-scale cohort geometric characterisation of different TCPCs and explore how the anatomic differences translate to the resulting haemodynamic outcomes.

**Methods:** 100 patient-specific TCPC 3-D anatomies were reconstructed from magnetic resonance images (MRI). Diameter, shape factor (SF, minimum/maximum diameter) and tortuosity (vessel length/shortest distance between 2 ends of a vessel) were quantified at each vessel. Angles between connecting vessels and caval offset (distance between the superior vena cava (SVC) and the Fontan pathway) were also quantified. Computational Fluid Dynamics simulations were carried out using time-averaged flow boundary conditions (obtained from phase contrast MRI) to quantify the resulting TCPC energy dissipation index (TCPC-EDI)<sup>(1)</sup> and HFD, which were then correlated to the geometric parameters.  $p < 0.05$  was considered significant for all statistical correlations.

**TABLE 1:** Significant correlations between haemodynamic results and geometric parameters of 100 Fontan patients<sup>#</sup>

Haemodynamic endpoint	Geometric parameter <sup>*</sup>	Correlation coefficient	p-value	
TCPC-EDI	Normalised Fontan pathway diameter (mm/m)	Min.	-0.460	<0.001
		Mean	-0.358	<0.001
		Max.	-0.225	0.02
	Normalised SVC diameter (mm/m)	Min.	-0.388	<0.001
		Mean	-0.270	0.007
	Normalised LPA diameter (mm/m)	Min.	-0.355	<0.001
		Mean	-0.308	0.002
	Normalised RPA diameter (mm/m)	Min.	-0.353	<0.001
		Mean	-0.248	0.01
		Shape factor (mm/mm)	Fontan pathway	-0.434
SVC	-0.314		0.001	
RPA	-0.218		0.03	
Cardiac index (L/min/m <sup>2</sup> )	Normalised min. diameter (mm/m)	Fontan pathway	0.248	0.01
		SVC	0.278	0.005
	Shape factor (mm/mm)	Fontan pathway	0.355	<0.001
Total venous Return/BSA (L/min/m <sup>2</sup> )	Normalised min. diameter (mm/m)	SVC	0.214	0.03
	Shape factor (mm/mm)	Fontan pathway	0.248	0.01
HFD (% towards LPA)	Pulmonary flow distribution + (% towards LPA)		0.308	0.002
		Normalised LPA diameter (mm/m)	Min.	0.216
	Mean		0.334	0.001
	Max.		0.225	0.03
	Angle between (degree)	Fontan pathway & LPA	0.315	0.001
		SVC & LPA <sup>†</sup>	-0.312	0.002
		SVC & RPA	0.327	0.001
	Normalised caval offset (mm/m)	Anterior-posterior <sup>‡</sup>		0.340
Left-right <sup>§</sup>			0.402	<0.001

<sup>#</sup>Spearman correlations were used unless specified otherwise. <sup>\*</sup>Length-scale parameters were normalised by square root of the body surface area (m) of each patient. <sup>†</sup>Pearson correlation was used. <sup>‡</sup>Caval offset was defined negative when the Fontan pathway was posterior of the SVC. <sup>§</sup>Caval offset was defined negative when the Fontan pathway was at the right of the SVC.

**Results:** TCPC-EDI correlated negatively with normalised diameters at all TCPC vessels, and also SFs of SVC, RPA (right pulmonary artery) and the Fontan pathway. Cardiac index was found to have positive significant correlations with Fontan pathway SF, and normalised SVC and Fontan pathway diameters. HFD was found to correlate with the pulmonary flow distribution, normalised PA diameter, caval offset and angles across the connections. No significant correlations were found between tortuosity and the haemodynamic endpoints.

**Conclusions:** Small vessel diameters and SFs are correlated with higher TCPC-EDI and lower cardiac index. This highlights the importance to dilate stenosis in any associated vessel of the connection. Also, PA diameter, caval offset and connection angles together can impact HFD.

**References:** I. L.P. Dasi, et al., Functional analysis of Fontan energy dissipation. Journal of Biomechanics, 2008;41:2246-2252.

**Abstract no: I599**

**Case report: Management of dextrocardia, situs inversus totalis, TAPVC-mixed, complete AV canal defect, common atrium, bilateral SVC, PDA and severe PAH**

**Sivasubramanian Muthukumar<sup>\*</sup>, Charles Jebaraj<sup>†</sup>, Ranjith Karthekayan<sup>#</sup>, Dheeraj Reddy<sup>\*</sup> and Periyasamy Thangavelu<sup>\*</sup>**

<sup>\*</sup>Department of CTVS, Sri Ramachandra University, Chennai, India

<sup>#</sup>Department of Cardiac Anaesthesia, Sri Ramachandra University, Chennai, India

<sup>†</sup>Department of Paediatric Cardiology, Sri Ramachandra University, Chennai, India

**Background and objective:** To present the management of dextrocardia, situs inversus totalis, “mixed type” TAPVC, complete AV canal defect Rastelli Type A, common atrium, bilateral SVC, PDA and severe PAH.

**Diagnosis:** A 9-month-old baby girl was admitted with failure to thrive and respiratory distress. Echo evaluation revealed the above mentioned diagnosis. After pre-operative evaluation, baby was subjected to surgery. Operative finding included “mixed type” TAPVC: Right superior and middle pulmonary veins and left inferior pulmonary vein draining into common venous chamber; Vertical vein from the common chamber joining RSVC; Left superior vein to LSVC; Right upper lobe vein to RSVC; Morphologic RA was on right side receiving RSVC and hepatic vein; Morphologic LA on the left side receiving LSVC and IVC; Unroofed Coronary sinus; Complete AV canal defect; Rastelli Type A with large inlet VSD; severe MR-AML cleft; Common Atrium.

**Result:** The procedure included TAPVC repair and AV canal repair. Rerouting of pulmonary veins was done with wide anastomosis between the common venous chamber and mLA. Vertical was ligated. Since innominate vein was present, LSVC was interrupted, hence diverting left superior vein to mLA. A double-patch technique was used to repair the AV canal. VSD closure, MV repair with AML cleft repair, Tricuspid valve repair, ASD closure with pericardia patch creating an atrial baffle diverting IVC, SVC and hepatic vein to mRA. Core cooling done to 28°C. Total CPB time was 325 minutes and aortic cross-clamp time 235 minutes. Total ventilatory support for 86 hours and inotropes for 6 days. ICU stay was till 8th POD.

**Conclusion:** Baby was discharged on 16th POD with stable haemodynamics and normal oral feeds.

**Abstract no: 1602****Delayed recognition of unilateral pulmonary vein obstruction after repair of cor triatriatum****Jae Hyeon Yu, Nan Yeol Kim, Yeun Ju Lee, Yu Young Jeong, Hyun Kong Oh, Min Woong Kang, Myung Hun Na and Seung Pyung Lim**

Department of Cardiac Surgery, Chungnam National university hospital, Jung-gu, Daejeon, Korea

**Background:** Cor triatriatum is a rare condition where the left atrium (LA) is subdivided by a membrane. Although cor triatriatum can be an isolated lesion, in many cases, it is associated with other congenital cardiovascular anomalies: atrial septal defect, a left SVC, and pulmonary venous anomalies, etc. We report a case of unilateral pulmonary vein stenosis diagnosed 7 years after the repair of cor triatriatum.

**Methods:** A 2-month-old boy was diagnosed with cor triatriatum and severe pulmonary hypertension (PAH). The LA was divided by an obstructing membrane of 2mm opening. The membrane was excised and the post-operative course was uneventful. We recognised that the vascular markings and the volume of the left lung were markedly decreased 7 years after the operation. An examination was done to get detailed information.

**Results:** On the Echocardiogram, the left pulmonary artery (LPA) is relatively small (7mm, Z=-3), but there is no PAH. The lung perfusion scan shows the markedly decreased perfusion on the left (95% vs. 5%). In the cardiac catheterisation, LPA is small and left pulmonary vein is not recognised. RV Pressure is 25/0/7, MPA 25/15/13, wedge pressure of RPA 10, LPA 15 and mean LA pressure is 10mmHg. The 3-DCT shows there are small LPA and no left pulmonary veins.

**Conclusion:** In this case, unilateral pulmonary vein obstruction was hard to diagnose because pulmonary hypertension and symptoms did not appear. We assume that the pulmonary vein stenosis may have been associated with cor triatriatum at the time of surgery, and progressed slowly during the follow-up period. Thus, particular care should be taken to identify such anomalies in patients with cor triatriatum before and after repair.

**Abstract no: 1615****End-point separation between mitral valve and septum over the diastolic left ventricle diameter (EPSS/LVDd) of the long-axis view of echocardiography: An easy, reliable means to assess global LV dysfunction in thalassemia major****Jeng-Sheng Chang\*, Yu-Chin Huang\*, Kang-Hsi Wu# and Ching-Tien Peng#**

\*Paediatric Cardiology, Children's Hospital, China Medical University Hospital, Taichung City, Taiwan

#Paediatric Oncology, Children's Hospital, China Medical University Hospital, Taichung City, Taiwan

**Background:** Myocardial dysfunction is a common complication in patients of Thalassemia major<sup>TM</sup>. Although echocardiography is a convenient instrument, none of its parameters so far has been considered as an ideal surrogate for predicting impending heart failure in TM. However, our previous study using the long-axis, M-mode echocardiographic measurements in TM patients with acute heart failure proved that the distance of endpoint separation between mitral valve and septum (EPSS) and the ratio of EPSS divided by diastolic diameter of the LV (EPSS/LVDd) was significantly correlated with their clinical and laboratory improvements. This study intended to extend the clinical application of EPSS and EPSS/LVDd to TM patients of long term follow-up.

**Material and method:** January 2003 - July 2012, the Thalassemia Registry of China Medical University Hospital, Taiwan, enrolled 34 TM patients for daily oral iron chelation therapy and echocardiographic studies, which were performed every 6 months - 1 year by 2 double-blinded technicians. The normal ranges of EPSS (2.5±1.7mm) and EPSS/LVDd (0.08±0.06) were used as references. Compared to the baseline data at enrolment, the trends of changes in EPSS, LVDd and EPSS/LVDd were assessed by the Generalised Estimation Equation (GEE).

**Result:** Their average age was 13.4±3.7 years at enrolment and the average follow-up duration was 7.2±2.0 years. In accordance with improvements of EPSS/LVDd, favourable response to iron chelation occurred in 24 of the 34 patients (13 with direct improvement, 5 stayed as normal and 6 showed delayed improvement after initial worsening), and unfavourable response occurred in 10 patients (5 remained as abnormal and 5 even deteriorated). GEE analysis showed a general trend of rising EPSS/LVDd after 5 years of follow-up (p<0.01).

**Conclusion:** EPSS/LVDd is a convenient parameter to monitor the global cardiac function of TM patients.

**Abstract no: 1616****Outcomes of implantable cardioverter-defibrillator use in paediatric and congenital heart disease patients: The North West experience****Atul Kalantre, Abbas Khushnood, Arjamand Shauq, Adam Fitzpatrick, Derrick Todd, Mark Hall and Edmund Ladusans**

Alder Hey Children's Hospital, Liverpool, United Kingdom

**Objective:** We reviewed our experience over 20 years of the use, complications and outcomes of implantable cardioverter defibrillators (ICD) in a large paediatric population.

**Method:** Retrospective analysis of 35 patients requiring ICD insertion from 1993 - 2012 at a single tertiary paediatric cardiac centre. Indications for implant were: long QT syndrome (n=16), catecholaminergic polymorphic ventricular tachycardia (n=5), hypertrophic obstructive cardiomyopathy (n=5), idiopathic ventricular tachycardia (VT) (n=3), VT with underlying congenital heart disease (n=4), Brugada (n=1), Arrhythmogenic right ventricular dysplasia (n=1).

**Results:** There were 38 ICD implantations in 19 males and 16 females. Median age at presentation was 10 years (range 5 - 20 years) and median weight 37kg (range 21 - 70 kg). The patients presented as follows: cardiac arrest (15), syncope (17), pre-syncope (1), palpitations (1) and seizure (1). Concomitant antiarrhythmic drug therapy was with beta-blocker alone (10), beta-blocker and amiodarone (2) and amiodarone only (1). A single chamber ICD was used in 18 and dual chamber ICD in 20. Median age at implant was 11 years (range 6 - 23). On follow-up (median 4 years, range 0.5 - 12 years), monitoring revealed 7 episodes (18%) of appropriate shocks and 9 (24%) of inappropriate shocks due to lead failure (n=4), oversensitivity (n=2) and sinus/atrial tachycardia (n=3). Twenty two (58%) patients did not receive any shocks. There were 9 lead replacements, 3 of which were elective due to box change. There were 2 episodes of infection; 1 minor and 1 needing box change. 2 (5%) patients died on follow-up, 1 with a head injury after collapse secondary to VT successfully cardioverted and 1 with catecholamine storm and repeated cardioversions.

**Conclusion:** ICD implantation in the paediatric population can be life-saving but is associated with a significant incidence of inappropriate shock and requirement of generator and electrode replacement.

**Abstract no: 1619****Aetiology of pulmonary hypertension in Africa: Preliminary data analysis after 1-year of recruitment: The Pan African pulmonary hypertension cohort study (PAPUCO)**

**Friedrich Thienemann<sup>\*,#†</sup>, Lori Blauwet<sup>‡</sup>, Anastase Dzudie<sup>§</sup>, Kamilu M. Karaye<sup>¶</sup>, Sani Mahmoud<sup>¶</sup>, Amam Mbakwem<sup>Δ</sup>, Patience Udo<sup>Δ</sup>, Ana O. Mocumbi<sup>∇</sup>, Karen Sliwa<sup>\*,#\*</sup> and PAPUCO Research Group\***

\*Institute of Infectious Diseases and Molecular Medicine, University of Cape Town, Observatory, South Africa

#Department of Medicine, University of Cape Town, Observatory, South Africa

†Infectious Diseases Referral Clinic, G.F. Jooste Hospital, Manenberg, South Africa

‡Division of Cardiovascular Diseases, Mayo Clinic, United States of America

§Douala General Hospital, Doula, Cameroon

¶Aminu Kano Teaching Hospital, Kano, Nigeria

ΔLagos University Teaching Hospital, Nigeria

∇Cardiology Unit, University of Uyo Teaching Hospital, Nigeria

∞Instituto do Coração, Maputo, Moçambique

\*Hatter Institute for Cardiovascular Research in Africa, University of Cape Town, Observatory, South Africa

**Background:** Pulmonary Hypertension (PH) is a devastating, progressive disease, with increasingly debilitating symptoms and, usually, shortened overall life expectancy. The epidemiology of PH in Africa has not yet been determined, but limited reports suggest that the incidence is higher than that reported from developed countries. Many known factors for PH are hyper-endemic in this part of the world, including HIV/AIDS, rheumatic heart disease, schistosomiasis and Sickle Cell Disease. We aim to describe the aetiology of PH in Africa.

**Materials and methods:** A prospective observational study of patients with newly diagnosed and previously untreated PH based on echocardiography. Fifteen cardiovascular centres from Sub-Saharan Africa participate in this study. Preliminary data analysis after 1 year of recruitment is presented.

**Results:** A total of 63 patients were recruited within the 1st year of the study. Median age was 40 years (range 11 - 86) and 62% were female. 24% of patients were living below the WHO poverty line of 1 US\$ per day. 37% of patients were known HIV positive with a median CD4 count of 401 cells/μL (IQR 226-537 cells/μL) and 83% were on antiretroviral therapy on presentation. 32% of patients had documented previously treated TB with site of TB being pulmonary in 85% of documented cases. 50% of those had at least 2 or more documented episodes of TB. At presentation, median pulse at rest was 94bpm (IQR 78 - 100bpm) and pulse oximetry was 95% (IQR 91 - 98%); respiration rate was 26bpm (IQR 21 - 32bpm). On Echocardiography, median RVSP was 56mmHg (IQR 48 - 68mmHg), TAPSE 14mm (11 - 17mm) and LV-EF 50% (35 - 65%). 33% of patients died within 6 months after presentation.

**Conclusion:** PH in Africa is an acquired cardiovascular disease of high mortality and of multiple aetiologies. HIV seems to be an important risk factor for PH in Africa.

**Abstract no: 1620****Impact of surgery by a visiting team on quality of life indicators in patients with congenital or acquired heart disease in nations with limited healthcare resources**

**Aubyn Marath<sup>‡</sup>, Laura Schiff<sup>¶</sup>, Chrisanna Gustafson<sup>Δ</sup>, Janine Henson<sup>§</sup>, Van-Trang Nguyen<sup>†</sup>, John Connett<sup>#</sup>, Kyle Rudser<sup>#</sup>, Susan Lowry<sup>#</sup>, Lazaros Kochilas<sup>\*</sup> and James St. Louis<sup>\*</sup>**

\*Amplatz Children's Hospital, University of Minnesota, Minneapolis, United States of America

#Biostatistics and Informational Technology, University of Minnesota, Minneapolis, United States of America

†University of Minnesota, Minneapolis and CardioStart International, United States of America

‡Oregon Health & Sciences University, Oregon and CardioStart International, United States of America

§Fairview Hospital, University of Minnesota, Minneapolis, and CardioStart International, United States of America

¶Enloe Medical Centre, Chico, California and CardioStart International, United States of America

ΔCalifornia State University, Chico, California and CardioStart International, United States of America

**Hypothesis:** Surgery by a visiting team improves quality of life indicators in patients with congenital or acquired heart disease in nations with developing healthcare programmes.

**Method:** CardioStart International's visiting surgical teams interviewed patients via translator (n=198) in Peru, Ghana, Vietnam, and Grenada from February 2008 - November 2011. Ages ranged from 24 days - 80 years old. Data was collected regarding symptoms, income, perception of health, and emotional status. Parents/guardians were interviewed if the child was unable to answer questions. Follow-up interviews were conducted with patients who had undergone surgery in prior years (n=33).

**Results:** For surgical candidates, the most common symptoms were shortness of breath (46.5%, 92/172) and fatigue (46.0%, 91/172). 57.1% (76/133) of patients had their symptoms for >1 year. Half (50.4%, 71/141) reported a household income loss of 50% or greater since diagnosis of the heart condition. In comparison with others, 51.6% (79/153) reported their health as fair or poor. 20.6% (29/141) felt fears and 36.4% (43/118) felt concern for the future related to the heart condition often or almost always.

**Follow-up data:** After surgery, 63% (17/27) revealed no symptoms; 11.1% (3/27) reported shortness of breath; 11.1% (3/27) reported fatigue. For income, 72% (18/25) reported no loss or an improvement in income since the operation. 81.5% (22/27) reported their health as good, very good or excellent compared to others; 8.7% (2/23) felt fears and 20% (4/20) felt concern for the future related to the heart condition often or almost always.

**Conclusions:** Surgery to correct congenital and acquired heart disease has the potential to improve quality of life in these diverse populations. Lack of healthcare infrastructure in these countries made it difficult to obtain follow-up data. More follow-up data is needed to make reliable inferences, and a more efficient implementation of follow-up is in progress.

**Abstract no: I626**

**Prevalence and clinical impact of major aortopulmonary collaterals in post-operative patients with transposition of the great arteries**

**Vikas Satwik, Raimonda Stukiene, James Gnanapragasam, Kevin Roman, Gruschen Veldtman, Tony Salmon, Nicola Viola, Markku Kaarne and Joseph Vettukatil**

Southampton University Hospital, Southampton, United Kingdom

**Background:** Transposition of the great arteries with intact ventricular septum (TGA) is rarely associated with clinically significant aortopulmonary collateral arteries (MAPCA). The known associated complications are: congestive heart failure, pulmonary volume overload, left ventricular dysfunction and respiratory failure. The aim of this study is to evaluate the prevalence and clinical impact of MAPCA's in post-operative patients with TGA.

**Methods:** All patients with TGA admitted after arterial switch procedure for a period of 7 years from July 2005 - August 2012 were included in the study. Data was analysed from PICU database regarding duration of ventilation, length of stay and outcome. Data from the cathlab was obtained to identify infants with TGA who had undergone catheter occlusion of MAPCA.

**Results:** A total of 48 cases were identified with TGA. Three haemodynamically significant cases requiring catheter occlusion of MPACA identified on echocardiography were identified. 2 patients had difficulty in weaning ventilation and 1 presented with pulmonary haemorrhage. The details are in Table 1.

**TABLE 1:** Outcome of TGA in relation to ventilation, PICU stay and mortality.

	d-TGA with IVS and no MAPCA	d-TGA with IVS and MAPCA
Total no: 49	45	3
Mortality	1	0
Mean ventilation in days (range)	3.8 (1- 6)	10 (5 - 14)
Mean PICU stay in days (range)	5 (2 - 9)	14 (12 - 22)

The mean duration of ventilation post coiling was 2 days (range 1 - 3 days). No coronary abnormality or ischaemic changes were identified on ECG in any of the 48 cases except in 1 who died.

**Conclusion:** TGA with MAPCA's is associated with prolonged ventilation, pulmonary hypertension (PHT) and pulmonary haemorrhage. There is a higher than expected incidence of clinically significant MPACA in our series and we report the 1st case of post-operative pulmonary haemorrhage in this setting. TGA with prolonged ventilation, persistent PHT, or pulmonary haemorrhage warrants careful evaluation for MAPCAs.

**Abstract no: I633**

**Pulmonary valve growth following valve-sparing Tetralogy of Fallot repair**

**Adrian Dyer, Alan Nugent, Kristine Guleserian, Matthew Lemler, Shannon Blalock, Claudio Ramaciotti and Joseph Forbess**

University of Texas South western Medical Centre, Dallas, Texas, United States of America

**Background:** There has been an increased focus on pulmonary valve (PV) preservation in Tetralogy of Fallot (TOF) repair. Valve-sparing (VS) techniques utilise a combined transatrial and transpulmonary approach, a limited right ventriculotomy, sharp commissurotomies and valvar/annular balloon dilation. The objective of this study is to describe the VS ToF repair experience in terms of valve growth and performance.

**Materials and methods:** Retrospective review of pre-operative echocardiograms in all patients who underwent repair of isolated ToF from 2004 - 2012. PV annulus (PVA) dimensions were assessed on immediate post-repair and most recent echocardiogram studies in those undergoing a VS approach.

**Results:** Ninety seven patients were identified. Fifty nine (61%) underwent a VS repair at a median age of 4.1 months and 38 had a transannular patch (TAP) at a median age of 3.5 months (p=0.5). Mean pre-operative PVA diameter was 7.4mm (Z-score, -1.7) in the VS group vs. 5.5mm (Z-score, -3.2) in the TAP group (p<0.0001). In the VS group, the mean post-operative PVA was 9.7mm (Z-score of -0.2; p<0.0001). Mean post-operative peak RVOT velocity (Vmax) was 2.1m/s and 93% had <mild pulmonary insufficiency (PI). Intra-operative pulmonary balloon valvuloplasty was performed in 10 patients. In this group, mean PVA increased from 6.9mm (Z-score, -2.1) - 9.1mm (Z-score, -0.8) and post-operative Vmax was 2.3m/s with 80% having <mild PI. Nineteen patients are followed at this institution with a mean follow-up of 3.7 years. In this subset, the mean post-operative PVA grew from 9.6mm - 14.9mm (p<0.0001), while maintaining normal Z-scores (-0.3 - -0.4), and no increase in the minimal pulmonary stenosis (PS) or PI.

**Conclusions:** VS ToF repair maintains the integrity of the PV and allows ongoing annular growth with minimal PS or PI. This technique may prevent the long term sequelae of right ventricular volume overload secondary to chronic pulmonary insufficiency.

**Abstract no: 1635**

**Diagnosis and assessment of subclinical rheumatic valve disease**

**P.G. Herbst, A.J.K. Pecoraro, Z. Cassimjee and A.F. Doubell**

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

**Background:** The pathophysiologic hallmark of rheumatic valve disease is valvulitis which causes commissural fusion. The sequelae of commissural fusion give rise to the morphology and physiology of rheumatic valve disease. This is neglected in the current WHO and recently proposed combined criteria (CC) for the diagnosis of subclinical rheumatic mitral valve disease.

**Methods:** Patient selection was based on a keyword search of echocardiographic reports and prospective enrolment of the first 15 patients fulfilling criteria of each of 5 cohorts. Cohort 1: Mitral Stenosis (MS); Cohort 2: Rheumatic Heart Disease (RHD) as defined by CC but without MS; Cohort 3: Dilated cardiomyopathy (DCM) with leaflet restriction due to tethering; Cohort 4: Normal controls; Cohort 5: Mitral valve prolapse (MVP). All patients had transthoracic echocardiography with direct commissural assessment and assessing secondary features of commissural fusion. We also tested the sensitivity and specificity of the WHO and CC against our commissural fusion criteria (CFC).

**Results:** Features of commissural fusion was observed in all cohort 1 and 2 patients (n=30). All patients with features of commissural fusion had AMVL diastolic bowing. No AMVL diastolic bowing was observed in cohort 3 - 5. Compared to cohorts 3 - 5, cohort 2 patients had significantly smaller mitral valve area (5.68cm<sup>2</sup> vs. 5.72cm<sup>2</sup> vs. 6cm<sup>2</sup> vs. 3.36cm<sup>2</sup>; p=0.01). The CFC had superior sensitivity and specificity in detecting RHD in cohort 1 - 5 when compared to the WHO and CC.

**Conclusion:** Commissural fusion and the secondary feature of AMVL bowing during diastole are typical morphologic features of rheumatic mitral involvement. Addition of these features should improve the sensitivity and specificity of current criteria for the diagnosis of subclinical rheumatic mitral valve.

**Abstract no: 1643**

**Concerning ocular side effect possibly associated with PDE5 inhibitor in an infant affected by complex congenital heart disease**

**Alessandra Cristofaletti<sup>\*</sup>, Marcella Gaffuri<sup>#</sup>, Maria Antonia Prioli<sup>\*</sup>, Caterina Mansoldo<sup>†</sup>, Luca Barozzi<sup>‡</sup>, Giorgio Marchini<sup>†</sup>, Giovanni Battista Luciani<sup>‡</sup> and Paolo Biban<sup>#</sup>**

<sup>\*</sup>Department of Medicine, Section of Cardiology, University of Verona, Verona, Italy

<sup>#</sup>Department of Pediatrics, Pediatric and Neonatal Intensive Care Unit, Azienda Ospedaliero, Parma, Italy

<sup>†</sup>Eye Clinic, Department of Neurological and Visual Sciences, University of Verona, Italy

<sup>‡</sup>Division of Cardiac Surgery, University of Verona, Verona, Italy

We report a case of bilateral blindness possibly associated with PDE5-inhibitor intake (sildenafil) in a 7-month-old baby affected by a complex congenital heart defect.

Cavo-pulmonary anastomosis was performed at 4 months of age for doublet inlet left ventricle, hypoplastic right ventricle, bulbo-ventricular foramen and moderate subpulmonary obstruction.

Post-op period was uneventful, except for the occurrence of chylothorax, which was treated with multiple drainages, parenteral nutrition and diuretics. Echocardiographic evaluation showed satisfactory results at the anastomosis level, with good left ventricle and atrioventricular valve function.

Nonetheless, intermittent episodes of chylothorax were still present after 2 months. Thus, despite the lack of obvious echographic signs of pulmonary hypertension, we raised the hypothesis that a relative increase of pulmonary vascular resistances might have been involved in the etiology of the persistent chylothorax. Then, ex adjuvantibus, we decided to start sildenafil at 0.2mg/kg TDS. Four weeks later a lack of visual focus on moving objects was noticed, with poor pupillary light reflex. An ophthalmologic examination revealed bilaterally: pendular nystagmus, absent papillary reflex, transparent crystalline lens, light pallor papilla, arterial venous tortuous vessels, peripapillary retinal haemorrhages and macular exudation. The sudden onset of visual loss associated with light pallor optic disc, in the absence of other neurological and orbital abnormality, prompted the diagnosis of posterior ischemic optic neuropathy (PION).

Sildenafil was immediately suspended, while a trial with methylprednisolone intravenously was attempted for the treatment of PION.

One month later, complete reduction of retinal hemorrhages, macular exudation and arterial venous tortuosity was noticed, however visual evoked potentials were absent. At one year follow-up no visual recovery was observed, with further worsening of bilateral optic disc atrophy.

This alarming case raises questions about the safety of sildenafil in young infants and the necessity of ongoing ophthalmologic evaluations before and during therapy with PDE5 inhibitors.

**Abstract no: 1647**

**Overview of paediatric dilated cardiomyopathy in a tertiary hospital**

**Joana Miranda<sup>\*</sup>, Esmeralda Rodrigues<sup>#</sup>, Teresa Vaz<sup>\*</sup>, Elisa Leão Teles<sup>#</sup>, Maria João Baptista<sup>\*</sup>**

<sup>\*</sup>Paediatric Cardiology Department, Hospital St. João, Porto, Portugal

<sup>#</sup>Metabolic Diseases Unit, Paediatric Department, Hospital St. João, Porto, Portugal

**Background:** Dilated Cardiomyopathy (DCM) is the most common form of cardiomyopathy in paediatric population and an important cause of heart transplantation in children.

**Objective:** To characterise the paediatric population diagnosed with DCM in the reference centre of Paediatric Cardiology in the north of Portugal.

**Methods:** Retrospective review of patients diagnosed with DCM from January 2005 - June 2012, including demographic data, clinical presentation, aetiology, treatment and outcome.

**Results:** We identified 61 patients with DCM (37 female; 24 male). Median age at diagnosis was 16 months. Heart failure was present in 83.6% (n=51) of patients and 44.3% (n=27) needed intensive care. Median left ventricular ejection fraction at clinical presentation was 32.0% (SD, 13.0%). The most prevalent causes were idiopathic (n=29 [47.5%]), viral myocarditis (n=11 [18.0%]) and inherited metabolic diseases (n=7 [11.5%]). In viral myocarditis Parvovirus B19 was the most common identified agent. Anti-congestive agents, angiotensin-converting enzyme inhibitor and anti-aggregant therapy were used in the majority of patients. Overall mortality was 16.1% (n=10) and 5 patients underwent heart transplantation. Age <1 year old was associated with higher mortality (p<0.001) and increased need of intensive care (p<0.05). Higher rate of transplantation was observed in children >1 year old (p<0.05) and with intensive care admission (p<0.05). We report no mortality in the 5 patients who underwent heart transplantation after 1-year follow-up.

**TABLE 1:** DCM aetiology and respective mortality.

DCM aetiology	Incidence		Mortality
	n	%	n
<b>Total</b>	<b>61</b>		<b>10</b>
Idiopathic	29	47.5%	7
Myocarditis Parvovirus B19 (8); CMV (2); Influenza-I (1)	11	18.0%	
Inherited metabolic diseases Mitochondrial citopathy (3); LCHADD (2); Mucopolysaccharidosis (2)	7	11.5%	1
Coronary artery anomalies	3	4.9%	1
Left ventricular non-compaction	3	4.9%	
Neuromuscular diseases	2	3.3%	
Tachycardiomyopathy	2	3.3%	
Polymalformative syndrome	1	1.6%	
Familial DCM	1	1.6%	
Maternal systemic lupus erythematosus	1	1.6%	
Doxorubicin toxicity	1	1.6%	1

**Discussion:** The most important prognosis factors identified were age at DCM diagnosis and requirement for intensive care. Although the majority of patients had an idiopathic form of DCM, a significant number of patients presented inherited metabolic diseases. This reinforces the importance of considering it in differential diagnosis of paediatric DCM, as an appropriate approach of inherited metabolic diseases could noteworthy alters the outcome of affected patients.

**Abstract no: 1653**

**Ventricular septal defect: An unusual sequel of blunt chest trauma in a 7-year-old boy**

**Ogunkunle Oluwatoyin, C.O. Duru, B.E. Adebayo and S.I. Omokhodion**

University College Hospital, Ibadan, Nigeria

**Background/hypothesis:** Ventricular septal defect (VSD) is the commonest congenital cardiac lesion encountered worldwide. Only very rarely is it acquired and causation through blunt injury is even rarer.

**Materials And methods:** A 7-year-old boy suffered blunt trauma to his chest while at play with his peers at school. He had been quite well before then, with no symptoms referable to the cardiovascular system and with no growth or developmental delay. He presented 2 days later at our centre with features of acute congestive cardiac failure. Two-dimensional and Doppler echocardiographic examination revealed a rent in the peri-membranous portion of the outlet ventricular septum with the avulsed flap still attached to the septum.

**Results:** Cardiac failure was refractory to anti-failure therapy and other stabilisation measures. His clinical condition took a rapid turn for the worse and he succumbed to the ensuing illness five days after the trauma, despite the stabilisation measures and before any surgical intervention could be undertaken.

**Conclusions:** Traumatic VSD, though rare, should be considered in cases of acute congestive cardiac failure in otherwise well active children with a history of blunt trauma to the chest and all such patients should undergo careful echocardiographic evaluation. Prompt surgical intervention has been reported to be life-saving in similar cases.

**Abstract no: 1655**

**Effect of spironolactone, furosemide and captopril on blood electrolyte levels in congenital heart defect infants with uncontrolled congestive heart failure**

**Alev Kiziltas, Nazan Ozbarlas, Sevcen Erdem and Osman Kucukosmanoglu**

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

**Objective:** The aim of this study was to evaluate the effect of spironolactone, furosemide and captopril on blood electrolyte levels in congenital heart defect infants with uncontrolled congestive heart failure.

**Material and method:** We examined 27 infants with congestive heart failure treated with furosemide and captopril for at least 1 month at Cukurova University Department of Paediatric Cardiology. Before the spironolactone treatment and after the 1st week of the treatment blood sodium, potassium, urinary creatinine levels were checked. The dosage of spironolactone was determined as 1-3mg/kg/day. Group 1 was treated with a dosage of spironolactone ≤2mg/kg/day; and Group 2 received 2-3mg/kg/day.

**Results:** Seventeen patients (63%) were classified as Group 1 and 10 (37%) in Group 2. After the 1st week of the treatment, mean blood sodium level was 134mmol/L, mean blood potassium level was 4.6mmol/L. Hypo-natremia occurred in 12 patients overall. There was a statistical difference in the mean blood sodium levels between Group 1 and 2 ( $p<0.02$ ). There was no statistical difference of the potassium levels between the 2 groups and hyper-potassemia occurred none of the patients. There was no statistical difference in the development of hypo-natremia between the 2 groups.

**Conclusion:** There is no safe dosage range or safe combination protocol of congestive heart failure treatment in paediatric patients. Because of the side-effects of natriuresis and hyperpotassemia occurring clinical and laboratory monitoring needs to be combined in therapy. We didn't encounter hyperpotassemia in different spironolactone dosages. There was no statistical upgrade of potassium levels between groups receiving different spironolactone dosages. Hypo-natremia improved quickly with spironolactone dose reduction or give up. In this prospective pilot study there was no statistical difference in potassium levels with spironolactone treatment, hyperpotassemia didn't occur even in cases with high dosages. But the occurrence of hypo-natremia in 44% of the patients was a remarkable result. In conclusion, frequent electrolyte monitoring is needed when treating congestive heart failure with spironolactone added to furosemide and captopril.

**Abstract no: 1657**

**Permanent cardiac pacing in children: A multicentre study on choosing the optimal pacing-site**

**Jan Janousek<sup>\*</sup>, Irene van Geldorp<sup>#</sup>, Sylvia Krupickova<sup>†</sup>, Eric Rosenthal<sup>‡</sup>, Maren Tomaske<sup>§</sup>, Andreas Frueh<sup>¶</sup>, Jan Elders<sup>▲</sup>, Anita Hiippala<sup>∇</sup>, Frits Prinzen<sup>∇</sup> and Tammo Delhaas<sup>\*</sup>**

<sup>\*</sup>Children's Heart Centre, University Hospital Motol, Prague, Czech Republic

<sup>#</sup>Paediatric Cardiology, CRIM, Maastricht, The Netherlands

<sup>†</sup>Children's Heart Centre, University Hospital Motol, Prague, Czech Republic

<sup>‡</sup>Evelina Children's Hospital, London, United Kingdom

<sup>§</sup>University Children's Hospital, Department of Cardiology, Zurich, Switzerland

<sup>¶</sup>Oslo University Hospital, Oslo, Norway

<sup>▲</sup>Radboud University Medical Centre, Department of Cardiol, Nijmegen, The Netherlands

<sup>∇</sup>Department of Pediatric Cardiology, Children's Hospital, Helsinki, Finland

<sup>∇</sup>Department of Physiology, CRIM, Maastricht, The Netherlands

<sup>\*</sup>Department of Biomedical Engineering, CRIM, Maastricht, The Netherlands

**Purpose:** We evaluated the effects of pacing-site on left ventricular (LV) synchrony and function in children requiring permanent pacing.

**Methods:** 178 children (age <18 years) from 21 centres with complete AV block and a structurally normal heart undergoing permanent pacing were cross sectionally studied. Median age at evaluation was 11.2 (inter-quartile range (IQR) 6.3 - 15.0) years. Median pacing duration was 5.4 (IQR 3.1 - 8.8) years. Data were analysed in a core lab. Pacing-sites were the free wall of the right ventricular (RV) outflow tract (RVOT, n=8), lateral RV (RVLat, n=44), RV apex (RVA, n=61), RV septum (RVS, n=29), LV apex (LVA, n=12), LV mid-lateral wall (LVLat, n=17) and LV base (LVB, n=7).

**Results:** LV synchrony, pump function (ejection fraction (EF), end-systolic volume index and change in shortening fraction as compared to pre-implantation values) and contraction efficiency were significantly affected by pacing-site and were superior in children paced at LVA/LVLat. LV dyssynchrony assessed by radial strain correlated inversely with LV EF ( $r=0.80$ ,  $p=0.031$ ). Pacing from RVOT/RVLat predicted decreased LV function (LV EF <45 %; OR 5.19 CI 1.74-15.50,  $p=0.003$ ) whereas LVA/LVLat pacing was associated with preserved LV function (LV EF >55 %; OR 6.97, CI 2.21-22.00,  $p<0.001$ ). Age at implantation, pre-implantation LV size and function, duration of pacing, DDD mode, QRS duration and presence of maternal auto-antibodies had no significant impact in a multivariable analysis.

**Conclusions:** LV mechanical synchrony, pump function and contraction efficiency may significantly deteriorate with RVOT/RVLat pacing and are best preserved with LVA/LVLat pacing.

**Abstract no: 1663**

**Outcomes of children and adolescents undergoing invasive testing for asymptomatic Wolff-Parkinson-White pre-excitation**

**Jan Janousek<sup>\*</sup>, Peter Kubus<sup>\*</sup>, Pavel Vit<sup>#</sup>, Roman Gebauer<sup>†</sup> and Ondrej Materna<sup>\*</sup>**

<sup>\*</sup>Children's Heart Centre, University Hospital Motol, Prague, Czech Republic

<sup>#</sup>Paediatric Cardiology, Children's University Hospital, Brno, Czech Republic

<sup>†</sup>Department of Paediatric Cardiology, Heart Centre, Leipzig, Germany

**Objective:** Results of a risk stratification strategy in asymptomatic (WPW) pre-excitation in the young were evaluated.

**Methods:** Eighty five consecutive patients aged <18 years with a WPW pattern (with persistent pre-excitation at maximum exercise) undergoing an invasive electrophysiologic study for risk stratification was retrospectively evaluated. Potentially dangerous accessory pathway (AP) properties were defined as any of the following: antegrade effective refractory period  $\leq 250$ ms, shortest pre-excited RR interval during atrial fibrillation/rapid atrial pacing  $\leq 250$ ms, inducible atrioventricular re-entrant tachycardia (AVRT) or presence of multiple APs.

**Results:** Median age at evaluation was 14.9 (IQR 12.5 - 16.6) years. Eighty two patients had a structurally normal heart and 3 had hypertrophic cardiomyopathy. A single manifest AP was present in 80 patients, 1 manifest and 1 concealed AP in 4 and 2 manifest APs in 1 patient. Potentially dangerous AP properties were present in 33/85 patients (38.8%) at baseline and in an additional 16/44 (36.4%) of those subjected to isoproterenol challenge. Ablation was performed in 41/49 patients with a potentially dangerous AP and deferred in the remaining 8 for proximity to the atrioventricular node. In addition, 18 low-risk patients were ablated based on patients'/parental decision.

**Conclusion:** 39% of patients evaluated with asymptomatic WPW pre-excitation persisting at peak exercise exhibited potentially dangerous AP properties at baseline. Isoproterenol challenge yielded an additional 36% of those tested at risk. Ablation was performed in 69% of patients.

**Abstract no: 1664**

**Using cMRI as primary inter-stage diagnostic screen prior to surgical completion of total cavopulmonary connection in patients with functionally single ventricles: A single institutional experience**

**Henning Clausen\*, Emma Stockton\*, Michael Broadhead\*, Oliver Tann\*, Vivek Muthurangu\*#, Troy Dominguez\*, Andrew Taylor\*# and Marina Hughes\*#**

\*Great Ormond Street Hospital for Children, London, United Kingdom

#University College London, London, United Kingdom

**Background:** Cardiac magnetic resonance (cMRI) is proposed as a sensitive diagnostic tool to guide decision-making after bidirectional cavopulmonary connection (BCPC) and prior to total cavopulmonary connection (TCPC). Our unit has adopted a protocolled, CMR-based screening method, performed under general anaesthetic with transduced central venous pressure (CVP) measurement from the internal jugular vein.

**Methods:** Retrospective review of single-centre, medium-term TCPC experience in relation to pre-operative cMRI data. Non-parametric statistical methods were used. Results expressed as median (interquartile range).

**Results:** The cohort included 192 local patients undergoing BCPC, since 2005. Of those, 86 have undergone TCPC at age 4.0 (3.3 - 4.6) years. 47 (55%) had right ventricular dominance. Weight at surgery was 15.1 (13.6 - 16.9) kg. 56 (65%) were non-fenestrated at the time of initial surgery. 76 (88%) operated patients are alive without heart transplantation, with duration follow-up 2.2 (1.1 - 3.7) years. Interstage cMRI was carried out in 55 (64%) of the 86 operated patients. The remaining patients required early investigation and intervention following BCPC, and underwent out-of-protocol CT scan or cardiac catheterisation. These patients experienced a 2.9 fold mortality; 6/31 (19.4%) deaths occurred in this group, compared with 4/61 (6.6%) deaths in the cMRI group. Of those undergoing cMRI median CVP was 12 (11 - 14) mmHg. Patients with CVP >13 had a greater proportion of pulmonary venous return contributed by systemic-to-pulmonary collateral flow (median 32% vs. 42%, p<0.05). The CVP measured at cMRI was not significantly related to pulmonary artery stenosis/hypoplasia, and did not predict length of hospital stay or mortality. However, mortality following TCPC was strongly associated with prominent venous channels visible on MR angiography, and off-loading superior vena cava (SVC) into inferior vena cava (IVC) territory (p<0.05).

**Conclusions:** Interstage cMRI prior to TCPC offers a comprehensive assessment of morphology, physiology, can elicit risk factors of post-operative outcome, and may identify confounding factors for pulmonary artery pressure, such as SVC to IVC collateralisation.

**Abstract no: 1669**

**Spot the difference: Can you transfer 1.5 T reference values to the 3 T Era?**

**Marcus Fischer\*, Hermann Korperich#, Deniz Kececioglu\* and Kai Thosten Laser\***

\*Centre of Congenital Heart Disease, Heart and Diabetes Centre., Ruhr University of Bochum, Bad Oeynhausen, Germany

#Institute for Radiology, Nuclear Medicine and Molecular Imaging, Heart and Diabetes Centre, Ruhr University of Bochum, Bad Oeynhausen, Germany

**Background:** Cardiac MRI is important in the treatment of children with congenital heart disease. It is reference standard for the assessment of ventricular dimensions and function. Most published reference values were obtained by 1.5 T MR scanner

**Methods:** Quantitative volumetric cardiac MR measurements were performed on a 3T TX MRT (Philips ACHIEVA) and a 1.5T MRT (Philips Intera) using a multi-slice multi-phase steady-state free precession gradient-echo acquisition in breath hold (TR/TE/flip=2.9ms/1.45ms/40°; Matrix=1.4 - 1.5x1.5 - .7mm²; 22 - 30 phases, 5mm slice thickness). Patient sample included 17 healthy persons (7 male, 10 female, mean age 13.5 years ± 4.3a; range 6 - 20a). Calculated stroke volume was controlled by flow-derived stroke volumes using phase-contrast MRI. Data were quantified by single expert. F-Test and unpaired T-Test was performed.

**Results:** There were no significant differences for both, left and right ventricle.

	LV-EDV	LV-ESV	RV-EDV	RV-ESV
3 T	79.2±14.6	29.0±5.4	78.7±16.1	30.5±7.4
1.5 T	82±12.4	29.8±6.9	80.9±13.6	30.9±8.1

All volumes indexed for BSA (ml/m²). RV: right ventricular, LV: left ventricular, EDV: end-diastolic volume, ESV: end-systolic volume.

**Conclusion:** There is no relevant difference in ventricular size when using a 1.5 T or 3 T scanners.

**Abstract no: 1672**

**Device closure of congenital ventricular septal defects: A single centre experience**

**Sivakumar Kothandam, Sreeja Pavithran, Anilkumar Singhi and Anpon Bhagyavathy**

MIOT Hospital, Chennai, India

**Background:** Routine transcatheter closure of perimembranous ventricular septal defects (VSD) is controversial due to atrioventricular nodal conduction disturbances. Muscular and post-operative VSD do not pose such risks, but are rare.

**Methods:** In a retrospective review of 80 patients, who underwent VSD device closure, perimembranous VSD (59); muscular defects (11), intraconal outlet VSD (2), subpulmonary VSD (1) and residual defects after surgery (2). Indications for closure were symptoms, pulmonary hypertension and cardiac enlargement. Asymmetric devices were chosen in membranous VSD without aortic margin, intraconal and subpulmonary VSD. Other devices were used in membranous defects with septal aneurysm.

**Results:** Seventy eight procedures were successful: age ranged from 8 months-years; body weight ranged from 6 - 79kg. Two patients had echocardiographic-guided periventricular closure; others had closure in catheterisation laboratory (cathlab). Asymmetric VSD occluder was used in 7 patients, symmetric VSD occluder in 2, muscular VSD occluder in 2, atrial septal occluder in 1, standard duct occluder in 30, and Amplatzer duct occluder II in 26 patients. With the exception of hybrid closures and duct occluder II devices, all others needed formation of arteriovenous loop. Two perimembranous defects needed 2 duct occluder II devices. One perimembranous VSD measuring 13mm where procedure failed was closed in surgery. Another post-operative residual VSD where procedure failed refused redo surgery. None had complete atrioventricular nodal block and need for pacing. One post-operative patient with aortic regurgitation and ventricular dysfunction had closure of residual large VSD with atrial septal occluder but died of progressive heart failure after 2 years. All others are free of any residual shunt at a mean follow-up of 2 years.

**Conclusion:** Closure of VSD with devices in various locations is safe and feasible. Atrioventricular nodal disturbances are not seen when duct occluders are used.

**Abstract no: 1673**

**Surgical correction of total anomalous pulmonary vein connection: A single centre experience**

**Ruben Movsesian, Natalia Fedorova, Nikolai Antsygin and Aleksey Shikhranov**

Paediatric Cardiac Surgery, Children's City Hospital, No. 1 Saint Petersburg, Russia

**Background:** Total anomalous pulmonary venous connection (TAPVC) is a rare cardiac anomaly often associated with other cardiac malformations and with a poor prognosis without surgical treatment. We represent a 15-year single centre experience in surgical treatment of different types of TAPVC.

**Methods and materials:** From January 1998 - May 2012, 58 patients underwent TAPVC surgery at Children's City Hospital No. 1 Saint Petersburg, Russia. Data collection occurred retrospectively. Mean age at the time of surgery was 7.2 days. TAPVC was supracardiac in 30 patients (51.7%), intracardiac in 19 (32.8%), infracardiac 4 patients (6.9%) and mixed in 5 cases (8.6%). TAPVC obstruction was confirmed by echocardiography and was evident clinically in 8 patients (13.8%). Median CPB time was 64 minutes (40 - 106 minutes). DHCA was used in all cases with a median time of 29 minutes (24 - 37minutes).

**Results:** Hospital mortality was 1.7% (n=1). Cause of death in this case was low cardiac output due to heart failure. Follow-up was available for all of the operative survivors for a median period of 6.8 years (2.5 months - 14 years). Five patients underwent re-do surgery, 3 of them had obstruction and 2 had residual anomalous pulmonary vein connection. The times to reoperation for anastomotic stricture (n= 3) were 13 days, 3 months, 4 years. Late mortality was 1.7% (n=1), this patient died of heart failure due to PV obstruction. The overall mortality of this series was 3.4% (n=2). Hospital mortality has not been observed since 2008 in all age groups.

**Conclusion:** TAPVC is a rare congenital heart lesion, which still remains a surgical challenge. Despite the good short-term results of surgical correction, we should focus on long term results, analysis of which will help us to reduce the number of late complications, the need for re-interventions and improve the quality of life.

**Abstract no: 1675**

**Anomalous left coronary artery-to-pulmonary artery (ALCAPA) in Red Cross War Memorial Children's Hospital, Rondebosch, Cape Town**

**Beyra Rossouw\*<sup>†</sup>, Liesl Zulke\*<sup>#</sup> and John Lawrenson\*<sup>##</sup>**

\*Red Cross War Memorial Children's Hospital (RCWMCH), Rondebosch, Cape Town, South Africa

<sup>#</sup>School of Child and Adolescent Health, University of Cape Town, Observatory, South Africa

<sup>†</sup>Tygerberg Children's Hospital, University of Stellenbosch, Bellville, South Africa

**Introduction:** Dilated cardiomyopathy (DCM) has a poor prognosis in the developing world where ICU beds, mechanical assist devices and cardiac transplants are limited. ALCAPA presents similar to DCM but is surgical treatable, therefore ALCAPA should be considered when investigating DCM.

**Aim:** To audit patients admitted to RCWMCH with a new diagnosis of ALCAPA.

**Method:** Retrospective folder review of ALCAPA patients admitted between July 2004 and August 2012.

**Results:** Twenty five newly diagnosed ALCAPA patients with median age of 5.3 (range 0.5 - 30) months at presentation. Presenting symptoms were tachypnoea (96%), cardiomegaly (92%), coughing (88%), failure to thrive (60%), feeding difficulty (52%) and gallop in 44%. Median length of symptoms was 24 days (range: 1 - 300). In 68% the diagnosis was made on Echocardiography. Echocardiography findings were pathological MR and dilated LV in 79% and bright papillary muscles in 78%. Median ejection fraction on admission was 26%. Median time from RCWMCH admission to surgery was 5 days (range: 1 - 20). All received coronary reimplantation. Median cross-clamp and bypass time was 71 and 140 minutes. 24% had delayed sternal closure. Median length of hospital stay, ICU stay, ventilation and inotropic support was 21, 10.8, 7.4 and 9.3 days respectively. Median Wernovsky inotrope score was 32 (5 - 85). Peri-operative complications included sepsis 68%, pleural effusion 24%, arrhythmias needing pacing 20%, bleeding 16% and renal replacement therapy 12.5%. 92% (23/25) survived to hospital discharge, 1 patient died pre-surgery and 1 during surgery. 3 patients died during subsequent admission for inter-current chest infection.

**Conclusion:** RCWMCH can expect 3 new ALCAPA cases per year presenting in congestive cardiac failure. Treatment is successful with mortality rates comparable to the developed world. The major complication is peri-operative-related sepsis.

**Abstract no: I683****Single vs. 2-stage repair for patients with pulmonary atresia and major aortopulmonary collaterals****Zeena Makhija and Rajesh Sharma**

Fortis Escorts Heart Institute and Research Center, New Delhi, India

**Background:** Both single and staged approaches are described for patients with pulmonary atresia (PA) and variable availability of major aortopulmonary collaterals (MAPCAs) for unifocalisation. We compared the outcomes between patients who underwent single stage vs. staged repair for this pathology at our institution.

**Materials and methods:** Between 2007 and 2012, 61 patients who underwent procedures for Ventricular septal defects (VSD), PA, MAPCAs were reviewed. Pre-operative Computed Tomographic angiogram was done in all patients to evaluate pulmonary vasculature, and MAPCAs. Twenty five patients (Group I) who underwent a staged repair (VSD was not closed in the 1st stage) were compared with 36 patients (Group II) who underwent single stage complete repair (VSD closed). 1st stage included a systemic-to-PA shunt (19), shunt and unifocalisation (6) and shunt and MAPCA ligation/coil embolisation (8). All patients of Group I had VSD closure and Right Ventricle (RV)-to-PA conduit interposition after a minimum follow-up period of 6 months. The number of MAPCAs unifocalised per patient ranged from 1 - 4 (median 2) in either group.

**Results:** There was no significant difference in the overall early complication rate between the 2 groups. Early mortality was significantly higher in patients who underwent staged procedures (n=8) vs. those who underwent single stage complete repair (n=0) (p=0.01). At a median follow-up period of 70 months, late mortality was similar between the 2 groups (p=0.45).

**Conclusions:** The data demonstrate that single stage complete repair has yielded acceptable early and mid-term results. The high mortality of the shunt group indicates the need for an alternative strategy to improve pulmonary blood flow other than a systemic to PA shunt. A palliative RV-to-PA conduit can be considered in these patients.

**Abstract no: I684****A novel syndrome of human pacemaking dysfunction: Lessons in personalised medicine from a rare disease****Philippe Chetaille\*, Jean-Marc Cote\*, Christine Houde\*, Michel Cameron#, Severine Leclerc#, Louise Gosselin\*, Maryse Thibeault#, Steve Jones† and Gregor Andelfinger#**

\*Cardiology Service, Centre Hospitalier Universitaire de Quebec, Quebec, Canada

#CV Genetics, Sainte Justine Hospital, University de Montreal, Quebec, Canada

†University of British Columbia, Vancouver, Canada

**Background:** Lifelong rhythmic contractions are the hallmark of the human heart and gut. They result from the pacemaking activity of the specialised tissue, namely the sino-atrial node in the heart and interstitial cells of Cajal in the gut. Here, we describe a new syndrome characterised by progressive loss of normal sino-atrial node function (sick sinus syndrome, SSS) and chronic intestinal pseudo obstruction (CIPO) in French-Canadians.

**Methods and results:** Detailed chart review, family history, physical exam, ECG and echocardiography were performed in 14 carriers with SSS/CIPO. Whole exome sequencing was performed in 3 affected and analysed under a model of recessive inheritance.

**Results:** Age at onset of SSS was between 6 and 27 years with all features of bradycardia-tachycardia syndrome. Pacemaker implantation was required in 5 individuals (age 6 - 21 years). Gastrointestinal symptoms appeared between 5 and 14 years of age, but not in any particular order to SSS/CIPO. Unexpectedly, we found that a mutation in shugoshin-like 1 (SGOL1), a component of the cohesin complex, leads to this hitherto undescribed syndrome. All surviving affected are homozygous carriers of a founder mutation in SGOL1 (c.67 T>C [p.Lys23Glu]) which results in a dramatic change of a highly conserved amino acid. Haplotype and genealogical analysis point to the introduction of a founder mutation at least 220 years ago. None of the unaffected 1st-degree relatives was homozygous for the mutation, and the mutation was not detected in 360 French-Canadian controls. A 6-year-old homozygous mutation carrier developed signs of SSS/CIPO during the course of the study.

**Conclusion:** Genetic analysis of the novel SSS/CIPO syndrome demonstrates a fundamental new link between the 2 main pacemakers in humans. A predictive value of SGOL1 testing was found in one case and highlights the need to develop preventive strategies based on molecular findings.

**Abstract no: I685****Presence and patterns of myocardial gadolinium enhancement in childhood dilated cardiomyopathy****Heiner Latus\*, Kerstin Gummel#, Stefan Rupp\*, Hakan Akintuerk#, Christian Jux\*, Juergen Bauer\*, Dietmar Schranz\* and Christian Apitz\***

\*Paediatric Heart Centre, University Children's Hospital Giessen, Germany

#Division of Cardiovascular Surgery, University Clinic Giessen, Germany

**Background:** Different patterns of myocardial gadolinium enhancement (MGE) including mid-wall fibrosis have been reported in up to 42% of adult patients with non-ischaemic dilated cardiomyopathy (NI-DCM). In these studies, MGE was associated with pronounced LV remodelling and predicted adverse cardiac outcomes. Accordingly, the purpose of our study was to determine the presence and patterns of MGE in children with NI-DCM.

**Methods:** Patients presenting with severe congestive heart failure who were admitted for evaluation of heart transplantation at our centre underwent CMR examination on a 3-T system that consisted of ventricular functional analysis and assessment of MGE for detection of myocardial scars. Ischaemic DCM was excluded by coronary angiography and right ventricular endomyocardial biopsies ruled out ongoing myocarditis.

**Results:** Fifteen infants and children (mean age 9.3±7.6 months) with severe LV dilatation (mean indexed LVEDV 160±55ml/m<sup>2</sup>) and LV dysfunction (mean LV-EF 16±7%) were examined. MGE was detected in 2 of the 15 patients (13%) appearing in patterns characterised as focal patchy and transmural in 1 patient respectively. None of the patients exhibited "classic" mid-wall enhancement.

**Conclusions:** Despite the small cohort size, the observed differences in frequency and type of myocardial fibrosis compared to adult patients might influence the therapeutic strategy in childhood NI-DCM. However, it remains unclear whether these findings reflect preserved endogenous repair mechanisms that enable favourable re-remodelling. Larger trials are needed to assess the prognostic implications of MGE in childhood NI-DCM and to determine whether MGE might be used for risk stratification as demonstrated in adult NI-DCM.

**Abstract no: 1687**

**Mid-term outcomes of a hand made trileaflet valved right ventricle to pulmonary artery conduit**

**Zeena Makhija and Rajesh Sharma**

Fortis Escorts Heart Institute and Research Center, New Delhi, India

**Background:** We have been using a new technique to make bovine pericardial valved conduits to overcome the shortage of cryopreserved homografts for right ventricle (RV) to pulmonary artery (PA) reconstructions. GORE PRECLUDE pericardial membrane (0.1mm thickness) is used for making the trileaflet valve. We reviewed our experience to analyse postoperative outcomes and their mid-term results.

**Materials and methods:** Between 2007 and 2012, 203 patients underwent primary right ventricle outflow tract reconstruction using bovine pericardial valved conduits in our center. 71.9% (n=146) patients were males. Their age ranged from 6 days - 42 years. Diagnoses included Ventricle Septal Defect (VSD), Pulmonary Atresia (PA), MAPCAs (n=61), VSD, PA (n=52), Truncus Arteriosus (n=20), Double outlet right ventricle, VSD, Pulmonary stenosis (PS) (n=18), Tetralogy Of Fallot/Absent Pulmonary Valve (n=12), Corrected transposition, VSD, PS (n=11), Transposition of great arteries, VSD, PS (n=7) and Ross procedure (n=11). 26.6% (n=54) patients had undergone a prior palliative shunt procedure. The sizes of the conduits implanted ranged from 12 - 24 (median 18).

**Results:** Conduit related early complication rate was 0.5% (n=1) (conduit revision for early pulmonary artery thrombosis). The mean hospital stay was 13.2±31.6 days. Early mortality was 7.4% (n=15). The mean follow up period was 61 months. 11.3% (n=23) patients underwent conduit replacement. Of these, acquired distal conduit stenosis was observed in 10.8% (n=22) and 0.5% (n=1) developed infective endocarditis. Late mortality was 0.5% (n=1). A gradient >30mm Hg was detected in 13.4% (n=27) patients, while 15.3% (n=31) developed conduit valve regurgitation greater than 2+.

**Conclusions:** The hand made valved conduit seems to offer equivalent results as compared to other conduits available today at midterm follow up. Long term results are awaited. Easy availability and low cost are additional advantages of this conduit.

**Abstract no: 1690**

**Congenital heart disease and the emission of developmental toxicants in Alberta, Canada**

**Deliwe Ngwezi\*, Alvaro Osornio Vargas\*, Brad Saretsky#, Ambika Mehta†, Eric White‡, Sujata Chandra§, Debbie Fruitman<sup>o</sup> and Lisa Hornberger\*<sup>o</sup>**

\*Department of Paediatrics, University of Alberta, Stollery Children's Hospital, Edmonton, Alberta, Canada

#Alberta Health Services, Alberta, Canada

†Faculty of Science, University of Alberta, Stollery Children's Hospital, Edmonton, Alberta, Canada

‡Faculty of Medicine and Dentistry, University of Alberta, Stollery Children's Hospital, Edmonton, Alberta, Canada

§Department of Obstetrics and Gynaecology, University of Alberta, Edmonton, Alberta, Canada

<sup>o</sup>Department of Paediatrics, University of Calgary, Alberta Children's Hospital, Edmonton, Alberta, Canada

**Background:** Congenital heart disease (CHD) is a significant global public health issue affecting 1% of all live births and the most common lethal congenital abnormality in infancy. Although CHD may occur in the presence of aneuploidy and single gene disorders the cause is unknown in most affected children. The role of environmental pollutants has recently received attention. We sought to explore the association of developmental toxicants (DTs) from industrial sources and CHD in Alberta, Canada through an interdisciplinary multistep study.

**Materials and methods:** In this ecologic study we collected the following data: (1) Chemical emissions from 2003 - 2010 from Canada's National Pollutant Release Inventory; (2) CHD cases (born between 1 June 2004 - 31 August 2011) from Stollery Children's Hospital Xcelera database; and (3) Annual births from Statistics Canada/Alberta Reproductive Health. We used Scorecard criteria to identify emitted DTs and corresponding toxic equivalent potential values in order to normalise emissions (risk score). The location of the emitting facilities and CHD cases were determined using the longitude and latitude coordinates. Data was aggregated by year and cases were assigned to the year when pregnancy occurred. Statistical analysis was done using STATA 12.

**Results:** We identified 1903 cases of CHD and 17 DTs emitted to air (99% of all emissions) during the study period. The average rate of CHD was 5.8±1.09/1000 live births with the most commonly encountered including septal (47.9%), left ventricular outflow tract obstruction (15.2%) and conotruncal (12.2%) defects. The average DTs emissions were 7 817 417±570 380 tonnes. Annual sulphur dioxide, ethylene oxide, 1,3-butadiene, hexachlorobenzene and carbon disulphide average risk scores strongly correlated with CHD rates (r=0.89; r=0.89; r=0.87; r=0.84 and r=0.84, respectively, p<0.01).

**Conclusions:** These findings suggest that DTs emitted to air in Alberta could have an impact on the development of CHD. An in-depth analysis using sophisticated statistical methods, modelling and GIS mapping is in progress.

**Abstract no: 1691**

**Comparison of shear stress rate distribution after Norwood with right ventricle-to-pulmonary artery conduit and Blalock-Taussig shunt for hypoplastic left heart syndrome: Mathematic modelling of haemodynamics**

**Tomasz Mroczek, Zbigniew Malota and Janusz Skalski**

Department of Pediatric Cardiac Surgery, Jagiellonian University, Krakow, Poland

**Objective:** The introduction of right ventricle to pulmonary artery (RV-PA) conduit instead of Blalock-Taussig shunt (BTS) for Norwood procedure for hypoplastic left heart syndrome resulted in a higher survival rate in many centres. Exact mechanisms and long term results are still under investigation. The main objective of this study was the comparison of shear stress rate (SSR) distribution in 2 models of Norwood physiology regarding different types of pulmonary blood flow sources.

**Method:** Based on anatomic details obtained from echocardiographic assessment and angiographic studies, 2 three-dimensional computer models of post-Norwood physiology with RV-PA 5-mm conduit and Blalock-Taussig shunt (BTS) 3.5-mm shunt were developed. The finite-element method was applied for computational simulations. Shear stress distribution was analysed in both models at basal level of pulmonary and systemic vascular resistances and also with decreased systemic vascular resistance and increased pulmonary vascular resistance.

**Results:** The highest values of shear stress rate were observed in shunts both in RVPA and BTS Norwood models during the peak systolic phase. The imperfect reconstruction of the aorta influences the local distribution of SSR. Changes of systemic and pulmonary vascular resistances do not change significantly the distribution patterns of SSR in both models.

**Conclusions:** The sources of pulmonary blood flow are the areas of the highest shear stress rate. The way of reconstruction of the aorta influences the local distribution of SSR.

#### Abstract no: 1716

### Complex approach to highly malignant long QT syndrome in a 2-year old girl

Viera Illikova\*, Dusan Poruban#, Vladimir Cingel† and Robert Hatala\*

\*Departments of Arrhythmias and Pacing and ICU, Children's Cardiac Centre and National Cardiovascular Institute, Bratislava, Slovakia

#Department of Oro-Maxillofacial Surgery, St Elisabeth Institute of Oncology, Bratislava, Slovakia

†Department of Surgery, Paediatric Faculty Hospital, Bratislava, Slovakia

**Background and objectives:** Clinical presentation and therapy of long QT syndrome (LQTS) depends on the genetic type of the syndrome. We present a case of a girl with clinical manifestation suggesting LQTS type 1 or 2, resistant to beta-blocker therapy.

**Methods:** 2-year-old girl was admitted after syncope. Prolonged QT interval and episode of Torsade de Pointes (TdP) was found.

**Results:** Beta-blocker therapy was started with metoprolol. A month later syncope recurred and frequent TdP occurred of few beats to 20 seconds in duration. Propranolol was initiated and increased to supra-maximal dose with no effect on TdP frequency and minimal effect on heart rate. Frequency of TdP increased to a few 100 a day with increased duration and frequent loss of consciousness. TdP were related to sympathetic discharge, completely disappearing during sleep. Lidocain and magnesium demonstrated no effect, analgo-sedation partial effect. Implantable cardioverter-defibrillator (ICD) was implanted and left cervical sympathetic denervation (LCSN) performed. Mexiletin obtained with the help of international pharmacy resulted in complete elimination of TdP episodes. One year later, patient was successfully defibrillated from life-threatening TdP recurrence. Genetic testing revealed SCN5A mutation P1332L.

**Conclusion:** Beta-blocker therapy of LQTS in a girl with clinical presentation suggesting LQTS type 1 or 2 was not successful. Complex approach including ICD implantation and LCSN was needed, with ICD serving as an emergency backup treatment. Surprisingly, lidocain was without effect but mexiletin led to disease control. Availability of mexiletin and genetic testing would allow earlier appropriate treatment.

#### Abstract no: 1718

### Cardiac involvement in patients with Becker muscular dystrophy

Joana Freitas\*, Silvia Alvares#, Marilia Loureiro# and Manuela Santos†

\*Paediatric Department, Central Hospital of Porto, Porto, Portugal

#Paediatric Cardiology Department, Central Hospital of Porto, Porto, Portugal

†Paediatric Neurology Department, Central Hospital of Porto, Porto, Portugal

**Background:** Cardiac involvement (CI) is an invariable feature of Becker Muscular Dystrophy (BMD), and can be even more pronounced than the skeletal muscle weakness. It may be subclinical and detectable only by instrumental investigation, or symptomatic. Asymptomatic CI occurs in most cases, but up to 1/3 of patients developed dilated cardiomyopathy (DCM) with concomitant heart failure. The degree of symptomatic CI in BMD varies greatly.

**Aim:** To assess the cardiac findings in patients with BMD from the outpatient clinic of our institution.

**Material and methods:** Retrospective review of clinical, electrocardiographic and echocardiographic assessments of patients with BMD.

**Results:** The study included 11 patients aged 1 - 16 years at referral to Paediatric Cardiology consult. Long term follow-up was 6.8±4.4 years. Patients were assessed annually from diagnosis or more frequently according to the clinical manifestations. Only 1 adolescent had complaints of poor exercise tolerance. Prevalence of preclinical and clinically evident cardiac involvement was 55%. Electrocardiogram was abnormal in 9% of patients. Echocardiogram was abnormal in 55% of patients. Mild left ventricle enlargement was present in 6 patients. Three patients were started on ACE inhibitors.

**Conclusion:** Though most patients were asymptomatic, a high percentage had evidence of cardiac involvement. Clinicians need to be aware of the importance of myocardial involvement in patients with BMD and the therapies available. Until etiology-specific genetic or cell-based therapies are developed, work in this area should concentrate on: (1) Implementing the complementary consensus recommendations for clinical care and research developed by neuromuscular and cardiovascular specialists; and (2) identifying children with cardiomyopathy so currently available treatments can be used appropriately.

#### Abstract no: 1719

### Treatment of patent ductus arteriosus in pre-term neonates: Surgery or indomethacin?

Carolina Capurco and Cleonice Mota

Federal University of Minas Gerais (UFMG), Brazil

**Objectives:** The haemodynamically relevant patent ductus arteriosus (PDA) impairs pulmonary and cardiac function. This prospective study evaluated the treatment and outcome in pre-term newborns.

**Methods:** From 2002 - 2008, 3043 newborns were enrolled, from whom 231 (7.6%) presented PDA and were subdivided into groups depending on the treatment. Spontaneous closure occurred in 39/231 (16.9%) and 17 (7.4%) died without treatment. Indomethacin therapy was successful in 100 (43.3%) patients, surgical closure was the 1st treatment in 55 (23.8%) and 20 (8.6%) undergoing ductal ligation after indomethacin failure. The proportion of neonatal morbidities (chronic lung disease (CLD), necrotising enterocolitis (NEC), haemorrhage intraventricular (HIV, pneumothorax) was compared between groups. The data collected was analysed with Chi-square, Mann-Whitney and multiple logistic regression tests.

**Results:** The age of diagnosis and echocardiographic initial findings were significantly different in neonates with spontaneous closure than those with significant PDA (p=0.00). PDAs with an internal diameter >2.0mm: reversal in diastolic flow in descending aorta and treatment after 8 days were associated with complications (death, indomethacin failure, CLD, NEC and HiV). Indomethacin and surgery gave similar benefits. There were no differences in deaths, CLD, NEC. Surgery was more effective in closing PDA (p=0.00) but it was associated with increased risk for HiV and pneumothorax (p=0.00).

**Conclusions:** Early PDA diagnosis and treatment in pre-term infants improves their outcome. Echocardiographic parameters predicted risk of complications and should be useful to guide the therapy. Indomethacin and surgical closure were safe and had no long term complications owing to surgery.

**Abstract no: 1722**

**Tetralogy of Fallot and meningitis complicated by brain abscess: Late fatal presentation in a child**

**Bosedede Adebayo<sup>#</sup>, Felix Ayun<sup>\*</sup>, Abel Alonge<sup>\*</sup> and Ade Omotayo<sup>\*</sup>**

<sup>\*</sup>Federal Medical Centre, Owo, Ondo State, Nigeria

<sup>#</sup>College of Medicine, University of Ibadan, Ibadan, Nigeria

**Background:** Brain abscess is a known complication of cyanotic congenital heart diseases. We report the case of a 4-year-old girl whose 1st presentation in the hospital was because of symptoms referable to the neurological system. The illness was however fatal due to the late presentation which is not uncommon in this environment due to various reasons.

In a resource-poor environment like ours, this is a reminder that proper history-taking, clinical examination and subsequent investigations will aid in early diagnosis and subsequent management of such cases to reduce childhood mortality.

**Abstract no: 1723**

**Comparison of the echocardiographic results in infants with ventricular septal defect and different NT-ProBNP and endothelin-I Levels**

**Bozena Werner and Jacek Skiendzielewski**

Department of Paediatric Cardiology and General Paediatrics, Medical University of Warsaw, Warsaw, Poland

**Background and aim:** To analyse the haemodynamic disturbances assessed by echocardiography in infants with ventricular septal defect (VSD) and different NT-proBNP and endothelin-I (ET-I) levels.

**Material and methods:** The study group consisted of 34 infants (aged 38 - 338 days, mean 130±81 days), 15 boys, 19 girls with VSD without pulmonary hypertension. In 2-D Echo the following parameters were analysed: pulmonary-t- systemic flow ratio (Qp/Qs), peak velocity of the pulmonary artery flow (Vmax PA), left atrial-to-aortic diameter ratio (LA/Ao) and indexed for body surface area, size of the defect (VSD/BSA), left atrial diameter (LAD/BSA), left ventricular internal diastolic diameter (LVIDd/BSA), right ventricular internal diastolic diameter (RVDd/BSA), main pulmonary artery diameter (MPA/BSA). After the analysis of NT-proBNP and ET-I levels, the following subgroups were selected: Subgroup I: 24 (70%) children with NT-proBNP<100fmol/ml; Subgroup II: -10 (30%) children with NT-proBNP≥100fmol/ml; Subgroup III: 25 (73%) children with ET-I<0,4fmol/ml; Subgroup IV: 9 (27%) children with ET-I≥0,4fmol/ml. Echocardiographic parameters were compared between subgroups I vs. II and III vs. IV. Statistical analysis was obtained using Student's t-test.

**Results:**

2-D Echo	Subgroup I - Mean±SE	Subgroup II - Mean±SE	p-value
Qp/Qs	2.1±0.2	3.3±0.3	p<0.01
VmaxPA (cm/s)	127±5.1	152±10.9	p<0.05
LA/Ao	1.5±0.05	1.8±0.1	p<0.01
VSD/BSA (mm/m <sup>2</sup> )	15.3±1.6	26.2±2.4	p<0.01
LAD/BSA (mm/m <sup>2</sup> )	56.3±3.0	83.9±4.4	p<0.01
LVIDd/BSA (mm/m <sup>2</sup> )	79±3.8	104±4.9	p<0.01
RVDd/BSA (mm/m <sup>2</sup> )	28.4±1.4	34.2±2.4	p<0.05
MPA/BSA (mm/m <sup>2</sup> )	35.3±1.6	52.4±3.7	p<0.01

SE: standard error.

No significant differences in the mean values of all echocardiographic parameters in Subgroup III compared to those obtained in Subgroup IV were found.

**Conclusions:** (1) Higher levels of NT-proBNP indicate more severe haemodynamic disturbances in infants with VSD; and (2) The determination of ET-I level seems to be useless in infants with VSD.

**Abstract no: 1726**

**Evolution of right ventricle-to-pulmonary artery conduit concept during the Norwood procedure: 10-years' single institution experience**

**Thomasz Mroczek and Janusz Skalski**

Department of Paediatric Cardiac Surgery, Jagiellonian University, Krakow, Poland

**Background:** The introduction of right ventricle-to-pulmonary artery (RV-PA) conduit in the Norwood procedure for hypoplastic left heart syndrome resulted in a higher survival rate in many centres. However, RV-PA conduit may result in more frequent unintended interventions due to stenosis at many levels.

**Methods:** We describe the evolution of RV-PA conduit concept for Norwood procedure in single institution, when more than 320 Norwood procedures with RV-PA conduit were performed from 2001 - 2012. The technical issue of RV-PA placement will be described including the way of proximal and distal implantation, choosing the right or left position according to neo-aorta, the diameter of shunt and applying of reinforced grafts.

**Results:** The current technique includes the reinforced, usually 5mm diameter shunt localised to the right according to the neo-aorta with introduction of proximal part of conduit into the right ventricle's wall.

**Conclusion:** The evolution of technical issues regarding RV-PA conduit for Norwood procedure results in less unintended interventions and facilitates the stage II performance.

**Abstract no: 1735****A rare form of congenital heart disease in pregnancy****Mamotabo Matshela**

University of KwaZulu-Natal, Durban, South Africa and Mayo Clinic, Rochester, Minnesota, United States of America

**Background:** Univentricule is a rare form of congenital heart disease. It is often associated with other congenital heart defects.**Objective:** To describe the clinical presentation and echocardiographic features of a patient presenting with a single atrium and single ventricle in pregnancy.**History and progress:** A 19-year-old female presented with a history of univentricular problems with left ventricular morphology, univentricular arteriovenous connection, absent right LV valve, pulmonary and tricuspid atresia and dextrocardia. In addition she had situs inversus and a single kidney. She had classic Blalock-Taussig shunt in 1991 at 6 weeks of age. She subsequently developed adhesions around the previous shunt and stenosis to the insertion site and on the either sides of the left pulmonary artery of the shunt insertion. Six years later she had total cavo-pulmonary connection. Thirteen years later she presented with an unplanned pregnancy and complained of dizziness and dyspnoea. She subsequently had episodes of syncope, cyanotic spells and worsening dyspnoea. She was admitted for close monitoring and 3 weeks later had an elective Caesarean section with no peri-operative complications. She was discharged a few days later and during her follow-up periods she remained haemodynamically stable. Her baby girl remained in a good condition.**Discussion and conclusion:** There is lack of literature on the prevalence and successful pregnancies in this form of a complicated congenital heart disease. The terms "complex single ventricle" and "univentricular" heart are used to describe this form of congenital heart disease. These terms are also used to describe a group of rare heart defects, which have in common, a large single pumping chamber or ventricle. It is a serious problem because without surgery, most children would not be able to survive the 1st year of life. These patients may present with congestive heart failure or cyanosis. Complex single ventricle can be diagnosed before birth by a fetal echocardiography and as early as 18 weeks into pregnancy. This should be suspected especially if there is a family history of congenital heart disease.**Abstract no: 1740****Prognostic implications of ventricular septal defects in pregnancy****Mamotabo Matshela**

University of KwaZulu-Natal, Durban, South Africa and Mayo Clinic, Rochester, Minnesota, United States of America

**Background:** Ventricular septal defects (VSDs) are among the commonest congenital heart disease. These can occur in isolation or as part of a more complex congenital heart disease. In adults they are usually small and restrictive, however if large they are commonly associated with Eisenmenger syndrome or forms part of complex congenital heart disease. In pregnancy isolated small or moderate sized ventricular septal defects or surgically closed with normal ejection fraction and without pulmonary hypertension are usually well tolerated.**Objective/aim:** Evaluate the prognostic outcome of ventricular septal defect in pregnancy.**Methods and design:** Thirty two patients who presented with ventricular septal defects during pregnancy were reviewed. Their clinical profile and echocardiography were retrieved and re-evaluated. The patients were divided into 3 groups based on the size of the defect i.e. small, moderate or large size ventricular septal defect. More than 70% of these VSDs were incidentally discovered during pregnancy. The New York Heart Association functional classification of dyspnoea was used to classify their functional capacity. All these patients were admitted and closely monitored during their peri-partum period. They were subsequently followed up 6 - 8 weeks post-delivery for full cardiac evaluation.**Results:** Of these patients 26 had small size, 4 had moderate size and 2 had large size ventricular septal defects. Two patients had associated dextrocardia; 1 had associated congenitally corrected transposition of great vessels and 12 had pulmonary hypertension (8 had mild pulmonary hypertension and 4 had moderate pulmonary hypertension). The majority of these patients presented with at least Class II symptoms (18) and 14 presented with Class I symptoms. None of these patients experienced any peri-partum complications related to either cardiac lesion or pregnancy. All patients received infective endocarditis prophylaxis during labour. At the 6 - 8 weeks follow-up; only patients with a large ventricular septal defect reported at least Class II symptoms and needed an earlier intervention.**Conclusion:** In this study we predominantly had patients with small ventricular septal defect which is associated with favourable overall outcome. One patient had complex congenital heart disease as part of the syndrome. No associated maternal or fetal complications peri-operatively or post-delivery.**Abstract no: 1741****Use of immunoglobulin for myocarditis in children: Variation in practice across the U.K.****Nitha Naqvi and Margerita Bartsota**

Royal Brompton Hospital, London, United Kingdom

**Background:** The U.K. Department of Health has produced updated guidelines for the use of immunoglobulin in 2011 which do not include myocarditis. A Cochrane review in 2009 found no evidence to support this indication but recent publications disagree. In the current financial climate British National Health Service funding of immunoglobulin is under review.**Objective:** To investigate variations in current U.K. practice for the administration of immunoglobulin (Ig) in children with acute myocarditis.**Method: Design and setting:** A questionnaire was emailed to paediatric cardiology consultants in all 13 U.K. paediatric cardiac tertiary units. Paediatric cardiac pharmacists were also contacted. **Main outcome measures:** Use of immunoglobulin and status of recipients at preferred administration time (early in illness vs. later when ventilated vs. only proven viral infection cases).**Results:** Eleven centres responded (85% response rate) with considerable variation in practice: 4 units do not use Ig in myocarditis; 2 units give only if a proven viral cause; 1 unit reported using only in desperately sick ventilated children; and the remaining 4 units were using Ig more liberally early in the illness in non-ventilated patients.**Conclusion:** Due to a paucity of evidence-based practice there is wide variation of opinion amongst management as to whether immuno-globulin is given or not. Furthermore in those units continuing to use Ig for myocarditis there is no agreement on timing of administration (early or later in illness). There is a need for consensus guidelines particularly as if a shortage occurs Ig will be withdrawn for this indication.

**Abstract no: 1746**

**Live real-time 3-D echocardiography in paediatric cardiology: Fulfilling the promise?**

**Uriel Katz, Yshai Salem, Amir Vardi and David Mishaly**

The Edmond and Lilly Safra Children's Hospital, Sheba Medical Centre, Tel Hashomer, Israel

**Background:** Three dimensional (3-D) imaging is one of the most significant developments in ultrasound technology, providing realistic and unique, comprehensive views of cardiac structures. When initially introduced, it held a great promise in imaging of congenital abnormalities. However, acquisition of full-volume 3-D data requires good patient cooperation, and lack of it in the paediatric population results in technically limited, often incomplete studies, causing in turn underuse of 3-D. Labour-intensive and time consuming off-line reconstruction further limits use of this technology.

**Methods and materials:** Live real-time three-dimensional echocardiography (LR-T3-DE) is performed by acquiring still images and short clips using the narrow-angle 3-D option, and applying appropriate views and angles. This readily available technique is practical in children, is requires short added examination time, and enables incorporating 3-D imaging into the daily clinical setting in a busy hospital clinic. Using LR-T3-DE routinely, and making it an integral part of the echocardiographic evaluation provides familiarity with its unique advantages as well as its limitations. We find it enhances understanding of anatomy and morphology if cardiac lesions, improves cardiologist-surgeon communication, and aids teaching.

**Results and conclusions:** We present examples of our experience in LR-T3-DE in both simple and complex congenital lesions and valvular heart disease, demonstrating its use and added value compared to 2-D imaging.

**Abstract no: 1749**

**Patterns of pre-natal cerebral growth among infants with congenital heart disease**

**Niels Bjerregaard Matthiesen\*, Peter Agergaard\*, Vibeke Hjortdal# and John Rosendahl Åstergaard\***

\*Department of Paediatrics, Aarhus University Hospital, Skejby, Denmark

#Department of Cardiothoracic Surgery, Aarhus University Hospital, Skejby, Denmark

**Background:** Microcephaly is more common in children with congenital heart disease (CHD), and the relation to neurodevelopmental abnormalities has been demonstrated. However confounders such as genetic syndromes and low placental weight were not sufficiently accounted for in existing studies. Additionally the focus has been on complex CHDs, and the impact of more common CHDs has yet to be evaluated.

**Materials and methods:** A register-based study was performed comprising a validated cohort of 2 947 Danish children born 2000 - 2008 with CHD. Genetic, newborn and maternal parameters were identified in national registries, and each child was randomly matched to 3 non-syndromic children without CHD (n=8841) according to gender and gestational age. By means of linear and logistic regression analysis, newborn head circumference will be adjusted to birth weight, maternal pre-pregnancy weight, smoking status, medical diseases, genetic abnormalities and placental weight.

**Results:** The cohort represents high numbers of diagnostic subgroups: e.g. transposition of the great arteries (n=189), hypoplastic left heart syndrome (n=98), Tetralogy of Fallot (n=210), pulmonary stenosis (n=178), coarctation of the aorta (n=207), aortic valve stenosis (n=88), atrioventricular septal defect (n=184), VSD (n=904) and ASD (n=535). Trisomy 21 (n=180) and 22q11.2-deletion/duplication (n=46) and other genetic abnormalities have been determined. Main outcome measure is newborn head circumference, non-adjusted and adjusted to newborn, maternal and genetic parameters comparing diagnostic subgroups of CHD to healthy controls. Preliminary comparison of unadjusted means revealed significantly smaller newborn head circumference in non-syndromic (-0.233cm, p<0.0001) and syndromic CHD cases (-1.299cm, p<0.0001) compared to controls. Analyses are ongoing and the results will be presented at the meeting.

**Conclusion:** The strength of this study, more than tripling the numbers of the largest study in the field, lies within the possibility to adjust head circumference to confounders such as genetic abnormalities and placental weight through data from unique national registries.

**Abstract no: 1764**

**Ethical analysis of HLHS**

**Mahmoud Elbarbary**

King Abdulaziz Cardiac Center, Riyadh, Saudi Arabi

**Case report:** In a limited-resources Middle Eastern country, a baby was diagnosed intra-uterine as HLHS (hypoplastic left heart syndrome) after 120 days of gestation. No paediatric cardiac surgery or heart transplant programme was available. It was also not feasible to send the patient abroad. Baby was born and required PGEI, mechanical ventilation for the severe cyanosis. Saturation improved with these actions but the baby developed NEC (started on TPN). Baby is obviously PGEI and ventilator-dependant. Parents agreed to "no code" status. ICU beds are full and there are 2 cases in ER of severe asthma that require urgent paediatric ICU admission with expected excellent outcome. The presentation will go through the following 5 steps:

1. **Collecting clinical data:** The starting point in the ethical analysis of a clinical case consists in gathering information related to:

- medical aspects (diagnosis, prognosis, potential treatments)
- personal and relational aspects
- cultural aspects (Islamic perspective)

2. **Assessing responsibilities:**

- What are the specific responsibilities of health care professionals in the given case?
- Has the patient (or his/her legal guardian) been adequately informed?
- What is the role of the family?
- What are the responsibilities of social bodies (social services, etc.)?

3. **Identifying ethical problems:** What ethical problems are involved in the evolution of the given case?

4. **Proposing alternative courses of actions**

- What are the possible courses of action for this case?
- What are the motivating reasons?

Courses of actions	Goals and reasons
Abortion vs. no abortion	No abortion >120 days: religious implications
	Yes <120 (Human life? Suffering?)
Asthma vs. HLHS priority	Yes asthma priority. HLHS futile: economic vs. societal benefits; self vs. governmental pay
No code vs. withdraw/withhold	No withdraw till new counselling; The pyramid of actions
TPN/hydration	No: Legal consequences
Active vs. passive	No: Intentions and immediate death vs. delayed death

## 5. Formulating and justifying ethical judgement.

### Abstract no: 1767

#### Changing pattern of rheumatic heart disease in Kano: Data from the Aminu Kano Teaching Hospital Echocardiography registry

**Mahmoud U. Sani<sup>\*,#</sup>, Muhammad S. Mijinyawa<sup>\*,#</sup>, Shehu A. Kana<sup>#</sup>, Nasir A. Ishaq<sup>#</sup> and Umar Abdullahi<sup>#</sup>**

<sup>\*</sup>Department of Medicine, Bayero University Kano, Nigeria

<sup>#</sup>Department of Medicine, Aminu Kano Teaching Hospital, Kano, Nigeria

**Background:** Rheumatic heart disease (RHD) remains a major public health problem in developing countries. Anecdotal reports across Africa show that the disease is becoming less prevalent in cities and that patients are surviving longer, though with a lot of morbidity. From 2002 - 2006 we found 9.8% of 1 312 patients with a mean age of 24 years on our Echo register have RHD. We set out to review our current data for RHD to see if there are any changes in patterns of presentation in our centre.

**Material and methods:** This is a retrospective analysis of a prospectively collected echocardiography data from August 2010 - July 2012. The study was conducted at the Aminu Kano Teaching Hospital, Kano Nigeria. The procedure was performed with Aloka SSD 4000. The standard techniques for depicting the anatomical structures of the heart were employed. All the procedures were performed by cardiologists. Information obtained from the records included the age, gender, clinical diagnoses and echocardiographic diagnoses. Prevalence and patterns were compared with previous findings.

**Results:** During this period, a total of 1 496 echocardiographic examinations were done. 104 (7.0 %) had rheumatic heart disease. There were 69 females (66.3%) and 35 males (33.7%) aged 30.71 ± 13.99 years (range 12 - 70 years). Forty (37.7 %) were aged 15 - 24 years. The commonest lesions were mixed mitral valve disease and aortic regurgitation (26.9%) followed by a combination of mitral and aortic regurgitation (25%) and pure mitral regurgitation in 17.3 %. Complications of RHD observed included secondary pulmonary hypertension, left ventricular dysfunction, atrial fibrillation and infective endocarditis.

**Conclusion:** Although there is an improvement in prevalence compared to previous findings, RHD is still an important cause of cardiac morbidity. Though patients are a bit older, they had more severe disease and still had complications at diagnosis.

### Abstract no: 1769

#### Peri-operative vasopressin results in reduced LOS in hospital after Fontan procedure

**Pooja Kashyap<sup>\*</sup>, Christopher Knott-Craig<sup>\*</sup>, Steven Goldberg<sup>\*</sup>, TK Susheel Kumar<sup>\*</sup>, Michele Harris<sup>#</sup>, Jean Ballweg<sup>\*</sup> and Mayte Figueroa<sup>\*</sup>**

<sup>\*</sup>University of Tennessee, Le Bonheur Children's Hospital, Memphis, Tennessee, United States of America

<sup>#</sup>Le Bonheur Children's Hospital, Memphis, Tennessee, United States of America

**Background:** The Fontan procedure is the final palliative surgery in patients with single ventricle physiology. Although early post-operative outcomes have improved over time, the length of hospital stay is still prolonged due to persistent chest tube output in the post-operative period. We hypothesised that the use of vasopressin in the peri-operative period would reduce chest tube output by maintaining vascular tone, thereby limiting rd spacing and the need for volume replacement.

**Material and method:** We retrospectively analysed 31 consecutive patients undergoing the Fontan operation from 2008 - 2012. In 2010 vasopressin was introduced as part of the standard management of patients undergoing this operation. The patients were grouped according to use (VP, n=24) or non-use (no-VP, n=7) of vasopressin (0.3 - 0.5mU/kg/min) in the peri-operative period. The endpoints analysed were hospital mortality, length of hospital stay (LOS), and chest tube output.

**Results:** The VP and no-VP subgroups were well matched for age and weight (14.9kg vs. 15.5kg, p=NS). There was no hospital mortality. The LOS in the VP group was 11.2±2.3 days compared to 18.4±3.6 days in the non-VP group (p=0.01). Daily chest tube output decreased significantly in the VP subgroup but not in the non-VP subgroup (p=0.01).

**Conclusions:** Use of vasopressin in the early post-operative period is associated with reduced chest tube output and length of hospital stay after the Fontan operation.

**Abstract no: 1771**

**Isolated left ventricular non-compaction**

**Bozena Werner and Malgorzata Golabek Dylewska**

Department of Paediatric Cardiology and General Paediatrics, Medical University of Warsaw, Warsaw, Poland

**Aim:** To analyse the clinical course and echocardiographic features of isolated left ventricular non-compaction (LVNC) in children.

**Methods and materials:** The study group consisted of 16 patients. Clinical evaluation, chest X-ray, ECG, Holter ECG and echocardiography were performed. In statistical analysis Student t-test was used.

**Results:** The age at diagnosis was 2 weeks - 18 years (average 6.44 years), follow-up 2 months - 9 years (mean 3.15 years). In 2 brothers Barth Syndrome was recognised. Signs of acute heart failure occurred in 4 children. In chest X-ray cardiomegaly was found in 7 with features of pulmonary congestion in 2. The ECG was in normal limits in 8, signs of LV hypertrophy (3), I degree atrio-ventricular block (1), QTc prolongation (1), and ST-T abnormalities in the form of fluttering and/or biphasic T (3). In 5 patients arrhythmia was diagnosed. In all cases evidence of 2-layered myocardium with prominent trabeculations was diagnosed by Echo, the non-compacted to compacted ratio ranged from 1.6 to 3:1, mean 2:1. Dilated left ventricle was found in 8 patients, reduced systolic function in 7 (EF 22% - 57%). The cardiac function progressively improved during follow-up in all of them. The mean age in patients with impaired left ventricular function was 5.9 months and the non-compacted to compacted ratio was 2.09:1 vs. those without left ventricular dysfunction where mean age was 60.8 months ( $p < 0.05$ ) and ratio 1.93 ( $p > 0.05$ ). Pharmacological treatment with ACEI and spironolactone was used in 8 patients and digoxin in 4 of them.

**Conclusions:** (1) The clinical course with severe heart failure and significant systolic dysfunction is observed in infants with LVNC; (2) Progressive improvement of the left ventricular function with age is characteristic for LVNC; and (3) The non-compacted-to-compacted ratio has not impact on left ventricular dysfunction.

**Abstract no: 1773**

**Paediatric cardioverter-defibrillator implantation: Still a challenge but safe and feasible**

**Fabrizio De Rita\*, Luca Tomasi#, Luca Barozzi\*, Alessandra Cristofaletti#, Lucia Rossetti#, Corrado Vassanelli#, and Giovanni Battista Luciani\***

\*Division of Paediatric Cardiac Surgery, University of Verona, Italy

#Division of Cardiology, University of Verona, Italy

**Background:** ICD implantation in pediatric age remains a challenge due to limited BSA in children, making implantation of percutaneous leads difficult, and due to adhesions generated by epicardial patches, which may interfere with normal development of the heart muscle. A new technique of subcutaneous AICD implantation in children with defibrillation coil is here reported.

**Methods:** With the patient supine decubitus, a left anterolateral thoracotomy is performed. The pericardium is opened and an epicardial bipolar electrode for sensing is placed on the left ventricle. The sensing wire is tunnelled subcutaneously and connected to the generator, placed in an abdominal box pocket. A stimulating lead linked to the generator is then tunnelled subcutaneously from the pocket to the back, with the distal end of the coil placed medially to the left scapula.

**Results:** The technique was successfully applied in 2 children (age 14 and 12 years at implantation) affected by LQTS1 and hypertrophic cardiomyopathy respectively, with history of resuscitation for cardiac arrest. Early after procedure, good values of sensing, pacing impedance, and excellent pacing threshold were found in both cases. Furthermore, the defibrillation threshold was optimal in both cases (14 J). The AICD-PM setting parameters were: brady therapy VVI 40 bpm and tachy therapy with a single VF-window from 194bpm. At follow-up of 9 and 23 months respectively, electrical parameters and position of subcutaneous lead remained stable. No events of inappropriate shocks were recorded during follow-up.

**Conclusions:** Placement of AICD with a surgical approach, epicardial pacing and sensing electrode and subcutaneous defibrillation coil, is a feasible, safe and effective procedure in pediatrics. This approach ensures better functioning of device at distance and does not interfere with child growth. Key to success is regular patient follow-up to ensure constant adjustment of sensing parameters for their growth in order to avoid inadequate shocks.

**Abstract no: 1777**

**Pharmacological treatment and long term outcome in patients with long QT syndrome**

**Beata J. Pietrucha\*, Maciej Pitakl, Beata Zaluska-Pitak\*, Jacek Kuzma\*, Jolanta Oko-Lagan\*, Agata Sulik\*, Artur Pietrucha\* and Andrzej Rudzinski\***

\*Children's Cardiology Department, Children's University Hospital, Jagiellonian University, Krakow, Poland

#Coronary Disease Department, John Paul Jagiellonian University Medical College, Krakow, Poland

**Case report:** We observed 17 children aged 6 months to 17.5 years (mean age 10 years), 10 girls and 7 boys, with diagnosed long QT syndrome. Patients were divided into 2 groups: Group I: 11 children (7 females, 4 males) with symptomatic long QT syndrome (syncope, dyspnoea); Group II: 6 patients (3 girls and 3 boys) with prolongation of QTc segment in ECG but without any clinical symptoms. All patients were in long term follow-up from 19 months - 7 years (mean observation time - 3.5 years). Standard 12-lead ECG with assessment of QT interval, corrected QT interval according to Bazett formula and QTc dispersion were done at the beginning and the end of the observation period. 24-hour Holter ECG monitoring with QT evaluation was also performed in all patients. All patients from Group I were administered with beta-blockers (propranolol or metoprolol) with mean dose 1.2mg/kg. Mean standard ECG QTc duration in patients in Group I was significantly > Group II both in the beginning and at the end of observation period (471.82ms. vs. 435ms. and 460.91 vs. 423.33ms.). A similar result was observed in QTc duration in Holter monitoring (535ms. vs. 455ms; 515.91ms. vs. 452.5 ms.). There were no significant differences between QTc duration before and after beta-blocker treatment in Group I (471.82ms. vs. 460.91ms.), although the frequency and intensity of symptoms in this group was expressively diminished. 3 patients (27.3%) from Group I were referred to ICD implantation.

**Conclusions:** (1) Beta-blockers diminished clinical symptoms in long QT syndrome; (2) Syncope can be evident risk factor of sudden cardiac death in children; and (3) Pharmacological treatment could decrease the SCD risk in children with long QT syndrome.

**Abstract no: 1778****Partial anomalous connection of pulmonary veins: Anatomic variants, surgical treatment and development****Jorge Luis Cervantes Salazar, Samuel Ramírez Marroquín, Alfonso Buendía Hernández, Juan Calderón-Colmenero and Lizbeth Gómez Martínez**

Instituto Nacional de Cardiología, Mexico

**Background and objective:** To make the Institute's experience in the surgical treatment of partial anomalous connection of pulmonary veins and its mid-term results in terms of morbidity and mortality available to the rest of the world.

**Method:** The study was designed a retrospective, longitudinal, observational, description of all patients with partial anomalous connection of pulmonary veins (PACPV) undergoing surgical correction from January 2000 - December 2010.

**Results:** Of all 86 patients, 62.5% were males, the average age was 19 years, and the vast majority (91%) was in functional Class I, II of NYHA (New York Heart Association). The most common variant of PACPV was the 2 vessels (64.28%), the surgical technique used the most was redirecting the flow through pericardium (66%), followed by the Warden technique (19.6%) and finally direct reimplantation of the anomalous vein (14.2%). There were 3 deaths in the early post-operative period; there were no deaths in follow-up later on.

**Conclusions:** Surgical results of PACPV at our institution were satisfactory in terms of morbidity and mortality, since they are low and are within the worldwide average. The mid-term prognosis is satisfactory as the remaining patients were in functional Class I of the NYHA.

**Abstract no: 1781****Comparison of exercise capacity in young patients after correction of Tetralogy of Fallot and atrial switch (Senning operation) of transposition of great arteries in long term follow-up****Beata J. Pietrucha\*, Artur Z. Pietrucha# and Andrzej Rudziński\***

\*Children's Cardiology Department, Children's University Hospital, Jagiellonian University, Krakow, Poland

#Coronary Disease Department, Jagiellonian University Medical College, Krakow, Poland

**Case report:** We observed 49 patients who were divided into 2 groups: Group I: 35 patients after correction of Tetralogy of Fallot (ToF) (6 females, 29 males) aged from 8.7 - 21.7 years (mean age 14.6 years). Group II: 14 patients after operation of transposition of great arteries (TGA) by Senning method (6 females, 8 males) aged from 7.3 - 17.8 years (mean age 13.1 years).

**Methods:** In all patients 24-hour Holter ECG monitoring was performed with assessment of heart rhythm, presence of arrhythmias. Also treadmill exercise testing (TT) was done in all patients and modified Bruce protocol was used. We estimated total metabolic equivalent (MET), total exercise time, presence of arrhythmias and changes in ST segment. Nine patients (25.6%) in Group I had ventricular arrhythmias, but complex arrhythmia was only present in 3 subjects. In Group II dysfunction of the sinus node was observed in 5 patients (35.7%). During TT significant changes of ST segment and dysfunction of right ventricle were present in 5 patients after ToF (14.3%), whereas these changes accompanied with additional chest pain were observed in 6 patients (42.9%) after the Senning operation. During TT the mean MET parameters achieved by patients after ToF were significantly > than in patients who had Senning operation (14.12 vs. 12.76). TT time duration was significantly longer in patients after ToF operation (13:12 vs. 10:39). In 3 patients (8.6%) reoperation and/or ICD implantation was considered after ToF, while an additional 3 patients (21.4%) required reintervention.

**Conclusions:** After Tetralogy of Fallot correction patients have a much better long term prognosis compared to patients who had the Senning operation for transposition of the great arteries. Systemic right ventricle dysfunction is frequently observed in patients after the Senning operation of transposition of great arteries. Exercise treadmill test could be very useful in identifying high risk patients after the correction of complex congenital heart defects.

**Abstract no: 1787****Evidence-based "decision analysis" and patient/family shared decision-making in management of CHD****Mahmoud Elbarbary**

King Abdulaziz Cardiac Centre, Riyadh, Saudi Arabia

**Background:** Decisions in critical care setting can frequently be difficult and involve several trade-offs and multiple risk-benefit ratios. In the face of this dilemma, the current most commonly used methods for decision-making in these circumstances are depending on the personal knowledge or experience, nihilism, defer to other experts, defer to patients, dogmatism or just blindly following a policy. These methods are not scientifically sound. These not only lack objectivity but, more importantly, lack supporting evidence and sometimes even diminish adequate and appropriate patient/family involvement. This situation is frequently encountered in making decisions for managing congenital heart diseases.

**Materials and methods:** In the recent era of evidence-based medicine, full arrays of decision support aids were developed. Amongst these evidence-based Decision Analysis (DA) is an important method which is unfortunately infrequently used in PICCU. In this presentation we will explain how this tool could help one make difficult decisions by (1) using available scientific evidence; and (2) also incorporating the patient/family's own preferences and values. DA is a scientifically-based methodology that is not only objective but also expressed numerical values of disutility and probability. The presentation will also explain the various tools that can be used to achieve proper patient/family shared medical decision-making.

**Abstract no: 1790****Chances of newborns and infants to survive to heart transplant in a low density population area: Impact of mechanical support and ABO incompatible transplant***Desiree Machado<sup>\*</sup>, Chloe Joynt<sup>#</sup>, Ernest Phillipos<sup>#</sup>, Ivan Rebeyka<sup>†</sup>, Kristin Simard<sup>‡</sup>, Lori West<sup>§</sup> and Simon Urschel<sup>§</sup>*<sup>\*</sup>Department of Paediatrics, Division of Neonatology and Pediatric Intensive Care, University of Alberta, Edmonton, Alberta, Canada<sup>#</sup>Department of Paediatrics, Division of Neonatology, University of Alberta, Edmonton, Alberta, Canada<sup>†</sup>Department of Paediatrics, Division of Cardiac Surgery, University of Alberta, Edmonton, Alberta, Canada<sup>‡</sup>Transplant Services, University of Alberta, Edmonton, Alberta, Canada<sup>§</sup>Department of Pediatrics, Division of Paediatric Cardiology, University of Alberta, Edmonton, Alberta, Canada

**Background:** Prolonged time on waitlist affects survival pre and post heart transplantation (HTx). We analysed outcomes of patients listed for HTx aged below 3 months in the low population density region of western Canada.

**Materials and Methods:** Review of patients listed below age of 90 days from 2006 - 2011, to determine waitlist mortality, outcomes after HTx and factors that could impact outcomes.

**Results:** Twenty seven patients were listed during the study period. Congenital heart disease was the predominant diagnosis in 20 patients (74%), and cardiomyopathy, myocarditis and others in the remaining 7. Twelve patients (44%) died or were delisted due to clinical deterioration after a median of 32 days (from 7 to 127 days); one was removed after 112 days due to clinical improvement. HTx was performed in 14 (52%) patients after a median waitlist of 51 days (from 2 to 215 days). Mortality in 14 patients who required extra-corporeal life support (ECLS) pre-Tx (9 bridged to transplant, 5 died on waitlist) was not different from patients without ECLS ( $p=0.61$ ) but time to death on the waitlist trended shorter ( $p=0.09$ ). In the transplanted group, 7 patients (50%) received an ABO-compatible (ABOc) heart, with 3 post-Tx deaths. The remaining 7 cases received an ABO-incompatible (ABOi) graft and are alive. ABOc patients waited median 51 days compared to 49 days for ABOi Tx ( $p=0.53$ ). Patient death was not associated with prematurity ( $p=0.61$ ) or birth weight below 2.5kg ( $p=0.71$ ). Cumulative survival post-listing was 44%.

**Conclusion:** HTx outcomes in early childhood are promising however high waitlist mortality has a negative impact on overall results. Despite having strategies such as ABOi HTx, waitlist mortality in western Canada exceeds rates reported from higher population density areas and is higher than other reported age groups. Further strategies to improve organ availability/ allocation are required.

**Abstract no: 1796****Ventricular septal defect: An unusual sequel of blunt chest trauma in a 7-year-old boy***Oluwatoyin Ogunkunle<sup>\*</sup>, Chika Duru<sup>\*</sup>, Bosede Adebayo<sup>#</sup> and Samuel Omokhodion<sup>\*</sup>*<sup>\*</sup>College Of Medicine, University College Hospital, Ibadan, Nigeria<sup>#</sup>University College Hospital, Ibadan, Nigeria

**Background/hypothesis:** Ventricular Septal Defect (VSD) is the commonest congenital cardiac lesion encountered worldwide. Only very rarely is it acquired and causation through blunt injury is even rarer.

**Materials and methods:** A 7-year-old boy suffered blunt trauma to his chest while at play with his peers at school. He had been quite well before then, with no symptoms of the cardiovascular system and with no growth or developmental delays. He presented 2 days later at our centre with features of acute congestive cardiac failure. Two-dimensional and Doppler echocardiographic examination revealed a rent in the peri-membranous portion of the outlet ventricular septum with the avulsed flap still attached to the septum.

**Results:** Cardiac failure was refractory to anti-failure therapy and other stabilisation measures. His clinical condition took a rapid turn for the worse and he succumbed to the ensuing illness 5 days after the trauma, despite the stabilisation measures and before any surgical intervention could be undertaken.

**Conclusions:** Traumatic VSD, though rare, should be considered in cases of acute congestive cardiac failure in otherwise well active children with a history of blunt trauma to the chest and all such patients should undergo careful echocardiographic evaluation. Prompt surgical intervention has been reported to be life-saving in similar cases.

**Abstract no: 1799****Complete repair of Tetralogy of Fallot in late diagnosed patients***Maria Joao Baptista<sup>\*†</sup>, Maria Ana Sampaio<sup>\*†</sup>, Monica Rebelo<sup>\*†</sup>, Manuel Ferreira<sup>\*†</sup>, Leonidia Airoso<sup>†</sup>, Rui Rodrigues<sup>#</sup>, Miguel Sousa Uva<sup>#†</sup>, Albino Pedro<sup>#†</sup>, Manuel Pedro Magalhaes<sup>#†</sup> and Antonio Filipe Junior<sup>†</sup>*<sup>\*</sup>St João Hospital, Porto, Portugal<sup>#</sup>Da Cruz Vermelha Hospital, Lissabon, Portugal<sup>†</sup>Girassol Clinic, Luanda, Angola

**Aim:** In most centres worldwide, correction of Tetralogy of Fallot is performed <9-months of age. However, in developing countries, early repair may be difficult due to many factors, including facilities for diagnosis. The purpose of this study was to evaluate the early results of surgical repair of Tetralogy of Fallot in patients older than a year, in a hospital performing congenital heart surgery in Angola.

**Methods:** It was performed as a retrospective analysis of early results of surgical repair in paediatric patients older than one year, in one single hospital from June 2011 - June 2012. Data were gathered from patients' records, pre-operative cardiac catheterisation or TC studies, operative intervention and pre- and post-operative two-dimensional echocardiographic. Patients submitted to systemic-to-pulmonary shunt were excluded.

**Results:** In our hospital, 55 patients were treated (ratio male-to-female 1.03:1) with a mean age of 6.4 years (range 13 months - 19 years). Early extubation occurs in the majority of patients (mean 9 hours, range 3.35 hours). Most patients didn't present significant post-operative lesions, except for residual interventricular shunt (4), mild or moderate pulmonary regurgitation (6), and mild obstruction of right ventricle outflow (14). Pleural effusion occurs in 5 patients, with good response to medical treatment. No deaths occurred.

**Conclusion:** We demonstrated good early results of complete repair of ToF in paediatric patients with late diagnosis in Angola. L Follow-up at a later stage must be evaluated, but this preliminary study reinforces the value of the establishment of local assessment to early diagnosis and treatment of congenital heart diseases in developing countries.

#### Abstract no: 1801

### Interrupted aortic arch: 10-year experience in surgical treatment

*Jorge Luis Cervantes Salazar, Samuel Ramírez Marroquín, Alfonso Buendía Hernández, Juan Calderón-Colmenero and Lizbeth Gómez Martínez*

Instituto Nacional de Cardiología, Mexico

**Introduction:** The interruption of the aortic arch is a rare malformation, representing less than 1% of cases of congenital heart disease, is associated with 90% mortality if not treated before one year of age; death occurs as a combination of the increase of a short circuit from the left to right, ventricular failure and closure of the ductus arteriosus, resulting in hypo-perfusion, renal failure and metabolic acidosis.

Initial treatment is to maintain a patent ductus arteriosus with prostaglandin administration. Surgical correction is the definitive treatment and must be performed to confirm the diagnosis.

**Method:** A retrospective, longitudinal, observational, descriptive diagnose of all patients with interrupted aortic arch was performed who underwent surgery for correction of this pathology in the NIC, in the period between January 2000 and December 2010.

**Results:** There was a total of 20 patients, there was no preference as to sex, 16 patients (80%) presented with type B interruption, 3 patients (15%) with type A and 1 patient (5%) with type C interruption. The average age at which surgery was performed was 2.9 months (range 3 days-7 months). The surgical technique used was end-to-end anastomosis. The mortality was 1 patient (5%). At follow-up, the rest of the patients are asymptomatic and without reintervention.

**Conclusions:** Despite late referral of many patients with interrupted aortic arch, surgical results and developments presented monitoring is similar to that reported in the world literature.

#### Abstract no: 1803

### Persistent left superior vena cava draining into left atrium with normal coronary sinus

*Zouzra Zahira, El Haouati Rachid, Boukaidi Yassine and Boumzebra Drissi*

Ibn Tofail Hospital, Mohammed VI University Hospital, Marrakech, Morocco

**Background:** The most common variation in the thoracic systemic venous system is a persistent left superior vena cava draining into a coronary sinus. A rare anomaly is a persistent left superior vena cava connecting directly to the left atrium. In this situation it is believed that the coronary sinus must be absent.

**Methods and materials:** We report an unusual case of a left superior vena caval drainage to the left atrium with normal coronary sinus, which was a pre-operative finding during surgical closure of an atrial septal defect in an 11-year-old patient. We rerouted left superior vena caval flow into the right atrium using intra atrial baffle. The post-operative course was uneventful. Through this case report, we discuss embryological development, clinical profile and surgical techniques to treat this condition.

#### Abstract no: 1806

### Normal cardiovascular responses to treadmill exercise test in healthy British children

*Rhiannon Davies\*#, Lindsay Antonio D'Silva\*#, Dirk G. Wilson\*, Amos Wong\* and Orhan Uzun\**

\*Department of Paediatric Cardiology, University Hospital of Wales, Cardiff, United Kingdom

#College of Engineering Swansea University, Swansea, United Kingdom

**Background:** Normal cardiovascular responses to exercise in childhood are not well defined. Maximum normal blood pressure response to exercise in childhood is overestimated which make assessment of hypertensive response in disease situation rather difficult.

**Aim:** To assess normal cardiovascular responses to exercise in healthy British children.

**Method:** A retrospective review was carried out on all children who underwent exercise testing (Bruce test using a treadmill protocol) at a tertiary institution from 2003 - 2010. 137 healthy children (80 males, 57 females) aged 9 - 16 years were included in the study.

**Results:** Minimum exercise duration was 12 minutes across the spectrum regardless of age and gender. Although lower  $\text{VO}_2\text{max}$  values were attained in females compared to male subjects, there was little change throughout adolescence years. All subjects achieved over 85% of maximum predicted heart rate for age. Younger subjects showed quicker heart rate recovery compared to older individuals. Maximum blood pressure did not exceed 155mmHg in any age group. Rate pressure product was lower in males compared to females in most age groups but similar in 13 - 14 years group. All values were summarised in the Table.

	9 - 10 years			11 - 12 years			13 - 14 years			15 - 16 years		
	All (n=22)	Male (n=9)	Female (n=13)	All (n=36)	Male (n=21)	Female (n=15)	All (n=46)	Male (n=29)	Female (n=17)	All (n=33)	Male (n=22)	Female (n=11)
Exercise duration (minutes)	13.1±0.67	14.8±1.1	11.9±0.72	13.3±0.50	13.7±0.64	12.6±0.80	13.1±0.33	13.9±0.40	11.7±0.39	14.3±0.49	15.2±0.53	12.5±0.83
Exercise capacity (METS)	14.7±0.61	16.6±0.83	13.4±0.65	15.5±0.45	15.9±0.49	15.0±0.83	15.4±0.41	16.3±0.51	13.7±0.49	16.5±0.51	17.4±0.56	14.7±0.86
VO <sub>2</sub> Max ml/kg/min	51.6±2.1	58.2±2.9	47.0±2.3	54.4±1.6	55.8±1.7	52.4±2.9	53.7±1.4	57.0±1.8	48.1±1.7	57.8±1.8	60.9±2.0	51.6±3.0
Recovery time (minute)	4.8±0.30	4.5±0.52	4.9±0.37	6.3±0.34	6.3±0.49	6.2±0.45	6.2±0.24	6.3±0.35	6.1±0.28	5.7±0.35	5.8±0.48	5.5±0.45
HR max (bpm)	194.3±2.8	193.0±4.6	195.1±3.6	195.2±2.0	194.0±2.3	196.7±3.8	196.4±2.4	195.3±2.9	198.3±4.3	198.6±2.2	198.5±3.0	198.9±2.5
% HR max reached (%)	92.3±1.3	91.6±2.2	92.7±1.7	93.6±0.99	93.1±1.1	94.3±1.8	95.1±1.2	94.6±1.4	96.1±2.1	97.1±1.1	97.1±1.5	97.1±1.3
85% target HR reached (%)	108.5±1.5	107.8±2.6	109.1±2.0	110.2±1.2	109.6±1.3	111.0±2.1	111.9±1.4	111.3±1.7	113.0±2.4	114.2±1.3	114.2±1.7	114.3±1.5
Change HR (max - rest) (bpm)	102.3±4.5	111.9±4.0	95.7±6.7	104.1±2.8	105.2±2.8	102.6±5.5	107.2±3.1	107.1±4.0	107.4±5.2	108.2±3.8	115.0±4.4	94.8±5.8
SBP max (mmHg)	136.1±3.5	134.8±5.1	137.1±5.0	139.8±2.0	140.8±2.1	138.3±3.8	146.5±2.4	148.3±2.9	143.4±4.0	151.5±2.9	153.6±3.6	147.1±4.6
Change SBP (max-rest) (mmHg)	28.7±2.6	27.0±4.7	29.9±3.1	32.5±1.8	32.1±2.0	33.0±3.4	32.9±2.2	32.7±2.7	33.2±3.9	36.1±2.8	37.6±3.8	33.1±3.8
DBP max (mmHg)	70.5±2.1	70.4±4.1	70.5±2.4	70.9±1.3	69.1±1.7	73.3±1.9	72.8±1.3	71.2±1.7	75.4±1.9	73.5±1.6	75.5±1.6	69.5±3.2
Change DBP (max - rest) (mmHg)	2.5±2.2	0.11±4.2	4.2±2.3	4.5±1.6	1.0±1.8	9.4±2.6	3.7±1.4	2.4±2.0	5.7±1.9	-0.30±1.5	0.0±1.8	-0.91±2.9
RPP max (rest HR x SBP max)	12655.2±776.8	11008.0±743.8	13795.5±1125.6	12714.4±368.3	12489.0±441.0	13029.9±642.4	13093.9±439.3	13140.9±632.9	13013.8±521.6	13581.0±504.9	12753.0±584.6	15237.1±774.5

**Conclusions:** Exercise duration in healthy children is a minimum of 12 minutes. 85% of maximum heart rate response is more achievable in children than maximum predicted heart rate hence it may be preferable in clinical decision-making. Maximum blood pressure response is much lower than historically quoted which may necessitate redefining hypertensive response to exercise in childhood.

**Abstract no: 1823**

**Proof of concept of a smart phone-based phonocardiographic system**

**Pouya Hemmati\*, Quan Ni†, Hanbo Tang#, Baishun Gao#, Bing Song#, Andreas Tsakistos‡ and Lee Pyles‡**

\*Medical School, University of Minnesota, Minneapolis, Minnesota, United States of America

#Department of Cardiac Surgery, First University Hospital of Lanzhou University, Lanzhou, Gansu Province, China

†Children's HeartLink, Minneapolis, Minnesota, United States of America

‡Amplatz Children's Hospital, University of Minnesota, Minneapolis, Minnesota, United States of America

**Introduction:** Clinical auscultation remains essential for the diagnosis of congenital heart disease in the developing world. In screening programmes of Lanzhou First University Hospital, Children's HeartLink, First Affiliated Hospital of Kunming Medical University and China-California Heart Watch in Peoples Republic of China, children were screened with phonocardiography (iAusc, n=52), standard auscultation (sAusc, n=19) and Echocardiography (echo). Our hypothesis is that iAusc detection of murmurs is sensitive to facilitate referral to a cardiac centre.

**Methods:** Children were examined with an electronic stethoscope connected to an iPhone® that stored, retrieved and transmitted the iAusc data, and then with a commercial stethoscope. Endpoints include pathologic vs. innocent murmur, location and intensity for iAusc and their overall concurrence with echo diagnosis. An echocardiogram was performed using standard techniques. Initial difficulty with high pitched vibratory innocent noises required different earphones.

**Results:** Organic murmur was differentiated from innocent murmur in each case with both iAusc (48/52, LAP) and sAusc (19/19). iAusc was recorded from the 4 primary cardiac exam areas on the chest, URSB, ULSB, LLSB, Apex. Principal diagnoses included: Ventricular Septal Defect (23), Pulmonary Stenosis (3), Atrial Septal Defect (5), Aortic Stenosis (2) and Normal (10). Sensitivity for murmur type was 48/52 or 92%. Specificity for innocent murmur was 10/14 (71%). Pulmonary hypertension was present in 3 of 4 murmurs in heart disease misdiagnosed as innocent.

Murmur	Number	Organic	Correct anatomic Dx	Innocent	Correct
Gansu	19	18	15	1	1
Yunnan	24	20	6	13	9
<b>Total</b>	<b>43</b>	<b>38</b>	<b>21</b>	<b>14</b>	<b>10</b>

**Conclusion:** Phonocardiography can be used to detect congenital heart disease. We report a proof-of-concept of iAusc to detect heart disease in underserved areas. The iAusc tracings can be emailed to remote experts from a cloud-based server, providing cost-effective access to care with reduced travel for disadvantaged children and a link to the cardiac referral centres. The methodology can provide a venue for international volunteerism for cardiologists based in advanced countries accounting for unfamiliar challenges such as Eisenmenger pulmonary hypertension.

**Abstract no: 1831****Reduction of radiation dose for coronary computed tomography using a 128-slice dual-source machine in children and young adults with coronary lesions after Kawasaki disease**

Shigeru Uemura\*, Kazuto Fujimoto\*, Hiroaki Kise\*, Hideshi Tomita\*, Nobuo Ooyama#, Madoka Sawada# and Soga Takashi#

\*Cardiovascular Centre, Showa University, Northern Yokohama Hospital, Yokohama, Japan

#Paediatrics, Showa University, Northern Yokohama Hospital, Yokohama, Japan

**Background:** Coronary artery lesions (CAL) from coronary aneurysm followed by Kawasaki disease (KD) have progressed to stenosis, occlusion and calcification. Coronary computed tomography (CCTA) has been providing much information on the diagnosis CAL after KD. CCTA using conventional 64-slice single-source machine provided high radiation exposure. A 128-slice dual-source CT (DSCT) can take coronary artery images quickly and reduce the radiation dose.

**Purpose:** The aim of this study is to evaluate the reduction of radiation dose and recognition of the CAL by DSCT.

**Patients and methods:** Thirty five patients (median age; 11 years 6 months, 2 years 5 months - 45 years) were examined. All patients were administered  $\beta$ -blockade to reduce the heart rate. 6 children were sedated and the other 29 patients could breath-hold. Scan parameters were as follows; mAs adapted body weight (100mAs - 310mAs) at 70Kv, 80Kv, 100Kv and 120Kv, with prospective ECG-gated. To assess radiation dose, we recorded the dose-length product (DLP) in mGycm and the effective dose in mSv estimated from the DLP. We used 2 modes to take CCTA, namely Flash Spiral Mode (FSM) and Adaptive Flash Cardio Mode (AFCM). The quality of images for proximal and middle segments of the right and left coronary arteries was evaluated.

**Results:** Twenty nine patients (median age; 11 years 6 months) were examined by FSM, and 6 patients (median age; 11 years 0 months) by AFCM. Total DLP was extremely low (median 45mGy cm) and effective radiation dose was 0.74mSv (median) by FSM. The total DLP was very low (median 128.5mGy cm) and effective radiation dose was 1.93mSv (median) by AFCM. 1 patient (aged 7) was not subjected to images by FSM because of movement.

**Conclusion:** Prospective ECG-gated DSCT can provide adequate CAL images in children and young adults. These methods are associated with highly reduced radiation doses.

**Abstract no: 1832****Analytic comparison of cardiovascular risk factors for adult congenital heart disease and normal control: A case-control study**

Ju Ryoung Moon

GUCH Clinic, Samsung Medical Center, Seoul, Korea

**Background:** Objectives of this study are to identify cardiovascular risk factors in adults with CHD and to provide basic materials for developing media to lower the risk factor levels.

**Methods:** This study conducted a survey of 240 people in total, including 120 adult patients with CHD and 120 patients as control group whose selection was based on age, sex, and body mass index (BMI). The survey was conducted regarding fasting blood glucose (FBS), lipid profile, Apoprotein A-I/B, occurrence of carotid stenosis, and environmental influences.

**Results:** Compared to the control group, the CHD group had significantly raised FBS, HDL, and Apoprotein A-I, but significantly lower total cholesterol, LDL, and Apoprotein B. The 2 groups had significant differences in the occurrence of carotid stenosis, exercise, and smoking. Regarding difference by sex between the 2 groups, males of CHD group had lower total cholesterol and LDH than males in the control group; females of CHD group had higher FBS and lower total cholesterol than females in the control group. However, there was no difference by gender in CHD group. Comparison of acyanotic and cyanotic CHD patients in CHD group showed that acyanotic CHD patients had higher FBS, total cholesterol, and LDH and lower Apoprotein than cyanotic CHD patients. When age and gender were adjusted, cyanotic CHD patients without surgery, cyanotic CHD patients with surgery, acyanotic CHD patients with surgery, and acyanotic CHD patients without surgery in this order had a high risk of developing metabolic syndrome ( $p < .001$ ). BMI and smoking were identified as variables influencing metabolic syndrome.

**Conclusions:** It is believed that regular follow-up of risk factors, BMI control, and non-smoking education may reduce the risk of developing metabolic syndrome in adult patients with CHD.

**Abstract no: 1836****Post-rheumatic cardiac valvular lesions management in a tertiary sub-Saharan centre: The experience of Shisong cardiac centre**

J.C. Tantchou Tchoumi\*, J.C. Ambassa\*, A. Giamberti#, S. Cirri# and G. Butera#

\*Shisong Cardiac Centre, St. Elizabeth Catholic General Hospital Kumbo, Cameroon

#Department of Cardiac Surgery, Instituto Policlinico Hospital, San Donato, Milan, Italy

**Abstract:** The aim of the study was to investigate the early post-surgical mortality and challenges in the care of operated children in St. Elizabeth Catholic General hospital's cardiac centre.

**Patients and methods:** This retrospective analysis included 56 patients aged from 6 - 17 years who underwent mitral repair or replacement and/or aortic valve replacement from November 2009 - June 2012. Data from patients' records, operative intervention, and pre-operative and post-operative 2-D echocardiographic studies were reviewed.

**Results:** Fifty six patients aged 6 - 17 years (mean age  $10 \pm 3.4$  years) underwent surgical correction of mitral and/or aortic valvulopathy. Mitral regurgitation was the commonest echocardiographic diagnosis present in 51.7% of the patients; 13.3% patients had mixed mitral valve disease and 35% had pure mitral stenosis. Before surgery, 8 patients were in class IV, 12 in class III, 25 in class II and 11 in class I according to the New York Heart Association's classification. Patients were extubated from 5 - 10 hours after surgery with low doses of inotropes. The mean stay in intensive care unit was  $1.5 \pm 0.5$  days. In the early post-surgical period, the ejection fraction (EF) changed from  $45.3 \pm 1.5\%$  -  $56.1 \pm 1.4\%$  ( $p < 0.005$ ) in 3 months and stayed almost the same after 6 months  $57.2 \pm 2.7\%$  ( $p > 0.05$ ); at 9 months it was  $55.1 \pm 1.8\%$  ( $p > 0.05$ ), at 12 months  $58.4 \pm 1.7\%$  ( $p > 0.05$ ); however the basal part of the interventricular septum was hypokinetic.

**Conclusion:** Post-rheumatic mitral valve regurgitation is the most encountered pathology. Post-surgical echocardiogram is characterised by motion abnormalities of the basal part of the interventricular septum. Due to financial limitation, poverty and illiteracy of parents, the post-surgical follow-up of patients is challenging.

**Abstract no: 1840****Anomalous origin of the pulmonary artery from the ascending aorta: 2 institutional reviews of cases from 1991 - 2012**

G. Dumani\*, P.A. Adams\*, B. Fourie\*, A.M. Cilliers\*, A. Nzimela#, E. Hoosen#, D. Reddy#, N.J. Buckles# and J. Ragadu#

\*Chris Hanu Baragwanath Academic Hospital, University of Witwatersrand, Johannesburg, South Africa

#Inkosi Albert Luthuli Hospital, University of KwaZulu-Natal, Durban, South Africa

**Introduction:** Anomalous origin of 1 pulmonary artery from the ascending aorta is a rare congenital anomaly associated with early onset of pulmonary hypertension and irreversible pulmonary vascular disease.

**Methods:** Retrospective clinical review of 18 cases presented to the Divisions of Paediatric Cardiology at Baragwanath Academic Hospital and Inkosi Albert Luthuli Hospital which are both tertiary care institutions in Southern Africa. Data collected included clinical features, diagnosis, operative procedures, pre-operative and post-operative follow-up.

**Results:** Sixteen infants, 1 child and 1 adult (11 males, 6 females) were diagnosed. The most common presenting features were respiratory distress, a cardiac murmur, congestive cardiac failure and failure to thrive. Median age at presentation was 67 days. Diagnosis was made with echocardiography and confirmed with CT angiogram (5) and angiography (5)s. There were 16 patients of anomalous origin with right pulmonary artery arising from the aorta (AORPA) and 2 cases of anomalous origin of left pulmonary artery (AOLPA). Patients were divided into 3 categories: isolated lesions (7), simple lesions with patent ductus arteriosus (8) and complex lesions (3). One patient with AOLPA had CATCH 22 and 2nd patient with AOLPA also had Mckusick-Kaufman syndrome. Five patients had successful direct re-implantation of the right pulmonary artery. One patient died the day after surgery following a pulmonary hypertensive crisis, 1 patient died 20 days after surgery from sepsis, and 3 patients remain well on follow-up. Three patients were deemed inoperable. The remaining 10 patients died before surgery could be undertaken.

**Conclusion:** There is a high mortality associated with anomalous pulmonary artery arising from the aorta without surgery. A good outcome can be expected with early surgery before pulmonary vascular disease.

**Abstract no: 1841****Infective endocarditis in children: Cross-sectional study in Ouagadougou at Burkina Faso**

N.V. Yameogo, K.J. Kologo, A.A. Yameogo, C. Yonaba, G.R.C. Millogo, B.J.Y. Toguyeni, A.K. Samadoulougou, S.Pignatelli, J. Simporé and P. Zabsonre

University of Ouagadougou, Ouagadougou, Burkina Faso

**Introduction:** Infective endocarditis is a transplant of a microorganism on a most often injured endocardium. It is rare in children. This work aimed to determine the frequency of endocarditis of the child, to describe clinical presentation, data from echocardiography, microbiological profile and clinical course.

**Patients and methods:** From 1 May 2010 - 30 April 2012, we consecutively included children received for infective endocarditis in 2 medical centres in the city of Ouagadougou: Saint Camille Medical Centre and Yalgado Ouedraogo teaching hospital. We investigated the functional and general signs and treatment already received. During physical examination we looked for an infectious syndrome, pneumonia, heart failure and entrance doors. Blood cultures, blood count, creatinine, blood chemistry, HIV status, electrocardiogram, chest radiography and cardiac Doppler ultrasound were systematic. The diagnosis of the disease was based on Duke's criteria.

**Results:** Thirty two cases of endocarditis in children (1.5% of admissions) were reported. Average age was  $4.6 \pm 2.5$  years (1-14). The sex ratio was 1.5 for girls. The clinical presentation was a common infectious syndrome. Impaired general condition and congestive heart failure were present on admission in 14 cases, respectively. The front door was dental in 14 cases (43.7%), skin and ENT in 6 cases (18.7%) respectively. A peripheral vein was implicated in 2 cases. In 4 other cases no front door had been found. HIV serology was positive in 6 cases. When the blood cultures were received 26 were positive: streptococcus (20) and staphylococcus (6). Echocardiography had revealed vegetations in all cases. These vegetations were localised on the mitral in 14 cases. Multiple locations were found in 8 cases. Underlying heart disease were dominated by rheumatic valve disease (19 cases), forms of heart health were found in 4 cases. Treatment consisted of antibiotics, antipyretic treatment as appropriate treatment for heart failure. Surgery was indicated in 3 patients in emergency but they were unable to benefit due to inadequate technical support. The evolution was marked by 4 deaths (25%) in an array of septic shock.

**Conclusion:** Infective endocarditis in children is common in our practice. The common clinical syndrome is infectious. Streptococcus and Staphylococcus are the two germs found. Surgical forms are immediately lethal. The global prognosis remains poor.

**Abstract no: 1843****Impact of SA Guidelines on the use of high sensitivity troponins – a pathology laboratory perspective**

R. Delpont\* and M. du Plessis#

\*Department of Chemical Pathology, University of Pretoria, Pretoria, South Africa

#AMPATH (Pathology laboratories in private practice), Pretoria, South Africa

**Introduction:** Guidelines on the use of high-sensitivity cardiac troponin (hs-cT) for the diagnosis of myocardial infarction (MI) in South Africa have recently been published (SA Heart, 2012). The aim of the study was to evaluate the effect of implementation of these guidelines on hs-cT laboratory measurements.

**Method:** A retrospective comparative study was conducted. Results were made anonymous by using codes to protect patients' privacy. Clinical presentation was not recorded. The 4th generation hs-cT assay (Roche Diagnostics, South Africa) was employed for hs-cT measurements.

**Results:** Total number of requests did not change significantly but a significant shift in distribution of values within hs-cT categories (<15ng/L; 15-52ng/L; 53-100ng/L;  $\geq 100$ ng/L) was observed (chi-square=23.7;  $p < 0.0001$ ). 14% of values exceeded 100ng/L for both groups. Significantly higher repeat rates were observed following implementation of the guidelines (33 vs. 42%; chi-square=5.9;  $p < 0.015$ ) as well as lower numbers of patients within hs-cT category 2 with a >50% increases in hs-cT (18% vs. 9%; chi-square=4.9;  $p < 0.026$ ). 35% of patients within hs-cT category 3 currently present with a >20% increases in hs-cT. Serial daily measurements (>3 days) were requested in 3% of all patients. A significant change was observed in time of repeat assessments ( $\leq 3$ hours,  $\leq 6$ hours,  $\leq 12$ hours, >12hours) following implementation of the guidelines (chi-square 8.5;  $p = 0.036$ ).

**Conclusion:** A significant change in laboratory observations is evident. Although this study does not specifically inform on the use of hs-cT to diagnose MI, implementation of the guidelines appears to be beneficial, irrespective of the indication for the test.