TRACK 4: ADULTS WITH CONGENITAL HEART DISEASE, AND THE PREVENTION OF ACQUIRED HEART DISEASE STARTING IN CHILDHOOD

Abstract no: 76

Elevated lipoprotein (A) in a newborn with thromboembolic disease and a family history of atherosclerosis

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Background: Thromboembolic disease is uncommon in young children. Catheters, sepsis and inherited diseases are important risk factors for thrombosis. Familial hyperlipoprotein A [Lp (a)] is an inherited condition that leads to higher levels of Lp (a). Lp (a) is a subclass of low-density lipoproteins, plasminogen-like with thrombogenic properties.
Case description: The male infant was born by Caesarean section at 37 week's gestation. The baby was cyanotic with little improvement in the delivery room. The 1st echocardiogram (Echo) demonstrated pulmonary arterial hypertension (PASP=80mmHg). During the 1st days of life, the newborn developed intracranial haemorrhage and worsening of cyanosis. The follow-up Echo showed 2 small clots within right ventricle and a larger one (13.4 x 9.2mm) near the left pulmonary artery with flow obstruction. Radionuclide scanning showed signs of pulmonary thromboembolism. An umbilical venous catheter was removed. Enoxaparin was administered to the patient (1.5mg/kg SQ q12h). The results of the collected blood tests were normal (thyroid hormones, liver enzymes, glucose, white blood cell and platelet count, haemoglobin electrophoresis, factors IX, V and VIII). Due to haemodynamical instability, the newborn didn't undergo cardiac surgery. The conservative treatment showed clinical improvement and gradual reduction of clots. He was discharged on oral anticoagulation (warfarin). On further investigation, the prothrombin gene mutation, the anti-cardiolipin antibodies and the proteins C and S were collected and were negative. The family history was positive for premature heart disease. Serum levels of lipoprotein (a) were obtained from the patient and his parents. The child (55mg/dl) and his mother presented higher levels of Lp (a). Anti-agregant (aspirin) was prescribed. After neonatal period, the patient presented no new thrombus.
Conclusions: Congenital thrombophilia needs to be strongly considered in neonates with a clinically significant thrombosis. The increased Lp (a) is a risk factor for thrombosis, coronary artery disease and cerebrovascular.

Abstract no: 108

Self-reported health-related quality of life in children and adolescent with heart disease: A Swedish registry study

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Background: During the last decades survival of children with congenital heart disease has improved significantly. It is important to assess HRQoL (health-related quality of life) in these patients. Some of the variables in the Swedish national registry of congenital heart disease, Swedcon, consists of HRQoL measurements.
Aim: To describe HRQoL in children and adolescents with heart disease.
Materials and methods: A descriptive study based on data from Swedcon, collected from 1 paediatric cardiac outpatient-clinic. The sample consisted of 104 girls and 154 boys, 9 - 18 years old with registrations of HRQoL variables and the questionnaire Disabkids short version. The patients were divided in 3 groups depending on the number of cardiac operations or catheter treatments they had undergone.
Results: Disabkids mean total score (max 100) in patient-group 1 (no cardiac procedures) was 92.9 in group 2 (1 cardiac procedure) 94.4 and in group 3 (2 or more cardiac procedures) 86.0. NYHA 2 and 3 were more frequent in patient group 2 and particularly in group 3. Patients in groups 2 and 3 had significantly more cognitive difficulties than those with no cardiac procedures. However >50% of all patients had more than 3 hours of physical activity/week.
Conclusions: In this unselected group of patients with congenital heart disease it is shown that HRQoL is better for children with fewer cardiac procedures and that HRQoL generally is better than for other chronically ill children, which confirms previous results on selected materials. In this single-centre study a web-based registry was used for evaluation of HRQoL in children and adolescents with heart disease. In the future it may be used for larger groups, such as the national population of children with heart disease.

Abstract no: 114

Compliance with lifestyle recommendations in children and young adults with hypertrophic cardiomyopathy

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Background: Hypertrophic cardiomyopathy (HCM) is the most common medical cause of sudden death during exercise. Previous studies suggest that restriction in competitive sports participation result in lower mortality rates. This study evaluates the effects of lifestyle recommendations on physical and leisure-time activities in HCM-patients.
Materials and methods: Twenty seven consecutively recruited asymptomatic patients with HCM diagnosed through family screening were asked to participate in the study. All received life-style recommendations according to international guidelines. 20 (median age 14.5 years, range 5 - 25) filled out a questionnaire regarding the frequency of strenuous physical activities (defined as becoming exhausted), sports participation and leisure time activities (such as spending time with friends, using a computer, watching TV, visiting the cinema, theatre or playing or listening to music), before diagnosis and after 1 year. Patients were classified in 5 - 6 groups according to the frequency of activities. Results were analyzed using Wilcoxon signed rank test.

Results: There was a significant decrease in the frequency of strenuous physical exercise from before diagnosis compared to 1 year later (p=0.002). 40% performed strenuous exercise >7 hours/week before diagnosis compared to 5% 1 year later. The number of patients who never participated in sports activities increased from 10% - 20% (p=0.007). No change was detected regarding leisure time activities. Time spent watching TV and using a computer did not increase significantly.
Conclusions: Life-style recommendations significantly affect the physical activity habits in HCM patients. Our results indicate a high level of compliance to the recommendations. Leisure time activities do not seem to be negatively affected neither did time spent on sedentary activities increase.

Abstract no: 126

Teenage cardiopulmonary exercise function changes with growth and increasing thoracic cavity volume

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Background/hypothesis: We looked at the importance of 3 years of normal teenage growth as well as the influence of surgically increasing thoracic cavity size on cardiopulmonary exercise function. Thoracic cavity enlargement was investigated in patients undergoing surgery for pectus excavatum (PE). We hypothesised that teenagers would increase cardiopulmonary function with growth and even more when acquiring a larger thoracic cavity.
Materials and methods: Healthy teenagers (n=26, age: 15.0±1.9 years) and patients (n=49, age: 15.5±1.7 years) were investigated at rest and during bicycle exercise 3 times each from the age of 15 - 18. PE-patients were investigated before operation, 1 year and 3 years postoperatively. Cardiac output, heart rate and aerobic exercise capacity were measured using a photo-acoustic gas-rebreathing technique during rest and exercise and indexed to body surface area. Sternal to spine diameter was measured using MRI.
Results: The 2 groups did not differ in age or BMI at any point during the investigational period. The sternal to spine diameter increased from 5.2cm - 8.9cm in the PE-patients undergoing surgery and from 8.2cm - 9.0cm in healthy teenagers during the same period. At age 15, healthy teenagers obtained a maximum cardiac index (CI_{max}) of 8.1±1.0 l/min/m^2 during exercise which was higher compared to age-matched PE-patients (6.6±1.2l/min/m^2), (p=0.0001). Three years later, teenagers had increased their CImax with 5.8% (8.3±1.6l/min/m^2). However, a larger increase of 24.5% (8.1±1.2l/min/m^2) was seen in the surgically corrected patients. No correlation was found between CImax and sternal to spine diameter. Maximum heart rate did not differ at any point between the two groups and VO_{2max} was unchanged.
Conclusion: Maximum cardiac index increases in healthy teenagers during growth from the age of 15 - 18 years. When age-matched teenagers with pectus excavatum undergo surgery with enlargement of the thoracic cavity a significantly larger increase is observed.

Abstract no: 128

Ventricular hypertrophy in adults operated for ventricular septal defect as toddlers

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Background: Cardiac morphology is expected to restore to normal after surgical closure of ventricular septal defects in early childhood. However, long-term abnormalities in cardiac morphology may exist and are the subject of this study.
Methods: Patients (n=15), median age at surgery of 2.6 years (1.5 - 4.1 years) and 21.1 years (19.8 - 23.2 years) at the time of examination, and age-matched control subjects (n=10) underwent Cardiac Magnetic Resonance scanning using a Philips Intera, 1.5T MR scanner. Stacks of 3 contiguous long-axis and 12 short-axis slices encompassing both ventricles were acquired. Quantitative flow measurements were made using phase-contrast gradient imaging. For data analyses OsiriX and Medviso Segment software were used.
Preliminary results: Compared to controls left ventricular ejection fraction was unaffected, median 63.2% (56.6 - 68.7%) vs. 65.9% (53.5 - 69.9%), p=0.99, and so was cardiac index, median 3.6 L (min m^2)-1 (3.2-4.1 L (min m^2)-1) vs. 4.0 L (min m^2)-1 (3.6-4.1 L (min m^2)-1), p=0.81. Ventricular mass indexes were larger in VSD patients, p<0.05 in both ventricles. Left ventricular peak ejection/filling rate was higher in patients; median 87.3ml sec-1 (72.5 - 111.3ml sec-1)/145.5ml sec-1 (82.7 - 174.5ml sec-1) compared to controls 69.3ml sec-1 (55.0 - 87.1ml sec-1)/87.4 ml sec-1 (76.3 - 126.2ml sec-1) in control subjects, p<0.05 and p=0.08, respectively. In contrast, right ventricular peak ejection/filling rate was lower in patients; median 57.7ml sec-1 (50.4 - 73.0ml sec-1)/50.4 ml sec-1 (37.0 - 60.2ml sec-1) as compared to controls 89.9ml sec-1 (48.5 - 131.0ml sec-1)/94.5ml sec-1 (51.1 - 137.2ml sec-1) in control subjects, p<0.01 for both parameters.
Conclusion: Twenty years after surgically closed VSDs, larger ventricular masses combined with superior peak rates of ejection and filling in the left ventricle are noticed. In contrast, right ventricular peak ejection and filling velocities were inferior compared to control subjects. The consequence for long term outcome is unknown and needs further studies.

Abstract no: 134

Menstrual bleeding after open heart surgery

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Background: We investigated whether open heart surgery with the use of extracorporeal circulation has an impact on menstrual bleeding (MB).
Material and methods: MB pattern was registered retrospectively and menstrual bleeding during admission was registered prospectively in fertile women undergoing heart surgery for congenital heart disease. Haematocrit and 24-hours post-operative bleeding were compared with men also undergoing congenital heart surgery in the period 2010 - 2011.
Results: Women (n=22), mean age 35 years (range: 17 - 60) were operated and hospitalised for 4 - 5 post-operative days. Mean post-operative bleeding in the 1st 24 hours was 312ml (range 50 - 1 442ml). 3 - 4 (16%) women were expected to have MB during their hospital stay. Unplanned MB (lasting 2 - 5 days) was

detected in 13 patients (60%). Six had expected MB and 3 had, as expected, no MB during hospital stay. Of the 13 unexpected MB, 4 were 1-7 days early; 4 were 8 - 14 days early; 3 were 1 - 7 days late; and 2 had MB despite having had MB within the last 2 weeks. None had unusual large or long-lasting MB. Ten women took oral contraceptives, 7 of whom had unexpected MB. Men (n=22), mean age 35 years (17 - 54) had a mean 24 hours post-operative bleeding of 331ml (range 160 - 796ml) which was not significantly different from the women. The mean pre-operative haematocrit was 40% (29 - 53%) among men, being not significantly different from women (mean 40% [32 - 60%]).
Conclusion: MB patterns are disturbed by open heart surgery in the majority of fertile women. Nevertheless, the unexpected MB is neither particularly long lasting nor of large quantity, and the post-operative surgical bleeding is unaffected. We recommend information about irregular menstrual bleeding, but no special precautions when operating women in fertile age.

Abstract no: 138

Limited knowledge among local cardiologists of the management of pregnant women with mechanical heart valves and minimal impact of focused education

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Background/hypothesis: There are significant risks to the mother and fetus in pregnant women with mechanical heart valves. While these patients are frequently managed by experienced multi-disciplinary teams, general cardiologists should be aware of the relevant issues. We assessed the knowledge of local cardiologists on these issues and evaluated the impact of focused education on the subject.
Methods: A questionnaire on the principles of management of pregnant women with mechanical heart valves was distributed to cardiology consultants and trainees from the U.K.'s Yorkshire and Humber region at an educational meeting. A lecture was then given and a leaflet summarising the issues circulated. At a further meeting 11 months later the participants were re-questioned.
Results: The questionnaire was completed by 35 doctors on the first occasion and 26 on the second. The responses are summarised in the Table.

Question	Accepted answer	% correct, 1st questioned	% correct, re-questioned
What is the risk of maternal death?	1 - 5%	48%	69%
What is the risk of fetal wastage?	20 - 40%	18%	31%
What is the risk of major bleeding?	1 - 4%	23%	23%
What is the risk of warfarin embryopathy?	5 - 6%	20%	31%
By what gestation should warfarin be stopped to avoid embryopathy?	6 weeks	3%	15%
The risk of warfarin embryopathy may be lower if daily dose is <? mg	5mg	49%	31%
How does pregnancy alter the pharmacokinetics of low molecular weight heparins?	Various	40%	38%
How would you determine the dose of low molecular weight heparin?	Anti-factor Xa levels	40%	35%
Rank in order thromboembolic risk during pregnancy: A) heparin throughout, B) warfarin throughout, C) heparin first trimester, warfarin thereafter	A>C>B	49%	35%

Conclusions: Knowledge among local cardiologists regarding the management of pregnant women with mechanical valves was limited. There was also a limited effect of improving knowledge with targeted education.

Abstract no: 139

Vascular function in adults with repaired coarctation of the aorta: Assessment by multi-modal MRI

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Background/hypothesis: Patients with surgically repaired coarctation of the aorta (CoAo) remain at increased risk of premature cardiovascular events including myocardial infarction and stroke. Magnetic resonance imaging (MRI) can be used to assess a number of measures of vascular function, including endothelial function (as flow-mediated dilatation (FMD) of the brachial artery) and large artery stiffness (as aortic pulse wave velocity (PWV) and distensibility).
Aim: Our aim was to gather preliminary comparative data on vascular function by MRI in adults with repaired coarctation and healthy controls.
Methods: Seven patients with previous coarctation repair and 14 age-matched healthy controls underwent multimodal MRI assessment of vascular function. Blood pressure was measured in the right arm. Distensibility of the ascending aorta was measured at the level of the right pulmonary artery. Phase contrast MRI was used to measure aortic arch PWV. The transit time between the arrival of the flow wave at ascending aorta and the proximal descending aorta was

calculated by a foot-to-foot method. Cine imaging of the right brachial artery was performed to measure brachial artery reactivity to reactive hyperaemia (FMD) and glyceryl trinitrate. Comparisons were made using Student's t-tests. Data are expressed as mean(SD).
Results: In 2 patients PWV data was of poor quality and thus excluded along with the corresponding data from controls. Ascending aortic distensibility was <in patients than in controls [4.3(2.2) vs. 7.2(2.1) mmHg-1.10 - 3, p<0.01], aortic arch PWV was higher [6.3(1.5) vs. 4.3(1.1) m/s, p<0.05]. There was no difference in brachial artery FMD [13(10)% vs. 16(6)%, p=0.3] or response to glyceryl trinitrate [22(12)% vs. 27(7)%, p=0.08].
Conclusions: In small numbers MRI appears to have identified differences in vascular function between patients with repaired coarctation and controls. Incorporation of these measures into surveillance MRI assessment of repaired coarctation may determine whether they have any prognostic value in the long term.

Abstract no: 153
Women with congenital heart disease: Their imperative towards conceiving

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Background: Advances in medical care have increased the number of women with CHD reaching child-bearing age and wishing to conceive. Pregnancy adds an additional stress on their compromised cardiovascular system, with risks to both themselves and their unborn child. Despite these risks, they still proceed to conceive.
Aim: A qualitative study was done to determine their motivations for conception.
Methods: A 2-group design of women with (n=20) or 20 without (n=20) CHD, >18 years of age who had completed 1 or more successful pregnancies were recruited from a tertiary centre and private clinics. The women completed a questionnaire and a semi-structured interview. Their medical records were reviewed. Thematic analysis was conducted on the interview data.
Results: CHD in the women ranged from minor lesions (e.g. atrial septal defect) to moderately severe lesions (e.g. Fontan circulation). The motivations for conceiving in women with CHD were similar to that of healthy women. These motivations included the influences of relationships with their partners, family and friends; concern for the reproductive changes associated with increasing age; their innate desires for motherhood; the women's personal goals; and cultural and social expectations. Women with CHD had a tendency to underestimate the severity and associated risks of their CHD. They exhibited a strong reliance on their treating clinicians and assumed advances in medical care would carry them through their pregnancies.
Conclusions: Motivations to conceive are similar for healthy women and women with CHD. Those with CHD had a tendency to harbour an unrealistic understanding of the severity of their CHD and its implications in pregnancy. This perception emphasises the importance for clinicians caring for these patients to be knowledgeable about the impact of CHD on the woman's pregnancy and on their CHD, as they carry great responsibility in caring for these patients.

Abstract no: 201
Surgery of Tetralogy of Fallot in adults in Sudan

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Background: Corrective surgery of Tetralogy of Fallot (rToF) beyond childhood is very rare due to improved methods of early detection and early repair. In our centre, 20% of ToF patients underwent repair were adults with good early results.
Method: This is a descriptive retrospective study. The data was collected by the authors from the records of the patients who underwent surgical repair of ToF from 2003 - 2011 in our centre. We include all patients >18 years. The data was collected manually and analysed by a computer program SPSS.
Results: Surgical repair in adults represent 20% of ToF surgery. 14 patients were male (75%). The mean age is 20.47 years. Eleven patients were in NYHA III. 60% of patients had pulmonary gradient >50mmHg. No previous palliative surgery or associated major cardiac anomalies occured. All patients underwent total surgical repair through right atriotomy and the VSDs were closed by synthetic patch. In 1 patient the pulmonary artery was opened and enlarged with pericardial patch. The mean follow up time was 4.5 years. Two patients died during their hospital admission and another 3 died follow-up period.
Conclusion: Although operating on adult patients with ToF is a challenge and rarely seen, we still had a reasonable number of patients with good early surgical outcome but poor in the mid-term follow-up period.

Abstract no: 233
Demonstration of strong feasibility of the iHeartChange website for transitioning patients with congenital heart disease

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Background: Published guidelines recommend initiatives to improve the rate of transfer and quality of transition of patients with congenital heart disease (CHD) from paediatric-t-adult cardiac care. Unfortunately, there are few published interventions for transitioning patients with CHD, proposed initiatives are often resource intensive, and strategies for engaging adolescents are unknown.
Aim: We created and evaluated the feasibility of the iHeartChange website targeting transitioning CHD patients.
Methods: Patients transferring from a paediatric to adult CHD program in a 1 year period were invited to participate in the study. Feasibility outcomes, specifically eligibility, participation rate, and patient feedback about the website, were investigated.

Results: A total of 207 CHD patients aged 16 - 19 were transferred, of whom 187 (mean age=17.7 years, 58% male, 75% with moderate/great defect complexity) met full inclusion criteria and were invited to participate. Thirty four percent (63/187) of eligible patients logged on and completed baseline surveys; there were no differences in age, sex, or defect complexity between patients who did and did not complete baseline surveys. Forty eight patients completed follow-up surveys at least 1 month after initial website access. The majority of patients mostly or strongly agreed that the website was: easy to use (95%); fun to use (82%); = gave information that was useful (91%); information could be trusted (95%); gave a better understanding of CHD medical issues (86%); should be offered to all patients moving to adult care (91%); and is something they would use again if available (87%).
Conclusion: Strong feasibility of the iHeartChange website was demonstrated. 1/3 of patients with CHD chose to access a website as part of a research study; higher usage is anticipated if offered in a clinical setting. Adolescent feedback in terms of design and content was extremely positive. In conclusion, a website targeting transitioning CHD patients holds significant clinical promise.

Abstract no: 258

Changing risk factors for arrhythmias and sudden death among adults with repaired Tetralogy of Fallot (rToF) in the current era

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Background: Among adults patients (pts) with repaired congenital heart, those with Tetralogy of Fallot (rToF) historically have exhibited increased risk for tachyarrhythmias (Ar), AV block (AVB) and sudden cardiac death (SCD) associated with right ventricular (RV) systolic pressures (RVSP) >60mmHg, outflow tract (OT) gradients >20mmHg and QRS duration >180ms. However, surgical practices have changed over the years. The purpose of this study was to evaluate Ar/SCD risk with symptoms, echocardiographic (ECHO), electro physiologic (EP), haemodynamic and RV volume on Cardiac MRI (CMRI) findings among the current generation of older rToF pts.
Methods: ToF pts seen in our Institution's Adult and Adolescent Congenital Heart programmes were divided into 2 groups: Group I: +Ar/SCD; Group II -Ar/SCD, and correlated with current pt age, gender, age at surgical repair, repair types (transannular patch; no patch; conduit; shunt), ECHO, CMRI, ECG/Holter, haemodynamic and EP results.
Results: Of 136 pts (66 male/70 female) ages 11 - 58 years (mean 26), 61 (45%) had Ar (22% atrial, 60% ventricular, 10% both) and 8% AVB. Of these, SCD occurred in 3 (5%) (Group I). These pts were chronologically older (mean 32 vs. 22 years) with repair performed at an older age (mean 50 vs. 22mos) than those without Ar/SCD (Group II) ($p<.05$). QRS duration (mean 158ms) and RVSP (mean 44mmHg) were persistent risk factors. However, there was no correlation with type of surgical repair, gender, RV pressure >60mmHg, RVOT gradient >20mmHg, or RV volume. Ar was induced in 91% of Group I pts studied, requiring ablation or device implant.
Conclusions: In the current era, rToF pts still remain at risk for Ar/SCD depending on their chronologic age and age at surgical repair. However, QRS duration is shorter (158ms) and RVSP less (44mmHg) than previously reported. Neither type of repair, RV outflow gradients, nor CMRI volumes correlate with Ar/SCD.

Abstract no: 362

Surgery for adults with congenital heart disease: An 11 year single-centre experience

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Background/hypothesis: Adults with congenital heart disease (ACHD) are increasing in number as a result of asymptomatic lesions during childhood or improved paediatric cardiac care. Some of these patients are not discovered in childhood, while others require subsequent cardiac interventions. We retrospectively reviewed the early and late outcomes of ACHD operations/re-operations over the last decade.
Materials and methods: From January 2001 - December 2011, 1 044 congenital lesions were repaired during 659 surgeries in 538 patients (287 males) (Table 1). Mean age at operation was 32.5±13.1 (range: 18 - 84) years. Excluding rhythm-related problems from post-operative analysis, 200/449 (45%) were 1st time operations, and 98/449 (21.9%) had NYHA Class symptoms 3 or 4 pre-operatively.

Procedure	Repair # (EM/LM)	Replacement # (EM/LM)	Total # (EM/LM)
Pulmonary Valve	8 (-/-)	185 (2/3)	193 (2/3)
Aortic Valve	17 (-/1)	131 (3/3)	148 (3/4)
Aortic Repair			115 (1/2)
Rhythm Related			262 (N/A)
Other			326 (5/12)
Total			**1 044**

Results: Early and late mortality (EM) (LM) was 1.3% (n=6) and 2.9% (n=13) respectively. Actuarial survival at 1, 3, 5, and 10 years was 98%, 96%, 94%, and 86% respectively. Major post-operative complications occurred in 54/449 (12.0%). Re-operations were required in 7.1% (n=32). Actuarial freedom from re-operation at 1, 3, 5, and 10 years was 98%, 92%, 87%, and 70% respectively. At mean follow-up of 33.1±35.7 (range: 0 - 137) months, 3.1% (n=14) had residual Class 3 or 4 symptoms, although 4.5% (n=20) had minimal follow-up.
Conclusions: A wide variety of CHD pathology is seen in adults. Surgical repair can be accomplished with low mortality and morbidity. Pulmonary valve, aortic valve, aortic pathologies and rhythm abnormalities dominate the spectrum of ACHD pathology.

Abstract no: 379

Mode of delivery and pregnancy outcome in a tertiary centre for adult congenital heart disease

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Background: As a result of the continuing improvement of congenital heart disease (CHD) therapy, a growing number of women are reaching childbearing age. Contemporary maternal risks of pregnancy and preferred modes of delivery are not well known.
Aim: To review all deliveries of CHD patients in our centre since 2000, regarding mode of delivery and maternal outcome.
Results: There were 391 deliveries. Mean age at delivery 30.3 (±5.5) years, age range 17.4 - 48.5 years. 208 women had 1 delivery (4 twins), 58 (2), 17 (3) and 4 (4). 231 (59%) deliveries were spontaneous, 94 (24%) by Caesarean section (CS) and 66 (17%) after medical induction of labour. 120/231 (52%) spontaneous deliveries were entirely uneventful. The rest had premature rupture of membranes (26), fetal distress (23), bleeding requiring transfusion (10), maternal arrhythmia (3), vacuum delivery (30), forceps (6) and perineal laceration grade II or more and/or uterine exploration. 56/66 successful medical inductions of labour were for cardiac indications: shunt lesions (18), valve disease (17), TGA complex (10), ToF (7), Fontan (2) and PFO post stroke (2). 58/94 (62%) CSs were elective (both term and pre-term). Seven were for very high risk CHD: Eisenmenger syndrome (4), PHT (2), TGA (1) – Rastelli, Fontan (1). 33 CS were urgent. 2/4 Eisenmenger patients died. 2 more patients died, both with repaired DSS: 1 crashed after induction and 1 died at home 3 days after an uneventful delivery. There were 7 more serious complications but with good outcome.
Conclusions: This large single centre series of deliveries in CHD, shows that, except for Eisenmenger patients, maternal outcome is very satisfactory with very low cardiac complications rate in all modes of delivery. A > expected rate of CS suggests that the threshold for this intervention was <in CHD patients.

Abstract no: 448

Sudden cardiac death in children and adolescents: It is preventable with a multi-disciplinary, multi-institutional approach

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Background/hypothesis: SCD in children and adolescents occurs with unclear frequency and is a devastating event. There are many etiologies of SCD in children/adolescents. We hypothesise that survival is achievable with a multi-disciplinary/multi-centre primary and secondary prevention approach emphasising awareness/education as well as advocacy for secondary prevention approaches (lay-public CPR, education, AED use and placement of school CPR-AED programmes.
Material and methods: Project Adam (PA) was initiated in the state of Wisconsin at Children's Hospital of Wisconsin in November 1999. Project ADAM is a primary and secondary prevention programme. Warning sign education, teaching importance of comprehensive pre-participation exam, advocacy for individual school emergency-preparedness plans and school CPR-AED programmes are all part of PA. Since the inception, affiliate PA programmes have been established in other states: Georgia, Pennsylvania, Florida, Illinois, Alabama, Washington, Tennessee, Texas and Michigan.
Results: Project Adam has saved 60 lives since November of 1999. Of the total, 24 lives of children/adolescents and 36 lives of adults have been saved: Wisconsin (total 23: children/adolescents (10), adults (13)); Georgia (total 29: children/adolescents (11), adults (18)); Pennsylvania (total 4: children/adolescents (2), adults (2)); Texas (total 2: children/adolescents (1), adults (1)).
Conclusions: PA has saved the lives of both children/adolescents and adults. Education/awareness and implementation of CPR-AED programmes across the country aimed at children and adolescents will have the ability to save many mores lives, adults included.

Abstract no: 460

Survey results of pregnancy and contraceptive advice provided to women with adult congenital heart disease

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Background: As more women with congenital heart disease survive to reproductive age, accurate education and counselling on the risks surrounding pregnancy and childbirth is important to enable a positive outcome for both mother and baby. All women with cardiac disease of reproductive age should have access to pre-pregnancy counselling.
Materials and methods: We report the information remembered following contraception and pregnancy counselling by physician or specialist nurse in an outpatient ACHD tertiary centre clinic. A telephone questionnaire was used to survey women of child-bearing age who had attended the clinic in the previous 18 months.

Results: Fifty three women completed the questionnaire. Twenty one women had no understanding or only a brief idea regarding their cardiac condition prior to attending the clinic, with an increase in understanding reported by all patients following their appointment. Prior to the clinic, 18 of 53 (34%) reported receiving contraceptive advice and 27 of 53 advice regarding pregnancy. Following their clinic appointment, a further 23 (43%) women remembered receiving advice about contraception and pregnancy. Thirty one women (59%) reported using contraception, including 11 women using oral contraception. Other methods included barrier contraception (n=6), progesterone-only pill (n=4), Implanon (n=2), Depo injection (n=1), coil (n=5) and 2 sterilisations. Twelve women (55%) had been seen in a cardiology clinic during pregnancy with the same number remembering having a fetal echocardiography. Twelve women were planning a pregnancy; 8 (67%) had been offered a specialist pre-pregnancy clinic appointment and 6 had already received pre-conceptual counselling. Three women found their counselling extremely helpful and 1 slightly helpful.
Conclusions: Counselling by a physician/specialist nurse is found to be beneficial and increases the understanding of the cardiac patient. Providing good patient information to women continues to present a challenge to the outpatient clinic, and more women would benefit from being given advice regarding pregnancy and contraception.

Abstract no: 505

Metabolic syndrome-related characteristics are associated with an increased carotid-femoral pulse wave velocity in children

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Background/hypothesis: The origins of cardiovascular disease are in childhood, with changes to vessel structure and function often apparent from early life. Carotid-femoral pulse wave velocity (PWV) is a validated non-invasive measure of arterial stiffness - a determinant of vascular function. We hypothesised that features of Metabolic Syndrome – obesity, insulin resistance and hypertension – would be associated with higher PWV in children.
Materials/methods: Carotid-femoral artery pulse wave velocity was performed on a population cohort of 147 twin pairs aged 7 - 11 years. Fasting bloods, blood pressure, anthropomorphic and other measures were collected concurrently. The participants were analysed as individuals by multiple linear regression, after adjusting for age and sex. Further between twin pair analysis was performed to investigate the effect of twin pair clustering.
Results: Of the 147 twin pairs, 47 were monozygotic and 100 were dizygotic. Mean age was 9.03 years, and half were male. There were no significant differences between the characteristics of monozygotic and dizygotic twins. Mean PWV was 5.95m/s, SD 0.655. In individual-based analysis, age (ß=0.262, 95%CI 0.170-0.348) and height (ß=0.015, 95%CI 0.004-0.027) were positively associated with PWV, as were markers of adiposity; weight (ß=0.012, 95%CI 0.002-0.023), truncal skin-fold thickness (ß=0.006 95%CI 0.001-0.010), waist circumference (0.011 95%CI 0.000-0.021) and hip circumference (ß=0.010 95%CI 0.000-0.020). Strong associations were found between markers of insulin resistance: HOMA (ß=0.116 95%CI 0.051-0.181) and c-peptide (ß=0.588 95%CI 0.248-0.927) - and increased PWV. Dyslipidaemia was also associated with increased PWV, most strongly with increased triglycerides (ß=0.278 95%CI 0.082-0.476). There was a strong association with increased systolic (ß=0.015 95%CI 0.008-0.021) and diastolic blood pressure (ß=0.034 95%CI 0.023-0.044). Relationships weakened within twin pairs – suggesting that the associations are at a familial level.
Conclusions: Accepted adult characteristics of Metabolic Syndrome are associated with increased PWV in pre-pubertal children.

Abstract no: 581

New predictors of sustained ventricular tachyarrhythmia in repaired Tetralogy of Fallot (rToF)

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Background: Repaired Tetralogy of Fallot (rToF) patients are at risk ventricular tachyarrhythmia and sudden cardiac death (SCD). Risk stratification for arrhythmia remains difficult.
Aim: Investigation of whether cardiac anatomy and function predict arrhythmia.
Methods: 154 adults with rToF, median age 30.8 (21.9 - 40.2) years, were studied with a standardised protocol including cardiovascular magnetic resonance (CMR) and prospectively followed-up over median 5.6 (4.6 - 7.0) years for the pre-specified endpoints of new-onset ventricular tachyarrhythmia (sustained ventricular tachycardia/ventricular fibrillation).
Results: Nine patients had ventricular tachyarrhythmia (6%) during follow-up. Patients who developed ventricular tachyarrhythmia were older (42.5 [34.9 - 50.2] vs. 29 [21 - 40] years; p=0.01), had a later repair (12.8 [6.2 - 13.9] vs. 4.4 [2 - 8] years; p=0.02), larger akinetic right ventricular outflow track (RVOT) region (length 55 [34 - 60] vs. 30 [20 - 40] mm; p=0.002) and a lower RV ejection fraction (42 [40 - 52] vs. 53 [51 - 55] %; p=0.01), compared to the other patients. On univariate Cox analysis, RVOT akinetic region length and RV ejection fraction were predictive of ventricular tachyarrhythmia. On stepwise Cox regression analysis, the RVOT akinetic region length was the only remaining predictor (Hazard ratio 1.05, 95% Confidence Interval 1.01 - 1.08 per mm; p=0.004). The survival ROC curve analysis indicated a cut-off value of 30mm as a predictor of VA during 6 year follow-up with an AUC of 0.77, sensitivity of 83% and specificity of 61%. RVOT akinetic area length >30mm predicted reduced VA-free survival (Logrank p=0.002).
Conclusions: RVOT akinetic region length predicts ventricular arrhythmia in late follow-up of rToF. This is simple, feasible measurements for inclusion in serial surveillance and risk stratification of rToF patients.

Abstract no: 583

Is grownup congenital heart disease (GUCH) different in a developing country? Experience from a tertiary care hospital in Pakistan

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Background: In the current, era the number of grownup congenital heart disease (GUCH) patients undergoing surgical intervention are increasing. There is considerable literature to suggest that most of the interventions in the developed countries are in the form of redo-operations in patients who had previously undergone repair, palliation or correction. However in the developing country like ours, most of the interventions are primary and corrective.
Methods: This is a descriptive study of 176 GUCH patients >16 years who underwent surgical intervention. Data was acquired from cardiac surgery database >6 years from July 2006 - June 2012. The majority of surgical procedures were performed by paediatric cardiac surgeon and as 1st time surgery.
Results: Out of 176 patients there were 54.3% males, with the age ranging from 16 - 76 years (mean 37.3 years). The majority of the patients underwent surgical interventions for: closure of atrial (n=77); ventricular septal defect (n=36),patients were operated for Tetralogy of Fallot (ToF: 16); NS coarctation of aorta (5). The average length of stay was 7.4 days. Overall mortality was 4% and morbidity of 30.7% including reoperations, prolonged ventilator, arrhythmias, heart block and others. Re-operations were performed in 1.7% patients as compared to almost to 50 % in the developed country. Similar trend was observed in literature from other developed countries along with a decline in primary procedures.
Conclusion: GUCH in our practice is for primary procedure with simple diagnosis. We at present do not have dedicated facilities for GUCH patients. This report highlights the fact that most of these simple lesions should have been treated before reaching adulthood. There is a need to focus on developing GUCH services in the developing countries to decrease the morbidity of untreated congenital heart disease.

Abstract no: 591

Aortopathy in adult patients with repaired Tetralogy of Fallot (rToF): To what an extent should we be worried?

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Background: We investigated the prevalence and progression of aortic dilatation (AD) late after repair of Tetralogy of Fallot (rToF) and sought to determine its predictors.
Materials and methods: Retrospective study of 110 rToF adults with native aorta studied with cardiovascular magnetic resonance (CMR) at baseline and at ≥1 year follow-up. Aortic measurements were performed in diastole at sinus and ascending aorta (AA) level. AD was defined as diameter 2 standard deviations larger than published normal.
Results: Age at baseline CMR was median (interquartile range); 30.9 (22.9 - 39.4) years and at repair was 4.5 (2.1 - 9.1) years. Time interval between CMR scans was 6.3 (5.1 - 7.6) years. 68 (61%) patients had AD, 65 had sinus dilatation (SD) (39(35 - 41) mm, range 31 - 55) and 24 had associated AA dilatation (AAD) (40(37 - 45) mm, range 36 - 51). Predictors of SD were: age at baseline (p=0.009); male gender (0.039); and previous palliation (p=0.0004) which remained an independent predictor (p=0.0004). Predictors of AAD were: age at repair (p=0.01); systemic hypertension (p=0.04); pulmonary atresia variant (PA) (p<0.0001); male gender (p=0.008); and sinus diameter (p<0.0001), with PA (p<0.0001) and male gender (p=0.004) being independent predictors for AAD. On ROC curve analysis, sinus diameter >39mm predicted AAD (AUC0.90, p<0.0001). Mean rate of sinus and ascending aortic diameter progression was respectively 0.05±0.1 and 0.12±0.26mm/year. No risk factor for increasing sinus diameter was identified. Predictors of ascending aortic diameter increase were age at repair (p=0.008) and previous Blalock-Taussig shunt (p=0.01). The latter was the only independent predictor (p=0.01). No patient underwent aortic surgery.
Conclusion: AD is common in patients with rToF but the rate of progression of aortic size is low in this large contemporary adult cohort. This should be taken into account when planning for scans in follow-up and/or prophylactic aortic replacement. Patients with sinus diameter ≥39mm need cross-sectional imaging to assess for ascending aortopathy.

Abstract no: 648

Clinical features of adult congenital heart disease: A Chinese institutional experience

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Background: A substantial number of children with congenital heart disease (CHD) now reach adolescence and adulthood as a result of advances in paediatric cardiology, surgery and other subspecialties over the past few decades. Consequently, there has been fast development in the management of this special group of patients in many countries. However, it remains a new phenomenon in China. We obtained clinical information about the adults with CHD at Beijing Anzhen Hospital, China.
Methods: Consecutive 431 patients aged 18 years or older with the diagnosis of CHD from January 2005 - March 2012 were enrolled. Demographics and disease characteristics including diagnosis, haemodynamics, treatments and outcomes were recorded.
Results: Patients' age was 30.3±11.0 years. Female was in 71%. There were 401 (92%) patients diagnosed with CHD for the first time. The majority had simple left-to-right shunt defects: including atrial septal defect (ASD) in 182 patients (42%); ventricular septal defect (VSD) in 90 (21%); and patent arterial duct (PDA) in 78 (18%). Other defects in the remaining patients included Tetralogy of Fallot (ToF) and total abnormal drainage of pulmonary veins. Cardiac catheterisation was performed in 82 patients with severe pulmonary hypertension. Systolic pulmonary arterial pressure was 79.6±14.7mmHg, and pulmonary vascular resistance 24.4±14.0 wood*m^2. Thirty five patients were diagnosed as Eisenmenger and received pulmonary vasodilator treatment. Surgical repair was performed in 129 patients, and cardiac interventions in 180 patients including VSD, ASD and PDA device closures. There was no in hospital death.
Conclusions: In our centre, the majority of patients was 1st diagnosed with CHD at adulthood, with severe pulmonary arterial hypertension and increased pulmonary vascular resistance. Patients who underwent surgical or cardiac interventional repairs had good early outcomes. A formal organisation to define treatment strategies and long term follow-up in adults with CHD is urgently needed in China.

Abstract no: 709

The value of a specialised psychological service for adults with congenital heart disease

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Background: Adult congenital heart disease (ACHD) patients are at increased risk of mood and anxiety disorders although there are few dedicated psychological programmes for this patient population. A specialised ACHD psychological service has not previously been described, but the information would be valuable to programmes considering expanding their scope to include psychological care.
Methods: A retrospective review of 100 ACHD patient psychology files was performed and the following data were abstracted: socio-demographic and medical variables, presenting psychological concerns, and a course of psychological treatment.
Results: Of 100 patients, the mean age was 33+11 years, 51% were female, and the majority were single (n=63), educated beyond high school (n=70), and had defects of moderate or great complexity (n=90). The most common psychological concerns were general anxiety (n=82), health/heart-related anxiety (n=71), depressed mood (n=60), and/or coping with a medical condition (n=49); 65 patients met diagnostic criteria for a psychiatric disorder. Following assessment, individual psychotherapy with the ACHD psychological service was recommended to 87 patients of whom 75 agreed to proceed with treatment. The median number of psychotherapy sessions was 8. Therapy most commonly included cognitive restructuring (94%), supportive listening (90%), and/or training in skills for relaxation (56%) or communication (46%). Of 36 patients who have thus far completed the recommended course of psychotherapy, 35 had reduced or absent psychological concerns at the end of treatment.
Conclusions: The majority of patients referred to a specialised ACHD psychology service have significant mood and/or anxiety problems; >70% present with health/heart-focused anxiety. Therefore, ACHD programmes are encouraged to integrate specialised ACHD psychological services in order to most effectively address patient needs. Reduced psychological distress can be achieved among ACHD patients who receive appropriate psychological intervention.

Abstract no: 720

Body mass index (BMI): Prevalence and predictors of obesity in urban and rural communities in South Eastern Nigeria

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Background/hypothesis: Obesity is assuming an epidemic dimension globally. Currently >1 billion adults are overweight and at least 300 million of them are clinically obese. There is no previous study on the prevalence and predictors of in Abia State, South Eastern Nigeria, necessitating this study.
Methods: The study was a cross-sectional aimed at ascertaining the prevalence and predictors of obesity in the state. Participations in the study were recruited from the 3 senatorial zones in the state. In each of the zones an urban and rural communities were randomly selected. Screening for obesity was carried out in these patients using the body mass index (BMI).
Results: Fifty three (2.1%) of the participants were underweight while 1456 (57.4% had normal weight. Furthermore 706 (28.2%) of the participants were overweight, while 313 (12.3%) were obese. 217 (8.6%) of the participants fell into class 1 obesity, 66 (2.6%) into class 2 obesity, while 30 (1.2%) had class 3 obesity. 180 (57.5%) of the obese patients were urban dwellers, while 133 (42.4%) were rural dwellers. These male-to-female and urban-to-rural differences in prevalence of obesity were statistically significant. In a multiple logistic regression analysis in which variables that were significant in the univariate analysis were entered into a model, two variables appear to independently predict obesity in our population. These were gender (OR=2.83, 95%CI 2.150-3.717), p=<0.001, and income (OR=1.51, 95%CI, 1.154-1.979, p=0.003).
Conclusion: The prevalence of obesity and overweight in the state is significantly high, and there is need for interventions to halt this trend.

Abstract no: 722

Prevalence and determinants of hypertension in Nigeria: Results of the Abia State non-communicable diseases and cardiovascular survey of risk factors

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Background/hypothesis: Hypertension is the commonest non-communicable disease in sub-Saharan Africa. In Abia state, Nigeria, no previous study has been carried out on the prevalence and correlates of hypertension among the populace. The purpose of this study is therefore to determine the prevalence and determinants of high blood pressure in Abia State, South Eastern Nigeria.

Methods: The study was a community based cross-sectional house-to-house survey conducted in rural and urban communities in the state. Participants in the study were men and women aged 15 years and above and were recruited from the three senatorial zones in the state.
Results: A total 2 983 consented to be interviewed. There were 1 430 men (47.9%); the mean age of the population was 41.7±18.5 years (range 18 - 96 years). Women had significantly >BMI than men. Similarly waist circumference was also >in women but waist-to-hip ratio was only significantly higher in women in the urban area. 31% of all the subjects had systolic hypertension (33.5% in men and 30.5% in women). This gender difference was statistically different in the urban area. On the other hand, diastolic hypertension was 22.5% in all the population (23.4% in men and 25.4% in women). Age and indices of obesity were the strongest predictors of blood pressure.
Conclusion: The prevalence of hypertension was high in our study both in rural and urban settings. The major determinants of blood pressure in our subjects included age, gender, indices of obesity and pulse rate.

Abstract no: 796

Partial anomalous pulmonary vein drainage, with atrial septal defect in adulthood: A case series of 4 patients in Indonesia

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Background: Partial anomalous pulmonary venous drainage (PAPVD) is defined as at least one pulmonary vein connected to the right (rather than the left) atrium. The connection can be directly to the right atrium or indirectly via either the (superior or inferior) vena cava. The most frequently lesion associated with PAPVD is an atrial septal defect. PAPVD is extremely common in sinus venosus defects (particularly superior) and is seen in approximately 2% of patients with secundum defects. Late presentation of PAPVD with ASD gives problems such as: atrial tachyarrhyrhmia, right heart failure and pulmonary hypertension.
Methods: To report 4 patients suffering from PAPVD and ASD with a mean age of approximately 42.5 years. Three of these patients were female, 1 patient was male. All patients admitted with severe pulmonary hypertension. Pre-operative diagnoses were established by echocardiography and cardiac catheterisation. All the patients underwent repair of PAPVD and PFO by using a pericardial patch.
Result: After the procedure none of the patients showed a residual defect, and with the proper pulmonary hypertension management in ICU, all these patients had good outcomes.
Conclusion: After 1 - 2 months follow-up, none of our patients had significant complications.

Abstract no: 824

Right ventricle in congenital heart defects with severe pulmonary hypertension: Anatomic and haemodynamic considerations

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Background: Right ventricular (RV) adaptation mechanisms to chronic pressure and/or volume overload are hypertrophy, dilatation and decreased function. Compared to other causes, in patients with severe pulmonary arterial hypertension associated with congenital heart defects (PAH-CHD) typical is long well-preserved RV function. The aim of the study was to evaluate specific RV features in these patients and in different subtypes of PAH-CHD.
Patients and methods: Analysed were 56 patients (40F/16M, median age 32 years) with PAH-CHD: 41 (73.2%) had post-tricuspid defects (post-TD); 9 (16.1%) pre-tricuspid defects (pre-TD); and 6 (10.7%) w/o residual shunt (no-shunt) after surgery. Anatomic/haemodynamic parameters were established by echocardiography and invasively; and compared to age-sex matched healthy controls (normal).
Results: (1) Echocardiography showed in PAH-CHD/normal significant RV dilatation (RV diameter (RVD) p<0.0001), RV hypertrophy (RV anterior wall (RVAW) p<0.0001) and lower RV function (fractional area change (FAC) p=0.024). Comparing post-TD/pre-TD/no-shunt showed difference in: median RVD 25/47/35mm (p=0.0001), with RVD >22mm/2 present in 24/100/60% patients (p=0.0001); median RVAW 11/7.5/8.2mm (p=0.009), with RVAW>10mm in 63.2/12.5/16.7% patients (p=0.01); median FAC 60/43/41% (p=0.003) with decreased RV function FAC<40% present in 5.3/37.5/50% patients (p=0.02). (2) Comparing post-TD/pre-TD/no-shunt showed significant difference in following invasive parameters: median mean pulmonary arterial pressure (mPAP) 78/41/49mmHg (p=0.0014); median systolic pulmonary-to-systemic pressure ratio (sPAP/sAoP) 1.06/0.61/0.6 (p=0.0011) with suprasystemic sPAP present in 72.2/0/16.7% patients (p=0.02); and median pulmonary vascular resistance (PVR) 18/7.4/8.2WU (p=0.08).
Conclusions: Compared to normal, in PAH-CHD right ventricle shows compensatory hypertrophy and dilatation. Though patients with various cardiac shunt locations demonstrate different haemodynamics. Post-TD patients are able to tolerate extreme PAP/ PVR, they show most dominant RV hypertrophy but less severe RV dilatation and good RV function. On the contrary, pre-TD/no-shunt present with a less favourable picture. Defect location might play an important role, enabling better RV off-load and thus preserving RV function - which is crucial to a patient's long term outcome.

Abstract no: 893

Neuro-psychological assessments of 40 adolescents after congenital heart defect repair

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Background: Congenital heart defects (CHD) are a risk factor in cognitive development. A growing number of post-CHD adolescents increases incidence of neurodevelopmental abnormalities.
Aim: Neuropsychological assessment of adolescent's post-surgical and/or interventional procedures due to CHD in childhood.

Patients and methods: Included 40 patients aged x-17.6±19.1 years: simple (28), complex CHD (12) who had to be transferred to adult centres because of age. Wechsler Intelligence Scales and clinical trials assessing memory, attention, praxis, abstract thinking and visuo-spatial functions.
Results: Only 8 pts (1 post-dTGA, 3 post-CoAo, 2 post-ASD2, 2 post-VSD repair) scored normal while the others demonstrated cognitive difficulties typical of organic CNS dysfunction/damage. 3 pts (1 post-PDA, 1 post-VSD, 1 post-CoAo repair) showed only visual memory impairment. 32 pts revealed frontal region dysfunctions, while 20 pts additionally showed visual memory impairment (characteristic of right temporal and frontal dysfunction). In 1 pt with complex CHD (post-TOF repair) and in 8 simple CHD pts (2 post-PVS, 2 post-CoAo, 2 post-ASD2, 1 post-VSD, 1 post-AVS), visuo-spatial impairment was noted, typical for temporo-parieto-occipital lesions. In 12 complex CHD pts, the number and severity of cognitive dysfunction were greater: 11/12 pts had executive and learning dysfunctions; 10/12 disorders with three error types (perseverative, confabulation, meaning changes); 8/12 visuo-spatial and visual memory disorders. Two pts had only frontal dysfunction, 1 temporal, while others had both types of dysfunctions. In the post-simple CHD repair group, 71.4% revealed disorders typical for frontal, 10.9% temporal, while 28.5% temporo-parieto-occipital dysfunction. Most of these pts (85.7%) committed only 1 type of error (confabulation).
Conclusions: (1) Of most commonly demonstrated neurodevelopmental abnormalities; GUCH patients show executive function impairment. (2) In post-simple CHD pts, frontal region dysfunctions are less severe than in pts after complex CHD and isolated types of cognitive dysfunctions are more common.

Abstract no: 910

The relationship between birth weights and other anthropometric parameters and aortic intima-media thickness at 1 month in unselected birth cohort

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Background/hypothesis: Evidence suggests that early-life fetal programming can have long term effects on the adult health. A number of small studies report an association between intra-uterine growth restriction and a recently validated and reproducible marker of early athero-sclerosis - aortic intima-media thickness (aIMT) in early life. However there are currently no aIMT data from an unselected population of infants, nor on the relationship between aIMT and body composition or growth parameters other than birth weight.
Materials and methods: Infants are assessed as part of The Barwon Infant Study, an unselected birth cohort of Australian mothers and babies, recruited before 28 weeks gestation (n=1.250). To date 651 babies have been recruited, with >80% participant retention to 12 months. Aortic IMT measurements have been obtained on >95% of those babies attempted at 4 weeks of age. Birth weight, length, head circumference and placental weight are recorded and adiposity is assessed by standardised skin fold thickness measurements in the 1st week of life. Maternal weight and weight change in pregnancy is recorded, together with co-morbidities such as gestational diabetes and pre-eclampsia. At 4 weeks of age aIMT of the posterior wall of the abdominal aorta is measured by trans-abdominal ultrasound using standardised and reproducible methods.
Results: Data on the 1st 500 babies will be available by early 2013: We will present data on the relationship between placental weight, birth weight, head circumference, length, skinfold thickness and the 4-week aIMT.
Conclusions: This will be the largest study of the association between birth weight and other growth parameters determined by the in utero environment and a validated early marker of atherosclerosis.

Abstract no: 917

Reactive hyperaemia index and detection of endothelial dysfunction in high-risk children

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Background: Reactive Hyperaemic Index (RHI) is considered as a new indicator of endothelial function (EF). This plethysmographic method is based on non-invasive assessment of endothelium-dependent changes in vascular tone (PAT) in patient fingertips. Type 1 diabetes mellitus (T1DM) is well-known a risk factor for development of premature cardiovascular disease. As a result of highly efficient treatment protocols of acute lymphoblastic leukaemia (ALL) in children the number of children leukaemia survivors has dramatically risen. Data on the premature manifestation of atherosclerosis (PMA) in ALL children are insufficient in the literature.
Materials and methods: Following approval by the institutional review board 73 eligible participants were enrolled in the study (33 T1DM (16±2.2 years) and 28 ALL patients (14±3.4 years) matched with 12 HC (16±1.7 years). Endo-PAT recorder was used for the determination of RHI as well as specific biochemical markers of endothelial function were assessed (hsCRP, ADMA, E-selectin, VCAM). RHI was evaluated in T1DM and ALL children and further compared with healthy controls (HC). In addition, RHI was correlated with anthropometric and biochemical parameters.
Results: Significantly lower RHI were revealed in T1DM and ALL patients in comparison with HC (1.44±0.41, 1.57±0.50 and 1.99±0.68; $p \leq 0.05$ respectively) implying impaired endothelial-dependent dilation. No association was revealed between RHI and anthropometric parameters, arterial blood pressure or glycated haemoglobin in both T1DM and ALL groups.
Conclusions: Our results support the hypothesis of impaired EF in T1DM children which is in agreement with previously published data in adults. Our study also showed impaired EF in ALL patients. We believe that a non-invasive method such as RHI is a promising future prospect for EF assessment in children with high risk of PMA as well as might aid tailoring their treatment strategies.
This work was supported by research project MSMT CZ- No. 002162081 6 and P36.

Abstract no: 961

Key points of chest pain in children and adolescents

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Objective: Paediatric cardiologists frequently come across chest pain in children and adolescent. This study is performed to characterise the common complaint and to emphasise the significance of the symptom.

Methods: The medical records of children and adolescent under 19-years-old who presented to Seoul National University Bundang Hospital Gyeonggi-do, Korea from 10 May 2003 - 30 June 2012 with a complaint of chest pain were reviewed retrospectively.

Results: Data of 479 patients were reviewed. Male/female ratio was 1.52 (289m, 190f) and median age 9.16 years (range: 2.44 - 18.79 years). Cardiac causes of chest pain were decided in only 4 cases (0.8%). The rest of the 475 patients had: chest pain of unknown origin (n=264); musculoskeletal (n=155); psychiatric problem related (n=28); pulmonary (n=14); or gastro-intestinal (n=12) origin. During follow-up period, only 34 patients (7.1%) had medication or an interventional procedure. Interestingly, 89.4% (n=428) patients were not more anxious about chest pain after reassurance from a doctor, however 49 patients (10.2%) still suffered from sustained chest pain, headache, abdominal pain and so on. There were no deaths during follow-up period and 2 patients were diagnosed with breast mass.

Conclusion: Chest pain in children and adolescents is a very common problem but it rarely has a cardiac cause. However, in about 10% of patients the problem remain unsolved. Although chest pain is not life-threatening condition - unlike in adults - more meticulous recording of patient history and a careful approach will be required.

Abstract no: 1072

Transition readiness among young adults with congenital heart disease

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Background: Given the lifelong nature of congenital heart disease (CHD), adolescents and young adults with CHD are expected to assume increasing responsibility for their health care management independent of their parents/guardians. The Transition Readiness Assessment Questionnaire (TRAQ) is a 29-item questionnaire with 2 domains (self-management, self-advocacy) designed to measure transition-readiness among adolescents and young adults with chronic health conditions. The TRAQ has not previously been evaluated in the CHD population. We investigated whether TRAQ scores differed by age, defect complexity (moderate vs. great), and/or clinic in which patients are followed (paediatric vs. adult).

Methods: Cross-sectional study of 18 - 25 year olds with moderate or complex CHD managed in Edmonton or Toronto, Canada. Subjects completed the TRAQ following CHD clinic visit. TRAQ scores have a range of 1 - 5, with higher scores reflecting greater independence.

Results: Of 128 patients, 80 (63%) had defects of moderate complexity and 48 (37%) had defects of great complexity; 55 (43%) were female. Self-management scores were significantly > among 23 - 25 year olds than 18 - 19 year olds (4.4±0.6 vs. 3.6±0.9, $p<0.0001$). Self-advocacy scores were also > among 23 - 25 year olds compared to 18 - 19 year olds (4.6±0.5 vs. 4.0±0.7, $p<0.0001$). Neither self-management scores nor self-advocacy scores differed significantly between patients with defects of moderate vs. great complexity or between patients followed in paediatric vs. adult CHD clinics.

Conclusions: Among young adults with CHD, transition-readiness does not appear to be influenced by the severity of the CHD lesion or whether patients are followed in a paediatric vs. adult CHD clinic. What is apparent, however, is that self-management and self-advocacy both improve with increasing age. This is consistent with the notion of a "successful" transition process in which young people with CHD gradually assume increasing responsibility for their health care.

Abstract no: 1078

Committed to life: Adolescents and young adults' experiences from living with Fontan circulation

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Background: Single-ventricle defects are among the most complex congenital heart defects and the development of advanced surgical procedures in the last decade has created the 1st generation of adolescents and young adults living with this condition. Yet little is known about how these individuals experience life and what impact the heart defect has on their life in general.

Aim: The aim was to illuminate and gain a deeper understanding of adolescents' and young adults' experiences of living with a surgically palliated univentricular heart.

Method: All adolescents and young adults operated <1995 according to the Fontan or the total cavo-pulmonary connection procedure at a single paediatric cardiology unit were included in the study. They were 17 - 32 years of age (mean age 22 years). Seven open ended in-depths interviews were conducted, transcribed and analysed according to the phenomenological hermeneutical method.
Results: The interpretation of the interview texts showed what the participants experienced living with a surgically palliated univentricular heart in terms of feeling exceptional, strong and healthy. This was supported by 2 structural analyses, where 3 themes emerged: happiness over being me, focusing on possibilities and being committed to life.
Conclusion: Living with a Fontan circulation included negative experiences but the analyses clearly demonstrated a feeling of being strong and healthy. An appreciation of having survived and being committed to life was found to be part in the development of the interviewees' existential growth. This probably strengthens them further in their ability to balance expectations and hurdles in life.

Abstract no: 1110
Technology and teenagers: Developing a mobile phone app for transitional care

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Background: Children are now surviving into adult life with conditions which were previously fatal in childhood. These young people often require complex interventions from numerous members of the multi-disciplinary team and there is an obligation to ensure that their health care needs are met as they move into adulthood. The National Service Framework for Children, Young People and Maternity services have highlighted transitional care as an integral component for all young people including those with chronic illness and/or disability (DoH 2003, 2004, 2006). Transition is further supported by the inter-collegiate report "Bridging the Gaps: Health Care for Adolescents" (RCPCH, 2003), guidelines from the Royal College of Nursing (2004), "You're Welcome" standards for adolescent-friendly services (DoH, 2005). There is now an evolving evidence base for development in this area (While, 2004) and the unmet transitional care needs of young people are well documented (McDonagh 2000, 2004, 2006a; Shaw 2004 a, b, c, 2005 b). There is an emphasis on increasing the knowledge of their condition in young adolescents, treatment and the personnel concerned with their care. Previously paper records had been given to young people however, the clinic nurses identified that young people did not return to clinic with these.
Method: Discussions were held with young people in transition and those in the adult clinic who had recently transitioned. These identified that a phone app for use by patients as a health passport would be beneficial to the young people during a difficult time.
Results and conclusion: Phase 1 was the development of the app and initial trial. Involving young people at all stages. Phase 2 will happen later in the year as we build on the initial design modify the app and roll out to all. We believe that technology is the way to engage young people in their own care and that this design can be shared with colleagues in other centres and across other specialities.

Abstract no: 1122
Aerobic capacity and exercise tolerance in children after Fontan operation

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Background: Due to standard surgical technique and perioperative care, survival of children with Fontan circulation was significantly improved over last 2 decades. Object of study was to assess aerob capacity and exercise tolerance in children after Fontan procedure.
Patients and methods: Ten children (age: 8 - 14 years, weight: 25 - 57kg) with complex congenital heart disease after Glenn and Fontan operation (postoperative time: 3-6 years) were included. In 1 case fenestration was not closed at the time of the study. Heart failure score was documented using New York University Pediatric Heart Failure Index (PHFI). ECG, echocardiography, MRI and exercise test were performed in all cases. Peak oxygen uptake (VO_{2max}), respiratory exchange ratio (RER), metabolic equivalent (MET) and ejection fraction (EF) were measured.
Results: Heart failure was not detected (PHFI:3-6). There was no severe AV valve regurgitation (>Grade II). VO2max (30.3±14.5), RER (0.95±0.13) and MET (8.65±4.14) were close to normal values in most cases. VO_{2max} was reduced (<25ml/min/kg) in 2 cases, one of them had opened fenestration. EF of systemic ventricle was not significantly reduced in any cases (59.9±8.93%). There was no stenosis at the Glenn anastomosis and in pulmonary branches found.
Conclusions: There is a good correlation between EF of systemic ventricle measured by MRI and VO_{2max}. Presumably due to a short postopetive time or/and young age, VO_{2max} and exercise tolerance were better, than expected. Further studies are required to assess exercise tolerance during long term follow-up after Fontan procedure.

Abstract no: 1135
Psychosocial maturity, quality of life and parental fostering of autonomy among young adults with congenital heart disease

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Background: Congenital heart disease (CHD) may impose psychosocial challenges on youth and their parents, especially during the transition to adult health services. We compared psychosocial maturity, quality of life, perceived health status, and parental fostering of autonomy among young adults with CHD of moderate vs. great complexity, and between those followed in a paediatric vs. adult clinic.

Methods: Cross-sectional study of 18 - 25 year olds with CHD of moderate or great complexity managed in Edmonton or Toronto, Canada. Subjects completed the Satisfaction with Life Scale (SWLS), Short Form 12 health status survey (SF-12), Erikson Psychosocial Stage Inventory (EPSI), and Kenny's Parental Attachment Questionnaire (PAQ; a measure of parental fostering of child autonomy). Student t-tests and Pearson's product-moment correlations were performed.
Results: 164 subjects participated, 106 (65%) with moderate and 58 (35%) with complex CHD. 71 subjects (43%) were female. Mean age was 21.4±2.4 vs. 21.9±2.3 years among those followed in a paediatric vs. adult setting, respectively (p=0.29). Mean SWLS score was 27.2±5.4 among patients with moderate CHD, compared with 25.8±6.7 among pts with complex CHD (p=0.17). SF-12, EPSI, and PAQ scores also did not differ by defect complexity. Patients followed by a paediatric cardiologist had more favourable PAQ scores compared to those followed by an adult cardiologist (55.7±10.9 vs. 50.9±12.3, p=0.02). Within the total sample, SF-12 Mental Component Summary scores correlated moderately with SWLS scores (r= 0.52, p<0.0001).
Conclusions: Somewhat unexpectedly, young adults followed in the paediatric setting reported higher parental fostering of their autonomy than those followed in the adult setting. Survey scores did not significantly differ between patients with defects of moderate vs. great complexity. These results suggest that defect complexity should not be a focus when considering the transition needs of young adults with CHD.

Abstract no: 1136

Preparing youth for transition from paediatric-to-adult congenital heart programmes: An intervention protocol from Edmonton, Canada

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Background: Many adolescents living with CHD have limited knowledge of their cardiac lesion or prior interventions, impairing their potential to transition successfully from child-centred to adult-oriented care. Although transition programmes have been introduced by some centres, there are no outcome data with respect to CHD transition.
Methods: A 1 hour nurse-led one-on-one clinic-based intervention preparing 15 - 17 year olds with moderate or complex CHD was developed in Edmonton, Canada and tested on 24 subjects. The aim of the intervention was to improve the knowledge level of the adolescent regarding their CHD. The intervention protocol included (1) review of a diagram depicting the subject's cardiac anatomy and prior surgical or catheter interventions; (2) review of potential late cardiac complications; (3) name, dose and rationale for the subject's cardiac medications; and (4) creation of a portable health summary (MyHealth passport). The intervention was conducted the same day as the teen's cardiology appointment in most cases. Email or texting (teen choice) was employed by the nurse as a follow-up in the week after the intervention. Intervention effectiveness was evaluated through a CHD knowledge questionnaire (MyHeart score) administered pre intervention and 1-month post intervention.
Results: The intervention lasted 67.8±17.9 minutes. The nurse was able to complete the intervention with every subject. The nurse reached 17 of the 24 teens for a follow-up contact; 13 by texting, 4 by email and 0 by phone. Mean MyHeart score improved from 70.5±16.5% correct responses pre-intervention to 79.3±13.3% correct responses 1-month post intervention (p=0.003).
Conclusion: This transition intervention was feasible in the outpatient setting. A modest improvement in CHD knowledge was demonstrated 1 month post intervention. Further study is required to demonstrate the impact of transition interventions on self-management skills among adolescents with CHD. A 2-stage, multi-centre intervention is being planned by our group.

Abstract no: 1186

Self-reported physical functioning may be misleading in predicting actual exercise capacity in adults with congenital heart disease

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Objectives: To compare self-reported physical functioning with the results of cardiopulmonary exercise test (CPX) in adults with congenital heart disease (ACHD).
Patients and methods: From November 2010 - October 2011, 76 ACHD (≥18 years) received a questionnaire survey regarding quality of life (QoL) and self-estimated exercise performance, as well as a symptom-limited CPX at the same day. The QoL was investigated using the Taiwanese version of the QoL questionnaire designed by the World Health Organisation, and the sub-scores of physical domain were also extracted. Self-estimated exercise performance was defined as the subjective prediction of exercise capacity compared to the general population and expressed as a percentage. Maximal oxygen consumption (O2max) in CPX was expressed as a percentage of predicted O2max for age and sex to represent the actual exercise capacity. Overestimation of exercise capacity was defined as the % of predicted O2max <self-estimated exercise performance.
Results: After excluding 10 patients (6 had premature termination of CPX and 4 had missing questionnaire data), a total of 66 patients (age: 31.8±11.7 years; 67% women) were studied. 70% of patients were classified as having moderate to severe CHD. Overall, ACHD had significantly decreased exercise capacity (O2max: 65.4±13.2% of the predicted value), which differed among different CHD severities (p=0.043). Although self-estimated exercise performance correlated with actual exercise capacity (r=0.345, p=0.005), overestimation of actual exercise capacity is common (59%), regardless of age, sex, and disease severity. In our ACHD cohort, physical QoL score was not related to actual exercise test results. Furthermore, a higher physical QoL score may paradoxically increase the probability of overestimation of the actual exercise capacity in multi-variate logistic regression (odds ratio: 1.34, p=0.039).
Conclusions: Overestimation of actual exercise capacity is common in ACHD. A higher self-reported physical functioning did not necessarily predict a better exercise test results, and may be even more frequently observed in patients who overestimated their own actual exercise capacity.

Abstract no: 1189

Substance use, dental hygiene and physical activity in adult patients with single ventricle physiology

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Background: The term single ventricle physiology (SVP) refers to a heterogeneous group of complex congenital heart defects, which are characterised by only one functional ventricle supporting both systemic and pulmonary circulation. The condition is associated with high morbidity and mortality. Due to improved surgical and medical treatment in recent decades, more patients are now surviving into adulthood. However, patients remain particularly vulnerable to additional health problems caused by detrimental health behaviour and lifestyle choices. Areas of concern include alcohol consumption, smoking, substance use, dental hygiene and physical activity
Materials and methods: A cross-sectional, case-control survey was conducted, including 59 adolescent and adult SVP patients and 172 matched healthy controls. Health behaviours were measured using 2 instruments: the Health-Behaviour Scale-CHD and the Godin Leisure-Time Exercise Questionnaire.
Results: In patients with SVP, 85% reported alcohol consumption (OR compared to controls =0.91; p=0.575); 26% admitted binge drinking (OR=0.56; p=0.041); 20% were cigarette smokers (OR=0.59; p=0.100); 12% had used cannabis over the past year (OR=0.80; p=0.596); 20% had had no dental visits during the last year (OR=1.07; p=0.684); 46% did not floss their teeth (OR=1.32; p=0.239); and a total of 39% were not physically active (OR=1.63; p=0.069).

TABLE 1: Comparison of health behaviours in patients with single ventricle physiology and healthy controls

Variables		Number of patients 59	Numbers of controls 172	Odds ratio (95% confidence interval)	p-value
Alcohol	Yes	49 (85%)	158 (92%)	0.91 (0.66 - 1.26)	0.575
	Binge drinking ≥ once a month	15 (26%)	71 (41%)	0.56 (0.32 - 0.98)	0.041
Smoking	Yes	12 (20%)	62 (36%)	0.59 (0.32 - 1.11)	0.100
	>10 cigarettes daily	4 (7%)	21 (12%)	1.00 (0.28 - 3.61)	1.000
Drug use#	Cannabis	7 (12%)	25 (15%)	0.80 (0.34 - 1.86)	0.596
	Ecstasy	0 (0%)	0 (0%)	-	
	Cocaine	1 (2%)	3 (2%)	0.86 (0.09 - 8.40)	0.896
	Hallucinogenic mushrooms	0 (0%)	0 (0%)	-	
	Speed	0 (0%)	0 (0%)	-	
	Sleeping pills	2 (3%)	4 (2%)	1.36 (0.25 - 7.52)	0.724
	Other drugs	0 (0%)	2 (1%)	-	
Dental Hygiene	No dental visit#	12 (20%)	42 (25%)	1.07 (0.76 - 1.51)	0.684
	Not brushing teeth daily	2 (3%)	2 (1%)	2.21 (0.31 - 15.82)	0.429

Conclusions: In general, health behaviours were better in SVP patients, who reported less smoking and significantly less binge drinking. Patients and controls reported similar behaviour regarding dental hygiene, although patients more frequently neglected to brush and floss their teeth daily. Not surprisingly, more patients than controls were physically inactive in their leisure time (p<0.001). We found no significant difference between the two groups on the total health risk score. As this study shows, there is a potential for interventions encouraging behavioral change in this group of vulnerable patients, especially in regard to physical exercise and the prevention of infective endocarditis, where good dental hygiene is a cornerstone.

Abstract no: 1201

Carotid elastographic elastic-pressure-modulus: A new predictive factor of vascular function in long term cardiovascular health status evaluation of patients born with intra-uterine growth restriction (IUGR)

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Introduction: Several studies have suggested that Intra Uterine Growth Restriction (IUGR) increases the risk of cardiovascular disease and early atherosclerosis. Early detection of vascular modulation is essential for implementing early intervention.
Objective: To assess cardiovascular health status of patients born with IUGR at an earlier stage than adulthood using known and novel non-invasive techniques.
Material and methods: We evaluated 18 adolescents born with IUGR and formerly evaluated in utero in our fetal echocardiographic laboratory. Data were compared to those obtained in 17 controls with normal fetal cardiovascular profile. Cases and controls were assessed at 13.96±0.51 vs. 14.72±1.02 years old.

Work-up included ascending aortic diameters and peak flows, pulse-wave transit time around the aortic arch with simultaneous blood pressure (BP) recording. Aortic pulse-wave velocity (PWV), input and characteristic impedances (Zi, Zc), stiffness index and elastic modulus of the ascending aorta (AA-Ep) were calculated. Common carotid intima media thickness (CC-IMT) and elastographic elastic modulus (CC-eEp) (a novel non-invasive method studying the carotid artery wall response to stroke volume) were calculated.
Results: The 2 groups were contrasted in the following table.

	IUGR (n=18)	Normal (n=17)	p-value
Birth-weight (g)	1438±614	3317±510	0.000
Umbilical art. pulsatility index	1.54±0.36	1.23±0.21	0.007
Weight (Z-score)	-0.18±1.05	0.48±1.14	0.089
Height (Z-score)	-0.52±0.88	0.43±1.23	0.013
BMI (Z-score)	-0.06±1.08	0.22±1.25	0.746
Systolic BP (Z-score)	0.38±0.72	-0.27±0.91	0.023
Diastolic BP (Z-score)	0.08±0.65	-0.26±0.57	0.103
PWV (cm/sec)	372±140	351±107	0.623
Zi (dynesxsec/cm)	176±22	236±34	0.000
Zc (dynesxsec/cm)	137±48	171±45	0.035
Stiffness index	4.3±1.6	4.4±1.4	0.801
AA-Ep (mmHg)	370.3±138	370±126	0.995
CC-IMT (mm)	0.46±0.05	0.46±0.04	0.980
CC-eEp (kPa)	93.88±26.6	68.43±31.5	0.016

IUGR subjects are shorter and smaller than controls, but comparable in BMI. IUGR subjects yielded higher systolic BP. In contrast to a previous report, classical biophysical properties of the aorta were comparable to controls; aortic impedance indices showed a supernormal adaptive response with a preserved stiffness index. From the carotid artery perspective and despite a normal CC-IMT, IUGR subjects had a significantly higher CC-eEp.
Conclusions: IUGR subjects present higher systolic BP and CC-eEp at adolescence, which is a probable precise predictive factor of vascular dysfunction. Our findings in the carotid arteries seem to detect intrinsic anomaly of the homeostasis of its arterial wall.

Abstract no: 1211

Surgical outcomes in adults with congenital heart disease: A single centre experience from the developing world

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Background: Lack of data exists on the spectrum and surgical outcomes of congenital heart surgery (CHS) in adults in the developing world. With the number of adults requiring CHS being projected to rise, we undertook this study to improve our understanding of this patient population.
Materials and methods: Data from patients 18 years and older with congenital heart disease, undergoing surgical procedure from January 1998 - May 2012 at our institution were collected. Categorisation of collected data was done according to the Society of Thoracic Surgeons Congenital Heart Surgery database data collection form. Outcomes data were analysed and compared with available European and developing world data.
Results: A total of 161 procedures were performed in 157 patients. Mean age at surgery was 30.1±11.3 years (47.3% male). Septal defects and right heart lesions constituted 2/3s of the diagnoses. Diagnostic pre-operative cardiac catheterisation was performed in 86.3% of patients. Incidence of pre-operative risk factors was 18.7%, with endocarditis and severe pulmonary hypertension being the most frequent. Operative mortality was 1.2% (n=2). Post-operative complications occurred in 24.2% of patients. Re-operative procedures constituted 26.3% of procedures performed, 1/3 of which were repeat reoperative procedures. Right ventricle-to-pulmonary artery conduit placement constituted 57% of the reoperations.
Conclusions: Although the incidence of complications is high, operative mortality is low in the setting of a tertiary referral centre with dedicated congenital cardiac surgeons. The clinical profile of our patient population was similar to that reported in a multi-centre European series. The spectrum of patients and incidence of reoperations did however differ from recently reported series from developing countries. Our utilisation of cardiac catheterisation as diagnostic modality was excessive.

Abstract no: 1217

Comparing the spectrum of heart disease in 2 cities undergoing socio-economic transition in sub-Saharan Africa

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Background: With rapid westernisation in sub-Saharan Africa, cardiovascular disease is gradually becoming the major cause of morbidity and mortality in this part of the world. Unfortunately, there is still a dearth of data on the pattern of heart disease in sub-Saharan Africa given the region's size and diversity and population. We therefore studied the pattern of heart disease in Abuja Nigeria and compared our findings with similar data derived from the Heart of Soweto Study.
Methods: We prospectively studied 1 515 subjects with confirmed cardiac diseases referred to the Cardiology Clinic of the University of Abuja Teaching Hospital over a 4 year period. We then developed a prospectively designed registry and gathered detailed clinical data.
Results: There were slightly >women (50.7%) than men and mean age of the study cohort was 49.0±13.7 years. Hypertension was the primary diagnosis in around 2/3s of the study cohort; comprising >women than men (OR 1.96 95% CI 1.26-2.65). Hypertension was also the commonest cause of heart failure (HF) accounting for HF in 60.6% of cases. The Abuja cohort were more likely to present with a primary diagnosis of hypertension (adjusted OR 2.10, 95% CI 1.85-2.42), hypertensive heart disease/failure (OR 2.48, 95% CI 2.18-2.83); p<0.001 for both comparisons and representing >2/3s of presentations in Abuja. Alternatively, they were far less likely to present with CAD (OR 0.04, 95% CI 0.02-0.11), DCMO (OR 0.35, 95% CI 0.26-0.46), right HF (0.09, 95% CI 0.05-0.17) and valve disease (predominantly RHD in both sites, OR 0.21, 95% CI 0.16-0.28); p<0.001 for all comparisons.
Conclusions: Hypertension and its associated complications is the commonest cardiovascular disease in the Nigerian population in Abuja, and unlike in Soweto, coronary artery disease is not common.

Abstract no: 1225

Prevalence of overweight and obesity among Turkish children and adolescents with heart disease

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Childhood obesity has increased in the last half of century. The aim of this study was to evaluate the frequency of obesity in the children with congenital or acquired heart disease.
Materials and methods: A total of 1 245 (674 female, 571 male; mean age 8.6±4.2 years, range 5 - 18) patients were evaluated in the outpatient clinic of paediatric cardiology department between March 2011 and July 2012. Patients were grouped into 4 categories. (1) "Clinic control subjects" were healthy patients referred to the outpatient clinics with a diagnosis of no heart disease (e.g., chest pain, palpitations, or functional murmurs); (2) "mild heart disease" that has not been treated with either surgical or catheter intervention; (3) congenital heart disease treated with surgical and/or catheter; and (4) "arhythmias" . Differences in the prevalence of obese (BMI ≥95th percentile), overweight (BMI 85th - 95th percentile.), and underweight (BMI<5th percentile.) children were compared with healthy control subjects.
Results: Of 1 245 patients assessed, 727 had heart disease. Diagnostic subgroups included 509 patients in group 2, 154 in 3, and 64 in 4. The prevelance of overweight, obesity and underweight was not different between the control subjects and the patients with heart disease (8.1%, 13.3% and 5.4%; 7.8%, 11% and 8.8% respectively, p=0.145). It was negative correlation between the follow up time and BMI percentile value in the group 3 (p= 0.045). The prevalence of obese and overweight children was significantly higher in the males in group 3 (p=0.033).
Conclusions: Obesity is common problem in children with heart disease at least as general population. It is an important additional risk factor for long-term cardiovascular morbidity and mortality. So precautions to prevent obesity should be parts of pediatric cardiologist's care of children with heart disease.

Abstract no: 1253

Exercise testing in coarctation of the aorta (CoAo): An update

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Background: Long term follow-up of C0Ao pose issues of optimal evaluation non-invasively. Routine clinical and ambulatory/home BP measurements, Echo may not provide enough answers.
Aim: To assess usefulness of ET in CoAo follow-up (fu).
Patient: Forty pts (7 - 32 years, median 21.06) male 29, female 11 followed-up to 20 years post-CoAo repair (surgical 29, native 11).
Methods: Modified Bruce treadmill protocol was done with VO2 uptake measured where possible. Tests were undertaken to evaluate BP, endurance and VO2max responses to exercise where there was suspicion of hypertension, aortic arch hypoplasia and ?response to medication. Pts had no residual pressure gradients. Seventeen pts had 2/> studies during individual follow-up. Response to stenting and growth of patients during fu were important factors needing repeat studies. Twenty three pts had 1 study -4 pre stents and 19 post stent/surgery.

Results: Nine pts had hypertensive peak BP response, 5 resolving post stenting, 1 unchanged post stenting (with weight gain) 3 waiting and 1 resolving with medication. Endurance time and VO2 max tended to improve post stenting. Seventeen pts had normal pre and peak BP response and 13 pts had borderline pre BP and normal peak BP response not requiring active management. One pt had marked improvement with endurance time post stent but still had hypertensive peak BP response (super athlete).
Conclusion: ET allows peak BP response to be assessed and appropriate interventions/surgery/medication undertaken. ET is a useful tool to evaluate optimal long term management for COA.

Abstract no: 1254

Coarctation of the aorta: 31 year follow-up

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Background: Coarctation of the Aorta (CoAo) though eminently treatable, continue to present with ongoing morbidity/mortality even after good treatment. This study explores potential areas of improvement in follow up (fu) by highlighting detection of hypoplasia(hao)/distortion of aortic arch(ao).
Aim: Analyse the fu of 70 pts with special emphasis on ongoing persisting hypertension (hbp).
Patients and methods: 70 pts (6 - 36 years median 19.96 years) 22 female/48 male fu to 31 years, surgery infancy/early childhood - 30 subclavian flap,12 end-to-end; 27 native ballooned (ba) - 7 infants. Records analysed for initial and subsequent adequacy of treatment. Upper and lower limb BP, periodic echo ambulatory bp/home bp, especially during periods of rapid growth (adolescence), exercise testing, and imaging of ao by CT/MRI with hbp and suspicion of hao/arch distortion. No pressure gradient at catheter does not mean good outcome in presence of hbp. Where hao (indexed) was present, stenting of ao undertaken to normalised (indexed) ao produce BP improvement. Stented pts. fu 1 - 4 years.
Results: Ba - 3 infants/toddlers had failed ba with resultant surgery; 2 still had hbp 2 - 4 years post surgery. Many pts. tend to develop hbp at puberty (22); post ba fu 10 pts have remained stable and 12 had stents. Post surgery 42 pts: stents (22), remaining normotensive (8), awaiting stents (5). Post stenting there is improvement in exercise endurance and BP control. One pt needed surgery, with adequate BP response. Stented pts. tend to maintain BP response thus far.
Conclusion: Persistent hbp warrants imaging ao. When grown, many coa pts. develop hbp with proven hao(indexed)and/or distortion and are improved with stenting and or surgery. Further long term fu necessary.

Abstract no: 1288

Late pulmonary valve replacement in patients with Tetralogy of Fallot repaired in infancy and childhood

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Background: Total repair of Tetralogy of Fallot (rToF) has a good long-term prognosis, but frequent chronic pulmonary regurgitation (PR) established after the correction can lead to exercise intolerance, Right Ventricular (RV) dilatation and failure, deleterious arrhythmias and even sudden death. Pulmonary valve replacement (PVR) may improve and/or avoid these situations.
Material and methods: As the ideal substitute for PVR remains a source of dispute, from January 2004 in our unit we are implanting a bovine pericardium biological prosthesis for PVR. This study was retrospective at its inception and prospective during the latest years. 82 patients were operated upon from January 2004 - June 2012. Median age was 22.15 years. The average interval between TOF repair and PVR was 17.2 years.
Results: Two patients died in the early post-operative period. Mean follow-up was 3.5 years (0.2 - 8.5 years). There was 1 late death after heart transplant for persisting biventricular failure. 95% of surviving patients are in NYHA class I. The post-operative (>1 year post-op.) assessment of RV volumes and function using Magnetic Resonance Imaging (MRI) improved significantly compared to MRI pre-operative data. Mean post-operative peak transprosthetic Echo-Doppler gradient at follow-up is 19.38±14.47mmHg.
Conclusions: PVR can be performed with low operative mortality. The bovine pericardium biological prosthesis presents good short and mid-term results, although a longer follow-up is necessary to confirm the initial results regarding its haemodynamics and durability. As it happens with the ideal substitute for PVR, the optimal timing of PVR remains controversial.

Abstract no: 1323

Adult congenital heart disease supervision in regional New Zealand

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Background: Patients born with congenital heart disease now commonly live into adult life (ACHD). Over the last 10 years the care model in New Zealand for ACHD has changed from follow-up centrally in Auckland to regional follow-up in conjunction with Starship ACHD service. Tauranga Hospital (TH) drains a population of just over 210 000 and is approximately a 3 hour drive from Auckland.
Aim: Our research describes the cohort of 103 patients with ACHD under active follow-up at TH at the end of August 2012 for the purposes of audit as well as service planning.
Patients and methods: Median age is 27 year (range 15 - 79 years), female sex 50.5%. The primary congenital cardiac lesion was classified as severe 46%, simple shunt 23%, other 31% in accordance with the schema used by Mackie, et al. In addition, 3 patients have Eisenmenger physiology and 1 patient has uncorrected cyanotic congenital heart disease.
Results: In the 2 years until August 2012, 196 clinic appointments were attended, 140 echocardiograms and 27 cardiac MRI performed. Whilst under local follow-up 14 patients have undergone cardiac surgery (including a single cardiac transplant) and there have been 16 pregnancies resulting in 13 live births. Five patients have died (age range 23 - 55 years) and are not included in the main analysis. Two of the 5 patients suffered from Eisenmenger syndrome.
Conclusions: A significant proportion of ACHD patients under local follow-up at Tauranga Hospital have severe or complex congenital heart lesions. ACHD patients although by number represent a small proportion of patients seen in the outpatient setting, utilise a significant amount of health resource, particularly during pregnancy. Furthermore given most ACHD patients require lifelong follow-up, clinical leaders should account for this group when projecting for growth in service demands.

Abstract no: 1326

TEVAR approach for adult patent ductus arteriosus with aneurysmal change

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Objectives: Patent ductus arteriosus (PDA) is relatively rare in adults, but if present, is often accompanied by calcification of vessels and heart disease. PDA closure in the elderly is a high-risk procedure due to fragility of the aorta and aneurysmal changes in the ductus. In older patients, simple ligation of PDAs very difficult because such cases require cardiopulmonary bypass (CPB). Endovascular treatment is a less-invasive procedure. We report 3 cases treated by stent-graft technique.
Methods: From September 2009 - September 2011, 3 patients underwent TEVAR for PDA with aneurysmal change. We kept the length of the proximal landing zone (PLZ) over 2cm and selected a straight line of PLZ. The number of neck vessels debranched were according to this strategy. **Case 1:** A 77-year-old women was admitted with recurrence of a PDA and lung congestion. She had a history of aortic valve replacement and 2 previous PDA closures via pulmonary artery with open operation and by coil embolisation with intervention. She required axillo-axillo artery bypass to keep enough PLZ before TEVAR. TEVAR was performed via common iliac artery from Zone 2 to T. **Case 2:** A 70-year-old women was admitted with heart failure and supraventricular tachycardia. She required 3 neck vessel debranching from ascending aorta under beating heart to keep enough PLZ. TEVAR was performed via ascending aorta simultaneously from Zone 0 to T. **Case 3:** A 55-year-old women was admitted with heart failure, pulmonary hypertension, and severe aortic valve stenosis. She required a bypass form left common carotid artery to left subclavian artery to keep enough PLZ. TEVAR was performed via common iliac artery from Zone 2 to T.
Results: Median follow up is 5 months (4 - 12). Thirty day mortality was zero. No cardiac or aortic events occured. Type 1 endoleakage was zero. Late mortality was identified in 1 patient who had an intracerebral hemorrhage 12 months after TEVAR.
Conclusion: We describe 3 cases in which this endovascular technique was successfully performed for closure of PDAs with aneurismal change in high-risk patients. TEVAR for PDA may increase the armamentarium for treating this pathology in adults.

Abstract no: 1359

The prevalence of hypertension and its associated risk factors in school-going children in the Phoenix Area, Durban

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Background: The aim of this study was to investigate the prevalence of hypertension and its associated cardiovascular risk factors in school-going children in a low income community.
Hypothesis: Risk factors including risky lifestyle behavior patterns have a significant influence on blood pressure and body morphometrics.
Materials and methods: The study was carried out on learners, stratified across 6 age groups. In the first stage, 1 200 learners received a risk factor questionnaire wherein demographic and behavioural information was recorded. Proportionate sampling from this population was used to select 419 candidates for physiologic, biochemical and anthropometric measurements.
Results: Indian, Black and mixed-race learners comprised 51.6%, 46.5% and 1.2% of the sample population respectively. Males comprised 49.4% of the sample population and females 50.6%. The average systolic blood pressure ranged from 81 - 180mmHg and the average diastolic pressure ranged from 42 - 121mmHg.

The prevalence of hypertension (systolic and diastolic pressures > 95th percentile for the age class) was 7.63% for the total population. The systolic and diastolic pressures were significantly ($p<0.05$) related to age and body mass index but not to gender or race. The average diastolic pressure was significantly correlated to both heart rate and triglyclerides. The mean waist circumference was greater than the 90th percentile for the age class in 27.1% of the total population and this was significantly correlated to BMI ($p<0.05$). Only 52.1% of parents were concerned about their child's weight while 71% of parents regarded their child's weight to be normal. The prevalence of the metabolic syndrome, estimated from the biochemistry and waist measurements was 3.8%.
Conclusion: The prevalence of hypertension was 7.63% in a sample of school going children in this community was closely related to BMI and metabolic syndrome parameters. Lifestyle patterns now account for the emergence of essential hypertension in children.

Abstract no: 1366

The late outcomes of surgical treatment of Scimitar Syndrome: No simple solution to a complex form of anomalous pulmonary venous drainage

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Background: Significant improvements in exercise capacity has been reported in Eisenmenger Syndrome with advanced pulmonary hypertension (PAH) therapies. There is limited evidence of safety and efficacy in complex congenital heart disease patients who do not have Eisenmenger physiology with shunt reversal.
Aim: To report the long term efficacy and safety data with use of advanced PAH therapies in patients with PAH associated with complex CHD.
Methods: Prospective, single centre study of 16 patients (11 F) with mean age 31.3 years (range 15 - 52 years), Fontan circulation (2), pulmonary atresia with VSD/MAPCAs (11), Truncus arteriosus (1), single ventricle with VSD/TGA (1), and TAPVD/ASD (1). 13 patients prescribed mono PAH therapy and 3 combination, from 2004 - 2012. Outcome measurements of oxygen saturation (SaO2), NYHA-FC, 6 minute walk test distance (6MWD) and adverse events were analysed.
Results: Mean duration of therapy was 33.7 months. Significant improvement NYHA-FC (baseline mean 3.19 - 2.4) and 6MWD (pre therapy 343.7m - 420.3m). There was no significant change in SaO2 or echocardiographic parameters. Two deaths due to failure of Fontan circulation, 2 switched to sildenafil because of worsening heart failure.
Conclusion: This single centre study suggests a significant improvement in functional class and exercise capacity after treatment of pulmonary hypertension in complex CHD with advanced PAH therapies.

Abstract no: 1367

Advanced therapies to treat pulmonary hypertension in complex cyanotic congenital heart disease without Eisenmenger physiology are an effective and safe long term treatment option

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Introduction: Significant improvements in exercise capacity has been reported in Eisenmenger Syndrome with advanced pulmonary hypertension (PAH) therapies. There is limited evidence of safety and efficacy in complex congenital heart disease patients who do not have Eisenmenger physiology with shunt reversal.
Aim: To report the long term efficacy and safety data with use of advanced PAH therapies in patients with PAH associated with complex CHD.
Methods: Prospective, single centre study of 16 patients (11 F) with mean age 31.3 years (range 15 - 52 years), Fontan circulation (2), pulmonary atresia with VSD/MAPCAs (11), Truncus arteriosus (1), single ventricle with VSD/TGA (1), and TAPVD/ASD (1). Thirteen patients prescribed mono PAH therapy and 3 combination, between 2004 and 2012. Outcome measurements of oxygen saturation (SaO2), NYHA-FC, 6 minute walk test distance (6MWD) and adverse events were analysed.
Results: Mean duration of therapy was 33.7 months. Significant improvement NYHA-FC (baseline mean 3.19 - 2.4) and 6MWD (pre therapy 343.7 m - 420.3 m). There was no significant change in SaO2 or echocardiographic parameters. Two deaths due to failure of Fontan circulation, 2 switched to sildenafil because of worsening heart failure.
Conclusion: This single centre study suggests a significant improvement in functional class and exercise capacity after treatment of pulmonary hypertension in complex CHD with advanced PAH therapies.

Abstract no: 1393

Ruptured sinus of Valsalva aneurysms in adult congenital heart disease: Clinical presentation, echocardiographic diagnosis and histopathological findings in 8 cases

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Background: Sinus of Valsalva aneurysms (SVAs) are rare anomalies, often associated with congenital cardiac malformations and connective tissue disorders. Aneurysm rupture usually triggers clinical deterioration.
Materials and methods: We searched our echocardiography, histopathology and electronic patient databases between 2004 and 2012. Clinical details and procedures were obtained from case notes.
Results: A total of 8 patients (5 females; mean age 35.7 years) were studied. Aneurysm of right coronary sinus was found in 5 and non-coronary sinus in 3 cases. Rupture into the right ventricle (RV) occurred in 5 cases (3 into RV body; 2 into outflow tract) and the right atrium in 2. Five patients had congenital defects:

bicuspid aortic valve (BAV) 2, ventricular septal defect (VSD) 1, resected subaortic stenosis 1. Another patient had Klippel-Feil syndrome. Acute onset of breathlessness or heart failure was the commonest manifestation (5/8). Clinical examination revealed a continuous murmur in 7/8 patients. Two patients were asymptomatic at time of diagnosis and 1 with Klippel-Feil syndrome died suddenly. Echocardiography demonstrated a continuous shunt across the rupture site in 7/8 cases. Seven patients had a successful procedure (6 surgical, 1 device closure) and remain well to date. Histopathological findings in the patients with concomitant BAV were focal disorganisation of the aortic wall elastic layers with increase in smooth muscle cells and medial and subintimal fibrosis. In the Klippel-Feil fatal case, a linear rupture was found in the non-coronary SVA with thinning of the aneurysmal wall and medial elastin replaced by fibromyxoid connective tissue.

Conclusions: SVAs arise mainly from congenital defects of the aortic media. Sudden onset of dyspnoea with continuous murmur suggests aneurysm rupture. Rarely can it result in sudden death. Commonly co-existing cardiac lesions are BAV and VSD. Echocardiography is the key to diagnosis. Timely surgical or device closure has a good long-term outcome.

Abstract no: 1431

Long term outcomes of myocardial revascularisation in adult patients with congenital heart disease: A single-centre experience

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Objective: coronary artery disease in adults with congenital heart disease (ACHD) is rare, and little is known of its impact on this population. We present the long-term outcomes of patients with a wide range of congenital cardiac diagnoses undergoing coronary artery bypass grafting (CABG) at a single institution.
Methods: Nineteen ACHD patients undergoing CABG between January 2001 and December 2010 were retrospectively reviewed.
Results: Median age was 58 years (range 30 - 82). Sixteen patients (84%) had severe atherosclerotic disease and one had congenital coronary anomaly. Eleven patients presented in CCS class 1 - 3, 7 had demonstrated reversible ischaemia. One, 2 and 3 vessels disease was diagnosed in 4, 7 and 5 patients, respectively. Risk factors included hypercholesterolemia in 9 patients (47%), hypertension in 6 (33%), smoking in 11 (58%) and diabetes in 1 patient (5%). Congenital diagnoses included ASD, AVSD, PAPVC, Tetralogy of Fallot, Ebstein's Anomaly and supra aortic stenosis. Eighteen patients underwent CABG with concomitant congenital repairs. The left internal thoracic artery was used in 9 patients (47%) and long saphenous veins in 16 (84%). Two patients died within 30 days (11%): one due to prosthetic valve endocarditis and one due to multiorgan failure. Complications included reopening for bleeding in 1 patient, atrial fibrillation in 6 (33%), permanent pace-maker in 1, renal impairment in 3 (16%). Mean length of stay was 10 days. Survival was 87%, 81% and 70% at 1, 5 and 9 years respectively. At follow up, 12 patients were in NYHA class 1 and 2 were in class 2.
Conclusions: In the ACHD population CABG is typically performed in concomitance with other congenital procedures. Morbidity and mortality do not seem to differ from those observed in the non-congenital population. Long term survival in this group is very good.

Abstract no: 1440

Pulmonary conduit function after the Ross operation: Longitudinal analysis of the German-Dutch Ross Registry experience

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Background: The Ross operation presents with excellent mid-term results. However the prevalence and predictors of late pulmonary conduit failure after the Ross procedure have been addressed only in few reports with small patient numbers. The objective of this study was to determine the natural dynamics of pulmonary conduit stenosis and regurgitation.
Materials and methods: Among 1 775 consecutive patients (mean age 43.7±12.0, range 16.1 - 70.5 years) that underwent the Ross procedure, 1 645 (93%) received an allograft (pulmonary n=1612, aortic n=12, unknown=21),120 (6%) received a bioprosthesis, and 5 (0.3%) received a bovine jugular vein for the reconstruction of the right ventricular outflow tract. Using non-linear longitudinal models, serial echocardiographic records (n=6950) from these patients were studied with a maximum echocardiographic follow-up of 22.4 years (mean±SD, 5.5±4.3 years).
Results: A slight increase in pulmonary conduit regurgitation grade was observed during follow-up. Freedom from regurgitation grade ≥2+ was 95% after 14 years of follow-up. Female patient gender, the use of an allograft (compared to bioprosthesis), male donor gender, antibiotic treatment of the allograft, and specific surgical adjustments were associated with a significantly higher regurgitation grade. Mean conduit gradient increased from ±4.7mmHg at 1 month to ±10mmHg by 14 years after the procedure. Peak gradient increased from ±8.4mmHg at 1 month to ±18.5mmHg by 14 years after the procedure. Smaller conduit diameter, male patient gender, younger patient age, younger donor age, and use of bioprosthesis were associated with significantly higher mean and peak gradient. The changes in echocardiographic measurements were mainly observed in the first 2 years after surgery.
Conclusion: Echocardiographic follow-up of pulmonary conduits shows outstanding conduit durability. Clinically important conduit regurgitation and stenosis is rare in patients after the Ross operation. Consideration of risk associated predictors may improve pulmonary conduit outcome.

Abstract no: 1454

A very rare case of an unoperated adult with tricuspid atresia and successful pregnancy

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Background: A 46-year-old female with Tricuspid Atresia, VSD and d-Transposition of Great Arteries was diagnosed during infancy and her family deferred surgical palliation due to lack of symptoms (1966). The patient transferred her care to us in 2009 for routine yearly follow-up. Her review of symptoms included cyanosis, occasional palpitations, and dyspnoea on exertion (NYHA Class II) but no chest pain, dizziness or easy fatigability. She works at an office. Current medications include Digoxin 0.25mg once daily. Her obstetric history included G1P1L1A0. She had uneventful pregnancy and delivered a healthy baby girl at 36 weeks of gestation.

Material and methods: The imaging studies including a transthoracic echocardiogram and MRI studies revealed tricuspid atresia type IIb, d-transposition of great arteries and bilateral branch pulmonary artery stenosis, adequate size ventricular and atrial septal defects. The EKG showed sinus rhythm and normal QRS axis; which is usually seen in about 50% of cases with type II tricuspid atresia.

Results: The reasons for her long-term survival was very well balanced pulmonary and systemic circulations. This is facilitated mainly by adequate size of ASD which allowed blood flow from right atrium to left atrium and adequate size of VSD with free flow of blood in to aorta. She also had bilateral branch pulmonary artery, controlling amount of pulmonary blood flow.

Conclusion: Long-term survival is very rare in unoperated tricuspid atresia patients. Only 3 patients including our patient survived into 5th decade. Our patient had long term survival without surgery because of her favourable anatomy for this condition and well-balanced circulation. To our knowledge, this is the 1st report of a successful pregnancy in an unrepaired Tricuspid Atresia with Transposition of Great Arteries.

Abstract no: 1471

Association of emergency admission with hospital length of stay in adults with congenital heart disease

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Background: In the field of emergency care, the increasing needs of adults with congenital heart disease (CHD) are going to increase the burden on medical resources. The aim of this study was to investigate the reasons for emergency admissions and its associations with increased hospital length of sta y(LOS).

Methods: We evaluated 2 720 Adults CHD in a single tertiary cardiac centre database. Patients who required emergency admission from September 1994 and December 2011 were reviewed from medical records.

Results: 669 adult CHD patients (24%) required emergency admission, culminating in a total of 1 726 admissions. Mean age was 48.4±18.5 years, with a female predominance (57%). Most were either schooling or working (62%) and were married (64%). Atrial septal defect (43.8%), ventricular septal defect (21.5%) and Tetralogy of Fallot (7.2%) formed the majority of diagnoses, reflecting proportions in our subject. 1/3 of the admissions were for cardiac reasons including heart failure, endocarditis, arrhythmias, haemoptysis and thromboembolism. The remaining 2/3s were for non-cardiac reasons. Median hospital LOS was 14.5±5.6 days. Those who older ($p=0.02$), and neither employed nor schooled ($p=0.021$) had longer LOS. Endocarditis accounted for longer LOS ($p<0.001$). 41% of the admissions that required inter-disciplinary referrals had increased LOS ($p<0.001$), with utilisation of diagnostic evaluation of non-cardiac ($p<0.001$). Increased LOS was not associated with adverse clinical outcome ($p=0.72$).

Conclusions: Adult CHD patients require admissions for both cardiac and non-cardiac reasons. Older age group, endocarditis were associated with increased LOS. Non-cardiac conditions required inter-disciplinary resources and were associated with increased LOS. Understanding their diverse acute needs may be possible improve care and outcome for these patients.

Abstract no: 1484

Obesity in adults with congenital heart disease is predictive of lower functional capacity

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Background: Following repair of congenital heart disease (CHD), adult patients are at risk for reduced exercise capacity. This is likely due to multiple factors; obesity may be one of these factors. The aim of this study was to determine the prevalence of obesity in adults with repaired congenital heart disease and its impact on exercise tolerance.

Methods: One hundred consecutive adult patients with CHD, who underwent routine cardiopulmonary exercise testing were evaluated, BMI was determined by height and weight obtained at the time of testing. Clinical data was obtained by retrospective chart review.

Results: Patients from 10 major diagnostic groups were identified. The median age for the cohort was 31 years (range 18 - 63) and included 43 males and 57 females, 79%, had at least 1 previous surgical repair. Patients were classified using WHO BMI values as overweight (BMI ≥25) and obese (BMI ≥30.0). 55% of males were overweight or obese, 29% overweight, 26% obese. This was less than US national averages of 72.2% overweight or obese. 58% of woman were found to be overweight or obese, evenly divided between both groups. This compares to a national average of 64% overweight and obese, 35.5 % obese. Being overweight or obese with CHD was predictive of decreased functional capacity by cardiopulmonary exercise testing. .

Conclusion: Obesity is common in adults with CHD but not at the levels seen in the general population. Being overweight or obese was associated with decreased functional capacity. Many factors can affect functional capacity of adults with CHD, some which can not be improved such as cardiac status, presence of restrictive lung disease. Weight is a parameter which may be modified by diet and exercise; it should be a goal to reduce the incidence of obesity for this at risk population.

Abstract no: 1491

Microalbuminuria in children with congenital heart disease: Sensitive marker of cyanotic nephropathy

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Backgrounds: Due to advances in cardiovascular surgery in congenital heart disease (CHD), a large number of patients with CHD are now entering adulthood, and are facing many problems, renal dysfunction being one of these. Micro-albuminuria is a sensitive marker of early-stage renal impairment, and micro-albumin-creatinine ratio (μALB/Cre) is reported to be a useful predictor of future cardiovascular events. We investigated whether early stage of renal impairment assessed by μALB/Cre already exists in children with CHD, and examined the determinants inducing μALB/Cre elevation.
Methods and results: We examined urinary concentrations of Cre and microalbumin, serum levels of Cre and cystatin C, arterial oxygen saturation (SaO2) and haemodynamic variables in 150 patients with CHD (90 male/female = 90/60, median age 5.5) who underwent cardiac catheterisation between 2011 and 2012 in our hospital. Among these, patients (32%) showed abnormal levels of μALB/Cre (>30). There was no significant correlation between μALB/Cre and Cre or cystatin C. Multi-variate analysis with age, sex, systemic cardiac output, blood pressure, pulmonary artery pressure, central venous pressure, and SaO2 included independent variables demonstrated that only SaO2 significantly correlated with μALB/Cre ($p=0.019$). In a separate group of cyanotic CHD, there was also a statistically significant correlation between μALB/Cre and SaO2 ($r = 0.70$ $p<0.05$), independent of the levels of Cre and systatin C.
Conclusions: Low SaO2 is an important determinant of renal impairment even in children. μALB/Cre appears to be more useful to detect early phase of cyanotic nephropathy than serum Cre or cystatin C.

Abstract no: 1518

Successful peri-natal outcome in a patient with single ventricle physiology with Eisenmenger Syndrome

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Background: Pregnancy in patients with Eisenmenger's syndrome (ES) is associated with high maternal and fetal mortality as pulmonary hypertension (PAH) is aggravated leading to poor outcomes. Here we report a rare case of successful outcome of pregnancy in a patient with Double Inlet Left Ventricle (DILV) with ES. Although pregnancy is discouraged in such patients, a coordinated multi-disciplinary team effort for a patient presenting in late gestation is imperative.
Material and method: A 26-year-old primipara with short stature and torticollis presented at 32 weeks gestation with an ultrasound showing a single live fetus. She was haemodynamically stable, in NYHA class II and saturating at 85% in room air. A loud 2nd heart sound along with an ejection systolic murmur could be appreciated. Echocardiography diagnosed DILV with a large inlet VSD amounting to a single ventricle, unrestricted pulmonary blood flow and severe PAH. Fetal echocardiography revealed no major cardiac abnormalities. Chest X-ray alluded to established ES and she had a sinus rhythm on ECG. Inputs from the obstetrician, anaesthesiologist and neonatologist were taken. The patient was admitted for monitoring and oxygen inhalation. Heparin infusion was started and diuretics were minimised to prevent volume depletion. Intravenous betamethasone was administered for fetal lung maturity. A Caesarean section was undertaken at 35 weeks gestation along with tubal ligation. Spinal anaesthesia had to be converted to general anaesthesia for adequate analgesia. A healthy, appropriate for gestational age, baby girl was delivered. The post partum period was uneventful. The newborn's echocardiogram was normal. Mother was prescribed oral sildenafil and bosentan in the post partum period along with advice to refrain from excessive physical exertion.
Conclusion: A multi-disciplinary approach, judicious use of medications and anaesthesia and delivery at a centre having expertise in grownup congenital heart diseases is mandatory for favourable outcomes in an otherwise extremely high risk pregnancy in cases of ES.

Abstract no: 1566

Fontan operation in adults: Is it necessary to stage the procedure?

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Background: Data on immediate post-operative results of Fontan operation in adult patients is limited. We are presenting a single institution's experience with single-stage and staged Fontan procedure in adults.
Materials and methods: Case records of adults (>18 years of age) who underwent Fontan operation from February 2004 - July 2012 were reviewed.
Results: Among 28 patients, 15 underwent single-stage Fontan whereas 13 patients underwent completion of staged Fontan. Mean age at surgery was 21.96±1.37 years. The ventricular morphology was left in 16, right in 6 and biventricular in 6 patients. Mean pulmonary artery pressure was 13.14±1.13mmHg and mean ventricular end diastolic pressure was 11.7±1.15mmHg. Extracardiac Fontan was done in 14 and lateral tunnel in 14 patients. All of them had fenestration. Atrioventricular (AV) valve repair was performed in 4 patients. Two patients had pacemaker implantation during the procedure for pre-existing complete heart block. The mean cardiopulmonary bypass time was 185±26.26 minutes and cross clamp time 56±12.34 minutes. Among survivors, the mean duration of mechanical ventilation was 17±4.39 hours and that of inotropic support was 51±9.4 hours. Average ICU stay was 3.25±0.42 days and chest drain duration was 7.16±1.7 days. There was no statistical significance (unpaired t test) between those who had single stage Fontan and staged Fontan completion in terms of survival and above mentioned outcome measures. 4 patients (14%) died in the post-operative period. 2 patients died due to post-operative arrhythmias: 1 due to failing Fontan and 1 to severe anaphylaxis.
Conclusion: Single-stage Fontan operation can be performed safely in the adult age group, if PA pressure is normal. There seems to be no significant difference in the early post-operative results between single-stage and staged Fontan in adults.

Abstract no: 1582

Role of routine use of continuous positive airway pressure therapy (CPAP) in fast tracking after open heart surgery in adults with congenital heart diseases

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Introduction: Prolonged elective ventilation after open-heart surgery in Adults with Congenital Heart Disease (ACHD) patients for respiratory complications, high pulmonary artery pressures, poor respiratory reserve and on high inotropic support is a routine practice. Mandatory elective ventilation also has its risks and complications such as sudden tube displacement, mucus plug blockages, atelectasis, infection and bleeding due to tracheal trauma. Sudden deaths in the ICU mostly occur in patients on ventilator due to one of the complications mentioned above but are under-reported. Fast-tracking emerged as one of the major advances in the recent era but has limitation, requiring extremely vigilant and expert intensive care teams which most cardiac surgical ICUs lack in India. We have evolved a strategy employing routine use of CPAP therapy in all elective for early weaning from mechanical ventilation and instituting CPAP mask ventilation with secured airway preserving normal swallowing, speech and feeding. Exclusion criteria being renal failure, low cardiac output state or state of confusion or non-cooperation.

Material and methods: From January 2010 - April 2012, CPAP mask ventilation was prospectively used in 125 patients who had high respiratory or cardiac risk factors. All patients were counselled in the pre-operative period and were given short trial of CPAP ventilation a day prior to their surgery. Analysis included retrospectively operated age, sex, disease and NYHA matched population of 135 patients operated earlier. Patients' cooperation and absence of chest infection were mandatory requirements.

Results: Incidence of lung atelectasis, retained secretions, poor ABGs were significantly less in the CPAP ventilation group. Cardiac arrhythmia (AF) did not differ in both groups. Only 1 patient required re-intubation in CPAP group except 1, who had cardiac arrest due to acute severe mitral insufficiency after a failed mitral repair. Two patients in non-CPAP group required re-intubation. Total ICU and hospital stay was significantly less in CPAP group, who also complained less postoperative pain due to lesser need for physiotherapy and coughing. More importantly there was significant cost reduction in CPAP group. Detailed data and group analysis will be presented.

Abstract no: 1608

Patient satisfaction at an adult congenital heart disease cardiac tertiary spoke centre

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Background/Hypothesis: Majority of ACHD patients need lifelong follow-up. Patient satisfaction plays key-role in adherence to medical recommendations and compliance. We present results of Service Evaluation Survey in a new ACHD clinic at a Tertiary University Cardiology Centre, which is a Spoke of the Supra-Regional Hub-and-Spoke model of delivery of ACHD service.

Methods: The new clinic is ACHD consultant-led service, where patients are initially seen by the physician followed by the ACHD-Specialist Nurse and detailed advice is given by both. Seventy three consecutive patients were given the survey to complete and return before they left the out-patients. Patients had the choice to remain anonymous. The survey included 25-short questions and a box for suggestions/comments.

Results: Male/female ratio: 1.2. On a scale of 1 - 10, 1 being very poor and 10 being excellent, the average ratings were as follows: The courtesy of the clinic nurse: 9.7, the doctor: 9.8, having more understanding of the heart conditio: 9.6; Feeling more involved in the decisions about care/treatment: 9.6; Rating of doctor in terms of knowledge/friendliness/courtesy: 9.9; Overall rating of visit to the ACHD clinic: 9.7.

	% of Yes
Were you given information on how to get to the hospital?	90%
Were you given information about parking?	77%
Did the doctor listen carefully to what you had to say?	100%
If you saw the ACHD specialist nurse, did she explain about the service?	100%
Did she give you enough time to discuss your condition?	100%

Conclusion: In the new Spoke-ACHD clinic, majority of patients were highly satisfied with the service, largely due to the "joint" nature of the clinic run by the Consultant and the ACHD Specialist Nurse, whereby the patient received very detailed information and had ample time/opportunity to have their queries answered. This model can be further improved by providing more information regarding public transport/parking.

Abstract no: 1631

Frequency of miscarriage, stillbirth and pregnancy termination in women with congenital heart defects in Germany, Hungary and Japan

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Background: The 2011 ESC Guidelines on the management of cardiovascular diseases during pregnancy define maternal predictors for neonatal events (preterm birth, small for gestational age, respiratory distress syndrome, cerebral haemorrhage, fetal/neonatal death) in pregnancies in women with heart disease. It is unknown whether these predictors also lead to an increased number of miscarriages, stillbirths and terminations of pregnancy (TOP), particularly regarding patients with congenital heart defects (CHD). In the general population, miscarriages and stillbirths occur in 15 - 20%. In the participating countries, stillbirth occurs in <0.5% of all pregnancies.
Material and methods: In a multi-centre cross-sectional questionnaire-based study, 634 women with CHD (Germany: 61%, Hungary: 24%, Japan: 15%) were surveyed over a period of 12 months concerning courses of pregnancy. 309/634 women were pregnant at least once (578 pregnancies). Patients were grouped into those with and those without existing maternal predictors for neonatal events. The predictors were NYHA >II or cyanosis, maternal left heart obstruction, smoking during pregnancy, multiple gestation, use of oral anticoagulants during pregnancy, and mechanical valve prosthesis. The outcomes were miscarriage/stillbirth and TOP (miscarriages and stillbirths were grouped together).
Results: In 75 women with predictors, a total of 141 pregnancies occurred (group 1, 24%); In 234 women without predictor, a total of 437 pregnancies occurred (group 2, 76%). There have been no significant differences between the participating countries.

	Group 1 n=141 (%)	Group 2 n=437 (%)	p
Miscarriage/stillbirth	27 (19.1)	70 (16.0)	0.536
TOP	22 (15.6)	24 (5.5)	0.002
Combined	49 (34.8)	94 (21.5)	0.042

Conclusion: Underlying maternal predictors for neonatal complications do not lead to a significantly higher number of miscarriages or stillbirths. However, TOP occurred significantly more frequently in this group. In presence of maternal predictors for neonatal events pregnancies in women with CHD are less likely to be successful.

Abstract no: 1641

Evaluation of contraceptive methods in women with congenital heart defects in Germany, Hungary and Japan

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Background: The 2011 ESC Guidelines on the management of cardiovascular diseases during pregnancy recommend that maternal risk assessment for women with heart diseases is carried out according to the modified World Health Organisation (WHO) risk classification. Particularly women with congenital heart defects who are at a higher pregnancy-related risk for cardiovascular complications or in which pregnancy is contra-indicated an early and effective contraception is necessary.
Material and methods: In a multi-centre cross-sectional questionnaire-based study, 634 women with CHD (Germany: 61%, Hungary: 24%, Japan: 15%) were surveyed over a period of 12 months concerning contraception. Median age was 30 years. According to the modified WHO classification of maternal cardiovascular risk patients were grouped into 3 risk groups (low; medium; high/pregnancy contra-indicated). The contraceptive methods (CM) used by each group were determined. In this study CM with a Pearl index ≤2 (at ideal use) was classified as "safe".

Results: In all 3 risk groups almost 1/3 of the women was using a CM classified as unsafe. In 29% of all cases an unsafe CM was used. There has been no significant difference between the participating countries.

	Low risk group n (%)	Medium risk group n (%)	High risk group n (%)	Total n (%)
CM safe	220 (72.1)	146 (69.5)	47 (70.1)	413 (71.0)
CM unsafe	85 (27.9)	64 (30.5)	20 (29.9)	169 (29.0)
Total	**305**	**210**	**67**	**582**

Missing values = 52

Conclusion: Alarmingly, almost 1/3 of the women with CHD and increased pregnancy-related risk of cardiovascular complications or contra-indication for pregnancy is using a contraceptive method deemed as unsafe. A more efficient education regarding contraception in women with congenital heart defects is necessary.

Abstract no: 1713

The fate of young people with familial hypercholesterolemia II/A treated by HELP

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Background: Familial hypercholesterolemia represent an important group of disease responsible for significant cardiovascular morbidity and mortality. Sustained elevation of serum cholesterol levels lead to early onset of acquired atherosclerotic heart disease. Type II/A familial hyperlipoproteinemias are inherited autosomal disorders with incomplete penetrance. The manifestation of the homozygous form is 1:1 000 000. Serum LDL plasmapheresis technics such as the Heparin-induced Extracorporeal LDL-cholesterol Pheresis (HELP) method are useful for controlling serum cholesterol and LDL levels. The HELP system completely removes LDL-Ch, fibrinogen and Lp(a) from the blood. The treatment is tolerable, safe and effective.
Material and method: Two cases of homozygous form of familial hypercholesterolemia was confirmed in childhood in Hungary.
Case 1: Authors present a recently 31-year-old young man whose valvular aortic stenosis was discovered at age 2. In 1989 he was examined because of xanthomatous skin alterations and familial hypercholesterolemia was diagnosed. The extended examination of lipid metabolism revealed normal level of receptors but decreased in function. His aortic stenosis was operated at Munich in 1994 and was followed by regular plasmapheresis and oral statin treatment. He has been on this combined treatment from 9 years of age and recently he has no sign of cardiovascular disease! **Case 2:** A recently 18-year-old girl was examined 1st at 7 years of age because of granulomatous and xanthomatous tendon lesions. The extended examination of lipid metabolism confirmed the homozygous form of familial hypercholesterolemia. Her treatment was started with cholesterol plasmapheresis using HELP system combined with esetimide and rasorvastatin therapy. She is on this combined treatment without any sign of cardiovascular disease. Authors considered it worthwhile to present the 2 cases because of the young age of the patients, the unusual presentations of the disease and the long term successful therapy by Heparin induced Extracorporeal LDL-cholesterol Pheresis (HELP).

Abstract no: 1721

Long term advanced therapy in Eisenmenger Syndrome: Serial right heart catheterisation and 6 minute walking distance

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Background: Despite similarities in pulmonary vascular changes, the survival is markedly different between patients with idiopathic pulmonary arterial hypertension (IPAH) and Eisenmenger syndrome (ES). Furthermore, while patients with IPAH slowly deteriorate in spite of advanced therapy (AT) with pulmonary vasodilators, few or no data exist regarding long term effect of AT on haemodynamics and 6 minutes walking distance (6 MWD) in ES. The aim of this study was to examine this by serial right heart catheterisation (RHC) and 6 MWD.
Methods: Nineteen adult patients with ES, 15 patients with ventricular septal defect and 4 patients with atrial septal defect were followed for 5 years with RHC before and after 3 months of AT, and then yearly. 6 MWD was performed at baseline and then every 6 months.
Results: None of the patients died or were transplanted during the study period. RHC revealed a continuous, significant improvement in pulmonary vascular resistance (PVR) (before AT 29±12 vs. 5 years 14±5.2 Wood, $p<0.0001$) and pulmonary blood flow (PBF) (before AT 2.6±1 vs. 5 years 4.7±1.8 L/min; $p<0.0001$) over time after initiation of AT. However, the 6 MWD improved over the 1st 2 years, and then showed a more fluctuating pattern.
Conclusions: This study showed that AT is a beneficial but also long-lasting treatment, which improves both haemodynamic as well as 6 MWD in ES. These data furthermore suggest that despite inconsistent performance as measured by 6 MWD, a long lasting improvement in haemodynamics is obtained by AT. The variation in 6 MWD may be due to conditions such as iron deficiency, which is known to influence 6 MWD. Finally, RHC is the gold standard in evaluating the effect of AT, but since ES on AT seems to be stable, yearly RCH is not necessary, but may instead be performed when clinical deterioration is present.

Abstract no: 1729

Life expectancy and causes of death in a cohort of Fontan conversion patients

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Background/hypothesis: The development of late atrial arrhythmias following Fontan palliations is associated with 3 year mortality approaching 40% in a recent multi-centre series. Fontan conversion and arrhythmia surgery (FCAS) is performed with the expectation that life expectancy would be improved, but the durability of this circulation is not known. We sought to determine mid-term survival and causes of death in a large series of patients (pts) following FCAS.
Materials/methods: Current status of 137 consecutive pts who had 138 FCAS at a single institution from 1994 - 2012 was reviewed. Current status was ascertained by phone interview with patients or review of physicians' records. Age at last follow-up, time to heart transplantation (OHT) and causes of death were assessed.
Results: Median age at original Fontan surgery was 5.7 years (range, 1.1 - 34.9 years), and at FCAS was 23.6 years (range, 2.6 - 47.3 years). Ventricular morphology was single left in 100, single right in 13 and complex/indeterminate in 24 pts. Early operative mortality was 1.5%. Of 135 early survivors, 16% (22/135) died (13) or underwent OHT (9). OHT occurred at a mean post-FCAS interval of 31 months; 4/9 died in the peri-transplant period. Of the remaining 13 deaths, 2 died of non-cardiac causes. Circulatory causes of death included: Sudden (3); congestive heart failure (3); sepsis following urologic procedure or defibrillator change (2); renal failure (1); liver failure (1); and hepatocarcinoma (1). At mean follow-up of 8.6±4.2 years, total post FCAS transplant-free survival is 83%. The mean current age of 113 transplant-free survivors is 32.4±6.6 years.
Conclusions: Survival into the 4th - 6th decades of life was achieved in this population of Fontan pts with predominantly single left ventricular morphology, posing challenges for ongoing medical surveillance and management of co-morbidities.

Abstract no: 1731

Congenitally corrected transposition of great vessels in a young pregnant woman

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Background: Transposition of great vessels is a rare form of congenital heart defect and occurs in <1% of patients with congenital heart disease. Normally the development of ventricular situs occurs during the 5th week of gestation, when twisting of the primordial heart tube to right (d-looping) places morphologic right ventricle on right side of heart and morphologic left ventricle on left side of heart. Normal development of the great arteries occurs during the 5th - 6th week of gestation and this is genetically influenced by neural crest cells. Abnormal development [congenitally corrected transposition of the great arteries (ccTGA) or (l-TGA)] results from looping of the primordial heart tube to the left instead of the right.
Aim: To show a rare case of a congenitally corrected transposition of great vessels in a 27-year-old pregnant patient.
History and progress: The patient was 1st seen at our combined obstetrics and cardiology clinic at 33 weeks gestation. On her initial presentation she reported symptoms of grade 2 dyspnoea and intermittent episodes of palpitations. The examination revealed a dextrocardia with a 3/6 pansystolic murmur along her right parasternal area. In addition she had a palpable and loud pulmonary component of the 2nd heart sound. Her echocardiography revealed a dextrocardia, L-type TGA and a small restrictive ventricular septal defect. The patient subsequently had an elective Caesarean section and her peri-operative period was uneventful. She and her baby remained haemodynamically stable. The patient declined any form of invasive stratification or intervention for her heart. She was discharged and then followed-up at the cardiology outpatient department.
Discussion and conclusion: There is limited literature on transposition of great vessels in pregnancy; however it can be associated with disastrous outcomes in pregnancy. The main recommendations in these patients include: (1) consultation with a cardiologist who has an experience with adult congenital heart disease before pregnancy; (2) scheduled cardiology evaluation and follow-up during pregnancy; and (3) a multidisciplinary coordination for labour, delivery and post-partum periods. Long-term effect of pregnancy on right ventricular function is unclear; however there is an increased risk of heart failure and arrhythmias with tricuspid valve regurgitation. Long-term outcomes/problems depend on presence/severity of associated lesions and right ventricular (systemic ventricle) failure.

Abstract no: 1737

Comparative evaluation of normal weight and obese children with essential arterial hypertension

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Aim: To compare normal weight and obese children with essential hypertension.
Materials and methods: 114 patients diagnosed with primary hypertension - 87 boys and 27 girls, aged 10 - 18 years, mean 15.2±1.8 years, were analysed. Patients were divided into 2 groups according to body mass index (BMI): group I with normal BMI (<85 percentile), group II - overweight and obese patients (BMI >85 percentile). Analysed parameters included: systolic and diastolic blood pressure (SBP, DBP), fasting glucose, total cholesterol, HDL cholesterol, triglycerides levels, heart rate at rest (HR). Mean values of the parameters in both groups were compared using Student's t test. The correlations between studied parameters and blood pressure in the selected groups were assessed by Pearson's correlation coefficient (r).

Results: Statistically significantly higher SBP, triglycerides levels, HR were observed in overweight and obese children, and positive correlations between SBP - triglycerides levels (r=0.34), HR-DBP (r=0.4), DBP-BMI (r=0.2), BMI-triglycerides levels (r=0.25) were found. In children with normal BMI, no relationships between the studied parameters were observed.

	Age (years)	SBP(mmHg)	DBP(mmHg)	BMI(kg/m²)	Glucose (mg/dl)	chol(mg/dl)	HDL(mg/dl)	TRIGL(mg/dl)	HR/min
Group I n=43	15.5±1.9	136.6±11.6	75.7±11.1	20.7±1.9	95.4±17.2	161.8±39.8	54.0±13.0	97.0±60.3	77.1±16.0
Group II n=71	15.0±1.8	141.0±10.8	75.7±10.8	27.2±3.0	94.6±10.6	171.5±43.7	49.1±16.1	128.5±70.3	83.8±16.0
Student t-test	p=ns	p=0.04	p=ns	p<0.00001	p=ns	p=ns	p=ns	p=0.01	p=0.04

Conclusions: (1) The overweight and obese children with hypertension are characterised by higher blood pressure values, serum triglyceride levels and heart rate compared to children with hypertension and normal BMI. (2) The moderate apparent correlations between blood pressure values and BMI, serum triglycerides levels and heart rate are observed in children with overweight and obesity.

Abstract no: 1782

Can arrhythmia surgery at the time of pulmonary valve replacement protect from future arrhythmia?

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Background/hypothesis: Longstanding pulmonary regurgitation after surgery for Tetralogy of Fallot (ToF) or pulmonary stenosis leads to progressive RV dilatation and arrhythmia. Pulmonary valve replacement (PVR) is often required in adult life. Arrhythmogenesis is complex, relating both to scars and ongoing haemodynamic problems. Our unit has performed surgical cryo-ablation in selected patients at the time of PVR since 2007.
Materials and methods: Retrospective analysis of case-notes for all patients undergoing PVR by a single surgeon at our institution between 2007 and 2010.
Results: Fifty operations were performed in 47 patients; 42 had underlying ToF. The main indication for PVR was significant pulmonary regurgitation. Thirty three patients with arrhythmia were also referred for surgical cryo-ablation to the RA and RVOT (right atrium and RV outflow tract). Mean age at operation was 27.8 years (range 14 - 61). Twelve patients had atrial fibrillation/flutter pre-op, all had cryo-ablation performed. Three (25%) had a recurrence of AF post-op. In 22 cases, RVOT cryo-ablation was performed: in 11 there had been pre-op ventricular tachycardia. Four patients developed VT post-op after a mean time of 12 months (range 1 - 30); only 2 required long term medication. One already had an Implantable Cardioverter Defibrillator (ICD); 1 required a new device insertion years later. Three further patients had ICDs inserted post-op: this had been planned electively pre-op and was not due to new arrhythmia. 3 patients who underwent cryo-ablation required insertion of a pacemaker post-op for new bradyarrhythmias at a mean time of 5 months (range 0 - 12). Three deaths occurred; 2 in patients post-ablation. None appear related to arrhythmia.
Conclusions: Surgical cryo-ablation at time of PVR appears to reduce arrhythmia recurrence in the short term although long term outcome data is required. Patients selected for this have already demonstrated a clinical arrhythmia burden and although initial results appear promising, whether this provides effective future protection remains to be seen.

Abstract no: 1792

Treatment of primary arterial hypertension in children

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Aim: Evaluation of influence of obesity on the severity of primary arterial hypertension (PHA) and its pharmacological treatment in children. Study group consists of 45 patients (pts) (35 boys and 10 girls) age 9 - 17 years (mean age - 13.8 years) with arterial hypertension diagnosed as primary after specific diagnostic evaluation.
Methods: Systolic and diastolic pressure were evaluated before (SBPpre, DBP pre) and after (SBPpost, DBPpost) at least 3 months of introduction of hypotensive treatment. All patients were treated with beta-blocker and ACE-inhibitor. In case of insufficient control of blood pressure, calcium antagonist or diuretic were added to the treatment. Anthropometric parameters (weight, height and body mass index BMI) as well as serum cholesterol and triglycerides concentration were evaluated at the initiation of the treatment. The values of measured SBP and DBP and effectiveness of pharmacological treatment of PHA were analysed in relation to presence of overweight or obesity.
Results: In all pts satisfactory blood pressure control was achieved. There were no significant differences between blood pressure values before treatment in pts with normal and increased values of BMI. Significant correlation between systolic blood pressure before and after treatment and BMI was noticed (SBPpre vs. BMI: r=0.51; SBPpost vs BMI: r=0.50 ; p=0.01). Effectiveness of the hypotensive treatment was related to the presence of overweight or obesity. In pts with BMI >25kg/m^2 SPBpost and DBPpost were significantly higher than in patients with normal weight (normal weight - 122.6/76.1mHg; overweight - 130.0/82.9mmHg;

obesity 136.3/81.3mmHg). Serum cholesterol concentration was higher in patients with obesity then in patients with overweight or normal BMI (respectively: 4.7; 3.7 and 3.4mmol/l). There were no correlation between BMI and serum triglycerides concentration.

Conclusions: Overweight or obesity has an important influence in the development of primary arterial hypertension in children. Overweight or obesity negatively influenced the effectiveness of pharmacological treatment of primary arterial hypertension in children. Reducing diet should be basic in treatment of primary hypertension in children.

Abstract no: 1816

Services for grown-up congenital heart disease (GUCH): Past, present, and future. A report from a GUCH special unit in Tokyo, Japan

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Background: The number of adults with congenital heart disease (CHD) is drastically increasing in Japan so that this population exceeded the one of children with CHD in 2005, and has reached approximately 500 000 recently. This is a report from one of the oldest GUCH units in the world established in 1975.
Materials and methods: GUCH patients admitted into the special unit were reviewed through medical records in order to know the difference in their medical profiles and services with time, the years of 1991, 2001, and 2011. Future recommendation in the system would be made with consideration of the changes of
Results: (1) The number of admission increased rapidly; 86, 154, and 305 in 1991, 2001 and 2011 respectively. The patients over 40 years old were of 2%, 14%, and 34% in 1991, 2001 and 2011 respectively, which showed that age of patients became older. (2) Surgical admission decreased; 20% of all admissions in 1990s compare to of 6% in 2011. Emergency admissions were 59 (19%), 12 in the 59 required the stay in intensive care unit, and 5 died in 2011. Those emergency admissions consisted of severe heart failure (39%), arrhythmia (17%), haemoptysis (10%), and infection (10%). (3) Consultation to non-cardiac departments were often needed; 123 times in 2011. Various specialty of support in 2011 such as liver-gastro unit (13%), neurology (12%), dental department (11%), and Liaison (8%) were unchanged from 1990s. New problems have occurred such as liver fibrosis in Fontan patients, amiodarone induced thyroid dysfunction, haemodialysis in complex CHD. (4) There were more old families demanding the visiting nurse system at home.
Conclusions: Specialised GUCH units require adequate manpower and highly advanced medical services in a multidisciplinary facility. This requires optimal centralisation of services.