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O1-1
The Brugada Syndrome in children: presentation and outcome of proband patients.

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Background: Brugada syndrome (BrS) is a potentially serious channelopathy that usually presents in adulthood and less frequently in children. We sought to examine the presentation and outcome of Brugada syndrome proband patients in childhood.
Methods and Results: A review from two institutions, Brussels and Barcelona collected retrospectively from 1996 and prospectively from 2006. The criteria for inclusion were documentation of spontaneous or drug induced ⩾2 mm coved (type 1) Brugada ECG pattern at age ⩽16 years and no previous diagnoses of BrS in the family. 15 patients were identified. Presenting age ranged from 2 to 16 years (median age:13 years). 10 patients were older than 12 years. Three patients (aged 12, 13 and 16 years) presented with aborted sudden cardiac death. In these 3, diagnosis was made retrospectively based on presentation and earlier ECG records. 5 patients presented with an aborted sudden death (age range 8 to 16 years). Syncope was the presentation in 6 other patients, aged from 2 to 16 years. 3 of these were previously diagnosed with Sick Sinus Syndrome or atrial fibrillation. 1 asymptomatic 16 year had coincidental Brugada ECG pattern. At presentation 12 patients had a type 1 ECG pattern. Ajmaline testing was performed in the 3 other patients and was positive. 1 patient required CPR following ajmaline infusion. An ICD was implanted in 7 patients. 4 patients received appropriate ICD shocks. 2 patients received inappropriate shocks. During the follow up of the remaining 12 patients, 1 patient with ICD died at the age of 18 years. 1 patient died 3 years after a first aborted sudden cardiac death because of misdiagnosis. One patient was lost from follow up after one year.
Conclusion: In this series of proband children with BrS, onset of symptoms occurs from a very young age. The presenting symptom is often aborted sudden death or syncope. Most patients presented with a coved type ECG in rest. The other patients had positive ajmaline testing. Sinus node dysfunction and atrial conduction disorders are commonly associated and Brugada syndrome needs to be considered as an etiology.

O1-2
Loss of sodium channel function-like phenotypes: Diagnostic and therapeutic aspects in children

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Introduction: Loss of sodium channel function-like phenotypes are a spectrum of diseases including Brugada Syndrome (BrS) and cardiac conduction disease, ranging in presentation from asymptomatic to sudden cardiac death (SCD) in young individuals. This study aimed to analyze the diagnostic and therapeutic aspects of these disorders in children.
Methods: Patients (n = 37) were included if they presented below 18 years of age with (a) pertinent cardiac symptoms plus abnormal ECG (BrS ECG and/or prolonged conduction intervals), (b) abnormal ECG during family-screening for confirmed sodium channelopathy or (c) incidental observation of abnormal ECG with family history of sudden unexplained death. Conduction intervals were considered prolonged if longer than 2 standard deviations for age.
Results: Age at presentation was 6 ± 5 years and 13 (35%) were female. Of the symptomatic patients (n = 18, 49%), 4 presented in the first year of life. Syncope (n = 9) was the most common symptom followed by arrhythmias (n = 8). Symptoms were associated with fever in 6 (33%) of the symptomatic patients, 2 of which occurred during vaccination-related fever episodes. SCD occurred in a 3 month old male following vaccination. Heart-rate on the earliest available ECG was 90 ± 25 bpm, PR interval 163 ± 37 ms, QRS duration 110 ± 17 ms and QTc 411 ± 27 ms. BrS ECG was present in 11 (30%) patients, prolonged PR in 21 (57%) and prolonged QRS in 28 (76%) patients. Of the genetically tested patients (n = 35), loss-of-function SCN5A mutations were present in 30, results were pending in 2 and 3 patients were genotype-negative. Genetic testing was not performed in 2 patients due to parental unwillingness Treatment was instituted in 11 (61%) of the symptomatic patients and

included beta-blocker alone (n = 3), ICD alone (n = 3), beta-blocker and ICD (n = 4) or beta-blocker and pacemaker (n = 1). During follow-up (4 ± 4 years), 2 patients had recurrence of ventricular tachycardia, there were no deaths.
Conclusions: Loss of sodium channel function-like phenotypes present with varying severity in infants and children. Fever and vaccination are the commonest arrhythmia triggers. Prolonged conduction intervals are seen more often than BrS ECG. Management includes careful monitoring during fever and vaccination, beta-blockers in patients with tachycardia-related arrhythmias and ICD for the resistant cases.

O1-3
Tachyarrhythmia following Norwood Operation: A Single Center Experience
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Introduction: Bradyarrhythmias are known risk factors for death following Norwood operation. Tachyarrhythmias are common following repair of congenital heart disease, and may be associated with increased morbidity and mortality. The purpose of this study was to determine the incidence of tachyarrhythmia in patients following Norwood operation.
Methods: Retrospective chart review of all patients who underwent stage I Norwood procedure from 1/1/2002 to 9/1/2011 (n = 100). Data collected included demographics, shunt type (modified Blalock-Taussig shunt (mBTS) or Sano shunt) cardiopulmonary bypass (CPB), cross clamp and circulatory arrest times, type of tachycardia and need for antiarrhythmic medication at discharge.
Results: Data regarding arrhythmia was available for 98 patients. Operative mortality after Norwood was 20% (19/98). Tachyarrhythmia occurred in 33/98 patients (34%). Operative mortality was 9% (3/33). There was no difference in mortality in those with tachycardia compared to those without (p = 0.33). Causes of mortality in those with a tachycardia included cardiac arrest (n = 2) or circulatory collapse requiring ECMO (n = 2), respiratory arrest (n = 1) and dural sinus venous thrombosis (n = 1). There was no significant difference in the incidence of arrhythmia based on shunt type (p = 0.23) where 8/32 underwent palliation with mBTS, and 25/66 with a Sano shunt. There was a significant association between male gender and duration of circulatory arrest for the development of arrhythmia (p = 0.003 and p = 0.02 respectively). Of the 33 patients with arrhythmia after Norwood operation, the distribution was as follows: atrioventricular reentry (AVRT): n = 17; junctional ectopic tachycardia (JET): n = 6; atrial flutter (AFL): n = 2; atrial ectopic tachycardia (AET): n = 4; wide complex tachycardia: n = 1; ventricular tachycardia (VT): n = 2. One patient had AVRT and JET. Thirty patients survived to hospital discharge with 23 receiving anti-arrhythmic therapy. Twenty received monotherapy with: Amiodarone (n = 1), Digoxin (n = 9), Sotalol (n = 9) and Propranolol (n = 1). Three patients received dual therapy with digoxin and flecainide (n = 1) or amiodarone (n = 2).
Conclusion: Post-operative tachyarrhythmia is common, occurring in 34% of our patients after Norwood operation. Shunt type was not associated with tachycardia. Male gender and longer circulatory arrest times are significant risk factors associated with the development of a tachyarrhythmia. Tachycardia was not associated with increased risk of death.

O1-4
Cryoablation with an 8-mm Tip Catheter for Supraventricular Tachycardia Substrates in Children
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Introduction: Cryoablation is utilized increasingly in children due to its safety profile. Recently, larger catheter tips are used more commonly in order to improve the long-term success rate. The aim of this study was to assess the safety and efficacy of 8-mm tip cryoablation of supraventricular tachycardias (SVTs) in children.
Methods: Electrophysiological procedures were performed using the EnSite electroanatomic system (St.Jude Medical Inc, St.Paul, MN) guidance. Procedures performed with an 8-mm tip cryoablation catheter were reviewed.
Results: Between July 2010 and November 2011 a total of 33 patients (mean age of 12.2 ± 3.8 years) underwent catheter ablations using an 8-mm tip cryoablation catheter. In 18/33 (%55) of these patients prior catheter ablation attempts using RF ablation or 6-mm tip cryoablation failed. The acute success rate was 13/18 (72%). In 15 patients, 8-mm tip catheter was the first choice of ablation and acute success rate was 14/15 (93%). The overall acute success rate was 27/33 (82%). Fluoroscopy was not used in 16/35 procedures. The mean fluoroscopy time in rest of the procedures was 4.1 ± 6.6 minutes (range: 0.1 to 25 minutes). The arrhythmia substrates and acute success rates were as follows: AV reentrant tachycardia:19/24 (79%), AV nodal reentrant tachycardia: 6/6 (100%), focal atrial tachycardia: 1/2 (50%), intraatrial reentry tachycardia:1/1 (100%). The overall recurrence rate was 8/27 (30%) at a mean follow-up of 6.7 ± 5.1 months. Two patients underwent successful second procedures. There were no complications.
Conclusions: The use of an 8-mm tip cryoablation catheter for SVTs in children appears to be safe and acutely effective in procedures where conventional ablation choices fail. However, recurrence rate is relatively high. Further studies are needed to assess the long-term efficacy.

O1-5
Cryoablation of Antroseptal Accessory Pathways in Children with Limited Fluoroscopy Exposure
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Introduction: Cryoablation is utilized increasingly in children due to its safety profile especially for the septal accessory pathways (APs). Recent advances in electroanatomic mapping technologies resulted in a decrease and even in elimination of fluoroscopy exposure during these procedures. This study was done to assess the efficacy and safety of cryoablation of anteroseptal APs using limited fluoroscopy guided by an electroanatomical mapping system.
Methods: A total of 15 patients underwent cryoablation for anteroseptal APs between August 2010 and September 2011. Cryomapping is performed with an 6-mm tip catheter before placing ablation lesions. The procedures were performed using EnSite system (St. Jude Medical, St Paul, MN) guidance.
Results: The mean age was 13.3 ± 4.7 years. The acute success was 93% (14/15). The mean procedure and cryoablation times were 175 ± 68 mins and 1524 ± 651 secs, respectively. Flouroscopy was used only in 4 patients and the mean flouroscopy time was 0.7 ± 0.6 mins (range: 0.2-2.1). The recurrence was noted in 1 patient (7%)

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at a mean follow-up of 9.6 ± 4.3 months. Temporary AV block was observed in 1 patient. No other complications noted.
Conclusions: Cryoablation of anteroseptal APs can be performed efficiently and safely in children using limited fluoroscopy exposure with the help of electroanatomical mapping systems.

O1-6
Outcome of Complete Heart Block Diagnosed in Fetal and Postnatal Life
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Introduction: Congenital complete heart block (CCHB) occurs in approximately 1 in 14,000-20,000 live births. Complete heart block can have a diverse aetiology and carries a significant morbidity and mortality when associated with structural heart disease.
Method: Review of all patients presented to our institution with diagnosis of complete heart block over the past 10 years. We investigated aetiology and outcome of complete heart block in fetal life, infancy and childhood.
Results: 49 patients were diagnosed with complete heart block of which 19 had antenatal diagnosis; seven were diagnosed in infancy, and 23 in childhood. 9 patients had an associated structural heart disease. Among fetal cases, maternal anti-Ro or anti-La antibodies were present in nine (45%). Ten (52%) fetuses had received prenatal dexamethasone and/or beta mimetic treatment, of which one recovered but four died. Total mortality rate was 16% (8/48) of which intrauterine death was responsible in 5 (62.5%). The presence of structural heart disease (55%), fetal diagnosis, maternal antibodies (28%), hydrops, and fetal heart rate lower than 55bpm were associated with higher mortality. Fetal diagnosis showed statistically significant mortality compared to infant and childhood diagnosis ($p < 0.0001$, $p = 0.005$ respectively). Eight (40%) fetuses received pacemaker insertion after birth. Freedom from pacemaker insertion was better in infant and childhood group.
Conclusions: Complete heart block diagnosed antenatally has strong association with maternal anti-Ro and anti-La antibodies and has poorer outcome compared to that of diagnosed during infancy or childhood. Beta-mimetics and steroids may offer help in selected cases to alter significantly higher mortality of antenatal AV block.

O1-7
Minimal- Invasive Transcutaneous Epicardial Pacemaker-Lead Lmplantation–A Novel Approach To Avoid Thoracothomy
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Introduction: In congenital heart disease (CHD) cardiovascular abnormalities, small vascular size and surgical procedures may preclude transvenous pacing. Surgical epicardial lead implantation is commonly used in these conditions. The objective of this study is to assess the feasibility of a newly developed minimal-invasive transcutaneous peacemaker-lead implantation technique to avoid thoracotomy and associated morbidity.
Methods: A preliminary pilot study using 5 German pigs was conducted. After femoral puncture RA-, LV- and aortic-root angiographies were performed to localize target sites. Epicardial access was established via a modified subxiphoidal approach: under fluoroscopy guidance a Tuohy-needle (18-22 G) was advanced retrosternally into the pericardial space. Contrast dye proofed correct location. Using coronary guidewires three 4-Fr. short-sheaths were introduced. Deflectable catheters (Medtronic C315) with a modified flat tip were positioned at the atrial and ventricular target site and stabilized by negative suctioning force. Through these 4.1 Fr. transvenous screw-in bipolar leads (Medtronic 3838) were implanted. After achieving acceptable sensing- and pacing-properties, sheaths and excessive air/fluid were removed. Leads were tunneled subcutaneously to a subcutaneously implanted 2-chamber pacemaker (Medtronic). Pacemaker was set to 0D0 to prevent interferences with intrinsic heart rhythm. First follow-up was at day 3-5.

Mean (Range)	Impedance (Ohm)	Pacing-Threshold @1ms (V)	Sensing (mV)
Atrium			
unipolar	340 (240-430)	3,53 (0,5-7,5)	1,31-2,1 (0,25-1,4 to 2-2,8)
bipolar	465 (327-651)	3,17 (2-4)	1,6-2,4 (0,25-0,75 to 2,8-4,0)
Ventricle			
unipolar	313 (232-464)	4,3 (3-6)	3,1-4,4 (2,8-4 to 4-5,6)
bipolar	423 (347- 568)	2,9 (1,25- 4)	1,6-2,5 (1-1,4 to 2,8-4,0)

Results: At implantation mean bodyweight was 28,2 (range 21, 5-31) kg.
Implant procedure was successful in all animals with establishment of a functional DDD-pacemaker system. Neither pericardial hemorrhage nor myocardial perforation occurred. One animal died unexpectedly in the post-procedural night. Remaining animals were euthanized due to pacemaker-pocket infection (without impairment of general-condition) (day 17, day 34), hind-leg fracture (not procedure-related) (day 18) and end-of-study (day 49). Lead-performance at first follow-up see table:
Conclusion: Epicardial 2-chamber-pacemaker can be successfully implanted transcutaneously by this newly developed minimal-invasive pericardial approach. Satisfactory sensing- and stimulation-properties could be obtained during implant and short-term follow-up. Further research and procedural refinement may enable selective-site-pacing in CHD-patients avoiding thoracotomy.

O1-8
Clinical profile and follow-up data of patients included in the European Registry for ICD and CRT devices in Pediatrics and Adults with Congenital Heart Disease (Euripides): An Interim Report
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Introduction: The European Registry for ICD and CRT Devices in Pediatrics and Adults with Congenital Heart Disease (Euripides)

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has been founded as an initiative of the AEPC, ESC and the German Competence Network Congenital Heart Disease in 2007.
Methods: Demographic and medical data including indication and implantation details as well as obligatory yearly follow-up of treatment efficacy, complications and therapy termination were prospectively entered into the registry since 2007 using a pseudo-anonymized web-based data entry.
Results: A total of 72 ICD and 25 CRT patients from 12 centres (8 European countries) aged median 13.8 (IQR 9-17.5) years at implantation have been included. Of the ICD patients 33% had congenital heart disease, 14% had hypertrophic cardiomyopathy and 31% suffered from primary electrical disease. 40% of the indications were primary preventive. CRT was aimed at resynchronization of the systemic/sub-pulmonary/single ventricle in 88/4/8% of patients. 33% of the CRT patients were in NYHA class III or IV and 67% had prior conventional pacemaker implantation. At least one yearly follow-up was entered in 51% of all eligible patients yielding a median (IQR) follow-up period of 1.1 (IQR 0.9-2.0) years. In the ICD group actuarial freedom from adequate/inadequate therapy was 81.5/97.1% at 1 year. Of the CRT patients 91% were regarded clinical long-term responders. 9 surgical system revisions have been necessary in 7/72 ICD patients (9.7%) with an actuarial freedom from revision of 83% at 1 year.
Conclusions: Euripides is the only registry enabling prospective follow-up of ICD/CRT therapy in paediatric/congenital heart disease worldwide. Data entered allow for detailed analysis of therapy indications, efficacy and complications. Major problems are slow data volume growth and low adherence to annual follow-up reports. (JJ was supported by a grant of the Internal Grant Agency of Ministry of Health of the Czech Republic NT 12321-3/2011).

O2-1
Significance of Premature Restriction or Closure of Foramen Ovale in the Fetus

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Aims: To review the frequency and consequences of restrictive foramen ovale (RFO) in foetuses with and without structural heart disease.
Methods: 10 year review of 2324 foetuses that were referred for cardiac screening to the University Hospital of Wales.
Results: Premature restriction or closure of foramen ovale was encountered in 35 fetuses, of which 25 had isolated restrictive foramen ovale (IRFO) and right ventricular dilatation but the remaining 10 foetuses had associated congenital heart defects (CHDs). In fetuses with CHDs there were four with hypoplastic left heart syndrome, two with critical aortic stenosis, two with transposition of the great arteries, one with congenitally corrected transposition, and one with pulmonary atresia and intact ventricular septum. Two fetuses died in utero, three died after surgery, and four fetuses are alive after surgery. In fetuses with normal hearts, dilated right heart structures, redundant-aneurysmal primum atrial septum, and posterior angulation of the ductus arteriosus were the most consistent and striking features. A small aortic isthmus mimicking coarctation of the aorta, relatively small left ventricular cavity imitating hypoplastic left heart, partial obstruction of left ventricular inflow, and premature atrial contractions were other additional findings. One fetus who was born prematurely at 26 weeks died after birth, two foetuses had to be delivered early at 37 weeks of gestation due to severe restriction of mitral inflow but remaining 22 fetuses were delivered at term (mean 38.68 +/- 0.46 weeks). Appearance of redundant-aneurysmal atrial septum, and right heart dilatation both resolved rapidly in all newborns with no significant cardiac problems occurring in the follow up.
Conclusion: IRFO results in disproportionately large right ventricle and imitates serious congenital heart disease in the foetus. IRFO should be considered among the causes of right ventricular dilatation. Although restrictive foramen ovale confers a significant risk to foetuses with structural heart disease, IRFO appears to be well tolerated with favourable outcome in foetuses with structurally and functionally normal hearts.

O2-2
Relief of early post-operative Sano Shunt stenosis in neonates with hypopolastic left heart syndrome

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Introduction: Severe symptomatic shunt stenosis may occur at the proximal and at the distal shunt anastomosis after a Norwood Sano operation for hypoplastic left heart syndrome (HLHS). We report on successful relief of the stenosis by transcutaneous stent implantation. Hence, high risk repeated cardiac surgery was avoided.
Methods and Results: Since may 2000, a Norwood operation was performed in 138 newborns with HLHS at our centre at a median age of 8 days (1 – 38d). In the early experience a 5 mm Goretex tube was used for the Sano modification. Three patients developed a severe proximal Sano shunt stenosis 2-7 weeks postoperatively and were successfully treated by transcutaneous stent implantation. After this experience a ring enforced 5 mm Goretex tube was used for the subsequent Sano shunts and no further proximal shunt stenosis occurred. However, since then 4 additional patients developed severe (cyanosis SaO2<60%) early postoperative distal Sano shunt stenosis 1,2,5 and 6 days post-operatively. All patients were treated successfully by stent implantation. Five mm x 12 mm premounted balloon expandable stents (EV3, Genesis) were delivered through the femoral veines (5F short sheath). Two early postoperative catheter interventions were emergency procedures (arterial oxygen saturation was 53% and 58% respectively). The arterial oxygen saturation improved significantly (> 75%) and in three patients a partial cavopulmonary shunt was done so far (at the age of 9 weeks, 3 $\frac{1}{2}$ months and 5 $\frac{1}{2}$ months respectively). One patient is still waiting for this operation. There were no periprocedural complications.
Conclusion: Proximal Sano shunt stenosis may be prevented by a ring enforced Goretex tube. Distal Sano shunt stenosis may occur after a modified Norwood operation leading to profound early postoperative cyanosis. Transcutaneous stent implantation is a safe and effective alternative to high risk repeated cardiac surgery in these patients.

O2-3
Freedom from Antihypertensive Medication after Balloon Dilatation with Stent Implantation in Patients with Coarctation of the Aorta

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We assessed determinants of freedom from medication at long term follow up after stent implantation for coarctation.

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65 consecutive patients with native CoAo and ReCoAo underwent stent implantation between January 1998 and November 2011. Six patients with other causes for hypertension, complex heart or arch disease were excluded. Resting blood pressure, medication, imaging, Doppler and invasive data pre and post-procedure were studied. The remaining 59 patients were included. Mean age (standard deviation) at stent implantation was 28 (10.5) years, 56% male, 63% had native CoAo. 54 patients (92%) were on antihypertensive therapy before stenting, with 33 (61%) on multiple drugs (2 to 7).
Minimal diameter of coarctaction was 6 (2,7)mm. Twenty patients (34%) had transverse aorta/aorta diaphragm level (Tao/DiaphAo) <0.8.
A total of 64 stents were implanted, of which 39% were covered. A second procedure was performed in 9 patients (15%) because of multistage procedure (n = 4), growth (n = 2), stent fracture (n = 2) and neointima (n = 1). Invasive gradient decreased from 46 (18) mmHg to 5 (5) mmHg. There were no major complications nor mortality.
It was possible to discontinue one or more antihypertensive drugs in 39 patients (66%) and 22 patients (37%) became free of medication. Patients who remained medication free were younger 21,6(7,7) versus 31,4(15,9) years, p < 0,009; had a lower Doppler gradient 38,9(19,1) vs 58,3(19,9) mmHg, p < 0,001 and a lower invasive gradient before intervention 33,8(14,4) versus 52,8(23,5) mmHg, p < 0,001. Gradients immediately after stenting were also lower in this group 2,3(3,6) versus 6,7(8,5) mmHg, p = 0,026. In medication free patients, final stent diameter correlated with BSA (p = 0,035). In patients with Tao/DiaphAo > 0,8, 46% remained medication free, but with Tao/DiaphAo<0,8 only 23% did not require medication at long term. Results were similar for native CoAo and ReCoAo.
In a mean follow up of 4.8 (2,9) years, one patient died of stroke 4 years after the procedure.
Percutaneous stent implantation in patients with coarctation is safe, reducing the need for multiple antihypertensive drugs in most of the patients. Patients who became medication free were younger, with lower initial gradients, larger Tao/DiaphAo and lower immediate residual gradients.

O2-4
Mitral valve replacement in children: long-term results and risk factors influencing outcome

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Introduction: The aim of the study was to analyse the results of mitral valve replacement (MVR) in children less than 18 years of age.
Methods: Between 1991 and 2011, 71 consecutive patients underwent MVR at a median age of 1.5 (IQR 0.5 – 4.5) years. A retrospective analysis of echocardiographic, surgical and follow-up records was performed.
Results: Following diagnoses were the reason of the MVR: AV septal defect (n = 27), congenital mitral insufficiency (n = 18), Shone syndrome (n = 14) and other (n = 12). Forty eight patients required 1 to 3 previous operations (MV plasty in 33). In another 4 patients MVR was performed after an unsuccessful MV plasty during the same procedure. Eighteen (25%) patients died 0 days to 13.8 years (median 29 days) after the first MVR with an actuarial survival at 5/10 years of 77/77%. Univariate risk factors for early (<30 days) death were the absolute MV annulus size (median 13 vs 22 mm, P = 0.019) and the mechanical prosthesis vs MV annulus diameter ratio (1.33 vs 0.95, P = 0.003). Seventy one patients underwent a total of 103 MV replacements (102 mechanical/1 biological), from whom 20 (28%) required one re-MVR, 4 (6%) patients 2 and 1 (1%) patient four re-MVRs. Freedom from re-MVR was 76/49% at 5/10 yrs. No risk factors for re-MVR could be identified in the total group. However, early freedom from re-MVR (<6 years) was significantly better in patients >10 kg of weight (89 vs 65%, P = 0.028). Re-MVR did not influence late mortality (P = 0.491). From the 53 survivors with a follow-up of median 8.86 (IQR 4.8 – 13.0) years from the first MVR, 26 (49%) patients are in NYHA class I, 24 (45%) in class II, 1 (2%) patient in class III and 2 patients were not classified. Pacemakers have been implanted in 17 (24%) of patients (surgical AV block due to MVR in 13).
Conclusions: MVR has a significant mortality and low freedom from re-MVR. Low MV annulus size and prosthesis/annulus mismatch is a risk factor for early death. Patients <10 kg require earlier re-MVR. Most survivors have a good quality of life.

O2-5
Over-the-wire-technique device implantation: extended experience in difficult cases.

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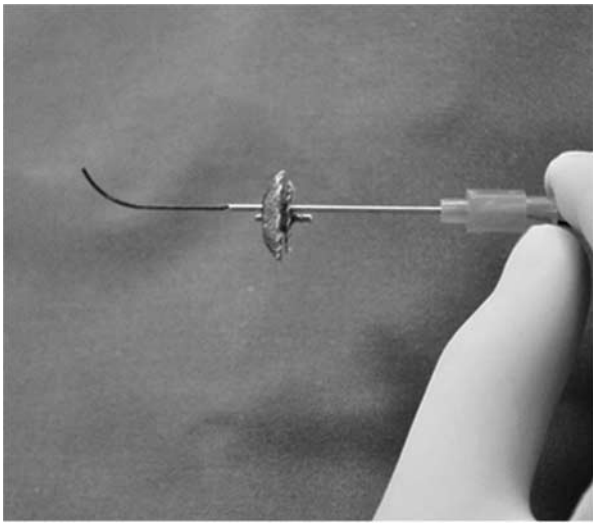
Transcatheter closure of cardiac defects is a mainstay of treatment in congenital and structural heart disease. Occasionally the devices used are useful in nonstandard or difficult positions but device embolization and malposition can be complicating factors necessitating percutaneous retrieval or emergency surgery. We describe a new 'over-the-wire' technique which allows guided safe deployment and easy retrievability if required.
Methods: We describe 10 cases in which Amplatzer devices were delivered over a wire in challenging anatomy in high risk patients. These cases included baffle leak in complex congenital heart disease (1 pt), paravalvular leak (2 pts), a large patent ductus arteriosus (1 pt), complex ventricular septal defects (2 pts), patent foramen ovalis (2 pts), atrial septal defect (2 pts). In each case the device was punctured close to the release mechanism and a guide wire fed through it (Figure upper), the device and guidewire then being loaded into the appropriate delivery system (Figure middle). The procedure allowed for perfect device delivery control and placamento (Figure bottom). Furthermore, the guidewire acted as a safety net in case of malposition/embolization that occurred in one case keeping the device in an harmless position and favouring its retrieval.



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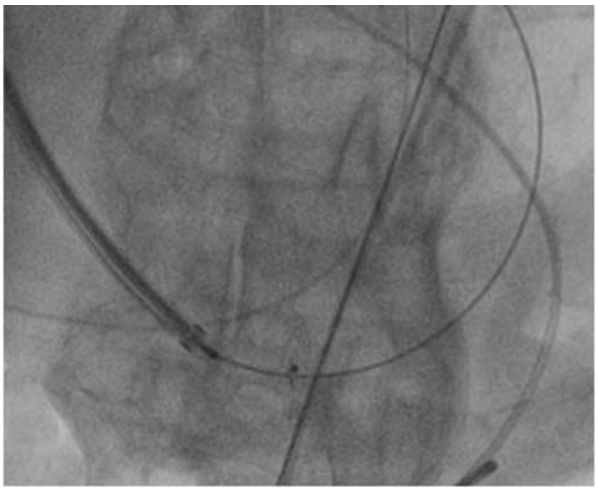

Conclusion: We describe the further experience with a novel 'over-the-wire' technique that is useful in deployment of Amplatzer devices in difficult and challenging anatomy, allowing careful controlled delivery and easy retrieval. This will help minimize procedural patient risk in complex cases.

O2-6

Hemocompatible Carbon Based Biosurfaces for Cardiovascular Implants

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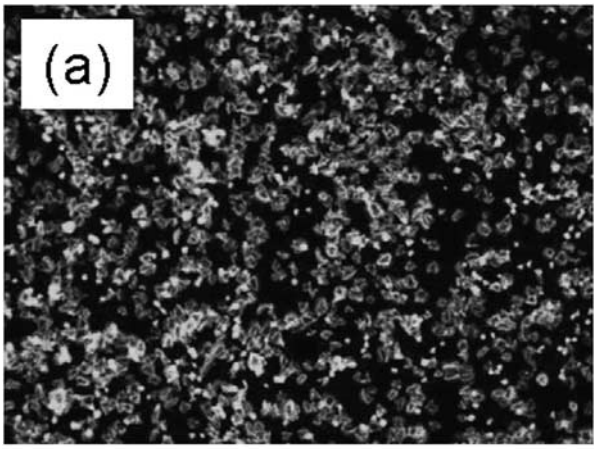
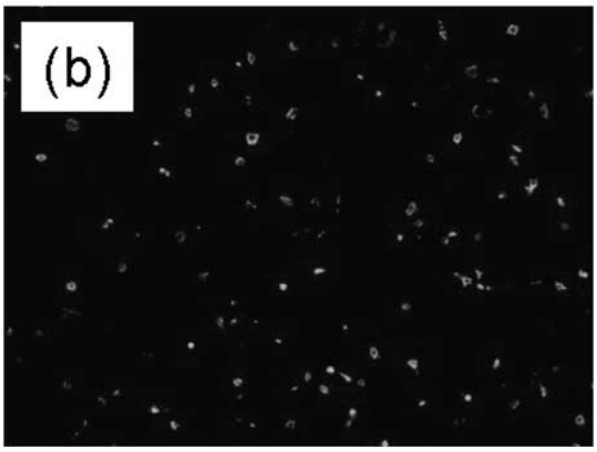
Introduction: Thrombus formation is one of the common problems observed at cardiovascular implants. Various coatings composed of material/material combinations have been developed to encounter this problem. Carbon biomaterials is one of the early tested materials in this field. More recently Diamond Like Carbon (DLC) has been considered an alternative material in cardiovascular medical devices. Here, we present development of a new type TiO_2-DLC bi-layer coating for reduction of the thrombosis formation.





Methods: A double layer coating was developed using two different types of techniques. The first layer consists of a thin TiO_2 film obtained by sol-gel method and using a dip-coating procedure. Afterwards, Si-DLC films were deposited on TiO_2 coated substrates using radio frequency (RF) plasma enhanced chemical vapor deposition (PECVD) technique. Acetylene (C_2H_2) and tetramethylsilane ($Si(CH_3)_4$, TMS) were used as the precursor gases.
For biocompatibility analysis, in-vitro monoculture models was established for Human Umbilical Artery Endothelial Cells (HUAEC) and Human Umbilical Artery Smooth Muscle Cells (HUASMC). Cells were seeded on prepared substrates and incubated under standard conditions. After reaching the desired confluence ($\sim$80%) each cell type was fixed and stained at different time points respectively after 24 h, 72 h and 96 h in order to visualize morphology and early adhesion behavior. Furthermore, scanning electron microscopy (SEM) was used for a detailed observation of the cytoskeletal behavior and orientation over the different surfaces. In addition platelet adhesion testing was performed to investigate the morphology, quantity and aggregation of the adherent platelets using fresh blood drawn from a healthy adult volunteer.
Results: While used cell types; HUAEC and HUASMC exhibit similar behaviours to those observed on control substrates, platelet activation is reduced TiO_2-DLC bi-layers (Fig. 1). In comparison to control substrates, no fibrin networks or platelet aggregation was observed on TiO_2-DLC bi-layers.
Conclusions: We have shown that TiO_2-DLC bi-layer is an effective surface modification method for reducing thrombosis. Such coating systems may play unique roles in next generation cardiovascular implants. On the other hand, detailed surface analysis

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(e.g. XPS) and cytotoxicity will be carried out to explore the mechanism of the platelet interaction with the surface.

O2-7
Left Pulmonary Artery Stenting in the Palliated Hypoplastic Left Heart
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Introduction: Left pulmonary artery (LPA) stenosis is a well recognized complication following palliation for hypoplastic left heart syndrome (HLHS). These lesions are a significant factor increasing resistance in a circulation in series. Therefore they are an important negative impact on haemodynamics and flow. Over the last decade we have taken an aggressive approach to stenting LPA stenoses. This is a single-centre experience looking at medium term outcomes.
Patients and Methods: Between 2000 and 2011, eighty patients palliated for HLHS underwent LPA stenting. Thirty-one underwent stenting prior to Fontan at a median age 51.1 (16.2-95.8) months and weight 15.5(9.3-19.5) kg. Four stents were inserted intra-operatively at Fontan completion (age 31-71 months, weight 13.1-17.5 kg). Fourty-five were placed following Fontan at median age 61.1(26.3-185.3) months, weight 17.2(11.5-55.2) kg and 12.9(0.3-1348) months-post. Indications for stent implantation were angiographic stenosis, long segment hypoplasia and/or LPA-SVC pressure gradient. This is a retrospective case-note review.
Results: Uncovered stents were used with median diameter of 10(8-15)mm and length 28(12-48) mm. Thirty-five patients have been re-catheterized at median 26(1-83) months after stent implantation. Twenty (57.1%) required no intervention with fourteen (40.0%) undergoing stent re-dilation to compensate for somatic growth. Re-stenosis due to neo-intima was rare, seen in only one (2.9%) case. There were no significant complications during any of these procedures.
Conclusion: LPA stenting is an important and effective therapy in patients palliated for HLHS to reduce overall vascular resistance in an effort to improve haemodynamics. Re-stenosis due to neo-intimal proliferation is rare and stents can be re-dilated to accommodate for somatic growth.

O2-8
Is stenting of pulmonary artery branch stenoses helpful to reduce the amount of pulmonary regurgitation in patients with Tetralogy of Fallot?
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Introduction: In patients with Tetralogy of Fallot (TOF) the extent of pulmonary regurgitation (PR) and the right ventricular volume are the main criteria for longitudinal evaluation of these patients. Weather an interventional treatment of stenoses of the left or right pulmonary artery has any impact on the timing or necessity of pulmonary valve replacement is under discussion.
Methods: Retrospective analysis of the indexed end-diastolic right ventricular volume (RV-EDV) and pulmonary regurgitation volume (PRvol) in 31 patients with TOF by cardiac MRI Sequential analysis over a period of 2 years. Group A: 16 patients (9 female, median age 15 years, RV-EDV $135.7 \pm 37.4\,ml/m^2$, PRvol $19.1 \pm 14.3\,ml/m^2$) had an CMR before and after stenting (mean interval 1.5 years). Group B: 15 patients served as control group (9 female, median age 12.5 years, RV-EDV $126.5 \pm 21.8\,ml/m^2$, PRvol $23.3 \pm 12.5\,ml/m^2$) and had no intervention.
Results: The RV-EDV in the control group increased significantly about 7.4% to $135.8 \pm 23.8\,ml/m^2$ ($p < 0.05$) without a significant change of the PRvol. Whereas in patient with intervention, even if not statistically significant, the right ventricular and pulmonary regurgitation volume decreased to $134.34 \pm 29.9\,ml/m^2$ and $15.2 \pm 19.5\,ml/m^2$, respectively.
Conclusion: Patients with Tetralogy of Fallot, PR and stenoses of the pulmonary artery branches may benefit from stent implantation already after a short period of time. Right ventricular enddiastolic volume remained stable whereas in the group without afterload reduction via stenting it increased significantly. These results may indicate that mechanical afterload reduction may delay the need and timing of pulmonary valve replacement, too.

O3-1
Reversible Pulmonary Trunk Banding. VIII. Glucose-6-Phosphate Dehydrogenase Activity of Adult Goat Myocardium Submitted to Ventricular Retraining
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Objective: Traditional pulmonary artery banding (PAB) is not always suitable for mature sub pulmonary ventricle retraining, with disappointing results in older patients. This study sought to assess myocardial function and glucose 6-phosphate dehydrogenase (G6PD) activity of the subpulmonary ventricle hypertrophy submitted to traditional versus intermittent PAB protocol in an adult animal model.
Methods: 18 adult goats were divided into three groups: Sham ($n = 6$, wt: 26.42 ± 2.63 kg, loose PAB, with no systolic overload), Traditional ($n = 6$, wt: 26.33 ± 2.32 kg, continuous systolic overload with fixed PAB), Intermittent ($n = 6$, weight = 25.17 ± 2.48 kg, daily 12-hour systolic overload with adjustable PAB). During a 4-week protocol, systolic overload was adjusted to achieve a 0.7 RV/systemic pressure ratio. RV, PA and aortic pressures were measured throughout the study. All animals were submitted to echocardiographic studies on a weekly basis, while hemodynamic evaluations were performed three times a week. After the study period, the animals were humanely killed for morphological and G6PD activity assessment.
Results: A 55.7% and 36.7% increase occurred in the Intermittent and Traditional RV masses, respectively, when compared with the sham group ($p < 0.05$), despite less exposure of Intermitent group to systolic overload. No significant changes were observed in RV water content in the 3 groups ($p = 0.27$). A worsening RV myocardial performance index occurred in the Traditional group throughout the protocol, compared with the Intermittent group ($P = 0.024$). Compared with the sham group (1.36 ± 0.14 nmol/min/mg protein), RV G6PD activity was elevated 55.15% in the Traditional group (2.11 ± 0.88 nmol/min/mg protein, $p = 0.05$) and only 10.29% in the Intermittent group (1.50 ± 0.24 nmol/min/mg protein).
Conclusions: Traditional continuous systolic overload for adult subpulmonary ventricle retraining causes upregulation of myocardial G6PD activity and RV dysfunction. It may suggest that the undesirable "pathologic systolic overload" is influenced by activation

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of penthose pathway and cytosolic NADPH availability. This altered energy substrate metabolism can elevate levels of free radicals by NADPH oxidase, an important mechanism in the pathophysiology of heart failure. In contrast, intermittent systolic overload promoted RV hypertrophy with better preservation of myocardial performance and smaller G6PD activity.

O3-2
An activable adhesive for patch closure of cardiovascular defects

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Background: Tissue adhesives have many advantages over sutures, namely in the reduction of operative times and procedural simplification. However, FDA approved tissue adhesives are associated with low adhesive strength, especially in the presence of blood and under dynamic conditions. Thus, we developed a light-activated adhesive, poly(glycerol-co-sebacate) acrylate (PGSA) that works efficiently on the beating heart in an in vivo model. The aims of this study were to evaluate in vitro adhesion in the presence of blood, obtain a quantitative evaluation of the long term adhesion on the epicardium, and the potential use for vascular anastomosis.
Methods: Adhesion strength was evaluated in pull-off tests using fresh epicardial tissue in the presence of saline or blood. Cyanoacrylate was used as control. A biocompatible patch was attached with PGSA or cyanoacrylate on the epicardium in a chronic in vivo rat model (n = 30). Hearts were explanted after 2, 7 and 14 days for histology and adhesion testing performed. A longitudinal incision (2 mm) of a porcine carotid artery was closed with a patch and PGSA in vitro (n = 4). Burst testing was performed after closure.
Results: In vitro adhesive strength was 1.2 ± 0.8 N/cm2 for PGSA in the presence of saline. Attachment of a biocompatible patch with PGSA and cyanoacrylate was shown to be feasible on the epicardium of a beating rat heart. The patches were well attached upon sacrifice 14 days after implant in the majority of the cases. Adhesive strength after 2 days was 0.34 ± 0.44 N/cm2 for PGSA. Histology revealed favorable biocompatibility of PGSA. The adhesive strength for PGSA did not decrease in the presence of blood (1.1 ± 0.7 N/cm2; $p = 0.7$). In contrast, cyanoacrylate was associated with a pronounced inflammatory reaction and the adhesive strength significantly decreased by 50% in the presence of blood. The PGSA patch on the carotid artery could withstand pressures of 300 mmHg before leaking.
Conclusions: PGSA is biocompatible and exhibits sufficient adhesive strength for attachment of patches or devices on dynamic tissues over the longterm and does not lose its adhesive strength in the presence of blood. Furthermore, it provides excellent anastomotic bursting strength in a model of vascular anastomosis.

O3-3
Deficiency of the cardiac potassium channel TASK-1 results in a LQTS phenotype and alters heart rate variability

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Introduction: TASK-1 is a potassium channel predominantly expressed in heart and brain. We have previously shown that TASK-$1^{-/-}$ mice have a prolonged QT interval in surface ECGs in sedation (ketamine). Heart rate variability (time and frequency domain) is significantly impaired in TASK-$1^{-/-}$ mice pointing to a sympathetic preponderance. The baroreflex sensitivity in TASK-$1^{-/-}$ mice is unknown and can be evaluated by heart rate turbulence (HRT). Although TASK-$1^{-/-}$ mice show a significant prolongation of monophasic action potential duration in isolated perfused hearts the electrophysiological role in vivo is unknown. First, we analysed rate corrected QT intervals (QT_c) in wake mice and after different drugs. Second, we evaluated the baroreceptor reflex by analyzing HRT. Third, TASK-$1^{-/-}$ and TASK-$1^{+/+}$ mice were characterized by programmed electrical stimulation.
Methods: Surface ECGs using different sedating drugs (avertin, pentobarbital, isoflurane) and telemetric ECGs by implanted transmitters were recorded and analysed. The HRT parameters turbulence onset and turbulence slope were determined after paced ventricular extrasystole and after ischemia/reperfusion. Programmed atrial and ventricular electrical stimulation using a transjugular octapolar catheter were performed to determine sinus node recovery time, Wenckebach point, atrial, atrioventricular and ventricular refractory periods. Atrial and ventricular vulnerability by burst stimulation before and after pharmacological stimulation with isoprenaline were studied.
Results: ECG analysis by telemetry showed a significantly prolonged rate corrected QT interval in TASK-$1^{-/-}$ mice (TASK-$1^{+/+}$ 43 ± 3 ms vs. TASK-$1^{-/-}$ 49 ± 5 ms, $n = 6$, $p < 0.05$). In surface ECGs QT interval was significantly prolonged in TASK-$1^{-/-}$ mice sedated with avertin and pentobarbital, two widely used drugs in mice (e.g. avertin: QT_c in TASK-$1^{-/-}$ 48 ± 4 ms vs. TASK-$1^{+/+}$ 37 ± 8 ms, $n = 13/16$, $p < 0.0001$). Of interest, isoflurane, known for its stimulatory effects on the TASK channel family, attenuates the QT_c prolongation in TASK-$1^{-/-}$ mice. Programmed electrical stimulation revealed normal values for electrical conduction and refractoriness. No significant arrhythmia after atrial and ventricular burst stimulation was induced in TASK-$1^{-/-}$ mice. However, turbulence onset is significantly altered in TASK-$1^{-/-}$ mice.
Conclusion: TASK-$1^{-/-}$ mice exhibit a phenotype of QT prolongation. The heart rate response after ventricular extrasystole is significantly abolished indicating an altered baroreceptor reflex. TASK-1 deficiency does not alter conduction velocity, refractoriness and vulnerability after electrical stimulation.

O3-4
Cardiovascular malformation spectrum of CHD7 mutations in CHARGE syndrome

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Background: CHARGE syndrome consists of a combination of congenital malformations including coloboma, heart defects, atresia of choanae, retardation of growth and developmental delay, genital anomalies and ear/semi-circular canal anomalies. A mutation of CHD7 is found in 70% of CHARGE patients.

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Cardiovascular malformations are considered a minor criterion for diagnosis because of lack of specificity.
Objectives: Study the panel of congenital heart defects (CHD) among CHARGE patients with a CHD7 mutation; discuss genotype-phenotype correlation and embryological basis of cardiovascular malformations.
Methods: We report on the spectrum of cardiovascular malformations in 75 CHARGE patients (including 15 fetuses) with CHD7 mutation. Cardiovascular phenotype was precisely assessed by echocardiography, catheterization and CT scan.
Results: Sixty-five CHARGE patients with CHD7 mutation (87%) have a CHD. Conotruncal malformations (25%), and atrioventricular septal defects (23%), are the most common. Aortic arch anomalies associated or isolated occur frequently. A rare association of atrioventricular septal defect and discordant ventriculo-arterial connections is described in 2 patients. Our data suggest that hypomorphic mutations are less frequently associated with a heart defect than nonsense and frameshift mutations.
Conclusions: Congenital heart defects are a frequent feature of CHD7 mutated CHARGE patients. The broad observed panel suggests the involvement of neural crest cell as well as cardiac progenitor cells of the second heart field and endocardium in embryological mechanisms leading to CHDs in CHARGE syndrome. More cardiac regulator genes have yet to be identified as potential targets of CHD7.

O3-5
Identification of novel genetic risk loci associated with fetal outcome in congenital heart block

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Objective: Congenital heart block (CHB) is a life threatening heart condition where the fetal atrioventricular conduction system is disrupted. The block may occur during fetal development after placental transfer of autoantibodies from Ro/SSA positive mothers. The association with maternal Ro/SSA antibodies is well established; however the recurrence risk is only 11-20% despite persisting autoantibodies in the mother, suggesting that additional factors other than the maternal autoantibodies determine disease development and fetal outcome. We hypothesized that the fetal genetic composition is a major factor influencing susceptibility to CHB, and therefore conducted a genome-wide association study to map potential genetic risk loci involved in CHB pathogenesis.
Methods: Genotyping of 561,490 SNP markers was accomplished in DNA samples from 389 individuals of 104 Swedish Caucasian families with at least one case of congenital heart block and a maternal serum positive for Ro/SSA autoantibodies. A family-based association analysis was performed using the transmission disequilibrium test (PLINK). The Cochran-Mantel-Haenszel statistical test was used to investigate SNP associations with CHB in index cases (n = 88) compared to Swedish Caucasian population-based healthy controls (n = 1710).
Results: A family-based analysis (n = 348) revealed that 37 SNP markers were associated with CHB ($p \leq 9.88 \times 10\text{-}5$, OR 0.18-6.50). In a case-control analysis between the index cases and population-based healthy controls 6 of the 37 SNP markers were confirmed. The CHB risk loci are located at the chromosomal positions 1p31.3, 2q35 and 3p25.1 and genes encoded within these regions are involved in cellular stress responses, energy metabolism and tissue repair.
Conclusion: Our study identifies three novel genetic risk loci associated with development of CHB after exposure to maternal Ro/SSA autoantibodies. Functional exploration of the genes encoded within these risk loci will help to shed light on the cellular mechanisms underlying CHB pathogenesis.

O3-6
Propofol administration to the fetal-maternal unit reduced myocardial injury in late-preterm lambs subjected to severe prenatal asphyxia and cardiac arrest

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Introduction: Cardiac dysfunction is reported in the majority of cases after severe perinatal asphyxia. We hypothesized that the maternal anaesthesia during an emergency caesarean section offers a therapeutic window and that propofol administration to the maternal-fetal-unit can diminish cardiac injury and dysfunction in preterm fetuses exposed to global severe asphyxia in utero. We proposed that propofol would interfere with the fetal mitochondrial apoptosis pathway by activating anti-apoptotic cellular mediators, such as the kinase family AKT and the signal transducer and activator of transcription-3 (STAT-3) with its effector molecule B-cell lymphoma-extra large (Bcl-xL). Bcl-xl can reduce the cytochrome c release from mitochondria which results in less activation of apoptosis initiating and effector caspases, such as caspase-3 and -9.
Methods: 36 late-preterm lambs (133d gestational age (GA), term 150d) underwent standardized total umbilical cord occlusion (UCO) or sham-treatment in utero. Eight animals served as GA controls. UCO resulted in global asphyxia and cardiac arrest. Mothers were randomized to either propofol or isoflurane anaesthesia. After emergency Caesarean section, the fetuses were resuscitated and subsequently anaesthetized the same way as their mothers. Cardiac function and injury was assessed by determination of left ventricular ejection fraction (LVEF) and troponin T release into the plasma.
Results: Propofol treatment resulted in higher median LVEF of 84% in comparison to isoflurane treatment (LVEF = 74%) after cardiac arrest, resuscitation and 8 h of intensive care. Cardiac troponin T increased after asphyxia only in isoflurane treated lambs. Fetal propofol treatment before and after UCO resulted in reduced activation of caspase-3, caspase-9, and cytochrome c release when compared to asphyctic isoflurane animals. On the other hand, Bcl-xL increased to 269%, its transcription factor pSTAT-3 to 655% and the AKT kinase family by nearly 5-fold if compared to isoflurane animals suffering from asphyxia.
Conclusions: Early propofol administration to the maternal-fetal unit preserved cardiac function of late-preterm lambs after cardiac arrest and resuscitation better than isoflurane. The underlying mechanism is probably an activation of anti-apoptotic STAT-3 and AKT pathway.

O3-7
Echocardiographic assessment of normal embryonic and fetal mouse heart development

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Background: The identification of several genetic mutations involved in congenital heart disease has led to the generation of specific genetically mutated mouse models to study normal and abnormal cardiac development. Up till now studies in embryos/fetuses of these models mainly focused on the cardiac morphology and the availability of functional i.e. hemodynamic data is limited. In this study we assessed the morphological and hemodynamic parameters of normal developing mouse embryos/fetuses by using a high-frequency ultrasound system.
Methods: A timed breeding program was initiated with a wildtype mouse line. All recordings were performed in sedated (Isoflurane 1.5%) pregnant mice under stable vital parameters (heart rate, respiratory rate and body temperature). Trans-abdominal echocardiographic assessments were performed in individual embryonic hearts at 12.5 days post conception (dpc), which were also assessed at early and late fetal stages of development, 14.5 and 17.5dpc respectively ($n = 105$).
Results: At the three stages assessed we could clearly visualize the individual compartments of the embryonic/fetal hearts: the left and right atria and ventricles and the developing inter-ventricular septum. Furthermore, at 14.5 and 17.5dpc the aorta and pulmonary artery were identified. Between 12.5 and 17.5dpc the embryonic/fetal heart rate increased significantly from 125 ± 9.5 to 219 ± 8.3 beats per minute. Reliable echo-Doppler recordings were made in the common atrioventricular canal and outflow tract (12.5dpc) and across the developing mitral, tricuspid, pulmonary and aortic valves orifices (14.5 and 17.5dpc). Furthermore, M-mode recordings (short/long axis) were made of the developing ventricles in order to calculate the shortening fractions (SF%) and diameters of the ventricular walls and inter-ventricular septum.
Conclusion: High-frequency echocardiography is a promising and useful imaging modality for structural as well as hemodynamic analysis of embryonic/fetal mouse hearts at subsequent stages of development. The implementation of this technique in embryos and fetuses of genetically mutated mouse models will provide important new data to unravel the etiology of congenital heart diseases. Furthermore, these data will help in the detection of human congenital heart disease at early stages of pregnancy.
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O3-8
Mesenchymal stem cells improve functional and morphological integration of induced pluripotent stem-cell derived cardiomyocytes into ventricular tissue

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Introduction: Transplantation of induced pluripotent stem cell-derived cardiomyocytes (iPS-CMs) into damaged myocardium might become a therapy to improve contractile function. However, current knowledge on the mechanisms of cell integration and processes of physiological reconstitution as well as mechanical and electrical coupling after transplantation into the host tissue is still fragmentary. There is cumulating evidence reporting beneficial effects of cell transplantation strategies combining a source for CMs with other cell types. From these observations we hypothesized that non-myocytes might be necessary for an improved functional integration.
Aim: To investigate whether murine mesenchymal stem cells (MSCs) improve functional and morphological integration of iPS-CMs into cardiac tissue.
Methods: Murine ventricular slices (diameter 300 μm) were cocultured with iPS-CMs and MSCs for 4 days. Integration was evaluated by visual methods, intracellular recordings via sharp electrodes and fieldpotential recordings and propagation maps via multi-electrode array (MEA) measurements. Data are represented as mean ± standard deviation, level of significance is set to $p < 0.05$
Results: IPS-CM clusters had an average beating frequency of 333 ± 140 ($n = 32$). Vital slices with ($n = 28$; 77.5 ± 54.8 bpm) or without ($n = 29$; 77.7 ± 43.7 bpm) iPS-CM clusters served as controls and displayed similar average frequencies after 4 days of co-culture. Co-cultures of vital slices with IPS-CMs and MSCs ($n = 16$; 237.6 ± 132.6 bpm, $p < 0.001$) showed a significant increase in beating rates compared to the controls indicating an improved electrical integration of the iPS-CMs. The improved integration of rapidly beating iPS-CM cluster induce a raised frequency of the slice, meaning that the IPS-CM cluster serves as a pacemaker for the cardiac slice. Morphological observations as well as propagation spread studies indicate that the improved engraftment is mediated by MSCs.
Conclusion: We conclude that non-cardiac cells like MSCs support morphological and electrical integration of iPS-CMs. MSCs are an easy accessible cell source and could be used in future as mediator cells for a successful transplanatation of iPS-CMs.

O4-1
New generation decellularized biological valve as pulmonic conduit in congenital heart surgery

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Objective: Establishment of right venticle to pulmonary continuity with a valve is indispensable in correction of many complex cardiac pathologies. In an attempt to creat an ideal conduit tissue engineers developped decellulerized biologic valved conduits. We report our surgical experience with last generation completly decellularized biological valved conduit in a heterogenous group of pediatric patients .
Methods: Since October 2010, 21 patients (median age 60 months; range:7 months to 29 years; median weight 15 kg; range 7 to 71 kg) with different diagnosis had implantation of decellulerized valved conduit in our hospital. 15 patients had Rastelli type operation, and in rest of the group; 2 patients undergone Ross procedure, 2 patients had total correction of VSD and pulmonary atresia, 1 patient had Truncus arteriosus repair and there was only one patient who had previously undergone right ventricle outflow tract conduit replacement.
Results: The implanted valve sizes ranged from 15 to 21 mm (median size was 17 mm). No mortality occured at perioperative or during follow up period (mean 8,5 ±4,8 months). The only valve related morbidity was revision for excessive drainage in one patient, the other 20 patients had no valve related problems during their follow up. In one patient who had pulmonary branch arterioplasty during corrective surgery, postoperative echocardiography showed right ventricular outflow gradient

more than 20 mmHg but it was measured at distal segments of pulmonary bed and valve fuction was normal.
Conclusion: The short term good results of this new generation valve is encouraging for the future. We believe that this conduit can be a good alternative for homografts in pulmonic position with its resistance to degeneration and can be more advantageous because of its growth potentiel especially in small age population.

O4-2
Incidence of interventions after Norwood operation: Comparison of Sano and modified Blalock Taussig shunt

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Background: With improved results after the modified Norwood procedure (NP), midterm outcome is mainly determined by interstage hazards leading to mortality, emergent interventions or premature superior cavopulmonary connection. This study compares incidence of interstage cardiac catheter interventions of the Sano and modified Blalock Taussig shunt (BT).
Methods: Incidence, location, interval and weight at interventions of 107 neonates (Sano, n = 37 and BT, n = 70) undergoing NP within the same interval from October 2002 through December 2009.
Results: 46 (43.0%) subsequently underwent interventions, most commonly for dilatation of the aortic arch (DAA, n = 26, 24.3%, Sano n = 10, BT n = 16, p = 0.6), of the shunt (DS, n = 15, 14.0%, Sano n = 11, BT n = 4; p = 0.002) or for closure of aortopulmonary collaterals (APC, n = 15, 14.0%, Sano n = 3, BT n = 12; p = 0.08). Mean interval and bodyweight at intervention (DS, DAA, and APC) was 72.4 ± 18.9, 108.5 ± 15.8, 110.7 ± 17.8 days and 4.5 ± 1.3, 4.9 ± 1.9, 5.3 ± 1.2 kg, respectively. Interventions were not related with death, but with increased rate of complications (9/46 = 21.4%), if compared to diagnostic catheterization (0/45, p = 0.03). Complications included closure of femoral or subclavian artery (n = 5), cerebral embolic or bleeding events (n = 4), cardiopulmonary resuscitation (n = 3) and temporary heart block (n = 2). Actuarial survival was similar from the 8th postoperative month onwards at 78.6 ± 4.9% (95% CI, 67.0–86.5%) for Sano and 78.4 ± 6.8% (95% CI, 61.4–88.6%) for BT: p = 0.95.
Conclusion: Interventions after NP were common, irrespective of shunt type. However a significantly higher rate of shunt interventions was noted in the Sano subgroup. Especially interventions addressing the aortic arch and the Sano shunt were related with significant rate of complications.

O4-3
Does posterolateral thoracotomy with cardiopulmonary bypass play a role in the surgical management of aortic coarctation?

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*Objectives:*The aim of this study was to evaluate the surgical treatment of aortic coarctation (CoA) through posterolateral thoracotomy using cardiopulmonary bypass (CPB) to increase the safety margin as part of spinal cord protection.
Methods: Between 1997 and 2011, 15 patients underwent surgical repair of CoA through a left thoracotomy utilizing CPB. CPB cannulation was performed at aorta descendens distal from the CoA site and the main pulmonary artery for venous return. The clinical outcome regarding the development of restenosis, major neurologic complication as well as spinal deformities was studied.
Results: There was no mortality. One patient acquired rethoracotomy for hemorrhagic control. At a mean follow-up of 4.0 years (range from 13 days to 14 years), two patient developed a recurrent stenosis at the CoA repair site. In the remaining 13 patients, echocardiography and MRI showed a widely patent anastomosis with no evidence of a hemodynamically significant gradient. None of the patients developed paraplegia, 3 patients demonstrated left diaphragmatic paresis and 3 patients developed transient left recurrent nerve palsy. No development of scoliosis as a result of current surgeries, no postoperative renal failure and visceral ischemia was observed. One patient developed chylothorax.
Conclusions: CoA without hypoplasia of the proximal aortic arch and intracardiac anomalies can be repaired with low mortality and morbidity via a left thoracotomy with CPB. The use of CPB reduces spinal cord and lower body ischemia. It provides a sufficient amount of time for the anastomosis, which allows a better anastomotic quality. In addition, CPB offers a possibility to carry out hypothermic circulatory arrest in management of complex anatomy and intraoperative bleeding. We strongly recommend the use of CPB in complex CoA repair.

O4-4
Early surgical outcome of a new modification for Senning procedure

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Objective: Since it was first described original Senning procedure was modified technically to prevent longterm problems such as baffle obstruction and leak and arrhythmia. According to lessons learned from different patient groups in litterature we developped another modification of this architectural surgery. The aim of this study is to evaluate early results of this modified technique.
Methods: Between 2009-2011 17 patients had modified Sennig procedure with diagnosis of simple TGA except one patient with baffle stenosis of previous Mustard operation. Median age and weight of our patients were 12 months (range: 3months-27 years) and 8,3 kg (range:3,5-79 kg).
All operations were performed with hypotermic cardiopulmonary bypass and under cold cristalloid antegrade cardioplegia. Senning repair was performed without patch augmentation of the septum and creation of pulmonic venous chamber was done with sutureless technique as described by LaCour Gayet. Mean cpb and ischemic times were 76,3 ± 14,2 and 61,6 ± 31,8 min respectively.
Results: All patients were follwed and mean follow up time was 12,76 ± 7 months. There is no mortality and none of the patients had rhythm disturbance and baffle problems, mitral dysfunction or left ventricle outflow obstruction at hospital discharge or during follow-up
Conclusion: We suggest that this modification of Senning procedure is a good technical alternative with encouraging early outcome. This method can prevent baffle obstruction by avoiding patch material and can have growth potential of venous chambers according to the age of patient.

O4-5
Transposition of the great arteries with intact ventricular septum. Arterial Switch Operation in patients one month of age or older.

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Objective: Neonatal arterial switch operation (ASO) is the procedure of choice for the repair of transposition of the great arteries and intact ventricular septum (TGA-IVS). Classically, after 3 weeks old the ability of the Left ventricular (LV) to sustain a systemic output remains uncertain and LV retraining could be necessary.
The aim of this study was to review our experience with patients with TGA-IVS and late referral to surgery.
Methods: Between 1993 and 2011, 78 patients with TGA-IVS older than 30 days old were referred to our institution for an ASO. The median age was 62 days (30 to 1070 days). Decision to perform a primary ASO was roughly based on the values of LV mass ⩾35 g/m2 and when between 25 and 35 g/m2 on the value of LV mass/volume ratio > 1.2. Below these levels, a rapid two stage approach was favoured. Primary ASO was performed in 40 (Group I)(median age 43, 30 to 187 days) while38 (Group II) (median age 112, 32 to 1070 days) underwent a rapid 2-stage (median delay 9days) management with initial pulmonary artery banding associated or not with a systemic to pulmonary shunt.
Results: Overall mortality was 3, 8% (3/78). It was 5% in group I versus 2.6% in group II. Mechanical LV support was required in 2 patient of Group I versus 4 in group II. Patients in group II were older than in group I (145 versus 57 days, $p < 0.0001$). Mortality and post operative morbidity were not influenced by age, LV geometry, LV mass index, LV posterior wall thickness index, LV volume index, LV mass/volume ratio, patent arterial duct or patterns of coronary anatomy.
Conclusions: Despite late referral, and initially inadequate left ventricular quality, patients 30 days of age or older with TGA/IVS can successfully be treated with the arterial switch procedure, provided that the left ventricle is adequately prepared. Either primary ASO followed by mechanical LV support or rapid two stage approach are adequate managements which provide good results.

O4-6
Outcome of the Ross procedure in 100 Children and Adults : Low mortality, excellent survival but frequent reinterventions during mid-term follow-up

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Background: Ross procedure (RPR) offers excellent hemodynamic and clinical outcome but questionable long-term durability. There are little data on long-term outcome and predictors of reintervention after this procedure.
Methods: Between 1993 and January 2011 (89 interventions after Jan 1, 2000), 100 children and adults (76 males; mean age 17 ± 12 years) underwent a RPR consisting in a root replacement at our center. In all patients (pts), pre- and postoperative clinical and echocardiographic data were analyzed as well as surgery reports, and mid-term follow-up (survival, NYHA class, frequency of reinterventions or endocarditis).
Results: Aortic valve (AV) pathology leading to RPR were congenital heart disease (including 64 bicuspid AV,12 monocuspid AV, 12 tricuspid, 3 quadricuspid, 9 indeterminate); a history of prior endocarditis (6 pts) and rheumatic heart disease (2 pts). 52 pts had previous cardiac interventions, including coarctation surgery (5 pts).
RVOT replacement was made with a pulmonary homograft (66 pts) or a Contegra graft (31 pts) in most. Procedures included reduction surgery of the ascending aorta (19 pts), and resection of subaortic stenosis/myectomy (9 pts).
Perioperative mortality was 1%: one 8 year old pt with postoperative stroke died 3 weeks postoperatively of ventricular fibrilllation.
Mid-term follow-up was available in 97 pts (98%) after 5.6 ± 3.8 years. 94 of 95 pt were in NYHA class I or II. Any dilatation of the aortic root or ascending aorta (Z score>4) was observed in 32 of 94 pts (34%). Postoperative endocarditis occurred in 2 pts (1x Contegragraft, 1x autograft). Reinterventions were necessary in 23 pts (24%): most frequently valvuloplasty of the RVOT (7 pts), percutaneous pulmonary valve replacement (6), aortic root procedures (3) and homograft replacement (3 pts). Death occurred in 2pts (heart failure in both). 5 year freedom from reintervention was 83.8 + 4.5%.
Conclusion: Ross procedure in pts with predominantly congenital aortic valve disease has low morbidity and mortality. Mid-term follow-up shows an excellent functional class, however, besides aortic dilatation (34%) also reinterventions are frequent (24%) especially in the RVOT.
This necessitates regular postoperative surveillance after RPR.

O4-7
Valved patches for closure of ventricular septal defects in patients with severe right ventricular pressure overload

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VSD closure in patients with severe RV pressure overload secondary to pulmonary hypertension or pulmonary artery (PA) hypoplasia is often associated with significant morbility and mortality.
The records of 11 patients operated with VSD closure with valved patches since January 2000 were reviewed. Ages ranged from 1 to 23 years (median 8 years); 6 were male. RV pressures ranged from 87 to 120 mmHg (90% at systemic level).
Six patients had large VSDs (5 perimembranous, 1 muscular) all with pulmonary hypertension (PAH). In this group mean PA pressure was 60,4 mmHg, mean PVR was 6,9 Wood units (4,2 to 11,4 in air, decreasing to 4,3 (1,3 to 9,7) after O2/NO testing). QP/QS ratio ranged from 1.2 to 3.3 in air and after vasorreactivity test 3,0 to 6,2.
The remaining 5 patients had Tetralogy of Fallot (TF) or PA atresia and hypoplastic PAs, with previous BT shunts or RV to PA conduit (5), stenting of PAs (2) and MAPCAs embolization (3). All patients underwent VSD closure with a unidirectional valved patch constructed with a dacron sauvage patch with a 5 to 7 mm circular hole and a monocusp valve of 0,1 mm thick PTFE or pericardial patch on the left side. In TF/PA group surgery for RVOT and PA bifurcation was performed in 2 patients.
There was no mortality. Mean follow up was 3,8 years (1 to 11 years). In the VSD group, 4 patients had episodes of right to left (RL) shunt in the post-operative period, well tolerated hemodinamically, requiring NO during 8 to 30 hours. There was no evidence of late RL shunt. In the TF/PA group, the immediate postoperative period was well tolerated without evidence of RL shunt. One patient developed high RV pressure, tricuspid regurgitation and severe systemic congestion 4 years after surgery, with no shunt at patch level at this time. Another patient had bacterial endocarditis 2,5 years after surgery.
Closure of large VSD's with valved patches in patients with PAH or PA hypoplasia provided encouraging results. The possibility of

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surgical correction in this group of patients should not be dismissed without consideration of this alternative approach.

O4-8

ECMO in the perioperative course of pediatric heart surgery

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The aim of this study was to assess the results of ECMO as a perioperative support in children with congenital heart disease (CHD) during and after open-heart surgery.
Material and methods: All patients aged<18years at surgery who needed ECMO support from 2004 to 2011, were included in the study. Clinical and biological data, demographics and outcomes were retrospectively reviewed.
Results: Twenty-seven patients (19 males) (i.e. 0.5% of total pediatric cardiac surgical procedures performed per year), aged 3days to 18years (mean 1.6years) were placed on ECMO, per-operatively in 10 (37%, for failure to wean off bypass) or during the early postoperative course in 17 (8 < 24th hour, 9 > 24th hour) for cardiac arrest (33%) or low cardiac output (30%). Surgical repair included: severe form of tetralogy of Fallot (4), complex arterial switch operation (7), complex left heart obstruction (5), Rastelli (4), ALCAPA (1), cavopulmonary anastomosis (3) and miscellaneaous (3). Twelve cases were in-hospital preoperatively, of whom 7 were dependent on mechanical ventilatory support.
Five patients died while on ECMO because of multi organ failure (4) or pulmonary hypertension (1). Main complications during support included hemorrhages (15 cases), renal failure requiring peritoneal dialysis (14), hemolysis (13), canulas thrombosis (6), and strokes (4). Only 3 cases were free from complication. Duration of ECMO was 5.4 ± 3.6days (1 to 16, median 5), of CICU stay 26 ± 16days (10 to 69, median 22). Survival to ECMO was 81.5% (22 patients) and overall survival was 59% (16). Significant predictive factors for mortality were: preoperative clinical status (in-hospital: 25% in alive patients versus 73% in deceased cases, $p = 0.01$), lactates level at onset of ECMO (mean 6 versus 10, $p = 0.004$) and duration of aortic clamp (mean 70 versus 110mn, $p = 0.05$).
Conclusion: This study shows that post-cardiotomy ECMO in children is a valuable therapeutic option as a bridge to recovery, despite high frequency of complications on support.

O4-9

Subsequent nitric oxide with inhaled iloprost versus inhaled iloprost alone in pulmonary hypertension following cardiac surgery in children with congenital heart disease

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Background: One of problems sometimes encountered following pediatric cardiac surgery is Pulmonary Hypertension that associated with significant mortality and morbidity. It is desirable to treat the condition and return haemodynamics to normal as quickly as possible, and enable a prompt removal of ventilator support. In our centre, the management of Pulmonary Hypertension consist of deep analgesia and sedation, controlled ventilation and inhaled Nitrous Oxyde (iNO) continued with inhaled iloprost.
Aim: The primary end-point of this study was to compare the efficacy of inhaled nitric oxide (NO) plus inhaled iloprost versus inhaled iloprost alone in reducing pulmonary artery pressure after cardiac corrective surgery in children with congenital heart disease (CHD) and pulmonary hypertension. The secondary end-point was to assess the safety of inhaled ilosprost and inhaled NO.
Method: A prospective, randomized, open label study was conducted in Harapan Kita National Heart and Cardiovascular Center, Jakarta, between March 2008 and March 2009. Children with CHD and secondary pulmonary hypertension were recruited. After surgery, patients were randomized to receive inhaled NO+iloprost or inhaled iloprost alone. Mean pulmonary artery pressure (mPAP), mean systemic arterial pressure (MAP), and the ratio of mPAP/MAP were measure at baseline, during treatment, and at the end of the study.
Results: Thirteen children were eligible for this study. Treatment with inhaled NO plus iloprost and iloprost alone were equally effective in reducing mPAP. The mPAP was lower in inhaled NO+iloprost group than iloprost alone, but the difference was not statistically significant (23.0 ± 7.7 vs. 30.4 ± 15.9 mmHg; $p = 0.359$). An adverse event of hypotension occurs in one patient in the iloprost alone group.
Conclusion: Inhaled iloprost alone or inhaled nitric oxide plus iloprost were equally effective in lowering pulmonary artery pressure in children underwent corrective cardiac surgery for CHD. Inhaled iloprost alone is more preferable due to its simpler administration.

O5-1

Analysis of Immunological Function of Infants with Congenital Heart Disease after Cardiopulmonary Bypass Surgery: Evaluation the Optimum Timing of Vaccination

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Background: The optimal timing of vaccination of infants with congenital heart disease (CHD) after cardiopulmonary bypass (CPB) surgery is not clear. They have to be received a number of vaccines earlier, such as *Haemophilus influenzae* Type b vaccine, pneumococcus vaccine, influenza vaccine. The purpose of this study is to examine immunological function of infants with CHD after CPB surgery for evaluating the optimal timing of vaccination.
Methods: The 14 infants with CHD after CPB surgery were examined. The age ranged from 2 months to 24 months (9.3 ± 6.7months). We analyzed immunological function of infants with CHD 1 month to 3months after CPB surgery (2.1 ± 0.7months). The IgG, IgM, IgA, IgD, B cell surface immunoglobulin(Sm-Ig), CD3, CD4, CD8, CD4/8, CD20, CD56 were measured using flow cytometry. Lymphocyte transformation test was also performed. We investigated measles and rubella(MR) virus antibody value in infants at before and after CPB surgery who were already received MR vaccine before CPB surgery.
Results: All of both humoral and cell mediated immunological function in infants with CHD were normal value at 1 month to 3 months after CPB surgery (IgG 587.7 ± 299.5 mg/dl, IgA 34.5 ± 28.0 mg/dl, IgM 68.8 ± 40.5 mg/dl, IgD 0.6 ± 0 mg/dl, Sm-IgG 1 ± 0%, Sm-IgA 1 ± 0%, Sm-IgM 15.6 ± 7.3%, Sm-IgD 14.9 ± 7.2%, Smκ 8.5 ± 4.2%, Smλ 6.8 ± 3.3%, CD3 59.2 ± 14.7%, CD4 39.3 ± 11.5%, CD8 24.7 ± 7.3%, CD4/CD8 1.8 ± 1.0, CD20 19.2 ± 7.7%, CD56 17.1 ± 7.9%, PHA 43424 ± 14092cpm). The MR virus antibody in 2 infants were was normal value even at after CPB surgery.

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Conclusions: Both humoral and cell mediated immunological function of infants at 1month to 3 months after CPB surgery were normal value. These results suggested that optimal timing of vaccination in infants with CHD was 1month to 3 months after CPB surgery.

O5-2

Diagnostic value of parental electrocardiographic screening in congenital and childhood, non-immune, isolated atrioventricular block.

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Introduction: The etiology of congenital or childhood non-immune, isolated AV block remainsunknown. Wehypothesizedthatthis conduction abnormality in the youngmaybe a heritabledisease.
Methods: A multicenterretrospectivestudy (13 French referralcenters, from 1980 to 2009) allowed inclusion of 141 childrenwith AV block diagnosedin utero, atbirth or before 15 years of age, without structural heartabnormalities and withoutmaternalantibodies. Parents and matchedcontrolswereinvestigated for familyhistory and for ECG screening.
Results: In parents, familyhistory of suddendeath or of progressive cardiac conduction defectwasfound in 1.4% and 11.1% respectively. Screening ECGsfrom 130 parents (meanage 42.0 ± 6.8 years, 57 couples) werecompared to 130 matchedhealthycontrols. All parents wereasymptomatic and in sinus rhythm, except one withunknowncomplete AV block. Conduction abnormalitieswere more frequent in parents than in controls, respectivelyfound in 50.8% versus 4.6% ($p < 0.001$). Long PR intervalwasfound in 18.5% parents but never in controls ($p < 0.001$). Complete or incomplete right bundle branch block wasobserved in 39.2% parents and 1.5% controls ($p < 0.001$). Complete or incompleteleft bundle branch block wasfound in 15.4% parents and 3.1% controls ($p < 0.001$). Heritabilityestimate for isolated conduction disturbanceswasveryhigh, calculatedat 91% (standard error = 1.019, p = 2.10-16).
Conclusion: ECG screening in asymptomatic parents fromchildrenaffected by idiopathic AV block revealed a highprevalence of conduction abnormalitieswith prolongation of intra-atrial, AV and/or intra-ventricular conduction delay. Heritabilityestimateconfirmed a high contribution of geneticfactors. Theseresults support the hypothesis of an inheritable trait in congenital and childhood non-immune, isolated AV blocks.

O5-3

Impaired TGF-β mediated cardiac healing in patients with right ventricular dilatation after repair of tetralogy of Fallot

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Introduction: The transforming growth factor (TGF)-β pathway plays an important role in the setting of left ventricular pressure overload and post-myocardial infarction compensatory processes with regard to cardiac hypertrophy, fibrosis, and remodeling. Our aim was to evaluate the role of TGF-β in patients with right ventricular (RV) volume overload after tetralogy of Fallot (TOF) repair.
Methods: 96 patients (22 (7–66) years) were included and underwent magnetic resonance imaging to assess RV size and function. Serum levels of TGF-β were assessed using ELISA kits; inflammatory factors (e.g. tumor necrosis factor-α and interferon-γ), cardiomyocyte growth factors (e.g. epidermal growth factor), and metalloproteinase inhibitor 2 (i.e. tissue inhibitor of metalloproteinase (TIMP) 2) were measured using protein array analysis. In 36 patients, serial follow-up measurements were available with a 5-year interval (14 ± 5 years at baseline). Results were compared to 70 healthy controls (21 (12–64) years).
Results: TGF-β tended to be lower (3.8 ± 3.3 ng/ml vs. 6.3 ± 5.7 ng/ml, $p = 0.10$) in patients with a large RV (RV end-diastolic volume (EDV) 171 ± 22 ml/m2) than in patients with a small RV (RVEDV 102 ± 12 ml/m2). In the serial follow-up patients, RVEDV increased (137 ± 33 ml/m2 to 148 ± 38 ml/m2, $p = 0.001$) over 5-years time, while TGF-β decreased (4.6 ± 2.6 ng/ml to 3.0 ± 1.9 ng/ml, $p = 0.005$) during this period. In controls, TGF-β remained unchanged with increasing age. Contrary to results in patients with a small RV and controls, inflammatory factors increased with increasing follow-up duration in patients with a large RV, while cardiomyocyte growth factors and TIMP 2 decreased.
Conclusions: We have linked the process of progressive RV dilatation with declining serum levels of TGF-β in patients after TOF repair. Lower TGF-β levels coincided with increased markers of immune activation and reduced markers of cardiac protection, making this subgroup of patients more susceptible to adverse RV remodeling and eventually RV failure.
A better understanding of the molecular biology that governs this process might aid in earlier recognition of patients at risk for heart failure and better treatment options.

O5-4

Abnormal vascular load relates to impaired relaxation in patients with Fontan circulation

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Introduction: Today, Fontan procedure is the last staged operation for children who can not reach to two ventricle repair. To modify operation method, mortality rate in patients with a single ventricle are dramatically reduced. Therefore, a lot of Fontan patients are entering adulthood, and these patients are facing an uncertain future. Concretely, patients are prone to development of late –onset supraventricular tachyarrhythmias, heart failure, progressive rise of their pulmonary vascular resistances, protein losing enteropathy, hepatic dysfunction, pulmonary arteriovenous malformation, and so on.Previous studies consistently demonstrated delayed relaxation in Fontan ventricle regardless of its systolic function. Mechanisms of delayed relaxation remain poorly understood. Relaxation is prolonged by acute increases in animals and human studies.(Hypothesis) We hypothesized that abnormal loading, which also demonstrated previously, has an important mechanism of delayed relaxation in Fontan patients too. (Methods) Fifty-five patients with fontan circulation(Age5.2 ± 3.9, 0.5-15years) underwent cardiac catheterization to check for residual pulmonary stenosis indicated by echocardiography.Impedance moduli were computed from Fourier components of pressure and flow data.

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Results: Time constant of relaxation(tau) of hybrid logistic model was significantly increased in Fontan patients than in controls with small ventricular septal defects(21.9 vs 17.8, $P < 0.01$). Resistances were in proportion to tau, and strong correlation($r = 0.71$). Reflect wave time, which were derived from fourth derivative of aorta pressure wave, were inversely with tau.($r = 0.35$) Characteristic impedance were in proportion to tau, but weak correlation($r = 0.49$).Compliances mean, PWV and LVP max did not have significant correlation.
Multivariate regression analysis have done. Resistance and wave reflection were correlated with prolonged tau. ($p < 0.05$)
Conclusion: Abnormal ventricular systolic-afterload interaction, abnormal afterload profile has detrimental effects on ventricular relaxation in fontan circulation. Thus afterload modulation may be benefical to improve long-term prognosis of patients with this circulation.

O5-5
Staged Fontan procedure ameliorates Ventricular hyper-fibrogenesis in the patients with single ventricular circulation

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Background: Preserving ventricular function before and after Fontan procedure is a key element in establishing successful Fontan circulation. We tested the hypotheses that 1) volume overload and/or cyanosis generally observed in the single ventricle (SV) circulation before Fontan surgery is associated with ventricular hyper-fibrogenesis and 2) Fontan procedure effectively ameliorates ventricular fibrogenesis, but sustained hyper-fibrogenesis in the Fontan ventricle is associated with ventricular diastolic dysfunction.
Methods: Serum levels of amino-terminal propeptide of type III procollagen (PIIIP), a marker of fibrogenesis, were measured in 63 patients with a single ventricle (SV) and in 111 healthy children (control). The children with SV were divided into three groups according to clinically relevant stage: (1) Unoperated children or children who underwent first-stage repair (Blalock-Taussig shunt or pulmonary artery banding: BTS/PAB group, $n = 63$). (2) Children who underwent second-stage palliation (Glenn group, $n = 59$). (3) Children who underwent Fontan surgery (Fontan group, $n = 36$). For the children with SV, serum levels of Renin, Angiotensin, and Aldosterone were also evaluated.
Results: The PIIIP levels decreased significantly after the Glenn procedure, and further decreased after the Fontan procedure, although the level was still significantly higher than that of the control (BTS/PAB group: 5.3 ± 2.1, Glenn group: 1.8 ± 1.0, Fontan group: 1.5 ± 0.5, Control: 1.1 ± 0.3 U/ml). Increased ventricular volume load assessed by pulmonary to systemic flow ratio (Qp/Qs) was significantly associated with increased PIIIP levels in children with BTS/PAB ($r = 0.674$, $p < 0.01$), while PIIIP levels in the Glenn group correlated significantly with the severity of cyanosis ($r = 0.459$, $p < 0.01$). In the Fontan group, high PIIIP levels correlated significantly with increased ventricular diastolic stiffness assessed by end-diastolic pressure-area relationship ($r = 0.44$, $p < 0.05$). The serum levels of Aldosterone remarkably decrease in parallel with the surgical stage, same as the trends of PIIIP. After the Fontan completion, serum levels of Renin, angiotensin and aldosterone were significantly correlated with that of PIIIP.
Conclusions: Our results suggest ventricular hyper-fibrogenesis in patients with SV circulation. Staged-Fontan procedure can ameliorate this process, and additional therapies aiming at anti-fibrogenesis, such as inhibition of Renin-Angiotensin-Aldosterone system, may be additively effective in improving the patient's prognosis after the Fontan procedure.

O5-6
Fetal laterality disturbance: Single centre experience of 227 cases spanning three decades

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Introduction: Disturbances of laterality in the fetus represent some of the most complex forms of congenital heart disease. This is a heterogenous group associated with a spectrum of cardiac and extracardiac abnormalities. In many cases single ventricle palliation is the only management option after birth. The cumulative effect of associated lesions means that many do not achieve Fontan completion long-term. We report a large prenatal series of right (RAI) and left atrial isomerism (LAI), describe their associated malformations and review outcome.
Methods: Retrospective search of our fetal cardiology database between 1980 and 2010 for cases of RAI and LAI. Fetal and postnatal management and long-term outcome were reviewed.
Results: A total of 230 cases of laterality disturbance were identified. Three cases were excluded as outcome data was not available. There were 145 cases of LAI of which 32 patients (22%) remain alive, 6 have a Fontan circulation and two have undergone cardiac transplantation. In 76 (52%) cases parents opted for termination of pregnancy (TOP), 13 pregnancies resulted in intra-uterine death (IUD), 18 patients died in the neonatal period (NND) and a further 6 died in childhood (INFD). Eighty-two patients had RAI of which 14 remain alive (17%) with 7 reaching Fontan circulation. In this group 43 (52%) opted for TOP, 5 resulted in IUD, there were 9 NND and 11 INFD. The number of parents opting for TOP has fallen from 66% in the first decade (1980-1990) to 60% in the second decade (1990-2000) and 42% in the recent cohort (2000-2010). Outcome by type and era is presented in Table 1. Examining the data on an 'intention to treat' basis reveals survival of 0%, 50% and 59% for LAI and 0%, 36% and 37% for RAI across the three decades respectively.
Conclusion: Survival for those with LAI has improved with many undergoing biventricular repair. Congenital heart block remains a concern in this group resulting in IUD and NND. Prognosis remains poor in those with RAI with high attrition rates in those with single ventricle physiology.

O6-1
Multiparametric assessment of the Right Ventricle by echocardiography in adult patients with repaired tetralogy of Fallot undergoing pulmonary valve replacement: a comparative study with MRI.

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Purpose: Evaluation of the right ventricle (RV) using transthoracic echocardiography is challenging in patients with congenital heart disease affecting the right ventricular outflow tract such as Tetralogy of Fallot (TOF). MRI is commonly used to determine the best timing for pulmonary valve replacement (PVR) but accessibility remains limited. The objective of this study was to evaluate the feasibility and the accuracy of a multiparametric echographic approach including 2D strain and 3D for RV volumes and function assessment, in comparison with MRI.
Methods and results: we performed a complete echocardiographic study including 2D parameters (TAPSE, S' TDI, Tei indice,

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index Fractional Area Change (FAC)), 2D strain and 3D and unsedated cardiac MRI in 26 consecutive patients with repaired TOF before pulmonary valve replacement and one year after surgery. Correlation between echography and MRI RVEF was poor for TAPSE, S' TDI and 2D strain but good for FAC ($r = 0.70$, $p < 0.01$and $r = 0.68$, $p < 0.01$, before and after PVR respectively) and 3D assessment ($r = 0.96$, $p < 0.01$ and $r = 0.98$, $p < 0.01$ before and after PVR respectively). Despite RV volume underestimation by 3D echography, correlation for RV volume assessment between 3D analysis and MRI was excellent in both pre and post-operative assessment ($r = 0.88$, $p < 0.01$ and $r = 0.91$, $p < 0.01$ respectively for RV end-diastolic volume; $r = 0.92$, $p < 0.01$ and $r = 0.95$, $p < 0.01$ respectively for RV end-systolic volume).
Conclusion: Global approach of RV function using 2D (FAC) or (3D) parameters seems reliable in patients with repaired TOF. The commonly used TAPSE and S'TDI focused on segmental analysis of RV inflow are less sensitive probably because RV inflow is less affected by RV remodelling related to initial surgical repair.

O6-2

Determinants and Clinical Significance of Flow via the Fenestration in the Fontan Pathway: A Multimodality Study

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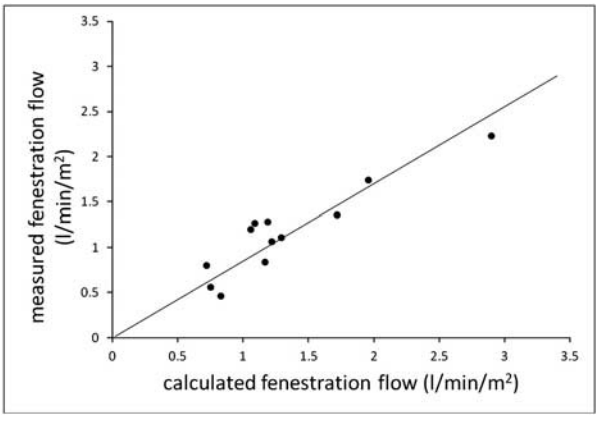
Introduction: The use of a fenestration in the Fontan pathway remains controversial, partly because its hemodynamic effects and clinical consequences are insufficiently understood. The objective of this study was to quantify the magnitude of fenestration flow and to characterize its hemodynamic determinants approximately 1 year after surgery.
Methods: Twenty three patients with a fenestrated extracardiac conduit prospectively underwent investigation by cardiac magnetic resonance (CMR), echocardiography, and invasive manometry under the same general anesthetic 12 ± 4 months after Fontan surgery. Fenestration flow was determined using phase contrast CMR either by subtracting flow in the Fontan pathway above the fenestration from Fontan flow below the fenestration or by direct measurement (12 patients, Figure).



Results: Measured and calculated fenestration flows showed an excellent agreement ($r = 0.92$, $p < 0.0001$, Figure). Fenestration flow constituted a mean of $31 \pm 12\%$ (range 8-50%) of ventricular preload. It was associated with a lower Qp/Qs ($r = -0.64$, $p = 0.001$) and oxygen saturation ($r = -0.74$, $p < 0.0001$). Fenestration flow volume was correlated with pulmonary vascular resistance ($r = 0.45$, p-0.04) and markers of ventricular diastolic function (early diastolic strain rate $r = 0.57$, $p = 0.008$ and ventricular untwist rate $r = 0.54$, $p = 0.02$).
In 14 patients (61%) all of the net inferior vena cava flow and part of the superior vena cava flow were diverted into the systemic atrium and did not reach the lungs. The magnitude of contribution of fenestration flow to ventricular preload was the most important predictor of ability to close the fenestration.
Conclusions: Fenestration flow can be measured accurately with CMR. In most patients the fenestration delivers most or all of the inferior vena cava flow to the systemic circulation, and in two thirds of patients some of the superior vena cava flow is also diverted through the fenestration. Therefore, the pulmonary blood flow increases only minimally and sometimes even decreases after the fenestrated Fontan operation. The amount of fenestration flow is related both to the pulmonary vascular resistance and systemic ventricular diastolic ventricular function, and may be used to predict hemodynamic suitability for fenestration closure.

O6-3

Assessment of intraatrial lateral tunnel anatomy and venous blood flow in children with hypoplastic left heart syndrome in Fontan circulation

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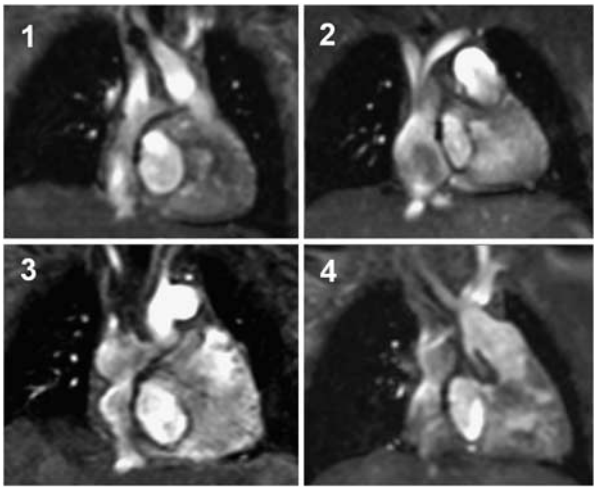
Introduction: In patients (pts) with hypoplastic left heart syndrome (HLHS) post completion of the total cavopulmonary connection (TCPC) with an intraatrial lateral tunnel, deviations of the tunnel from an ideal tubular shape are common. However, little is known about frequency and potential adverse effects of such shape deviations. We sought to analyze tunnel anatomy, dimensions and blood flow with magnetic resonance imaging (MRI).
Methods: Sixty-one HLHS pts (mean age 6.4 ± 2.7 years) underwent 3.0-T MRI with gradient-echo cine sequences, 2D- and flow-sensitive 3D-phase-contrast MRI. We analyzed anatomy, diameters, cross-sectional areas and volumes of the tunnel. Tunnel blood flow was measured at the level below the connection of the inferior vena cava (IVC) with the pulmonary arteries.



Results: 23 pts had a tubular-shaped tunnel (Figure 1). In 28 pts bulging and/or narrowing at different locations of the tunnel was

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present (Figure 2-4). In 10 pts a classification was not possible because of artifacts from implanted devices. Cross-sectional areas, tunnel volumes, the mean blood flow and the mean and maximal flow velocity were not significant different between pts with a tubular tunnel and pts with shape deviations of the tunnel. We found a relation between the normalized tunnel volume and age ($r = 0.44$; $p = 0.002$), body surface area ($r = 0.42$; $p = 0.005$) and time after TCPC ($r = 0.42$; $p = 0.001$). The mean tunnel blood flow correlated with age ($r = 0.75$; $p = 0.001$) and body surface area ($r = 0.83$; $p < 1.0e-4$). Flow-sensitive 3D-phase-contrast MRI showed retrograde flow at the junction between the IVC and the tunnel and a non-linear tunnel blood flow (e.g. reflux, vortices) below the fenestration.
Conclusion: 1) The volume and the mean blood flow of the intraatrial lateral tunnel correlated with age and body surface area of HLHS pts in Fontan circulation, suggesting that the conduit capacity of the tunnel adjusts to body growth, unlike an extracardiac tube. 2) Flow-sensitive 3D phase-contrast MRI showed a non-linear blood flow in the lower part of the tunnel. Follow-up MRIs are needed to detect long term effects of irregular tunnel shapes on flow dynamics.

O6-4
Abnormal blood flow pattern in the aorta and pulmonary trunk of patients with Transposition of the Great Arteries operated with Atrial Baffle Switch

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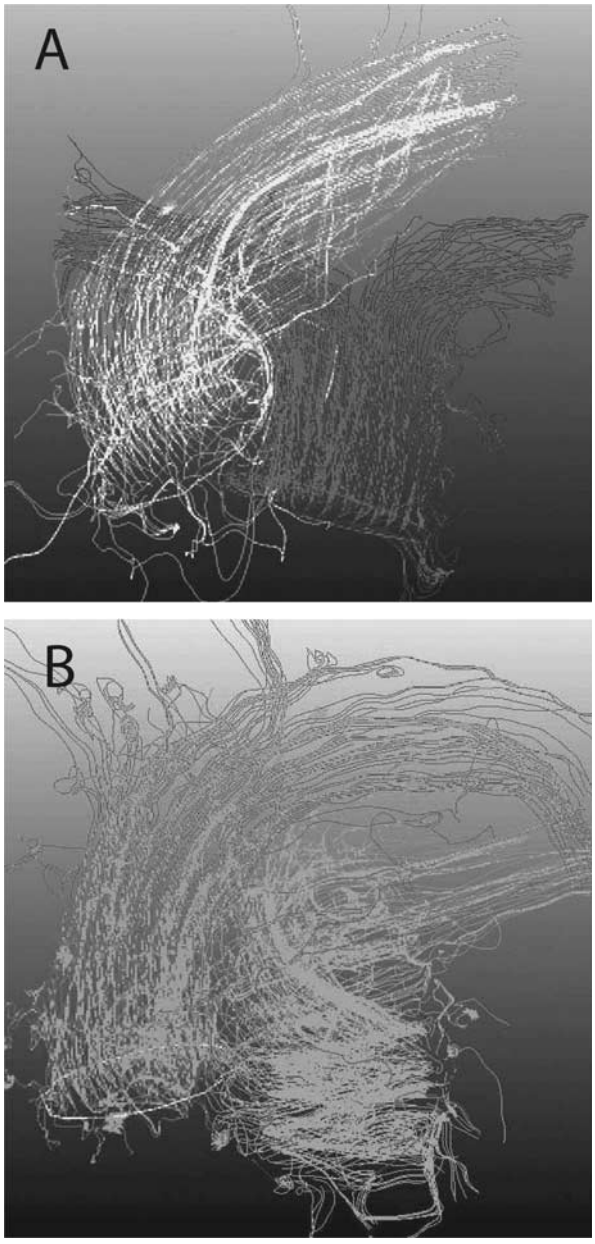
Background: Patients with transposition of the great arteries (TGA) after atrial baffle switch operation show differences in ventricular torsion and outflow tract geometry compared to healthy volunteers and TGA patients after arterial switch operation. We sought to investigate if these differences in cardio-mechanics translate into abnormal blood flow patterns in the pulmonary trunk and the aorta.
Methods: Blood flow patterns were assessed with flow-sensitive four-dimensional velocity-encoded magnetic resonance imaging (4D VEC MRI), using a 1.5T Phillips MRI system. Measurements were made in the pulmonary trunk and the aorta of TGA patients after atrial baffle switch operation ($n = 10$) and compared to healthy volunteers ($n = 7$) and TGA patients after arterial switch operation ($n = 7$). Blood flow was analyzed for vortex formation and helical blood flow pattern using custom-made software.
Results: There were clear differences in blood flow patterns between TGA patients after atrial baffle switch operation and healthy volunteers and TGA patients after arterial switch operation in both the pulmonary trunk and the aorta. In healthy volunteers and TGA patients after arterial switch operation, flow was laminar, parabolic in the pulmonary trunk and showed left helical flow pattern in the aorta. In TGA patients after atrial baffle switch operation we observed opposite flow patterns with predominant parabolic flow in the aorta but helical flow and vortex formation in the pulmonary trunk. Helical blood flow pattern was present in the ascending aorta in healthy volunteers and patients after arterial switch and was less pronounced in patients after atrial baffle switch ($p < 0.05$).
The figure shows streamlines of the ascending aorta and pulmonary trunk in a healthy volunteer (A) and a representative patient with TGA after atrial baffle switch operation (B).
Conclusions: There are abnormal flow profiles in the aorta and pulmonary trunk in TGA patients after atrial baffle switch operation compared to healthy volunteers and TGA patients after arterial switch operation. The data of this study provide evidence that differences in left and right ventricular cardiomechanics directly translate into different flow patterns.



O6-5
The role of Dipyridamole stress echocardiography in the diagnosis of coronary pathology after the arterial switch operation for Transposition of the great arteries

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Objectives: To evaluate Dipyridamole stress echocardiography (DSE) as a diagnostic tool in paediatric patients with Transposition of the great arteries (TGA) treated with arterial switch operation (ASO), and to test its accuracy in screening myocardial ischemia.

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Methods: One-hundred consecutive patients (mean age 5 ± 3,8 years, range 3 months -21 years; mean weight 21 ± 13 Kg, range 5,6-89 Kg) with TGA (71 simple TGA, 25 TGA and VSD and 11 DORV-Taussig Bing type) and ASO, underwent DSE and selective coronary angiography, 4,9 ± 3,6 years (range 3 months-21 years) after the intervention. Before the test, the history of perioperative ischemic events and the clinical/instrumental signs of ischemia at the time of the study were noted (pre-test evaluation). DSE was performed in the catheterization laboratory under general anaesthesia, and once it was completed was followed by coronary angiography.
Results: DSE and coronary angiography were well tolerated without any complication. Dipyridamole provoked myocardial ischemia in 4% of patients and coronary stenosis were demonstrated in 5% of subjects. Two patients with coronary stenosis and no dipyridamole-inducible ischemia showed a well developed collateral circulation. In all pts with negative pre-test evaluation, no stress-ischemia and no angiographic coronary stenosis were detected.
Conclusion: Dipyridamole stress echocardiography is feasible, safe and is a reliable marker for ischemia in children with Transposition of the great arteries after anatomical correction. It is unable to detect coronary abnormalities when collateral circulation fully compensates the coronary stenosis. Stress echocardiography is an important adjunctive tool in the management of patients with coronary abnormalities after the arterial switch operation.

O6-6
Temporal and spatial accuracy of 3D real time echocardiography in the neonatal and pediatric setting–Validation studies using small moving and pulsative phantoms

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Objective: Purpose of the study was to verify the accuracy of RT-3DE for the assessment of small cardiac volumes and high frequencies similar to newborn and pediatric hearts.
Material & Methods: For the assessment of spatial and temporal resolution of 3D-matrix transducer, customized, small test phantoms were moved linear in a water basin. The accuracy of 3D-volume calculations and measurement of time periods was tested by pulsatile balloon-phantoms. The calibrated phantoms were controller-operated to ascertain the exact end-diastolic and end-systolic volume, cardiac cycle length, isovolumetric phases and volume time curves. Artificial cardiac cycles with different volumes (5-17 ml) and variably frequencies (60-150/min) were acquired by a commercial RT-3DE-System (iE33, Philips, X7-2, X5-1). 3D-volumes were calculated with two semi-quantitative software algorithms (QLab, Philips and LV Analysis, TomTec).
Results: Spatial and temporal resolution: 3D-scans of moved test objects showed a spatial distortion which is attributed to the inherent sequential scanning mode of the matrix transducer. Different parts of the test objects are not scanned simultaneous; they are acquired with a time delay between 22-45.2 ms, which is equally to the time duration of a single 3D-frame.
Assessment of 3D-volumetry: 3D-data sets did not include the complete cardiac cycle, 15% of the end-diastolic period were missing (Fig 1). Prolonged isovolumetric phases were not recognized. Volumes were underestimated.
Conclusion: RT-3DE is an established tool to assess ventricular function and volume, but different segments of moving objects are scanned with delay. This can result in significant misinterpretation of segmental synchrony and averaging of volumes. Quantification of volume time curves by software systems follows strict algorithms which are adapted to a normal cardiac cycle. If time flow (prolonged isovolumetric phases) or geometry deviate from this, imprecise calculations of the volume changes over time can be caused. Clinicians have to consider these limitations, when using 3D-RTE for volumetry, synchronicity analysis or assessment of valvular movement in fast beating and small hearts.

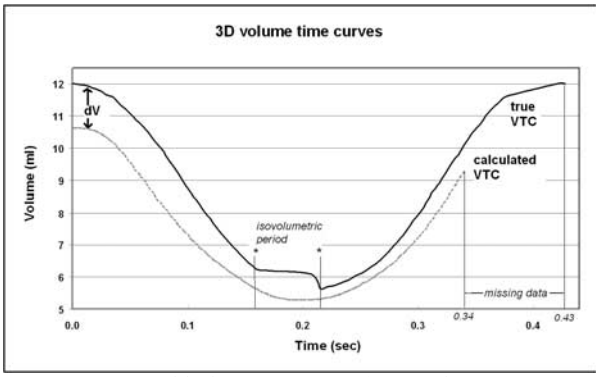

Fig: True (upper, black) vrs. measured 3D-volume time curve (dotted, grey): calculated volumes are smaller than true volumes (dV), cardiac cycle length is incomplete (0.34 vrs 0.43 s; "missing data"), prolonged isovolumetric periods are not recorded (*)

O6-7
Tissue motion annular displacement (TMAD) of the mitral valve using two dimensional speckle tracking echocardiography predicts left ventricular ejection fraction in normal children.

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Introduction: The gold standard for determining left ventricular ejection fraction is cardiac magnetic resonance imaging (CMRI). This is time consuming, costly and often requires general anaesthesia in children. Other echocardiographic parameters for determining ejection fraction such as MMode are operator dependant and often inaccurate. Assessment of the displacement of the mitral valve annulus using two dimensional speckle tracking echocardiography potentially provides an accurate and simple method of determining left ventricular ejection fraction in children.
Method: A total of 70 children aged 9 years were assessed using cardiac magnetic resonance imaging and two dimensional trans-thoracic echocardiography. These children are part of the Southampton Women's survey and have no history of cardiovascular disease. A total of 48 patients were included in the study, exclusions were for poor image quality and incorrect image acquisition. All echocardiograms were analysed using the Tissue motion annular displacement (TMAD) feature of the cardiac motion quantification (CMQ) plug in. (Phillips QLAB version 9.0). TMAD is based on speckle tracking technology and calculates the valvular annular displacement over time. Three points on the apical four chamber view of the left ventricle were identified, the lateral and medial mitral valve annulus and the apex, and tracked using this feature. The mid point displacement of the mitral valve was automatically calculated and using a quadratic formula a predicted ejection fraction was calculated. This predicted ejection fraction was compared with CMRI and MMode derived ejection fractions.
Results: The ejection fraction from CMRI (64.5 +/- 4.6) was not significantly different from that derived from TMAD mid point

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(60.9 +/- 2.7) or that derived from Mmode (61.9 +/- 7). The TMAD mid point correlated strongly with the CMRI ejection fraction ($r = 0.69$ $p < 0.001$) as did the predicted ejection fraction ($r = 0.67$ $p < 0.001$). The MMode ejection fraction showed a poor linear correlation with both CMRI and TMAD values ($r = 0.33$ and 0.02).
Conclusion: TMAD of the mitral valve is a simple, effective and highly reproducible method of assessing left ventricular function in normal children. It shows a strong linear correlation with CMRI derived ejection fraction and is superior to MMode derived ejection fraction.

O6-8
Evaluation of complex congenital heart disease and associated complications in newborns, infants and small children using multi-detector CT (MDCT) – An 11-years-experience at 5 centers

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Introduction: MDCT with multiplanar and 3D-reconstructions has become an important first-line imaging tool in diagnosis of complex congenital heart (CHD) and airway diseases in children. Aim was to assess the diagnostic value of MDCT in newborns and infants at 5 centers of 3 countries.
Materials and Methods: Between 2000 and 2011, 281 patients (mean age: 7 ± 7 months, range: 6 hr to 2 y) were examed using varying scanners (4 up to 256 slices, resolution isotrope 0.4-1.25 mm; scan-time 2-20 s, 80-120 kVp, 60-80 mA) under ventilation or free breathing. Diagnoses: in-stent stenosis ($n = 72$), pulmonary stenosis (48) and atresia (32), arterial rings and slings (32), aortic arch anomalies (21), bronchoscopy revealed stenosis (24), abnormal pulmonary venous return (17) and others (35). The image quality was rated using a 5-point score. Image findings were correlated to ECHO, conventional angiography, bronchoscopy, and intraoperativ findings. The effects of dose on image quality were also evaluated, retrospectively.
Results: MDCT data were almost free of cardiac and respiratory motion. Images were scored in >95% of all cases as excellent or good, showing a significant improving with increasing number of detectors. Radiation exposure was mostly less than 2 mSv (range 0.3 to 3.2). High radiation exposure settings did not improve image quality. Morphology and topography could be assessed exactly and the final diagnosis was allowed. Even smallest vessels (diameter<1 mm) could be identified and excel¬lently visualized. 83% (232/281) of all patients had benefited from MDCT: Catheter was neither necessary to perform treatment planning nor to exclude an anomaly, or radiation doses and sedation time due to interventional procedures could be reduced markedly.
Conclusions: A 3D submillimeter evaluation of CHD can be achieved routinely in a matter of seconds with little motion artefacts, without general anesthesia and with low radiation exposure. The use of MDCT may result in a net decrease in overall radiation decreasing the number of diagnostic catherizations. MDCT can now be regarded as the modality of choice as a minimally invasive, robust, and accurate technique. This advance should have the greatest impact in the smallest, youngest, and most critically ill children.

O7-1
Efficacy and impact of a nation-wide prenatal cardiac screening program

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Introduction: The aim of the study was to describe the evolving efficacy of a prenatal ultrasound screening program for congenital heart disease (CHD) and its impact on pregnancy management and outcome.
Methods: All pregnant women the Czech Republic undergo fetal ultrasound scanning provided by obstetricians trained by paediatric cardiologists. Every prenatal CHD is confirmed in a single tertiary referral centre and/or by a post mortem evaluation, which is obligatory in all terminated pregnancies. A prenatal registry was created of all foetal patients with detected CHD over a 25-year period from 1986 to 2010.
Results: Most common lesions detected prenatally were atrioventricular septal defect ($N = 384$, 15.2%), hypoplastic left heart syndrome ($N = 379$, 15.0%), ventricular septal defect ($N = 230$, 9.1%) and double outlet right ventricle ($N = 220$, 8.7%). As compared to estimated postnatal incidence at given birth rate detection of all/critical heart disease increased significantly between 1986 -1999 and 2000-2010 from 7.3/20.8 to 29.4/76.5% ($p < 0.001$ for both). In the latter period, the detection rate reached 95.1% for hypoplastic left heart syndrome and 40% for transposition of great arteries. Families opted for early termination in 1403/2528 foetuses with prenatally detected CHD (55.5%). Extracardiac malformations were observed in 705/1403 (51%) and the aneuploidy rate was 24.2% (338 cases, trisomy 21/18 in 57/24%). Of the respective prenatally detected lesions the most frequently terminated was the hypoplastic left heart syndrome HLH (22.7%), atrioventricular septal defect AVSD (15.8%), double outlet right ventricle DORV (12.3%) and pulmonary atresia PA (7.7%) leading to a significant decrease of expected postnatal incidence (HLH from 3.4 to 1.3%, AVSD from 4 to 2.6%, DORV from 1.4% to 0.3% and PA form 2.2 to 1.5%, resp.). From the continuing pregnancies, 73 (6.7%) of 1125 foetuses died in utero, and 1052 (93.3%) babies were born alive.
Conclusions: The nation-wide prenatal ultrasound screening program showed an increasing efficacy for the detection of congenital heart disease. High rate of pregnancy termination leads to a significant change of postnatal incidence in selected lesions.

O7-2
The differential insertion of the atrioventricular valves: a plane-dependent echocardiographic feature for normal fetal hearts

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Introduction: The differential insertion of the atrioventricular valves (DIAVV) is an echocardiographic feature in a four-chamber view of structural normal fetal hearts. DIAVV is an ultrasound image of the more apical attachment of the tricuspid valve (TV) to the atrioventricular septum compared to the mitral valve (MV). A linear insertion of the atrioventricular valves (LIAVV) is suggested as a marker for atrioventricular septal defects (AVSDs). The aim of this study is to evaluate the anatomical substratum of DIAVV and LIAVV in normal hearts and AVSDs.

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Methods: Serial sections of 17 normal human hearts, ranging from 10 to 36 weeks gestation, were studied with various histological and immunohistochemical tissue markers with special attention to the extent and position of the fibrous skeleton and the attachment of the leaflets of the AV valves. For each trimester a 3D AMIRA reconstruction was made. Sectional planes equal to the echocardiographic four-chamber view were studied. In addition, this anatomical region was compared to a case with AVSD.
Results: In normal fetal hearts the DIAVV is visible in the four-chamber plane just caudal to the aortic outflow tract. In this plane the offset of the TV is located more apical than the MV. In a more caudal four-chamber plane there is a lack of the DIAVV, resulting in a linear appearance of the offset of the AV valves. In a trisomy 21 fetus with an AVSD, a linear appearance (LIAVV) was observed in the four-chamber plane. However, more caudally to the aortic outflow tract a differential insertion (DIAVV) was found.
Conclusions: In structural normal fetal hearts and in a heart with an AVSD both DIAVV and LIAVV can be found dependent on the plane where the four-chamber view is visualised. For demonstration of the DIAVV it is important to obtain the four-chamber view just caudal of the aortic outflow tract. The knowledge that such an accurate positioning of the four-chamber view is necessary, explains differences in reference values described in the literature. Therefore, the DIAVV is a useful echocardiographic tool, however, caution should be exercised when interpreting the DIAVV as characteristic for a normal heart.

O7-3
Congenital heart defects in twin pregnancies

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Background: The relative risk for congenital heart defects (CHD) is increased in monochorionic twins. The respective contribution of genetics of cardiac development and environmental risk factors such as twining per se are scarcely known.
Aim: To analyze CHDs in twin pregnancies in which one or two foetuses were diagnosed to have a CHD and to identify the role of chorionicity and amnionicity on CHD type and concordance between twins.
Methods: 226 consecutive twin pregnancies with one or two foetuses with CHD were reviewed. Chorionicity, amnionicity, twin-twin transfusion syndrome (TTTS), cardiac phenotypes, anatomical/CHD type (7 groups of CHD), concordance between twins and outcomes were analyzed.
Results: Pregnancies were bichorionic-biamniotic (BCBA) in 103 cases and monochorionic in 112 (biamniotic 92-MCBA, monoamniotic 20-MCMA) and the remaining 11 were unknown. Overall the 2 foetuses were affected in 35 cases (15.4%) with the two CHDs belonging to the same group in 23 (65.7%). The two foetuses had a CHD in 35% of MCMA, 14.1% of MCBA and in 10.7% of BCBA. The most frequent defects were conotruncal defects (n = 71) with concordance in 13/71 (8/20 MCBA, 3/5 MCMA). For other groups concordance in all pregnancies, in MCBA and in MCMA were respectively : right outflow tract obstructions 5/48, 0/33, 0/0; obstructive left heart diseases 7/42, 2/13, 1/1; laterality defects 2/36, 0/6, 2/12; ventricular septal defects 5/19, 1/9, 0/0; atrioventricular septal defects 3/9, 1/2, 0/0; functionally univentricular heart 1/15, 0/5, 0/1. MCBA pregnancies were more frequent in right outflow tract obstructions (33/48; 69%- $p < 0.01$) and discordance between twins was constant and was associated with proven TTTS in 22/33. MCMA were more frequent in laterality defects (12/36; 33%- $p < 0.01$) and discordance between twin of was observed in 10/12. The outcome of pregnancy was: termination of pregnancies 47 foetuses, intrauterine foetal deaths 20 foetuses, neonatal death 24/387 live births. 116 patients underwent cardiac surgery within the first year of life with 13% mortality.
Conclusion: Concordance for CHD between twins is low even in monochorionic-monozygotic twins. Discordance in monochorionic twins might be related to twining per se for laterality defects in MCMA and pulmonary stenosis in MCBA-TTTS.

O7-4
Cardiovascular function after successful treatment of intrauterine twin-to-twin transfusion syndrome – a 10-year long-term follow-up

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Introduction: TTTS is associated with a high incidence of fetal and neonatal cardiovascular morbidity including myocardial hypertrophy and cardiomegaly; hypertension and increased vascular stiffness are attributed to pathological vascular fetal programming. Reports of long term cardiac function have focussed only on survivors after serial amnioreduction.
Objective: To assess cardiac and vascular function of survivors 10 years after intrauterine laser coagulation (LC) of severe TTTS (median gestational age at LC: 20 + 5 wks; 3 hydrops, 6 severe tricuspid, 3 severe mitral regurgitation)
Methods: 31 surviving twin pairs with a median age of 9.99 [8.58-11.42] years and an age matched control group of 15 healthy probands were assessed by 2D, Doppler-, 3D-echo and speckle tracking including measurement of blood pressure and vascular stiffness (PWV, augmentation index AI).

Selection of parameter examined

Parameter (mean ± SD)	Donor	Recipient	Control	p-value
N=	31	31	15	
RR sys (mmHg)	109 ± 7	113 ± 11	109 ± 6	0,161
RR dia (mmHg)	60 ± 6	62 ± 11	62 ± 5	0,669
MAD (mmHg)	78 ± 6	80 ± 9	79 ± 5	0,443
AI % Sphygmocor	-16,7 ± 1,0	-22,1 ± 0,6	-20,4 ± 0,4	0,443
PWV Sphyg.(m/s)	4,37 ± 0.5	4,72 ± 1,7	4,11 ± 0,1	0,484
Tei LV	0,32	0,35	0,34	0,45
Tei RV	0,31	0,35	0,37	0,86

Results: Cardiac function parameter showed no significant differences between donor and recipient twin. There was no significent difference between the cohorts regarding hight corrected blood pressure, PWV and AI (table).
Conclusions: Follow-up until the age of 10 years after successful intrauterine LC prove that both donors and recipients have normal cardiac and vascular function in the longer term. This underlines the reversibility of even severe cardiovascular intrauterine dysfunction by causative laser coagulation.

O7-5
Features and outcomes of fetuses with tricuspid atresia

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Background: Tricuspid atresia (TrA) has variable anatomic features and a rather severe prognosis, requiring a univentricular correction. *Objectives:* Our aims were to analyse retrospectively the features and outcomes of cases diagnosed in our Center with this lesion. *Material and methods:* Out of around 1250 fetal cases with CHD studied in our Centre between Jan.1998 and Dec. 2011, 55 (4.4%) were found to have TrA, at 16-34 weeks' gestation (w.g.) The characteristics and outcomes of 53 cases with known follow-up were retrospectively analysed.
Results: 1/ Characteristics: Four fetuses were twins; 23 fetuses (43.4%) had associated pulmonary stenosis (PS); 9 (17%) had pulmonary atresia (PAtr), 2 with sinusoids; 14 (26.4%) had associated one or more ventricular septal defects (VSD), one with coarctation of aorta; 7 (13.2%) had transposition of great arteries (TGA) with VSD, 1 with interrupted aortic arch. One fetus had a minor chromosomal anomaly–inversion (2) p11.2q13; 8 had associated extracardiac anomalies (ECA). Three fetuses had heart failure already at presentation, due to restrictive foramen ovale.
2/ Outcomes: Sixteen fetuses (30.2%) were terminated, 5 with ECA. Out of 37 fetuses that continued pregnancy, one died in utero at 24 w.g., 4 died shortly after birth (2 in heart failure, 1 in severe cyanosis and 1 fetus with PAtr and sinusoids had cardiac arrest). The remaining 32 cases were operated, all with palliative procedures (shunt or banding pulmonary artery), 11 underwent Glenn operation and 6 the Fontan procedure. A total of 5/32 cases (15.6%) died postoperatively, 3 after shunt operation (1 with PAtr and sinusoids), 1 (with interrupted aortic arch) after the Norwood 1st step and 1 after Glenn operation. Total intrauterine and postnatal mortality was 10/37 cases (27%). The remaining infants are alive, a half of them on medications, at 2-12 years.
Conclusions: Despite an improvement in surgical results and in perinatal management the outcomes of TrA remain still poor, almost one third dying postoperatively or spontaneously. Negative prognostic factors were restrictive foramen ovale, heart failure and PAtr with sinusoids.

O7-6
Hypoplastic Left Heart Syndrome with Restrictive Atrial Septum: Echocardiographic Criteria for the Need of In-Utero Catheter Intervention and First Results of Human Fetal Atrial Stenting

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Background: Fetal pulmonary venous (PV) Doppler flow has been correlated with the severity of atrial septal restriction and the need for emergent atrial septostomy in newborns (NEAS) with hypoplastic left heart syndrome (HLHS). Despite NEAS, many infants with severely restrictive atrial septum (RAS) will eventually die from persistent severe pulmonary vascular disease.
Methods: To develop selection criteria for fetal intervention, we reviewed 67 HLHS fetuses with active postnatal care and studied the correlation between PV Doppler, NEAS at day 1, and survival to infancy. PV Doppler assessments included S-, D- and A-wave peak velocities (cm/s), A-wave duration (ms), and forward/reverse PV VTI ratios. Moreover, the outcome of four recent RAS fetuses with in-utero atrial stenting is reported.
Results: Four (6%) of the 67 cases without fetal intervention required NEAS. Two other fetuses with the same PV Doppler findings developed fetal hydrops and died during the last trimester. The table indicates the median (range) pre-delivery PV Doppler indices of HLHS cases with vs. without NEAS:

Parameters	NEAS (n = 4)		No NEAS (n = 63)		P-values
Fetal age (last echo; wks)	36	(35-38)	35	(28-39)	NS
S-wave (cm/s)	48	(25-56)	43	(15-91)	NS
D-wave (cm/s)	0	(0-11)	19	(8-52)	NS
A-wave (cm/s)	-43	(-19-57)	-20	(-1-72)	0.006
A-wave duration (ms)	107	(90-113)	65	(30-88)	<0.0001
Forward/reverse PV VTI	1.5	(1.3-2.3)	8.1	(1-50)	<0.0001

All NEAS cases had a forward/reverse PV VTI<2.5 and PV A-wave duration >90 msec. This was the case in 0/63 (0%) cases without NEAS ($p = 0.0001$; sensitivity 100%; specificity: 100%). The need of NEAS was associated with 0% survival during the first 2 months due to severe pulmonary vascular disease, while 83% without NEAS were alive (hazard ratio 7.7; $p < 0.001$). Of 4 recent RAS cases with fetal atrial stenting at 27, 31, 32 and 35 weeks, 2 did not require NEAS, 4 underwent stage 1 surgery, and 3 are alive.
Conclusions: Two PV Doppler parameters (A-wave duration >90 ms; PV VTI <2.5) accurately predicted the need for NEAS, but none survived beyond early infancy. Fetal atrial septal stenting improved the postnatal outcome of our latest cases.

O7-7
Outcome after Fetal Diagnosis of Ebstein's Anomaly and Dysplasia of the Tricuspid Valve: A Single Center Experience

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Background: Ebstein's anomaly (EA) and tricuspid valve dysplasia (TVD) are serious fetal observations with survival rates to infancy below 25% in all major studies. There is little doubt that the worst disease spectrum is detected in utero, with many affected fetuses notable for significant cardiomegaly, severe tricuspid regurgitation and absent antegrade pulmonary blood flow. To survive, the fetal left ventricle needs to compensate for the dysfunctional right ventricle (RV) while postnatal survival rather depends on the RV to restore adequate pulmonary flow. Potential treatment strategies to improve outcomes include preterm delivery at signs of heart failure, and postnatal pharmacological lowering of the pulmonary vascular resistance and ligation of the arterial duct to avoid circular shunts. We reviewed the institutional experience with this entity.
Methods: Fetuses diagnosed with EA/TVD between 1999 and 2010 were identified from our prospectively acquired database. Echocardiography data were correlated with the established outcome of all cases.
Results: EA was diagnosed in 33 and TVD in 13. There were 4 pregnancy terminations with EA, 5 intrauterine deaths with EA, 8 neonatal deaths (mean age 10.1 days) and 29 survivors >1 months. Survival of the entire cohort was 80% at birth and 63% at 1 month, or 88% and 69%, respectively, on an intention-to-treat basis. Parameters associated with mortality (n = 15) included earlier gestational age at diagnosis (25.5 + 4.7 vs. 31.4 + 6.8

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weeks; p <0.005), larger right atrial area index (0.95 + 0.1 vs. 0.57 + 0.2; p < 0.0001), increased cardio-thoracic circumference ratio (0.6 + 0.1 vs. 0.49 + 0.1; p < 0.005), absent pulmonary forward flow (73% vs. 22%; p < 0.003) and severe tricuspid regurgitation (67% vs. 30%; p = 0.03). No associations were found between EA vs. TVD (11/33 vs. 4/13; p = 0.74), with fetal hydrops (33% vs. 27%; p = 0.73), additional cardiac abnormalities (13% vs. 33%; p = 0.27) and era of diagnosis (1999-2004: 4/12 (33%) vs. >2005: 11/30 (33%); p = 1.0).
Conclusion: Survival rate of prenatally diagnosed EA/TVD in our experience is significantly better than previously reported likely because less severe forms were more commonly detected in our catchment area during the past decade. Previously established predictors associated with adverse outcomes were confirmed in this study.

O7-8
Neonatal management of critical aortic stenosis after in-utero valvuloplasty: role of Ross-Konno operation

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Neonates with critical aortic stenosis (AS) after in-utero valvuloplasty usually present with borderline left ventricular size, function and varying degrees of endocardial fibroelastosis. The decision towards a biventricular circulation remains a challenge. The purpose of this study was to investigate the role of an early Ross-Konno (RK) operation on a biventricular outcome.
Between 12/2001 and 1/2012 we attempted 38 fetal aortic valvuloplasties in 35 fetuses (median GA 26 + 4 weeks; 21 + 4 to 32 + 1), 5 of these with advanced end stage heart failure and hydrops. The procedure was successful in 27/35 fetuses (77%), there were 3 IUDs in this group, so 24 children were live-born in 9 different centers in Austria, Germany, Poland, Denmark and Italy. According to the respective criteria of these centers, 18/24 (75%) newborns were managed towards a biventricular circulation either with aortic balloon dilation alone or followed by a RK operation. Balloon dilation alone was effective as a first line therapy in only 5/18 newborns, two of them needed later a RK operation at 1 and 4 years respectively, 3 patients died due to left heart failure and 2 had to go on to a Norwood procedure with 1 death. The remaining 8 patients underwent a neonatal RK at a median age of 14 days (7-19 days). One child died 37 days after successful surgery due to NEC. At a median follow-up of 2,3 years (0,3-6,8 years) all children are biventricular, with sufficient LV function and normal pulmonary artery pressures. Children born in centers offering neonatal RK surgery had higher probability to achieve a biventricular circulation, than children born in centers without this option: 10/16 (62,5%) vs 2/8 (25%)
Conclusions: In neonates with critical AS born after successful in-utero valvuloplasty and non-effective post-partum aortic valvuloplasty, early aortic valve replacement with a RK operation appears to be effective in improving LV function and increasing the chance for a biventricular circulation.

O8-1
Towards a fully absorbable percutaneous ASD closure device

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Introduction: Transcatheter closure of hemodynamically significant atrial septal defects (ASDs) with adequate rims has become the standard treatment modality. While this treatment is generally safe and effective, there are several reports of complications of a chronic foreign body. In addition, the permanent implant hampers transseptal access to the left atrium should this be required during alter life. We investigated the feasibility and biological short-term effects of a fully absorbable septal occluder in an animal model.
Methods: ASD were created interventionally in a sheep model by transeptal puncture and subsequent balloon dilatation of the defect (n = 12). Using a 12F sheath, the defects were closed under fluoroscopic and intracardiac echo control using a (patially) radio-opaque, fully retrievable and repositionable septal occluder consisting of a completely absorbable polymer. Residual leakage was assessed by color Doppler and bubble contrast studies and devices explanted for gross pathology and histology after 1, 3 and 6 months.
Results: Transcatheter ASD closure was possible in all animals. In 4 sheep the device was either repositioned or retrieved and replaced without problems. Devices showed excellent echogenicity and good visibility under fluoroscopy. All defects were closed effectively and showed no or small residual shunting immediately after implantation. No residual leaks were observed after 1-6 months follow up. Histology showed a very benign healing response with less cellular response compared to permanent implants and first signs of polymer resorbtion.
Conclusions: Transcatheter ASD closure using fully absorbable, completely repositionable and retrievable ASD occluder is feasible. In an experimental sheep model the devices showed an excellent echogenicity and good radiopacity. During short-term follow-up there were no residual or recurrent leaks. Histology showed a favorable and remarkable inert healing response. While longer term results are awaited, the material and technology holds promise for percutaneous ASD closure without permanent foreign material.

O8-2
Self-expanding stent Sinus superflex Visual to create an unrestrictive atrial communication in infants

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Background: In infants with complex CHD an unrestrictive atrial septal communication may be necessary. However balloon septostomy and dilation may be inefficient in creating an adequate atrial shunt; blade septostomy requires a big sheath and balloon expandable stents may slide and embolise. We evaluated the use of a 5F self-expanding "hooked" stent for this indication.
Patients and methods: Procedure done in 6 infants requiring a stable atrial shunt after failed Rashkind or thick atrial septum predicting poor result. Access through a 5 Fr sheath in the femoral vein. A 0.014" stiff coronary wire is preferably curled in left atrium; balloon interrogation of atrial septum with a soft balloon (10 mm Tyshak, Numed); optimizing the beamer angulation (LAO-cranial) perpendicular to the atrial septum; positioning of Optimed Sinus superflex Visual® 5F 10/20 mm; opening the distal part in the LA, hand injection through sidearm of the sheath to delineate relationship with the septum; pull back if required; opening the stent completely; post dilation if required. Low dose acetylcalicylic acid is given at 2 mg/d.
Results: 6 infants age median 3.7 months (range 1.0–7.8); CHD: complex TGA (1), tricuspid atresia (1), small left heart (4). In all patients the stent was successfully deployed; the anti-jump

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technique and visual markers allowed perfect placement; the open cell design provides the stent to hook at the septum, preventing sliding or embolisation. In all patients the stent provided an adequate unrestrictive atrial communication until the next surgical step [complex switch (1) or Glenn (5)]. The timing of the next operation was elective and not imposed by inadequate atrial shunt. All stents could easily be removed by the surgeon. No evidence of any shift or thrombo-embolic event during a follow-up of 4.7 months (range 1.4–7.0). The stent was firmly attached to the atrial wall.
Conclusions: Sinus superflex Visual® stent delivered safely through a 5F sheath allows to obtain an adequate atrial connection for several months in infants with complex congenital heart disease.

O8-3
Multimodality 3D-Imaging for 3D-Guidance of catheter-based interventions in congenital heart disease

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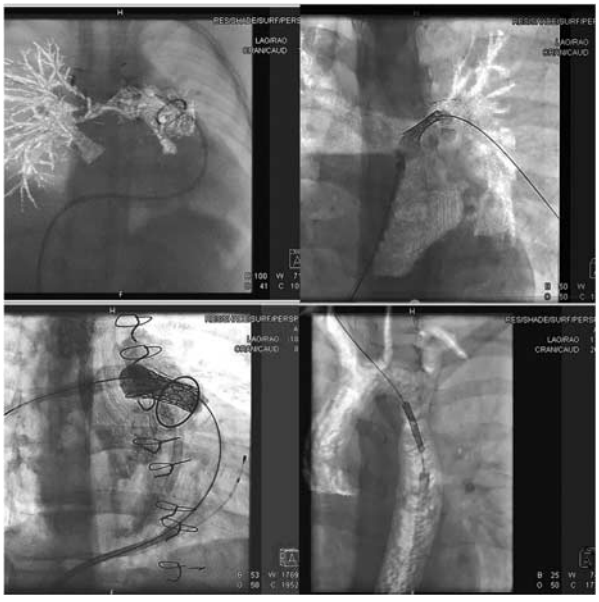
Introduction: Cardiac catheterization in congenital heart disease is a procedure in a 3D-space and limited by standard 2D-fluoroscopy. 3D-guidance is possible by three-dimensional rotational angiography (3D-RA): 3D-models from intra-procedural 3D-RA as well as 3D-reconstructions from former MRI- and MDCT-scans can be used. We evaluate possibility and significance of 3D-guidance.



Methods: Assessment of all cases with 3D-guidance in a 26 month period. Benefit for intervention and accuracy of 2D-3D registration were evaluated. Determination of fluoroscopy time, radiation-dose and used contrast dye.
Results: More than 90 cases at this time. Sufficient image fusion over 95%; 3D-guidance rated superior to 2D-guidance in over 94%; total dose-area product (μGym^2): 171.2 (range 38.6–1276.6), used contrast-medium: 4.2 ± 2.5 ml/kg.
Conclusions: Benefit of 3D-guidance: vascular structures are always visible in any c-arm angulation and table position. Surrounding tissue can be displayed (e.g. coronaries by pulmonary valve implantation). This technique enables a safer and faster catheter-based intervention in congenital heart disease with reduced need of radiation dose and contrast dye.

O8-4
A 10-year single-centre experience in percutaneous interventions for multistage treatment of hypoplastic left heart syndrome

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Introduction: Constant progress in surgical treatment of congenital heart defects in the last decade has significantly improved the prognosis for children with hypoplastic left heart syndrome (HLHS). However, due to specific anatomy and hemodynamics, complications still pose serious challenge. Interventional procedures complement or occasionally replace surgical treatment. The purpose of this paper is to report our 10 years of experience of interventional treatment of patients with HLHS.
Methods: Between 01/2001 and 10/2010, 161 percutaneous interventions were performed in 88 patients with HLHS between different stages of surgery. Ten interventions were performed as initial treatment of HLHS: isolated balloon atrial septostomy (5) or hybrid procedure (5). After stage I operation, 38 patients underwent 47 interventions. Stenosis of the aortic arch and isthmus was treated in 20 patients, stenosis of right ventricle-to-pulmonary artery shunt in 8 and proximal stenosis of the left (4) or the right (2) pulmonary artery in 6, secondary restriction of interatrial communication in 4. After second stage of surgical treatment 64 patients received 85 interventions: balloon angioplasty and stent implantation of stenosed pulmonary arteries (38 patients), balloon angioplasty of stenosed bidirectional Glenn shunt (14), closure of veno-venous collaterals (9), of systemic-to-pulmonary artery shunt (2) and of Sano shunt (1). In patients after Fontan operation closure of extracardiac fenestration was performed

Stage of treatment/ Indication		Before	After
Prior to surgical treatment/ Restriction on IAS	Sat. O2/%/	53% ± 9,4%	77% ± 9,3%
	Diameter of communication/mm/	1,75 ± 0,289	3,63 ± 0,479
	Pressure gradient/mmHg/	25,50 ± 5,802	9,75 ± 2,986
After I stage/ Restriction on IAS	Diameter of communication/mm/	4,2 ± 1,15;	9,9 ± 3,17
	Pressure in the LA/ mmHg/	19,4 ± 4,1	13,8 ± 2,4
Stenosis of Sano shunt	Sat. O2/%/	52 ± 12	75 ± 4
	Diameter/mm/	2,28 ± 0,48;	4,14 ± 0,69
Stenosis of aortic arch/ isthmus	Diameter/mm/	2,87 ± 0,82	5,15 ± 0,82
	Pressure gradient /mmHg/	29,38 ± 15,40	7,14 ± 4,28;
Stenosis of pulmonary arteries	Diameter/mm/	2,33 ± 0,51	3,58 ± 0,49
	Pressure gradient/mmHg/*	3,33 ± 1,633	2,5 ± 1,049
After II stage/ Stenosis of BDG	Sat. O2/%/	71% ± 7,2%	77% ± 4,9%
	Diameter/mm/	4,70 ± 1,38	7,32 ± 1,73
	Pressure gradient/mmHg/	3,29 ± 2,785	0,57 ± 0,646
Stenosis of pulmonary arteries	Sat. O2/%/	75 ± 9	77 ± 6
	Diameter/mm/	3,6 ± 1,33	8,65 ± 1,62
	Mean pressure/mmHg/	17,82 ± 4,73	15,18 ± 3,18
Closure of Veno-venous collaterals	Sat. O2/%/	74% ± 8%	79% ± 5%
	Mean pressure in VCS/ mmHg/*	17,08 ± 3,87	15,92 ± 3,42
After III stage/ Closure of fenestration	Sat. O2/%/	81 ± 5	96 ± 2
	Mean pressure in tunnel / mmHg/	13,85 ± 1,91	15,62 ± 1,26

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in 14, widening of stenosed extracardiac tunnel in 3 and self-expandable stent implantation in a stenosed left pulmonary artery in 1. One patient, with signs of failing Fontan, required interventional widening of extracardiac fenestration.
Results: See Table. All but 2 (*) results were statistically significant.
Conclusions: Patients with HLHS require additional percutaneous interventions between different stages of surgery, with the largest number of interventions being performed in those patients after bidirectional Glenn shunt and before Fontan operation. Percutaneous interventions result in haemodynamic stability, reduction in the number of operations and significant changes in pulmonary artery pressures, vessel diameters, O2 saturation.

O8-5
Off-label Use of Percutaneous Pulmonary Valved Stents in the RVOT
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Introduction: Percutaneous pulmonary valve implantation is now considered feasible and safe in selected patients with dysfunctional RVOT conduits. The "native" RVOT, smaller conduits (less than 16 mm) and the relatively large RVOT with dynamic outflow aneurysms are considered off-label use for percutaneous valve implantation.
Aim of study: To report the safety and feasibility of extended (off-label) application of percutaneous pulmonary valve implantation in patients with RVOT dysfunction.
Design: Retrospective analysis of prospectively collected data.
Setting: Tertiary pediatric and adult congenital heart cardiac centre.
Patients and Methods: Off-label use was defined as valve implantation in patients with a native RVOT (pulmonary valve or patch), RVOT conduits smaller than 16 mm or larger than 24 mm or the final valve diameter ⩾110% of the nominal conduit diameter. Successful valve implantation was defined as sufficient relief of RVOT obstruction (if present) and valve competence.
Results: Twenty Melody® valves and 2 Sapien® valves were successfully implanted in 22 patients at a mean age of 17 years (range 6.1–80.4years). Pre-stenting was performed in 21 patients 4.8 months (range 0–69.2) before valve implantation (14 covered stents; 12 bare stents). In 10 patients valves were implanted in the native RVOT after transannular/infundibular patch (n = 8) or pulmonary valvoplasty (n = 2). Mean diameter of the native RVOT was 18.8 mm (range 14–24) and mean final valve diameter was 22 mm (range 16–26). Twelve patients had their valves implanted in existing conduits ranging from 10 to 20 mm in size (mean 16 mm) with a mean final valve diameter of 20 mm (range final size 18–22; increase of diameter from nominal 4 mm (range 2–6 mm)). The implantations were uneventful and all patients were hemodynamically stable throughout the procedure.
Conclusions: Percutaneous pulmonary valve implantation is safe and feasible even in patients with unfavourable anatomy according to traditional protocol. Creating an adequate "landing zone" by pre-stenting is recommended. Larger studies are needed to identify the "ideal timing" for pre-stenting of the native RVOT.

O8-6
Melody transcatheter pulmonary valve implantation. Results from the Registry of the Italian Society of Pediatric Cardiology (SICP)
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Introduction: We aimed to analyze current practice in Italy of patients treated by using the Melody Medtronic valve.
Methods: Prospective, observational, multi-centric survey by means of a web-based database.
Results: Six centres participated and 63 patients were included in the registry (49% female; median age of 24 years (range 11-65 years). Subjects included had a history of a median 3 previous surgeries (range 1-5) and a median of 1 previous cardiac catheterization (median 0-4).
A tronco-conal disease was present in 39 patients, previous Ross operation in 9, other diagnosis in 15. Right ventricular outflow tract was reconstructed with homograft in 33 pts, biological valved conduit in 28 and other types in 5. Indication to valve implantation was pure stenosis in 21 patients (33%), pure regurgitation in 12 (19%), association of stenosis and regurgitation in 30 (48%). In all, except four, a femoral approach was used. Implantation was performed in 61 subjects. Pre-stenting was performed in 85% of cases. Ensemble delivery system was used (18 mm in 31%, 20 mm in 24%, 22 mm in 45%). The system was post-dilated in 63% of the procedures. Median procedure time was 170 minutes (range 85-360). No significant regurgitation was recorded after procedure while the trans-pulmonary gradient reduced significantly. Early complications occurred in 7 subjects(11%).Minor complications occurred in three subjects. One death occurred in the early post-operative period in a severely ill subject. Hospital stay was a median of 5 days (range 3-45 days). At a median follow-up of 30 months (range 12-48 months) three patients died due to underlying disease. Complications occurred in 6 patients: external electric cardioversion due to atrial fibrillation (1 pt), herpes virus encephalitis (1 pt), Melody valve endocarditis needing surgical explantation (2 pts), major fractures of the stent needing a second Melody valve implantation (2 pts). Freedom from valve failure at latest follow-up was 81.4% ± 9%.
Conclusion: Results of the SICP registry on transcatheter Melody pulmonary valve implantation shows that the procedure is safe and successful. Major concerns are related to the occurrence of stent fracture and bacterial endocarditis. Longer follow-up and larger series are needed.

O8-7
Percutaneous arterial duct stabilization in low-weight newborns (<2.5 kgs) with congenital heart disease and duct-dependent pulmonary circulation.
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Introduction: Arterial duct (AD) stenting is nowadays considered as an effective alternative to surgical systemic-to-pulmonary artery shunt in neonates with congenital heart disease and duct-dependent pulmonary circulation (CHD-DDPC). This option might be even more advisable in low-weight neonates who are at higher risk for surgical shunt and in whom repeat stent dilatations might adapt the shunt magnitude to the patient's growth.
Methods: Between April 2003 and December 2011, 88 neonates underwent AD stenting as palliation of CHD-DDPC at our Institution. Among them, 20 patients were at high-risk for surgical

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shunt because of low-weight (2.1 + 0.3 kgs, range 1.4-2.5, median 2.2)(Group I). Procedural success and complication rate of AD stenting in this subgroup were compared to the normal weight neonates (Group II).
Results: The procedure was successfully completed in all patients. Procedural and fluoroscopy times did not significantly differ with respect to normal weight neonates (103 + 34 vs 114 + 50 min and 28.1 + 14.9 vs 21 + 20 min, respectively, p = NS for both comparisons). Complication rate and need for emergency surgical shunt were 15.7% and 9.3%, respectively (p = NS vs Group II, for both comparisons). In-hospital mortality was 10% (2 patients)(p = NS vs Group II), unrelated to the stenting procedure. After stenting, the duct size increased from 2.3 + 1.2 to 3.7 + 1.4 mm ($p < 0.01$) and percutaneous O2 saturation increased from 80.5 + 11.3 to 91.8 + 5.5% ($p < 0.0001$), respectively. Over a mid-term follow-up, 3 patients underwent stent redilatation and 5 were submitted to successful corrective surgery. At pre-surgical cardiac catheterization, the Nakata index significantly increased from 120 + 61 to 295 + 121 mm/mm2 ($p < 0.05$), without any difference with respect to the Group II.
Conclusions: As already reported in normal weight newborns, AD stenting is a feasible and cost-effective palliation also in low-weight newborns with CHD-DDPC, supporting the spontaneous clinical improvement process or promoting significant pulmonary artery growth in view of corrective surgery at lower risk.

O8-8
Stent Fenestration of the Failing Fontan Circulation – a single centre experience
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Introduction: Elevated systemic venous pressures after Fontan completion are a risk factor for early or late complications or failure. Fenestrations allow for off-loading of the Fontan circuit and can provide relief of symptoms. Catheter creation of diabolo stent fenestrations was introduced at BCH in 1998.
Setting: Retrospective case review of patients who underwent stent fenestration of a Fontan circulation over a 10 year period at a single institution.
Patients and Methods: Thirty-four patients underwent transcatheter stent fenestration 1day – 10 years (median 22 days) after Fontan completion. De-novo fenestrations were created in 16 patients using either the stiff end of a guidewire or a Brockenborough needle. Median age was 5.5(3.4-14.1) years and median weight 17 (11.5-63) kg. Seven patients underwent concomitant LPA stenting.
Results: Three patients underwent stent fenestrations for LCOS within 5 days of operation as an alternative to Fontan take-down – all three survived. Twenty-one were treated for prolonged pleural effusions with resolution within a median of 10 days. Five patients each underwent creation of stent fenestration for bronchial casts or PLE late post Fontan with resolution of symptoms after a mean of 3.5 (2-6) months. Overall Fontan pressures decreased from mean 18.5 to 16 mmHg ($p < 0.001$) and saturations decreased from 93% to 81% ($p < 0.001$). All diabolo stents remained patent over the F/U period.
Conclusion: Stent fenestration of the failing Fontan circulation provides good symptomatic relief and a significant drop of Fontan pressures at the cost of systemic desaturation. Long-term patency of the diabolo stent is excellent.

O9-1
Clinical utility of Egami score system in patients with Kawasaki disease to predict coronary artery abnormality
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Background: The Egami score is used to predict responsiveness to initial intravenous immunoglobulin(IVIG) in patients with Kawasaki disease (KD). However, clinical utility of predicting coronary artery lesions (CAL) is unclear.
Methods: This study included 156 patients with KD who were treated at Kitasato University. We investigate the clinical utility of Egami score system for predicting CAL before IVIG treatment. The Egami score system use age, days of illness, platelet count, C-reactive protein, and alanine aminotransferase level to identify IVIG-resistant patients before treatment (cut off:3 points; 78% sensitivity and 76% specificity). Patients were divided into 2 groups using Egami score: group A(≤ 2 points) and group B(≥ 3 points). Coronary artery Z-score was evaluated in the left main trunk (LMT), left anterior descending artery(LAD), and right coronary artery(RCA) before initial treatment and at 1 month after treatment. The p values were corrected by using Student's t test.
Results: The coronary artery Z-score in group B was significantly higher than that in group A before initial treatment and at 1 month after treatment($p < 0.05$ for LMT, LAD, and RCA).
Conclusions: The Egami score predicted coronary artery abnormalities before initial treatment and might predict coronary artery outcomes.

O9-2
Vulnerability of Coronary Circulation after Norwood Type Operation
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Background: Surgically reconstructed aorta after Norwood (ND) type operation can be stiff and hence associated with reduced aortic reservoir function. This could in turn adversely affect coronary perfusion and thereby induce right ventricular (RV) dysfunction, which is often observed after the ND procedure. We hypothesized that ND operation leads to a reduction in subendocardial coronary perfusion due to its hemodynamic characteristics and that the impaired coronary perfusion is associated with poor outcome after this procedure.
Methods: We studied 29 consecutive patients who had undergone ND operation and RV-PA shunt. The subendocardial viability ratio (SEVR: Buckberg index), a marker of subendocardial ischemia, was calculated as time tension index divided by diastolic pressure time index, using the ascending aortic pressure waveform recorded during cardiac catheterization. Data were compared to those in 31 control subjects who had VSD or PDA with negligible shunt flow.
Results: The mean SEVR of ND patients was significantly lower than that of the controls (0.71 ± 0.21 vs. 1.04 ± 0.24, mean ± SD, $p < 0.001$). The mean ascending aortic stiffness was significantly higher in ND patients than in the controls. Importantly, the SEVR in ND patients with poor outcome (defined as death, progressive atrio-ventricular regurgitation, or severe symptomatic heart failure) was significantly lower than that in the remaining ND patients (0.62 ± 0.04 vs. 0.82 ± 0.05, $p < 0.01$). In addition, the lower SEVR in ND patients correlated significantly with higher circulating levels of BNP, angiotensin and aldosterone ($p < 0.01$, each). In this study, cardiac index, Qp/Qs, and the anatomical feature of the native aorta, have no correlation with SEVR. Multivariate regression

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analysis showed that increased aortic stiffness, faster heart rates and severity of residual aortic arch stenosis (% stenosis compared to descending aorta) correlated significantly with the lower SEVR ($p = 0.018$, 0.011, 0.008, respectively).
Conclusions: There is high risk of reduced coronary perfusion after ND operation due to increased aortic stiffness and abnormal morphology of the reconstructed aorta and vulnerability to tachycardia. These results emphasize the importance of the surgical design of the re-constructed aortic arch, and highlight the importance of rate-control in the postoperative management of patients with ND operation.

O9-3

Implications of Early Aortic Stiffening in Patients with Transposition of the Great Arteries after Arterial Switch Operation.

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Objectives: In patients with transposition of the great arteries (TGA) the elastic function of the transposed aorta after arterial switch operation (ASO) is suspected to be important for long-term prognosis. Therefore, the goal of this study was a comprehensive assessment of the aortic bioelastic properties in children and adults with TGA after ASO.
Methods: 51 patients and 34 age-matched controls were studied at 3.0 Tesla with magnetic resonance imaging. 43 patients (12.8 ± 6.9 years) underwent one-stage ASO, 8 patients (23.8 ± 6.9 years) had prior pulmonary artery banding (two-stage ASO). Aortic dimensions, distensibility, pulse wave velocity (PWV), aortic arch angle, left ventricular (LV) mass, LV function and left atrial (LA) volumes as a surrogate marker of diastolic dysfunction were assessed.
Results: Compared to controls, patients had increased aortic root areas (602.6 ± 240.5 vs. 356.8 ± 113.4 mm2/m, $p < 0.01$) and reduced distensibility of the thoracic aorta most pronounced at the aortic root (3.2 ± 2.0 vs. 9.1 ± 4.7 10-3 mmHg-1, $p < 0.01$). Ascending and descending aortic distensibility correlated negatively with the aortic areas at the same level ($p < 0.01$). PWV was significantly higher in two-stage ASO (5.3 ± 1.0 vs. 3.5 ± 0.6 m/s, $p < 0.01$). In patients PWV and aortic distensibility correlated with age ($p = 0.04$-<0.01). LV mass was higher in patients than in controls ($p = 0.02$). LA volumes correlated negatively with aortic root and ascending aortic distensibility and positively with PWV ($p < 0.05$).
Conclusions: Reduced aortic bioelastic properties and aortic root dilatation are present in TGA patients post ASO and are likely to contribute to LV diastolic dysfunction.
Impaired aortic bioelasticity was strongly associated with age suggesting the need for routine monitoring for early onset of degenerative cardiovascular disease.

O9-4

Coronary orifices patterning in outflow tract defects: a marker of outflow tract rotation during cardiac development.

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Background: Coronary arterial anatomy is often abnormal in cardiac outflow tract defects (OTD), particularly in common arterial trunk (CAT). Our hypothesis is that the position of the coronary orifices on the aortic/truncal circumference in OTD could be related to the location of a coronary-repulsive subpulmonary myocardial domain that influences the epicardial course of the main stems and their final connection to the aorta.
Material: We analyzed 101 heart specimens with OTD from the anatomic collection of the French Reference Center for Complex Congenital Heart Defects: 46 CAT, 15 tetralogy of Fallot (TOF), 29 TOF with pulmonary atresia (TOF-PA), 11 double-outlet right ventricle with subaortic ventricular septal defect (DORV), and 17 controls.
Methods: Hearts were analyzed in anatomic position with the extremities of the right and left atria on the intersecting line of a horizontal and vertical plane. The position of left and right coronary orifices (LCO, RCO) was measured in degrees on the aortic/truncal circumference, and their configuration analyzed. We calculated the anterior angle between LCO and RCO (α) that represents the area devoid of coronary orifices.
Results: The LCO was more posterior in OTD compared to control, mean position 0° in controls, 31° in TOF, 47° in TOF-PA, 44° in DORV, 63° in CAT ($p < 0.005$). The LCO was more posterior in CAT than in other OTD ($p < 0.05$).
The RCO was more anterior in TOF (242°), TOF-PA (245°) and DORV (271°) than in controls (213°, $p < 0.05$), but not in CAT (195°).
The α angle was similar in TOF, TOF-PA, DORV and control (respectively 149°, 162°, 133° and 147°) but significantly larger in CAT (229°, $p < 0.0001$).
Coronary orifices were abnormal in 87% CAT (LCO 72%, RCO 42%), 20% TOF, 17% TOF-PA, 9% DORV, 23% controls.
Conclusion: In all OTD but CAT, the posterior displacement of LCO and anterior displacement of RCO, while the α angle remains constant, might be due to incomplete rotation of the myocardium at the base of the outflow tract, leading to abnormally positioned subpulmonary myocardial domain. The larger α angle, and the abnormal configuration of LCO found in CAT, could reflect its dual identity, aortic and pulmonary.

O9-5

Assessment of pulmonary endothelial function during invasive testing in children and adolescents with idiopathic pulmonary arterial hypertension

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Introduction: IPAH may be associated with pulmonary endothelial dysfunction, however data regarding the impact of endothelial dysfunction on severity and prognosis of this disease are limited. The purpose of our study was to assess pulmonary endothelial function by vasodilator response to Acetylcholine (Ach) administered in segmental pulmonary arteries in children with idiopathic pulmonary arterial hypertension (IPAH). We hypothesized a relation between pulmonary endothelial response to Ach, severity of the disease and clinical outcome.
Methods: 43 children and adolescents (mean age 10.4 ± 5.5 years) with IPAH were included in the study. Changes in pulmonary blood flow in response to Ach were determined using intravascular Doppler flow measurements. Pulmonary flow reserve (PFR) was calculated as the ratio of pulmonary blood flow velocity in response to Ach relative to baseline values.
Results: Mean PFR of all patients was 1.58 ± 0.1. Mean follow up after catheterization was 55.7 ($\pm$ 41.9) months. Freedom from serious cardiovascular events (lung transplantation or death) was 83%

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after 2 years, 76% after 3 years, and 57% after 5 years. PFR correlated with WHO functional class and long-term response to calcium channel blocker therapy. Receiver-operating characteristic curve revealed a PFR of 1.4 as best cut-off value. Kaplan-Meier analysis demonstrated PFR <1.4 as highly predictive for cardiovascular events (Log rank (Mantel Cox) Chi square 12.49; $p < 0.0001$).
Conclusions: Our study demonstrates a strong relation between pulmonary endothelial response to Ach and prognosis of children with IPAH. As an adjunct to the usual testing protocol, this method provides additional information for therapeutic guidance.

O9-6
Effect of pulmonary vasodilatators on clinical and Echocardiographic parameters in children with primitive pulmonary hypertension
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This study was to assess the influence of anti-PHT medications on clinical and echocardiographic parameters in children.
Methods: Patients <18years diagnosed with primitive PHT were retrospectively reviewed. Functional class, RV and LV diameters, tricuspid regurgitation grade, systolic and mean pulmonary pressure, TAPSE and TAC were recorded before and after anti-PHT treatment. Deceased or transplanted patients (group I) were compared to survivors (group II).
Results: 16 patients (11 males), were diagnosed with PHT (11 primitive and 5 illegitime association: closed VSD = 2, closed PDA = 2, TGA = 1), at mean age of 6.7years (0.7 to 16.2). All received anti-PHT treatment: monotherapy (IV prostacycline = 4, bosentan = 2), or bitherapy (IV prostacycline+bosentan = 2, bosentan+sildenafil = 7) or tritherapy (IV prostacycline+bosentan+sildenafil = 1). Mean FU was 4.3years (0.7 to 10.5). Five patients died, 1 underwent heart-lung transplantation (group I = 6) at mean FU 3.4y (1 to 4.1), and 10 survived (group II) at mean FU 4.7y (0.8 to 9.5).
NYHA class changed from 50% class II, 37.5% III and 12.5% IV before treatment, to 50% class I, 12.5% II and 37.5% IV after.
Overall mean RV/LV did not change (1 to 0.95). RV/LV decreased in group II: 1.14 to 0.51, and increased in group I: 0.87 to 1.57. The proportion of TR grade 0-I increased from 47% to 60%, and of TR grade II-III decreased from 53% to 40%. All patients with TR grade III at end FU ranged in group I. Systolic and mean pulmonary pressure did not change: respectively 92.1 to 99.9 mmHg and 65.8 to 56.2 mmHg, and were similar between groups I and II.
TAPSE and TAC were not available in group I, and increased in group II: respectively from 14 to 17.9 mm and 76.8 to 101.6 ms.
All patients in group I were either in NYHA class IV at first presentation (2cases), or received first therapy more than 6 months after diagnosis (4cases).
Conclusion: Anti-PHT therapy contributes to ameliorate functional class and RV function despite no significant effect on pulmonary pressure level. Persistent TR grade III and increasing RV/LV seem to be factors of bad prognosis.

O9-7
Current medical treatment in Pediatric Pulmonary Hypertension (PH)–Insights from the Global Registry Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension (TOPP)
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Objectives: The TOPP global registry provides demographic, clinical and outcome data in pediatric PH. One of the primary objectives was to describe current treatment. Treatment decisions were made by site clinicians without TOPP involvement.
Methods: 31 sites from 19 countries enrolled patients, diagnosis on/after January 2001, enrollment 2008-2010, age 3 months–18 years at confirmatory right heart catheterization). PH was defined as: mPAP ⩾ 25 mmHg, PCWP ≤ 12 mm Hg and PVRI > 3 units x m2.
PH targeted therapy (PHTT) included prostacyclin and its analogs (PGI2s), endothelin receptor antagonists (ERAs) and phosphodiesterase inhibitors type 5 (PDE5Is). Calcium channel blockers (CCBs) were considered as PHTT in responders to acute vasodilator testing. Supportive therapy included anticoagulation, oxygen, diuretics and/or digitalis.
Results: Of the 456 patients enrolled, 362 (79%) met all entrance criteria with 357 (99%) based on right heart catheterization and 5 (1%) on independently reviewed echocardiography. Of the PH confirmed patients, 102 (28%) were incident (defined as diagnostic right heart catheterization <3 months prior to enrollment) and 260 (72%) were prevalent patients. For the present analysis only incident patients were included.
At diagnosis 10% (n = 10) of incident patients were on PHTT, 76% (n = 77) were treatment naïve and 15% (n = 15) had incomplete data regarding treatment. PDE5Is were used in 52% (n = 53), whereas 17% (n = 17) of the patients received ERAs and 11% (n = 11) PGI2s. CCBs for PHTT were used in 13% (n = 13). Supportive therapy in form of anticoagulation was provided in 17% (n = 17) of the patients, oxygen in 27% (n = 27) and diuretics in 30% (n = 31). Digitalis was used in 16% (n = 16) of the patients. Of the incident patients, 66% (n = 51) were started on PHTT monotherapy at diagnosis and 16% (n = 12) started with dual therapy.
Conclusion: The largest proportion of pediatric PH patients enrolled in TOPP and recently commenced treatment start on PDE5Is (as monotherapy) at diagnosis. Long-term follow-up should provide invaluable data on how treatment patterns affect outcomes.

O9-8
Characteristics of PAH associated with pretricuspid shunts in the registry of the French PAH network
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Background: Pulmonary arterial hypertension (PAH) associated with pretricuspid shunts is commonly delayed until adulthood but PAH may reveal the underlying unknown congenital heart defect (CHD).
Objective: To review and analyze patients with PAH and pretricuspid shunts enrolled in prospective French PAH registry.
Patients and results: CHD-PAH accounted for 9.8% of PAH in this registry. 145 patients (101 female-44 male)had a pretricuspid

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shunt (80% of CHD-PAH diagnosed after 30 years of age). Thirty had their shunt previously closed. Mean age at first symptom and at diagnosis of PAH were respectively 37.2 years and 40 years, males being much older at time of diagnosis 48.0 vs. 34.3 for females. In the majority of cases, the diagnosis of the shunt was made concomitantly with PAH diagnosis but the diagnosis of the CHD was posteriorly made in 8 cases. Functional class at entry was I (2%), II (33%), III (60%), and IV (5%). Resting oxygen saturation was below 92% in 7%. Mean 6 minutes walk distance at entry was 358 ± 109 m. Total lung capacity (TLC) was below 80% of the predicted value in 38%, and the median value of the ratio of the forced expiratory volume in one second to the forced vital capacity (FEV1/FVC) was below 70% of predicted value in 40%. Mean pulmonary artery pressure at RHC (n = 136) was 55 ± 16 mmHg, cardiac index 2.9 ± 1.1 L/min/m2 and PVR were 1565 ± 978 dynes.s.cm-5. At entry, 91% received a monotherapy, 8% a combined oral therapy, and 1% a tritherapy including prostacyclin. At last follow-up (mean 3.7 ± 2.2 years), functional class was improved by +1 in 71% of class IV, 28% of class III, and 13% of class II; 6 minutes walk distance increased by +35 m; and control RHC (n = 66) showed increase in cardiac index ($p < 0.05$) and decrease in PVR ($p < 0.05$). Survival at 6 years was 76%. No difference could be found between open and closed shunts in this series.
Conclusion: PAH at diagnosis was only moderately less severe than in idiopathic PAH and outcome was not as favorable than expected in CHD-PAH.

O10-1
Evaluation of Coronary Artery Abnormalities and Sudden Death Risk in Williams Syndrome Patients Using Myocardial Perfusion Scintigraphy and CT Angiography

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Introduction: Sudden death risk in Williams syndrome (WS) patients has been shown to be 25-100 times higher than in the general population. This study aims to detect coronary artery anomalies and myocardial perfusion defects in WS patients using noninvasive diagnostic methods.
Patients and Methods: This study features 38 patients diagnosed with WS. In addition to physical examination, electrocardiography, and echocardiography, computed tomography (CT) angiography and rest/dipyridamole stress technetium-99m sestamibi (99mTc-sestamibi) single photon emission computed tomography (SPECT) myocardial perfusion scintigraphy (MPS) were performed.
Results: Twenty-one patients (55%) were male; 17 (45%) were female. The average patient age was 12 ± 5 years (2.5-26 years); the average follow-up period was 7.2 ± 4.2 years (6 months-18 years). Cardiovascular abnormalities were found in 89% of patients, the most common one being supravalvar aortic stenosis (SVAS). CT angiography revealed coronary anomalies in 10 patients (26%), the most common ones being ectasia of the left main coronary artery and proximal right coronary artery as well as myocardial bridging. SVAS was present in 80% of patients with coronary artery anomalies. 99mTcsestamibi SPECT MPS revealed findings possibly consistent with myocardial ischemia in 29% of patients, and ischemia in 7 out of 10 patients (70%) with coronary anomalies shown on CT angiography ($p = 0.03$).
Conclusion: Coronary artery abnormalities are relatively common in WS patients and are often accompanied by SVAS. CT angiography and dipyridamole 99mTc-sestamibi SPECT MPS seem to be less invasive methods of detecting coronary artery anomalies and myocardial perfusion defects in WS patients.

O10-2
ECG risk score: a significant advance in the risk stratification of paediatric patients with hypertrophic cardiomyopathy

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Introduction: In hypertrophic cardiomyopathy (HCM) the risk of sudden unexpected arrhythmia death in an asymptomatic individual is at its highest in between 8-16 years of age. In adult HCM patients we have shown that abnormally large ECG-voltages, and repolarisation abnormalities indicate increased risk, and have published an ECG risk score which improves risk stratification. The objective of this study is to evaluate this risk score for paediatric HCM-patients.

ECG risk score

Any deviation in QRS-axis		1 point
Pathological T-wave inversion limb leads		1 point
Pathological T-wave inversion precordial leads*		2 points
ST-segment depression ≥ 2 mm		2 points
Dominant S in V_4		2 points
Limb-lead QRS-amplitude sum	≥ 7.7 mV	1 point
	≥ 10.0 mV	2 points
	≥ 12.0 mV	3 points
12-lead amplitude-duration product	≥ 2.2 mV.s	1 point
	≥ 2.5 mV.s	2 points
	≥ 3.0 mV.s	3 points
QTc	≥ 440 ms	1 point
		Max score = 14

*Total score available for T-wave abnormalities is 2 points, i.e. 1 limb lead point is not added on top of precordial points. QTc = corrected QT-interval (Eur Heart J 2010;31:439-49).

Methods: All Sweden's centres of paediatric cardiology collaborated on identifying all patients that had died suddenly from HCM between birth and 19 years of age in the last 30 years. 33 patients were identified, with 29 patients where ECGs had been recorded prior to death (mean follow-up 9.8 + 5.9 (SD) years). For comparison we used ECGs from the total geographical cohort of HCM-patients age 0-19 years from West Götaland region where systematic cascade screening has been used to identify symptom-free children with HCM (n = 48, follow-up 6.8 + 8.1 years).
Results: A high risk ECG-score limit set at 6 points or above gives a relative risk for sudden death of 24.3 [95% CI 3.5-169; $p < 0.0001$], sensitivity of 96% [80-100%] and specificity of 78% [62-89]. The positive predictive value was calculated within the West Götaland cohort where there were 7 deaths (giving an annual sudden death mortality of 2.1%). Sensitivity was here 100% [54-100%], positive predictive value 40% [16-68%] and negative predictive value 100% [89-100%].
*Conclusions:*ECG risk score is applicable to children, and the hitherto most specific individual risk factor described.

O10-3
High Prevalence of Sarcomeric Mutations in a Pediatric Cohort of Hypertrophic Cardiomyopathy

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Introduction: Hypertrophic cardiomyopathy (HCM) is, in most adults and adolescents, a genetic disorder of sarcomeric proteins inherited as an autosomal dominant trait. In contrast, a genetic origin of the disease in infants and children is considered unusual (commonly associated with metabolic disorders, neuromuscular diseases and congenital malformation syndromes). The objective of this study is to describe the genetic origin and clinical characteristics of a cohort of pediatric patients with HCM.
Methods: Prospective observational cohort study from June 2010 to November 2011 of children with idiopathic HCM. Demographics, family tree, genetic test for sequences of sarcomere protein genes (MYH7, MYBPC3, TNNI3, TNNT2, TPM1, MYL2, MYL3, ACTC and TNNC1), ECG and echocardiography were performed.

Gene	Mutation
MyBPC3	2 x R502Q/g10952G>A
	R470W/g10770C>T
	R502W/g10951C>T
	IVS22-1/g14970G>A
MYH7	H251N/g6648C>A
	R453H/g9124G>A
	T1759M/g22083C>T
MYL3	A57D/g2629C>A
TNNT2	R278C/g18433C>T
ACTC	L106V/g2278C>G

Results: 15 patients were included in the cohort. 73,3% were male. Mean age 10,7 years (2,3 months to 17,7 years), being 9 (60%) under 13 (Mean age 7,1 years ± 3,5). 66,7% of the patients included (10/15) were positive for a mutation, this proportion was equal in both age groups. A patient had 2 concurrent mutations in 2 different genes. The mutations were found in 5 different genes (see table). Three mutations (30%) had never been described before. At present, 7 of the 10 positive families have been studied, 2 being denovo mutations and 5 inherited mutations. The interventricular septum thickness was similar in both mutation and no-mutation groups. 3 patients with mutations had an ICD implantation. 1 patient with mutation was diagnosed after an aborted sudden death episode, and died for severe neurological complications. 1 patient with no mutations has a myomectomy performed. The youngest patient, with MYL3 mutation, was a prenatal HCM diagnosis and also a Noonan Syndrome.
Conclusions: Mutations in cardiac sarcomere proteins are a common cause of pediatric HCM, also in the group of infants and children. Therefore, systematic screening of relatives of HCM patients, even in those aged younger than 13 years, is needed. The description of novel mutations will expand the range of reported sarcomeric mutations, improving the knowledge and management of pediatric HCM.

O10-4
Origin, genotype and clinical phenotype of the Long QT Syndrome R518X/KCNQ1 mutation in Sweden

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Introduction: In the Long QT Syndrome (LQTS), associated with syncope and sudden death at a young age, mutation-specific risk stratification could be of clinical importance. Here we investigate the clinical phenotype and origin of the worldwide common R518X/KCNQ1 mutation, in Sweden, causative of both recessive and dominant type LQTS.
Methods: Clinical data, including manually measured ECGs, were collected from medical records, a personal interview and a questionnaire. Genealogical investigations were performed, using parish records and genealogical databases. A haplotype analysis, including 14 microsatellite markers flanking the KCNQ1 gene, was performed in index families and healthy controls. Mutation age was estimated using ESTIAGE and DMLE computer software.
Results: We identified 97 mutation-carriers (59 females) in 19 Swedish R518X index families, whereof 11 cases (6 females) with double mutations (4 homozygotes) and concomitant hearing-loss, i.e. Jervell-Lange-Nielsen (JLNS) cases.
JLNS cases presented with a severe phenotype (QTc 576 ± 61 ms; age at debut 2 ± 1 years; syncope 73%; (aborted) cardiac arrest 55%).
In heterozygous carriers, during a mean follow-up before beta-blockers of 30 ± 20 years, phenotypic variability was evident (QTc 464 ± 31 ms, range 383-537 ms; age at debut 15 ± 11 years, syncope 17%; (aborted) cardiac arrest 2%), with poor correlation between QTc prolongation and symptomatic phenotype ($p = 0.651$). Four symptomatic cases presented with a normal QT interval. One previously asymptomatic case (QTc >500 ms) died suddenly during physical exertion in combination with a diet-induced hypokalaemia.
A common geographic origin in the upper northern region with focus in Pite River Valley was found for 17/19 families. A common haplotype (4-14 markers, median 8) was identified in 16/17 tested families. The age of the founder haplotype was estimated to 650 years (95% CI 425;1000) and 600 years (95% CI 400;950) by ESTIAGE and DMLE, respectively.
Conclusions: R518X is associated with a severe phenotype in JLNS cases and a highly variable, albeit mild, clinical phenotype in heterozygotes. Importantly, in R518X heterozygotes, QTc prolongation did not identify at-risk individuals.
The majority of Swedish R518X/KCNQ1 cases share an upper northern origin around the 14th century. Founder effects have resulted in a high frequency of R518X in Sweden, and a notably high JLNS prevalence.

O10-5
Unexpected Sudden Cardiac Death Under School Supervision in Japan

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Objectives: Approximately half of sudden cardiac death (SCD) of children occurs unexpectedly. In Japan, cardiac screening has been continued for 40 years. To inform school caregivers of necessary knowledge about SCD, recent reports of death cases were analyzed.

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Methods: Ninety-eight percent of all primary, middle and high schools are affiliated with compensatory system for accidents managed by Japan National Agency for Sports and Health, and they have to report precisely when SCD occurred. SCD student whose life-threatening cardiac disorder was already diagnosed before fatal event is classified as 'expected' group, and SCD student whose risk was unknown before fatal event but was judged as cardiac disorder by autopsy or emergency examinations as 'unexpected' group. Causal disease, relation with exercise, and use of automated external defibrillator (AED) were analyzed with their medical reports and compared them.
Results: Ninety-two SCD were reported from schools between 2006-2009. Forty-four were classified as expected group, and 48 as unexpected group. Causal diseases in expected group were 14 congenital heart diseases, 13 cardiomyopathies, 4 Wolff-White-Parkinson syndromes, 2 long QT syndromes, 2 aortic dissections, and 9 others. In unexpected group, causal diseases revealed by emergency examinations and/or autopsies were 5 acute myocarditis, 5 congenital coronary artery anomalies, 4 aortic dissections, 4 cardiomyopathies, 2 incomplete right bundle branch blocks, 1 commotio cordis, however, causes of 27 cases (56.3%) including 7 autopsies in unexpected group were unknown. In expected and unexpected group, 24 cases (54.5%) and 39 cases (81.3%) occurred during exercise, respectively, with significant difference ($p = 0.04$). AED was used in 29 cases in every both groups with no significant difference. The incidence of SCD decreased to 23.0 per year during 2006 and 2009, comparing to 73.2 per year during 1989 and 1998.
Conclusions: Though improvement of incidence is supposed to be multifactorial, cardiac screening and management has possibility to prevent SCD. Utilization of AED by schoolteachers or students as bystanders also has possibility to save a part of fatal cases. It may be worth discussing to induce echo screening of aorta width and origin of coronary arteries at least for students who will be competitive athletes.

O10-6
Neurodevelopmental outcome at one year of age after neonatal cardiac surgery for severe congenital heart disease in relation to perioperative cerebral magnetic resonance imaging

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Objectives: To analyse impact of neonatal cardiac surgery for severe congenital heart disease (CHD) on neurodevelopmental outcome at one year of age in relation to perioperative cerebral magnetic resonance imaging (MRI) and spectroscopy (MRS) .
Methods: Cerebral MRI was performed before and after cardiac surgery in 30 neonates with d-transposition of great arteries, hypoplastic left heart syndrome and other severe CHD. Follow up included standardized neurological examinations at neonatal age and the Bayley Scales of Infant Development III at 1 year of age.
Results: Before surgery, in 26 patients (87%) MRI showed signs of generalized hypoxia with hyperintensity of the white matter (WM) on T2, with punctuate WM lesions in 5 (17%). Ten patients (33%) showed subdural and 8 (27%) had choroid plexus hemorrhages. Four patients had small cerebral strokes. After surgery WM hyperintensities (T2) were observed in 24 patients (82%). Two patients developed new WM lesions. New subdural hemorrhages were found in 5, new plexus choroid hemorrhages in 2. No new cerebral strokes were seen after surgery, while all cerebral strokes detected before surgery decreased in size. MRS was abnormal in all patients with elevated lactate in WM and basal ganglia and decreased N-acetyl-aspartate (NAA) values. Before and after surgery the standardized neurological examination (Zurich Neuroscore, range 0-18 points) was similar with mild abnormalities (median score = 2 before/after surgery) with predominantly muscular hypotonia, but no focal neurological deficit. There was one patient with tonic-clonic seizures before surgery (no WM lesion or cerebral stroke). So far, twelve patients had a one-year neurodevelopmental examination. Median cognitive score was 105 (range 60-120), language score was 94 (65-106) points and motor score was 91 (46-103).
Conclusions: Signs of generalized hypoxia in the WM with pathologic MRS values are the predominant findings in more than 80% of infants, while intracranial hemorrhages are observed in half of infants, and punctuate WM lesions and cerebral strokes in less than 20%. Neurological findings in the neonatal period are mostly mild and non-focal and at one year of age, the cognitive and motor outcome is within the normal limits, but with a very wide variability.

O10-7
Outcomes associated with occlusive thrombosis and its management in pediatric patients after cardiac surgery

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Introduction: Thrombosis is an important cause of morbidity and mortality after pediatric cardiac surgery. Multiple treatment options are available for these patients, with progressive intensity associated with increased risk of adverse events. We sought to explore the balance of adverse outcomes of thrombosis versus adverse effects of therapy.
Methods: Patients with ⩾1 radiologically-confirmed occlusive thrombus after cardiac surgery (2002-2009) were reviewed. Information collected on each thrombus included patient characteristics, clot characteristics, hemostatic system activity before surgery and at the time of clot diagnosis, treatment details and outcomes.
Results: A total of 400 occlusive clots in 203 patients (median age 2 months) were included. Clots were identified in veins (n = 281;70%), arteries (n = 58;15%), brain (n = 18;5%), heart (n = 14;4%), surgical shunts (n = 20;5%) and pulmonary circulation (n = 9;2%). Mechanical/surgical clot removal was performed to address 43 clots in 27 patients. Indications for removal included occlusion of a cardiac structure/shunt/pulmonary vein/artery in 17(63%), occlusion of venous return in 9(33%) and hepatic infarction in 1(4%). Most patients (85%) who underwent mechanical/surgical clot removal did not require further antithrombotic therapy; however, 7(26%) did not survive to hospital discharge. Thrombolytics were given to 19 patients. Indications included multiple (>5) clots for 6(32%), valve thrombosis for 5(26%), occlusion of a cardiac structure/shunt/pulmonary vein/artery in 3(16%) and occlusion of peripheral vessels non-responsive to unfractionated heparin in 5(26%). Major bleeding complications were seen in 8/19(42%) patients who received thrombolytics, and 6/19(32%) patients did not survive to hospital discharge. For patients who survived to hospital discharge, the prevalence of clot resolution was 13% at 6 weeks after diagnosis, and 37% and 58% after 3 and 6 months.

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Factors associated with clot resolution included non-venous clot (HR:2.43, $p < 0.001$), lower fibrinogen level at diagnosis (HR:1.23, $p = 0.03$), higher enoxaparin dose at discharge (HR:1.74, $p = 0.03$) and use of thrombolytics (HR:2.29, $p = 0.002$). Severe bleeding complications occurred in 30 patients (15%). Associated factors included older age diagnosis (OR:1.15/year, $p = 0.04$), higher fibrinogen level at diagnosis (OR:1.67/unit, $p = 0.009$), longer duration of unfractionated heparin treatment (OR:1.05/day, $p = 0.02$) and use of thrombolytics (OR:5.84, $p = 0.001$).
Conclusions: Escalation of antithrombotic treatment was associated with both increased effectiveness and increased risk of bleeding complications. Both outcomes should be considered when selecting therapy for these patients.

O10-8
Long-term survival after univentricular heart surgery

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Objective: To collect reliable and complete data for evaluation of long term survival after surgery for univentricular heart defects.
Methods: All 304 patients (126 girls/178 boys) operated due to a univentricular heart defect in our institution before the age of 18 years from Jan 1st 1994 to Jan 1st 2009 were included. Patient files were cross-checked as of Jan 1st 2012 against the National Population Registry in Sweden allowing for reliable and complete data on survival. Two patients (0,6%) emigrated and were lost to follow up.
Results: Median age and weight at first surgery was 10 days (0-15.1 years) and 3.6 kg (1.2 – 30.0). Median age of survivors at follow up was 11.4 years (3.0–30.7). 83 (27.3%) deaths occurred with a median age at death of 69 days (0.01-22.6 years). Median survival time in the deceased patients was 29 days (0-11.2 years) after the last major surgery. Mortality in 271 patients with their first surgery in 1994-2008 were 51/110 (46.3%) in patients with classic hypoplastic left heart syndrome or an unbalanced atrioventricular septal defect with left ventricular hypoplasia (A), compared to 9/87 (10.3%) in patients with tricuspid atresia, double inlet left ventricle or pulmonary atresia with intact ventricular septum (B) ($p < 0.001$). Mortality in children with an indeterminate ventricular morphology and/or other complex congenital heart defects (C) was 18/74 (24.3%). Mortality in patients with their first surgery in 1994-1998, 1999-2003, and 2004-2008 was 41/95 (43.1%), 19/87 (21.8%) and 18/89 (20.2%) respectively ($p < 0.05$). Such an improvement was found in all three subgroups and in patients with their first surgery in 2004-2008 mortality was 13/33 (39%), 2/32 (6%) and 3/24 (12.5%) in group A, B and C respectively. Heart transplantation was performed in 16 patients (5%) with four late deaths.
Conclusion: Long term survival in patients with a systemic ventricle of left ventricular morphology was high (89.7%), while survival in patients with a systemic ventricle of right or indeterminate ventricular morphology was worse, i.e 53.7% and 75.7% respectively. An improvement of survival was seen over time in all groups.

O11-1
The impact of acoustic radiation force impulse sonoelastography to assess the liver stiffness in patients after Fontan procedure

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Introduction: Although late hepatic dysfunction and cirrhotic change were often seen in Fontan patients, not only the prevalence and progression of cirrhotic changes but also the non-invasive diagnostic tools for hepatic fibrosis have not been clearly clarified. In this study, we aim to clarify the clinical impact of the novel echographic imaging: sonoelastography using acoustic radiation force impulse (ARFI) to assess the liver stiffness in Fontan patients.
Methods: The study subjects were 20 patients with Fontan procedure (age: 12.7 +/- 5.3 years) and 75 patients either with minor cardiac anomalies or after uncomplicated definitive surgeries, having no hemodynamic compromise, (age: 11.4 +/- 6.2 years) as a control. The imaging apparatus was Acuson S-2000 (Siemens AG, USA) with 9L4 and 4C1 probe. The liver stiffness was measured and estimated by shear propagation velocity (m/s) (Vs) by Virtual Touch TM tissue quantification (VTTQ) which provided accurate numerical measurements related tissue stiffness at user-defined location using ARFI. The value of Vs was proportional to the degree of tissue stiffness (Young elastic modulus). The patients were in prone position and Vs at the 2-3 cm inner portion of the right lobe of liver was measured at 5 times consecutively and averaged.
Results: Vs of Fontan patients (2.52 +/- 0.63 m/s) was significantly higher than those of the control patients (1.25 +/- 0.23 m/s) ($p < 0.001$). In Fontan patients, Vs was weakly proportional to the ratio of mean pulmonary artery and aortic pressure : PA/AO ($r2 = 0.29$), the ratio of pulmonary and systemic vascular resistance: Rp/Rs ($r2 = 0.31$), and the multiplication of central venous pressure(CVP) and interval from Fontan ($r2 = 0.27$), but not associated with age at measurement, the interval from Fontan procedure, CVP, cardiac index, and biomarkers (BNP, r-GTP, platelet). Vs of Fontan patients were lower than those of patients with end-staged liver cirrhosis (> 4.0).
Conclusion: The liver stiffness after Fontan patients measured as Vs by VTTQTM is higher than control from shortly after Fontan operation and weakly associated with PA/Ao, Rp/Rs, and CVP*Interval. The Vs by VTTQTM could be a powerful tool for early non-invasive detection of liver fibrosis and cirrhosis in Fontan patients.

O11-2
Right atrial size is a strong predictor of arrhythmia in patients with repaired tetralogy of Fallot.

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Introduction: Patients with repaired tetralogy of Fallot (rtoF) are at risk of arrhythmia, right ventricular (RV) dilatation and dysfunction, and sudden death during long term follow-up. Frequently, pulmonary valve replacement (PVR) and/or defibrillator implantation are indicated. To date, risk stratification focuses on QRS duration and right and left ventricular dysfunction. Recent studies reveal an increasingly important role for atrial size and function in outcomes prediction in non-congenital cardiomyopathies. We hypothesised that atrial dilatation would also relate to outcomes in rtoF.
Methods: Retrospective analysis for atrial size was performed in 154 rtoF patients attending cardiovascular magnetic resonance (CMR) imaging from 2002 to 2008. Atrial area was measured on cine CMR 4-chamber view at end-systole. Clinical and echocardiography contemporaneous to CMR were collected. The endpoints were new onset of clinically documented new onset of arrhythmia (atrial and sustained ventricular tachycardia) occurring during follow-up.

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Results: Median [IQR] age at CMR was 30.8 [21.4-40.2] years. Median follow-up was 6.0 [4.6-6.9] years. During follow-up there were 3 deaths, 26 new onset arrhythmias : 16 atrial tachycardia of which 2 atrial fibrillation, 14 atrial flutters; and 9 sustained ventricular tachycardia. Atrial arrhythmia was correlated with maximal right atrial area indexed to body surface area (RAAimax) (ROC analysis, AUC 0.72 [0.64-0.79], $p = 0.003$), with a cut-off value of 16 cm^2/m^2. On survival curve with this cut off value, there is a strong difference in new onset atrial arrhythmia ($p < 0.0001$) (Figure 1). RV end-diastolic volume index (RVEDVi) was not significantly related to atrial arrhythmia but as expected correlated to PVR ($p < 0.0001$). Interestingly, the RAAimax significantly decreased after PVR ($p = 0.0001$). Patients with restrictive physiology showed no difference regarding the endpoints but they had a higher RAAimax ($p = 0.01$) and more tricuspid regurgitation ($p = 0.0007$). There was no difference in RVEDi, pulmonary regurgitation or pulmonary stenosis compared to the non restrictive physiology population.
Conclusions: RAAimax is a strong predictor of atrial arrhythmia in rtoF. Right atrial area measurement is feasible and widely available to inform clinical decision-making.

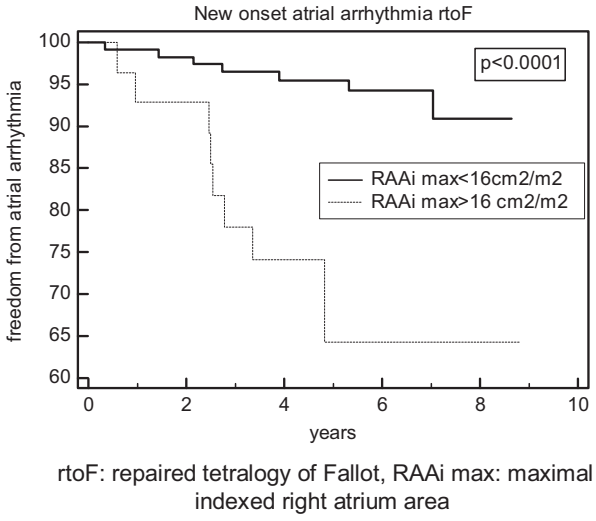

Figure 1: Freedom from atrial arrhythmias survival curve with cut off value of 16 cm2/m2 for the RAAimax.

O11-3
Closure of Atrial Septal Defect in the adult.

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Introduction: Treatment of atrial septal defect (ASD) in the adult is still controversial and with older age the likelihood of treatment is decreased. Closure affects subjective symptoms but not atrial fibrillation. The aim of this study was to investigate the effect of ASD closure in adults and especially in the elderly in our institution in a retrospective review.
Methods: Patients (N = 203) were operated for an isolated ASD at Aarhus University Hospital from 1999 to 2008. Ten were lost to follow up and thus 197 were included in the study. Hospital records were reviewed and symptoms and echocardiographic findings preoperative and at 3 months follow up registered. Patients were divided into; Group I (N = 118): less than 50 years old and group II (N = 79): more than 50 years. Results within and between the 2 groups were compared
Results: One patient (0.5%) died during follow up. Complications occurred in 18% in group I and 22% in group II. There was a relative risk reduction of 66% and 53% respectively in RV dilatation after operation. Atrial fibrillation was noticed in 6% of the young and 47% of the elderly with a reduction after treatment to 26% in group II with a relative risk reduction of 45%. Subjective symptoms occurred in 75% in group I and 99% in group II with a postoperative reduction to 43% and 67%, respectively. In group I 70% felt improvement of symptoms while it was 86% in group II.
Conclusions: Symptoms and RV dilatation is more pronounced in the elderly (>50 years) and reversibility is higher in the young (< 50 years) patients. However, the elderly benefit substantially from ASD closure. Improvement of symptoms was reported by 86%. In 50% RV dilatation was reduced after 3 months and almost half of the patients with atrial fibrillation recovered from it. ASD closure after the fifth decade is therefore recommendable.

O11-4
Long-term outcome of discrete subaortic stenosis in adults: a multicenter study

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Introduction: Discrete subaortic stenosis (DSS) is often diagnosed early in life and notable for its rapid haemodynamic progression during childhood. However, little is known about the evolution of DSS in adulthood. Therefore, our goal was to evaluate the long-term outcome of DSS, both the natural course as well as after surgical treatment in a large cohort of adults.
Methods: All adult patients with a pre-existing diagnosis of congenital fibromuscular DSS seen between 1980 and 2011 were included in this retrospective multicenter cohort study. Clinical and surgical data were obtained from chart abstraction. Patients were classified into 2 (overlapping) groups: patients who had no surgery over time (natural course group) and patients who underwent at least 1 DSS operation (surgical group).
Results: A total of 427 patients (51.3% male) were included in this study: 149 patients in the natural course group, and 313 patients in the surgical group (412 operations). Additional congenital lesions were found in 48% of patients and 63% had aortic regurgitation. Median age at baseline was 19.3 (IQR 14.5-29.0) years. Follow-up duration was 6.1 (IQR 3.0-12.4) years in the natural course group and 12.9 (IQR 6.2-20.1) years in the surgical group.Peak left ventricular outflow tract (LVOT) gradient at baseline was 32 ± 17 mmHg in the conservative group, rising to 47 ± 29 mmHg after follow-up ($p < 0.001$). Progression of LVOT obstruction was predicted by: higher baseline LVOT gradient ($p < 0.001$), greater left ventricular mass ($p = 0.001$), presence of aortic regurgitation ($p = 0.007$), younger age at baseline ($p = 0.001$) and younger age at diagnosis ($p = 0.004$).In the surgical group, the peak LVOT gradient decreased from 76 ± 28 mmHg pre-operatively to 15 ± 14 mmHg post-operatively, but increased to 31 ± 22 mmHg after follow-up ($p < 0.001$). Re-operation was predicted by higher residual LVOT gradient (HR 1.04(1.03-1.06)) and smaller post-operative aorto-septal angle (HR 0.94 (0.91-0.97)).

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Conclusions: DSS in adulthood progresses slowly in adulthood. In particular younger patients with a higher baseline peak LVOT gradient, aortic regurgitation and left ventricular hypertrophy are at risk for faster disease progression and should be monitored cautiously. Post-operatively the risk of re-operation is higher in patients with a high residual LVOT gradient and smaller aorto-septal angle.

O11-5

Ross Procedure: Prevalence and predictors of aortic autograft dysfunction and aortic dilatation in 97 patients during mid-term follow-up

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Background: As the ideal prosthesis for aortic valve replacement is controversial, the Ross procedure (RPR) using pulmonary autograft implantation remains a good alternative. However, there are concerning reports on autograft dysfunction and aneurysmal dilatation of the neo-aortic root and ascending aorta. Data on incidence and predictors of these complications are scarce.
Methods: Between 1993 and 2011, RPR was performed in 100 patients (pts; mean age 17 ± 12 years; 41 pts<14 years old), 97 of the 99 survivors (98%) had a clinical and echocardiographic follow-up after 5.6 ± 3.8 years. In 89 pts, measurement of the aortic root including Z scores were available. Z score of >4.0 defined aortic root dilatation.
Results: In 78 of the 97 pts, congenital aortic valve disease was present: bicuspid (63 pts), monocuspid (12 pts), quadricuspid (3 pts), tricuspid valves (10 pts), indeterminate morphology in 9 pts. Associated congenital heart disease included subaortic stenosis/complex left ventricular outflow tract obstruction (12 pts), aortic coarctation (9 pts). Previous surgery included aortic valve surgery in 26 and/or balloon valvuloplasty in 24 pts. Preoperative aortic dilatation was described in 28 pts (29%).
In the 97 pts RPR included concomitant reduction plasty of the ascending aorta (19 pts) and subvalvular resection of membrane (9 pts). At mid-term follow-up, moderate or severe aortic regurgitation was present in 7 pts (7%), moderate or severe aortic stenosis in 3 pts (%), and any aortic dilatation (root and/or ascending aorta) in 32 pts (33%). Median Z-score of the aortic root was 2.4 ± 1.7, of the ascending aorta 2.7 ± 1.9. In 23 of 89 pt. (26%), at least one Z score of >4.0 was observed.
Predictors of aortic dilatation were previous coarcation surgery ($p = 0.02$) and complex left ventricular outflow tract obstruction ($p = 0.04$). Reoperation on the autograft was necessary in 7 pts (7%) including autograft replacement in 5 pts.
Conclusion: Although aortic dilatation during mid-termfollow-up after RPR is very frequent (at least 26%), reoperation due to autograft dysfunction is more rare (7%). Besides assessment of the right ventricular outflow tract, careful examination of the aortic root after RPR is important, especially in patients with complex LVOT and prior cardiac surgery.

O11-6

Long term outcome of arterial switch operation performed in neonates with the transposition of the great arteries.

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Introduction: Arterial switch operation is nowadays the treatment of choice for children with transposition of the great arteries. Although the good early and mid-term postoperative results are well described, long term fate of patients still remains uncertain. The major complications occurring after surgery are supravalvular pulmonary stenosis, neoaortic regurgitation, and coronary artery insufficiency.
The aim of the study: was to assess the long term outcomes after switch operation performed in neonatal period.
Methods: Between years 1992-2011 611 patients with transposition of the great arteries have had a arterial switch operation in Cardiosurgery Department of Polish Mother's Memorial Hospital with total mortality 6,7%. From this group 172 consecutive patients were qualified for this study. The inclusion criteria were: switch procedure performed in the first 30 days of life, over 10 years of follow up in our institute and at least one full echocardiographic examination performed over 10 years after surgery. Patients with two stage operation (pulmonary artery banding prior to the switch procedure) were excluded from this study.
Results: Early and late mortality of arterial switch operations performed between years 1992-2000 was 9,7%. Mean follow up was 13,5 years(SD ± 2,4). Significant pulmonary stenosis(PG over 25 mmHG) occurred in 13 patients(7,5%). Independent risk factors for pulmonary stenosis were patch reconstruction (OR = 14,04; CI95%:4,7-41,7; $p = 0,001$) and non-facing commissures (OR = 3,96; CI95%:1,18-12,03; $p = 0,009$). Pulmonary valve insufficiency was observed in 147 patients(85%) but in majority of cases it was trivial or mild(92,5%). Neoaortic regurgitation increase with follow up to the 76% of patients (27%-trivial; 42%-mild; 7%-moderate; 0,6%-severe) at the end of observation. Significant factors for neoaortic insufficiency development were non-facing commissures (OR = 4,05; CI 95%:1,34-11,9; $p = 0,01$) and pulmonary and aortic valves discrepancy(OR = 2,05; CI95%:1,04-4,02; $p = 0,031$). Coronary anomalies were observed in 58 patients(34%) – this was mostly Cx inverted(54%). Three patients(1,7%) had LCA stenosis in routine coronarography, two of them had myocardial perfusion disturbances in exertion scintigraphy. 4 patients required reoperations (2,3%): because of recoarctation of aorta(2), LVOTO(1), and mitral insufficiency(1). In 7 patients (4,1%) percutaneous interventions was necessary because of supravalvular pulmonary stenosis(9 procedures) and recoarctation of aorta(2 procedures).
Conclusions: Arterial switch operation performed in neonatal period ensures good postoperative effect, however because of possible complications all patients still need to be followed.

O11-7

Can the echocardiography and NT-proBNP replace magnetic resonance imaging to study the right ventricle in volume overload?

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Objective: To detect in adult patients, late after repair of tetralogy of Fallot (TOF) or pulmonary atresia+VSD with volume overload, possible correlation between myocardial parameters assessed at rest by clinical status, ECG, echocardiography, serum NT-proBNP levels, cardiopulmonary test and cardiovascular magnetic resonance (CMR).

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Methods and results: The study included 50 patients (50% male, mean age 30.64 ± 13.30 years, minimum age 16 years, maximum 70 years) with prior cardiac surgery intervention of TDF (90%) or pulmonary atresia+VSD (10%). 82% of the patients had functional class according to the New York Heart Association I.
A standard examination was performed using a transthoracic echocardiogram.
The right ventricular systolic function was assessed using: the fraction of the systolic-diastolic, dp/dt, TAPSE, TDI, S wave of the tricuspid valve, right ventricular MPI, right ventricular systolic pressure, peak velocity of the wave E ', and wave A', maximum slope of systolic blood pressure and the pulmonary regurgitation index.
We performed cardiac MRI with gadolinium in 49 adult patients.
At baseline we determined also maximal exercise capacity (V'O2peak), serum NT-proBNP levels, quality of life by means of the SF-36, and the TAAQOL Congenital Heart Disease questionnaires.
The analysis of these data shows a significant correlation beetwen the TAPSE, the S wave, the fraction of the right ventricle and the right ventricular ejection fraction estimated by CMR.
A TAPSE value of 15.9 mm and S-wave value of 9.4 cm/s correspond to 45% of cardiac EF estimated by CMR.
The NT-proBNP has a negative correlation with VO2 max ($r = -0.19$, $p = 0.045$), and positive correlation with the general conditions (NYHA) ($r = 0.31$, $p = 0, 0021$), with the area of the right atrium ($r = 0.46$, $p = 0.0001$).
The QRS duration, which corresponds to the right ventricular end-diastolic volume of 150 ml/m2 is 160 ms c. Right ventricular end-diastolic area which corresponds to 150 ml/m2 is 38 mm2.
Conclusion: all echocardiographic parameters, the serum level of NT-proBNP, ECG and clinical assessment can be a valuable a support in management of this patients, but need further validation studies.

O11-8
Cerebrovascular findings on MRI compared to medical history in patients with cyanotic congenital heart disease

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Background: Patients with cyanotic congenital heart disease (CCHD) are known with an increased risk of thrombosis, especially cerebrovascular events. CCHD patients can present with various neurological symptoms due to hyperviscosity, why it can be dificult to differentate whether the patient has thrombosis or not.
Therefore the aim of this study was to examine with MRI the incidence of cerebrovascular events in CCHD patients. Furthermore to examine whether clinical reported events were similar to the MRI findings.
Methods: In a prospective study 72 clinical stabile CCHD patients were examined with MRI. The radiological findings were compared with medical information and a medical questionnaire, in order to obtain all previously relevant medical history.
Results: MRI revealed that 34 (47%) of the 72 patients had had a cerebrovascular event, and that more than one cerebral infarction was seen in 25% of the patients. According to medical history only 22% of these patients had a history of stroke. Furthermore only 58% of the patients with a medical history of stroke had an infarction on MRI.
Conclusion: Patients with CCHD seems to have a higher prevalence of cerebrovascular events than previously reported/assumed. In order to evaluate whether a patient has had a cerebrovascular event imaging should be used, since there is large discrepancy between symptoms and imaging.

PW1-1
Genetic background of long QT syndrome in infants, children, and adolescents

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Introduction: Little data are available for the prevalence of genotypes of long QT syndrome (LQTS) in pediatric patients. A school-based ECG screening program to screen cardiovascular diseases uncovered many children and adolescents with LQTS in Japan. The aim was to determine the genetic characteristics of childhood LQTS in Japan.
Methods: The study population included 102 unrelated probands (0–17 years, median: 9.8 years; M:F = 57:45) who were referred to our centers from 1993–2011. Total family members were 197. Genomic DNA was isolated from blood and direct sequencing for LQT1–LQT12 (LQT4 & LQT11 excluded) was performed. When multiple mutations were present, each was counted in each genotype. Of 102 probands, 57 were screened by the program, 35 visited hospitals because of symptoms (Symptomatic) and 13 subjects were diagnosed by family study or by chance.
Results: Genotypes were identified in 63 of 102 probands and in 100 of 197 family members. *KCNQ1* was found in 31 probands (48 family members), *KCNH2* in 19 (26), *SCN5A* in 11 (22), and others in 7 (14). The prevalence of *SCN5A* in probands (11/57) and in family members (22/91) among three main mutations was significantly higher ($p = 0.006$ and $p = 0.0004$, respectively) than for the adult population (11/192 in probands and 82/812 in family members in the literature). Of 102 probands, the screened subjects showed a higher rate of genotypic determination (40/55) than symptomatic subjects (15/34, $p = 0.01$). Conversely, the symptomatic group showed a higher rate of multiple mutations than the Screened group (4/15 vs 2/40, $p = 0.04$).
Conclusions: Clinical and genetic analysis in fetus, neonate, and infants revealed that LQT3 patients diagnosed during these periods presented critically ill and needed intensive care. A high prevalence of the *SCN5A* genotype in the pediatric population suggests progress in the medical management of these patients during infancy and childhood. School-based ECG screening and genetic testing may help prevent LQTS-related symptoms and improve order-based medicine in Japan. Numerous undetected mutations exist in symptomatic patients.

PW1-2
Cryoablation of Mahaim Pathways in Children

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Introduction: Cryoablation is a safe and effective alternative to radiofrequency ablation (RFA) for many cardiac arrhythmias. Main advantages of cryoablation include cryomapping and increased catheter stability. Mahaim pathways (MPs) are characterized by decremental atrioventricular node like conduction properties and are involved in antidromic AV reciprocating tachycardia. The aim of this study was to report the initial experience of cryoablation of MPs in children.

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Methods: Between January 2010 and November 2011, 358 patients underwent electrophysiological studies. Patients with the MPs were reviewed in this study. These patients underwent either RFA or cryoablation. A three-dimensional, surface electrode-based navigation system (EnSite NavX^{TM}, St.Jude Medical Inc., St. Paul, MN, USA) was used in all procedures.
Results: A total of 8 patients underwent catheter ablation for MPs. The median age was 10 years (range: 8-16). The median weight was 55 kg (range: 31-80). There were 6 males and 2 females. Locations of the MPs were at the right lateral (n = 2) and right posterolateral (n = 6) tricuspid annulus. Mapping was based on the finding of a specific Mahaim potential in 5/8 patients. One patient underwent successful ablation with RFA. Cryoablation was performed in 7 patients. In 2 of these patients initial attempt with RFA failed and cryoablation was successful. In 1 patient neither RFA nor cryoablation was effective. The acute success rate was 88% (7/8). A 6-mm tip cryoablation catheter was used in 2 patients and an 8-mm tip cryoablation catheter was used in 5 patients. The median fluoroscopy time was 2.9 minutes (range: 0–14.6). The median procedure time was 250 minutes (range: 170–420). There was one minor procedure-related complication. No major complications were observed. Arrhythmia recurrence was noted in 2/7 patients. One of these patients was successfully treated using an 8-mm tip cryoablation catheter resulting in a long-term ablation success rate of 84% (6/7 patients).
Conclusions: Cryoablation appears to be a safe and effective alternative therapy in children with MPs. Further studies are needed to assess the efficacy and safety of cryoablation in comparison to RFA.

PW1-3
Histopathological characterization of cryolesions at growing myocardium

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Introduction: Animal studies and clinical observations have demonstrated that radiofrequency current (RF) application at growing myocardium may result in coronary artery obstruction. Experimental data of the histopathological effects of cryoenergy application at growing myocardium is limited.
Methods: Cryoablation was performed in ten piglets (body weight 14-18 kg) at -75 °C for 4 minutes with a 6-mm-tip electrode (Freezor Xtra®) at the posterior and lateral atrial aspect of the tricuspid valve annulus. Additional cryoenergy lesions were induced at the lateral and posterior atrial and ventricular aspect of the mitral valve annulus, respectively. After 48 hours and 6 months the hearts were removed in 5 piglets for further histological work-up, respectively. The results were compared with our previous results after RF application.

	Cryoenergy [mm^3]	RF energy [mm^3]	P-value
Acute atrial lesions	52 ± 45	49 ± 14	Non significant (N.s.)
Chronic Atrial lesions	40 ± 20	27 ± 5	N.s.
Acute ventricular lesions	144 ± 203	150 ± 50	N.s.
Chronic ventricular lesions	192 ± 118*	97 ± 90*	P < 0.05

Results: In contrast to RF lesions, acute and chronic cryolesions were sharply demarcated, no affection of the adjacent coronary artery was noted. As RF lesions, the acute hemorrhagic cryolesions were bordered by a layer of neutrophile leucozytes. Thrombus formation was noted in 2/3 of the acute lesions. In chronic cryolesions myocardium was replaced by fibrous tissue in the upper endocardial layer and by fat cells in the deeper layers. Lesion size is displayed in the table:
Conclusions: Due to these results cryoenergy appears favorably in tachycardia substrates close to the coronary arteries. However, the increase of ventricular lesions size over time after cryoenergy long-term effects of cryoenergy application should be further evaluated.

PW1-4
Sensing malignancy: Prodromal signs of post-transplant lymphoproliferative disease in pediatric heart transplant recipients

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Background: Post-transplant lymphoproliferative disorder (PTLD) contributes to morbidity and mortality after transplantation. Early and adequate diagnosis can be challenging due to unspecific and heterogeneous clinical presentation. The aim of this review was to determine characteristic prodromal signs of PTLD development.
Methods: Medical records of 106 pediatric heart transplant patients at our institution were retrospectively reviewed for PTLD development, their clinical course including B symptoms, lymphoma-like changes in blood cell counts, EBV antibody titers and immunosuppression levels prior to PTLD diagnosis.
Results: Between 1990 and 2010 seven of 106 children (6,6%) developed PTLD within five month to fourteen years post heart transplantation (HTx) (mean 7,7 ± 5,1 y). Biopsies revealed polymorphic B cell lymphoma (five), monomorphic diffuse B cell lymphoma (one) and plasmacytic hyperplasia (one). Five of seven recipients diagnosed with PTLD were under one year of age at time of HTx and all but one (6/7) were initially EBV seronegative. Our patients presented from four month to two days before diagnosis with recurrent fever of unknown origin (5/7), decrease in general performance (7/7) and lymphadenopathy (4/7, 4 cervical, 1 submandibular, 2 axillary). Lab results showed rising EBV-antibody titers (5/7, two cases EBV-negative even after PTLD diagnosis), increasing lactate dehydrogenase (LDH) levels up to 447-1072 U/l (4/7) and remarkable dropping in blood cell counts (microcytic anemia and leukopenia, 4/7). Immunosuppression regimen and drug levels showed no relevant changes before PTLD onset.
5/7 cases were treated successfully with reduction of immunosuppression (and/or conversion to everolimus /monotherapy) and chemotherapy (NHL-BFM 95, Ped-PTLD 2005). One case with monomorphic lymphoma showed low response to Rituximab and developed a relapse of PTLD. An older recipient died of pulmonary deterioration during chemotherapy.
Conclusions: Tight follow-up with a close look for PTLD prodromal signs in anamnesis and clinical examination like recurrent fever of unknown origin, lymphadenopathy and reduced general condition, is crucial for patients at risk for PTLD, especially in the infant recipient group. Rising EBV-load and correlating changes of blood cell counts and LDH peaks are further hints for a looming PTLD development.

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PW1-5

The three year expierence in treatment of dilated cardiomyopathy by bone marrow derived cell intramyocardial implantation in childhood

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Introduction: Dilated cardimyopathy is a serious disease in pediatric age due limited conservative therapy and heart transplantation as a final option. Due to the lack of transplantation we have looked for another possibilities. Bone marrow derived progenitor cell (BMCs) transplantation is becoming a promising method of treatment in adult population and there are a few cases in pediatrics described. Based on this, we have done the BMCs transplantation in seven patients since May 2009.
Methods: The seven patients had been admitted for the BMCs transplantation in age 4 month to 17 years. The inflammatory basis was excluded by laboratory investigations. Seventeen to ninety million BMCs were isolated and as suspension of physiologic saline with heparin given to patients by intramyocardial injection in the interventricular septum under echo control. There is complete 1 year follow-up for six patients. The results of method were controlled by echo EF measurements by Simson's, ECG changes, CTR at X-ray and NT-proBNP levels. The data analysis was made by descriptive and mathematical statistic methods. The statistical significance was determined by t-Test ($p = 0.05$).
Results: One year following BMCs transplantation we observed increase of ejection fraction, The average basal EF was 33.5%. We observed increasing up to 54% (=9.54, $p = 0.00154$) in 6 month period and up to 54,5% (=10.82, $p = 0.00315$) after one year. The difference between basic data and results after 6 and 12 months was statisticaly significant, but there was no difference between 6-month and 12-month data. The results were within the confidence interval in all measurements. Also there was correlation observed in ECG and echo improvement and CTR and BNP level decrease.
Conclusions: We found the improvement of patient's condition after intramyocardial administration of bone marrow cells. We can do the first judgement abuot efficiency of procedure after six month evaluation.

PW1-6

Aetiology, Presentation, and Outcomes of Hypertrophic Cardiomyopathy in Childhood in Wales

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Background: Hypertrophic cardiomyopathy (HCM) is commonly caused by a mutation in a sarcomeric protein, or secondary to a neuromuscular disorder, malformation syndrome or inborn error of metabolism. The purpose of this study was to establish the causes, management and outcomes of HCM in the Welsh paediatric population.
Methods: Patients diagnosed with HCM in Wales between 1984 and 2011 were included in the review.
Results: Of 58 patients diagnosed with HCM, 17 (29.3%) cases were familial while 41 cases (70.7%) were secondary to another cause. 21 of these secondary cases (36.2%) resolved with treatment while the remaining did not. The most common identification of familial HCM was through genetic testing of patients with a family history, with the majority (64.7%) being diagnosed over 15 years old. While secondary HCM was most commonly identified by neonatal screening with 80.5% being diagnosed during infancy. All patients with non-resolving secondary HCM had an abnormal baseline ECG, abnormal echocardiogram or both. Of the patients with familial HCM three (17.6%) of genotype positive patients had completely normal investigations. Five patients (29.4%) with familial HCM received an ICD to prevent sudden cardiac death (SCD). The four (6.9%) patients from the secondary HCM group died; only two of these deaths were cardiac related, one as a result of heart failure, the other due to arrhythmic SCD.
Conclusions: Despite the eminent risk of SCD in patients with familial HCM, no patients in this cohort died as a result of this and most remained asymptomatic. The prognosis for secondary HCM was highly variable depending upon the cause. Those born to diabetic mothers had an excellent prognosis with all cases resolving in time; in contrast those secondary to metabolic diseases had much poorer outcomes.

PW1-7

The natural history of patients with scimitar syndrome: an Italian multi-centric study of the Italian society of pediatric cardiology

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Objective: Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies. Surgical correction of this malformation is usually performed in symptomatic patients or in patients with an increased pulmonary blood flow. The aim of this study was to analyze the natural history of patients with scimitar syndrome who didn't require surgery.
Materials: Between January 1990 and December 2011, 36 patients from 7 Italian centers with diagnosis of scimitar syndrome who didn't require surgical correction were included. Excluded were patients with associated severe forms of congenital heart disease. Primary outcomes included the evaluation of their clinical status at follow-up.
Results: Median age of patients at diagnosis was 9 months (range 1 day-41 years). There were 20 females and 16 males. Twenty-one patients (58%) were symptomatic at the time of diagnosis (recurrent respiratory infections in 16 patients, congestive heart failure (CHF) in 10 patients, cyanosis in 1 patient, emophtoe in 1 patient. Seventeen patients (48%) had associated cardiac anomalies mainly including an atrial septal defect type in 13 patients, a ventricular septal defect in 5 patients and a patent ductus arteriosus in 5 patients.

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Thirty patients underwent cardiac catheterization (84%). The mean pulmonary artery pressure was 26 mmHg (range 13-50 mmHg) and the mean Qp:Qs ratio was 1,6:1 (range from 1:1 to 3:1). Systemic arterial supply to the right lung was demonstrated in 20 patients (67%); 15 of which were treated by coil embolization (50%) (Table 1). Patients presenting with CHF have a higher mean pulmonary artery pressure and had a higher association to cardiac anomalies other than atrial septal defect ($p < 0.05$).
Median age at last follow-up check was 7,5 years (1month-46years). One patient died at the age at 14 months for severe untreatable pulmonary hypertension. At the last clinical check, 18 patients are still symptomatic (51%) without reported worsening of symptoms and 15 patients (43%) are asymptomatic.
Conclusions: Scimitar syndrome is not a benign congenital heart malformation, especially when associated to other congenital heart disease and pulmonary artery hypertension. However it can present as an isolated lesion in almost half of the patients which in the majority of cases are asymptomatic.

PW1-9
Comparison of the effect of inhaled with intravenous anaesthetic on pulmonary vascular resistance measurement at cardiac catheterisation
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Introduction: Children with pulmonary hypertension (PH) require assessment of pulmonary vascular resistance (PVR) to determine the effectiveness of medical therapy and whether cardiac surgery is safe to perform. It is crucial that anaesthetic drugs have minimal effect on PVR. Propofol has been evaluated in children with congenital heart disease, and isoflurane in adults with PH, but the effect of these anaesthetic drugs on haemodynamics in PH is unknown. We aimed to undertake a randomised cross-over pilot study of 10 children to evaluate the equivalence of isoflurane and propofol on PVR in children with pulmonary hypertension
Methods: After ethical and MHRA approval we recruited 10 children undergoing cardiac catheterisation for assessment of PVR. All children had PH (tricuspid regurgitant velocity of more than 2.8 m/s) with evidence of raised PVR and were randomised to receive propofol or isoflurane. Blinded measurements were taken at baseline, response to Nitric oxide (NO) 10 parts per million (ppm), NO 20 ppm, and NO 20 ppm with 100% oxygen. Baseline measurements were repeated using the other anaesthetic drug after wash in/wash out of 15 minutes. BIS (alertness) monitoring was used to reduce the risk of awareness.
Results: 10 children were studied, median age 20 months, median weight 9.5 kg.6 children received propofol (Pro) then isoflurane (iso), 4 children received Iso then Pro. There was no significant difference in baseline PVR (Iso: 3.5 U.m2, Pro: 3.2 U.m2, $p = 0.16$) or SVR (systemic vascular resistance) (Iso: 14.1 U.m2, Pro: 16.4 U.m2, $p = 0.24$) or shunt fraction (Qp/Qs) (Iso: 0.32, Pro: 0.32, $p = 0.86$) when measured under isoflurane or propofol anaesthesia. There was no significant difference in BIS values (Iso: 50, Pro: 47, $p = 0.12$) or lung compliance (Iso: 11.8 ml.cmH2O, Pro: 11.6 ml.cmH2O, $p = 0.64$) between isoflurane and propofol anaesthesia. There were no adverse incidents.
Discussion: Propofol and isoflurane are equivalent with respect to their effects on PVR in children with pulmonary hypertension. The effects on SVR and shunt fraction are also equivalent. This pilot study suggests that either can be used for the assessment of children with PH.

PW1-10
Clinical classification of congenital heart disease associated pulmonary hypertension. Does it work for pediatrics? Analysis of the TOPP registry (Tracking outcome and practice in pediatric pulmonary hypertension)
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Objectives: Current guidelines for pulmonary hypertension (PH) include a subclassification of congenital heart disease (CHD) into 4 groups: Eisenmenger (A), PAH-CHD associated to left to right shunt and high pulmonary vascular resistance (PVR) (B), PAH with small septal defects (C) and PAH after corrective surgery (D). We used a pediatric PH registry (TOPP) to evaluate the feasibility of classifying the pediatric patients.
Methods: TOPP was designed before publication of these guidelines and did not use the PAH-CHD subclassification. However, specific details were collected to describe the anatomy, hemodynamics and surgical status. Two investigators reviewed independently all data and classified the patients. When agreement was obtained the patient was coded as A, B, C or D. If there was a discrepancy a consensus was reached by discussion. If consensus was not obtained, the patient was considered "not classified" (NC).(A) was defined as a large unrestrictive shunt defect with a Sa O_2<90%, (B) as a defect with SaO_2 >90% and indexed PVR >3 WU*m^2, (C) as small defects as described by the investigator or moderate defects and patient considered too young for the development of pulmonary vascular disease and (D) as PAH after complete repair .
Results: Demographics, anatomy and indexed PVR are shown in the table. 27 patients diagnosed as idiopathic PAH had some type of CHD and were reclassified in (B) (3/27), (C) (19/27) and (D) (5/27).The largest group was PAH after corrective surgery 48/142 but a significant number of patients were not classified(NC) 18/142.

Group (n)	A (24)	B (25)	C (27)	D (48)	NC (18)
Age (years) Mean ((SD)	10.0 (6.3)	7.2 (5.9)	6.0 (5.4)	8.1 (4.9)	6.4 (4.9)
ASD	7	7	18	13	6
VSD	17	15	6	27	6
PDA	5	7	9	14	7
AVC	2	2	0	4	1
Other	3	5	3	3	4
PVRi WU*m2 Mean (SD)	21.0 (8.6)	13.2 (6.6)	14.4 (8.9)	17.1 (13.3)	15.5 (7.5)

Conclusions: The subclassification of PAH-CHD seems feasible for pediatric patients in group A, B and D but was difficult to apply to group C with the current description of absolute defect size which does not always relate to children. The largest group was D, characterized by high PVR raising concerns about the appropriateness of surgical repair. Several participating sites classified patients with very small shunts as idiopathic PAH. In addition several patients with complex CHD or patients who never had shunts were difficult to classify. A more tailored description appears necessary in particular for small defects or complex forms of CHD.

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PW1-11

Circulating Endothelial Cell Levels Decrease after Vasodilator Therapy and are a Biomarker of Deterioration in Pediatric Pulmonary Hypertension.

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Background: Pulmonary vasodilators in general and prostacyclin therapy in particular have markedly improved the outcome of patients with pulmonary arterial hypertension (PAH). Endothelial dysfunction is a key feature of PAH and we previously described that circulating endothelial cells (CECs) could be used as a biomarker of endothelial dysfunction in PAH. We now hypothesized that PAH-specific vasodilator therapy might decrease CEC numbers.
Methods: We quantified CECs in peripheral blood from children with idiopathic PAH (iPAH, n = 30) or PAH secondary to congenital heart disease (PAH-CHD, n = 30), before and after treatment and during follow up. CEC were enumerated by immunomagnetic separation with mAb CD146-coated beads.
Results: CEC counts were significantly decreased in children after treatment with oral endothelin antagonists and/or PDE5 inhibitors. In 10 children with refractory PAH despite combination oral therapies, SC treprostinil was added and we found a further significant decrease in CEC count during the first month of treatment in every patient. We quantified CEC during 6 to 36 months follow-up after initiation of SC treprostinil and found that CEC count is modified according to clinical status.
Conclusions: CEC counts fall with vasodilatator therapy in PAH and could also be used as biomarker of deterioration in refractory pediatric pulmonary hypertension treated with SC treprostinil.

PW1-12

Hospitalization for Lower Respiratory Tract Infection Increases the Risk of Childhood Respiratory Morbidity Among Children with Congenital Heart Disease

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Introduction: Congenital heart disease (CHD) is a risk factor for severe respiratory syncytial virus (RSV) lower respiratory tract infection (LRTI) in infancy. Whether infants with CHD are at increased risk of RSV-related sequelae (e.g. asthma) is unknown. The objective was to estimate the incidence and increased risk of chronic respiratory morbidity among CHD infants hospitalized for LRTI.
Methods: A retrospective population-based study was conducted using the Régie de l'Assurance Maladie du Québec administrative databases. Children <2 years of age with CHD (by ICD-9 code diagnoses on physician billing or hospital discharge abstracts) born in 1996 or 1997 were included. Hospitalizations for LRTI before age 2, and chronic respiratory morbidity (asthma, chronic bronchitis, or chronic lung disease) before age 10, were identified by ICD codes. Incidence rates and incidence rate ratios (IRRs) with 95% confidence intervals (CIs) comparing risk among LRTI-hospitalized and non-hospitalized CHD children were calculated. Logistic regression models estimated the adjusted odds ratio (95% CI) of chronic respiratory morbidity based on hospitalization for LRTI in infancy, among CHD children without other known risk factors.
Results: Of the 3,223 CHD children, 19 (0.6%) and 417 (12.9%) were hospitalized for RSV or LRTI, respectively, before 2 years old. Before age 10, 58.5% (244/417) of CHD children who were hospitalized for LRTI in infancy were diagnosed with chronic respiratory morbidity; compared to 31.5% (884/2,805) of CHD children not hospitalized for LRTI in infancy. IRRs for chronic respiratory morbidity were 2.0 (1.8-2.2) and 1.6 (1.3-1.9), for males and females aged 2-10 years, respectively. The adjusted odds of developing chronic respiratory morbidity after LRTI hospitalization was 3.0 (2.3-3.9); and of hospitalization for chronic respiratory morbidity after LRTI hospitalization was 5.7 (4.0-8.1).
Conclusions: CHD children hospitalized for LRTI in infancy are at almost twice the risk of childhood chronic respiratory morbidity, compared to CHD children not hospitalized for LRTI. Among CHD children, LRTI hospitalization was associated with a 3-fold increase in the risk of childhood chronic respiratory morbidity – and a 6-fold increased risk of hospitalization. The impact of LRTI hospitalization is therefore not limited to the perinatal period among CHD infants; but extends throughout childhood.

PW2-1

Laser capture microdissection and comparative microarray expression analysis identified vessel-specific molecular markers of the ductus arteriosus and aorta in fetal rats

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Introduction: Closure of the ductus arteriosus (DA) is a crucial step in the transition from the fetal to the postnatal circulation. Patent DA is one of the most common cardiovascular anomalies in children causing morbidity especially in premature infants. Little is known about the molecular mechanisms regulating the unique remodeling process of the DA.
Objective: We aimed to identify genes that specify the DA in the fetus and differentiate it from the non-closing aorta.
Methods and results: Comparative microarray analysis of laser-captured microdissected endothelial (ECs) and vascular smooth muscle cells (SMCs) from the DA and aorta of fetal rats identified vessel-specific transcriptional profiles. The linear model of microarray analysis (LIMMA) revealed a strong age-dependency of gene expression in the samples from embryonic day 18 and 21. Among the DA-dominant genes the regulator of the G-protein coupled receptor 5 (*RGS5*) and the homeobox transcription factor *DLX1* exhibited the highest and most significant level of differential expression. The aorta showed a significant preferential expression of the Purkinje cell protein 4 *(PCP4)* gene. The results of the microarray analysis were validated by real-time quantitative PCR. Finally, immunohistochemistry at day 21 documented

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differential expression of the proteins encoded by the three newly identified genes in DA and aorta.
Conclusion: In conclusion, our study confirms a DA and aorta-specific transcriptional profile in ECs and SMCs. For the first time we recognized the preferential expression of *RGS5* and DLX1 in the fetal DA. These genes may represent novel molecular targets for the regulation of fetal DA maturation and postnatal DA closure.

PW2-2
Pediatric Usage of a Newly Available Handheld (Einthoven Lead I) Web Based Event Recorder [ER] (Zenicor EKG-2 TM): First Results
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Introduction: ER recordings in children are difficult due to direct positioning of the device on the chest, limited storage capacity and difficult technical handling. Recently a hand held, thumbs activated ER has become available using GSM network messaging after automatic recording and a web based analysis tool.
Purpose of the study: To investigate Zenicor EKG-2 ER [ZER] in pediatric patients [pts].
Methods: ZER ECGs in pts with and without tachycardia, palpitations, pacemakers and ICDs were performed and compared with 12 lead ECGs.
Results: 100 ECGs in 98 pts were recorded (male: n = 49; structural heart disease: 54%) in pts with sinus rhythm [NSR] (age 0-11 month [INF], n = 20; age 1-4 yrs. [TODDL], n = 20; age 5-17 yrs. [CHILD], n = 20), paced rhythm [PACE] (n = 20) and in tachycardia [TACHY] (n = 20). Successful ZER recording and data transmission was possible in all cases and considered easy or very easy in 92.6%. In 96% QRS complexes were visible and heart rate could be calculated. R was 0.975 ($p < 0.001$) for matching of ventricular cycle length. P wave detection was possible in 82.1%. 24% of the pts had arrhythmia, which could be detected by ZER in 95.8%. For NSR P-wave detection rate was significant lower in CHILD as compared to TODDL, but not to INF. For heart rate detection there was no difference between these subgroups. For PACE, detection of pacing mode was possible in 70% for AAI/AVI and in 99.3% for VVI, VAT or DDD pacing. For TACHY, SVT (n = 13) as well as sinus- or atrial tachycardia (n = 6) could be detected in all cases.
Conclusion: 1. ZER recording is technically easy for children of all age groups. 2. Data transmission is perfect and web analysis can be almost always obtained just minutes after the ECG recording has been taken. 3. Heart rate detection is possible in >95% for NSR in all children, as for paced rhythms and tachycardia. 4. Ventricular pacing can be identified nearly always. 5. Tachycardia detection is excellent and even classification of tachycardia is possible. 5. P wave and atrial pacing detection remains challenging, esp. in the older age group.

PW2-3
Long-term survival after paediatric cardiac surgery in patients with congenital heart defects
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Objective: To collect reliable and complete data for evaluation of long-term survival after paediatric cardiac surgery.
Methods: All 2439 patients (1125 girls/1314 boys) operated due to congenital heart defects in our institution before the age of 18 years from Jan 1st 1994 to Jan 1st 2009 were included. During the study period nearly 50% of all paediatric cardiac surgery in Sweden was performed at our institution. Patient files were cross-checked as of Jan 1st 2012, against the National Population Registry in Sweden, allowing for reliable and complete data on long-term survival. 23 patients (0.9%) emigrated during the study period and were lost to follow up.
Results: Median age at first surgery was 0.35 years (0-17.8 years). 2135 patients (87.5%) had surgery for biventricular correction and 304 patients (12.5%) had univentricular heart palliations. Median age of survivors at follow up was 12.9 years (3-34.7). 184 deaths (7.5%) occurred with a median age at death of 0.5 years (0-30.1). Median survival time in the deceased patients was 51 days (0-17.1 years) after the last major surgical procedure. 101 deaths (4.7%) occurred after surgery for biventricular correction and 83 (27.3%) after palliation for univentricular heart defects ($p < 0.001$). 51/109 (46.7%) patients died after surgery for classic hypoplastic left heart syndrome or unbalanced atrioventricular septal defect with left ventricular hypoplasia, compared to 32/195 (16.4%) after surgery for other univentricular heart defects ($p < 0.001$). 22 patients had a heart transplantation of whom 6 (0.3%) had previous biventricular and 16 (5,2%) previous univentricular heart surgery. Seven late deaths occurred in patients who received a heart transplant (32%).
Conclusions: Total survival from birth was 92.5%, with more than 50% of all deaths occurring later than 30 days after the last major surgical procedure, reflecting the need for long-term follow up. Survival after surgery for biventricular correction was 95.3%, compared to 53.3% after surgery for classic hypoplastic left heart syndrome or unbalanced atrioventricular defect with left ventricular hypoplasia and 83.6% after surgery for all other types of univentricular heart patients.

PW2-4
Evidence of Seasonality in Births of Patients with Congenital Heart Disease Who Require Subsequent Surgical Repair
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The Hospital for Sick Children, Toronto, Canada

Introduction: Patients born with multiple congenital defects are known to occur in seasonal clusters. Anecdotal and limited evidence of such an effect for patients with congenital heart disease (CHD) has been reported. We sought to determine whether such seasonality might be observed for patients born with CHD who require subsequent surgical repair, and to explore variations across major anatomical types.
Methods: All patients who underwent cardiac surgery for a CHD between 2002 and 2010 were included. Patients were classified into major anatomic groups based on their primary diagnosis. Duplicate entries (i.e. multiple surgeries on the same patient) were removed. The total number of births per month associated with each diagnosis was calculated. This was compared to the expected number of births for each month using moving average methodology adjusted for natural monthly variation in the birth rate as determined by Census Canada population statistics. The relative difference between the observed and expected number of CHD cases (ratio) for each calendar month was calculated and compared to the null hypothesis of no seasonal effect.
Results: N = 3,775 patients were included. For all patients, the ratio of observed and expected number of CHD cases ranged from 0.87 in February to 1.10 in May; this was statistically

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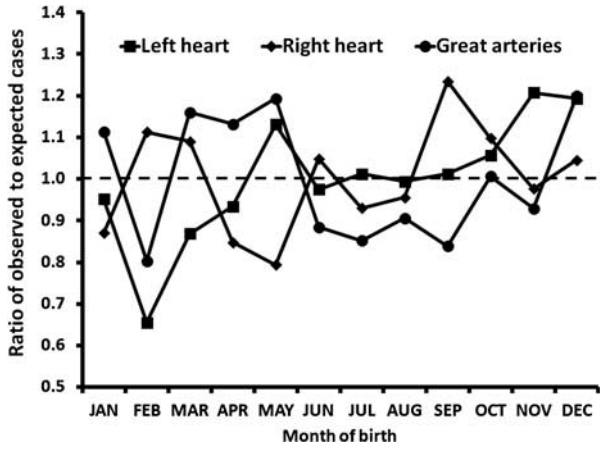
significant for February (lower than expected, $p = 0.002$), November and December (higher than expected, $p = 0.03$ and $p = 0.008$ respectively). Seasonal variation was significantly associated with anatomical type of CHD. For patients with left heart lesions and septal defects (N = 1,716), February and March had a deficit of cases compared to expected, while May, November and December were associated with an excess of cases. For right heart lesions (N = 463), April and May were associated with a deficit of cases, while excess cases were noted in September. Finally for patients with abnormalities of the great arteries and the aorta (N = 742), February and June-September were associated with a deficit of cases while December and March through May were associated with an excess of cases.



Conclusions: Differences in seasonal patterns between the 3 major anatomical groups of CHD exist, and may indicate a potential etiologic effect of environmental influences or possible gene-environment interactions.

PW2-5

Social and health related coping strategies of parents of children with operated congenital heart disease

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Giving a birth of a child with congenital heart diseases (CHD) profoundly affects the whole family system. Parents have to learn how to live in a different situation and to adapt their life style to the new conditions.

The aim of the research is to study the specific active or passive types of social and health related coping strategies, which parents of CHD operated children use.

Methods: A special four area (health related, social, family and psychological) questionnaire was developed.The health related sphere includes medical information sources; knowledge about the disease, it's treatment and follow up; changes in the family life, after having a "cardiac" child. The social sphere involves: changes in friendship circle; upbringing of the child; and changes in the parental employment status. Active coping is related with a direct and rational approach, whereas passive coping involves avoidance, withdrawal and wishful thinking.

Target group consists of 109 parents of 68 children with at least one open-heart operation, aged 3 to 14 years, 33 with "simple" and 35 with "complex" CHD. The analysis was made on the basis of the demographic data.

Results: The health related area analysis revealed that parents from the capital and cities significantly more often use active coping strategies compared to the parents from towns and villages: more than one source for information ($p = 0.003$); change in attitude to the child ($p = 0.05$).

Parents of the children with "simple" CHD more often use passive coping strategies. There is a tendency for a change in the strategy type from active to passive during the progress of the disease and duration of the follow up.

The social sphere analysis demonstrates that most of the parents use active strategies: change in the friendship circle (60%) and no change in the employment status (76.1%). The way of upbringing a CHD child depends mainly on the disease severity and the parental residency.

Conclusions: The parents of CHD children need special support, training and education for acquiring new and different skills for coping with the stress, caused by child's disease. Those needs are higher among parents from villages and towns.

PW2-6

Reduced pulmonary diffusing capacity is highly increasable in Fontan patients

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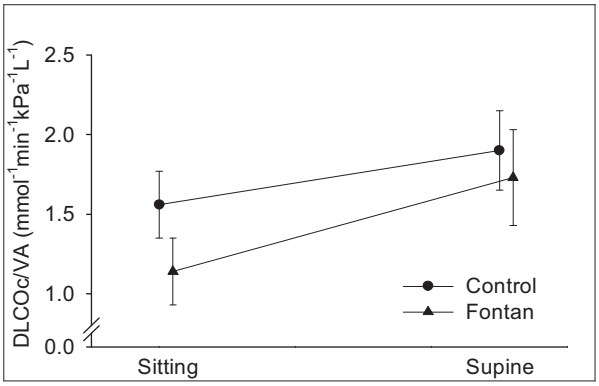
Introduction: Fontan patients have reduced pulmonary diffusing capacity, however the cause remains unclear. We aimed to assess the etiology of this reduction, and in particular to distinguish between reduced alveolar capillary membrane diffusing capacity and/or reduced pulmonary capillary blood volume. Furthermore, we aimed to assess potential reversibility of the reduced diffusing capacity and to search for independent predictors of reduced diffusing capacity.

Methods: Advanced pulmonary function test were performed in the sitting position in 87 Fontan patients (mean age: 16.3 years, SD 7.6 years) using the single breath method. Of the 87 patients, 72 patients performed a symptom-limited cardiopulmonary exercise test. Before the exercise test cardiac index and stroke volume index were measured at rest using the inert gas-rebreathing method. Furthermore ten Fontan patients and nine healthy matched controls performed a supine pulmonary function test after a supine rest.



Results: The single breath test showed a mean diffusing capacity corrected for concentration of hemoglobin and alveolar volume (DLCOc/VA) of 70.3% of predicted, equal to a z score of minus 2.38. Alveolar capillary membrane diffusing capacity was normal (z score: minus 0.14) while pulmonary capillary blood volume was reduced (z score: minus 2.04). In a multiple linear regression analysis cardiac index at rest significantly predicted DLCOc/VA (regression coefficient: 0.18, $p < 0.001$). In the supine compared

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to sitting pulmonary function test, DLCOc/VA increased 51.8% in Fontan patients compared to 23.3% in the control group ($p < 0.001$) (Figure). Pulmonary capillary blood volume increased 48.3% in the Fontan group compared to 20.2% in the control group ($p = 0.001$) as well as alveolar capillary membrane diffusing capacity (14.1% in the Fontan group compared to 6.6% in the control group, $p = 0.008$).
Conclusions: We found a marked reduction in diffusing capacity in Fontan patients. In assessment of the etiology of this reduction we found a reduced pulmonary capillary blood volume while function of the alveolar capillary membrane appeared normal. The diffusing capacity was highly increasable in Fontan patients compared to a healthy control group, mainly due to increase in the pulmonary capillary blood volume. Cardiac index at rest was a highly significant independent predictor of reduced diffusing capacity.

PW2-7
Structural brain changes in teenagers with essential arterial hypertension
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Objective: To study the prevalence of structural changes of brain with the help of MRI data in teenagers with essential AH and to assess their connection with data of 24-hour blood pressure monitoring.
Methods: 150 youths with essential AH at the age of 12-18 years old were examined. Average age of patients was 14,9 ± 2,0 years old. The control group involved 12 healthy youths comparable with the examined group by sex and age. Every patient was made BP daily monitoring (BPDM), according to results of which study groups were formed: 1 group – patients with phenomenon of "white coat hypertension" (WCH) – 44 persons; 2 group – youths with labile atrial hypertension (LAH) – 50 persons; 3 group – stable atrial hypertension (stable AH) – 56 persons. Magnetic resonance imaging (MRI) of brain was made by tomography Magneton – OPEN (Simens AG, Germany).
Results: Linear sizes enlargement of liquor contained structures of brain was disclosed in 74% of pts in the study group. Difference of average size figures of subarachnoid cavity (SAC) of postcranial fossa in all study groups in relation to control as amended to sex and age was clinically significant. In the group of teenagers with stable AH SAC size of convexital brain space were more than in healthy counterparts ($p = 0{,}031$). 0.14 mm increase (0,02:0,46), $p = 0{,}032$) of SAC size of posterior cranial fossa is marked with 1 mm Hg increase of average BP daily. 1% increase of time index of systolic BP at nights causes the same parameter increase in 0,09 mm (0,01:0,16), $p = 0{,}028$).
Conclusion: Sizes enlargement of liquor contained structures of brain can be interpreted as early markers of its disturbances along with essential AH formation in teenagers. Signs of average BP daily and time index of SBP at nights make a great contribution into formation of structural brain disturbances in the form of enlargement of subarachnoid cavity of occipital zone, which is initial substrate in the chain of disturbances of liquor contained areas with BP increase.

PW2-8
Exercise Training Improves Activity and Psychosocial Wellbeing in Adolescents with Congenital Heart Disease (CHD)
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Recently, exercise training has emerged as a method of improving activity and psychological health in some patient groups and adolescence may be an ideal opportunity to introduce such positive lifestyle changes. We aimed to ascertain if motivational techniques and a structured exercise program could increase activity and improve wellbeing in patients with CHD.
Patients aged 12-20 years were identified using the Northern Ireland regional database (HeartSuite). Participants completed standard psychological questionnaires and underwent evaluation of exercise ability (formal exercise stress testing and measurement of free-living activity using an ActiGraph accelerometer). Following randomisation the intervention group attended an activity day where they were given a personal exercise programme. The control group received their usual level of care. Patients were followed up at 6 months for reassessment and results obtained were analysed using parametric methods.
One hundred and forty three patients (mean age 15.6 years) consented to participate, 86 were male (60%) and 105 had major CHD (73%). Psychological health appeared well preserved at baseline with few differences across study groups. On formal exercise testing, complex patients performed worse at peak exercise. However, patients with major CHD had significantly higher activity counts. 101 (71%) attended for reassessment. There was a significant increase in duration of exercise test (Pillai's Trace 5.34 ($p < 0.05$)) and average activity counts per minute (Pillai's Trace 46.55 ($p < 0.001$)) for the intervention group at reassessment. This group also had trends toward improved mood and self esteem.
An exercise program to promote activity and healthy lifestyle is both feasible and beneficial for young people with CHD. Exercise training significantly improves peak exercise capacity and free-living activity in this group. Increased activity also appears to have a positive effect on self-esteem and mood parameters. Future interventions targeted around this area may considerably improve outcomes for this population and should be incorporated into formal transition programs.

PW2-9
Quality of life in school-age survivors of severe congenital heart disease: results from the UK Collaborative Study of Congenital Heart Defects (UKCSCHD)
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Introduction: As mortality due to congenital heart defects (CHDs) falls, long-term outcomes including health-related quality of life (QoL) attain greater importance. Paediatric QoL has been measured in cardiac outpatient populations using the PedsQL™ questionnaire and impaired psychosocial QoL scores demonstrated. However, previous studies were often limited to instrument validation, comparisons with reference norms or specific CHD types.
Our aims were to (1) compare QoL scores, for a UK-wide cohort of children born with serious CHDs, with a unaffected classmate comparison group and (2) investigate factors predicting lower PedsQL™ scores that may be modifiable through improving care.
Methods: Children aged 8-12 years were recruited through the UKCSCHD cohort, a multi-centre study of CHD outcomes

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involving all 17 UK paediatric cardiac centres. Each child (and their parent) was sent a generic PedsQL^{TM} 4.0 questionnaire and was asked to give a questionnaire to two age- and sex-matched classmates. Information on children with CHDs was also available from a case-note review.
Characteristics of respondents with CHDs were compared with all cohort survivors and unaffected children. Using Generalised Additive Models for Location, Scale and Shape (GAMLSS) based on the sinh-arcsinh (SHASH) distribution, univariable and multivariable models were constructed to investigate predictors of PedsQL scores, including individual and family factors, illness severity and participation in peer activities.
Results: Data were obtained from 479 affected children (271 boys; mean age 12.1 years) and 467 unaffected children (256 boys; mean age 12.0 years). Compared with unaffected peers, children with CHDs reported significantly lower QoL scores on the PedsQL^{TM} physical, psychosocial functioning and total scales in univariable models ($p < 0.001$). In univariable models, predictors of lower QoL scores in children with CHDs included measures of cardiac severity, non-cardiac illness and special educational needs.
Discussion: Children with CHDs in this prospective multi-centre study reported significantly lower physical as well as psychosocial QoL compared to their healthy classmate peers. Clinical management should seek to optimise children's ability to participate fully in peer activities, particularly within the school environment.
Funding: British Heart Foundation PG/02/065/13934.

PW2-10
Truncal valve function in children following primary correction of common arterial trunk. A long term longitudinal single centre study
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Kardiocentrum, University Hospital Motol, Prague, Czech Republic (1); Great Ormond Street Hospital for Children and Institute of Child Health, London, Great Britain (2)

Objectives: To identify predictors of development of severe truncal valve insufficiency (TrVI) after primary correction in infancy.
*Methods and Results:*A retrospective longitudinal echocardiographic study of 61 consecutive patients after primary correction of common arterial trunk in infancy. Severe TrVI was observed before repair in 8 (13%) out of 61 patients. The mean Z–score of truncal valve annulus and sinuses dimension in patients with severe preoperative TrVI compared to patients with absent to moderate TrVI was 7.6 ± 0.7 vs 6.4 ± 1.5; $p = 0.03$ and 7.1 ± 2.0 vs 5.2 ± 1.6; $p = 0.003$ respectively. The truncal valve was subsequently replaced in 9 out of 50 early survivors with actuarial freedom from the valve replacement 91%, 78% and 69% at 5, 10 and 15 years after the correction. Severe TrVI developed in 10 out of 40 patients with absent to moderate TrVI postoperatively, the actuarial freedom from severe TrVI was 89%, 85% and 53% at 5, 10 and 15 years after the correction. The major independent predictor of postoperative development of severe TrVI was the presence of disproportional enlargement of truncal annulus (HR 11.6; CI 1.4–95.2; $p = 0.023$) or sinuses (HR 10.1; CI 1.3–83.9; $p = 0.033$).
*Conclusion:*The truncal valve insufficiency after successful correction in infancy is progressive in patients with the disproportional truncal root enlargement.
Supported by Ministry of Health of the Czech Republic, grant No. MZOFNM 2005.

PW2-11
Exercise capacity and biventricular function in adult patients with repaired tetralogy of Fallot.
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Introduction: Adult patients with repaired tetralogy of Fallot (rTOF) often have diminished right ventricular function and /or significant pulmonary regurgitation. Our aim was to examine whether these abnormalities play a role in diminuation of exercise function in patients with rTOF.
Methods: This was a retrospective review of 46 adult patients with rTOF. Right ventricular function (RV) and pulmonary regurgitation (PR) were assessed echocardiographically and by cardiovascular magnetic resonance (CMR). Peak oxygen consumption (peak VO(2)), predicted Vo(2)max for age and sex and ventilatory efficacity was measured by cardiopulmonary exercise test. All patients were clinically stable, their investigations were done within 1 year.
Results: The mean age of the cohort was 28 +/- 9years (48% females). Seven patients had PM/ICD. Seventy two percent of patients (n = 33) were asymptomatic (NYHA class I). The mean peak Vo(2)max was 24 +/- 6 ml/hg/min, predicted peak Vo(2)-max 69 +/- 17% and VE/VCO2 slope was 30 +/- 5. There were 27% of patients with ⩾ moderate right ventricular dysfunction and 72% with ⩾ moderate pulmonary regurgitation. There was no significant difference between mean peak Vo(2)max (25 +/- 5 vs. 23 +/- 6 ml/hg/min, p = 0,43), predicted peak Vo(2)max (69 +/- 17% vs 67/18%, p = 0,86) and VE/VCO2 slope (30 +/- 5 vs 29 +/- 6, p = 0,87) in patients with or without RV dysfunction and /or PR.
Conclusions: Our data suggest that in a young, mostly asymptomatic cohort of patients with repaired tetralogy of Fallot at least moderate RV dysfunction and/or moderate pulmonary regurgitation exercise capacity may be preserved.

PW2-12
Reference Values of Aortic Augmentation Index in a Large Healthy Population Aged 3-18 Years
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Basics: Aortic pressure augmentation caused by pressure wave reflection is a physiologically important phenomenon. The aortic augmentation index (Aix_{ao}) increases with age in adults and the increased Aix_{ao} is a strong predictor of cardiovascular morbidity and mortality. However, the pressure waveform in infants and children are similar to those in older adults, and thus the Aix_{ao} is higher in this group. Consequently the determination of reference values of Aix_{ao} in a large population of healthy children and adolescents is very reasonable.
Aims: To determine the reference values of Aix_{ao} in children and adolescents and to find the possible physiological mechanisms of the enhanced aortic pressure wave reflection during childhood.
Methods: AIx_{ao} were measured by a new non-invasive, occlusive, oscillometric method (Arteriograph, TensioMed Ltd., Hungary) in a healthy population aged 3-18 years with normal BMI and with normal blood pressure (1802 males, 1572 females). Smoothed percentile curves from 3th to 97th were determined using LMS method. Results were analyzed by Student's t-test.
Results: The physiological changes of Aix_{ao} measured in healthy population are shown in Figure 1. and 2. The Aix_{ao} decreased

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with age in both genders. From the age of 14 years the Aix_{ao} were significantly lower in males than in females ($p < 0.02$), and this difference had become even more pronounced from age of 15 years ($p < 0.001$). Assessing the background of these gender differences in Aix_{ao} we have found that the changes of the median of Aix_{ao} are exactly identical to the changes of the median of body height.

Figure 1.

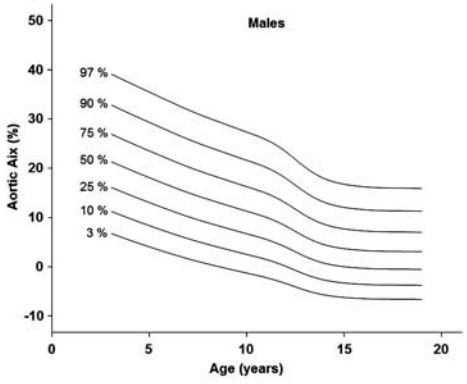
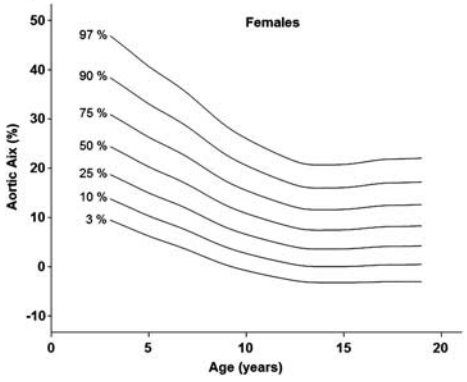


Figure 2.



Conclusions: This is the first large population study involving 3374 healthy subjects aged 3-18 years which describes the physiological changes of Aix_{ao}. Our data provide supporting evidences that the pressure waveforms in infants and children are markedly elevated and similar to those with advanced age. This very interesting phenomenon can entirely be explained by the differences in body height (aortic length) with ageing. In children with the shorter body height (aortic length) the reflected wave returns earlier and increases the augmentation index (Aix_{ao}), not because of the stiffer arterial system.

PW3-1

Long-term follow-up of patients with left lung perfusion abnormalities following transcatheter closure of patent ductus arteriosus

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Background: This study presents the long-term follow-up of patients who developed left lung perfusion (LLP) abnormalities following patent ductus arteriosus (PDA) closure with various device types.

Methods: The study includes 23 adult and pediatric patients who had undergone transcatheter PDA closure and were shown to have decreased LLP (<40%) by the first scintigraphy performed within the average follow-up period of 14,0 ± 8,12 (2,0-30) months. For PDA closure, the Amplatzer duct occluder was used in 12 patients and coils were used in 11. Within the average period of 58,91 ± 12,93 (37-85) months after transcatheter PDA closure a second lung perfusion scintigraphy was performed.

Results: In 13 of 23 patients (56.5%) having impaired LLP improved by the time of the second scintigraphy. Improved and unimproved patients didn't differ with regard to age, weight, body surface area, PDA diameter, ampulla diameter and PDA length at the time of PDA closure and the second scintigraphy. There was no significant difference with regard to the percent of improved patients between the different device types ($p = 0.88$). Also the left pulmonary artery indexes was insignificantly different ($P = 0.446$). Patients with persistent LLP abnormality have significantly higher mean DVI [(LPA blood flow velocity–RPA blood flow velocity)/MPA blood flow velocity] x 100 values ($p = 0.007$) and PDA diameter/length. If the DVI ⩾50% is taken as the cut-off value it is possible to predict patients with persisting LLP abnormality with 80% sensitivity and 76% specificity. The abnormality could persist in patients having PDA diameter/length ⩾0.5 with 80% sensitivity and 92.3% specificity.

Conclusions: The LLP abnormalities seen after PDA closure with various devices eventually improve to normal in the majority of patients during long-term follow-up. Patients whose PDA length is shorter than its diameter are at risk of developing LLP abnormalities that persist long-term.

PW3-2

Use of Covered Cheatham-Platinum (CCP) stents in congenital heart disease

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Objective: Evaluation of possibilities and safety of covered Cheatham-Platinum (CCP) stents congenital heart disease (CHD).

Methods: Single-center retrospective study of all CCP stents implanted 2003-2011. Three subgroups were identified: stent implantation for aortic coarctation (CoA), RVOT pre-stenting for percutaneous revalvulation and miscellaneous. Indication, effectiveness and safety of stent implantation were assessed. Narrowed segments were expanded using moderate inflation pressures (4-6 atm) opposing the CCP stent against the vessel wall. In case of subtotal relief of the obstruction or if a tear was anticipated, time was allowed for "ingrowth" of the stent until full expansion at a 2nd procedure.

Results: 78 CCP stents were implanted in 80 patients.

CoA group: 45 CCP stents were implanted in 44 patients: 2/44 for exclusion of aneurysms, 42/44 "prophylactically" (due to atresia requiring puncture in 1, filiform stenosis in 21). Stent was dilated to desired dimension at implantation in 35 patients, stent re-dilation after 6.7 ± 7.5 months in 9 patients. Final CCP stenting resulted in significant increase in the CoA diameter from 6 ± 4 (range 0–12) to 15 ± 2 (range 12–20) mm with a decrease in PTP gradient from 28 ± 18 to 3 ± 5 mmHg.

RVOT group: 27 CCP stents in 25 patients with stenosed conduits. In 6 patients CCP stents were used out of caution due

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to rupture of pre-dilation balloon. Dilation was performed up to "nominal" in 12/25 patients and 2–6 mm beyond "nominal" in 13/25 (52%) patients.
Miscellaneous group: 15 CCPstents implanted in 11 patients for: closure of Fontan-circuit fenestration (n = 3), restoration of caval vein (n = 2), stenosis of cavopulmonary connection (n = 2), to preserve pulmonary artery patency (n = 2) and relief of supra-pulmonary stenosis (n = 2). A hybrid stent implantation in 2 patients to obtain a sutureless connection between a conduit and minute intrapulmonary arteries. CCP stenting was necessary as rescue treatment in 2 patients.
The desired result was obtained in all patients; no extravasation was encountered despite significant expansion and presumed tears of narrow segments.
Conclusion: Where vessel tear and extravasation can be expected, the use of covered stents is safe and opens new opportunities with more complete dilation. Therapeutic use includes aneurysm exclusion and control of bleeding.

PW3-3
Transcatheter PDA-closure in (pre-term) newborns below 3 kg of weight
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Introduction: Surgical ligation is the standard treatment for a hemodynamically significant patent ductus arteriosus (PDA) in preterm infants who remain unresponsive to medical treatment. The Amplatzer ductal occluder II-AS (ADO-II-AS; AGA/St. Jude) is a symmetrically shaped device delivered through a 4F proprietary catheter, CE-marked for transcatheter ductal closure in children older than 6 months of age and above 6 kg of weight with a ductal diameter of $\leq$4 mm and length $\leq$8 mm.We report on our experience with PDA closure in premature children with a weight of$<$3 kg.
Patients and methods: In 2011, 13 patients with a body weight below the recommended weight of 6 kg underwent transcatheter PDA closure for hemodynamically significant ductus using the ADO-II-AS at our institution. PDA closure was attempted in 5 pre-term infants with a body weight of less than 3 kg (range 2.1 to 2.8 kg). Percutaneous vascular access was from the femoral venous side only (4F short sheath) in all patients. A hand-injection of contrast through the side-arm of a Touhy-Borst adapter on a diagnostic catheter placed over a floppy wire through the duct visualized the ductal morphology. An ADO-II-AS size chosen according to the manufacturer's instructions was implanted through a 4F delivery catheter. Transthoracic echo and hand-injection of contrast through the side-arm of the delivery catheter confirmed correct device position prior to release.
Results: Transcatheter PDA closure by antegrade implantation of an ADO II AS was technically feasible without any problems in all children. All infants except one showed complete ductal closure within 24 hrs after the procedure. No obstructions of the aortic arch or the left pulmonary artery were observed. In one child (2.6 kg) the device tilted after release from the delivery system resulting in significant residual shunting. This device was retrieved percutaneously and uneventfully through a long 4F sheath during the same procedure.
Conclusions: Using the ADO II AS, transcatheter PDA-closure is possible in selected (pre-term) newborns below 3 kg of weight. The device that can be delivered transvenously through a 4F catheter preserves the arteries in small children and extends the previous limits of percutaneous PDA closure.

PW3-4
Stenting of the Right Ventricular Outflow Tract
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Objective: To assess the indication, technical aspects and outcome of stenting of the right ventricular outflow tract (RVOT) in the management of symptomatic patients with severely limited pulmonary blood flow.
Methods: Retrospective case note and procedure review of patients undergoing stenting of the RVOT over a 7 year period.
Patients: Between 2005-2011, 38 selected patients underwent cardiac catheteriza¬tion with a view to stent a very narrow RVOT to improve pulmonary blood flow. In all, cardiac surgical intervention was deemed high risk due to presenting condition, weight, associated defects, underlying anatomy, or co-existing syndromes. In 3 patients the procedure was abandoned due to unsuitable anatomy. Median age at stent implantation was 64(range7-406) days and median weight was 3.8(1.7-12.2) kg.
Results: Thirty-five patients underwent stent implantation. Median procedure time was 60(29-260) and fluoroscopy time 16(8-73) minutes. There was one procedural death (2.9%) and one requiring emergency surgery (2.9%). Saturations increased from 70(52-83)% to 91(81-100)% [$p < 0.001$].
Eleven further catheter interventions were carried out (balloon in 6, further stent in 5). Twenty patients underwent delayed surgery (complete repair in 15, palliative in 5) at a mean of 264(10-758) days post stenting. Thirteen patients remain well palliated after a mean of 202(14-508) days.
Conclusion: Stenting of the RVOT is an effective treatment option in the initial management of selected patients with very reduced pulmonary blood flow.

PW3-5
Closure of extracardiac Fontan fenestration with covered stents
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Introduction: In patients after total cavopulmonary connection (TCPC) fenestration allows to improve acute postoperative morbidity. After a stabilized chronic hemodynamic state is achieved, an interventional closure of fenestration can be performed with various devices to increase the systemic arterial saturation and improve the exercise tolerance.
The aim of this paper is to review our experience in the use of covered stents for transcatheter closure of fenestration in patients after TCPC.
Methods: We implanted CP covered (6) or Advanta V12 (3) stent in 9 patients after fenestrated TCPC. Median patients age and weight were 8 years and 27.5 kilograms respectively and median interval after the surgery was 22 months. Stenosis of the homograft was additional indication for stent implantation in 2 patients, rhythm disturbances during occlusion test in 2 and unfavorable localization of the fenestration not allowing for proper implantation of Amplatzer Septal Occluder in 1 patient. Femoral approach was used in all but 2 patients having bilateral thrombosis of femoral veins. The Advanta V12 was premounted on a balloon catheter whereas the CP stent was crimped on a BiB balloon.

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Results: Mean oxygen saturation and internal caval vein pressure increased acutely from 81,1% +/- 4.7% to 96.1% +/- 2.6% ($P < 0.001$) and 14 +/- 2 mm Hg to 14.7 +/- 2.4 mm Hg ($P < 0.02$), respectively. In 2 patients control angiography showed trivial leak through the fenestration, in the remaining 7 there was no residual flow. Discharge echocardiography showed no flow at the level of the fenestration in all patients. No procedural or intra-hospital complications occurred. One patient died 8 months after the intervention due to thromboembolic complications after arm fracture. The remainder patients are symptom-free at median follow-up of 8 months, and the mean oxygen saturation is 95.8% +/- 1.3%.
Conclusions: The covered stents can be safely and effectively used for closure of TCPC fenestrations. Apart from avoiding protrusion of prosthetic material into the left atrium, this method has additional advantage in patients with stenosis of the homograft, rhythm disturbances or inappropriate anatomy of the atrium or localization of the fenestration to safely deploy other devices.

PW3-6

Retrograde transcatheter closure of ventricular septal defects using the Amplatzer duct occluder II device

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Objectives: To describe a new technique of transcatheter closure of VSDs in children. Routine anterograde closure of VSDs is well established, but required the formation of an arteriovenous loop. We describe the retrograde single catheter approach.
Methods: 77 symptomatic children (42 males, median age 46 months, range 0.6 to 240 months, median weight 14 kg, range 4.8 to 38 kg) with various types of perimembranous ($n = 61$) or muscular defects were selected for VSD closure. The VSD diameter had to be <6.5 mm, as this is the maximum available waist diameter of the Amplatzer Duct Occluder II (ADO II). An ADO II device was chosen with a waist diameter equal to or 1 mm greater than the minimum VSD diameter. Using a 5F or 6F Judkins right coronary catheter (internal diameter 0.056 to 0.070") and 0.035" Terumo wire combination, the VSD was crossed retrogradely via the femoral artery, and the appropriate ADO II device delivered. The distal (RV) disc was initially deployed, followed by the waist and LV disc, under transthoracic ($n = 56$) or transesophageal echocardiographic guidance.
Results: The median VSD diameter was 4.5 mm (4 to 6.5 mm). The mean fluoroscopic time was 11 ± 8 minutes. Two devices embolized, but were successfully retrieved, and the VSD closed with a larger device. Preexisting tricuspid regurgitation in children with perimembranous VSDs invariably improved. At a median follow-up of 14 months, 3 patients had a right bundle branch block; complete closure rate was 90%.
Conclusions: Retrograde VSD closure using a single standard guiding catheter is safe, feasible and simplifies the procedure. It should be considered in young, symptomatic patients.

PW3-7

Femoral vein occlusion in infants undergoing cardiac catheterisation pre-cavopulmonary shunt procedure

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Introduction: Femoral venous thrombosis and femoral vein access issues after catheterisation is a well recognised complication. There are no studies describing the extent of the problem, predisposing factors and possible preventive strategies in infants with univentricular heart physiology. We set out to look at this high risk group of infants, undergoing routine cardiac catheterisation prior to their cavopulmonary shunt procedure.
Material and Methods: Retrospective analysis over 2 year period from 2007 till 2009, conducted on 101 children undergoing cardiac catheterisation pre-cavopulmonary shunt procedure. We analysed procedure records and angiograms of each patient. Five were excluded for lack of data and 96 included.
Results: Routine cardiac catheterisation performed on 96 children prior to cavopulmonary shunt procedure at median age of 4.2 months (2.3 -17 months) and median weight 5.9 kg (4–11 kg). The commonest underlying diagnosis was hypoplastic left heart syndrome in 83(86%) with the remainder comprising of other univentricular physiologies. All patients underwent a surgical procedure; either Norwood or PA banding initially followed by stay in intensive care. In addition to this, 15 infants also had cardiac catheterization. Right femoral vein (RFV) access was obtained in 42(45%) and failed in 53(55%). Elective right femoral artery (RFA) access was used in one unstable infant. In those with failed RFV access; access was achieved using left femoral vein (LFV) in 18(34%), internal jugular vein in 4 (7%) and either right or left femoral artery was used when both femoral veins were not accessible in 31(58%).
Angiographic evidence of occluded veins was seen in 34(35%), occluded RFV in 11(11%), occluded bilateral femoral veins in 19(20%) and occluded inferior vena cava in 4(4%). One patient had ultrasound evidence of blocked right femoral vein.
Conclusions: Venous access during cardiac catheterization of infants with univentricular heart with prior surgery can be challenging. One third of these patients have angiographic evidence of one or more femoral vein occlusion. Routine ultrasound evaluation prior to procedure may help reduce procedure time by identifying vessel patency. Other options such as heparin coated central lines need to be considered to minimize extent of this problem.

PW3-8

Importance of coronary artery anatomy and variation in percuraneous pulmonary valve implantation

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Introduction: The percutaneous implantation of pulmonary valves (PPVI) has become standard in many centres. The coronary anatomy and variability has significant impact on the suitability of this procedure. We report on our first 50 patients with right ventricular outflow-tract obstruction and the attempt to implant pulmonary valves.
Methods: Prospective study in all patients scheduled for PPVI. A total number of 50 patients was enrolled, there were 27 male and 23 female. In 34 patients PPVI was successfully completed (22 Melody® valves, 12 Edwards Sapien® valves). Our standard protocol included a maximum balloon inflation using high pressure balloons to ensure complete dilatation of the stenosed RVOT and simultaneous selective coronary angiography in all patients. In the remaining 16 patients PPVI was not possible. This was due to a subpulmonary stenosis in 1 patient, an extensive size of the RVOT after standard balloon sizing/dilatation in 1 patient, but due to coronary anatomy in 14 (= 28%). Compression of the

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coronary blood flow occurred in 10 patients during balloon dilatation/sizing, and coronary anatomy was found to close to possible stent location in 3.
Discussion: Coronary anatomy and variation prevents successful PPVI in a significant proportion of unselected patients with RVOTO. A standardised protocol to simulate stent implantation is compulsory to perform the procedure safely.

PW3-9

Transcatheter closure of secundum atrial septal defect associated with deficient rims other than the antero superior using Amplatzer devices.

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Introduction: We aimed to assess the feasibility of transcatheter closure for secundum ASDs with deficient rims (<5 mm), other than the antero-superior because it is well demonstrated that deficiency in the anterior rim toward the aorta does not influence the success rate for transcatheter ASD closure.
Method: Between January 1st, 2008 and december 31, 2011, 192 patients underwent percutaneous closure of ASD in our institution under transesophagal echocardiography guidance in children and intracardiac echocardiography guidance in adults. Amplatzer devices (Amplatzer Septal Occluder or Cribriform) were used in 191 whereas one case without deficient rim was closed with intrasept ASD occluder. We retrospectively analyzed the outcomes of the 43 patients (22,4%, 26 children) with one or more deficient rim.
Results: The median age and weight was 16(1,1 to 85) years and 56(8,8 to 99)Kg, respectively. Deficiency of the inferior (toward the AV valves, n = 11), inferior- posterior (toward the inferior vena cava, n = 15), or of the superior -posterior rim (toward the superior vena cava, n = 17) was confirmed by transesophagal echocardiography in all the cases. Transcatheter closure was successfully accomplished in 37 (86%) of the cases with a median ASO size of 28(10 to 40) mm. A modified method of implantation (sizing balloon technique)was used in 30 patients (70%).In 6 patients (5 children) the ASD could not be closed.Four other children experienced device embolization few hours after successful transcatheter closure and underwent surgical ASD closure without further complication. By Fisher's exact test, deficiency of the inferior-posterior rim was associated with failure or embolization($p = 0.0496$)and there was a trend for adult age to be associated with a low risk of embolization or failure ($p = 0,06$).
Conclusion: Transcatheter closure of secundum ASD is feasible in patients with deficient rims. However, this cannot be recommended because of an intolerable rate of embolization. Possibly, transcatheter closure of such secundum ASDs with deficient rims may be more successful in the adult population.

PW3-10

Outcome after Fetal Diagnosis of Hypoplastic Left Heart Syndrome: The Toronto Experience of 267 Patients

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Background: Hypoplastic left heart syndrome (HLHS) is characterized by variable degrees of underdevelopment of the left heart, such that is unable to support the systemic circulation after birth. Available management options offered to parents at our center include termination of pregnancy (TOP), compassionate care at birth (CC), and active management either with staged single ventricle surgery (including two stage 1 options: hybrid procedure vs. Norwood operation) or cardiac transplant. Studies on HLHS outcomes are usually focused on postnatal outcomes and suggested a rather wide range of outcomes among centers and sometimes among surgical options. The aim of this study was to review the overall outcome of a large cohort of HLHS babies from the time of fetal diagnosis to 1-year of life.
Methods: 267 fetal cases diagnosed with HLHS (1990-2011) were identified in our cardiac database. Clinical/echocardiography data were correlated with outcomes of actively managed babies.
Result: Mean age at referral was 23.7 + 5.4 gestational weeks with mitral atresia/aortic atresia (MA/AA: 49%) being more common than mitral stenosis /aortic atresia (29%) and mitral stenosis/ aortic stenosis (22%). Additional major extracardiac abnormalities were detected in 13% of cases, while genetic disorders affected 19% of 119 tested cases, with Turner syndrome as the most common anomaly (7%). 160/267 (60%) resulted in TOP or CC and 13 (4.9%) underwent spontaneous intrauterine demise (IUD). Of 94 live-births with intended active management, 75 (80%) survived to infancy and only 56 (60%) to 1 year. Risk factors for IUD were a highly restricted atrial septum ($p < 0.001$), extracardiac structural abnormalities ($p < 0.001$), at least moderate tricuspid regurgitation ($p = 0.002$) and pericardial effusion ($p < 0.001$). Postnatal mortality was significantly associated with MA/AA ($p = 0.05$) and a highly restrictive atrial septum with a need for urgent neonatal balloon atrioseptostomy ($p = 0.02$). Other variables including the choice of surgery and era of diagnosis did not significantly affect outcomes.
Conclusion: The overall survival to 1 year of life after a fetal diagnosis of HLHS was 21% in this study. Main reasons for the high attrition rate included frequent associations with genetic/ extracardiac disorders, parental decision of TOP/CC, and surgery-related complications.

PW3-11

Prenatal cardiac diagnosis and the 'Postcode Lottery'; has UK NHS FASP (fetal anomaly screening protocol) made any difference yet?

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Introduction: There has been marked regional and intra-regional variability in prenatal cardiac anomaly detection rates in the UK. Although the East Midlands has historically had higher detection rates than much of the UK, as a Regional Cardiac centre we are aware that rates within the East Midlands itself have also varied widely. New UK standards for fetal cardiac (and general) screening (UK NHS FASP) were launched in early 2010, & a national training programme to improve cardiac screening was provided. There was a stated expectation that 'all units would meet minimum standards' by April 2011. We have therefore evaluated the proportion of babies (<1 year) undergoing surgery or interventional cardiology procedures at EMCHC diagnosed prenatally, by calendar year, over the last 3 years, to assess progress, and also to review local differences using postal (ZIP) code area within the East Midlands
Methods: Retrospective case note review. 6 Main post code areas identified; mainly (although not exactly) corresponding to catchment areas for referring obstetric centres. Procedures to

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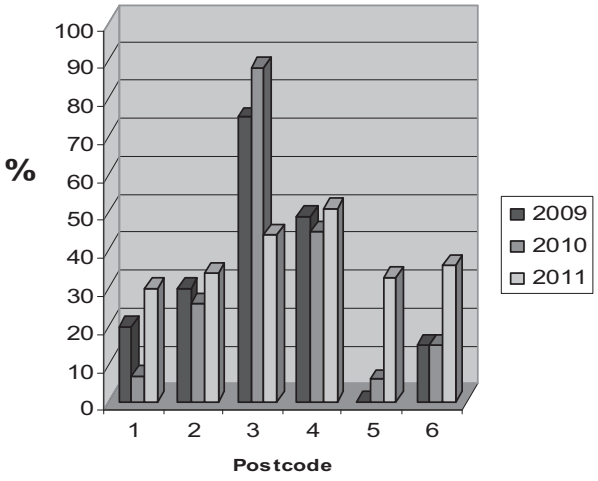
close ASD or PDA were excluded as were cases from 'non-main' postcode areas.



Results: In 2009, overall percentage of cases with a prenatal diagnosis was 33% (42/126), in 2010 was 32% (43.134) & in 2011 it was 39% (53/137). By Postcode area, percentages ranged between 0 -75% in 2009, 6 – 88% in 2010, & 30-51% in 2011.

Post-Code Area	2009 % (total n)	2010% (total n)	2011% (total n)
1	20% (10)	7% (14)	30% (23)
2	30% (37)	26% (43)	34% (44)
3	75% (4)	88% (16)	44% (9)
4	49% (49)	45% (31)	51% (35)
5	0% (3)	6% (17)	33% (15)
6	5% (13)	15% (13)	36% (11)
Total	33% (126)	32% (134)	39% (137)

Conclusions: Although the change for postcode area 5 also corresponds with the closure of an adjacent surgical unit & consequent change in prenatal referral pattern, there is a clear overall increase in percentages of cases with prenatal diagnosis in 2011, especially in the 'lower scoring' areas. However there is clearly significant room for improvement in some areas and further work is need to identify and remedy the relevant local issues.

PW3-12

Effect of changes to UK fetal cardiac screening protocols across the spectrum of congenital heart disease requiring intervention in the first year of life

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Introduction: New UK standards for fetal screening (UK NHS FASP) were launched in early 2010, & a national training programme to improve cardiac screening was provided. Prior to 2010, standard 18-20 week screening principally required a 'normal 4 chamber view'. The new standards have added views of the aorta/LV outflow tract & the pulmonary artery/RV outflow tract or a 3 vessel view showing the aorta, pulmonary artery and superior vena cava. Use of Colour flow is encouraged but not mandatory. There is a stated expectation that 'all units would meet minimum standards' by April 2011. We evaluated the proportion of babies (<1 year) undergoing surgery or interventional cardiology procedures at EMCHC diagnosed prenatally, by calendar year, over the last 3 years, in broad morphologic groups, to assess progress and identify areas for further improvement
Methods: Retrospective case note review. Patients with ASD, PDA and ALCAPA excluded. Lesions grouped as 'isolated'; VSD, Coarctation, Aortic & Pulmonary stenosis (AS+PS), TGA, AVSD, and 'complex'; functionally single ventricle (V), 'Conotruncal' abnormalities (ToF variants, Truncus Arteriosus and IAA with VSD), 'balanced' ccTGA variants. No cases of isolated TAPVD were diagnosed prenatally.

	2009		2010		2011	
Group	Diagnosed (% of total)	Missed	Diagnosed (% of total)	Missed	Diagnosed (% of total)	Missed
VSD	4 (25%)	12	1 (5%)	18	0 (0%)	21
Coarct'n	1 (8%)	12	1 (10%)	9	3 (18%)	14
AS+PS	3 (25%)	9	2 (20%)	8	3 (18%)	13
TGA	5 (33%)	10	5 (38%)	8	5 (41%)	7
AVSD	0 (0%)	3	5 (36%)	9	11 (73%)	4
Single V	11 (73%)	4	14 (70%)	6	11 (79%)	3
'Conotruncal'	10 (36%)	18	8 (44%)	10	8 (40%)	12
ccTGA	2 (100%)	0	1 (33%)	2	2 (100%)	0
Total	36 (35%)	68	37 (35%)	70	43 (37%)	74

Results: In 2009, overall percentage of cases with a prenatal diagnosis was 33% (42/126), in 2010 was 32% (43.134) & in 2011 it was 39% (53/137). Main diagnostic groups shown below;
Conclusions: Overall percentage of cases diagnosed prenatally did rise in 2011, but varied for different lesions. Prenatal diagnosis rate for AVSD was poor but has increased dramatically, detection of TGA is slowly improving, but it appears that detection of isolated VSD has in fact deteriorated. Percentage of prenatal diagnosis for single ventricle lesions remains high (reflecting a regionally low rate for termination of pregnancy), but some remain undetected. Detection rate of 'milder' obstructive valve disease remains constant, but detection of 'conotruncal' defects, which the new FASP standards should facilitate, has not yet changed dramatically. At least in the East Midlands, the 2010 FASP standards have improved 4 chamber screening for AVSD but have not yet impacted for other lesions. More training work is needed.

PW4-1

Hypothermia after hypoxia is neuroprotective possibly via upregulation of cold shock proteins RBM3 and CIRP

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Introduction: Ischemic insults during cardiac arrest or resuscitation can lead to a hypoxic state in the brain tissue and cause neurological sequelae. Therapeutic hypothermia is known to be an effective cytoprotectant, possibly improving the neurological outcome of these patients. Two RNA-binding proteins, the cold-inducible RNA-binding protein (CIRP) and the RNA-binding motif protein 3 (RBM3) have been observed to be upregulated during hypothermia. This observation prompted us to hypothesize a possible participation of CIRP and RBM3 in the hypothermia-induced neuroprotection.
Methods: The SK-N-SH neuroblastoma cell were exposed to either mild or severe hypoxia (8% or 0% oxygen) for 24 hrs and

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afterwards kept at 33,5°C for 72h. LDH-assay for cell death, western-blot for quantification of the expression of the cold shock proteins RBM3 and CIRP were performed.
Mild and severe hypoxia for 24 hrs increased the LDH-release in neurons, when incubated at 37°C afterwards, compared to a normoxic control group. Mild hypothermia for 24hrs, 48hrs or 72hrs could reduce the LDH-release significantly at all time points in both cells exposed to 8% and 0% hypoxia for 24 hrs. These results support the theory of hypothermia function as a cytoprotectant after a hypoxic insult.
RBM3 is significantly upregulated by mild hypothermia in a time-dose-dependent manner in SK-N-SH cells at all time points, that were exposed to mild or severe hypoxia for 24 hrs before. RBM3 is furthermore significantly upregulated in SK-N-SH cells that were not exposed to hypoxia, but incubated at 33,5°C for 24 hrs and 48 hrs. CIRP is upregulated by hypothermia in SK-N-SH cells, that were exposed to mild or severe hypoxia for 24 hrs before. CIRP is furthermore upregulated in SK-N-SH cells that were not exposed to hypoxia, but incubated at 33,5°C for 48hrs.
Conclusion: Mild hypothermia after hypoxia reduces neuronal cell death .RBM3 and CIRP upregulation in response to hypothermia could be one possible mechanism for hypothermia-induced neuroprotection.

PW4-2

Weaning from ventricular assist device in non – myocarditis pediatric heart failure patients.

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Retrospective analysis of a single-center experience with successful weaning procedures in children supported by the BerlinHeart–Excor paracorporal ventricular assist device. Weaning was completed in 5 of 19 VAD applications. Patients: age: 3 months- 15 yrs. (median 8 months); indication: DCMP (3), ischemic heart failure (1), DCMP and congenital Heart defect (1); myocarditis ruled out by biopsy in 5/5; all pts. had been listed for urgent Heart Tx at time of VAD implantation. VAD mode: 3 LVAD, 2 BIVAD, LV-apex canulla in 5/5. Duration of MCS until weaning: 12 – 42 days (median 25 days). Preconditioning instruments (ie. intracoronary stem cell infusion, pulmonary artery banding), evaluation of possible myocardial recovery and the weaning procedure itself are described in detail. In 2/5 forced weaning was mandatory due to VAD related infection and risk of thromboembolic events.
Results: weaning was completed in 5/5 patients: 2/ 5 need a 2nd VAD 15 days/6 months resp. before transplantation; in 3/5 myocardial recovery sustained.
Conclusions: Weaning is always an option even for non – myocarditis pediatric patients; Weaning is possible after a short time unloading on VAD. Temporary weaning off MCS is reasonable to avoid VAD related complications.

PW4-3

Cardiac catheterization yields significant information and improves outcome in otherwise unidentifiable hemodynamic instability early after congenital heart surgery

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Objective: Hemodynamic instability in the early post operative period is a challenge after surgery for congenital heart disease (CHD). In some patients a cause is not identifiable by clinical and non-invasive means.
Aim of the study: evaluation of therapeutic options and outcome of patients undergoing an early cardiac catheterization (CC) within 30 days after surgery for CHD.
Methods/results: Between 2008 and 2011, 1940 patients (pts) with CHD were operated at our centre. Of these 151 (7.8%) needed an early CC including 37 patients with HLHS. Mean age of these pts was 2.8 yrs (SD 7.8; range:1day–72 yrs), mean weight 11.4 kg (SD 19.4; range: 1.7–127 kg).
Indications for CC were low cardiac output syndrome (LCOS) or hemodynamic instability in 50 patients, cyanosis (44 patients), chronic effusions (17 patients), respiratory insuffiency (6 patients), thrombosis (17 patients), and miscellaneous causes (18 patients). 20 patients were on ECMO during CC. Mean time between operation and CC was 9 days ± 10.
Treatment: In 64 (42%) pts CC was followed by medical treatment only, but in 31 of these 64 pts CC findings lead to essential changes of medical treatment. 46 (30%) pts had catheter interventions (pulmonary arterial interventions: 19, closure of aorto-pulmonary or veno-venous collaterals: 11, angioplasty or stentimplantation because of acute aortopulmomary/Sano-shunt stenosis or closure: 11, angioplasties +/-lytic therapy of thrombosed caval veins: 5, two interatrial septum interventions, one coronary artery-stenting, one LPSVC-closure after TCPC, and one valvuloplasty of the pulmonary and aortic valve). In some patients more than a single intervention was done. There was no intraprocedural mortality during CC. 33 pts (22%) had reoperations, and nine pts a catheter intervention and a reoperation. Clinical improvement was achieved in 123 pts. (80%). 23 (15%) pts (10 on ECMO) died: 3 after intervention, 8 after reoperation, 2 after intervention and reoperation, 10 under medical treatment/compassionate care.
Conclusion: Cardiac catheterization is an important diagnostic tool in the early post operative period to decide on further treatment options. Even in these hemodynamically compromised patients CC can safely and effectively be performed. In 78% of our patients CC findings led to causative treatment.

PW4-4

Going down, going slow–Esmolol as potent myocardial protector in rescue cardiac extracorporal membrane oxygenation (ECMO)

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Introduction: Cardiac failure or arrest post elective cardiac surgery in neonates and children are rare events. However their occurrence during a highly vulnerable period of myocardial recovery imply immediate expert support by specialized intensive care unit staff. Most of the time ECMO has to be initiated. Timing and efficiency of resuscitation as well as duration of cannulation for ECMO are crucial for the survival of affected patients. Equally important are subsequent cardiovascular management of those patients to optimize myocardial recovery. Beside volume unloading by ECMO to prevent excessive myocardial stretch, optimal coronary perfusion has to be

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maintained to protect cardiomyocytes from oxidative stress and acute cell death. Recently it could be shown that beta blockers combine cardioprotective mechanisms not only by improving myocardial relaxation and simultanously coronary perfusion but also by increasing antioxidative activity (gluthatione peroxidise, superoxide dismutase).
Methods: Patients (n = 6) requiring rescue ECMO post elective cardiac surgery due to acute heart failure or sudden cardiac arrest were started on Esmolol infusion as soon as stabilized. Complete cardiac unloading was achieved by full flow ECMO (≥150 ml/kg/min). Serial transthoracic echocardiography was performed to assess myocardial contractility and cardiac unloading.
Results: 6 patients (2 male, 4 female), age 2.2 ± 4.1y with single ventricle physiology (n = 3), complex cyanotic heart disease (n = 2), coronary anomaly (n = 1). All patients demonstrated myocardial stunning. ECMO duration 8.8 ± 1.9 days, maximum dose Esmolol 106.7 ± 50.1 μg/kg/min, maximum heart rate (HR) prior to Esmolol 168.3 ± 11.7 beats per minute (bpm), maximum heart rate during Esmolol infusion 73.3 ± 8.2 bpm, fractional shortning (FS) prior to Esmolol 9.2 ± 4.9%, FS post Esmolol 33.3 ± 7.5%. Weaning of ECMO was successful in 4 patients. 2 patients died on ECMO due to neurological issues.
Conclusions: In this small pilot study without case control, all patients showed significant improvement in myocardial contractility. Esmolol appears to provide cardioprotection for pediatric patients post cardiac failure/arrest requiring ECMO due to stunning myocardium. We speculate that its combined potential antioxidative effect may further support recovery of the affected myocytes by increased activity of gluthation peroxidise and superoxide dismutase.

PW4-5

Topsy-Turvy Heart: Very rare rotational heart anomaly with tracheobronchial anomalies

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Introduction: Topsy-Turvy heart is characterized by approximately 90 degree clockwise rotation of the entire heart as a block around the axis from apex to base, leaving the right ventricle in a superior spatial relationship to the left venticle, while the great arteries are displaced inferiorly and posteriorly into the distal mediastinum resulting elongation of the brachiocephalic arteries. As a result of the rotation trachea and bronchi become stretched and elongated. To date, only 4 alive cases have been described in the literature. We report 3 cases with morphologic details, clinical presentation, additional anomalies and their management.
Methods: The patients were 1 month, 5 months and 4 years of age. Two girls and a boy. Last two patients were siblings born to consanguineous parents. In addition to the characteristic rotational anomaly of the heart and the great vessels, all patients had large aorto-pulmonary window defects, some degree of left main bronchus stenosis and systemic pulmonary hypertension. First two patients with severe heart failure symptoms, underwent open heart surgery. Aorto-pulmonary window defects were closed by using pericardial patch under total circulatory arrest (18°C) in case 1 and under selective antegrade serebral perfusion (26°C) in case 2.
Results: Severe respiratory failure and air trapping in the left lung developed in the first case. Sternum was left opened. ECMO support was initiated in postoperative day 10 due to resistant respiratory failure. She died of multiorgan failure in postoperative day 16. The second case had a prolonged intensive care unit stay due to pulmonary hypertensive crisis and postoperative sepsis. He was discharged from the hospital on the 30th postoperative day in good clinical condition. Cardiac catheterization of the third patient revealed high pulmonary vascular resistance (12 Wood U). Bosentan treatment was started and second hemodynamic evaluation was scheduled. Genetic counseling is pending.

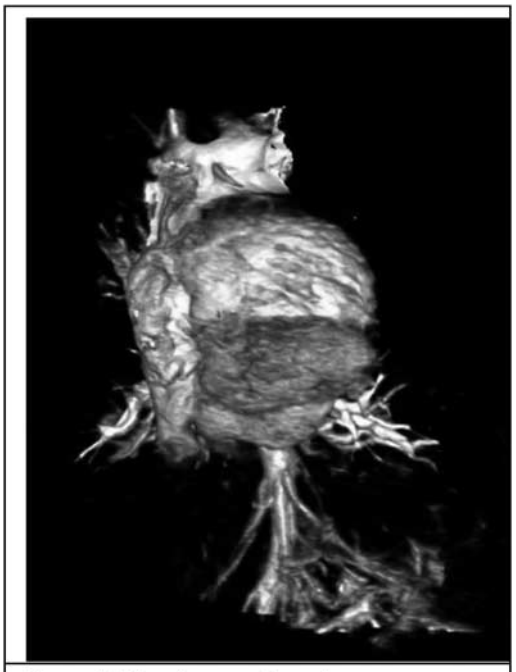

Figure 1:CT angiogram of case1
3D reconstruction, anterior view

Conclusion: Our findings support the previous reports in the literature. Patients have no intracardiac anomaly except a large aorto-pulmonary communication. History of consanguinity, like other report, support a single gene disorder with a recessive mode of inheritance. Respiratory complications which may be fatal in postoperative period, necessitate multidisciplinary team approach.

PW4-6

Non-invasive three-dimensional pressure maps by flow-sensitive MRI: comparison of measurement accuracy compared to invasive catheterization in patients with aortic coarctation

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Background and Objectives: State-of-the-art MRI provides superb anatomic and functional information but still lacks assessment of pressures. Data from four dimensional velocity-encoded cine magnetic resonance imaging (4D VEC MRI) can be computed into relative 3D pressure fields. In this study, we sought to investigate the accuracy of this method.

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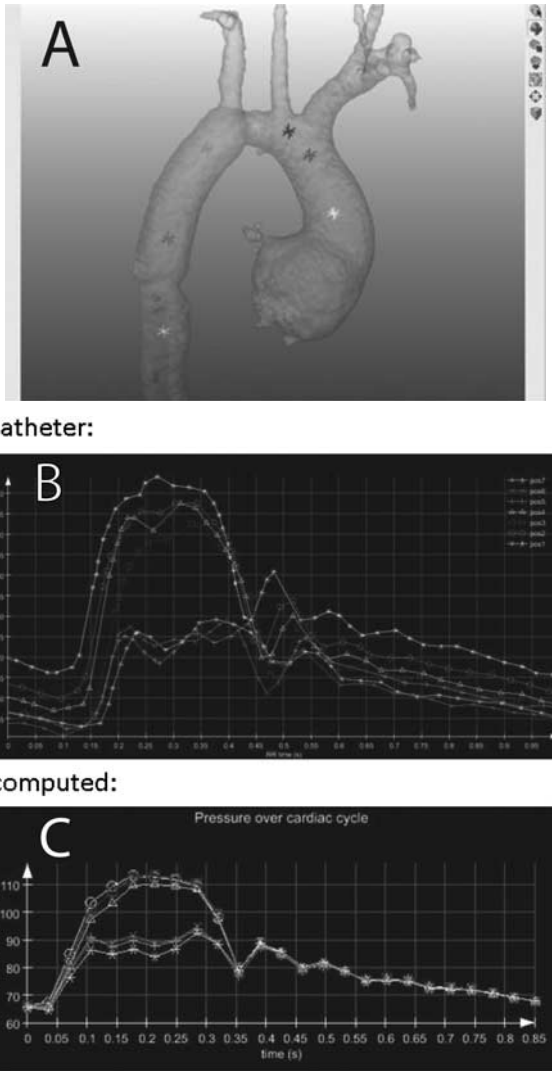
Methods: Five patients (age range 14 to 40, mean 20 years, n = 1 male, n = 4 female) with re-coarctation of the aorta were referred to invasive diagnostic catheterisation. Pressures were acquired at several predefined locations along the thoracic aorta. Prior to catheterization, patients were studied by MRI including 4D VEC MRI of the aorta. Relative 3D pressure maps were computed based on the Navier-Stokes equation. Data was calibrated with one invasive pressure point obtained in the ascending aorta for computing of absolute pressures. Agreement of MRI and catheter derived peak systolic pressures were compared by Bland-Altman test.
Results: All MRI data were of good quality for analysis. Bland-Altman test showed good agreement of peak systolic pressures with a bias of 3,3 ± 3,3 mmHg. Maximal gradients were 20 mmHg (mean, range 10 to 25 mmHg) across the localized coarctation in four patients.
One patient had a summation-gradient of 25 mmHg between ascending and descending aorta due to several mild stenoses along the thoracic aorta. The figure shows a representative case with a reconstructed MR angiography (A) and six locations for pressure measurements via catheter (B) and computed pressure curves (C).



Conclusion: The assessment of non-invasive pressure maps in aortic coarctation can be accurately obtained by non-invasive 4D MRI. This technique might evolve to an alternative to invasive diagnostic catheterisation for assessment of pressures.

PW4-7

The influence of respiration on blood flow in children – validation by realtime MR velocity mapping at 3 Tesla

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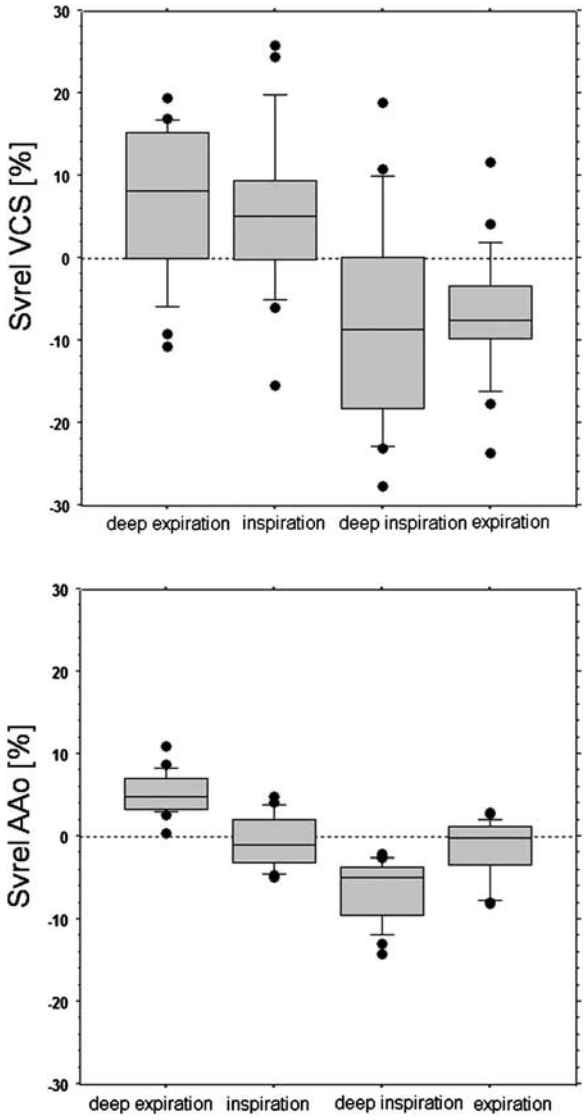
Introduction: Respiration can strongly influence cardiac preload which is especially important for children with CHD. We investigated realtime quantitative flow (QF) in healthy children with high temporal resolution to get unique hemodynamic information during respiratory phases.



Methods: QF was recorded on a 3T-TXMR scanner (Philips) using a non-triggered, free-breathing, real-time phase-contrast EPI sequence (RT-QF, In-plane resolution 2.7×2.7 mm^2, slice thickness 6 mm, acquisition time 12s). Temporal resolution below 25 ms was achieved using a SENSE factor of 4 combined with half-Fourier. Measurements were performed in the

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ascending aorta (AAo) and vena cava superior (VCS) in 23 healthy kids (13 male, age = 13.6 ± 3.6y) and compared with a validated conventional QF sequence (Ref-QF). Respiratory volumes were assessed using a Spirostik (Fa Geratherm) simultaneously.
Results: High agreement was observed comparing "mean" SV provided by RT-QF and Ref-QF in AAo (mean ± SD; limits-of-agreement: 3.0 ± 11%; -18.9 to 25, r = 0.934), whereas some higher overestimation was found for RT-QF in VCS (10.0 ± 10.6%; -11.1 to 31.1, r = 0.882) using Bland Altmann statistics. RT-QF SV varied much more in the VCS than in the AAo during respiration (mean ± SD [%]; deep expiration, inspiration, deep inspiration and expiration. VCS: 6.7 ± 8.9; 5.6 ± 9.6; -7.3 ± 12.5; -6.7 ± 7.6, AAo: -5.4 ± 2.4; -0.6 ± 3.1; -6.5 ± 3.6; -1.2 ± 3.4). ANOVA demonstrates that all volumes belonging to the various respiratory phases are statistically different of each other for VCS and AAo ($p < 0.001$) except the relation "deep expiration/inspiration" ($p = 0.69$, VCS), "deep inspiration/expiration" ($p = 0.85$, VCS) and "inspiration/expiration" ($p = 0.52$, AAo). With forced in- and expiration this differences in respiration-dependent flow augmented parallel to the respiratory volumes.
Conclusions: Impact of the dynamics of respiration on blood flow pattern in thoracic vessels can precisely be quantified in healthy children using RT-QF. This may especially be important for the assessment of children with a Fontan's circulation. Furthermore it raises the question to what extent flow in the great thoracic vessels is attenuated by breath-holding if fast flow mapping methods are applied. Using RT-QF, no respiratory gating or ECG triggering are needed. Furthermore, cardiac arrhythmia should no longer preclude safe and reliable flow measurements.

PW4-8
Assessment of Diffuse Myocardial Fibrosis Using Small-Animal Look-Locker Inversion Recovery T1 Mapping

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Background and Objectives: Myocardial fibrosis is an important determinant of heart failure. Patchy focal fibrosis can be visualized by late-enhancement techniques of gadopentetate dimeglumine. We hypothesized that diffuse myocardial fibrosis can be quantified by measuring myocardial extracellular volume (ECV) of gadopentetate dimeglumine in a small-animal model using Look-Locker inversion recovery T1 mapping.
Methods and Results: Sprague-Dawley rats (n = 10) were subjected to continuous angiotensin-2 (AT2) infusion for 2 weeks via a subcutaneously implanted minipump system. Magnetic resonance imaging (MRI) was performed both before and after AT2 infusion. The MRI protocol included multislice cine imaging and before-and-after contrast small-animal Look-Locker inversion recovery T1 mapping and late gadolinium enhancement imaging. Myocardial ECV was calculated from hematocrit and T1 values of blood and myocardium. During the course of AT2 infusion, the mean SD systolic blood pressure increased from 122 ± 10.9 to 152 ± 27.5 mm Hg ($P < 0.003$). Normalized heart weight was significantly higher in AT2-treated animals than in control littermates ($P < 0.033$). Cine MRI documented concentric left ventricular hypertrophy. Postcontrast myocardial T1 times were shortened after treatment (median [interquartile range], 712 [63] versus 820 [131] ms; $P < 0.002$). Myocardial ECV increased from 17.2% (4.3%) before to 23.0% (6.2%) after AT2 treatment ($P < 0.031$), which was accompanied by perivascular fibrosis and microscarring on myocardial histological analysis. There was a moderate level of correlation between ECV and collagen volume fraction, as assessed by histological analysis ($r = 0.69$, $P < 0.013$).
Conclusions: In a small-animal model of left ventricular hypertrophy, contrast-enhanced T1 mapping can be used to quantify diffuse myocardial fibrosis.

PW4-9
The role of Semi-Supine Cycle Ergometry Stress Echocardiography in Decision Making for Surgery in Children with Aortic Insufficiency

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Introduction: The criteria for surgery for children with aortic insufficiency (AI) have not been empirically developed, although the response of left ventricular (LV) function to exercise is well documented in adults. Semi-supine cycle ergometry (SSCE) stress echocardiography can be used to assess exercise capacity and cardiac function during staged exercise. The aim of this study was to assess the responses to SSCE in children with AI.
Methods: We retrospectively reviewed stress echocardiograms performed in 15 children with AI (Patients) and compared them to a healthy control group (Controls) (n = 29). Subjects exercised on a semi-recumbent cycle ergometer to volitional fatigue. Workload was progressively increased every three minutes by 20-40 Watts, depending on the size of the patient. Echocardiography, Doppler, heart rate (HR) systolic (SBP) and diastolic (DBP) blood pressures were taken at rest, 1.5 minutes into each stage, and post exercise. LVED, LVES, LVPWs, shortening fraction (SF), rate corrected mean velocity of circumferential fiber shortening (MVCFc), wall stress at peak systole (sPS), left ventricular stroke volume index (SVI), and left ventricular cardiac index (CI) were obtained.
Results: Aortic cross sectional area was higher in patients ($p = 0.02$). At rest, HR and SBP were similar, DBP was lower ($p < 0.001$), LVED and LVES were larger ($p = 0.003$; $p = 0.009$, respectively) and LVPWs was thicker ($p = 0.003$) in patients. SF, MVCFc, and sPS were similar between groups. SVI and CI were higher ($p < 0.001$) in patients. Cumulative work was similar between patients and controls. At peak exercise, LVED and LVES remained larger ($p < 0.001$). Although the percent change in LVES, SF, sPS from rest to peak exercise was lower ($p = 0.004$; $p = 0.03$; $p = 0.002$, respectively), SF and MVCFc were similar and sPS was higher in patients ($p = 0.001$). SVI and CI remained higher at peak exercise ($p = 0.005$; $p = 0.02$, respectively).
Conclusions: AI patients had normal exercise capacity, with appropriate rises in SBP and HR. The increase in SF and MVCFc indicates adequate recruitable myocardial reserve. SSCE may be helpful in decision-making for surgery in children with AI.

PW4-10
Assessment of Ventricular Pressure-Volume Relations and Myocontractility by Real Time Three Dimensional Transesophageal Echocardiography Coupled with Diagnostic Catheterization

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Introduction: In physiology, pressure-volume relations (pv-loops) are still the gold-standard for studying inotropic myocardial systolic and diastolic performance. Technical challenges for simultaneous measurement of ventricular pressures and volumes are, however, a barrier for their introduction into the clinical context.
The objectives of this study were to determine the feasibility and accuracy of measuring left ventricular (LV) pressure volume relationships and derived parameters of myocontractility in a large animal model using real time three-dimensional transesophageal echocardiography (RT3DTEE) coupled with catheterization. Conductance catheter (CC) pv-loops served as gold standard.
Methods: Six pigs (median weight 34 kg) were studied. CC (6 F, Millar, TX) were positioned in the LV apex via right carotid artery access (MPVS Ultra, Millar and PowerLab, AD Instruments). Simultaneous CC and RT3DTEE ('3D Opt' single beat mode, X7-2 X matrix, iE33, Philips) data were obtained with the animal ventilated (end-expiration), and paralyzed (vecuronium) at 3 different conditions: baseline, inferior vena caval (IVC) occlusion, and IVC occlusion during dobutamine (5 μg/kg/min). All measurements in each condition were performed thrice. CC derived LV pressures (Labchart) were integrated with RT3DTEE volumes (TMQ, QLab) on a beat-to-beat basis using an ECG trigger signal. From CC and RT3DTEE derived pressure volume loops, the end systolic pressure volume relations (Emax) were determined.
Results: Bland-Altman analysis showed excellent agreements between the RT3DTEE and CC for parameters derived at baseline and with dobutamine. At baseline, the mean ± SD were (mmHg/mL) Emax-CC 1.86 ± 1.1 and Emax-RT3DTEE 1.78 ± 1.2 ($p = 0.502$). On dobutamine, the means were Emax-CC 3.43 ± 1.5 and Emax-RT3DTEE 3.60 ± 1.23 ($p = 0.171$). Emax (mean ± SD) normalized to end diastolic volumes were 0.034 ± 0.02 for CC and 0.033 ± 0.02 for RT3DTEE at baseline ($p = 0.433$); the respective values were 0.081 ± 0.04 and 0.084 ± 0.03 on dobutamine ($p = 0.133$).
Conclusions: Pv-loops and Emax can accurately be assessed by RT3DTEE and there is good agreement with CC methods. Thus, the use of RT3DTEE might facilitate the introduction of intrinsic myocardial functional parameters in the clinical context.

PW4-11

An child-focused educational program allows INR stability and safety in children treated with VKA.

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Background: Improvement of anticoagulation control in children using a point-of-care International Normalized Ratio (POC INR) monitor in the home setting requires a robust parent/patient education program (EP).
Objective: To evaluate prospectively the efficiency of a child-focused EP based on group sessions for the children and their families using specific educational tools including a specially adapted game.
Patients and Methods: All children receiving VKA and their parents were proposed to follow a standardized training program (initial and reinforcement sessions) with medical support. Program efficacy was assessed by the analysis of (i) the time spent in the therapeutic range (TTR), (ii) the compliance with the treatment and INR control, (iii) VKA adverse events, (iv) patient satisfaction and (v) the accuracy of POC INR vs. laboratory INR.
Results: We enrolled 104 children (median age 8 years [3 months to 17 years]) receiving VKA for cardiac diseases in 96%. Median follow-up was 481 days (range 70-1001). The median TTR was 81.41% (36 to 100%). During reinforcement session, the game evaluated a very good knowledge of VKA treatment of patients and their family and their excellent capacity to react under numerous given situations. There were one severe haemorrhage and no thrombosis. 102 families were satisfied and wanted to continue this EP. Observance was adequate as POC INR was performed only when requested and parents did not perform additional INR controls. POC INR and laboratory INR were identical.
Conclusion: No patients of our series were excluded for any reasons and we consider that this inclusiveness is a valuable asset of our self-testing EP associated to an effective system of medical supervision. The TTR is among the highest TTR observed for home monitoring in paediatric studies and it is sustainable between 2008 and 2011. The key factor in the success of our EP is the empowerment conferred upon the children and their families considered as mature individuals responsible for their own health concerns. Moreover, the game allows the medical team to evaluate not only the patient's knowledge about his treatment and the practice of self-testing, but also the patient's adaptability to specific situations.

PW4-12

Clinical Experience with Thrombolytic Therapy using Low Dose of Tissue Plasminogen Activator in the Management of Intracardiac and Major Vessels Thrombosis

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Introduction: Incidence and associated morbidity to intravascular thrombosis are an increasing problem, especially patients admitted to pediatric cardiac intensive care unit (PICU). Experience with tissue plasminogen activator (t-PA) and dosage recommendations are limited.
Objective: To analyze the efficacy and side effects of low doses of t-PA for the management of acute intracardiac and major vessel thrombosis in children.
Patients and Methods: Prospective observational study. 34 children with congenital heart disease admitted to PICU, between 2 days of life and 13 years (53% <1year), treated with low doses (0,01-0,06 mg/kg/h) of continuous intravenous thrombolytic therapy with t-PA were studied between 2008-2011. Indications for treatment were: evidence or clinical suspicion of intravascular and intracardiac thrombosis, based on ultrasound and angiographic diagnosis. Treatment was ended when: clinical improvement, partial/complete resolution of thrombus and/or important side effects was achieved.
We collected demographic, clinical(ex, recent surgery), and laboratory(ex, D-dimer) data; treatment(ex, anticoagulants); t-PA dosage and duration; administration method(local-systemic); efficacy(complete or partial thrombus resolution based on ultrasound or angiography) and side effects: minor bleeding(ex, epixtasis), major bleeding(ex, retroperitoneal bleeding). Clinical and laboratory surveillance were performed before, during and 12-24h after treatment.
Results: 5 patients presented venous thrombosis and 9 arterial. 9 patients Fontan conduit thrombosis, 8 right atria thrombosis,

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one systemic-pulmonary fistulae thrombosis, one valved conduit(Labcor) thrombosis in mitral position and one left ventricle thrombus. In two patients, thrombus appeared, during ECMO support, and in one during mechanical biventricular support. 30 patients underwent previous surgery. 17 patients had catheter related thrombosis,12 postop, and 5 post-catheterization. Ultrasound diagnosis was performed in 25 and angiography in 8. The 34 patients received low doses of t-PA, medium 0,036, SD(0,014), medium duration 30 hours(range 5-148);20 patients received previous intravenous bolus;26% received local administration by placing a microcatheter under angiographic guidance;26 patients received previous treatment (heparin, antiplatelet). Treatment was effective in 30 patients(43% partial resolution). Major bleeding appeared in 5 cases(2 with mechanical cardiac support, 4 with systemic administration), and minor bleeding in 10. Mortality was 6%. There were no differences between laboratory values pre versus post treatment.
Conclusion: Low dose t-PA treatment is effective for acute intracardiac and mayor vessel thrombosis treatment, with a low range of serious complications.

P-1
Pharmacologic cardiac resynchronization in children.
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We present the case of 1 3/12 years old male toddler with Wolff-Parkinson-White syndrome (WPW) and severe LV dysfunction due to early activation of interventricular septum via the septal AP and delayed activation of the left ventricular free wall. This resulted in intraventricular wall motion delay with impairment of LV function (LV fractional shortening (FS) of 14% and a biplane LV ejection fraction (EF) of 31%). 2D strain analysis revealed intraventricular dyssynchrony with septal to posterior wall motion delay (SPWMD) of 350 ms. Interventricular mechanical delay (IVMD) was 65 ms. After propafenone administration (1 mg/kg i.v.) preexcitation disappeared due to conduction block in the AP. On oral propafenone (275 mg/kg/m2) no recurrence of preexcitation was seen. LV function improved significantly with FS of 29% and EF of 55%. No interventricular dyssynchrony was measured in 2D strain with SPWMD of 40 ms and IVMD of 5 ms.Preexcitation is still absent after 24 month of follow up and cardiac function is normalized with EF of 81% and FS of 48%.
Conclusion: Septal AP′s are a rare cause of intraventricular dyssynchrony and severe LV dysfunction in children. Cardiac resynchronization can be achieved by induction of conduction block in the AP using propafenone as a safe and effective alternative to catheter ablation in very young children.

P-2
Optimal Dose Of Adenosine Effective For Treating Supraventricular Tachycardia In Children
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Introduction: Supraventricular tachycardia (SVT) is the most common tachyarrhythmia in children. These tachyarrhythmias are poorly tolerated and potentially fetal in children. The dose of adenosine, although well defined in adults, is not adequately established in children.
Methods: All children presenting to the department of paediatric cardiology, The Children's Hospital, Lahore with SVT from July 2008 to June 2011 were administered adenosine in rapid boluses according to PALS guidelines using incremental doses of 100, 200 and 300 μg/kg. The response was recorded on 12 lead ECG. Reversion to sinus rhythm for 3 min was considered as effective response. Pre-excitation was documented and echocardiography performed on all children after attaining sinus rhythm.
Results: Eighty five patients were treated for 110 episodes of SVT with adenosine. M:F ratio was 2.2:1. The mean age ± SD was 27.9 ± 48.1 months. Adenosine was effective in reverting 97 episodes of SVT to sinus rhythm (88.2%). A dose of upto 100 μg/kg was only effective in 36.4% episodes of SVT. Two hundred μg/kg was effective in 44.3% of those not responding to 100 μg/kg dose (n = 31/70, cumulative 71/110, 64.5%). A dose of 300 μg/kg was effective in further 25 patients not responding to lower doses (n = 25/38, 65.8%; cumulative 97/110, 88.2%). Mean ± SD effective dose of adenosine was 185.3 μg/kg ± 81.0 μg/kg with median effective dose of 200μg/kg. Significantly higher dose of adenosine was required in children with underlying pre-excitation, n = 18/97 (220.8 μg/kg ± 67.6 μg/kg vs 177.2 μg/kg ± 81.9 μg/kg, p = 0.039). The effective dose was lower in patients with underlying dilated cardiomyopathy or congenital heart disease but difference was not statistically significant.
Conclusion: Adenosine is an effective drug in treating SVT in children but a dose of 100μg/kg may not be adequate as first bolus. Higher starting dose of 200μg/kg may be administered in majority of children specially with pre-excitation.

P-3
Effects of flecainide therapy on arrhythmias and inappropriate shocks in children with catecholaminergic polymorphic ventricular tachycardia
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Purpose: Current recommendations for therapy of catecholaminergic polymorphic ventricular tachycardia (CPVT) include ß-blockade and implantable cardioverter defibrillators (ICD). Patients may experience recurrent arrhythmias, ICD shocks and sudden death despite optimal medical therapy. It was recently shown that flecainide directly targets the molecular defect in CPVT by inhibiting premature Ca++ release. This study evaluated the efficacy and safety of flecainide in our CPVT children.
Methods: We collected data from the genotype-positive CPVT children started on flecainide in our tertiary pediatric cardiac center.
Results: The first case is a boy who was diagnosed with CPVT at 3 years of age. He was managed with nadolol (75 mg/m^2 BSA daily) with no further occurrence of syncope. At age 11, we noted bigeminal ventricular premature beats and couplets during exercise testing and one episode of sustained ventricular tachycardia on Holter recording despite nadolol titrated to maximum tolerable dose, so that an ICD was implanted. A few months later, inappropriate shocks were delivered due to episodes of sinus tachycardia. Holter monitor showed multiple episodes of sinus tachycardia and non-sustained ventricular tachycardia. Flecainide (100 mg = 4 mg/kg BW daily) was added to the ß-blocker regimen. At 18-months follow-up, no ICD shock occurred and repetitive Holter recordings and exercise testings excluded sinus tachycardia and arrhythmias. The second case is a girl who was diagnosed with CPVT at 8 years of age. At age 13, we noted bigeminal ventricular premature beats and couplets during exercise testing and on Holter

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recording despite nadolol titrated to maximum tolerable dose. Flecainide (100 mg = 2 mg/kg BW daily) was added to the ß-blocker regimen. At 6-months follow-up, the patient has complete suppression of exercise-induced ventricular arrhythmias and Holter recordings also excluded arrhythmias.
Conclusions: Recent experimental evidence suggested that flecainide can suppress CPVT. The presented cases support that novel strategy and show that flecainide is effective in preventing exercise-induced ventricular arrhythmias and also inappropriate shocks in children with CPVT.

P-4

Prevalence, Mutation Spectrum and Cardiac Phenotype of the Jervell and Lange-Nielsen Syndrome in Sweden

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Objectives: To explore the prevalence, mutation spectrum and cardiac phenotype of the uncommon Jervell and Lange Nielsen Syndrome (JLNS) in Sweden.
Methods: We performed a national inventory of clinical cases. Genotype and geographical origin was investigated. Clinical data, including information regarding timing between medical treatment and/or interventions and cardiac events, was collected from medical records and a personal interview.
Results: 19 cases in 13 Swedish JLNS families were identified. The mutation spectrum consisted of eight KCNQ1 mutations, whereof p.R518X in 12/24 alleles. Geographic clustering of four mutations (20/24 alleles) and similarities to Norway's mutation spectrum were seen. A high prevalence of heterozygous mutation-carriers in the Swedish population was suggested.
Three pediatric cases on β-blockers since birth were as yet asymptomatic. Seven out of 16 symptomatic cases had suffered an aborted cardiac arrest and four had died suddenly. Differences between the sexes were apparent, with regards to age at debut (earlier for males) and frequency of cardiac events in adulthood (higher for females). QTc prolongation was significantly longer in symptomatic cases (mean 605 ± 62 ms vs. 518 ± 50 ms, p = 0.016). Relation between cardiac events and medical interventions; β-blocker therapy, including data on type and dosage (n = 15), left cardiac sympathetic denervation (LCSD, n = 3) and implantation of a cardioverter-defibrillator (ICD, n = 6) was described and efficacy discussed.
Conclusion: A JLNS prevalence >1:200 000 in preadolescent Swedish children was revealed, and Scandinavian founder effects could explain 83% of the Swedish mutation spectrum.
Due to the severe cardiac phenotype, the importance of optimal β-blocker therapy and compliance, and consideration of ICD implantation in case of therapy failure is stressed.

P-5

Gastrointestinal symptoms, signs and associated iron-deficiency anaemia in the Jervell and Lange-Nielsen Syndrome

Clinical Phenotype Beyond the SurdoCardiac Syndrome
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Introduction: The KCNQ1 gene is essential for gastric acid secretion. The Jervell and Lange-Nielsen Syndrome (JLNS), originally described as a surdocardiac syndrome, is characterized by impaired KCNQ1 function. Here we investigate possible symptoms and signs related to loss of gastric acid secretion in genotyped JLNS cases with KCNQ1 mutations.
Methods: We investigated 14 genotype-ascertained JLNS cases with KCNQ1 mutations (age 31 ± 24 years, range 4-87, whereof 9 females), initially by personal interview and medical record review.
Current testing for iron-deficiency anaemia and gastrointestinal function/inflammation (fasting levels of gastrin and pepsinogen, and faecal calprotectin) was performed.
Results: Previous anaemia (12/13 tested cases), subjective gastrointestinal symptoms (13/14 cases) and a previous positive faecal haemoglobin test (n = 4) was revealed. Endoscopy had been performed in 5 cases revealing no case of coeliac- or inflammatory bowel disease but 2 cases of mucosal hyperplasia and one ventricular tumour. One sibling with JLNS had died from gastric carcinoma.
At current testing signs of iron-deficiency anaemia or iron-substitution was present in 9/12 tested cases. Elevated levels of gastrin (in 7/9 cases, mean 379 ± 426 pmol/L, range 22-1285, normal<60), pepsinogen (in 6/7 cases, mean 366 ± 332 μg/L, range 121-892, normal<130) and faecal calprotectin (in 9/9 cases, mean 322 ± 195 mg/kg, range 77-641, normal<50) were present. A significant correlation between elevated gastrin levels and current anaemia was found (p = 0.028), and in no case was anaemia present without elevated gastrointestinal markers.
Conclusion: We propose that JLNS phenotypically includes iron-deficiency anaemia and gastrointestinal symptoms/signs, secondary to hypochlorhydria on the basis of KCNQ1 mutations. The resultant hypergastrinaemia is a risk factor for gastrointestinal cancer. Further studies are needed to elucidate the importance of hypochlorhydria, hypergastrinaemia and gastrointestinal inflammation in JLNS. Monitoring of JLNS cases with regards to developing anaemia and gastrointestinal cancer should be considered.

P-6

Diagnosis of Congenital Long QT Syndrome: Do all Clinically Suspected Patients warrant Genetic Testing?

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Introduction: Diagnosis of congenital long QT syndrome (LQTS) involves clinical, electrocardiographic (ECG) and genetic evaluation. Genetic testing however has the disadvantage of not being uniformly available and is costly and time-consuming. This study aimed to investigate if all clinically suspected patients warrant genetic testing.
Methods: Study population included 214 unrelated probands referred for genetic counselling and predictive testing (KCNQ1/KCNH2/SCN5A/KCNE1/KCNE2/KCNJ2). Demographic and clinical characteristics, resting heart-rate and QTc from the earliest available pre-therapy ECG, Schwartz score (SS) and genotyping results were documented for all patients. Maximum QTc on Holter (QTcH) was analyzed for patients with a SS ≤3. QTc/QTcH >450 ms were considered prolonged.
Results: Age at diagnosis was 31 ± 21 years (range 0-85 years), heart-rate 73 ± 19 bpm, QTc 478 ± 64 ms, 68% were female and

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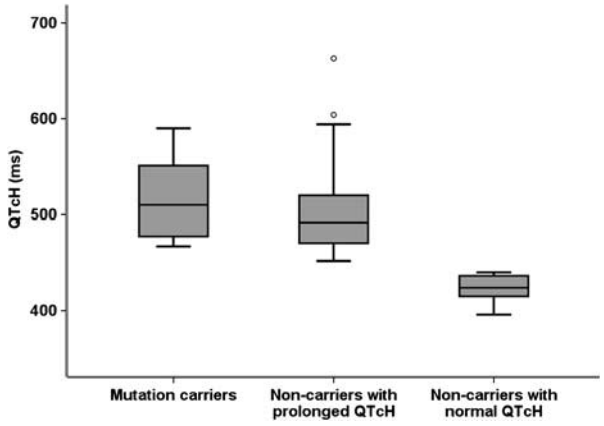
78% had symptoms suggestive of LQTS. KCNQ1/KCNH2/SCN5A/KCNJ2 mutations were present in 34/49/15/2 patients respectively. Among patients with SS ⩾3.5 (n = 103), 75% were mutation-carriers. Among patients (n = 111) with SS ⩽3, QTc was prolonged in 32 (477 ± 20 ms, 28% mutation-carriers) and normal in 79 patients (417 ± 19 ms, 9% mutation-carriers). Among the latter 79 patients, QTcH was normal (425 ± 13 ms) in 16, all of whom did not carry a putative pathogenic mutation; QTcH was prolonged (506 ± 44 ms) in 63 patients, 7 of whom were mutation-carriers (Figure). Sensitivity and specificity were 83% and 79% for SS ⩾3.5 in the total cohort, 56% and 76% for QTc cut-off 450 ms among subjects with SS ⩽3, and 100% and 22% for QTcH cut-off 450 ms in subjects with SS ⩽3 and normal baseline QTc.

Conclusions: This study demonstrates that in the challenging group of patients with a clinical presentation suggestive of LQTS and a normal resting QTc, Holter QTc cut-off of 450 ms was 100% sensitive and can potentially be used to rule-out LQTS, especially when genetic testing is not readily available or feasible.



Maximum Holter QTc (QTcH) based on LQTS mutation-carriership in patients with normal baseline QTc (Open circles indicate outliers).

P-7

Outcome of children and adolescents undergoing invasive testing for asymptomatic Wolff-Parkinson-White preexcitation

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Introduction: Results of a risk stratification strategy in asymptomatic WPW preexcitation in the young were evaluated.

Methods: 85 consecutive pts with a WPW pattern undergoing from 10/2000 to 8/2011 an invasive EP study for risk stratification were studied. All were without antiarrhythmic medication and had persistent preexcitation during exercise. Potentially dangerous accessory pathway (AP) properties were defined as follows: antegrade effective refractory period (AP ERP) ⩽250 ms, shortest preexcited RR interval during atrial fibrillation/rapid atrial pacing (SPERRI) ⩽250 ms or inducible AV reentrant tachycardia (AVRT).

Results: Age at evaluation was median 14.9 (IQR 12.5 – 16.6) yrs (<18 yrs in all). Structurally normal heart was present in 82 pts, hypertrophic cardiomyopathy in 3 pts. A single manifest AP was found in 81 pts, two APs in 4 pts. At least one risk factor was present in 35/89 APs (39.3%) at baseline and in additional 14/89 (15.7%) after isoproterenol: rapid antegrade conduction in 36/89 (Table) and inducible AVRT in 25/89 APs. Ablation was performed in 40/49 potentially dangerous APs (81.6%) and was deferred in 9/49 (18.4%) because of proximity to the AV node. In addition, 23 low risk APs were ablated based on patient/parental decision.

AP properties (N = 89)	baseline	isoproterenol
AP ERP median (IQR)	295 (270-320)	270 (248-290)
SPERRI median (IQR)	295 (256-330)	240 (230-283)

Conclusions: Using a currently accepted risk stratification strategy including isoproterenol challenge, 55% of the evaluated clinically asymptomatic APs exhibited either rapid antegrade AV conduction or AVRT inducibility. Safe ablation (remote from AV node) could be performed in 81.6% of these APs. (PK and JJ were supported by a grant of the Internal Grant Agency of Ministry of Health of the Czech Republic NT 12321-3/2011).

P-8

Management Of T-Wave Oversensing After Extracardiac ICD Implantation In A Pediatric Patient With Brugada Syndrome

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Introduction: The extracardiac system offers a safe and effective approach for ICD implantation in infants and small children. The implantation technique includes placement of the defibrillation lead subpleurally along the course of the 6th rip, attachment of bipolar steroid-eluting sensing and pacing leads to the anterior wall of the right ventricle, and implantation of the device subdiaphragmally as "active can". This ICD configuration allows together with modern algorithms to overcome T-wave oversensing due to alternative sensing options.

Case report: An extracardiac ICD (Medtronic Protecta VR) was implanted in 18-months-old patient (body weight 9.8 kg) due to familial Brugada syndrome. Intraoperative DFT was ⩽10 J, intraoperative RV amplitude was 7.5-5.5 mV, and amplitudes during VF varied from 3.0 to 7.0 mV during DFT testing. RV sensing was programmed to 0.45 mV with the auto adjustment sensitivity algorithm. 6 months later, on routine follow-up ICD interrogation recurrent episodes of T-wave oversensing were observed. Up to 10 inappropriate shocks had been avoided by the novel T-wave oversensing (TWOS) algorithm (SmartShock TechnologyTM, Medtronic Inc.). By discrimination of R- and T-waves through differential filters together with R- to T-wave beat to beat pattern analysis therapies were withhold. In addition, R-wave amplitude was reduced to a mean of 3.5 mV with a variance from 1 to 5 mV. After reprogramming RV sensing to 0.6 ms T wave oversensing was still present due to comparable R and T-wave amplitudes. With programming sensing to 1 mV, intermittent undersensing of R-wave was noted. Therefore, ventricular sensing was programmed between the RV leads

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and the subpleural shock electrode with an estimated sensing distance of 8 cm. With this configuration sensing was 9.8 mV and no further T wave oversensing was observed with a RV sensing at 0.6 mV. Induced VF was appropriately detected and the DFT was stable at $\leq$ 10 Joule. After a follow-up of 2 months, no further T wave oversensing was noted.

Conclusion: Alternative sensing options of the extracardiac ICD together with modern algorithms may prevent T wave oversensing and avoid traumatic inappropriate shocks and reoperations.

P-9

Paroxysmal tachycardia with wide QRS complexes, right bundle branch block pattern and left axis deviation: A seldom electrophysiological mechanism.

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Introduction: Atrioventricular pathways with slow and exclusively anterograde decremental conduction (Mahaim fibers) are almost exclusively located at the tricuspid valve annulus. Left-sided Mahaim fibers have only rarely been described.

Case presentation: An electrophysiological study was performed in a 15-year-old boy due to recurrent episodes of a wide QRS complex tachycardia with an inferior axis and a right bundle branch block pattern. During sinus rhythm no preexcitation was present. During atrial pacing widening of the QRS complexes was noted exhibiting a right bundle branch block pattern and left axis deviation identical with the morphology of the clinical tachycardia. There was no change of the AV interval but fusion of the His potential with the onset of the right bundle branch potential of the His catheter which was still preceded by the onset of the wide QRS complex of the surface ECG verifying preexcited QRS complexes. Right ventricular pacing demonstrated earliest atrial retrograde activation at the His bundle catheter.

Clinical tachycardia with a cycle length of 292 msec and retrograde 1:1 ventriculo-atrial activation was induced by programmed atrial pacing. Earliest ventricular activation was noted at the left posterior aspect of the mitral valve annulus and earliest retrograde atrial activation was noted at the His bundle catheter. Tachycardia could be entrained by atrial extrastimuli.

Subsequently, detailed mapping was performed at the atrial aspect of the mitral valve annulus during tachycardia. In the left posterior region local ventricular electrograms preceded the ventricular electrograms of the coronary sinus catheter leads by 5 msec. RF application at this location terminated the tachycardia by antegrade conduction block. Subsequently, there was no further evidence of preexcitation during repeat atrial pacing and tachycardia was not inducible anymore. During a follow-up of 2 years no tachycardia recurrence was noted.

Conclusion: Left-sided decrementally conducting accessory pathways are rare. In patients with atrioventricular or atriofascicular Mahaim fibers successful radiofrequency ablation can be performed by mapping the earliest ventricular activation at the mitral valve annulus.

P-10

Handheld ECG in analysis of arrhythmia and HRV in children with Fontan circulation.

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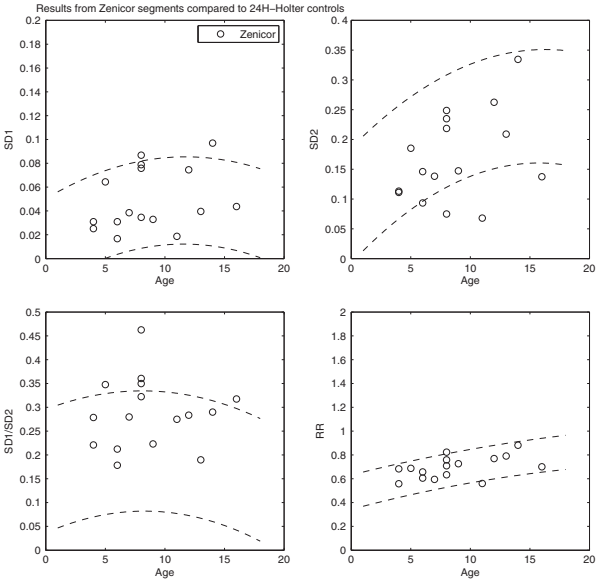
Objectives: Although modifications have improved the Fontan operation, arrhythmia remains a frequent problem.

The Fontan patient is, due to her/his significant hemodynamic abnormalities, susceptible to recurrent atrial tachycardia. Atrial tachycardia could be presented only with vague symptoms, as the heart rate will not be dramatically increased in presence of a 2:1 atrioventricular block. It is known that Fontan patients have reduced heart rate variability (HRV).

The aim of this pilot-study was to use an ambulatory short-term ECG system to study the prevalence of arrhythmia and to perform HRV analysis, using Poincare′plots.

Methods: The device, Zenicor-ECG®, is a small hand-held device with a display and two thumb sensors, providing for a bipolar extremity lead I. In the study the patient (and the parents) was instructed to apply the thumbs onto the sensors for 30 seconds for ECG registration. The assessment was performed during 14 day period–twice a day and if there were symptoms.



ECGs were analyzed for rhythm and arrhythmia. Each RR interval was plotted as a function of the previous RR-interval (Poincare′plot). Results were presented as SD1, SD2 and the ratio SD1/SD2. Two patients with a pacemaker were excluded from HRV-analysis. The results were compared to 24-h ambulatory ECG recordings in a control group of 66 healthy children.

Results: 18 patients, 5 girls and 13 boys aged 9.5 years (range 4.1-16.5) underwent short-time ECG-registration.

Fifteen of the patients were in sinus-rhythm, 1 patient had ectopic atrial rhythm, and 2 patients had intermittent pace-maker induced rhythm. We found no brady- or tachy arrhythmia, apart from in one patient where we found very frequent ventricular extra systoles in pairs and in bigeminia and 2 patients with frequent nodal replacement beats.

Poincare′plot shows the nature of RR interval fluctuations: four patients had lower SD2 than controls.

Conclusions: Arrhythmia was not commonly found by handheld ECG. However, this pilot-study is too small to draw conclusions of the prevalence of silent arrhythmia. Four patients with Fontan circulation showed a decreased SD2 which may indicate reduced

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long-term variability. It seems like hand-held ECG can be used for this purpose, but larger studies are needed.

P-11
Radiofrequency catheter ablation for atrial tachycardia in patients with complex congenital heart disease after intracardiac operation

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Background: Atrial tachycardia (AT) is known as a difficult complication in patients with complex congenital heart disease (CHD) late after surgical correction.
Purpose: We retrospectively analyzed the results of radiofrequency catheter ablation (RFCA) for AT.
Methods: We divided patients into three groups; group A: atrial switch (Mustard or Senning) procedure in 17 cases, group B: Fontan (atrio-pulmonary connection) procedure in 36 cases, group C: Fontan (total cavo-pulmonary connection) procedure in 3 cases.
Results: Induced ATs were isthmus-dependent atrial flutter, intra atrial reentrant tachycardia and atrial fibrillation. In group A, tachycardia substrate existed in the pulmonary venous atrium (PVA) in 15 of 17 cases. Catheter access to the PVA was transaortic approach in 2 patients, transseptal approach in 10, and buffle leak 3. We could not puncture the Senning route and could not eliminate the tachycardia in 4 cases. In group B, the mean number of induced tachycardia was four. RFCA for atrial fibrillation and unmappable AT in 15 cases were unsuccessful. We could not reach the PVA in 3 patients in group B, and could not ablate the tachycardia. Three cases in group C had isthmus-dependent atrial flutter in the PVA. RFCA was successful in all 3 cases, by a transaortic approach, using a puncture of conduit, and via a fenestration. The success rate was 76% (13/17) in group A, 50% (18/36) in group B, 100% (3/3) in group C.
Conclusion: There were many difficulties and limitations in RFCA for the patients with complex CHD after operation. ATs after atrial switch operation and TCPC procedure can be managed with catheter ablation with favorable success rate.

P-12
Predictive factors for future symptoms and dropout in patients with long QT syndrome in a single pediatric cardiovascular center in Japan

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Introduction: Long QT syndrome (LQTS) is a leading cause of sudden cardiac death due to arrhythmia in pediatric patients. A school-based ECG screening program to screen cardiovascular diseases uncovered many children and adolescents with LQTS. The aim of the study was to determine predictive factors for both the presence of symptoms and future dropout. We used data from a single pediatric center to unify the follow-up strategies.
Methods: Subjects were 143 consecutive LQTS patients (M:F = 72:71) who visited our center between April 2005 and December 2010, who were $\leq$ 20 years old, and included subjects who were previously followed by other hospitals. Thirty-seven subjects (26%) did not visit our hospital during the preceding 24 months (Dropout group). There were 106 final subjects (M:F = 49:57; mean age: 9.5 $\pm$ 5.1 yr; age range: 0–20 yr; mean follow-up period: 5.2 $\pm$ 5.6 yr). Subjects included 66 patients who were screened in the program (Screened group), 16 who visited clinics because of symptoms, and 24 who were diagnosed by family study or by chance.
Results: Four subjects (6%) in the Screened group experienced LQTS-related symptoms before diagnosis and 20 (30%) had a family history of LQTS. One subject died. She had a history of aborted cardiac arrest at 2 months, and died suddenly during sleep at 5 years of age. Logistic and multivariate regression analyses showed that the sole predictive factor for the presence of new symptoms after diagnosis was longer QTc values ($p = 0.02$) and that predictive factors for frequent symptoms were a longer follow-up period ($p < 0.0001$) and drug noncompliance ($p < 0.0001$). Logistic regression analysis showed that the presence of a family history ($p = 0.004$) was a significant predictor for the absence of dropout.
Conclusions: A school-based screening program is an effective method to identify children and adolescents with LQTS. Continued follow-up is important because the risk of a new episode increases with age in this group. Good drug compliance is crucial to prevent recurrent episodes. A new strategy is needed to prevent subjects without a family history from dropping out of visits.

P-13
Assessment of Performance of various QT correction formulae in Infants and Children

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Introduction: several formulae have been used to correct QT interval for heart rate. These are however known to over- or under-correct the QT interval. The purpose of this study was to ascertain which formula best corrected for heart rate in children of all ages
Methods: we enrolled a cohort of 600 healthy children. While in a quiet state a digital 12 lead electrocardiogram (50 mm/second) was recorded and stored. Subsequently, the QT and RR intervals were measured digitally from lead 2. The QT interval was corrected for RR interval by applying 6 formulae, Bazett, Fridericia, Framingham, van de Water, Hodges and Tabo. Linear regression equations for the corrected QT (QTc) intervals against RR intervals were obtained by the method of least squares and the slope and R square (R2) of equations were calculated. When both the slope and R2 were close to zero, the formula was judged to eliminate the effect of heart rate on QT interval.
Results: Mean age: 2,1 years, SD+3,5 (range 0–18 years). Mean QT 295 +29 ms, RR mean 514 + 121 ms; QTc Bazett 414 + 17 ms; QTc Fridericia 370 + 16 ms, QTc Tabo 384 + 15 ms; QTc Framingham 370 + 16 ms; QTc Hodges 403 + 20 ms, QTc Van De Water 337 + 20 ms.
Linear Regression plots of QTc against RR intervals: QTc Bazett slope – 0.06, R2 0.16; QTc Fridericia slope 0.053, R2 0.15; QTc Hodges slope -0.07, R2 0.193; Qtc Tabo slope 0.017, R2 0.019; QTc Framingham slope 0.065, R2 0.3; QTc Van de Water slope 0.13, R2 0.65.
Conclusion: the many QT correction formulae attests to the importance of adjusting QT interval for heart rate particularly in infants with increased basal heart rate. From linear regression analysis of the QTc interval against RR interval, this study illustrates that, of the published formulae, Tabo's formula best corrected the effect of RR on QT interval, even in infants, and showed superior dissociation of the QTc interval from RR interval (least slope and lowest R2). Further evaluation is in progress.

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P-14

Assessment of AV Conduction Following Cryoablation of Atrioventricular Nodal Reentrant Tachycardia in Children

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Introduction: Cryoablation is an effective and safe treatment for children with atrioventricular nodal reentrant tachycardia (AVNRT). Early-onset transient atrioventricular block (AVB) can be seen rarely following cryoablation of AVNRT, despite lack of any AVB at the end of the ablation procedure. The purpose of this prospective study was to assess AV conduction shortly after successful cryoablation of AVNRT in children.
Methods: From October 2010 to December 2011, a total of 33 children (14 females, 19 males) underwent cryoablation for AVNRT. A 6-mm tip cryoablation catheter was used in 30 patients (91%). An 8-mm tip catheter was used in 3 patients. The efficacy of the procedure was assessed in terms of disappearance of dual-AV node physiology and/or lack of inducibility of AVNRT. Inducibility of AVNRT was checked 30 minutes following last ablation lesion on and off metaproterenol. ECGs and 24-hour ambulatory Holter monitoring were performed prior to and following the ablation procedure. All procedures were one using the EnSite system (St. Jude Medical, St. Paul, MN).
Results: The mean age and weight was 12.3 ± 3.6 years and 44.5 ± 15.1 kg, respectively. Congenital heart disease was present in 3% of the patients. The acute success rate was 91%. The mean procedure time was $124.9 + 50$ minutes. Total cryoablation time was 2219 ± 1206 sec. Fluoroscopy was used only 3 patients and the mean fluoroscopy time was $4 + 2.9$ minutes. No major complications occurred. Two (6%) patients had variable degrees of transient AV block despite having normal AV conduction at the end of the procedure. One of these patients developed 2nd degree AVB which lasted for 2 hours and second patient experienced 2:1 AVB which lasted for 8 hours. Both patients had complete resolution of the AVB which was also documented at follow-up Holters.
Conclusion: Transient AVB can rarely occur shortly after cryoablation of AVNRT. Patients should be monitored at least for 24 hours in the hospital following cryoablation of AVNRT.

P-15

Catheter Ablation of Ventricular Tachycardia in Children Using a Limited Fluoroscopy Approach

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Introduction: Ventricular tachycardia (VT) can cause significant morbidity and mortality in the pediatric population. Curative therapy of VT can still be challenging in interventional electrophysiology. The aim of this study was to review our experience in catheter ablation of VT in children using the EnSite (St. Jude Medical, St Paul, MN) system.
Methods: Ten consecutive children with VT underwent radiofrequency catheter ablation using the EnSite system guidance in addition to fluoroscopy. EnSite system allowed us to either eliminate or significantly decrease fluoroscopy exposure besides its help in electroanatomical mapping.
Results: The origin of VT was in the right ventricular outflow tract in 6 patients, left ventricle in 3, and left aortic cusp in 1. The mean age and weight were 15 ± 2 years (range: 12 to 17) and 57.8 ± 6.4 kg, respectively. The mean procedure and fluoroscopy durations were 170.6 ± 53.2 minutes and 12 ± 14 minutes, respectively. No fluoroscopy was used in 3 patients. The acute success rate was 8/10 (80%). In 2 failed procedures, VT focus was epicardial. Recurrence was noted in 3 patients at a follow-up of 9 ± 7 months. Two of these patients underwent successful second procedures. The focus was epicardial and adjacent to the left main coronary artery in the third patient who had recurrence. Further procedure was not done to prevent coronary artery injury.
Conclusions: Catheter ablation of VT can be performed safely and effectively and with low fluoroscopy exposure in children using the EnSite system.

P-16

Combination of sinus node dysfunction and intestinal pseudo-obstruction in children and young adults: about 11 cases of this undiscribed association

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Introduction: Sinus node dysfunction (SND) is rare in children and is encountered mainly as a late complication of congenital heart surgery. It is sometimes found in patients with a structurally normal heart and can be inherited. Chronic intestinal pseudo obstruction (CIPO) is a rare disabling isolated condition in children. It is idiopathic in most cases. The combination of CIPO and SND has not been described.
Results: We report 11 French Canadian children (5) and young adults (6) that manifested a combination of symptomatic CIPO and SND (between 01/1998 and 03/2010).
In the pediatric population (non related patients), first signs of CIPO developed at mean age of 11.1 yrs (6.6–17 yrs). CIPO preceded SND by 4.2 and 5.5 yrs in 2 patients, CIPO and SND were simultaneous in 2 and SND preceded CIPO by 2 yrs in 1 patient. There were 4/5 boys. Three patients with severe symptomatic bradycardia, junctional rhythm and pauses required a pacemaker (PM) 1-6 months post diagnosis. They remain well 0.5-7.5 yrs post PM implant. The other 2 patients manifested tachy/brady syndrome requiring antitachycardia medication and did not need a PM. The 5 patients had dilated left atrium and ventricle on echo with normal systolic function, normal anatomy and no mitral regurgitation. One patient had left diastolic dysfunction. One patient died at 18.8 yrs in the post-operative period of multiorgan transplantation. During the same period, 6 other patients (2/6 boys), were diagnosed with CIPO in our institution without manifestations of SND. No family members of our affected pediatric patients suffered of CIPO or SND.
Among the adults (mean age 35 yrs, range 23-43 yrs; 4/6 men), all have CIPO (all diagnosed before age of 15 yrs) and have cardiac symptoms. All but 2 have PMs and tachycardias and the 2 others have sinus bradycardia and supra-ventricular tachycardias. Within these adults, there are 2 pairs of siblings. Genetic testings of patients and families are currently underway.
Conclusions: SND developed in 11 patients diagnosed with CIPO in our institution, mainly during childhood (7/11) or young adulthood (4/11). Genetic investigations are currently underway and a founder effect is so far most probable.

P-17

Idiopathic ventricular arrhythmias in children: factors determining the state of intracardiac hemodynamics

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Aim: To disclose factors which determine the state of intracardiac hemodynamics in children with idiopathic ventricular arrhythmias.
Methods and materials: 139 children at the age of 11,8 ± 4,21 years old with idiopathic ventricular arrhythmias were included into the study. The control group involved 18 children with previously registered no arrhythmia and cardial conduction disturbances, and also without structural heart anomalies and myocardium disease. Doppler-echocardiography and quantitative bloodpool SPECT were performed to assess the state of intracardiac hemodynamics.
Results: Increase of both left and right ventricles were disclosed in patients with ectopic activity more than 20%. Ejection fraction also decreased in comparison with the control group. Significant increase of both ventricles, and also decrease of their ejection fraction in comparison with the control group and with the group of single premature beats were defined in patients with group premature beats, and also with runs of nonsustained ventricular tachycardia. Enlarge of end systolic volume of right and left ventricle was discovered in patients with specific arrhythmia symptoms. In this case the ejection fraction was lowered. Increase of right and left ventricle volumes in comparison with the control group was disclosed in patients with negative arrhythmia dynamics for exercise test. Decrease of left ventricle ejection fraction was marked in patients with arrhythmia localization in left ventricle. With right ventricle localization increase of its end diastolic and systolic volume was discovered.
Conclusion: Thus, degree of ectopic activity, presence of group premature beats in a patient, symptoms and type of arrhythmia reaction for exercise test are master factors of intracardiac hemodynamics state. Disclosed in the study factors which influence upon intracardiac hemodynamics in patients with idiopathic ventricular arrhythmias may be the basis of algorithm of treatment indications for the given patient group.

P-18

Minimally Invasive Method for Epicardial Implantation of Implantable Cardioverter Defibrillators in Children

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Introduction: Epicardial implantation of Implantable Cardioverter Defibrillators (ICDs) is considered in the presence of an intracardiac shunt, venous access issue or small body size. More common technique of epicardial ICD implantation involves placement of epicardial patches through sternotomy. In the present study, our experience with epicardial ICD coil implantation using a minimally invasive method is reported.
Methods: Subjects who underwent epicardial ICD implantation between January 2010 and December 2011 were included. Clinical and procedural data of these patients were retrospectively evaluated.
Results: A total of 6 patients were included in the study. The median age was 6.2 years (range 3.8 -9.5). The median weight was 15.8 kg (range: 12-24). Implantation diagnoses included long QT syndrome (n = 3), catecholaminergic polymorphic ventricular tachycardia (n = 1), and hypertrophic cardiomyopathy (n = 1) and fast monomorphic ventricular tachycardia (n = 1). Minimally invasive method involved a subxiphoid incision in order to place the epicardial pacing leads. ICD coil was placed in the transverse sinus in 4 patients using an access path posterior to the heart. Second approach (anterior approach) involved a path anterior to the heart to reach the epicardial location posterior to the left atrial appendage and this was performed in 2 patients. The median defibrillation threshold (DFT) at implant was 10 J (range: 5 to 10 J). A lower DFT was observed in patients who had leads placed using the anterior approach (10 J vs. 5 J). Appropriate ICD shocks were observed in 3 patients during a median follow-up of 13 months (range: 5-18). No inappropriate shocks were noted. One patient suffered pericardial tamponade and required surgical drainage and medical therapy. A patient with long QT syndrome developed ICD storm and was successfully treated with left cardiac sympathetic denervation. No other complications were observed.
Conclusions: Epidardial ICD coil implantation using a minimally invasive method is effective in children in whom transvenous approach is challenging. Anterior implantation approach appears to result in lower DFT values. Further studies are needed to assess for the optimal technique for epicardial ICD and coil implantation in children.

P-19

Cardiac Resynchronization in a $1\frac{1}{2}$ Year Old Girl With Dilated Cardiomyopathy

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Introduction: Cardiac resynchronization has become an established tool for end stage heart failure in adults. In children experience with this kind of therapy is limited to a few series and case reports. Criteria of responders are not well defined in the pediatric age group. We report a case of successful CRT in a toddler with dilated cardiomyopathy (DCM) who was not eligible for heart transplantation (HTX).
Patient and Methods: A girl, 1 month of age, presented with heart failure and was diagnosed with DCM caused by HHV6 infection. She remained stable under medication for 5 months, when repetitive administration of Levosimendan was started. HTX was refused by the parents. At the age of $1\frac{1}{2}$ years (height 74 cm, weight 8.9 kg) she underwent mitral valve repair (downsized 28 mm Carpentier prosthetic ring) due to progressive annulus dilatation and additional volume load of the left ventricle. At the same time epicardial leads for biventricular pacing were implanted (right atrium: Medtronic 5071; right and left ventricle: Enpath Myopore). After a honeymoon of 6 months following mitral valve reconstruction the left ventricular function deteriorated and the leads were connected to a CRT device (Medtronic Consulta^{TM}), placed intraabdominally. AV delay was optimized and interventricular delay was programmed to 20 ms according to echocardiographic evaluation.
Results: Echocardiography revealed regression of left atrial area (18 to 5.4 cm^2) as well as left ventricular diameter (56 to 41 mm) within 9 months following CRT onset. Ejection fraction rose from 23 to 50%. Functional class improved from NYHA III to I, the values of B natriuretic peptide dropped from >35000 to less than 600 pg/ml. Postcapillary pulmonary hypertension was no longer present. The patient is showing excellent physical and mental development.
Conclusion: Additionally to pharmacological treatment, CRT is a viable option for the pediatric heart failure patient. Epicardial lead implantation makes this strategy applicable for younger patients, starting in infancy. CRT allows for bridging till HTX and yields potential as definitive therapy of heart failure.

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P-20

Electrophysiological properties of the atrioventricular node, slow and fast pathway in children at long term follow up after slow pathway radiofrequency catheter ablation

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The slow pathway radiofrequency ablation (RFA) is effective method of treatment in children with atrioventricular nodal reentrant tachycardia (AVNRT).
The aim of our study was to evaluate anterograde conduction, fast and slow pathway electropsysiological properties in children before and after slow pathway RFA, to determine the efficacy and safety of this method.
Material and methods: Noninvasive transesophageal electrophysiological investigation was performed in 32 patients (mean age 16.6 (2.5) years) at the follow up period (mean duration 3.2 (1.7) years) after the slow pathway RFA.
Results: The slow pathway function was observed in 24 patients at the follow up period, and in 28 patients after atropine sulfate. No one had paroxysms due to AVNRT.
Atrioventricular node conduction decreased significantly at the follow up to 147.9 (28.3) bpm versus the preablation 190.2 (31.4) bpm, $p < 0.001$. The atrioventricular node effective refractory period (ERP) prolonged significantly at the follow up to 347.9 (80.6) msec versus the peablation 251.3 (39.7) msec, $p < 0.001$. Slow pathway ERP prolonged significantly at the follow up to 406.3 (123.4) msec in comparison with preablation 321.8 (53.8) msec, $p = 0.001$. Fast pathway ERP prolonged at the follow up to 481.2 (132.9) msec in comparison with preablation 408.0 (60.4) msec, $p = 0.03$. The prolongation of the slow pathway ERP was more significant than the fast pathway, $p < 0.001$.
Conclusion: Anterograde conduction, ERP of atrioventricular node, slow and fast pathway changed significantly after long term follow up. RFA of slow pathway is effective and safe method of treatment AVNRT with preservation of dual atrioventricular nodal physiology.

P-21

Radiofrequency Ablation and Criyoablation in Patients with Multiple Arrhythmic Focus

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Introduction: Multiple arrhythmic focus (MAF) are frequent in children with arrhythmia. In this report, we aimed to present our experience about impact of ablation in MAF.
Methods: After detection of multiple arrhythmic focus (MAF) with electrophysiological studies, radiofrequency ablation (RFA) and cryoablation applied to 18 patients were enrolled in our department between October 2009 and December 2011. Twenty-eight ablation procedure was performed in 18 patients.
Results: During this period, the ablation procedure was made in 210 patients and 18 (9%) had MAF. Atrioventricular nodal tachycardia (AVRT) was diagnosis in 69 patients (54 with WPW, 15 concealed accessory pathway) and 15 (22%) had multiple accessory pathway (MAP). Twelve patients were male, mean age was 12.2 years (4-18 years), and mean weight was 44.8 kg (15-70 kg). Follow-up period was 1 month-2 years. Thirteen patients diagnosed with Wolf Parkinson White syndrome (WPW), 1 patient had WPW plus atrioventricular nodal reenteran tachycardia (AVNRT), 1 patient had concealed accessory pathway (AVRT), 2 patient had atrial tachycardia, and 1 patient had AVRT plus AVNRT. One patient had 3, others had 2 focus. Recurrence was observed in one patients (WPW) after 24 hours. Transient complications developed in 3 patients (right bundle branch block, ST depression and atrial fibrillation). Procedure time was 147.5 ± 67.5 minutes, fluoroscopy time was 54.6 ± 36.6 minutes. RFA and criyoablasyon was performed in 10 patients at the same session, criyoablasyon plus RFA was successful in 9 patients. In 1 patient RFA failed but criyoablasyon was successful. RFA alone was applied in 8 patients; in 5 patients complete, and in 3 patients partial success was achieved. Complete success was % 89, partial success was 11%, recurrence was 4%, and transient complications was 11%.
Conclusions: The rate of MAP in WPW has been reported 3.1-13.8%. Success rate 81-95%, and recurrence rate 2.1-11% has been reported after RFA. Multiple accesory pathway and MAF rates are higher in children with arrhythmia. Their main success and recurrence rates are similar to patients with single-arrhythmic focus. In patients when RFA can not be applied because the focus is close to the AV node, criyoablasyon can be applied in the same session succesfully.

P-22

Ablation of right-sided accessory pathways in children and patients with congenital heart disease using a new 3D Mapping system (NavX- Velocity®)

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Introduction: Right sided accessory pathways (AP), are readily approached for catheter ablation by venous access but still have the lowest acute success rates, the highest incidences of recurrence and often require long fluoroscopy times. We describe our experience using a new 3D mapping system (NavX-Velocity®) in children and patients with congenital heart disease (CHD) for mapping and ablation of right-sided APs.
Methods and Results: Seven patients (age 10–41, mean age 17,6 years, 3 males) with right-sided accessory pathways (AP) underwent catheter ablation. One patient had structural heart disease (Ebstein's anomaly). Atrioventricular reentrant tachycardia was induced in 3 patients. The anatomy of the tricuspid ring and the coronary sinus ostium was reconstructed and mapping of the AP performed during sinus rhythm using the NavX Velocity® system. The AP was located at the lateral right free wall ($n = 1$), anterior right free wall ($n = 3$) or in the right posteroseptal area ($n = 3$). Acutely successful catheter ablation was performed using a long sheath (Agilis NxT Steerable Introducer) in all patients (irrigated tip catheter $n = 6$, solid tip $n = 1$). Mean fluoroscopy time was 17,8 min (10–33 min) and mean procedure duration was 145.8 min (69 to 259 min). No procedural complication occurred. Mean follow-up time was 6 months (2 – 10 months). During this time a recurrence of the accessory pathway occured in one patient (1/8 pts, 12,5%) with a right anterior free wall AP, who underwent successful reablation.
Conclusions: In pediatric and CHD patients with right-sided APs, the use of the NavX-Velocitiy 3D mapping system for catheter ablation improves acute ablation success and can help to reduce fluoroscopy time. Furthermore nonfluroscopic guidance (NavX) seems to reduce the recurrence rate compared to a conventional approach.

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P-23

Multidisciplinary approach in patients with supraventricular arrhythmias late after Fontan surgery: Transcatheter and surgical ablation.

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Introduction: Classical atriopulmonary Fontan connections tend to fail in the long term due to progressive anastomotic site obstruction, right atrial enlargement, and refractory atrial arrhythmias. Timing to conversion to total cavopulmonary connection and arrhythmias management are controversial issues.
Methods: We reviewed data from 22 Fontan patients (pts) referred to our centre for refractory supraventricular arrhythmias. All pts underwent complete clinical and instrumental evaluation. After multidisciplinary discussion pts were referred to cavopulmonary connection with concomitant arrhythmia surgery when hemodynamic indication to surgery was present or when they were not eligible for transcatheter (TC) ablation. All others pts underwent TC ablation.
Results: TC ablation was performed in 8/22 pts. Multiple arrhythmias were present in 5/8 pts. The most common form of arrhythmia was intra-atrial reentrant tachycardia (IART); focal atrial arrhythmia was present in 2 pts and atrioventricular nodal reentry in 2 pts (in one case using a twin atrio-ventricular node). TC ablation was acutely effective in all patients. At a median follow-up of 30 months (8-45) two pts experienced arrhythmia recurrence. After clinical revaluation one was referred to a second TC ablation, the other patient underwent surgery for the occurrence of new hemodynamic indication. Conversion to total cavopulmonary connection and surgical ablation was performed in 14/22 pts. Atrial fibrillation was present in 8 pts; IART in 5 pts and atrial flutter in 1 pt. Maze ablation was performed in the right atrium or in both atria according to the arrhythmia diagnosis obtained by EKG or, when needed, by electrophysiological study. There were two patients who died in the early post-operative period. At the mid-term follow-up, late atrial arrhythmias had recurred in 2/14 pts.
Conclusions: TC ablation is effective in Fontan patients not having hemodynamic indication to surgery. In the presence of a failing Fontan surgical ablation during conversion to cavopulmonary connection reduces the incidence of atrial arrhythmias. A multidisciplinary evaluation needs to be performed to identify patients who might benefit from TC ablation and in order to optimize the timing for surgical approach.

P-24

Management of neonates with congenital complete heart block – a tertiary paediatric cardiothoracic unit experience

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Background: Reported risk factors for poor outcome in neonates with congenital complete heart block (CCHB) include prematurity, low birthweight, hydrops, low ventricular rates, and structural heart disease.
Objective: By reflecting on experience in managing neonates with CCHB admitted to a tertiary paediatric cardiothoracic intensive care unit, to identify prognostic factors for outcome.
Methods: Retrospective study (January 2003–May 2011). Data harvested from clinical records and electronic databases.
Results: Sixteen neonates included (11 male (69%)). Median (range) gestational age was 35 (30–40) weeks; 69% were preterm (<37 weeks). Mean (range) birthweight was 2.5 (1.5–3.2) kg. CCHB was diagnosed antenatally in 13 (81%), with median (range) ventricular rate in utero of 58 (40–85) bpm. In 9 cases, maternal anti-Ro antibodies were present: 6 received antenatal steroids, 2 of whom were antenatally hydropic. Seven (44%) had underlying structural heart disease. Ventricular function (via 2D Echocardiography) on admission was good in 11 (69%), moderately impaired in 4 (25%), and very poor in 1 patient.
Nine (56%) patients were admitted within 24 hours (75% within 72 hours) of birth. Prior to transfer, one patient required temporary pacing; 7 (44%) required isoprenaline. Four (25%) patients required emergency cardiac surgery (all non-bypass). All 16 patients ultimately required permanent pacemaker (PPM) implantation; 6 (31%) were temporarily paced beforehand.
Two early deaths occurred: one arrested during PPM implantation, the second, 3 days post-PPM implantation with progressive LV dysfunction and multiorgan failure. Two patients died later (aged 4–5 months): one with rhinovirus infection and ventricular decompensation, the other, following progressive ventricular dysfunction. All four deaths occurred in children born at ≤33 weeks gestation with birthweight <2 kg.Structural heart disease coexisted in 3 who died (all 3 had good ventricular function on admission).
Conclusions: In this cohort, prematurity and low birthweight in association with CCHB were significant risk factors for mortality; underlying structural heart disease, hydrops, and antenatal low ventricular rates were not. Good ventricular function at birth did not predict favourable outcome, especially when structural heart disease coexisted. Early and late mortality remains high in CCHB, making close surveillance essential following PPM implantation.

P-25

Arrhythmia characterization in catecholaminergic polymorphic ventricular tachycardia and CPVT-related phenotypes in children – a single centre experience

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Introduction: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare, inherited disorder associated with emotion/exercise-related syncope and sudden cardiac death. It is characterized by adrenergically induced ventricular and supraventricular arrhythmias in structurally normal hearts.
Objective: Electrocardiographic characteristics of resting ECG and exercise- or isoproterenol-induced arrhythmias were investigated in our patients with CPVT and CPVT-related phenotypes. Demographic data and clinical presentation were also summarized in association with ECG findings.
Method: Patients were diagnosed with CPVT or CPVT-related phenotypes according to exercise-induced or isoproterenol-provoked polymorphic or bidirectional ventricular tachycardia (VT) in 2004-2011 at our centre.
Results: 9 patients (5/4 male/female) were identified at the age of 2-15 years (median: 11.2). Syncope and sudden cardiac arrest (SCA) and other manifestations, such arrhythmic heart sounds or atypical spells, as presentation showed equal distribution among our patients. Baseline ECG showed sinus bradycardia and U waves in most cases, but ventricular ectopy appeared, also spontaneously in 2 cases. Mean QTc was 422.8ms (380-480ms; STD: 36.8ms). VT was provoked by exercise in 4 patients and by isoproterenol in 3 (small or disabled) patients, respectively. In 2 cases with Andersen-Tawil syndrome non-sustained VT occurred spontaneously, but not on exercise tests. Ventricular ectopy started to arise at a mean heart rate of 116.8/min (STD: 11.6/min).

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Non-sustained VT was bidirectional in 55% and polymorphic in 45%. The initiating beat of VT was late-coupled (mean coupling interval: 446ms, STD: 87ms) and wide (mean QRS duration: 126ms, STD: 14ms), and QRS morphology suggested an outflow tract origin in 66%. Multiple episodes of VT in 1 patient were not common, so reproducibility of VT morphology could not be investigated. The more serious presentation of SCA was not associated with a distinct type of VT or initiating ventricular beat, or with earlier age of presentation. Supraventricular arrhythmia was detected only in 1 patient during the median follow up of 3.9 years (IQR:1.3-5.6).
Conclusion: In our patient population with CPVT ventricular tachycardia was often polymorphic, late-coupled, and initiated from the outflow tract.
Resting ECG was usually unremarkable with sinus bradycardia and prominent U waves.
Appearance of supraventricular arrhythmia was not common in childhood.

P-26
Lead extraction – growing problem in children with endocardial pacing
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Introduction: Removal of endocardial pacemaker or implantable cardioverter-defibrillator (ICD) leads in children is difficult clinical problem. In young patient lead failure is more often observed then in adults, non functional endocardial lead may increase risk of venous obstruction or infection and makes new lead implantation more difficult. The indication for lead extraction in pediatric population has to be individualized. The purpose of this study was to sum up our experience in removal of endocardial leads.
Material: From January 2007 to December 2011 we extracted 25 leads (16 right ventricular, 5 atrial and 4 ICD leads) in 21 children age from 5 to 18 (mean 13,3) years. The implant duration ranged from 0,1 to 11,4 (mean 4,9) years. The lead failure was the main reason for extraction -19 leads (76%), in 4 (16%) children, with long lasting permanent pacing, ventricular leads were dangerous tight because of the patients' growth. In 2 cases with ICD leads were removed because of endocarditis or sepsis. In all children the lead extraction procedure was performed under general anesthesia by the same experienced team (cardiologists and one cardiac surgeon), the extracorporeal oxygenation and circulation system was available. In all children the Cook Medical system was used during the procedure.
Results: All the procedures were successfully completed. 19 leads (76%) were totally removed. In 6 children we had clinical success (a short fragments of 2 atrial lead and 4 ventricular leads were abandoned). In 1 patient subclavian venous bleeding occurred as a main our complication. None required cardiac operation. In 18 children we implanted a new endocardial pacing system during the same procedure.
Conclusion: In children the main indication to remove an endocardial lead is its failure. Extraction of en endocardial lead in children is difficult but possible even after long pacing duration, but experienced team and proper facilities are necessary.

P-27
Effects of Umbilical Venous Catheters on Arrhythmia and Heart Rate Variability in Premature Newborns
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Introduction: Cardiac rhythm abnormalities associated with umbilical venous catheters in newborns are limited to anecdotal case reports. The present study intended to evaluate true incidence and association, if there is any, between umbilical venous catheter, arrhythmic potential, and changes in heart rate variability.
*Methods:*The study consisted of two groups; 26 preterm newborns with a umbilical venous catheter (group 1), and 26 control group without (group 2). In all babies the following parameters were recorded prospectively: gender, gestation at birth, birth weight, working diagnosis for admission, medications, complete blood count, serum electrolytes, calcium, glucose, liver and kidney function tests, and clinical assessment scores (SNAP-II and SNAPPE- II). Holter recordings were fitted on day 2 of life in all newborns. The heart rate variability study was performed over a 24-h period by utilizing time-domain and frequency-domain analyses.
Results: There was no statistical difference between the two groups for gestational age, birth weight, SNAP-II and SNAPPE-II scores, hemoglobin, mean calcium, potassium, and sodium levels.In spite of any type of arrhythmia ratio appeared to be higher in infants with catheter (65%) than the control subjects (42%), this difference did not reach statistical significance ($p = 0.095$). Life-threatening cardiac arrhythmias were not found in any patients on Holter recordings. None of the heart rate variability parameters were found to be statistically different between the two groups.
*Conclusions:*Our study reassuringly demonstrated that umbilical venous catheter does not have any significant effect on arrhythmia or heart rate variability in preterm newborns.

P-28
An insight into the autonomic mechanisms underlining the reflex syncope in children and adolescents
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Introduction: It has been estimated that around 15% of children and adolescents experience, at least, one episode of syncope until adulthood. Excluding a minority of cases that are due to a potential cardiac disease, the majority of syncopal episodes are of reflex origin and benign in nature. In this situation, a tilt-test (HUT) is performed to reproduce the clinical symptoms, allowing the evaluation of the progressive cardiovascular adaptations to orthostatism. However, the mechanisms underlining a HUT-induced syncopal episode, in these patients, are not yet well defined.
Aim: To investigate the modifications of autonomic activity that occur during a diagnostic HUT for reflex syncope in young patients.

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Methods: Seventy-three pts (12 ± 3.5 years, 66% females) with unexplained syncope were enrolled in this study. HUT was performed following a standard protocol. A positive response was defined as a sudden development of syncope or presyncope associated with hypotension, bradycardia, or both. Beat-to-beat RR-intervals (RRI) and arterial pressure (BP) were continuously recorded and data were used to heart rate variability (HRV) analysis.
Results: Thirteen pts (17%, 11 females) experienced syncope after 18 ± 10.2 min of tilting. According to VASIS classification responses included mixed (53%), vasodepressor (30%) and cardioinhibitory (23%) types. Systolic BP (SBP) was significantly different between tilt-positive and tilt-negative pts at 10 min (SBP 115 ± 10 vs. 126 ± 12 mmHg, P = 0.02), 15 min (SBP 112 ± 10 vs. 120 ± 10 mmHg, P = 0.04) and 20 min after tilting-up (SBP vs. 98 ± 12 vs. 120 ± 10 mmHg, P = 0.001). No differences on HR were found between groups. Significant differences were noted in HRV parameters before syncope. In tilt-positive pts, LF changed dynamically during HUT, showing a sudden and initial rise of sympathetic tone (LF 8.5 ± 6.27 vs. 19 ± 15.3 mmHg2, P = 0.037) followed by a second overshoot of activity (LF 7.7 ± 5 vs. 15 ± 8.2 mmHg2, P = 0.041), continued by a steady fall-off 1 to 2 min prior to syncope. This pattern occurred in every fainter and was absent in all non-fainters.
Conclusions: This study shows a strong effort by the autonomic nervous system to adapt to orthostatic stress through modifications of sympathetic tone of different magnitudes, before the syncopal event, in tilt-positive patients. These changes suggest an exhaustion of the sympathetic reserve not enabling a time-progressive adaptation leading the individual to faint.

P-29
Heart Rate Variability Alterations in Children with Euthyroid Hashimoto Thyroiditis

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Introduction: Hashimoto thyroiditis (chronic autoimmune thyroiditis) is the most common form of thyroiditis in childhood. Previous studies have found autonomic dysfunction of varying magnitude in patients with autoimmune diseases, which in turn is a considered a cardiovascular risk factor. The aim of the present study was to evaluate the heart rate variability (HRV), a measure of cardiac autonomic modulation, in children with euthyroid Hashimoto thyroiditis (eHT).
Materials and Methods: The study included 32 patients with eHT (27 girls and 5 boys; mean age 11 ± 4.1 years, range 8-16; body mass index 0.47 ± 0.69 kg/m2), as judged by normal or minimally elevated serum TSH levels (normal range: 0.34-5.6 mIU/l) and normal levels of free thyroid hormones (FT4 and FT3) and 38 euthyroid healthy controls. Subjects with cardiac, metabolic, neurological disease or any other systemic disease that could affect autonomic activity were excluded from the study. Patients with eHT and control subjects underwent a full history, physical examination and 24-h ambulatory ECG monitoring. Time domain parameters of HRV were evaluated for cardiac autonomic functions.
Results: Children with eHT displayed statistically significantly lower values of time domain parameters of SDANN, RMSSD, NN50 counts and pNN50 for each 5-min interval ($p < 0.05$) as compared to healthy controls ($p < 0.05$). Only the SDNN in children with eHT did not significantly differ from those in the controls ($p > 0.05$).
Conclusion: These results demonstrate that eHT is associated with decreased sympathovagal modulation of the heart rate. Hashimoto thyroiditis may increase cardiovascular risks in children even when they are in the euthyroid state.

P-30
Cryoablation for PARAHİSSİAN Accesory pathways

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Introduction: Cryomapping and cryoablation is prefered for parahisian accessory pathways since close ralationship to his bundle. We report 20 patients with Wolff Parkinson White syndrome who underwent cryoablation of the parahissian accesory pathways .
Methods and Results: From September 2010–December 2011, 20 patients between 8-31 ages underwent cryoablation. Body weigths were above 25 kg.All of the patients' accesory pathway was determined during electrophisiologic study (EPS) as parahissian. Cryoablation performed in -80° degree for 4-5 minutes on avarge 4 episodes. Radiofrequency (RF) catheter ablation used in six patients who had multiple accessory pathways. Cryoablation performed on places where tachycardia stoped during cryomapping, preecitation dissappeared by catheter pressure or cryomapping. In all patients except one acute success achieved and determined by unable to provocate tachycardia and disapperance of preexitation. In one patient temprorary complete AV block obsereved during 3rd minute of cryoablation, two patient developed right bundle branch block. Avarge procedure time 144 minutes, flouroscopy time was 36 minutes. After avarage seven mounths follow up, jthere were two recurrences. In one of these patients transeosofageal EPS performed but, tachycardia couldn't be induced.
Conclusion: Cryoablation can be used safely in pediatric patients whose accessory pathway close to the conduction system. Complication develops rarely compared to the RFA. Complete AV block can be observed but it is temporary. Although recurrency is reported higher compared to RFA, the present study represents an acceptable recurrence rate.
Key words: cryoablation, Wolff Parkinson White, parahissian.

P-31
Acute Effectiveness of RF Ablation for Idiopatic Ventricular Arrhythmias in Children

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The idiopathic ventricular arrhythmia (IVA) in children is a rare disease with benign clinical outcome but there is still some risk of congestive heart failure, syncope or sudden death. The RF ablation has been widely used for supraventricular tachycardia treatment in children, but limited data exist on this therapy for idiopathic ventricular tachycardia in children.
Among 505 children ablation procedures 30 were done for idiopathic ventricular arrhythmias in our Institute. Children were qualified for ablation if they have symptomatic ventricular arrhythmia or the number of ventricular beats exceed more than 30% of total QRS complexes in Holter ECG. Based on the clinical charts the early effectiveness data were collected. We analyzed the recurrence of arrhythmia within 24 hours after procedure in Holter monitoring.

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There were 30 pts (15 girls and 15 boys) with IVA. The mean age at the procedure was 14 years 2 months (ranged from 21 months to 18 years). All children had normal heart anatomy. Fourteen were symptomatic (palpitations, syncope and aborted sudden death-1 pts), 3 pts with RV and 11 with LV arrhythmia. Thirteen children had runs of ventricular tachycardia with mean heart rate 160/' (ranged 130-195/'), 17 pts had ventricular extrasystole with the number of ventricular beats from 30-70% (mean 40%) of rhythm in Holter monitoring. The procedures were done with CARTO system, under general anesthesia in 8 pts, in local in 22pts. The origin was: RVOT 10 pts, RV 3 pts, LVOT 3 pts, LV 8 pts and fascicular tachycardia 6 pts. At the end of procedure ablation was successful in 24 (80%) children, failed in 6 pts (20%). In Holter ECG done after 24 hours 17 pts (57%) were still free of arrhythmia, 5 (17%) had less than 5% of arrhythmia. All ablation done for fascicular and LVOT tachycardia were successful. In 8 (26%) children IVA was still present: RVOT-3pts, RV-1pts and LV -4pts. There were no complications.
In 74% of our children with IVA RF ablation was highly effective in early observation, although the recurrence rate within 24 hour after procedure was 6%. RF ablation for idiopathic ventricular arrhythmia is safe procedure.

P-32
Long-term single centre study on epicardial pacing leads in children and patients with congenital heart disease

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Introduction: Data on permanent epicardial pacing in young patients is limited with respect to long-term lead-survival, types of pacing leads and study size. We report on a 17-year single centre experience with various epicardial pacing leads in 215 patients.
Methods: We retrospectively reviewed all epicardial leads implanted in our centre between 1993 and 2010. A total of 215 patients (median age at implantation 5.2 years, range 1 day to 28 years) underwent 371 epicardial lead implantations (105 atrial and 266 ventricular leads). Various leads (157 [42%] screw-in, 214 [58%] suture-on leads) were used. The number of former cardiac surgeries, the number of former lead implantations, the age at implantation and a Fontan-type of palliation were investigated as predictors for lead failure.
Results: During a median follow-up of 3.9 years (range 1 day to 15.6 years), lead failure was documented in 61 leads (16%) with exit block/elevated pacing thresholds being the most common cause (n = 42). The only predictor for lead failure was ventricular lead position (HR 2.2, CI 1.1–4.2, p = 0.02). The overall 1-, 5-, 10- and 15-year lead survival was 98%, 84%, 73% and 49%, respectively. There was no difference in lead survival of screw-in and suture-on leads. There were no deaths related to lead dysfunction.
Conclusions: In our study of 371 epicardial leads, lead survival was good, with no difference in screw-in versus suture-on leads.

P-33
Postoperative junctional ectopic tachycardia in paediatric patients

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Junctional ectopic tachycardia (JET) is a serious postoperative complication with a mortality rate up to 14% in patients after heart surgery. In this study we evaluated the incidence, risk factors and outcome of junctional tachycardia.
Methods: We analysed retrospectively the data from 1345 patients who underwent open cardiac surgery in our institution from 2006 to 2010.
Results: The incidence of junctional ectopic tachycardia was 2,5%. The patients with JET were significantly younger (median 0,25 vs 1,04 years; p = 0,001). They had also a longer cardiopulmonary bypass time (median 117,5 vs 91 minutes; p = 0,001). Complete atrioventricular block (CAVB) after surgery was diagnosed in 33 patients, 19 of them developed postoperatively also a junctional ectopic tachycardia. In all of the patients with a combination of CAVB and JET recovered the AV conduction in the postoperative course spontaneously.
Conclusions: The incidence of junctional ectopic tachycardia was in our cohort low (2,5%). The patients with JET were younger and had a longer cardiopulmonary bypass time.

P-34
Catecholaminergic Polymorphic Ventricular Tachycardia and Left Ventricular Noncompaction Cardiomyopathy: a case report and review of the literature.

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Introduction: The case of a 14 year old girl affected by catecholaminergic polymorphic ventricular tachycardia (CPVT), sick sinus syndrome, left ventricular noncompaction cardiomyopathy (LVNC) and slight cognitive impairment will now be described.
Case report: In 2006, a 9 year old girl experienced exertional syncope. Investigations revealed polymorphic ventricular tachycardia on exertion, sinus disfunction with pauses up to 3.59 sec and atrial tachycardia. Cardiac RM performed in 2006 was normal. A dual chamber epicardic pace-maker was implanted and anti-arrhythmic prophylaxis with propranolol and sotalol was started with little benefit.
In 2010, the patient was revalued in a reference center, and therapy was switched to nadolol and flecainide with good antiarrhythmic control. Genetic testing revealed a "de novo" hRyR2 gene mutation.
In 2011, she experienced monomorphic ventricular tachycardia without hemodynamic impairment. ECG revealed negative T waves in lateral precordial leads and echocardiogram showed a noncompaction-like morphology of the left ventricle with normal size and function. Plasmatic levels of flecainide were on therapeutic range. Dual chamber ICD with monocoil, active-fixation ventricular lead and 3830 active-fixation atrial lead (Medtronic, Inc.) was implanted. Flecainide was increased with better control of exertional atrial arrhythmias including atrial fibrillation. The evolution of cardiomyopathy will be monitored by close echocardiographic follow-up.
Discussion: CPVT is a genetically determined disorder associated with syncope and sudden death that manifests predominantly in children and teenagers with a structurally normal heart. RyR2

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mutations are the first reported gene alterations associated with the autosomal dominant form of CPVT. Whether or not ryanodine receptor mutation may induce structural abnormalities has been under debate since the first description of the ryanodine receptor involvement. Genetic mutations that give rise to CPVT phenocopies have been reported in the context of other arrhythmia syndromes with structural heart disease, such as arrhythmogenic right ventricular dysplasia and dilated cardiomyopathy. We describe a CPVT phenotype associated with sick sinus syndrome, LVNC and slight cognitive impairment, related to a RyR2 mutation. In our knowledge, no association between CPVT, sinus disfunction and LVNC has previously been reported. Further follow-up is required in order to assess evolution of the cardiomyopathy and arrhythmic profile.

P-35

Possible Involvement of the Natural Immune Systems in the Pathogenesis of Kawasaki Disease

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Introduction: Kawasaki disease (KD) is a pediatric vasculitis, which sometimes involves aneurysm as a sequel of coronary arteritis. Although its etiology remains to be elucidated, several preceding studies have indicated that altered immune responses could be involved in the pathogenesis of the disease. By employing an animal model for KD, we aimed at shedding light on the pathological mechanisms underlying the disease, from the viewpoint of natural immunity.
Materials, Method and Results: To induce KD, DBA/2 mice were administered Candida albicans water-soluble fraction (CAWS). Biochemical and immunohistochemical analyses revealed (1) increased levels of self-reactive immunoglobulin M (IgM)-type antibodies, (2) extravascular deposition of IgM around the aorta, (3) decreased levels of mannose binding protein-C (MBL-C) in plasma, and (4) deposition of MBL-C in the aortic intima, all suggesting that the natural immune system could be involved in early-stage CAWS-induced vasculitis. IgM and MBL-C have been known to activate the complement classical and lectin pathways, respectively. Interestingly, preliminary results showed that inhibition of the lectin pathway partially suppresses CAWS-induced vasculitis.
Conclusion: Taken together, these findings indicate that the pathogenesis of KD could be associated with dysregulation of natural immune systems, including the lectin pathway. This and other immune pathways may be possible therapeutic targets for KD.

P-36

Infant with tacrolimus induced hypertrophy after heart transplantation – increased expression of CRTC-1, but not CRTC-2 and -3.

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Introduction: The role of calcineurin (CN) in the development of cardiac hypertrophy is controversial. In humans CN expression is increased in hypertrophied non-failing hearts, while tacrolimus induced hypertrophy (TIH) can occur in humans after solid organ transplantation and immune-suppression with CN antagonists. Lately, the CREB-regulated transcriptional co-factor (CRTC), was described the heart. It is closely tied to the cAMP-responsive element-binding protein (CREB) which is activated by CN.

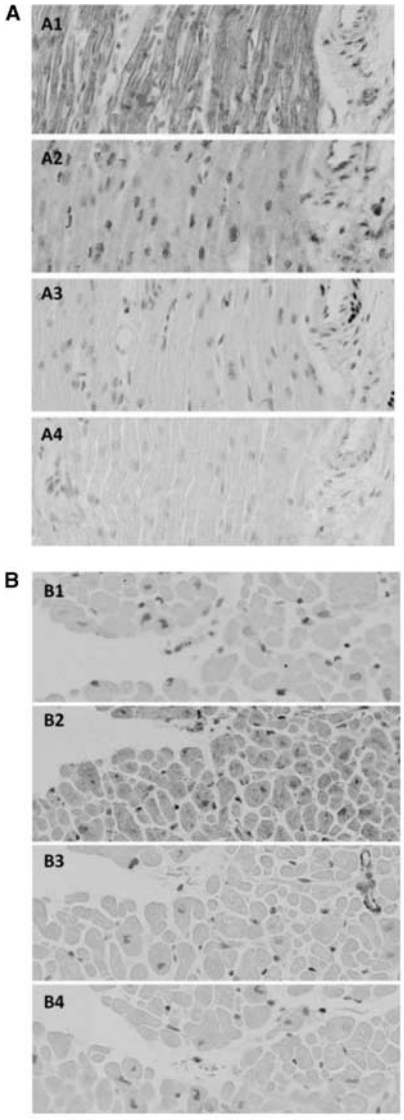

Figure 2: A: EMBs at d 5, B: EMBs at 8 Weeks post HTX. 1-3: CRTC 1-3, 4: control.

We hypothesized that the CN antagonist tacrolimus influences the expression of CRTC in TIH.
Case Report and methods: A now 20 months old male infant underwent heart transplantation (HTX) due to dilatative cardiomyopathy at the age of 10 months. Immunosupression consisted of anti-IL-2 antibody, methylprednisone, mycophenolatemofetil and tacrolimus (TAC). Initial target TAC through levels (TL) were 13-15 [micro]g/l. Endomyokardial biopsies (EMB) were obtained after 5 days, no rejection was detected (ISHLT 0R). Due to high metabolic turnover, stable tacrolimus TL of 13[micro]g/l were reached only with high doses of TAC (0,5 mg/kg/d).
Marked hypertrophy occurred after 4 weeks. Steroids were tapered and ended after 12 weeks, hypertrophy persisted. EMBs

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were obtained again, histology revealed diffuse hypertrophy, no signs of rejection (ISHLT 0R), establishing the diagnosis of TIH. TAC TL were adjusted to 6 [micro]g/l with tacrolimus 0,06 mg/kg/d. Cardiac hypertrophy decreased remarkably and function improved. EMBs before and after onset of TIH were subjected to CRTC immunohistochemistry with CRTC antibodies [Figure 2].
Results: In non-hypertrophied samples at d 5 after HTX, CRTC-1 and -2 are found in the cytosol of most cardiomyocytes (Fig. 2 A1 and A2). For CRTC-1 some, and CRTC-2 most nuclei are stained. In the TIH samples, only CRTC-2 is found (Fig. 2 B2). CRTC-3 is unchanged and only found in vascular endothelial cells (Fig. 2 A3, B3).
Conclusion: TIH occurred due to initially high doses of TAC within a few weeks after HTX and resolved upon reduction of TAC. Immunohistochemistry revealed decreased CRTC-1 expression in TIH samples. CRTC-2 expression decreases slightly with TIH development. The change in CRTC-1 and -2 expressions are a hint that the CN dependent CREB/ CRTC pathway is involved in the development of TIH.

P-37
Creation of a single-center DNA-bank of congenital heart disease with focus on cardiac phenotype

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Background: The creation of DNA-banks is the necessary basic requirement for developing genetic research in Pediatric Cardiology. Precise cardiac phenotyping is mandatory in order to exploit the samples. The main future objective of such programs will be to test different candidate genes derived from animal and human research in different groups of CHD.
Methods: We started a single center DNA-bank called CARREG (CARdiacREGulation genes) of patients' blood samples with congenital heart disease. This DNA-bank is not restricted to a specific research project. Initially this program was limited to CHD of the outflow tract (OFT) and then extended to all types of CHD after informed consent of patient and legal representative. Samples are also taken from consenting parents of an affected patient. Congenital heart disease of each participant is fully phenotyped according to clinical exam, echocardiography, imaging and surgery protocol. Each patient is grouped in one of the 10 major groups according to the recently published "Anatomic and clinical classification of congenital heart defect" classification (ACC-CHD). Once grouped, each CHD is described precisely according to the International Pediatric Cardiac Congenital Code (IPCCC) with the number of necessary codes.
Results: Starting from April 2009 1243 blood samples have been collected and stored in a dedicated space of the institution's DNA-bank. 698 are patients' blood samples. 545 blood samples come from parents or relatives. According to ACC-CHD there are 7 samples in group 1, 6 samples in group 2, 13 samples in group 3, 16 samples in group 4, 18 samples in group 5, 30 samples in group 6, 14 samples in group 7, 551 samples in group 8, 39 samples in group 9, 4 samples in group 10.
Practical outcomes: In order to investigate genetic foundations of CHD, DNA-banks have to contain a representative number of all possible CHD. The initial restriction to patients with OFT disease explains the present predominance of group 8 in our gene bank. To make international comparison and data exchange easier, cardiac phenotyping in CARREG relies on IPCCC and the newly developed ACC-CHD. In the future this program will be extended to a national level.

P-38
Predicted upper limits for left ventricular internal diameter at end diastole in children and adolescent athletes

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Objectives: The quantitative of LV cavity enlargement may be significant in the differentiation between athlete's heart and dilated cardiomyopathy.
The aim of the study was to assess the possibility of predicting cutoff values for left ventricular internal diameter at end diastole in athletic children and adolescents.
Methods: The study procedure was approved by regional bioethics committee. Subjects were one hundred sixty seven male athletes aged 14.8, (SD 1.6, range 12-17 years) participating in basketball, rowing and cycling and 168 healthy sedentary controls matched for age, sex and body surface area (BSA). Athletes had competed for a mean of 3.2 (SD 2.2) years with training volume 8.1(SD 2.3) hours/week. Standard two-dimensional guided M-mode and Doppler echocardiography were employed to evaluate left ventricular morphology and function. Predicted limits of LVIDd were calculated based on the mean LVIDd values +2 SD for the control subjects for every year of age.
Results: Compared with controls, athletes had a larger LV cavity (LVIDd) (50.16 (3.97) v 47.59 (2.89) mm), a difference of 5.4%. LVIDd exceeded predicted limits in 47.9% of athletes; there were 55.6% of cyclists and 51.6% of basketball players. LVIDd was > 54 mm in 9.6% athletes. 3.2% of athletes had an LVIDd 60 mm commensurate with a diagnosis of dilated cardiomyopathy. None of athletes had an LVIDd >60 mm.Systolic and diastolic function were within normal limits for all athletes.
Independent factors determining the value of LVIDd exceeding the predicted upper limits were older age (16–17 years old), higher body weight (OR = 1.08; CI = 1.03–1.13, $P < 0.001$) and cycling sporting discipline ($P < 0.01$).
Conclusion: Almost half (47.9%) of athletes left ventricular internal diameter at end diastole exceeded the predicted limits, but minority (3.2%) of athletes LVIDd was 60 mm.Upper normal limits of LVIDd was 60 mm.Values in excess of these should prompt further investigation to identify the underlying mechanism. Age, body weight and cycling sporting discipline determined LVIDd exceeding the predicted upper limits.

P-39
Experimental Study on Etiological Participation of Oxidative Stress in Vasculitis by Kawasaki Disease

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Background: Because it is recently showed that oxidative stress and inflammation amplifying each other have been implicated in the onset and progress of atherosclerotic lesion, it has been hypothesized that oxidative stress might also be related to the

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acute vascular disorders associated with Kawasaki disesase (KD). Therefore, we have clinically investigated the dynamics of oxidative stress in the acute KD, and have reported that oxidative stress plays a very important role in the KD vasculitis. However, the etiological relationship between oxidative stress and KD vasculitis has not yet been clarified. In this study, we examined the dynamics of oxidative stress in KD model mice to reveal the etiological participation of oxidative stress in KD.
Methods: Candida albicans water-soluble fraction (CAWS), which induces vasculitis such as that seen in KD, was intraperitoneally injected to DBA/2 mice for five consecutive days. At 1, 7, and 14 days after CAWS injection, mice were sacrificed. Autopsy was performed to obtain serum, hearts and spleens. We assayed reactive oxygen metabolites (ROM) as the oxidation marker, the biological antioxidant potential (BAP) as antioxidant potency, and sulfhydryl (SH) groups which are based on the capacity of thiol groups have antioxidant effect in the serum of mice. Hearts were fixed and prepared in paraffin blocks. Tissue sections were stained with H&E stain. We defined the onset of vasculitis as the presence of inflammatory cell infiltration.
Results: The ROM level significantly increased on day 1 after CAWS administration and subsequently decreased. In contrast, The BAP gradually increased until day 7 but the SH groups decreased until day 7 and recovered on day 14. The Heart weight did not change. The spleen weight significantly increased on day 1 and gradually decreased. Inflammatory infiltration was observed in the adventitia of the aorta and proximal coronary arteries from day 7.
Conclusions: In the KD model mice, oxidative stress increased before the onset of vasculitis. Therefore, oxidative stress may be related to the pathogenesis of vasculitis in KD. In contrast, the antioxidant potency increase with activation of oxidative stress, and a lot of thiol groups may be used in acute KD.

P-40
Simulator training improves beginner's skills in performing echocardiography in congenital heart disease.
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Objectives: Although simulator based training is increasingly used in medical training, there is paucity of studies proving its effectiveness. We have studied the effectiveness of our previously described simulator EchoCom for training echocardiography in congenital heart disease (CHD).
Methods: The simulator consists of a life-sized neonatal manikin, an electromagnetic tracking system and a computer application. The application is linked with a data base of 3-dimensional echocardiographic data sets of CHD. For the present study we have chosen a pre-/post-test design with nine pairs of data-sets. The first set was presented to 10 beginners in echocardiography without clinical information. Participants were asked to scan the data and come up with a presumed diagnosis. All participants underwent a structured simulator based echocardiography training session by an expert afterwards. After training the study was repeated using different data sets of identical lesions. Pre- and post-training results were compared to evaluate the effectiveness of simulator based training.
Results: Post-training results were significantly better than pre-training results with number of correct diagnoses improving from a median of 3.1 to 8.3.
Conclusion: Simulator based training improves beginner's skills in echocardiography in CHD. We have not compared simulator based training with traditional learning like bed-side teaching or lectures. Bed-side teaching however is often impractical for hands-on training in critical lesions.

P-41
Developmental abnormalities of the fourth primordial pharyngeal arch
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Introduction: Anatomical variants of the aortic arch and its supraaortic branches occur repeatedly. They might be incidentally discovered on imaging or present with respiratory or digestive symptoms when forming a vascular ring and compressing the trachea and esophagus. Moreover, these vascular abnormalities could produce challenges during conventional angiography, endovascular and surgical procedures, especially if such anatomic variations are previously not well-known. Our aim was therefore to undertake a study, analyzing the structural variations of the aortic arch and its supraaortic vessels according to their embryological basis with an emphasis in the development of fourth primordial pharyngeal arch.
Methods and Results: A retrospective study of medical records and our surgical database identified various anatomic variants of aortic arch and its supraaortic branches. Imaging modalities include echocardiography, CT, MRI as well as invasive catheterization. In this study, the embryologic development and imaging appearance of the aortic arch system and its various malformations are reviewed. Diverse aortic arch anatomic variants such as left/ right aortic arch, bovine/ equine arch, aberrant/ isolated subclavian artery, interrupted arch and cervical aortic arch are discussed based on their embryological basis.
*Conclusions:*Various congenital malformations of the aortic arch vessels have been reported in the literature, which reflects the complexity of their embryologic development. Most of these malformations are incidental findings and remain asymptomatic until adulthood. However, they can also be responsible for a variety of clinical symptoms and frequently associated with congenital cardiac diseases. Familiarity with the spectrum of the possible aortic arch malformations is essential for their accurate diagnosis, classification and management. This could be simplified by understanding the cardiovascular embryology.

P-42
Exome Sequencing in Syndromic Patients with Congenital Heart Disease performing Trio Analysis
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Introduction: Congenital heart defects (CHD) are a major cause of infant morbidity and mortality. Reaching an etiological diagnosis in patients with a syndromic heart defect is important, not only to gain insight into their pathogenesis and genetic counseling on recurrence risks, but especially with regard to providing information on the future prospective, based on knowledge of the natural course of the disorder.

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In syndromic cases, an exact etiological diagnosis can be reached in an estimated 50-60%, following careful clinical evaluation, complemented by various genetic tests, including array-CGH. With the advent of exome sequencing, it is now feasible to perform a trio analysis, i.e. sequencing of the coding parts of the genes in both parents and the child, where only the child is affected, in order to identify a candidate gene.
For syndromic cases, we hypothesize that these patients have a thus far not recognized monogenic condition, responsible for both the intelligence deficit and the heart defect. Since the vast majority of syndromes featuring CHD are dominant, it is likely that at least in a subset of these, a de novo dominant mutation is present.
Methods: In-solution capture (Nimblegen target enrichment system) and sequencing will be done on the Illumina HiSeq2000 platform (Genomics Core KULeuven/ UZLeuven). Data analysis will be done using commercial and in-house developed software. Afterward, the variants will be annotated by Annovar to allow filtering of all found variants in order to identify potential causal mutations.
Results and Conclusion: Preliminary results on the exome sequencing of five sets of trio's, with the child presenting with congenital heart disease, dysmorphic features and mental retardation, will be discussed.

P-43

A family with Myotonic Dystrophy (DM1) associated with Sudden Death, Long QT and a Brugada-like ECG pattern in different affected relatives.

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Myotonic dystrophy is a multisystem condition inherited in an autosomal dominant manner. In addition to myotonia and muscular dystrophy, affected individuals are also at risk of cardiac dysrhythmias and conduction defects. A 17 year old male died suddenly whilst riding a bicycle having been previously well. Post mortem examination was normal and cause of death was considered to be due to a primary underlying arrhythmia. His parents and 13 year old brother were referred for cardiac screening. His father's resting ECG showed evidence of R waves starting from V1. His echo showed some septal hypokinesia with a normal MRI. Following an ajmaline test, there was asymptomatic sustained ventricular tachycardia. His mother had a normal echo, MRI and ajmaline test but had a corrected QT interval of 450ms associated with hypoparathyroidism which resolved with treatment. Initially, his brother had a normal resting ECG and echo but subsequently was found to have a corrected QT interval of 500ms. At the time of cardiac screening his mother and brother were well with no other medical history. However, his father was concurrently being assessed for dysphagia. On further examination, he had frontal balding, facial muscle weakness and myotonia. On direct questioning there was also a history of myotonia in the deceased. Gene testing identified a DM1 gene expansion from leucocyte DNA in the father and brother and in DNA extracted from pathological blocks from the deceased, confirming Myotonic Dystrophy. Long QT and Brugada gene testing (KCNQ1, KCNH2, KCNE1, KCNE2, SCN5A) in the father and brother was normal. This family highlights the variability of cardiac dysrhythmias associated with Myotonic Dystrophy and that this condition should be considered in families with an otherwise unexplained sudden arrhythmic death.

P-44

Serotonin receptor-related coronary vasomotor function in young miniaturized pigs is additively impaired by streptozotocin-induced diabetes and Chlamydia infection but protected by simvastatin

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Background: Children with diabetes mellitus are at risk of increased recurrence of respiratory infections. Although both infection and diabetes have been suggested to accelerate atherosclerosis already in childhood, to date there is no proof of an eventual interplay of these two in the early development of vascular disease. In addition to this, we investigated whether statin therapy initiated shortly after diabetes induction could coronary vasomotor dysfunction.
Methods: Coronary vasomotor function was evaluated in vivo by intracoronary Doppler velocimetry in 2-month old nondiabetic and diabetic miniaturized pigs in response to intracoronary bolus of serotonin, bradykinin and adenosine. Diabetes was induced by injection with streptozotocin. Animals from each group (diabetes and non-diabetes) were randomly assigned to 3-time inoculation with either saline or Chlamydia pneumoniae (CPn) at 1 –week interval. Another subgroup of diabetic animals received only one inoculation with CPn. Simvastatin therapy was initiated in 4 diabetic & 3-time CPn inoculated animals 1 week after diabetes induction and continued until the end of experiment. The responses to intracoronary agonists were expressed as % change in average peak velocity from baseline.
Results: Intracoronary bolus of bradykinin resulted in similar APV responses in all animals regardless of diabetes and infection status ($p > 0.2$). In contrast, serotonin-induced responses were worst in animals with both diabetes and multiple CPn inoculations ($p = 0.04$ vs controls, and 0.05 vs mono-inoculated animals). The responses in this group were however lessened by simvastatin therapy ($p = 0.1$ vs simvastatin-nontreated animals).
Conclusion: In minipigs, streptozotocin-induced diabetes and infection with CPn appeared to have additive adverse effects on serotonin receptor-related vasomotor function of coronary circulation, while statin therapy seemed to exert some protective effect.

P-45

Zebra fish heart slices: A new model to investigate contractile properties of the heart

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Introduction: Zebra fish are a popular model organism for cardiovascular research. They are easy accessible and allow insights into different development changes. Their genome is almost entirely sequenced and genetical modifications can be easily applied. Action potential morphology and ECG signals are similar to the human heart. However there are no data in regard of the contractile properties of Zebra fish hearts. Our aim was it to develop a model, which allows the functional characterization of contractile properties of zebra fish hearts.
Methods: Hearts of adult Zebra fish were excised, low-melting agarose maintained was poured over the hearts. The agarose-embedded hearts were fixed onto a specimen holder. Ventricles were sectioned into 300-μm-thick slices along the short axis.

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The slices were mounted on an isometric force transducer. Length was increased stepwise to the length of maximal force development. Contractions were recorded from spontaneously beating as well as from electrically stimulated preparations. Preparations were field-stimulated by silver electrodes. Force frequency protocols as well as pharmacological tests (Isoproterenol, Carbachol and Nifedepine) were applied. Electrical stimuli and analog signals from the force transducer were amplified with a bridge amplifier and analog signals were transferred to an analog to digital board.
Results: From every heart 3 to 4 ventricular slices could be generated. More than 50% of the slices were beating spontaneously. All slices developed force of contraction after electrical stimulation. The slices displayed a negative force-frequency-relationship, (less amplitude at higher stimulation frequencies). Hormonal modulation of zebrafish slices by Isoproterenol lead foremost to an increased beating frequency and only to a slight increase in force of contraction. Nifedepine reduced the force of contraction significantly.
Conclusion: Generation of ventricular slices from Zebra fish hearts is possible. We developed successfully a model to characterize the contractile properties of Zebra fish hearts. The differences of these properties compared to human hearts have still to be analyzed.This method could serve as basis to evaluate the role of different genes and their influence on the physiology during cardiac development by using hearts of "morpholino"-Zebra fishes, where almost every gene can be directly knocked down.

P-46
Effects of Hypothermia on RBM3 and CIRP Cold-Shock Proteins Expression in Murine Organotypic Hippocampal Slice Cultures

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Therapeutic hypothermia has emerged as a highly effective neuroprotective therapy for cardiac arrest survivors. There are a number of purported mechanisms, but the exact mechanism still remains to be elucidated.
Although hypothermia generally down-regulates protein synthesis and metabolism in mammalian cells, a small subset of homologous (>70%) cold-shock mRNAs and proteins (RBM3, RNA-binding motif protein 3 and CIRP, cold-inducible RNA-binding protein) are induced under these conditions. In this study, we compared the effects of moderate (33.5°C) and deep (17°C) hypothermia with standard normothermia (37°C) on the regulation of RBM3 and CIRP mRNA and protein expressions in organotypic slice cultures from mouse hippocampus (OHSC).
Methods: Organotypic hippocampal slice cultures (OHSC) were prepared from C57BL/6 mice postnatal 5 days old. Brain slices were incubated under moderate (33.5°C) and deep (17°C) hypothermia in a specially developed incubator in an atmosphere of 5% CO2 at 100% humidity for a maximum of 48 h. Normothermic control cells were incubated at 37°C throughout the experiments. Real-time RT-PCR, Western Blotting, and MTT Assay were also performed.
Results: Exposure to moderate hypothermia resulted in a significant up-regulation of both RBM3 and CIRP mRNAs in the murine OHSC, as compared to normothermia control. Up-regulation in mRNAs started after 4 hours cooling and continued to increase till experimental end (48 hours). Interestingly, exposure to deep hyperthermia did not resulted in a significant increase in RBM3 or CIRP transcripts. RBM3 protein expression was also significantly up-regulated by moderate hypothermia, but starting after 24 hours exposure and remained elevated till experimental end. No significant up-regulation of CIRP protein expression was observed in the slice cultures at any time points.
Conclusion: We observed that RBM3 gene and protein expressions in brain slices were significantly up-regulated upon exposure to moderate hypothermia. These findings further support the implication of RBM3 as a potential effector for hypothermia-induced neuroprotection.

P-47
Hypothermia reduces inflammation in a co-culture model of neuronal and microglial cells

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Background: Deep Hypothermia is a standard method for neuroprotection during cardiac surgery in children. However, the cellular mechanisms which are induced by deep hypothermia have not been clearly understood in vitro. Therefore we investigated the effects of deep hypothermia and rewarming on the co-culture model of a murine BV-2 microglial cell line and a murine HT-22 neuronal cell line.
Methods: HT-22 neuronal cells and BV-2 microglial cells were co-cultured 24 hours before the experiment at 37°C. The co-culture cells were exposed to 17°C for 2 hours and then slowly rewarmed to 37°C within 2 hours. Non-specific microglial cell activation was performed by incubation of cells for 4 hours with 1μg/ml Lipopolysaccharid (LPS). In addition, the cells were damaged with 100 mM Glutamate to imitate neuronal damage for 4 hours. Morphology was documented using phase- contrast microscopy. The viability of the co-culture model was quantified by MTT assay. Flow cytometric analysis was performed with CD11b to differentiate both cell types. IL-6, MCP-1 and TNFα levels were measured as markers inflammation.
Results: There was no significant difference in cell viability under hypothermic conditions in comparison to the control cells kept at 37°C for 24h.
Furthermore deep hypothermia led to morphological changes. from a ramified and resting status under 37°C to amoeboid shaped cells under 17°C even without LPS stimulation. Neuronal cells change their network stucture. Under 37°C the long neurites are connected, while under 17°C neuritis become short and cell bodies touch directly.
The secretion of the pro-inflammatory cytokine IL-6 was significantly decreased after the induction of deep hypothermia and rewarming in comparison to normothermic controls. After 2 and 4 h the IL-6 concentration assimilated for normothermic and hypothermic cells. Moreover, MCP-1 release was significantly decreased after 2 and 4h in the normothermia and hypothermia groupe.
Conclusion: Deep hypothermia has no influence on the cell viability but reduces the release of the proinflammatory cytokines IL-6, MCP-1 and TNFα in our co-culture model of neurons and microglial cells

P-48
"Idiopathic" cardiomyopathy in children is highly associated with infectious genome presence in the myocardium

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Introduction: Viral infections are major causes of myocarditis, sometimes progressing to myocardial dysfunction, but incidentally found cardiomyopathy (CM) is usually deemed "idiopathic". The purpose of this study was to test endomyocardial biopsies (EMBs) and blood from "idiopathic" CM children for infectious genomes using polymerase chain reaction (PCR) to assess the role of cardiac infections in the pathogenesis of CM.
Methods: Blood and EMBs were obtained for PCR and histologic analysis from 26 consecutive paediatric patients with unexplained CM aged 8.5 ± 7.3 years. Workup included ECG, ECHO or MRI with decreased EF present in only 58% of patients. PCR and reverse transcription–PCR were performed to detect the genomic sequences of enterovirus (EV), adenovirus (ADV), cytomegalovirus (CMV), herpes simplex virus (HSV), Epstein-Barr virus (EBV), human herpesvirus (HHV), parvovirus B19 (PVB19), influenza A and B viruses, chlamydia and mycoplasma.
Results: Infectious genomes could be amplified from EMBs of 24/26 (92.3%) of patients: EV in 2 (7.6%), CMV in 2 (7.6%), HSV in 7 (26.9%), EBV in 4 (15.4%), HHV in 4 (15.4%), PVB19 in 3 (11.5%), chlamydia in 12 (46.2%) and mycoplasma in 1 (3.8%) including 9 (34.6%) cases with multiple agents. Blood PCR was positive in 17/26 (65.4%) patients, in 13 with exactly the same and in 4 with fewer agents. No case had active or borderline myocarditis according to the Dallas classification. Patients with positive EMBs were treated with respective antibiotic or antiviral agents. Over 1.2 ± 0.9 years follow-up, 2 patients died from heart failure, 1 received left ventricular assist device and the rest are clinically stable. Repeat testing was performed in 7 patients 0.4 ± 0.3 years after the initial evaluation, with PCR showing persistence of infectious genomes in EMB in all 7 patients (fewer agents in 3) and in blood in only 1 patient.
Conclusions: Infectious genomes were very frequently detected in EMBs of patients with unexplained CM without active or borderline myocarditis irrespective of the presence of systolic LV dysfunction, suggesting that infectious myocardial involvement, often with multiple agents, may be important in the pathogenesis of CM. Further research is needed concerning possible antiinfectious treatment and management of these patients.

P-49

Pulmonary CT number demonstrates congestive lung in patients with simple total anomalous pulmonary venous connection

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Background: Pulmonary CT number is reported to help in determining the severity of emphysematous lesion. But little is available on lung congestion. In patients with total anomalous pulmonary venous connection (TAPVC) lung congestion occurs because of pulmonary venous obstruction (PVO). We investigated pulmonary CT number to evaluate the lung conditions in patients with TAPVC.
Methods: The medical records of 20 patients with simple TAPVC were reviewed. PVO was judged by echocardiography. Between 2005 and 2009 they underwent High-resolution computed tomography (HRCT) by contrast medium both before and after repair. HRCT scanning was performed with 1.0-mm-thick axial sections. Pulmonary CT number was estimated on three horizontal slices (upper, middle, lower). Regional of interest (ROI) was established to avoid large blood vessels and bronchi. CT number was computed in the range of 30 pixels and represented by Hounsfield units (HU).
Results: Nine patients had preoperative PVO and seven had postoperative PVO. All pulmonary CT numbers were distributed from -214 to -686 HU. Preoperative pulmonary CT numbers in patients with infracardiac type TAPVC were significantly larger than those in patients with supracardiac type (-328 vs -437 HU). In early postoperative period pulmonary CT numbers in patients with infracardiac type were still higher. Before operation pulmonary CT numbers in patients with PVO tended to be larger than those in patients with non-PVO (-353 vs -429 HU; $p = 0.083$). In middle post-operative period CT numbers in patients with PVO tended to be larger (-483 vs -580 HU; $p = 0.095$). Three patients underwent surgery of PVO release. Two patients with pulmonary CT numbers decreasing before re-operation achieved success. But a patient with CT number increasing before each re-operation died.
Conclusion: In pulmonary congestion lung water volume increases and pulmonary CT number becomes larger. That CT numbers in infracardiac TAPVC with all preoperative PVO were significantly larger might suggest pulmonary CT number reflected pulmonary congestion by PVO. That patients with CT number improving before re-operation had good progression showed pulmonary CT number served as evaluating intra-individual change of lung conditions. Pulmonary CT number may reflect congestive lung comprehensively in patients with total anomalous pulmonary venous connection.

P-50

Heart Transplantation in Adult patients with Congenital Heart Disease

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Since 85% of patients with Congenital Heart Disease (CHD) can currently survive and reach adulthood, Heart Transplantation (HTx) is becoming a novel challenge in adults with CHD (ACHD). The aim of this study was to assess results and outcomes of HTx in ACHD.
Material and methods: Clinical data, demographics and outcomes of patients with underlying diagnosis of CHD transplanted at ⩾18years of age, were retrospectively reviewed.
Results: Among – adult heart transplantations performed from 1991 to 2011, 20 recipients were ACHD (13 males). Age at transplant was 21.6 ± 4years (median 19.6). Sixteen of 20 (80%) were performed since 2000 and only 4 (20%) before. CHD included: double inlet single ventricle (10 cases), tricuspid atresia (2), transposition of great arteries (2), unbalanced atrioventricular septal defect (2à and miscellaneaous (4 cases: 1 Ebstein anomaly, 1 HLHS, 1 ALCAPA, 1 double outlet right ventricle). Seventeen had at least one previous cardiac surgery (85%), i.e: Fontan procedure in 5 (25%), palliative procedures in 10 (50%) and Mustard operation in 2 (10%). Three cases had no previous cardiac surgery (15%), i.e: 1 Ebstein, and 2 unbalanced AVSD.
Two early deaths occurred at day-1 and day-23 post-transplant, due to pulmonary hypertension and graft RV failure. One late death occurred at 9.6 year FU post-transplant, and one patient

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underwent re-HTx at 6-year FU, due to graft coronary artery disease. Follow-up post-transplant was 6.4 ± 5.3 years (1 to 20.9, median 5.3). Survival rates were 90% at 1 and 5-years post-transplant and 72% at 10 and 15 years. All survivors (17 cases = 85%) are doing well in NYHA class I. Overall<1year mortality. Fontan procedure, gender, pre-transplant status did not impact significantly on survival.
Conclusion: The results of this small sample size study shows that HTx is an acceptable option for ACHD patients with end-stage failure of myocardial function and no other therapeutic alternative.

P-51
Ventricular assist device as a bridge to heart transplantation in children with an end-stage heart failure–own experience.
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Introduction: Extremely high mortality in children awaiting heart transplantation (OHT) in numerous centers triggered the development of ventricular assist device (VAD) application program. The majority of potential heart transplant recipients develop severe circulatory failure requiring continuous catecholamine infusion and following VAD use.
Aim: Presentation of our own experience regarding VAD application in children with end-stage heart failure.
Material and methods: From November 2009 to December 2011 VAD was used as a bridge to heart transplant in 6 children. Their clinical records were analyzed retrospectively. There were 4 males and 2 females with average age of 10,25 (1,5 – 17) yrs old, average weight–25,9 (8,2-54,0) kg. Every patient was diagnosed with dilated cardiomyopathy. In two of them VAD type Polcas-Religa (FRK, Zabrze, Poland) was used. One of them required left ventricle assist device (LVAD) and the second–biventricular assist device (BiVAD), Remaining four patients received Berlin-Heart (Mediproduct, GmbH, Berlin, Germany) LVAD. The mean assist time was 148 days (12 to 888 days). During the VAD therapy motoric rehabilitation was implemented.
Results: Significant body mass increase was observed during VAD application: mean-9,25 kg (range: 4-17 kg) and improvement in general condition (motoric and psychological). In one patient with BiVAD major complications were observed–bleeding from the cannulas' site, ischemic stroke, systemic infection (sepsis). In four patients OHT was performed in our center. Mean time of VAD applications before OHT was 209 days. In three of them good outcome was observed. Mean follow-up after OHT is 281 days. One death occurred in early post-transplant period. In 2 children VAD is still used, they are awaiting a donor heart. In one patient after OHT standard biopsies show grade 3A ISHLT cellular rejection, which is treated according to the protocol.
Conclusions: Our preliminary observations confirm effectiveness of VAD application as a bridge to OHT in patients with end-stage heart failure. Improved clinical condition with body gain of children during VAD use facilitates subsequent OHT.

P-52
Extensive myocardial infarction (MI) in a 11year old girl – case report
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MI in childhood is a very rare event. It is almost always associated with structural (i.e. Bland-White-Garland, coronary fistulas), hereditary (i.e. hyperlipidemia/hypercholesterolemia, metabolic or other storage disorder) or acquired (i.e. Kawasaki, Kounis, diabetes, tumor) disease.
We report on 11year old girl who was presented after a near drowning episode to a peripheral hospital. The child was in cardiopulmonary shock and had to be ventilated. Despite volume and start with catecholamins blood pressure remained low. A severely depressed myocardial function was stated in a quick echocardiogram. Initial blood work showed white blood cell count of 27'000 with an abnorm I/T ratio, CK 7000U/I, CK-MB 688ug/l, Troponin 12 ug/l. The child's family was suffering from an upper respiratory tract infection with cough and rhinitis. Otherwise the girl's history was unremarkable. After stabilization the child was transferred to our center.
Repeated echocardiogram revealed a severely reduced left ventricular function (ejection fraction (EF) 25-30%) with significant dyskinesia/hypokinesia along the free left ventricular wall while the right ventricular systolic function remained normal. ECG showed sinus tachycardia, deep Q-waves in V3-V5, ST elevation in V1-V2. Based on these findings we primarily diagnosed an acute myocarditis (differential diagnosis acute vasculitis with coronary vasospasm, transient hypoxic ischemia after near drowning).
Within the following 2 days the girl's left ventricular function recovered (EF 50-55%). A sudden episode with nausea and vomiting on day 3 preceeded ventricular tachycardia and subsequent ventricular fibrillation. After resuscitation and ECMO cannulation a coronary angiogram was performed. It revealed a widely spread thin network of arteries along the left coronary artery with a discrete narrowing of the main stem. The right coronary artery was normal. A thin slide CT scan finally revealed an anomalous orifice of the left coronary artery coming from the non-coronary cusp.
Surgical unroofing of the left main coronary artery was performed. Unfortunately despite regained normal coronary artery flow the left ventricular function remains severely depressed (EF 20-25%). The girl is listed for heart transplantation.
Conclusion: Presence of segmental myocardial dyskinesia/hypokinesia always implies further diagnostic examination regardless the patient's age.

P-53
The relationship between intima media thickness of pulmonary artery and biochemical parameters in children with Eisenmenger syndrome
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Introduction (or Basis or Objectives): Eisenmenger Syndrome is defined as irreversible pulmonary hypertension, resulting from inversion of the left to right shunt at aorta-pulmonary, ventricular or atrial level. This study was aimed to determine the relation o intima media thickness of pulmonary artery with N-Terminal pro-brain natriuretic peptide (NTpBNP), uric acid, endothelial micro-particle levels, atherosclerotic risk factors. Also echocardiographic risk factors and hemodynamic data in children with

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Eisenmenger syndrome were studied according to emphasize their importance for prognosis of disease.
Methods: 16 patients (10 male and 6 female) aged 3-18 years with Eisenmenger syndrome were included in this study. Thickness of pulmonary artery intima was measured ultrasonograhically in order to detect vascular changes. Levels of NTpBNP, uric acid, endothelial micro-particles (CD144, CD146) and atherosclerotic risk factors were studied biochemically. Echocardiography was performed fort the assessment of ventricular functions. Hemodynamic evaluation was done in patients with right atrial catheterisation. The data gathered except those obtained through hemodynamic assessment; was compared with 37 healthy children.
Results: Significant increase in NTpBNP, uric acid, CD144 and CD146 levels in addition to significant decrease in HDL cholesterol, total cholesterol and fasting blood glucose levels were observed. Echocardiographic examination of intima media thickness of pulmonary artery was found significantly high Also body surface area, body mass index, systolic blood pressure and mean blood pressure were found significantly low.
Conclusion: Even processes starting the pathological differentiation in pulmonary arterial hypertension are still not known however it is accepted that multiple factors including various biochemical pathways, cell types take role in pathogenesis. The mechanisms take part in pulmonary vascular resistance are vasoconstriction, obstructive remodelling, inflammation and thrombosis. Effect of NTpBNP, uric acid, endothelial microparticles and atherosclerotic risk factors on intima media thickness of pulmonary artery and interrelationships with each other were aimed to be discussed in this study.

P-55
Prenatal diagnosis of isolated total anomalous pulmonary venous connection: a series of fourteen cases

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Background: Prenatal diagnosis of total anomalous pulmonary venous connection (TAPVC) diagnosis is possible but remains a challenging issue in foetal echocardiography. The objective of this study was to determine accuracy and outcome of 14 foetuses diagnosed with isolated TAPVC.
Methods: We review our ten-year-experience of prenatal diagnosis of isolated TAPVC and describe the most relevant echocardiographic features. Medical records and echocardiographs of foetuses with a prenatal diagnosis of isolated TAPVC were retrospectively assessed. Special attention was paid to the foetal echocardiographic features motivating referral for expert echo scan.
Results: Expert foetal echocardiography identified 14 foetuses with isolated TAPVC. Prenatal diagnosis was made at a mean gestational age of 30 weeks (25 – 35 weeks). One woman was lost to follow-up before birth. There was one termination of pregnancy (TOP) for chromosomal abnormality. Two infants had a normal heart after delivery. Prenatal diagnosis of isolated TAPVC was confirmed in ten infants at postnatal assessment. All infants with confirmed TAPVC underwent corrective surgery. There was one in-hospital death after 1 $\frac{1}{2}$ months due to respiratory failure. At the time of data collection, all patients were in good clinical condition without pulmonary hypertension or stenosis of pulmonary venous anastomosis after a mean follow-up of 30.5 months (1, 2-103, 8).
Conclusion: Foetal echocardiography can diagnose isolated TAPVC even in absence of complex congenital heart disease but remains challenging. The diagnosis has to be made by direct visualization of the pulmonary venous confluence behind the left atrium and absence of pulmonary veins connecting to the left atrium.

P-56
Risk factors associated with in utero demise in fetuses with congenital heart disease: A case/control study

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Basics: The objectives of this study are to define the prevalence of in utero demise (IUD) in fetuses diagnosed with congenital heart disease (CHD) and to identify prenatal echocardiographic predictors of IUD. IUD occurs in all types of CHD. Factors associated with IUD are largely unknown.
Methods: Between Jan 2004-Dec 2010, cases of IUD were identified from the Hospital for Sick Children database. Control echocardiograms were cases of fetal CHD diagnosed within 48 hrs of the case. For multiple possible controls, the control with more severe CHD was chosen. Multivariate regression models were used to determine echocardiographic predictors of IUD.
Results: Of 1584 fetal cases, 74 IUD were identified (4.6%). IUD cases were diagnosed earlier (mean gestational age was $23.8 \pm 5/3$ weeks vs. 26.7 ± 5.8 weeks controls, $p = 0.002$) and were smaller (biparietal diameter 5.6 ± 1.6 cm vs. 6.58 ± 1.6, $p = 0.001$). The mothers were healthy in 80% of both groups. Main diagnoses were: hypoplastic left heart ($N = 10$, $N = 11$ control), hypertrophic cardiomyopathy ($N = 8$, $N = 0$ control), isolated ventricular septal defect ($N = 6$, $N = 9$ control). The majority demised in the 2nd trimester ($N = 39$, 53%). Cause of demise was hydrops in 17, arrhythmia in 11 (tachycardia in 8, complete heart block in 2 and 2:1 atrioventricular block in 1) and unknown in 46. Echocardiographic variables associated with IUD after multivariate analysis were: cardiomegaly ($p = 0.0005$, HR 26.7), extracardiac abnormalities ($p = 0.002$, HR 10.2), umbilical artery absent end diastolic flow ($p = 0.05$, HR 7.8), and any pericardial effusion ($p = 0.02$, HR 5.8).
Conclusions: Basic fetal echocardiographic parameters such as cardiomegaly and pericardial effusion are associated with IUD.

P-57
Fetal ventricular aneurysms and diverticula: Nine new cases and a review of the current literature

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Introduction: Congenital ventricular aneurysms (CVA) and diverticulum (CVD) are rare conditions with a variable prognosis when diagnosed in fetal life. There have been isolated case reports but little evidence to predict survival or outcome. Here we present a large fetal case series and review a total of eighty-two cases to ascertain prognostic factors that may aid with prenatal counselling.
Methods: Retrospective search of our fetal cardiology database between 1991 and 2011 for cases of CVA or CVD. Patient

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notes, fetal echocardiograms and relevant postnatal investigations were reviewed. A literature search identified 73 previously published cases.
Results: Two fetuses had CVD, one arising from the left ventricle (LV) and one from the right ventricle (RV). There were seven cases of CVA with 5 arising from the LV and 2 from the RV. Two cases of CVA had poor ventricular function and parents opted for termination of pregnancy (TOP). One patient with a rapidly expanding LV aneurysm suffered an intrauterine death (IUD). All six live-born infants remain well on follow-up (range 3 months – 18 years). Reviewing all published cases, there were 57 survivors, 10 TOP, 11 IUD and 4 neonatal deaths. Fetal hydrops, ventricular dysfunction, increasing size and cardiomegaly were all independently associated with non-survival ($p < 0.05$). These poor prognostic signs were more frequent in those with a LV aneurysm. Eleven fetuses had associated structural heart defects (predominantly septal defects), overall this did not affect outcome. Presentation in later gestation and those with only fetal dysrhythmia tended to have a favourable outcome ($p < 0.05$). Pericardial effusion was present in 33 cases and associated with a poor outcome in the LV group ($p < 0.05$). Prenatal intervention with pericardiocentesis was performed most frequently in those with RV diverticulum who had a favourable outcome with or without intervention.
Conclusion: CVA and CVD are a heterogenous group of conditions where prognosis depends on location and type of lesion. Our review demonstrates that detailed fetal cardiology assessment to look for associated sequelae can further help to predict prognosis and guide prenatal counselling. Intervention should be discussed on a case by case basis. The aetiology of these lesions remains uncertain.

P-58
Various coexisting forms of fetal arrhythmias associated with fetal atrial septal aneurysm

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Premature atrial contractions (PACs) are a common form of cardiac arrhythmia, which are usually benign and associated with good outcomes. We report a fetus with PACs detected in the third trimester and complicated by episodes of fetal bradycardia and sinus tachycardia secondary to an atrial septal aneurysm (ASA) originating in the region of fossa ovalis and extending into the left atrium.
Fetal echocardiography performed at 33 weeks' gestation showed an ASA and recurrent blocked bigeminal PACs causing paradoxical bradycardia of 100-120 bpm. Redundant atrial tissue was seen hitting the left atrial free wall and the mitral valve annulus. The patient was monitored with weekly fetal heart rate assessment. At 36 weeks' gestation, fetal echocardiography showed PACs, intermittent sinus tachycardia of 180-190 bpm, mild tricuspid regurgitation and minimal pericardial effusion. Elective caesarian section was performed at 37 weeks of gestation, and the newborn was in good condition. Neonatal electrocardiography revealed only rare PACs. PACs resolved spontaneously at 2 months of age, when ASA was observed to adhere to the septum secundum.
While PACs are generally known to be benign phenomena, coexisting ASA may complicate the clinical picture and cause more significant forms of fetal cardiac arrhythmias, requiring a closer follow-up.

P-59
Role of decreasing maternal ingestion of polyphenol-rich foods on fetal ductus arteriosus dynamics in normal pregnancies: an open clinical trial.

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Basics and objective: We have recently demonstrated that reversal of fetal ductal constriction occurs after dietary maternal restriction of polyphenol-rich foods (PRF), such as herbal teas, dark chocolate, grape and orange juices, olive oil, berries and others (J Perinatol, 2011). Other clinical and experimental evidences have already corroborated the association of maternal ingestion of PRF to fetal ductal alterations, due to interference on prostaglandin synthesis. This study tested the hypothesis that normal third trimester fetuses also improve ductus arteriosus dynamics after a polyphenol-poor diet for a period superior to 2 weeks.
Methods: An open clinical trial was designed, in wich 46 fetuses with gestational age (GA) $\geq$ 28 weeks were submitted to 2 Doppler echocardiographic studies with an interval of at lest 2 weeks. Systolic and diastolic ductal velocities (SDV and DDV), pulsatility index (PI) and right to left ventricular dimensions ratio (RV/LV) were assessed. The examiners were blind to maternal dietary habits at the first examination. After the first study, a detailed food frequency questionnaire was applied and a diet based on polyphenol-poor foods (<30 mg polyphenols/100 mg) was reccomended. A control group of 26 third trimester fetuses in which no dietary intervention was offered was submitted to the same protocol. Statistical analysis used t test for independent samples.
Results: Mean GA was 33 ± 2weeks (28-38 weeks). Mean daily maternal estimated polyphenol intake was 1277 mg.After dietary orientation, mean daily polyphenol consumption decreased to 126 mg($p = 0.0001$). Comparing the 2 echocardiographic studies, significant decreases in SDV, DDV and RV/LV ratio, as well as increase in ductal PI were observed [DSV = 1.2 ± 0.4m/s (0.7–1.6) to 0.9 ± 0.3m/s (0.6-1.3)($p = 0.018$); DDV = 0.21 ± 0.09m/s (0.15-0.32) to 0.18 ± 0.06m/s (0.11-0.25)($p = 0.016$); RV/LV = 1.3 ± 0.2 (0.9–1.4) to 1.1 ± 0.2 (0.8-1.3)($p = 0.004$); ductal PI = 2.2 ± 0.03 (2.0 – 2.7) to 2.4 ± 0.4 (2.2–2.9)($p = 0.04$)]. In the control group, with GA of 32 ± 4weeks (29-37 weeks), there were no significant differences in mean daily maternal ingestion of PRF, mean SDV, DDV, PI and RV/LV ratio after a period of 2 weeks.
Conclusion: The oriented restriction of third trimester maternal ingestion of polyphenol-rich foods for a period of 2 weeks or more improve fetal ductus arteriosus flow dynamics and right ventricular dimensions.

P-60
10 Year Review of Outcome of Fetal Arrhythmias in Wales

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Objectives: To evaluate the frequency, diagnostic features, clinical management and outcome of fetal arrhythmia.

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Method: 2084 fetuses referred to the University Hospital of Wales for the last 10 years were included in the study.
Results: 208 fetuses had provisional diagnosis of fetal arrhythmia (10%). 194 fetuses had normal heart, and 14 showed structural cardiac abnormality. In 80 patients fetal arrhythmia had resolved spontaneously by the time of specialist cardiac scan. Arrhythmia had persisted in 128 patients; of which complete heart block (CHB) was detected in 16; supraventricular tachycardia (SVT) in 31; premature atrial contractions (PAC) in 67, and premature ventricular contraction in 7 fetuses. PAC later triggered sustained re-entrant tachycardia in two fetuses. 5 patients with PAC had adverse outcome; three with oligohydramnios died suddenly at term, and two with restrictive foramen ovale required emergency caesarean section at 38 weeks. 29 patients with sustained tachycardia received antiarrhythmic medications; and except for one all responded to digoxin and flecainide combination. 11 of 16 fetuses with CHB survived the pregnancy giving an intrauterine mortality rate of 30%. 8 patients had permanent pacemaker following birth.
Conclusion: Fetal arrhythmias are a common referral indication for fetal cardiac assessment. PACs are generally benign but if they are associated with oligohydramnios or restrictive foramen ovale outlook may be unpredictable requiring close follow up. SVT is better responds antiarrhythmic combination treatment, but CHB carries a substantial mortality.

P-61
Features and outcomes of cases with agenesis/dysplasia of the pulmonary valve diagnosed in fetal life

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Background: Agenesis of the pulmonary valve (APV) or dysplastic pulmonary valve (DPV) is a rare anomaly, occurring mostly as a variant of Tetralogy of Fallot (TF). Its features are pulmonary stenoinsufficiency of a variable degree with dilated pulmonary branches, causing cardiomegaly and potential lung hypoplasia.
Material – methods: Out of around 800 cases with congenital heart disease examined by fetal echocardiography during the past 10 years, 8 were found to have a pattern of APV/DPV at 21–32 weeks' gestation (w.g.). Six cases (group 1) had TF, while in 2 other cases (group 2) the anomaly was isolated. A global score was introduced to summarize the echocardiographic findings: z-score of PV anulus+degree of other findings in points 1,2,3.
Results: Group 1: Three fetuses had microdeletion 22q11.2, they developed at 25-26 polyhydramnios, progressing to fetal hydrops in 2; all died after preterm birth, one after operation. All these cases had a large pulmonary anulus (z-score >4), marked pulmonary insufficiency (PI), large dilated pulmonary branches, cardiomegaly and high global score. Three other cases had smaller pulmonary anulus (z-score 1.5 -3), major degree of pulmonary stenosis (PS), lesser degree of PI and cardiomegaly and lower global score; they were delivered at term and operated at 5 and 6 months, surviving. The ductus was absent in all.
Group 2: One case had a moderate PS and PI and is well, stable at 5 years. The second case had a peculiar form of valve dysplasia, with a marked PI and a markedly enlarged right ventricle, improved after birth, was operated at 2 years and is well at 5 years. The ductus closed spontaneously in both.
Conclusions: The data of our small series show a better outcome in the fetuses with APV associated to TF who had a major degree of PS than in those with a large pulmonary anulus and a marked PI. The isolated form presents probably a better prognosis.

P-62
Intracardiac Tumors in Neonates: Report of 2 Cases of Myxomas.

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Introduction: Cardiac tumors are rare in children, with primary benign tumors more frequent than malignant lesions. Although cardiac myxoma is the most common primary tumor in adults, rhabdomyoma is the most frequently encountered in infancy and myxoma are uncommon.
Methods and Results: We report 2 cases of intracardiac myxomas in very young patients, respectively diagnosed prenatally and in the early neonatal period.
A 29-year-old G1P0 woman was referred for evaluation of a heart mass detected at 21 weeks gestation. At fetal echocardiogram, the tumor appeared unique (7×5 mm), sessile and was localized at the apex of the right ventricle. The echogenicity was homogeneous without calcifications and slightly hyperechogeneous compared to the myocardium. According to power-Doppler, the tumor was not highly vascularized. The rhythm was normal and there was no hemodynamic compromise. At 27.4 weeks, the tumor was slightly larger with as only repercussion mild tricuspid valve regurgitation. A multi-organic disease was excluded by ultrasound and MRI. Fetal karyotype was normal and there was no évidence for tuberous sclerosis. A 4.4 kgs full-term boy was born uneventfully. Clinical examination and ECG were normal. Postnatal echocardiogram defined the tumor as a sessile multilobed mass in the apex of the right ventricle. At 1-month of age, MRI evaluation led to the diagnosis of an isolated myxoma (17×11×12 mm) without hemodynamic compromise.
Our second case was diagnosed in a 2-week-old girl, born full term with 3.5 kgs, without any relevant personal or family history. A workup for cyanotic breath-holding-spells revealed an hyperechogeneous, homogeneous, sessile cardiac mass in the apex of the left ventricle. The biventricular function, cardiac anatomy and electrical activity were normal. MRI suggested a myxoma of oval form, 8×9×3 mm.
In both cases, a conservative approach was adopted because of the tumor stability and the absence of symptoms, arrhythmia and hemodynamic compromise. Our two patients are currently well at respectively 5-month and 3-year follow-ups.
Conclusions: Myxomas are very uncommon tumors in the pediatric age. They may however already be present prenatally and should be part of the differential diagnosis of cardiac masses. MRI may confirm the diagnosis. Surgical abstention is conceivable, especially when the patient is asymptomatic.

P-63
Prenatal diagnosis and management of fetal arrhythmias

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Introduction: Fetal arrhythmias can occur in about 1-2% of gestations. Fetuses affected show higher prenatal and neonatal morbidity and mortality rates.
Methods: Since January 1995 to April 2011 we observed 6143 fetal heart scans. Among these, we diagnosed 98 (1.6%)

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consecutive fetuses with cardiac dysrhythmia, defined as: sporadic irregular beats (<1 every 10 sec); frequent irregular beats (>1 every 10 sec); sustained tachycardia (>95° centile for GA for more than 10 sec); sustained bradicardia (<5° centile for GA for more than 10 sec). Rhythm diagnosis was based on M-mode, pulsed wave Doppler, tissue Doppler imaging (TDI) and Strain-strain/rate imaging.
Results: The mean gestational age at diagnosis was 25.5 +/- 4.5 weeks. 51/98 (52%) fetuses had irregular beats: 37/51 (73%) were sporadic and 14/51 (27%) were frequent. In 2/51 (4%) cases the prenatal diagnosis (obtained with strain/strain-rate imaging) was premature ventricular beats; in both cases diagnosis was confirmed after birth. No one out of the 51 fetuses showed signs of heart failure, so the mothers were never gived therapy. In 7/51 (14%) cases, the neonate was given oral therapy with flecainide or sotalol after the birth. 34/98 (35%) fetuses had tachycardia with a 1: 1 atrial-ventricular (AV) conduction. Based on ventricular-atrial interval, prenatal diagnosis was: 16 atrial-ventricular re-entry tachycardia (VA'< AV); 7 permanent junctional reciprocating (VA'> AV), 11 atrial ectopic (VA'> AV). 1/98 (1%) had atrial flutter, 2/98 (2%) had ventricular tachycardia, 3/98 (3%) had sustained sinusal bradicardia and 7/98 (7%) had congenital AV block. Only 20 fetuses were treated using oral maternal drug therapy (digoxin, sotalol or flecainide). They had incessant tachycardia (> 12 h, > 200 bpm), signs of left ventricular dysfunction, or hydrops. The total success rate (sinus rhythm or rate control) was 18/20 (90%). 10 fetuses were hydropics. 4 of these died (one at 28 weeks of gestation, one at 35 weeks of gestation, two in the first week of life). No misdiagnosis was made using TDI and Strain-strain/rate imaging. At 7 year mean follow-up, 94/94 children are alive and well.
Conclusions: Fetal echocardiography could clarify the electro-physiological mechanism of fetal cardiac dysrhythmias and guide the therapy.

P-64
Congenital Heart Malformations in the fetus with intracardiac echogenic focus in low vs high risk pregnancies

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Background: To study the prevalence of congenital heart malformations in fetuses with intracardiac echogenic focus (ICEF) in low versus high risk pregnancies .
Methods: We performed a detailed fetal echocardiography for fetuses who were found to have one or more intracardial echogenic focus in sonographic (US) examination from January 2007 through July 2010. They were divided in two groups. low risk and high risk pregnancies. Risk factors included: Maternal age > 35 years, first degree relative with congenital heart disease, diabetes mellitus, nuchal translucency, single umbilical artery, maternal drugs that affects the fetus, maternal SLE or PKU. Intracardiac echogenic focus was defined as a discrete area of echogenicity noted in the left or right ventricle, that was as bright as bone. In the low risk group participated 72 women. In the high risk group participated 74 women .In 11 cases there was another extracardiac malformation. Materanl age was 19-33 years and the gestational age was 19-35 weeks.
Results: In the low risk group there was two cases of small muscular VSD, s. In the high risk group there was 11 cases of congenital heart diseases. Two cases of hypoplastic left heart, 3 cases with ventricular asymmetry, 5 cases with VSD, one case with ASD and VSD, two cases with moderate aortic arch, and one case with echogenic bicuspid aortic valve.
Conclusion: detailed fetal echocardiography is waranted in high risk pregnancies in the presence of intracardiac echogenic focus and other extracardiac malformation.

P-65
A questionnaire study of Ro/ SSA autoantibody mediated congenital heart block indicate need for increased information, support and highly specialized medical care

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Introduction: Congenital heart block (CHB) may develop in the fetus of women with Ro/SSA autoantibodies. The aim of this population based study was to generate quantitative data on findings from a previous hypothesis-generating interview study investigating how women expecting a child with CHB experienced their pregnancy and post-partum period.
Methods: Women with Ro/SSA autoantibodies giving birth to a child with CHB in Sweden were identified in a population-based manner (n = 88) and offered to participate in the study. A questionnaire was constructed based on findings from previous individual interviews in a subgroup of the women. The questionnaire response frequency was 89% (n = 78).
Results: More than half of the women (57%) did not think that information on CHB obtained from health care personnel was sufficient, and close to 1/3 did not understand the information given to them. Most women attended both the regular maternity health care program and visited at least one specialist for supervision of the pregnancy, but commonly more. However, half of the women did not know who had the medical responsibility for them or for the child (50% and 46%, respectively). Further, the medical care differed substantially depending on the experience of the caregiver of the situation. The majority of the women (60%) expressed that they felt afraid when thinking about the child, and 48% felt responsible for the child developing CHB. The women were supported during the pregnancy and postpartum period by several persons; both relatives and caregivers, but 1/3 did not feel that it was enough and 45% wanted additional support. Despite this and the rareness and seriousness of the condition only 18% of the mothers were offered professional psychological support.
Conclusions: Increased awareness and knowledge of autoantibody-mediated CHB are needed to provide adequate care. Information should be provided both to patients and professional care givers. Referral to specialized centers and offering psychological support could increase the quality of medical care and release the families from unnecessary stress and feeling of guilt.

P-66
Ro52 autoantibody-positive women's experience of being pregnant and giving birth to a child with congenital heart block

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Introduction: Congenital heart block may develop in the fetus of women with Ro/SSA autoantibodies. The aim of this study was to investigate how women expecting a child with congenital heart block (CHB) experienced their pregnancy and postpartum period.

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Methods: Women pregnant with a child with CHB in Sweden during 2000-2009 (n = 21) were identified in a population-based manner and individually interviewed post pregnancy using a semi-structured interview guide. The interviews were audio taped, transcribed verbatim and analyzed by qualitative content analysis.
Results: Three categories emerged from the responses; learning, suspense and facing.
The category learning contained both learning about the child's heart block, but frequently (16/21) also of autoantibody-positivity and a potential rheumatic diagnosis in the mother. The medical procedures and information differed considerably depending on area of residence and whom was encountered in the health care system. In many cases ignorance about this rare condition caused a delay in treatment and surveillance. The category suspense described the women's struggle to cope with the feeling of guilt and that the child had a serious heart condition and might not survive the pregnancy. The category facing included the postpartum period, leaving the hospital and adjusting to everyday life. It also describes contacts with child care welfare and relatives. Most of the women had tended to put their pregnancies "on hold", and some described that they needed prolonged time to attach to their newborn child.
Conclusions: Increased awareness and knowledge of CHB are needed to provide adequate and secure care. Offering psychological support by a health care professional to the women and families to give them tools to handle the situation with a potential rheumatic diagnosis for the mother and to facilitate the early attachment to the child should be considered.

P-67
Assessment of diastolic function after cardioversion of the fetal supraventricular tachycardia using pulsed Doppler and myocardial tissue Doppler.

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Objectives: To evaluate fetal cardiac diastolic function after cardioversion of supraventricular tachycardia (SVT) using pulsed Doppler (PD) and myocardial tissue Doppler (MTD).
Methods: 56 fetuses with SVT were diagnosed in the referral centre for fetal cardiology. Sinus rhythm was obtained in 90% after transplacental treatment with digoxin and/or amiodarone. In 13 fetuses with normal heart anatomy and SVT successfully treated in utero a long-term observation of the PD and MTD was performed. We analyzed the E/A ratio in the conventional pulsed Doppler tricuspid flows and the Em/Am ratio in the myocardial tissue Doppler with the sample volume placed at the basal segment of the right ventricular free wall.
Results: In 6 fetuses with SVT after cardioversion the E/A ratio was above 1,0 from 2 to 10 days (mean 4,3 ± 3 days). In 9 fetuses with SVT after cardioversion the Em/Am ratio was above 1,0 from 2 to 35 days (mean 15 ± 13 days). In 7 cases the ratio of Em/Am>1,0 lasted longer (mean 9,5 ± 13 days) than the ratio of the E/A>1,0. In the remaining 6 fetuses the normalization of Em/Am ratio took place simultaneously with the normalization of the E/A ratio.
Conclusions: After cardioversion of SVT the normalization of the E/A ratio occurred earlier than normalization of the Em/Am ratio. MTD may be more sensitive than atrioventricular spectral Doppler for the detection of transient ventricular diastolic dysfunction in fetuses after cardioversion of supraventricular tachycardia. This symptom may be associated with abrupt improvement in myocardial relaxation during sinus rhythm, pressure drop in end-diastolic phase and, consequently, increasing the pressure difference between the atria and ventricle.

P-68
Potential Added Value of Bidimensional Myocardial Strain in Prenatal Diagnosis of Aortic Coarctation

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Introduction: Prenatal diagnosis of coarctation of the aorta (CoA) is difficult. Strain (S) and strain rate (SR) imaging is a new non-invasive ultrasonic technique able to quantify regional myocardial deformation properties. It has a superior sensivity over that of standard echocardiography and myocardial velocity for non-invasive assessment of ventricular function. Aim of our study was to assess the ability of bidimensional myocardial S and SR analysis in prenatal diagnosis of CoA.
Methods: We studied 35 consecutive foetuses (mean gestation age 28 ± 4.9weeks) with RV and PA prevalence. In all foetuses we studied, left ventricle (LV) to RV ratio, Aortic (Ao) to PA ratio and average global peak systolic RV S and SR. RV prevalence and PA prevalence were defined as a ratio <5° percentile for gestational age. Normal value for S and SR were obtained comparing with normal value previously established by our group in 100 consecutive normal foetuses.
Results: Among 35 consecutive foetuses with RV and PA prevalence, 8 (22,8%) had CoA at echocardiographic evaluation in the neonatal period, 27 had no cardiac abnormalities at postnatal echocardiographic evaluation.
-LV/RV was not significantly lower in foetuses with CoA (mean value 0.68 vs 0.69; p = 0.13).
-Ao/Pa was significantly lower (mean value 0.60 vs 0.68; p < 0.05).
-LV/RV ratio inferior to 0.67 showed a sensitivity of 42% and a specificity of 72.9%.
-Ao/Pa ratio inferior to 0.68 showed a sensitivity of 68% and a specificity of 66.7%.
-Peak systolic RV S showed normal values (-24% ± 4) in the 27 foetuses with no CoA at post-natal evaluations, whereas it was significantly reduced in CoA group with (-15 ± 3.7, p < 0.0001), with a cut-off value of -18% (sensivity 85.7%, specificity 86.7% at ROC, Figure 1).
Conclusions: Precise diagnosis of CoA during fetal life remains difficult. Our preliminary data suggest that 2D-strain could be a helpful approach to improve the prenatal diagnose CoA.

P-69
Foetal Dysfunction of the Arterial Duct: Clinical Spectrum and Outcome

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Background: Prenatal ductal complete closure is a recognised entity causing pathology at different levels of the right heart and pulmonary vasculature. Outcome of milder dysfunction needs to be defined.
Methods: retrospective study; foetal (1008) and neonatal (5400) echo databases searched for evidence of prenatal ductal dysfunction (1998-2011). Inclusion criteria: prenatal closure – constriction-aneurysm of arterial duct, postnatal (<7d) excessive RV

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hypertrophy, cyanosis by atrial R < L and/or obvious ductal pathology.
Results: 31 patients: 13 prenatally (GA29w,21-38w), incidence 1.3% abnormal scans), 18 postnatally (D0-6).
Foetal ductal abnormalities: complete closure (n = 9), constriction (n = 3) or aneurysmatic dilatation (n = 1). 3 Mothers had taken NSAID. Clinical presentation at birth was cyanosis (sat.<85%) (74%), severe pulmonary hypertension (38%), heart murmer (6%); 20% were asymptomatic. On echocardiography patients had excessive RVH (28), a bipartite RV (4), RV aneurysm (1), significant TR (17), chordal rupture with functional PA (2), hydrops (1), PS (6) and PR (4) ranging from mild to « agenesis » of pulmonary valve (3), dilation of pulmonary trunk and branch pulmonary arteries (8), compression of airways with "air-trapping"(2), thrombosed aneurysm of the duct (4), 3 with extension of the into the left PA and PPS. In 7 patients premature delivery was chosen to avoid further damage of the right heart & lungs. Neonatal treatment varied from observation (5), oxygen administration only (13), ventilation with pulmonary vasodilators (12), ECMO (1), heparine IV (3) Late treatment (7): thrombectomy of the PA (2); balloon dilation PS. (3), RVOT plasty (1), homograft RVOT (1), mitral valve plasty for CCMP with severe MR (1). 3(10%) Pts died in the neonatal period: 2 with massive TR and functional PA, 1 with severe respiratory insufficiency due to microcystic lung disease – air trapping. (14)45% went to cardiopulmonary normalisation, 14(45%) had residual lesions: PS (5), supraPS-PPS (6), CCMP (1), PI (3), TR (2/31).
Conclusions: Fetal dysfunction of the arterial duct can stress different levels of the right heart and pulmonary vasculature, resulting in a very wide range of pathology. The clinical outcome ranges from normal to residual hemodynamic lesions or death. Premature delivery might be indicated in selected patients.

P-70

Assesment of Diastolic Ventricular Function in Fetuses of Gestational Diabetic Mothers Using Tissue Doppler

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Introduction: To evaluate of ventricular diastolic function by tissue doppler in fetuses of diabetic mothers and compare with healthy control groups.
Methods: Between November 2009, May 2011 38 mothers with gestational diabetes mellitus and 75 healthy control subjects were recruited prospectively to have fetal echocardiogram at 24, 28, 32 and 36 weeks of gestation to assess cardiac diastolic function and interventricular septum thickness. Diastolic function was evaluated by using tissue Doppler and pulse wave Doppler. Haemoglobin glycated (HbA1c) levels were obtained at 24 gestational week. Early (Ea) and late diastolic (Aa) peak tissue Doppler imaging at the base of right ventricular free wall, ventricular septum, left ventricular free wall and both atrioventricular valves early (E), late (A) inflow velocities were analyzed in 113 fetuses and compared between groups. General linear model repeated measure was used as a statistical analysis.
Results: In both groups fetal interventricular septum thickness increased progressively with advancing gestation. Fetal interventricular septum were significantly thicker in the presence of gestational diabetes mellitus ($p < 0.001$). Mean levels of HbA1c were $5{,}78 \pm 0.73\%$ in gestational diabetic mothers, $5.01 \pm 0.37\%$ in control group ($p < 0.003$). HbA1c was significantly higher in the gestational diabetic mothers. E, A, Ea, Aa, Ea/Aa and E/A ratios increased with gestational age at all sites in both group. Peak velocities of mitral E ($p < 0.001$), mitral A(0,007), tricuspid E(<0.001) and tricuspid A (0.002) were significantly higher in the presence of gestational diabetes mellitus. Ratios of E/A and Ea/Aa progressively increased in both group. Mitral E/A and tricuspid E/A of fetuses of diabetic mother were lower in third tremester. Ea: Aa ratio of right ventricle was higher in fetuses of mothers with gestational diabetes mellitus advancing gestation ($p < 0{,}03$).
Conclusions: Myocardial velocities obtained with TDE in conjunction with traditional Doppler methods provide an important non-invasive means of assessing diastolic function. We think that maternal diabetes mellitus is associated with impaired ventricular diastolic function in the fetus. İmpaired ventricular diastolic function seems as a result of decreased relaxation and compliance.

P-71

Impact of prenatal diagnosis of congenital heart disease on neonatal outcome in a regional case controlled study (Canton of Vaud, Switzerland)

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Introduction: Little epidemiologic evidence exists on the impact of a regionally organized fetal screening program. This study reports on the outcome of a fetal cardiac screening program in the Swiss canton of Vaud, population of 684'922 comparing the diagnostic data to the Eurocat register (ER) from 1.5.2003 to 31.12.2008
Methods: 40'567 births were registered with 572 cases of congenital cardiac pathology (CCP). The CCP were sorted in separate categories based on the severity.
Results: 126 of the 572 CCP could be attributed to 4 groups considered major cardiac congenital malformations. Prenatally diagnosed in this population were 83/126, 67%.
Group I (31), all CCP for which only palliative care is available, detected antenatally (28/31, 90.3%), resulting in TOP in 24 (85.7%). 7 born alive (4 diagnosed prenatally) of which 4 died (comfort care) 3 went on to be operated.
Group 2 (6 cases) severe heart disease requiring immediate postnatal intervention, TGA. 4 detected prenatally (66.7%), 2 TOP associated with chromosomal anomaly, 3 underwent arterial switch, 1 non-diagnosed TGA died.
Group 3 (heart disease requiring postnatal care but deferred surgical or interventional correction, conotruncal anomalies, AVSD), 47 cases in the ER, 33/47 detected prenatally (70.2%). 30/47 had a chromosomal anomaly, 20 TOP, 26 born alive, 1 died in utero. Of the 26 born alive, 24 had surgical correction, 1 died shortly after birth (pulmonary atresia type of Fallot) and 1 (with associated malformation/VACTERL) received palliative care and died subsequently.
Subgroup 3b (AVSD) 22 patients in the ER, 18 had a chromosomal abnormality (15 T21, 3 T18), 2 were dysmorphic, 2 were normal. Prenatal diagnosis 19/22, TOP in 15 cases (15/15 chromosomal abnormalities), 7 born alive with subsequent surgical correction.
Group 4 (very mild abnormalities) 466 included in the Eurocat register of which only a small percentage (no TOP) was diagnosed, but which had no impact on the outcome.
Conclusion: The study shows that in the most severe group of congenital heart disease interruption of pregnancy reaches 86% in the prenatal diagnosed group.
In the transposition group the number of cases is too small to draw conclusions.

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P-72

Heart failure evaluation using the Cardiovascular Profile Score in fetal right heart defects

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Introduction: Congestive heart failure in fetuses with congenital heart defects may be assessed by the cardiovascular profile (CVP) score. The aim of the present study is to determine the variables predicting the risk of perinatal death in congenital right heart defects (RHD) to achieve a disease specific CVP score.
Methods: Retrospective analysis of medical records of 28 fetuses with RHD, evaluated at our Perinatal Clinic for a 5 year period. Logistic regression analyses were performed to obtain Odds Ratios (OR) for the relationship between risk of death and CVP score, cardiothoracic ratio, right ventricle pressure, aortic peak velocity, umbilical artery and middle cerebral artery pulsality index and parameters of LV performance.
Results: Fetal echocardiograms (143) from 28 patients were analyzed. Mortality was 50% by 30 days postnatally. The CVP score predicted the risk of death in fetuses with RHD, OR 0.9252(95%CI 0.866-0.988). Width of the RV/LV was lower in the non-survivors, OR 0.360(95%CI0.190-0.715). A lower pressure gradient of the TR jet, RV pressure and RV/LV pressure were associated with mortality, OR 0.384(95%CI0.228-0.646), 0.959(95%CI 0.940-0.978) and 0.395(95%CI 0.237-0.659), respectively. Peak aortic velocity was a protective factor, OR 0.104(95%CI 0.020-0.529).
Conclusion: Fetal echocardiography is useful in predicting outcome in RHD. CVP score is associated with the risk of perinatal death. Estimated RV pressure and LV ejection velocity may useful as part of right heart defect specific CVP score.

P-73

Abnormal cerebral and umbilical doppler flow values in the fetuses with hypoplastic left heart syndrome

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Introduction: Recent publications show that many abnormalities exist in peripheral circulation in fetuses with structural heart defect.
Goals: Evaluation of cerebral and umbilical arterial flow parameters in the fetuses with selected CHD
Methods: Retro- and prospective analysis of 127 fetuses (from 2007-2010) divided into two groups:
1. With left heart structural anomalies with reversal blood flow to ascending aorta (R), n = 67: R1- with left ventricle hypoplastic syndrome (HLHS), n = 58, R2-with critical aortic stenosis (AS) n = 12
2. Control group – without structural and functional defects (N), n = 50
Peak systolic velocity (PSV), pulsative and resist inexes (PI, RI) and S/D and CPR for MCA and UA were analyzed.
The statistical correction was used to make indexes independent from gestational age (ANCOVA test and Z score indexes). The multiple comparison LSD and post-hoc GT2 Hochberg analysis were used.
Results: There were significant differences between groups for the UAPI and UAPS. The UAPI and UAPS values were higher in the R group compared with the control group ($P<,001$ and $P=,006$ respectively). The higher values of UAPI compared to healthy fetuses were observed both in the R1 and R2 subgroups. ANCOVA test in the main group demonstrated significant differences between MCARI, MCAPI, MCAPS and MCAS/D, for the Z-score for all the parameters without MCAPI. The comparison in subgroups with ANCOVA showed that the RI and PI indexes were lowered in the fetuses with HLHS compared to healthy fetuses ($P<,001$ for MCARI and $=,029$ for MCAPI). This dependence was not observed in the fetuses with critical AS. Similar results for the Z-score for MCARI, MCAS/D were received. Z-score for MCARI and MCAS/D were the lowest in the HLHS subgroup. Z-score for CPR was lower for the R1 vs. control group ($P<,001$)
Conclusions:
1. Fetuses with congenital heart defects with retrograde flow in ascending aorta, mainly with LV hypoplasia presented changes in the arterial cerebral and umbilical flow patterns.
2. Autoregulative mechanisms in the fetuses with the most impaired cerebral flow are insufficient and abnormal indexes of middle cerebral artery flow could be an important marker of unfavorable changes in the central nervous system.

P-74

Aneurysm Of Left Atrial Appendix Diagnosed In The Intrauterine Period

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Introduction: Aneurysm of the left atrial appendage (LAAA) is caused by dilatation of left atrial apendage and mostly diagnosed incidentally in asymptomatic individuals. Most of the cases are congenital, The diagnosis is usually delayed until the second or third decade of life. Supraventricular tachycardia, systemic tromboembolic events, congestive heart failure may occur, and patients with such complications need intervention. Intra-uterin diagnosis is exceedingly rare. We herein present a case diagnosed during fetal echocardiography and operated successfully after birth.
Case: Twenty-one-year old pregnant lady was referred at 36th week of pregnancy. She was at her second pregnancy with a healthy living child. The reason for referral was the visualization of a cystic lesion within the pericardial space during routine obstetric ultrasonography. Fetal echocardiography revealed the aneurysmatic dilatation of the left atrial apendix causing a large cystic lesion 3×2 cm in diameter adjacent to the left atrium and left ventricle, compressing the left ventricle. Aneurysmatic sac was connected to the left atrial cavity anatomically and blood flow between the two cavities was observed by color Doppler echocardiography. There was no evidence of fetal heart failure. She gave birth to a boy weighing 3000 grams at 39th week. Postnatal echocardiography revealed the diagnosis of LAAA compressing the mitral valve and the left ventricle, mild regurgitation of the mitral valve was observed. At one month of age echocardiography showed enlargement of the aneurysm and spontaneous contrast echoes were observed within the appendage. Acetyl salicylic acid 3 mg/kg/day was started in order to prevent tromboembolic events. The aneurysm was resected successfully. During 6th month, the patient was asymptomatic,

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thriving normally, echocardiography showed mild dilation of the left atrium and left ventricle and mild mitral regurgitation.
In conclusion, LAAA is a rare abnormality that should be considered in differential diagnosis of cystic lesions adjescent to the left sided cardiac chambers. Close clinical follow-up and surgical intervention is required in order to prevent life threatening complications.

P-75

Prenatal Diagnosis of Total Anomalous Pulmonary Vein Connection: Our Institutional Experience.

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Introduction: Prenatal diagnosis of total (TAPVC) anomalous pulmonary venous connection in isolation or associated with other cardiac disease is rare and may reflect the difficulty in detecting them in utero.
Objective: To describe and evaluate the spectrum of fetal cardiac anomalous pulmonary venous connections and associated cardiac defects, at our tertiary referral center.
Methods: Retrospective review of 1934 fetal echocardiograms, performed between 2010-2011. All pregnancies with a fetal diagnosis of TAPVC, prenatal and postnatal echocardiograms and medical records were reviewed.
Results: We identified 8 fetuses with prenatal diagnosis of TAPVC. Mean gestational age at diagnosis was 29 weeks. 3 supracardiac, 3 intracardiac(coronary sinus) and 2 infracardiac type. Seven had an additional major cardiac defect, including 3 atrio-ventricular channel defects with associated right heterotaxy syndrome, one congenital corrected transposition of large vessels, one hypoplastic left ventricle, one double outlet right ventricle and one interventricular septal defect. Visualization of a dilated superior cava vein(2/3) and a vertical vein(1/3) in the supracardiac type, a dilated coronary sinus(3/3) in the intracardiac type and a vertical vein draining into the portal sinus(2/2), were the most consistent echocardiographic clues. In five cases, prenatal diagnosis was only made at follow-up assessment. 2 had altered karyotype, one prenatal diagnosis of Cat-Eye-Syndrome and one postnatal diagnosis of Edwards-Syndrome (parents refused to undergo amniocentesis). Diagnosis was confirmed postnatally in 7/8, one suspected supracardiac TAPVC had left pulmonary drainage obstruction confirmed by MRI. In five fetuses with TAPVC and obstruction, confirmed postnatally, continuous turbulent flow in the drainage site were demonstrated by Color-Doppler. Complete TAPVC surgical correction was performed in 7/8 with an average life of ten days. Premature death(<2months) occurred in 3 cases(2 Heterotaxy syndromes and Edwards-Syndrome).
Conclusion: Detailed assessment of the PV connection, both at initial examination and in serial fetal studies, is important for the prenatal diagnosis of TAPVC. The high morbidity and mortality were largely due to the presence of severe additional pathologies. In these patients fetal diagnosis did not significantly improve the prognosis, although it was critical for parental counseling and perinatal management.

P-76

New national program in prenatal aortic valvuloplasty – preliminary experience

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Objectives: Aortic balloon valvuloplasty in fetuses has already been accepted as a method of treatment in fetuses with evolving hypoplastic left heart syndrome (HLHS) or critical aortic stenosis with heart failure. There are still many questions concerning indications for prenatal and the best way of postnatal treatment. The aim of this study is to evaluate indications for prenatal valvuloplasty and outcome of fetuses in whom percutaneous balloon aortic valvuloplasty was performed.
Methods: 14 fetuses underwent prenatal treatment in two different institutions. 8 interventions was performed abroad until June 2011 and 6 in the national center for fetal therapy since then. All neonates were treated in four different national centers for pediatric cardiology and cardiac surgery.
Results: There was one intrauterine death related to the prenatal intervention and one due to placenta insufficiency (mother was heavy smoker). There were not complications related to the intrauterine intervention on the national basis. 11 neonates were born in 3 different centers, 4 by CC, 7 vaginal delivery, 1 still in utero. There was not difference in neonatal conditions born by CC or vaginal delivery. In 5 patients Norwood operations was performed, hybrid procedure in one, neonatal aortic balloon valvuloplasty in 5. Just in one aortic valvuloplasty was necessary in the first day of life. 1 patient died afer Norwood procedure due to sepsis, two after balloon aortic valvuloplasty, with heart failure and pulmonary hypertension. All other are in good general condition. Children after aortic valvuloplasty did not require aortic valve replacement.
Conclusions: Preliminary results of the new national program for fetal cardiac interventions are promising. Underlying anatomy is the main factor influenced the method of postnatal treatment. Detailed discussion is needed to develop the best method for treatment of neonates and infants with severe aortic stenosis after prenatal interventions.

P-77

The experience of being diagnosed with hypertrophic cardiomyopathy through family screening in childhood and adolescence

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Introduction: Hypertrophic cardiomyopathy (HCM) is a hereditary disease and the most common medical cause of sudden death in childhood and adolescence. This is the reason for recommending screening in children with an affected first degree relative. A diagnosis of HCM implies lifestyle modifications, restrictions that may bring profound changes to the daily life of the affected individual. The aim was to describe the experiences of children and adolescents at being screened positive for HCM and how this impacts on daily life.

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Methods: Descriptive qualitative interview study. Thirteen asymptomatic children or adolescents diagnosed with HCM through family screening were interviewed 12–24 months after the diagnosis. Analysis was conducted with qualitative content analysis.
Results: Children described an involuntary change, which affected their daily life with limitations and restrictions in life, both in the individual and social context. Life-style recommendations had the most severe impact on daily life and affected their social context. They tried to navigate in a world with new references and after reorientation they felt hope and had faith in the future.
Conclusions: Children diagnosed with HCM through family screening went through an involuntary change resulting in limitations and restrictions in life. This study indicates that there is a need for support and health professionals have to consider the specific needs in these families. Our findings thus give guidance in how best to improve support to the patients and their family. Diagnosis in asymptomatic children should be accompanied by ideally multiprofessional follow-up, focusing not only on medical issues.

P-78
Normalization of Haemostatic Alterations in Overweight Children with weight loss due to lifestyle intervention

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Background: Obesity has been shown to be associated with a hypercoagulable state. However, the effect of weight loss on these haemostatic alterations has not been studied yet with an overall function test such as the thrombin generating test (TG) in obese children.
Methods: We prospectively determined weight status as SDS-BMI, fibrinogen, and performed TG determining time to peak (TTPeak), peak, time preceding the thrombin burst (lag-time), and 'endogenous' thrombin potential (ETP) in 27 overweight children (mean age 11.9 +/- 2.4 years, 45% female, mean BMI 27.5 +/- 5.6 kg/m^2, mean SDS-BMI 2.31 +/- 0.48) both at baseline and after 1 year lifestyle intervention based on nutrition education, exercise therapy, and psychological care. Furthermore, thrombin generating test and fibrinogen were determined in 50 healthy children of the same age.
Results: Overweight children demonstrated significantly ($p = 0.013$) higher fibrinogen levels, shorter lag-time ($p < 0.001$) and TTPeak ($p = 0.028$) as compared to normal-weight children. Furthermore, ETP ($p < 0.001$) and peak ($p < 0.001$) were significantly higher in overweight children than in normal-weight children. The overweight children reduced their degree of overweight significantly (-0.45 +/- 0.22 SDS-BMI, $p < 0.001$). At the end of the lifestyle intervention, all haemostatic alterations normalized (significant decrease of fibrinogen ($p = 0.036$), ETP ($p = 0.034$), and peak ($p = 0.001$); significant increase of lag-time ($p = 0.040$) and TTPeak ($p < 0.001$).
Conclusions: Thee alterations in the haemostatic system in obese children normalized after weight loss due to lifestyle intervention demonstrating their reversibility.

P-79
Left ventricular in obese and overweight asymptomatic adolescents may be affected by higher arterial blood pressure, as compared with lean adolescents

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Background and Aim: Structural and functional cardiac changes are known in obese adults but respective disturbances in children and adolescents are still controversial. The aim of this study was to assess the relationship between arterial blood pressure, body mass index (BMI) and echocardiagraphic measurements in overweight and obese asymptomatic adolescents
Methods: This study included 93 healthy subjects aged 10-15 years (mean 12.6 ± 1.2 years). In all, systolic and diastolic blood pressure, weight, height, BMI, waist, hips, waist/hips ratio, hematologic and biochemical blood tests were assessed. Based on BMI, subjects were divided into three groups: lean (L, n = 32), overweight (Ov, n = 33) and obese (Ob, n = 32). In the same day, a complete transthoracic echocardiographic examination was performed.
Results: Interventricular septal and LV posterior wall end-diastolic thickness was increased parallel to the BMI (L: 0.84 ± 0.09 cm, Ov: 0.88 ± 0.1 cm, Ob: 0.96 ± 0.1 cm, $p < 0.001$, and L: 0.78 ± 0.1 cm, Ov: 0.80 ± 0.09 cm, Ob: 0.94 ± 0.08 cm, $p < 0.001$, respectively). Relative wall thickness (RWT) and LV mass index (LVMI) adjusted to body surface area were similarly increased (L: 0.34 ± 0.05, Ov: 0.34 ± 0.05, Ob: 0.40 ± 0.04, $p < 0.001$, and L: 78 ± 13 g/m2, Ov: 77.6 ± 13 g/m2, Ob: 86.9 ± 16.3 g/m2, $p = 0.022$, respectively). Systolic blood pressure (SBP) values were significantly different (L: 107.5 ± 7.5 mmHg, Ov: 115 ± 11 mmHg, Ob: 118 ± 12 mmHg, $p < 0.001$), whereas diastolic blood pressure (DBP) values were not significant (L: 74 ± 6 mmHg, Ov: 76 ± 7 mmHg, Ob: 74 ± 9 mmHg, $p = 0.428$) between groups. SBP correlated with BMI ($r = 0.282$, $p = 0.006$) and with LV posterior wall thickness ($r = 0.243$, $p = 0.019$), whereas DBP correlate with LV ejection fraction ($r = -0.227$, $p = 0.029$), with deceleration time of E wave ($r = -0.223$, $p = 0.032$), with LV lateral early diastolic velocity ($r = 0.215$, $p = 0.039$).
Conclusion: The systolic blood pressure is higher in obese and overweight adolescent compared with lean adolescents. Both, high blood pressure and obesity, may affect left ventricular systolic and diastolic function in these patients.
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P-80
Tricuspid annular peak systolic velocity (TAPSV) in children and young adults with pulmonary artery hypertension secondary to congenital heart diseases and tetralogy of Fallot: comparison with MRI data

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The tricuspid annular peak systolic velocity (TAPSV), as echocardiographic index to assess right ventricular (RV) systolic

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function, has not been investigated thoroughly in children and young adults with tetralogy of Fallot (TOF) and pulmonary artery hypertension secondary to congenital heart disease (PAH-CHD). TAPSV values of patients with TOF (n = 185) and PAH-CHD (n = 55) were compared to age-matched normal subjects. TAPSV values were compared to RVEF and RVEDVi determined by MRI. TAPSV values become significantly reduced after an age of 10.4 years in PAH patients, and after an age of 13.6 years in TOF patients when compared to the lower bound of the ± 2 SD interval of normal subjects. A significant positive correlation between TAPSV with RVEF was seen in both, TOF ($r = 0.66$, $p < 0.001$) and PAH-CHD ($r = 0.82$, $p < 0.001$) patients. A significant negative correlation between TAPSV with RVEDVi was also seen in TOF ($r = -0.29$, $p = 0.002$) as well as in PAH-CHD patients ($r = -0.59$, $p < 0.001$). Although initially preserved, we found impaired TAPSV values with increasing age in patients with TOF and PAH-CHD. This indicates that persistent pressure overload in PAH-CHD patients as well as volume overload in TOF patients can lead to an impairment of systolic RV function and increased RVEDVi. The validity of TAPSV data could be confirmed by MRI data (RVEDVi and RVEF).

P-81
Interleukin-21 Receptor Gene Polymorphisms in Kawasaki disease

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Introduction: It has been known that interleukin-21 receptor (IL-21R) gene polymorphism is related with the development of systemic vasculitis. We explored the polymorphisms of IL-21R gene in patients with Kawasaki disease (KD).
Materials and methods: We genotyped the promoter region of IL-21R gene (-2500 bp to +1 bp) in 100 patients with KD and 100 healthy controls who are all Koreans.
Results: We found the 5 single nucleotide polymophisms (SNPs) of which minor allele frequency > 0.01 in the promoter region of IL-21R gene. Those are -1681 G > T (chromosome site 27411802), -380 G > A (27413104), -332 G > C (27413151), -237 A > T (27413246), and -53 G > A (27413430). There is no significant difference in minor allele frequency of each SNP between patients with KD and healthy controls except -237 A > T. Twenty five patients with KD had more than 1 SNP while 7 healthy controls had. There were more individuals who have any SNPs of IL-21R gene in patients with KD than the controls (odds ratio: 3.0, 95% CI: 1.6-5.6, $P = 0.0005$). There was no significant correlation between IL-21R gene polymophisms and the serum level of IL-21 in patient with KD. The serum level of total IgE was not affected by the existence of IL-21R gene polymorphisms in this study.
Conclusion: Our data suggest that the genetic susceptibility profile for KD may include IL-21R gene.

P-82
Chronic hypoxemia leads to reduced serum IGF-I levels in cyanotic congenital heart disease

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Cyanotic congenital heart disease in children commonly causes more pronounced growth retardation in comparison with acyanotic congenital heart disease. Chronic hypoxemia has been suggested as the cause of poor growth in these patients, but the relationship between serum IGF-I levels and chronic hypoxemia is unclear.Some studies shows Serum IGF-I concentrations, oxygen saturation and nutritional status were evaluated in patients with cyanotic congenital heart disease, and serum IGF-I levels were compared with a group of 20 well-nourished, age-matched control children to assess the relationship between IGF-I levels and chronic hypoxemia. The nutritional status of each patient was determined by using anthropometric parameters and calorie and protein intake ratios. The patients were divided into malnourished and well-nourished groups (21 and 8 patients, respectively) according to their nutritional status. Serum IGF-I concentrations were measured in the two patient groups and the controls. The malnourished group had the lowest IGF-I levels (48.14 +/- 21.8 ng/ml, $p < 0.05$). However, the well-nourished group's IGF-I levels were significantly lower than the control subjects' despite improved nutritional status (85.5 +/- 30.2 and 107 +/- 19.7 ng/ml, respectively, $p < 0.05$). In addition, they found a positive correlation between serum IGF-I levels and oxygen saturation of the patients ($r = 0.402$, $p < 0.05$). These findings indicate that chronic hypoxemia has a direct or indirect effect to reduce serum IGF-I concentrations and this may be a cause of the increased growth failure in patients with cyanotic congenital heart disease. chronic hypoxia plays a significant role in the pathogenesis of malnutrition and also believe that IGF-1 deficiency seen in CHD patients may be responsible in the etiology of the decrease in left ventricular mass independently from GH.

P-83
Mildly functional decline of left ventricle is detected by systolic time intervals in pediatric patients with chronic peritoneal dialysis

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Background: Cardiac depression often appears in adults with hemodialysis for chronic renal failure. Chronic peritoneal dialysis (CPD) is selected in pediatric patients with chronic renal failure. There are few reports to mention systolic time intervals (STI) in pediatric patients with CPD. We investigated cardiac functions by measurement of STI in pediatric patients with CPD.
Methods: The medical records of 12 patients with CPD were reviewed. Patients ranged in age from 1 year to 14 years (median 4 years). We used 12 healthy children as control who were matched sex and age. Echocardiographic data of left ventricle were compared between the two groups. On STI the ratio of preejection period to ejection time (PEP/ET) and isovolmetric contraction time-ET ratio (ICT/ET) were employed as index of systolic function. The ratio of isovolume relaxation time to ET (IRT/ET) was employed as diastolic function.
Results: Initiation age of CPD was between 4 days and 14.7 years (median 6 months). Duration of CPD was between 6 months and 4.2 years (meadian 2.3 years). The values of PEP/ET were higher in patients with CPD than those in controls (0.29 vs 0.24, $p = 0.0035$). There were no differences in ICT/ET. The values of IRT/ET were higher in patients with CPD than those in controls (0.15 vs 0.083, $p = 0.023$). The dimension of left ventricle, ejection fraction, the ratio of E wave to A and deceleration time of E wave were not different. In patients with prolong PEP/ET the levels of creatinine were elevated (6.8 mg/dl vs 3.3 mg/dl, $p = 0.027$); those of phosphate elevated (5.9 mg/dl vs 4.2 mg/dl, $p = 0.027$).
Conclusions: We showed abnormality of left ventricular function by STI in pediatric patients with CPD. Contrastingly left ventricular dysfunctions were not detected by M-mode and

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pulsed Doppler method. Myocardial contractility was mainly measured by STI. Even if myocardial contractility disturbed mildly, pump function was commonly reserved. Maybe we see petty dysfunctions of ventricle by STI. In pediatric patients with chronic failure dialysis mostly extends over a long period. We should follow up cardiac functions by systolic time intervals in pediatric patients with CPD, even if cardiac functions look normal by routine echocardiogram.

P-84
Mild extension of left ventricle engages with elevation of brain natriuretic peptide in patients with atrial septal defect

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Background: When patients with atrial septal defects (ASD) are candidate for surgery, dimension of LV becomes small. The levels of BNP sometimes increase in response to left ventricular dysfunction early after surgery of ASD. We predicted BNP was also excreted from small LV before repair. The present study investigated whether LV influenced the elevation of BNP in patients with ASD.
Methods: The medical records of 87 patients with ASD were reviewed. They underwent cardiac catheterization with operation in view. Blood test and chest X-p were performed at the same point in time. Levels of BNP ranged from 3.9 to 313pg/ml. The patients with levels of BNP in the top quartile (BNP$\geqslant$55.6pg/ml) were defined as the group of BNP elevation. Relation BNP elevation and factors were assessed which were obtained from catheterization and radiograph.
Results: Patients ranged in age from 5 months to 17 years (mean 4.1years). In the univariate analysis two factors were related to BNP elevation significantly on left-sided heart: expansion of end-diastolic volume of LV (LVEDV); increased cardiac index. Four factors were related to BNP elevation on right-sided: extension of EDV of right ventricle (RVEDV); extension of end-systolic volume of RV; elevated end-diastolic pressure of RV (RVEDP); increased cardio-thoracic ratio (CTR). Multiple logistic regression analysis revealed that four factors had independent relationship with high levels of BNP: mild enlargement of LVEDV ($\geqslant$107%); increased RVEDV ($\geqslant$280%); elevation of RVEDP ($\geqslant$10 mmHg) ; enlargement of CTR (>60%).These four factors explained 63% of high levels of BNP.
Conclusion: Same as previous reports burden to right ventricle were associated with BNP elevation in patients with ASD. Our study also showed mild expansion of LV was related to high-elevated BNP before operation. There were no reports that BNP levels got elevated preoperatively in patients with ASD. Small left ventricle of ASD might be weak against little load. We should attend left ventricular dysfunction in ASD patients with elevated BNP, if right-sided load was not so intensive.

P-85
Usability of QTc Dispersion for the Prediction of Orthostatic Intolerance Syndromes

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Introductions: Syncope is defined as transient loss of consciousness and muscle tone, usually of short duration. Noncardiac causes of syncope are classified as orthostatic intolerance syndromes (OIS). QT and QTc (corrected QT) dispersions are the measurements of myocardial instability and show predisposition to arrhythmias. In this study; clinical findings, QT and QTc dispersions of the patients who were diagnosed as OIS were evaluated retrospectively. Also, the aim of the study is to clarify the association of clinical characteristics of unexplained syncope with the outcome of the QT and QTc dispersions in children.
Methods: We designed a retrospective study including 152 children and adolescents who had repeated unexplained syncope or presyncope between June 2002-August 2010. Head-up Tilt table test (HUTT) were performed for all patients. Control group consisted of 67 healthy children. The QT and QTc dispersions were measured from the 12 ECG leads.
Results: 84 (55.2%) patients had positive and 68 (44.8%) had negative response to HUTT. QT and QTc dispersions were significantly higher in HUTT positive patients than in negative ($p < 0.01$, $p < 0.001$ respectively). Also, QTc dispersion was significantly higher in both vasovagal syncope and postural orthostatic tachycardia syndrome groups than in HUTT negative group ($p < 0.001$, $p < 0.05$ respectively). Specifity and sensitivity of QTc dispersion longer than 50 ms for predicting positive HUTT are 76.5% and 59.5% respectively. The positive predictive value of the test calculated as 75.8%.
Conclusions: These results revealed that we can use QTc dispersion measurement as a noninvasive electrocardiographic test to evaluate OIS for predicting positive result before performing HUTT.
Keywords: Syncope; ortostatic intolerance syndrome; QTc dispersion; head-up tilt table test; children

P-86
Pulmonary arterial hypertension in congenital heart anomalies with left-right shunt

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Introduction: Pediatric cardiology is mainly engaged in congenital heart anomalies (CHA), of which two thirds are anomalies with left-right (LR) shunt. Pulmonary arterial hypertension (PAH) present a secondary increase in pulmonary arterial pressure at rest > 25 mmHg or > 30 mmHg during loading, during the catheterization. In clinical practice elevation of pulmonary vascular resistance (PVR) leading to irreversible changes in pulmonary vasculature that are contraindications for operative treatment.
Aim: to evaluate incidence of the pulmonary arterial hypertension in congenital heart anomalies with left-right shunt
Methods: selective group patient with PAH based on clinical examination, ECG, X-ray, 2D echo and final diagnostical cathteterisation were divided in two groups : fixed and nonfixed PAH
Results: In the period between april 1997. and november 2011 was performed a total of 216 pediatric cardiac catheterization, (44% of all surgical–489 patients), 169 (79%) diagnostic, 47 (21%) of interventional catheterization. In 45 (26%) patients was to estimate PAH (girls 25). In 15 (33%) patients we have fixed (irreversible) PAH. From that group in 9 patients (60%) PAH was associated with Down syndrome. 30 (66%) patients after cardiac catheterization were with non-fixed (reversible) PAH is not operated on four children (13%). At this point, 15 patients with PAH is evaluating and treating.
Conclusion: Assessment of PAH is imperative for operative correction and inoperable patients require specific treatment prostanoids, endotelin receptor antagonists and inhibitors of phosphodiesterase type-5, alone or in combination.
Key words: pulmonary arterial hypertension, congenital heart anomalies, treatment.

P-87

Left ventricular function, natriuretic peptide type B and troponin T levels at onset of diabetes type I in children (a pilot study)

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The incidence of diabetes type I (DB1) have increased dramatically in the last decade in the Nordic countries, especially in the age group 5-9 years. In this age group, insulin resistance may be influenced by factors such as obesity, low grade of physical activity and infections. In animal models, Ljungan virus has been reported to be associated with DB1 and myocarditis. Therefore, one may speculate that there may be an association between onset of DB1 and myocarditis.
Aims: To evaluate left ventricular function, natriuretic peptide type B (BNP) values and potential cardio-myocyt damage at the time of onset of diabetes type I in children.
Methods: Left ventricular function was evaluated by echocardiography, and blood samples for analysis of BNP in plasma, S-troponin T (TnT), S-creatine kinase-MB (CKMB) and S-creatinine were taken consecutively, within 48 hours after admission to hospital at the time of the primary diagnosis of diabetes type I in children.
The reference interval for P-BNP was 0-18.4 ng/L, for CKMB <5 mg/L and TnT <0.03 mg/L.
Results: Ten children, 6 boys and 4 girls, with a median age of 8.9 years, range 4.4–13.3 years, with primary onset of diabetes type I were recruited to the study. Left ventricular dimensions and ventricular function were within the normal range in the vast majority of the children: left ventricular inner diameter in diastole Z-score, median 0.76 (0.28-1.92) intra-ventricular septum in diastole Z-score, 0.85 (0.26-2.63), left ventricular posterior wall Z-score, –0.15 (-1.69-2.15) left ventricular mass Z-score, 1.33 (-0.72-2.73) and fractional shortening, 36% (31-39). E/A ratio, median 1.6 (1.2-1.9), deceleration time 119 msec (75-168) and iso-volumetric relaxation time, 60 msec (43-69) indicated essentially normal diastolic function.
P-BNP was within the normal range in all the children but one, median 8.1 ng/L (3.2-25.8) and markers for myocytolysis and S-creatinine were normal in all, CKMB 3.0 mg/L (2.0-3.0), TnT mg/L <0.01 and S-creatinine 34 mmol/L (25-52). None of the children had pericardial effusion or any pathological valve regurgitation.
Conclusion: Left ventricular function, BNP and TnT values are essentially normal in children at the time of onset of DB1, indicating the freedom of myocarditis.

P-88

The Evaluation of the Frequency and Echocardiographic Features of Cardiovascular Anomalies Among the Families of the Patients with Bicuspid Aortic Valve or Other Left-sided Cardiovascular Anomalies

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Objective: To evaluate the frequency of the cardiovascular anomalies among the families of the patients with bicuspid aortic valve (BAV) or other left-sided cardiovascular anomalies (LSCAs) and the echocardiographic features of these anomalies.
Methods: The patients, evaluated with BAV or any LSCA (aortic stenosis without BAV, coarctation of the aorta (CoA), interrupted aortic arch, hypoplastic left heart syndrome, and isolated dilatation of ascending aorta) in our clinic between October 2010 and August 2011 and their first degree relatives were enrolled into the study. The patients with any known genetic abnormality were excluded. All the participants underwent an echocardiographic examination. The relatives of every individual with newly diagnosed cardiovascular anomaly were also sequentially included. The patients were assessed in three groups: BAV, BAV+CoA, and other LSCAs. The relatives were grouped and evaluated according to the diagnosis of the proband in their families. The echocardiographic measurements of the patients and their siblings were standardized by computing Z-scores.
Results: The numbers of the patients in BAV, BAV+CoA, and other LSCAs group were 52, 14, and 20 respectively. Any LSCA was determined in 19 of the 263 relatives (7.2%) of the patients with BAV or LSCAs. Fourteen of those (5.3%) had aortic dilatation and five (1.9%) had BAV. A second individual with a left-sided cardiovascular anomaly was observed in 11 of the 86 (12.8%) families investigated. Most of the 66 patients with BAV (81.8%) were male and 14 out of these 66 patients (21.2%) had CoA as an associated anomaly. The frequencies of aortic stenosis (AS), aortic regurgitation (AR), AS+AR, and aortic dilatation in the patients with BAV were found 37.9%, 53%, 25.8% and 48.5% respectively. The aortic measurements of the patients with BAV were larger than the healthy siblings' values. The valvular dysfunction and aortic dilatation were more commonly observed in the patients who had right coronary and noncoronary leaflet fusion.
Conclusion: Almost all the complications of BAV and other LSCAs are preventable. Because the clustering of LSCAs in some families is observed, we recommend echocardiographic screening of those relatives. If not possible, at least it should be achieved for BAV.

P-89

Nowadays, Children With Congenital Heart Disease are not Limited in Their Submaximal Exercise Performance

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Objective: Formerly, adolescents and adults with congenital heart disease (CHD) showed a reduced exercise capacity even in defects considered to be simple. Nowadays, the children might get a better medical management and less restriction concerning an active lifestyle or sport activities. The exercise performance of this new generation of children with CHD has to be evaluated.
Patients and Methods: In the year 2010, eighty-eight children (12.7 years, 52 male) eleven to fourteen years old with various congenital heart diseases performed a cardiopulmonary exercise test in our institution. These children were matched for age and gender with healthy subjects, who underwent the same procedure at a school survey.
Results: In comparison to healthy controls, children with CHD had a diminished peak oxygen uptake (CHD: 35.5 ml/min/kg vs. controls: 42.4 ml/min/kg; $p < .001$) corresponding to 87.1% (CHD) and 99.5% (Controls) of the reference value, respectively. Peak oxygen uptake decreased with the severity of the heart defect ($r = -.410$; $p < .001$). However, there were no differences in oxygen uptake at the ventilatory threshold (CHD: 20.6 ml/min/kg vs. controls: 21.5 ml/min/kg; $p = .675$).
Conclusions: Nowadays, children with CHD are not limited in their submaximal exercise performance. However, there is still a reduction in peak oxygen uptake.

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P-90

Pulmonary hypertension in scimitar syndrome : a series of 90 consecutive patients from a single centre

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Background: Scimitar syndrome (Scim.) combines congenital cardiopulmonary anomalies with anomalous drainage of one or more of the right pulmonary veins to the inferior caval vein. Pulmonary hypertension (PH) is a common finding but its causes are poorly understood.
Objective: To analyse the constellation of anatomic anomalies and their potential relation with PH and outcome in Scim.
Methods: We reviewed 90 consecutive cases of Scim for presenting symptoms, cardiac phenotype, extracardiac anomalies, surgical procedures and outcome. We also reviewed right heart catheterisation (RHC) for pulmonary hypertension (PH) when available.
Results: We identified 90 cases (53 females, 37 males) with Scim. Diagnosis was done in fetus in 10 pts, at birth in 25 pts, before 1 year of age in 34 pts, and after 1 year of age in 21 pts. 48/90 had an associated cardiac defect. The abnormal pulmonary venous return of the right lung was complete in 65 pts. Associated anomalies of systemic veins were present in 13 pts. Congenital stenosis of the scimitar vein was found in 10 pts and aberrant drainage in 2 pts. Systemic arterial supply to the right lung was present in 75 and considered significant in 60 pts. Extracardiac malformations were present in 21 pts. 73 RHC had been performed (the remaining 17 pts had normal estimation of pulmonary pressures on echo): 51 pts had PH at time of diagnosis. PH had different causes : PPHN in 17 neonates, PH due to massive overflow by the systemic supply in 10 pts; PH due to associated CHD in 7; postcapillary PH in 4; respiratory disease in 3; and finally, pulmonary arterial hypertension was observed in 26 pts with or without associated CHD. There were 24 deaths (18 neonates) that were directly related to refractory PH/cardiac failure in 9 pts, and to severe respiratory disease in 4.
Conclusion: Surgical outcome of scimitar syndrome has been described favourable but this certainly due to the fact that only survivors and children without PAH undergo surgery. In our series, mortality was high and beside associated CHD, the management of PH of multifactorial origin is a remaining challenge.

P-91

Respiratory outcome in children with scimitar syndrome

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Background: Scimitar syndrome is a rare congenital association of an abnormal venous return of the right lung into the inferior vena cava, and of various heart and lung abnormalities, including a virtually constant right lung hypoplasia.
Objectives and methods: Our aim was to evaluate respiratory morbidities and lung function tests in the cohort of patients evaluated at our center since 1976.
Data related to the hospital course and to the follow-up controls of patients, as well as most recent lung function tests were collected.
Results: Eighty-one children were investigated. Twenty-six patients died, all with the infantile form. The median duration of follow-up of surviving children was 5.9 years. A high rate of respiratory morbidities was measured, with 38% and 43% of children reporting pulmonary infections or wheezing episodes during the last 12 months of follow-up, respectively. One third of children have been re-hospitalized for a respiratory cause. Lung function tests were obtained in 19 children. The median value of total lung capacity (TLC) was 76.0% of the predicted value (IQR:67.5;89.0), and the median value of the ratio of the forced expiratory volume in one second to the forced vital capacity (FEV1/FVC) was -1.06 Z score (-2.13;0.14). Significantly lower TLC values were obtained in children with the infantile form ($p = 0.008$), or with a history of thoracic surgery ($p = 0.003$). FEV1/FVC Z score values were significantly lower in boys ($p = 0.05$), and in children with a history of wheezing ($p < 0.05$). Wheezing episodes were not associated to significant salbutamol-induced reversibility.
Conclusion: Respiratory complications are frequently observed in children with scimitar syndrome. Pulmonary hypoplasia appears as an independent marker of long term severity in these patients.

P-92

Are there any associations between the cardiac septal defects and ROCK2 gene polymorphisms at the childhood? : A case-control study

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Background: Rho/Rho-kinase pathway plays a critical role in the regulation of cellular functions such as proliferation and migration. One of possible theories of the development of ventricular septal defects is cell migration disorder. The aim of this study was to analyze the genotype distributions and allele frequencies for ROCK2 gene Thr431Asn, Asp601Val, and Lys1083Met polymorphisms among the cardiac septal defects patients in a Turkish population.
Methods: In this case-control study, 300 patients with cardiac defects (150 patients with ventricular and 150 patients' atrial septal defects) and control group (150 healthy control subjects) were investigated. A single-nucleotide polymorphism in ROCK2 gene Thr431Asn was analyzed by real-time polymerase chain reaction using a Light-Cycler, and Asp601Val, and Lys1083Met polymorphisms was detected by restriction fragment length polymorphism polymerase chain reaction.
Results: Neither genotype distributions nor the allele frequencies for the Thr431Asn, Asp601Val, and Lys1083Met polymorphisms showed a significant difference between the groups.
Conclusion: These results suggest that there were no evidence for an association of the ROCK2 gene Thr431Asn, Asp601Val, Lys1083Met polymorphisms with cardiac septal defects in pediatric patients

P-93

Could be useful to measurement short-term analysis of heart rate variability to access autonomic function in obese children and its relationship with metabolic syndrome?

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Cardiovascular autonomic nerve system in obese children is one of the main initiators of coronary heart disease and hypertension development and also may have close relationship with insulin resistance. Heart rate variability is one of the non-invasive methods for assessment of cardiovascular autonomic system. In this method, low frequency parameters reflect sympathovagal activity, high frequency reflects vagal activity and their ratio reflects sympathovagal balance.
In this study, short term analysis of heart rate variability was investigated in 66 obese children and 40 healthy controls. While high frequency parameter values are low in the obese compared to controls (16.02 ± 12.9 nu vs 21.45 ± 13.6 nu, $p = 0.046$), low frequency/high frequency ratio was found significantly high (3.79 ± 2.34 vs 2.25 ± 0.93, $p < 0.001$), a significant difference was not detected in low frequency values ($p = 0.787$). Insulin resistance was found in 33 (50%) patients, dyslipidemia was found in 39 (59%) and hypertension was found in 18 (27%). Metabolic syndrome in obese group was detected in 39.4% patients.
Conclusion: As the result of our study, we found that vagal activity decreased in obesity and autonomic nervous system balance was impaired in favor of sympathetic activity at the short term heart rate analysis. We put forward that metabolic syndrome frequency is too higher in obese children.

P-94
Determinant of increased type B natriuretic peptide (BNP) in acute phase of Kawasaki disease

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Background: The aim of this study was to identify the determinant of increased type B natriuretic peptide (BNP) in patients with acute Kawasaki disease (KD).
Methods: Subjects were 56 patients with acute KD and 21 febrile controls (FC) and they underwent multi-modal echocardiographic studies to determine functional parameters including LVEF, LVDd transformed into z-value (zLVDd), LV volume indexed to body surface area (LVVI), Tei-index, and E/e'. In addition to white blood cell count (WBC) and C-reactive protein (CRP), inflammatory cytokines including interleukin (IL)-1ß, IL-6, IL-8, IL-10, IL12p70, and tumor necrosis factor were determined in 47 of KD and 10 of FC. Echocardiographic and laboratory data were compared between KD and FC. Also, correlations between Log (BNP) and these data were determined.
Results: There was no significant difference in age, days of illness at study, WBC, CRP, but log (BNP) in KD was significantly higher than that in FC (1.58 ± 0.64 vs. 1.01 ± 0.44, $p = 0.0003$). Though, zLVDd (0.81 ± 1.00 vs. 0.22 ± 0.74), LVVI (58 ± 22 vs. 46 ± 12 ml/M2, $p < 0.03$), E/e' (9.6 ± 2.3 vs. 8.4 ± 1.6, $p < 0.05$) in KD were significantly higher than those in FC, there was no significant difference in LVEF and LV Tei-index. Among the inflammatory cytokines, only IL-6 in KD was significantly higher than that in FC (233 ± 241 vs. 46 ± 37 pg/ml, $p < 0.02$). Among the parameters that correlated with Log (BNP) including zLVDd ($r = 0.35$, $p < 0.005$), LVVI ($r = 0.33$, $p < 0.004$), LVEF ($r = -0.27$, $p < 0.002$), E/e' ($r = 0.30$, $p < 0.03$), WBC ($r = 0.38$, $p < 0.01$), CRP ($r = 0.51$, $p = 0.0003$), and IL-6 ($r = 0.76$, $p < 0.0001$), multiple stepwise regression analysis identified IL-6 as the single most significant predictor of Log (BNP) ($\beta = 0.77$, $p < 0.0001$).
Conclusion: In patients with acute Kawasaki disease, BNP is significantly increased with mildly dilated left ventricles and this increased BNP is associated with not only left ventricular systolic dysfunction but also inflammation itself represented by increased IL-6.

P-95
Ebstein' s anomaly: fetal presentation and neonatal outcome

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Objective: To identify predictors of poor outcome in neonates with Ebstein's anomaly (EB).
Background: EB is a rare congenital heart disease with variable neonatal presentation and outcome.
Patients and Methods: From January 2004 to January 2011, 16 newborns, 11 with a prenatal diagnosis, were referred to our centre.
Results: Among foetuses with EB 2 had a severe hydrops, 1 a tachyarrhythmia, 9 a cardio-thoracic ratio >0.6, 4 a reversed flow through the ductus arteriosus. At birth 14 patients had moderate to severe EB; 4 had congestive heart failure, 9 duct-dependent pulmonary circulation and needed PGE1 infusion, which restored an adequate pulmonary flow in 5 children.
3 patients with restrictive PFO and metabolic acidosis underwent an atrioseptostomy.
4 patients needed a BT shunt associated with plasty of the right atrium or closure of the tricuspid valve. There were 4 deaths, all occurred in infants with a prenatal diagnosis: 1 at birth, 2 postoperative and 1 sudden death 53 days after surgery. Patients with prenatal heart failure displayed a positive evolution under PGE1 and did not need any intervention.
At a median follow-up of 2 years, 2 patients had a Glenn palliation, the remaining 10 patients are symptoms free. The positive outcome was not related to the degree of tricuspid regurgitation, which showed the tendency to decrease with time, but with the area of functional right ventricle.
Conclusions: EB is a severe cardiac disease when it presents during the fetal or the neonatal period. In this small series fetal hydrops was not related with perinatal exitus. Mortality was high and the outcome, not influenced by prenatal diagnosis, was related to the presence of functional right ventricle. All babies who survived after the neonatal period had a good outcome at medium term follow-up.

P-96
Smart nature. Aortico-left ventricular (Ao-LV) tunnel bypassing congenital critical aortic stenosis

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Case report: A 3 day old neonate was transferred to our unit because of a heart murmur detected on routine examination before discharge from nursery. Apart from a systolic-diastolic murmur grade 4/6 the child was completely asymptomatic with normal vital signs, blood gases and without elevated lactate.
Echocardiography revealed a thickened, dysplastic and severely stenotic aortic valve with a Doppler derived peak gradient of 70 mmHg. The Ao-LV tunnel consisted of an aneurysmatic extracardiac component measuring 1,3×1,8 cm. The aortic orifice of the tunnel was 3 mm in diameter entering the Aorta just above the right coronary sinus. The ascending aorta was significantly dilated. The left ventricle was mildly dilated with increased apical trabeculations but without endocardial fibroelastosis. Based on the echocardiographic findings the child underwent successful Ross operation on the 9th day of life and was discharged 14 days thereafter.

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At that time echocardiographic findings showed normal LV function, no aortic stenosis with trace regurgitation.
Discussion: Ao-LV tunnel is a rare anomaly with an unknown etiology. In a substantial number it is associated with aortic valve anomalies including severe aortic stenosis. The few caess described in the literature presented with critical hemodynamic status calling for immediate action. Our case however was hemodynamically stable with preserved LV function due to the Ao-LV tunnel that served as a natural bypass. The aortic orifice of the tunnel was big enough to maintain cardiac output, but restrictive enough to limit regurgitation. Thus we could postpone surgery to the ninth day. To the best of our knowledge this is the first case of Ao-LV tunnel and critical aortic stenosis that did undergo neonatal Ross procedure.

P-97

Evaluation of the incidence and echocardiographic findings of congenital heart diseases in children with Down Syndrome in the Middle Anatolia of Turkey

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Objectives: Down syndrome is the most common chromosomal disorder in all countries and congenital cardiac disease is the greatest cause of mortality and morbidity in these patients. In the recent study, incidence and types of congenital heart diseases of children with Down syndrome were evaluated in the Middle Anatolia of Turkey.
Methods: Data of 239 patients diagnosed as Down syndrome who were consulted to our pediatric cardiology department between March 2007 and April 2011 were retrospectively reviewed. History, physical examination, echocardiographic and cardiac catheterization findings of all patients were listed. Chromosome analysis was performed in our genetic laboratory.
Results: Congenital heart diseases were detected in 86 (% 36) of 239 patients with Down syndrome. Congenital heart diseases were detected in 48 (%33.8) of 142 patients who were admitted during neonatal period while this ratio was % 39.2 (38/97) after neonatal period ($p = 0.041$). Evaluation of all single or multiple cardiac defects revealed that endocardial cushion defect (% 37.2) was the most common congenital heart disease. During the study period only two patients died due to the cardiac operation. These patients were diagnosed as complete endocardial cushion defect in older ages and cardiac operation was performed with high risk.
Conclusions: In our study the prevalence of congenital heart diseases in neonates with Down syndrome was lower than the current literature. To determine the real prevalence of congenital heart diseases in neonates with Down syndrome, prospective and multicenter studies were needed in our country. Appropriate-time-planned cardiac operations will decrease the high incidence of mortality and morbidity in Down syndrome with large ventricular septal defect or endocardial cushion defect.
Key words: Down Syndrome, Infant, Newborn, Congenital Heart Defects.

P-98

Dilatation of aortic root, arachnodactyly, and arthrogryposis in a newborn as early manifestation of a Marfan syndrome with a new splice acceptor mutation

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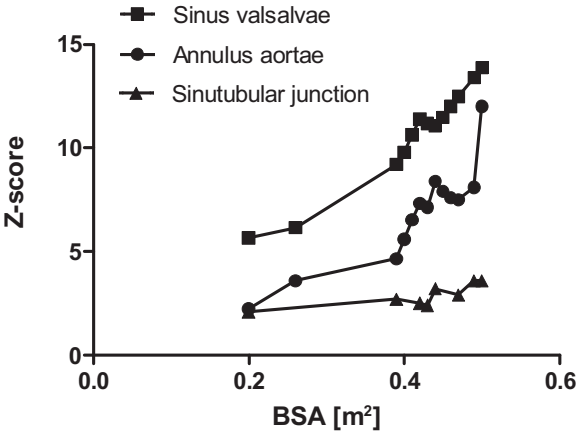
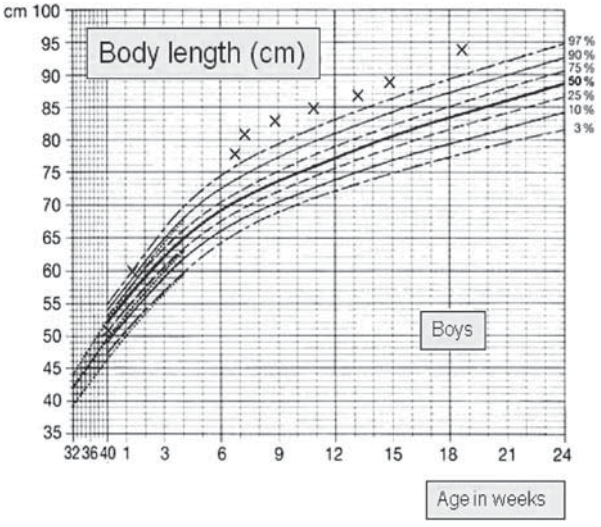
Introduction: Marfan syndrome is an autosomal dominant connective tissue disorder. The diagnosis relies on defined revised criteria (1, Ghent nosology, 2010) and is possible in over 95% due to detection of causative mutations in the fibrillin-1 gene. The revised criteria emphasize the cardiovascular manifestations and the ectopia lentis for diagnosis. The systemic manifestations are comprised in a new scoring system. However, the clinical phenotype is highly variable and the most severe forms get symptomatic in the neonatal period and early infancy. Therapy is medical and under certain conditions surgical. Prognosis depends on the progression of aortic root dilatation and consecutive aortic dissection or rupture.
Case: A term male newborn presented with a dilated aortic root (Z-score +5), arachnodactyly, joint contractures and facial progeria. In addition, a severe myopia (-17 dioptre) and a Morgagni hernia were detected. The score for systemic manifestation was nine. Molecular analysis showed a new heterozygous splice acceptor mutation in the fibrillin-1 gene (c.3965-1G > C). In the further course the patient developed a progressive dilatation of the aortic root in spite of medical therapy (ß-blocker and AT-1 antagonist), a severe thoracolumbar kyphosis and acceleration of the growth of his body length (see figures).





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Discussion: The relevance of the mutation, the differential diagnosis in respect to connective tissue disorders considering the revised Ghent criteria, therapeutic interventions and the further course and prognosis are discussed.
Summary: Early diagnosis by detecting a causative mutation in the fibrillin-1 gene is important for exclusion of other diagnosis, optimal treatment and genetic counseling of the families.
[1] *Loeys BL et al. (2010) The revised Ghent nosology for the Marfan syndrome. J Med Genet 47:476-85*

P-99

Right ventricular distress influences the elevation of brain natriuretic peptide in symptomatic infants with ventricular septal defects

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Background: In symptomatic infants with ventricular septal defects (VSD) volume overload for left ventricle occurs. The values of brain natriuretic peptide (BNP) uprise to high levels occasionally. In severe cases cardiac decompensation occurs. We attempted to identify the clinical characteristics in symptomatic infants with VSD who had high levels of BNP.
Methods: Between 2005 and 2011, consecutive 96 symptomatic infants with VSD were studied. Cardiac catheterization was performed with surgical intervention in view. Venous blood samples for analysis of BNP were obtained within a week before catheterization. We defined BNP levels 100 pg/ml or over as high-levels of BNP. First, we compared the clinical feathers between patients with high-levels of BNP and those with non-highly elevated BNP. Second, cardiac performances influencing high-levels of BNP were determined.
Results: Study age and cardio-thoracic ratio were not different significantly between two groups. But body weight in high-level BNP was significantly lighter that in low-level BNP group. Additionally patients in high-level BNP group had more histories of hospital care significantly. After multiple logistic regression analysis high levels of BNP was independently associated with odds ratio of 6.9 ($p = 0.004$) for expanded end-systolic volume of RV ($\geqslant$240%), 6.4 ($p = 0.038$) for elevated end-diastolic pressure of RV ($\geqslant$9 mmHg) and 3.9 ($p = 0.024$) for elevated end-systolic pressure of RV (RV/LV$\geqslant$93%). These three factors explained 48% of high-elevated BNP (R-square = 0.48). The extension of end-dyastolic and end-systolic volume of LV had a weak tie to high levels of BNP by univariate analysis ($p < 0.1$).
Conclusions: Our study showed heart failure worsened all the more in patients with high levels of BNP. This study also revealed that a variety of RV overload were related to high levels of BNP in symptomatic infants with VSD. The left-sided functions were not significantly with BNP elevation even by univariate analysis. Heart failure might not be caused by volume overload of left heart, more might be caused by excessive burden of fragile right ventricle. Dysfunction of RV is difficult be detected by echocardiogram. We could use high-levels of BNP as the method of picking out the exhaustion for right ventricle in symptomatic infants with VSD.

P-100

P-wave dispersion in children with rheumatic heart disease

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Introduction: Rheumatic heart disease (RHD) is a common cause of morbidity and mortality in poor and developing countries. Mitral regurgitation results in volume overload, then causes left ventricular and left atrial remodeling and eventually left ventricular systolic impairment can be seen. Arrhythmia may be seen in patients with mitral regurgitation. P-wave dispersion is an electrocardiographic marker that reflects left atrial enlargement and also has been reported to be associated with inhomogenous and discontinuous propagation of sinus impulse and paroxysmal atrial fibrillation. The purpose of this study was to assess any atrial arrhythmia risk in children with chronic mitral regurgitation caused by rheumatic fever in respect to P-wave dispersion.
Methods: A total of 43 children with rheumatic mitral regurgitation, with a mean age of 12.22 ± 2.28 years, and 25 healthy children, with a mean age of 11.21 ± 1.79 years underwent 12-lead surface electrocardiography and transthoracic echocardiography. Chidren with acquired and congenital heart disease other than chronic rheumatic mitral regurgitation were excluded from the study. P-wave dispersion was calculated as the difference between the maximum and the minimum P-wave duration. An SPSS 17.0 program was used for statistical analysis.
Results: Children with RHD had significantly larger left atrium and ventricle than control group. All patients and control subjects had similar ($p > 0.05$) and normal systolic functions (ejectional fraction and fractional shortening). Patients had significantly longer P-wave dispersion compared to control group (41.07 ± 7.16 vs 27.76 ± 3.28, $p < 0.001$). In patient group, maximum mean P-wave duration was significantly longer (86.46 ± 8.61 vs 77.36 ± 6.36, $p < 0.001$) and minimum mean P-wave duration was significantly shorter (45.48 ± 5.67 vs 49.60 ± 5.71, $p < 0.01$) than control subjects. In all subjects ($n = 68$), P-wave dispersion was found significantly correlated to maximum P-wave duration ($r = 0.768$, $p < 0.001$), left ventricular end diastolic diameter ($r = 0.306$, $p < 0.05$) and left atrial width ($r = 0.252$, $p < 0.05$).
Conclusion: Inhomogeneity of atial conduction and prolonged atrial conduction time may be related to left atrial dilation in children with rheumatic mital regurgitation. These children should be closely followed up for the risk of clinically important arrhythmias.
Key Words: P-wave dispersion, rheumatic heart disease, children.

P-101

Paediatric myocarditis: new aspect from a case series

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Introduction: Our knowledge regarding the clinical characteristics of myocarditis presenting in childhood is patchy and inconclusive. Prolonged follow up created an opportunity to gather additional important knowledge regarding the nature of this rare condition. The clinical characteristics of paediatric myocarditis treated in our institute were analyzed in order to generate new knowledge about the course of the illness and possible comorbidities.
Methods: A retrospective data collection was performed to identify patients with myocarditis treated in our institute between the 1st of January 1996 and 31st of December 2010. The median follow up time was 12.71 years (range: 3.57-15.2 years).
Results: Over the 15-year span the diagnosis of myocarditis were established in eight children based on the clinical presentation. The patients' age ranged from 3 months to 8 years. The median age was 1,16 (0,25-8,71) years. Among the primary symptoms

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poor feeding, lethargy were present in 7, tachypnoea in five and chest pain in one patient. On admission all had an abnormal electrocardiogram and mitral regurgitations. On echocardiography 7 patients had severe systolic dysfunction of the left ventricle and one patient had regional wall motion abnormality with pericardial effusion. Cardiac enzymes or troponin were checked in 6 patients and were positive in 5. Among the 8 patients 3 died. In the survivors the left ventricular dilatation and the ejection fraction normalized within 4-21 months. In one patient with recurrent perimyocarditis the regional wall motion abnormality disappeared within 11 and 18 days. None of the cases progressed to dilated cardiomyopathy. Myocarditis or recurrent perimyocarditis preceded the manifestation of coeliac disease in two patients. Cystic fibrosis was diagnosed in one patient after the improvement of cardiac function. Alström syndrome was diagnosed years after complete recovery from myocarditis in one patient.
Conclusions: Myocarditis may precede the manifestations of other chronic paediatric diseases. The association between coeliac disease and myocarditis is already known from the literature. No publication was found until now regarding the potential relation between cystic fibrosis or Alström syndrome and myocarditis. The results are suggesting that myocarditis may be a sentinel of non-cardiac chronic diseases based on genetic susceptibilities.

P-102
Assessment of left ventricular functions with Tissue Doppler, Strain and Strain Rate Echocardiography in children with Juvenile Idiopathic Arthritis: An Observational Study

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Objective: In Juvenile idiopathic arthritis (JIA) cardiac involvement is usually silent without typical symptoms. The purpose of this study was to assess left ventricular functions with tissue Doppler echocardiography (TDE), strain and strain rate in children with JIA.
Methods: Our study was designed as cross-sectional study. Thirty pediatric patients with JRA and 30 age- and sex-matched healthy controls were studied. In addition to standard echocardiographic methods, tissue Doppler, strain and strain rate were performed to assess left ventricular functions in all participants. The means of variables that did not distributed normally were compared with "Mann-Whitney U" test.
Results: In patients with JIA, E′ values of mid and apical regions of left ventricular lateral wall were significantly lower than those of the controls (15.76 ± 3.24 cm/s vs 17.91 ± 3.29 cm/s, 11.10 ± 2.96 cm/s 12.64 ± 2.42 cm/s, $p < 0{,}05$). In longitudinal strain reflecting left ventricular regional systolic functions, apical-lateral, basal and mid-septum peak S values, lateral peak S values in circumferential strain (-17.30 ± 6.22 vs $-21.97 \pm 4{,}32$, -18.23 ± 4.62 vs -21.53 ± 2.69, -20.35 ± 3.75 vs -22.75 ± 3.50, -9.68 ± 7.12 vs -13.70 ± 6.81, $p < 0.05$, r:0.42, 0.41, 0.42), in longitudinal strain reflecting diastolic functions, apical-lateral, mid-lateral, apical-septum, mid-septum peak E values (2.22 ± 1.00 vs 3.17 ± 0.87, 1.62 ± 0.84 vs 2.15 ± 0.72, 2.51 ± 0.76 vs 3.31 ± 0.87, 1.99 ± 0.64 vs 2.47 ± 0.57, $p < 0.05$, r:-0.39, -0.55,-0.43) and in circumferential strain lateral and posterior peak E values (1.32 ± 0.83 vs 1.88 ± 0.94, 1.31 ± 0.71 vs 1.85 ± 0.91, $p < 0.05$, r:-0.33, -0.22) were significantly lower than those of the controls.
Conclusion: Although marked myocardial involvement was not detected with TDE in JIA patients with subclinical cardiac disease, regional impairments in left ventricular strain and strain rates were found.
Key words: Juvenile idiopathic arthritis, tissue Doppler echocardiography, strain, strain rate

P-103
Physical Activity Mediates Blood Pressure in Overweight Adolescents Without Abdominal Adiposity

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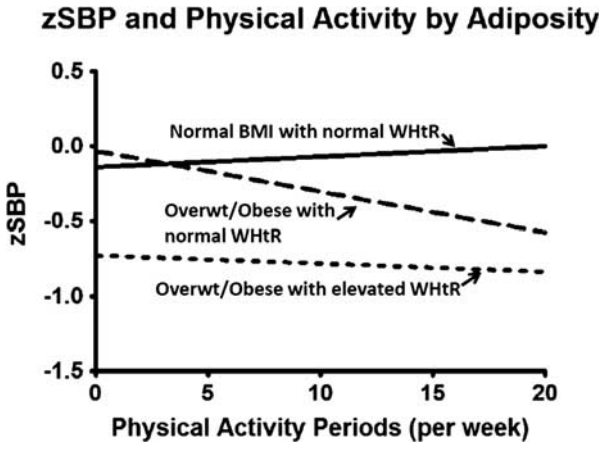
Objective: We sought to identify lifestyle factors that may mediate the association of adolescent blood pressure (BP) and hypertension (HTN) with adiposity and, hence, suggest targets for specific interventions.
Methods: N = 4,104 grade 9 students (14-15 years old, 52% males) were assessed as part of the Heart Niagara Healthy Heart Schools' Program. Measurements included BP, adiposity (body mass index [BMI] and waist circumference [WC]) and lifestyle assessment. WC to height ratio (WHtR) was classified as normal (<0.5), elevated (0.5-0.59) or abdominally obese (≥ 0.6). WC was also classified by percentile categories based on published normal values. BP was converted to age, gender and height specific percentiles and z-scores, and classified as normal ($<90^{th}$ %ile), pre-HTN ($<95^{th}$) and HTN ($\geq 95^{th}$). Associations between BP, adiposity and lifestyle factors were assessed in linear and logistic regression models adjusted for age at enrollment and gender.



Results: Median BP z-score was -0.6 (IQR: -1.3; -0.1) for systolic (zSBP) and $+0.0$ (IQR: -0.4; $+0.6$) for diastolic (zDBP), with 2.8% of subjects classified with pre-HTN and 0.8% with HTN. Based on BMI percentiles, 15% of subjects were overweight (85^{th}-94^{th}%ile) and 19% obese (95^{th}+%ile). WHtR > 0.5 was noted in 2.5% of normal BMI subjects, 38% of overweight and 95% of obese subjects. Higher WHtR category was associated with higher zSBP and zDBP within BMI categories. Higher WHtR category was weakly associated with HTN category within BMI categories. Higher WC%ile category was more significantly associated with higher HTN category within BMI categories. Higher zBP was significantly associated with family history of HTN but not cardiovascular disease. Higher zBP was associated with skipping breakfast, but no other dietary characteristic. Lower zBP was associated with greater weekly physical activity

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and participation in organized activities, but not with sleep duration. Subjects who were overweight/obese but did not have elevated WHtR showed lower zSBP but not zDBP with increasing level of weekly physical activity (Figure).
Conclusions: Waist measures in addition to BMI should be included in the anthropometric assessment of risk for obesity-related HTN. Physical activity interventions might be effective in managing BP for overweight/obese adolescents who do not have abdominal adiposity.

P-104
Hypoxic perinatal cardiomyopathy-diagnosis and evolution
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Hypoxic perinatal aggression induces appearance of manifestations of myocardial suffering with variable evolution, sometimes severe. Purpose: to present the main aspects of myocardial injury secondary to perinatal hypoxia.
Methods: Patients: 88 newborns aged 0 to 14 days, with normal birth weight, with perinatal hypoxia (Apgar score 3–7), receveing resuscitation, but without major congenital heart diseases. All cases were investigated by clinical exam, ECG, chest X-ray (Rx.CT), Doppler echocardiography (Echo). Most of patients were evaluated and after 6 months.
Results: The patients had mainly signs of neurological post hypoxic suffering, only 8 cases signs of severe heart injury(cardiomegaly, respiratory distress, cyanosis, peripheric hypoperfusion), other cases: systolic murmur (64 cases) and signs of PPHN (8 cases). Chest X-ray: cardiomegaly (32 cases). ECG: severe left ventricle (LV) repolarization disturbances and low voltage of QRS complexes (37), without ischemic changes. Doppler echo exam at 2-7 days of life revealed: ⋆ the absence of other severe congenital cardiac anomaly; ⋆permeability of foramen ovalae (100%) and forced foramen ovalae (gradient LA/RA > 8 mmHg); mild to severe tricuspid insufficiency and RV and RA dilation (29); sometimes right-left shunt through the FO ⋆myocardial hypertrophy (42 cases) mainly IVS(29), signs of PPHN(6); prolonged IVRT (35), increased myocardial performance index (44 cases), the systolic dysfunction in 5 cases and severe LV diastolic dysfunction in 45 cases. All the cases received spironolactone 1-2 mg/kg/day for 3 months. New evaluation at 6 months showed; reduction of the myocardial hypertrophy and of tricuspid regurgitation, normal LV systolic and diastolic function.
Conclusions: The perinatal hypoxia can induce a important myocardial injury as hypoxic ischemic myocardopathy or transient post hypoxic hypertrophic cardiomyopathy at more than 62,2% of patients, the signs of cardiovascular suffering missing often. Echo is the main method for diagnosis and follow up of perinatal hypoxic cardiomyopathy and is necessary performed from the first week of life. The research of cardiac biomarkers CPK-MB and cardiac troponin may amplify the value of cardiological investigation of hypoxic myocardial injury in newborn infant.

P-105
Cardiovascular Abnormalities in Williams Syndrome; 20 Years' Experience in Istanbul
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Introduction: Williams syndrome (WS) is a microdeletion syndrome affecting cardiovascular and connective tissue as well as the endocrine and central nervous systems in 1 in 10,000 live births. This study aims to identify and evaluate cardiovascular abnormalities (CVAs) in 45 WS patients.
Patients and Methods: We retrospectively reviewed a cohort of WS patients that were followed at our institution from January 1, 1990 through December 31, 2010. WS was clinically diagnosed by an experienced medical geneticist and confirmed by fluorescence in situ hybridization. CVAs were assessed using electrocardiography, echocardiography or cardiac catheterization.
Results: Twenty-seven patients (60%) were male; 18 were female (40%). The mean age at presentation was 4.6 ± 3.1 years (3 months-13 years); the follow-up period was 6.9 ± 4.4 years (6 months-18 years). CVAs were found in 86% of patients, the most common one being supravalvar aortic stenosis (SVAS) in 73% (isolated in 48%), peripheral pulmonary artery stenosis (PAS) in 42%, and mitral valve prolapse (MVP) in 22%. Less common were aortic insufficiency (15%), ventricular septal defect (11%), valvular pulmonary stenosis (11%), and aortic arch hypoplasia (8%) and coarctation (2%). Hypertension was present in 22% of patients. Surgical or catheter-based interventions were performed in 22% of cases. Two patients were lost in the postoperative period.
Conclusion: CVAs were found in more than four out of five patients, the most common ones being SVAS and PAS. Although surgery was performed in more patients with SVAS than with PAS, SVAS was minimal or mild in most patients and improved in few cases.

P-106
Cardiac involvement in infant of diabetic mother: diagnosis and follow up of evolution
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Background: Cardiac involvement is one of the most important clinical manifestations of newborn of diabetic mother.
Objectives: To present the main aspects of cardiac involvement in newborn of diabetic mothers (IDMs), the the diagnosis and follow-up their evolution.
Methods: Patients: 84 IDMs newborns investigated in the first week of life and 49 of them re evaluated at 6-12 month, by clinical exam, ECG, cardiothoracic radiography (Rx.CT) and Doppler echocardiography (Echo) for congenital and/or acquired cardiac diseases. Fetal echo was performed in 38 cases after 28 weeks of gestation.
Results: Fetal Echo showed: cardiomegaly and myocardial hypertrophy of left ventricle (LV)(11 cases), confirmed by postnatal Echo. Clinical exam in newborn: macrosomia (40% cases), in 32 patients a systolic murmur was found, only 3 cases with signs of heart failure and the other being asymptomatic or presenting signs for other pathology than cardiac. ECG: LV hypertrophy (14 cases) and disturbed ventricular repolarization in 30 cases. Rx.CT: cardiomegaly (12). Echo showed: non obstructive hypertrophic cardiomyopathy (HCMP) with asymmetric IVS hypertrophy (34 cases: 42%), arterial pulmonary hypertension(6), LV diastolic dysfunction with normal systolic function (52%) and congenital cardiac anomalies: PDA (6), VSD (3), coarctation of aorta (1), ASD (4 cases). LV myocardial hypertrophy was not significantly correlated with the type of mother's diabetes, before pregnancy or gestational, but rather to an inadequate control of disease. Control performed at 6-12 months (21 cases) revealed a

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normal morphological cardiac aspect (14 cases) or significant reduction of HCMP (7), all of them showing normal diastolic and systolic LV function.
Conclusions: Newborn of diabetic mother presents a high risk for cardiac involvement, either cardiac congenital malformations (17%) or acquired cardiac pathology: HCMP (43% of cases) and disturbances of diastolic function of LV (54%).This fact justify early cardiologic screening for all of these newborns with or without of cardiac suffering symptoms. Fetal echo provides useful data for diabetic pregnant women and should be made mandatory to all these patients. Echo is the most sensitive and noninvasive method of diagnostic, useful for primary diagnostic as well as for follow up.

P-107

Cardiac biomarkers for early diagnosis of anthracycline induced cardiotoxicity in children with malignant diseases

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Background: Cardiac biomarkers research in children treated with anthracyclines for malignancy is useful for the early diagnosis of anthracycline induced cardiotoxicity.
Objective: To establish the value of research of cardiac biomarkers for early diagnosis of anthracycline induced cardiotoxicity in children with malignant hemopathies
Methods: Patients: 46 children (aged 2 months-18 years), treated for malignant hemopathies with anthracyclines with various manifestations of cardiotoxicity. Control group: 20 healthy children without history of cardiac diseases. Patients and controls were investgated by: clinical exam, Doppler echocardiography (Echo), determination of plasma values of of cardiac biomarkers BNP(B natriuretic peptide) and cTnI(troponin).
Results: Determination of cardiac biomarkers showed: ⋆Increased plasma levels of BNP in 45.7% of patients, from a mean baseline of 89 ng/ml (0-117 ng/ml) to value 240 ng/ml (0-810 ng/ml),⋆ increasing cTnI values. plasma at 4.34% of cases, the initial values <0.04 pg/ml to values >0.04 pg/ml in 2 cases. Echo modifications: anthracycline induced cardiomyopathy or just only diastolic dysfunction of LV in majority of cases, often correlated with cumulative dose of anthracyclines. Biomarkers changes were correlated in most cases with the presence of clinical manifestations and echo modifications induced by anthracycline cardiotoxicity.
Conclusions: Increased levels of cardiac biomarkers: BNP and cTnI in children treated with anthracyclines ± other drugs with cardiotoxic efects positively correlates with installation of the cardiotoxicity with clinical or infraclinical manifestations, constituing an useful marker for the.cardiotoxicity. Changes in this parameters appeared early than echo modifications in anthracicline induced cardiotoxicity and is necessary to systematic monitoring these parameters during and after cytostatic therapy.

P-108

The 12-Lead Surface ECG in Patients with Congenital Defects of the Atrioventricular Canal: Insights into the Adaptation of the AV Conduction to the Variation of the Patients' Heart Rate

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Introduction: A diagnostic feature of common AV canal defect – affected Patients lies in their ECG. We describe here the characteristics of the ECGs of Patients with complete and partial CAV and compare them with a cohort of normal, age-matched subjects.
Methods: We retrospectively searched our ECG database for suitable tracings and found the pre-surgical ECGs of 51 patients aged 0- 4 months and the normal ECGs of 58 subjects, aged 0-4 months, with regular cardiac anatomy.We calculated the duration of the PQ, RR intervals and QRS complex and the QRS axis, the PQ/RR ratio, the mean PQ interval and QRS complex duration and the mean QRS axis in each group. The relationship between the PQ/RR (PQ duration normalized by the RR interval) and the RR interval was expressed as a correlation coefficient. Data were compared using appropriate Student's t-tests. A p value<0.05 was required to refuse the null hypothesis.
Results: In the case group, the mean PQ interval length was 130.00 ± 0.03 msec, the mean QRS duration was 69.76 ± 16.45 msec; the mean QRS axis on the frontal plane stood at 46.90°; 21 (45%) Patients had a leftward axis deviation (-83°–+18°), 3 (5.8%) subjects displayed incomplete right bundle branch block, 3 (5.8%) had signs of biventricular hypertrophy.
In the control group, the mean PQ interval and QRS duration were respectively 100 ± 0.02 msec and 59.22 ± 6.76 msec. The mean QRS axis was +99.54° (range: +30–+184).
Conclusions: Pre-surgery ECGs are significantly abnormal in Patients with CAV. Moreover, the PQ interval, which summarizes on the surface ECG the AV conduction, displays a significantly reduced increase along with that of the heart rate in children with CAV defects not surgically treated, when compared to normal, age-matched subjects. Differences in the morphology of the AV junction could possibly explain these differences, although we cannot exclude that in these Patients the conduction system may be congenitally different, and that this may also relate to their later tendency to non surgery–related AV blocks, as a possible expression of the wearing off of their conduction system.

P-109

Clinicopathological investigation on dilated cardiomyopathy in infants and children: The role of endomyocardial biopsy and biochemicalmarkers

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Purpose: Early diagnosis of dilated cardiomyopathy (DCM) enables the start of effective treatment with the purpose of stopping the progress of the disease and delaying the development of symptomatic heart failure. Regarding this fact, routine and widely accessible diagnostic methods in assessing the state of advancement of the disease and the prognosis are requiring. Current applications of biomarkers may offer a means of identifying high-risk patients before they develop symptoms not only in adults but also in the pediatric population, however there are some differences from adults in the approach to the diagnosis and management of cardiomyopathy in children. In this aspect, we try to make clear the prognostic factors in pediatric cardiomyopathy comparing clinical symptoms with biomarkers and ulstrastructural changes of pediatric patients with myocardial involvement by endomyocardial biopsy (EMB).
Patients and method: They included 28 DCM; 7 patients with idiopathic DCM, 3 dilated phase hypertrophic cardimyopathy

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(d-HCM), 2 noncompaction of left ventricule (NCVM), 7 myocarditis, 6 tachycardia induced cardiomyopathy(TIC), 2 hyperthyroidism and 36 Kawasaki disease(KD) as comparative subjects. Selected biochemical markers were high-sensitive CRP, myoglobin, Creatin Kinase MB, troponin T, heart-type fatty acid binding protein, ANP and BNP. Histopathology was evaluated with semiquantitative morphometry with an automated image analysis system.
Results: Resuscitated sudden death occurred in 3 of idiopathic DCM and NCVM. Elimination of focus following EPS was 1 for TIC. ICD implantation was one for d-HCM. Myocardial changes on EMB showed various abnormalities such as inflammatory cell infiltration and higher % fibrosis in cardiac death patients. Prevalence of abnormal biochemical markers was found in severely symptomatic DCM rather than KD.
Conclusions: Severely symptomatic DCM showed raised concentration of biochemical markers. Although clinical severity did not reveal statistic correlation with biochemical markers and histopathological severity, biochemical markers might be one of the plausible predictors for the severity of myocardial damage and ongoing ischemia. EMB may still be helpful to determine etiology in an otherwise undiagnose dilated cardiomyopathy in children. An aggressive diagnostic evaluation to detect severity of myocardial damage with biochemical markers and the use of a multifaceted treatment approach to prevent developing symptoms and sudden death should be required.

P-110
Rhythm and Conduction Abnormalities in Children with Acute Rheumatic Fever: Are They Specific to the Disease?

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Acute rheumatic fever (ARF) is a multisystem disease developing after upper respiratory tract infections with group A beta-hemolytic streptococcus (GABHS). It is common among children between 5-15 years.
During the course of the disease, some electrocardiographic (ECG) changes are seen, independent from carditis. First-degree atrioventricular (AV) block is the most common ECG abnormality, and it is used as a minor criterion in the diagnosis of ARF. Second- and third-degree AV block, ventricular tachycardia and junctional acceleration are also seen in ARF patients. In the present study, the specificity of these abnormalities for ARF was investigated.
The study consisted of patients with ARF (Group 1), children who had suffered from recent GABHS upper respiratory tract infection but had not developed ARF (Group 2) and patients who had diseases that may affect the joints and/or heart (Group 3).
The frequency of first-degree heart block in the surface ECGs of Group 1 patients was 21.9% (14/64), in Group 2 patients 0% (0/50) and in Group 3 patients 2.7% (1/37). First-degree AV block was significantly more frequent in Group 1 ($x2 = 17.877$, $p = 0.000$). Specificity of first-degree AV block for ARF was 96.7% and its positive predictive value was 88.8%.
In 10 patients in Group 1, accelerated junctional rhythm (AJR) was seen in surface ECG and/or Holter recordings; none of the patients from Groups 2 and 3 had AJR. Specificity and positive predictive value of AJR for ARF were both 100%.
No significant difference was detected between the groups in terms of presence of premature beats and corrected QT intervals. Escape beat/rhythm and ectopic atrial rhythm were significantly more frequent in Group 2 and 3 patients.
In conclusion, first-degree AV block in surface ECG and AJR in surface ECG and/or Holter recordings are specific for ARF. Although their frequencies are low, it seems that they can be used in the differential diagnosis of ARF, especially in patients with isolated arthritis.

P-111
Clinical course and the impact of interstage monitoring after Norwood I and Hybrid procedure

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Objectives: Infants with hypoplastic left heart syndrome (HLHS) are at risk for interstage morbidity and mortality especially between the first and second surgical stage after Norwood and Hybrid procedure.
Methods: We compared morbidity (defined as hospitalization/reinterventions) and mortality in patients treated for HLHS between the first and second stage, and the impact of interstage monitoring on outcome.
Results: Between June 2008 and December 2011 twenty-six (male = 14) infants with HLHS (n = 16) and other univentricular heart malformations with aortic arch anomaly (n = 10) were planned for interstage monitoring after Norwood I (n = 12) and Hybrid procedure (n = 14). Three (11.5%) infants died after first stage palliation (Hybrid n = 1; Norwood n = 2), 3 (11.5%) after second stage palliation (Hybrid n = 2; Norwood n = 1), all of them after early second stage (<90 days). There were no difference between Norwood I and Hybrid procedure regarding over-all mortality (23%) (Hybrid n = 3; Norwood n = 3). 7 (26.9%) infants could not be discharged from hospital due to hemodynamic instability and were referred to early second stage (<90 days). After first stage invasive re-evaluation rate before discharge was high with catheterizations in 14 (53.8%) (Hybrid n = 8; Norwood n = 6), with the need for re-interventions in 8 (30.7%) (Hybrid n = 5; Norwood n = 3) or cardiac surgery in 3 (11.5%) (Hybrid n = 2; Norwood n = 1).
14 infants had interstage monitoring for 89 (10-177) days. One infant (3.9%) died during interstage monitoring. Interstage monitoring was positive in 7 (50%) infants after 10 (4-68) days (Hybrid n = 5; Norwood n = 2), leading to rehospitalization and catheterization in 6 (Hybrid n = 4; Norwood n = 2), requiring interventions in 2 (PDA stent dilatation, ASD stenting, all in Hybrid). Overall, 3 of the 7 patients with positive interstage monitoring were candidates for an early second stage.
Conclusions: Morbidity in infants after first stage before discharge is characterized by a high reintervention rate (30.7%). After discharge interstage monitoring is positive in 50% indicating the need for catheter treatment in 29%. Despite retrograde aortic flow in infants with HLHS after Hybrid procedure mortality is comparable in both groups. Mortality after second stage is associated with early age (<90 days). This highlights the importance of interstage monitoring in both treatment groups.

P-112
Clinical Characteristics of Patients with Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease, Evaluation of the Long-Term Follow-Up Results

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The acute rheumatic fever (ARF) remains a significant health problem for our country. the aim of the study is, evaluate

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epidemiological, clinical and laboratory characteristics features of the disease and determine the prevalance according to the years and evaluate the long-term prognosis of the valve disease of the patients with carditis in bursa and its environs.
In this study, the medical records of 151 patients with the diagnosed of ARF between January 2000 and February 2011 in our Department, were retrospectively reviewed. Application date, initial complaints, major, minor and supportive signs, laboratory findings, echocardiographic findings and changes in valve involment during the follow-up time, were evaluated. To determine the valve insufficiency in patients with carditis, echocardiographic examination, in admission, after the inflammation recovered and in last control, were evaluated respectively. Patients age ranged between 4 and 15 years (9.6 ± 2.42), male and female ratio is 1.2 /10. Sixty-nine of the cases (45.7%) were 9 years or under and 82 (54.3%) were aged 10 years or over. The most common symptoms were fever (62.9%) and joint symptoms (36.6%) respectively. Carditis (76,8%) was the most common symptom according to the Jones major criteria. The most commonly association was, arthritis and carditis (52.9%) There was no difference in the distribution of the major criteria according to sex and age groups.
The most common valvular lesion in the first echocardiography was combined mitral (MR) and aortic regurgitation (AR) (59.8%). When evaluated separately, the most common valvular lesion was mitral valve insufficiency (%36,2). In the control echocardiographic examination after inflammation period, complete improved were deteced in the 15% of MR and %20,4 of AR according to the baseline examination. On the last echocardiographic examination total recovery rates were 37,5% for MR and %50 for AR, 39,5% of the cases, there were no change for valvular lesions. Subclinical carditis (silent carditis) was detected in 17.2% of the cases. surgical intervention was performed in six (5,4%) cases.
In conclusion, this study showed that, despite decrease in the number of cases over ten year period in Bursa and environs, ARF still remains a health problem for our region.

P-113

Echocardiographic Diagnosis of Acute Rheumatic Silent Carditis in Children and Comparison of the Features of Patients with Clinical and Silent Carditis

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Objective: Acute rheumatic fever and rheumatic heart disease continue to be an important public health problem in the developing countries. Doppler echocardiography is now widely used in most parts of the world for early detection and recurrences of clinical evident carditis (CC) and subclinic (SC) carditis. We aimed to determine the frequency of subclinic (silent) carditis and to compare clinical and echocardiographic features of the patients with silent and clinically evident carditis (CC).
Materials and Methods: 156 consecutive patients, diagnosed with acute rheumatic fever in our pediatric cardiology department between 2003-2009 were included into study. All the patients underwent echocardiographic evaluation. Clinical and echocardiographic features of the patients were recorded. Follow up data was also obtained. The patients without clinical evidence but with echocardiographic findings of carditis were diagnosed as SC. The features of those patients were compared with CC group.
Results: Acute rheumatic fever was diagnosed in 156 patients and 103 (66%) of them had carditis. The ratio of SC was 28.2% among these 103 patients. Among patients with chorea, 17(% 89.5) had evidence of carditis.The chorea was more frequently observed major criteria in the patients with SC (31%). Whereas, arthritis was more common in CC group. Isolated mitral regurgitation was the most common lesion and concomitant involvement of mitral and aortic valves was the next common finding in both groups. Seventy-four of the patients with carditis were followed up more than one year and 20 of those had SC. Valvular regurgitation disappeared completely in 14.8% and improved in 37% of the patients with CC. The recovery and improvement rates in SC group were 15% and 30%, respectively. No significant difference was determined for persistent valvular damage between the groups.
Conclusion: As the valvular involvement in both SC and CC group behave alike, they should be managed in the same manner. We suggest that Doppler echocardiography should be performed in all patients with suspected acute rheumatic fever for early detection of SC. Echocardiographic examination should be taken as a diagnostic criterion in order not to miss the diagnosis of SC, associated with significant morbidity.

P-114

Truncal valve dysplasia is related to abnormalities of the coronary arteries: a pathological study

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Background: The arterial valve of hearts with a common arterial trunk (CAT) may show dysplastic leaflets and be insufficient or, more rarely, stenotic. Marked truncal valve malformations appear to be an important causal factor in early deaths and may impact the postoperative prognosis of the affected children. The morphology of truncal valve deformities is still poorly defined and usually described qualitatively in the literature as nonspecific fibrous thickening of the nodular margins of the leaflets. Anomalies of the coronary arteries are also described as frequent findings in CAT. To date, however, no study has sought to establish objective relations between the truncal valve abnormalities and other morphological features of the defect.
Objective: Systematically assess, both qualitatively and quantitatively, using histo-morphometric techniques, alterations of the truncal valve and check for possible associations with the coronary anatomy.
Methods: Thirteen heart specimens with CAT were analyzed. The thickest truncal semilunar leaflet was determined on gross examination and sampled for histological analysis. Linear measurements were obtained: proximal, medial and distal thicknesses, length and total area of the leaflet. Gross features of the coronary arteries ostia (number, position and shape) were annotated.
Results: The thickest valvar segments were the medial and distal ones. There was a significant negative correlation between the distal thickness of the valvar leaflet and the linear distance from the coronary ostium to the valvar commissure ($R2 = 0.448$; $p = 0.024$ and $R2 = 0.697$; $p = 0.001$, respectively for the left and right coronary ostia). The prevalence of high take-off (coronary ostium above the sinutubular junction) was respectively 36.4% and 18.2% for the right and left coronary arteries. Anomalous shape (not rounded or elliptical) of the coronary ostium was significantly more prevalent for the left than for the right coronary artery ($p = 0.008$).
Conclusion: Anomalies of the position and shape of the coronary ostia are related to the dysplasia of the truncal valve. Also, there are significant differences between the anomalies of right and left

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coronary ostia. These malformations should call attention to the possible effects on the correspondent territory of irrigation of the coronary artery.

P-115

Myocardial performance during and after therapeutic hypothermia for perinatal asphyxia in term neonates

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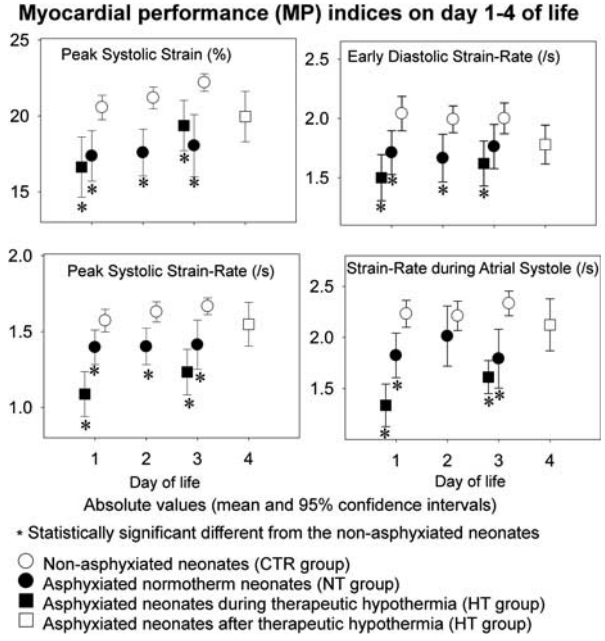
Introduction: Therapeutic hypothermia (HT) reduces brain damage after perinatal asphyxia. The effect of HT on the myocardial performance (MP) is poorly investigated. Strain-Rate (SR) and Strain by tissue Doppler (TD) are sensitive indices for MP after perinatal asphyxia. Higher absolute Strain and SR values reflect better MP. This study assess Strain and SR by TD as markers for MP in cooled term neonates as compared to historical control groups of asphyxiated term neonates treated with normothermia (NT) and non-asphyxiated term neonates (CTR).
Methods: Twenty asphyxiated neonates (HT) cooled to core temperature 33.5°C for three days were examined during hypothermia on day 1 and 3 and after rewarming on day 4. Twenty NT and 48 CTR examined on day 1, 2 and 3 were used as controls.
The MP was assessed as Longitudinal Peak Systolic Strain, Peak Systolic SR, Early Diastolic SR and SR during Atrial Systole
Results: The HT group were more severely asphyxiated than the NT group; the pH of the neonate was 7.07 (7.01, 7.14) (mean (95%CI)) vs. 7.20 (7.10, 7.31), the Base Excess -16.2 mmol/L (-18.4, -14.1) vs. -10.6 (-14.8, -6.3), the 5-min Apgar-score 4 (2, 5) (median (quartiles)) vs. 5 (4, 6) and the 10-min Apgar-score 5 (3, 6) vs. 7 (6, 8) ($p < 0.05$).



The MP was similar on day 1-3 within each of the groups, except for an increase in the Peak Systolic Strain in the CTR group ($p < 0.05$).
The MP was similar in the NT and HT groups during cooling on day 1-3, and lower than in the CTR group ($p < 0.05$).
After rewarming, the MP improved on day 4 in the HT group, approaching the MP in the CTR group on day 3.
Conclusions: Although the HT group was more severely asphyxiated than the NT group, the myocardial performance was equally depressed in both groups as compared to age matched controls. The myocardial performance in the hypothermic treated neonates improved after rewarming on day 4, approaching the levels in the non-asphyxiated neonates on day 3. Therapeutic hypothermia did not decrease the myocardial performance during treatment and might have had a positive impact after treatment.

P-116

Correlation between calcification and oxidative stress in chronic Kawasaki disease

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Introduction: Patients with acute Kawasaki disease (KD) sometimes develop coronary lesions despite undergoing various medical treatments. Such patients have to undergo appropriate medical therapy and undergo long term follow-up. These coronary lesions initially show dilatation or aneurysms and their appearance is altered over time because of remodeling. It is now thought that these lesions may finally lead to atherosclerosis. Oxidative stress is known to play an important role in vascular remodeling and arteriosclerosis progression. However, the dynamics of oxidative stress in KD are not known. In this study, we investigated the correlation between calcification (as the final stage of atherosclerosis) and oxidative stress in patients with chronic KD.
Methods: We measured the reactive oxygen metabolite (dROM) levels and biological antioxidant potential (BAP) in 23 patients with chronic KD; of these patients, 9 had coronary calcification, 9 did not have calcification, and 5 did not have coronary lesions. Oxidative stress indicates the imbalance of the redox system; therefore, we tried to evaluate the oxidative stress by determining the balance between dROM and BAP (adjusted ratio, BAP/dROM/7.541; normal range, 1.0 ± 0.13) and compared these ratios for each group.
Results: The patients who had calcification had lower dROM values and higher BAP values than the patients who did not have calcification. Thus, the adjusted ratio was significantly higher in the patients who had calcification ($p < 0.01$).
Conclusions: Calcification may be one of the factors responsible for the collapse of the balance of the redox system in patients with chronic KD. Calcification is a well-known risk factor for cardiovascular events; therefore, evaluating oxidative stress may aid in predicting cardiovascular events in patients with chronic KD.

P-117

The first treatment experience of a patient with functional single ventricle and homeostasis pathology

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Introduction: Homeostasis pathology increases significantly risk of death after CHDs correction. We observed 5 cases of development of venous thrombosis in patients with "functional single ventricle" after BCPC and TCPC. We assessed the methylenetetrahydrofolate reductase thermolabile variant (MTHFR C677T), the prothrombin mutation (FII G20210A), the factor V mutation (FV A4070G and G1691A), and the platelet glycoprotein receptor IIIA (GpIIIa PlA1/A2) as predictors of thrombosis.

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Results: We had a 6-year-old patient with functional single ventricle, heterotaxy with polysplenia syndrome, absence of subhepatic segment of inferior vena cava, arteriovenous malformation of pulmonary vessels and heterozygous carrier of allele PL-A2 of GpIIIa gene, presence of which determines increase of thrombocyte aggregation and resistance to antiaggregant therapy. At 1 year old Kawashima procedure, main pulmonary artery ligation were performed. At examination the patient's weight was 19 kilos, saturation– 53%-59%, antithrombin deficiency III – 73% (norm – 80-140%), protein C decrease-67% (norm – 10-130%), level increase of soluble fibrin-monomeric complexes to 7,5 mg% (norm – to 3,5%), erythrocytes level to 6.99*109 /l and Hb to 17,7 g/dL, hematocrit 57%. Performed surgery: redirection of hepatic venous into v. azygous by prothsesis «Gore-tex» 16 MM, left pulmonary artery banding, atrioventricular valve repair. Postoperative monitoring of homeostasis system included: determination of antitrombin III, activated partial thromboplastin time, D-dimer, INR, level of thrombocytes and hematocrit. Level of antithrombin III was defined daily during 9 days. When the level was lower, intravenous infusion of antithrombin III human was prescribed till target level 90% with simultaneous infusion of fresh frozen plazma in dose 10 ml/kl/daily. In 6 hours after surgery heparine infusion in dose 5 un/kg/hour with further dose increase to15 un/kg/hour was started. On 5th day after surgery heparine was canceled, fraxiparine was prescribed 0,3 ml twice per day. From 6th day and later the patient got warfarin. In two months after surgery INR was 2,2-2,5, antithrombin III – 75%, thrombocytes 373*109 / l, D-dimer – positive (>250 ng/ml), however, venous thrombosis were not disclosed.
Conclusion: Complex approach with use of modern medication allows improving treatment results of patients with congenital heart diseases with presence of homeostasis pathology.

P-118
Close and remote results of endovascular treatment of aorta coarctation in children

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Aims: To assess close and remote results of endovascular treatment of aorta coarctation (AoCo) in children and teenagers.
Methods: From 1998 to 2010 endovascular angioplasty of AoCo was performed to 59 patients with average age 6,4 ± 2,4 years old. All the patients were performed clinical instrumental examination: Echo – before and after endovascular intervention, balloon dilatation in X-ray operation room. Necessary balloon diameter was defined according to aorta diameter on the level of left atrium. 28 pts were examined in a year after the procedure.
Results: At the preoperative stage the most pts (80%) complained only on high blood pressure (BP). Only 28 children (47,5%) had AoCo without combination with other heart diseases. According to Echo, till endovascular intervention peak pressure gradient in a descending aorta was 30 mm Hg in 6,8% pts, in 37,3%-30-50 mmHg, in the rest – more than 50 mmHg. Manometry, performed before angioplasty, did not always coincide with Echo data. After balloon dilatation transstenotic gradient (TSG) was absent only in 8 children (13,5%). The rest TSG was 10 mmHg in 10 pts (16,9%), in 23 pts (38,9%) – 10-22 mmHg, in 14 pts (23,7%) – 20-30 mmHg and only in 4 children it was significant enough – more than 35 mmHg. High remaining TSG was marked only in patients with primary high pressure gradient in the AoCo field in combination with its hyperplasia. BP normalization was marked in the most pts after treatment, and only 14 children required antihypertensive drugs. All the pts were performed Echo after angioplasty in 2-3 days. Peak pressure gradient was up to 10 mmHg in 10 pts (16,9%), 20 mmHg – in 25 pts (42,4%), 30 mmHg – in 18 pts (30,5%), higher than 30 mmHg – in the rest. In a year the most examined children (60%) kept good treatment effect – peak pressure gradient – 10-20 mmHg, absence of arterial hypertension (AH). The rest pts had AH in spite of low TSG (15-30 mmHg) and good feet pulse.
Conclusion: In spite of AoCo abolition preserved arterial hypertension in 40% of children was disclosed. It requires follow-up and adequate therapy.

P-119
Tissue Doppler Imaging in very preterm infants. The alteration of myocardial performance during the first 24 hours of life.

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Introduction (or Basis or Objectives): Tissue Doppler Imaging (TDI) is a new tool for assessment of myocardial function which has not been much applied in preterm infants born before 32 weeks gestation. During the first day(s) of life, these infants go through a substantial circulatory transition, and some infants, especially the youngest ones, are in need of circulatory support in order to maintain adequate blood pressure and flow. More knowledge about cardiac performance during this transitional phase could potentially guide the choice of cardiac/circulatory support.
Methods: In a total of 50 newborn, preterm infants, echocardiographic examination for assessment of structural and haemodynamic parameters was performed at 5, 12 and 24 hours after birth. If feasibel, TDI was recorded for off-line measurement of annular systolic and diastolic velocities of the left and right ventricle, as well as displacement of both ventricles.
Results: One or more TDI measurements were obtained in 33, 20 and 22 infants at the 5, 12 and 24 hours exams respectively.

Age at exam	5 h	12 h	24 h	p
cTDI S' LV (cm/s)	1.8 (0.7)* n = 32	1.3 (0.4)* n = 18	1.5 (0.6) n = 22	0.002
cTDI S' RV (cm/s)	2.8 (0.9)* n = 33	2.2 (0.9)* n = 20	2.5 (0.9) n = 21	0.01
cTDI D' LV (cm/s)	2.3 (1.1)* n = 32	1.8 (0.8)* n = 18	2.1 (0.9) n = 22	0.035
cTDI D' RV (cm/s)	4.1 (1.6)* n = 33	2.8 (0.9)* n = 20	3.3 (1.5) n = 21	<0.001
Dis LV (mm)	1.9 (0.9)* n = 30	1.4 (0.7)* n = 18	1.8 (0.8) n = 22	0.024
Dis RV (mm)	3.3 (1.2)* n = 31	2.2 (1.2)* n = 20	2.9 (1.3) n = 21	0.002

cTDI S' = colour TDI peak systolic velocity; cTDI D' = colour TDI peak diastolic velocity; LV = left ventricle; RV = right ventricle; Dis = displacement. P value pertains to the difference between the 5 and 12 hour exams (*)

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We found a significant reduction of systolic (S') and diastolic (D') velocities as well as displacement of both ventricles from 5 to 12 hours age. There was a slight, but not significant increase of all parameters from 12 to 24 hours age.
Conclusions: By applying colour TDI to a group of very preterm infants, we found a significant decrease in several parameters of myocardial performance from 5 to 12 hours age. Larger studies are needed to confirm these findings.

P-120
Genetic approach to immunopathogenesis of acute rheumatic fever
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Introduction: Prevention of acute rheumatic fever is accomplished by adequate penicillin therapy. But in developing countries fluctuations are not rare. During the latent period of disease immune response to streptococcal antigens occur. It is important to investigate genetic influence in immunopathogenesis to prevent the disease.
Methods: 23 patients with arthritis and 43 patients with carditis were investigated in Ege University, department of pediatric cardiology. 29 were male and 37 were female, age between 7-12 years. Control group consisted of 31 male, 36 female cases, age between 9-13 years.
Antistreptolysin O (ASO), C reactive protein (CRP) levels, hemogram were evaluated and electrocardiography, echocardiography were performed in all patients before and after treatment. Also monocyte chemotactic protein 1 (MCP-1), angiotensin converting enzyme(ACE), interleukin 6 (IL-6) levels were evaluated as immonopathologic parameters and genotype and allele frequency were studied.
Results: In both groups no significant relation was found before and after treatment according to acute phase reactants ($p < 0.01$). In patient group; ACE homozygote DD, MCP-1 allele frequency was higher but not significant when compared with control group ($p > 0.05$). But IL-6 genotype frequency was significant ($p < 0.01$).
Genotype and allele frequencies were not statistically significant in patients with arthritis and carditis. ($p > 0.05$)
Conclusion: Indeed many factors play role in ARF immunopathogenesis, ACE, MCP-1 and IL-6 contributes development of inflamation.

P-121
Chest pain – the most common cardiac couses in pediatric age group
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Chest pain as a symptom is a very frequent reason for pediatric emergency department visits. Parents and patients as well are usually very frightened about the cause of the pain. However, most researches showed that heart disease is not the leading cause of chest pain. In the chest there are many other organs rather than the heart that can cause the pain. The etiology of chest pain can be idiopathic, musculoskeletal, psychological, respiratory, gastrointestinal, cardiac and any other. While examining a child with chest pain a doctor must be aware of the most frequent chest pain causes and in the same time be able to exclude conditions that require hospitalization or emergency interventions. Although very rare in children, acute myocardial infarction requires emergent intervention. Most of the published cases of myocardial ischemia in children are linked with specific conditions–anomalous origin of left coronary artery from pulmonary artery, nephrotic syndrome, familial hypercholesterolemia, Kawasaki disease and other.
The aim of this research was to determine the number of patients with cardiac causes of chest pain. The research included 96 patients admitted to pediatric cardiology ward in the last 5 years. Diagnostic processes included ECG, chest X-ray, cardiac enzyme levels, ECHO and other methods used according to patient's anamnesis and clinical findings. In 17 out of 96 patients we found that the cause of pain was cardiac, in 32 out of 96 was idiopathic, in 14 out of 96 was musculoskeletal, in 16 out of 96 was respiratory illness, in 8 out of 96 was psychogenic, in 4 out of 96 was gastrointestinal and there are some other causes found in 5 out of 96 patients. The most frequent cardiac cause of chest pain that we find was mitral valve prolapse (7/17) and arrhythmias (5/17). Less frequent causes were high arterial pressure (3/17) and myocarditis (2/17). Our results confirmed previously published data on this topic. Nevertheless pediatric patients with chest pain should be carefully examined and the cause of the chest pain should be found whether it is cardiac or non–cardiac type.

P-122
The Evaluation of P Wave Dispersion, QT Dispersion, QTc and QTc Dispersion On Early Diagnosis Of Autonomic Dysfunction in Children and Adolescents with Type-1 Diabetes Mellitus
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In this study, it was aimed to determine the sensitivity of p wave dispersion (Pd), QT interval, QT dispersion (QTd), QTc and QTC dispersion (QTcd) in early diagnosis of cardiac autonomic function disorder (CAFD) that was caused by diabetes in children and adolescents with type-1 Diabetes Mellitus (Type 1 DM) and to evaluate the association between these measures and other markers such as duration of diabetes.
A total of 150 children and adolescents admitted to Pediatric Endocrinology Department between June 2009 and June 2010 with Type 1 DM diagnosis and a control group consisting of 100 healthy children and adolescents with silent murmur admitted to Pediatric Cardiology Department were enrolled to this study. Electrocardiography (ECG) was performed in all cases and heart rate, Pd, QT, QTd, QTc and QTcd were calculated. The clinical and demographic features such as age, gender, age at admission, HbA1c levels of the patients were examined and the effects of these measures on Pd, QT, QTd, QTc, and QTcd were investigated.
There were no differences between the patient and control groups in terms of age and gender. According to the duration of follow-up, QTd and QTcd in groups 1, 2 and 3, according to HbA1c levels during follow-up QTd and QTcd in groups 2 and 3, according to blood glucose levels Qtd and QTcd in groups 2 and 3 were found to be sighificantly higher than the control group. In agreement with the literature, Pd was found to be significantly higher in children with Type-1 DM in our study.
Conclusion: CAFD may develop in the course of Type-1 DM and may cause to significant morbidity and mortality. CAFD can be diagnosed with ECG in these cases before the development of symptoms. In our patients, in agreement with the literature, Pd, QT, QTd, QTc and QTcd were found to be significantly higher in patients without CAFD symptoms compared with the controls.

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P-123

Myocardial TissueI Doppler Echocardiograhic evaluation of preterm infants with bronchopulmonary dysplasia at 2 to 4 year of age

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Objective: To investigate the role of myocardial tissue Doppler echocardiography in detecting cardiac pathology in BPD patients at 2 to 4 year of age and to find out possible risk factors related to cardiovascular sequela.
Study design: Infants born prematurely with BPD (N = 21, 4 severe BPD, 3 moderate BPD and 14 mild BPD) and without BPD (N = 20) were evaluated with conventional and myocardial tissue Doppler echocardiography at 2 to 4 year of age.
Results: In conventional echocardiography; right ventricular fractional shortening, tricuspid E/A ratio and mitral late diastolic inflow velocity, pulmonary acceleration time was decreased; mitral E/A ratio, left and right ventricular myocardial performance indexes were increased in BPD group compared to controls.
Early – to- late tissue Doppler velocity ratio (E'/A') was decreased at the tricuspid annulus and systolic velocity was increased at the interventricular septum with myocardial tissue doppler measurements in BPD group. Birth weight, disease severity and cumulative steroid dosage were related with echocardiographic changes.
Conclusion: BPD affects global cardiac performances at 2 to 4 years of age with regard to birth weight, disease severity and cumulative steroid dosage. Myocardial tissue Doppler examination did not have additional value in demonstration of these changes with conventional echocardiography.

P-124

Progression of ascending aorta dimensions in pediatric patients with bicuspid aortic valve

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Introduction: Bicuspid aortic valve (BAV) is the most common congenital cardiac defect. Apart from obstructive lesions, a significant part of patients develop dilatation of the ascending aorta. Natural history and significance of aortic dilatation in pediatric BAV patients is not well defined. Therefore, we studied progression of dimensions in these patients with and without coarctation.
Methods: Patients with BAV or with BAV and coarctation (BAV+coarc) were selected from institutional database. Inclusion criteria were: at least three echocardiograms over last 15 years, each at least one year apart. Aortic diameters were measured at 3 levels (annulus, sinus, ascending), and transformed into age- and gender-specific z-scores. Impact of gender, diagnosis (BAV vs. BAV+coarc) and valve function on changes of z-scores over time was analyzed by linear mixed-effects regression.
Results: 116 measurements were recorded in 28 patients (9 patients BAV, 19 patients BAV+coarc). Mean age at time of measurements was 12.0 years (range, 0.1 to 23.1 years). Eighty-five measurements were performed in male patients. Aortic regurgitation was present in 29, and mild valvular stenosis in 22 measurements. No deaths were recorded. Overall, mean z-score for aortic annulus was 1.14 (range, -1.72 to 3.72), for sinus 0.49 (-2.87 to 3.63), and for ascending aorta 1.86 (-2.02 to 5.54). Ascending aorta z-scores increased significantly over time in the entire sample (0.07/year; 95%CI, 0.04 to 0.10; $p < 0.001$); in both BAV (0.12/year; 95%CI, 0.05 to 0.18; $p < 0.001$) and BAV+coarc (0.06/year; 95%CI, 0.02 to 0.10; $p = 0.002$); in boys (0.08/year; 95%CI, 0.04 to 0.11; $p < 0.001$) but not in girls (0.03/year; 95%CI, -0.05 to 0.10; $p = 0.49$); in patients with (0.10/year; 95%CI, 0.03 to 0.17, $p = 0.006$) and without aortic regurgitation (0.07/year; 95%CI, 0.03 to 0.10; $p < 0.001$); and in patients without aortic stenosis (0.07/year; 95%CI, 0.04 to 0.11; $p < 0.001$) but not in patients with aortic stenosis (-0.02/year; 95%CI, -0.10 to 0.06; $p = 0.65$).
Conclusions: Ascending aorta z-scores progressed significantly, particularly in patients with isolated BAV; indicating intrinsic wall abnormalities. Progression was influenced by gender and aortic function. Patients will be further examined for molecular markers, aiming to identify patients at risk for severe progression, with potentially highest benefit from early medical treatment.

P-125

Pulmonary Artery Sling: A Rare Cause Of Recurrent Aspiration Pneumonia

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Introduction (or Basis or Objectives): Pulmonary artery sling (PAS), known as anomalous left pulmonary artery originating from right pulmonary artery, is a vascular anomaly in which the left pulmonary artery arises aberrantly from the proximal part of the right pulmonary artery and courses posterior to the trachea to reach the left hilum. We report an intriguing case who was presented for recurrent aspiration pneumonia caused by pulmonary arterial sling, and diagnosed by computed tomographic angiography.
Methods: 14 months old girl firstly admitted to emergency room with dyspnea when she was 6 months old. Inadequate ventilation was noticed in chest X-ray. Because of the infiltration in right paracardiac pulmoner region on X-ray, bronchoscopy was performed and solid food remnants were removed from her right bronchus, but the left main bronchus couldn't be accessed. CT was planned but informed consent couldn't be taken from the parents. But recurrent pattern of the dyspnea, respiratory distress attacks and chronic cough, stridor, feeding intolerance lead them to emergency room and CT scan was done: Left pulmonary artery was originating from right pulmonary artery (Fig1a), Lumen of left bronchus was obliterated due to compression and pulmonary artery calibration was decreased at this level (Diagnosis was PAS). After the diagnosis echocardiographic examination performed; Left Pulmonary artery was originating from right pulmonary artery, and there was no pressure gradient over pulmoner artery in high parasternal views. The surgical correction (left pulmonary artery was excised and anastomosed to the main pulmonary artery) was done one week after the diagnosis. After surgery the patients' respiratory problems and feeding intolerance were all recovered. Postoperative echocardiographic and tomographic examinations were in normal ranges (Fig 1b).
Conclusions: Infants with recurrent respiratory symptoms, such as chronic cough, stridor and wheezing, should be examined carefully and vascular rings and slings must be taken into consideration for differential diagnosis. PAS is a rare vascular anomaly which results in respiratory problems. Early surgical management of symptomatic patients is an effective way to

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treating PAS and relieving the symptoms. By this case, we want to emphasize a rare congenital anomaly, which results recurrent pulmonary problems.

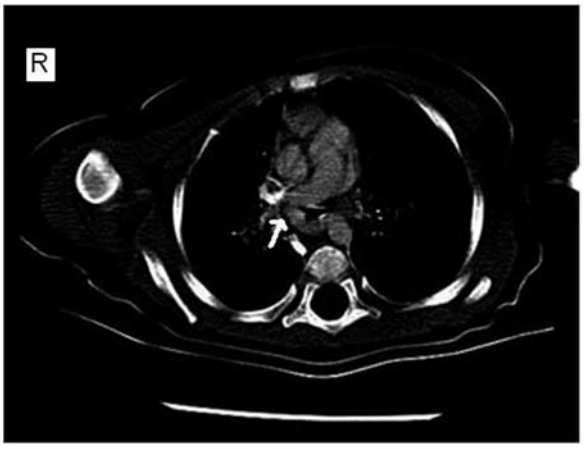

Fig1A: Preoperative image: Left pulmonary artery originating from right pulmonary artery

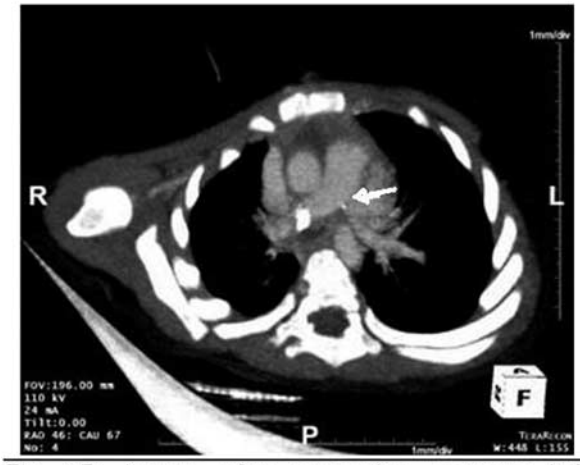

Fig 1B: Image after corrective surgery: left pulmonary artery originating from main pulmonary artery

P-126

Comparison between tilt table and active orthostatic testing for syncope simulation and hemodynamic pattern investigation in children

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Objective: to evaluate effectiveness of different orthostatic tests (tilt table test and active lying-standing test for syncope simulation and hemodynamic pattern's investigation in children suffering from recurrent syncope.
Methods: 205 children in age from 4 to 17 years old (mean age 13,08 ± 2,88) after at least the 3rd episode of syncope and having no contraindications revealed on physical examination, 12-lead electrocardiogram, underwent orthostatic testing: 72 – tilt table testing, 133 – active standing testing according to random selection. Blood pressure and electrocardiogram were recorded by monitors DASH 2000 and Datascope Duo™. Results were compared with those in control group. Control group: 92 healthy children (mean age 12,64 ± 3,04) never had syncope or presyncope episode, underwent physical examination, 12-lead electrocardiogram and orthostatic testing. Tilt testing was performed in 40, active standing testing – in 52 control group's children. There was no statistically significant difference of groups in gender and age division. Data were processed with SPSS 17,0 statistical packet. Comparison between groups was made using Student's t-test, F (ANOVA), and χ^2 test.
Results: Tilt table test was positive in 72,2%, and active test was positive in 71.4% of investigation group children. There was detected no statistical significant difference, $p = 0.904$, $\chi^2 = 0.015$. Tilt testing was positive in 27.5% children from control group, and active standing test – 23.1% accordingly. Statistical significance of control group was $p = 0.627$, $\chi^2 = 0.236$. Difference of positivity in orthostatic testing between children with recurrent syncope and children never had the episode was significant: $p = 0.000$, $\chi^2 = 56.6$. Patient's age had no significant influence for orthostatic test results ($p = 1.19$, $\chi^2 = 2.36$). No statistical significant difference for orthostatic test results was noticed in patient gender ($p = 0.34$, $\chi^2 = 0.89$) and reflex or atypical circumstances of syncope ($p = 0.31$, $\chi^2 = 1.03$).
Conclusions: Tilt table testing and active orthostatic testing showed no statistical significant difference for syncope simulation and it's hemodynamic pattern investigation in children suffering from recurrent syncope.

P-127

Electrocardiographic changes in children with pectus excavatum

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Introduction: Pectus excavatum is a dfeformity of the chest that consists of backward displacement of the sternum and costal cartillages giving rise to a depresssion in the area under sternum. In the absence of any cardiac anomalies in children with pectus excavatum, electrocardiographic changes are to be expected, because of alterations in the position of the heart.
Methods: Included in this study were 50 children with pectus excavatum seen at the pediatric clinic in Martin between 2008 and 2010. An electrocardiogram, chest roentgenograms and Frank vectorcardiograms had been obtained in all 50 children.
Results: The authors evaluated in detail electrocardiographic findings in 50 children with pectus excavatum, before surgically repairing their lesions. 68% of patients with pectus excavatum have electrical axis displaced to the right. Most frequently, but also the most typical, are complexes of rSŕ or rsŕ of the whole right precordium. Complexes of rSŕ or rsŕ were found in 41 per cent of children with pectus excavatum In the right precordial leads, evidence of the right ventricular hypertrophy has been not recorded. Quite often–in 43.2% of cases–there has been proven the absence of q waves in the left precordial leads. In 11.4% of the cases above the left precordium, there has been found the high waves (R waves), but with no changes on the T waves.
Conclusions: The authors refer to interesting electrocardiographic findings in children with pectus excavatum. They claim that electrocardiographic changes in pectus excavatum in children are relatively constant. The results of the authors confirm the claims of other authors that rSŕ or rsŕ complexes above the right precordium are caused by the distortion cardioelectrical field arousing from the cavities of the heart for abnormal changes in the heart by deformation of the thorax. Waves ŕ in the right precordial leads means the depolarization of the base part of the

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right ventricle and also the higher parts of the interventricular septum, if these areas are located at the front and right by the rotation of the heart in pectus excavatum.

P-128

Procedural safety, hemodynamic and histological findings of myocardial biopsy in children

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Background: Cardiac catheterization with endomyocardial biopsy is an invasive diagnostic technique for children with suspected cardiomyopathy and myocarditis.
Aims: To study the procedural safety, hemodynamic and histologic findings of cardiac catheterization with myocardial biopsy in children treated for cardiomyopathy and myocarditis.
Methods: All children with myocardial biopsy between 2003 and 2011 in our institution were analyzed regarding hemodynamics, periprocedural outcome (defined as inotropic support and catheter-related complications), and histological findings.
Results: Twenty-six patients at (mean ± SD) age of 8.5 ± 5.4 years with a transvenous endomyocardial biopsy in the right ventricle were included. There were no major catheter-related complications such as myocardial perforation, hemopericardium, or need of mechanical resuscitation. All cardiac catheterizations were performed under general anaesthesia with sevoflurane/atracurium. Inotropic support including use of milrinone/adrenaline had to be established during the procedure in 7 (27%), increased in 2 (7.7%), continued in 2 (7.7%) patients. Hemodynamic findings under inotropic support revealed elevated enddiastolic left ventricular pressure (20 ± 7 mmHg), left atrial pressure (18 ± 6 mmHg), borderline pulmonary artery pressure (24 ± 7 mmHg), normal pulmonary vascular resistance (2.3 ± 1.8 Woods units/m2), and low cardiac index (3.1 ± 0.8 ml/min/m2). Left ventricular biplane ejection fraction was reduced with 39 ± 20% (performed in 10 patients). Coronary artery angiography ruled out coronary artery disease in all patients. Histological findings of myocardial biopsy revealed dilated cardiomyopathy in 3, myocarditis in 16 (8 chronic, 6 borderline, and 2 acute) of all cases. Other etiologies were identified in four (failing Fontan, restrictive myocarditis, anthracycline-induced cardiomyopathy, endocardfibroelastosis). No pathologic changes were found in three samples. Myocardial and serological viral polymerase chain reaction (PCR) was positive in six and in eleven patients, respectively, with predominance of human herpes virus type 6 and 7, and parvovirus B 19.
Conclusions: Cardiac catheterization with myocardial biopsy can be performed safely, although increased inotropic support during procedure may become necessary. Hemodynamic invasive data and histologic findings complete clinical diagnosis of myocarditis and cardiomyopathy, and give important information stratifying further clinical management.

P-129

Recurrent syncope in the child – don't forget the coronary arteries.Atresia of the left coronary artery in a 9 years-old boy

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Background: Syncope is one of the most frequent reasons for referral for evaluation in pediatric cardiology. Whereas vaso-vagal syncope are usually harmless, potentially life-threatening aetiologies, including arrhythmias, structural heart disease, anomalies of the coronary arteries (CA) and myocardial ischemic disease need to be ruled out.
Methods: Case report and review of the literature
Results: This boy presented with recurrent syncope following physical activity since the age of 5. Repetitive cardiac evaluations including clinical examination, electrocardiogram (ECG), 24h-ECG, stress ECG, event recorder, echocardiogram and stress echocardiogram were unremarkable. Neurological and gastroenterological evaluations were normal. At the age of 9 years the boy experienced sudden cardiac arrest during a soccer game. Cardiopulmonary resuscitation was immediately started and was successful after threefold defibrillation of ventricular fibrillation. Again all non-invasive examinations were unremarkable, except a slightly diminished ventricular function, which quickly normalized under milrinone. Invasive electrophysiological study was normal. Coronary angiography showed a prominent right CA and retrograde perfusion of the left sided CA system, with small left anterior descending (LAD) CA and ramus circumflexus (Figure). The ostium and the first segment of the left main CA were missing and atresia of the left coronary artery was diagnosed. Myocardial perfusion studies consisting of SPECT and perfusion magnetic resonance showed no ischemia under adenosine stress and absence of scarring. Prophylactic medication with metoprolol was started and surgical revascularisation with a left internal mammary artery to proximal LAD bypass grafting performed without complications. The patient was discharged 10 days postoperatively in good clinical conditions with acetylsalicylic acid.
Conclusions: CA anomalies are rare, but a frequent cause of sudden cardiac death in adolescents and young adults. In patients with exercise related syncope, CA anomalies need to be ruled out. Atresia of one CA is exceedingly rare with only 28 reported cases. This is usually an isolated lesion, but can be related to other cardiac defects in up to 30% of the cases. Surgical revascularization using internal mammary artery is the therapy of choice providing good results and growth of the left CA system.

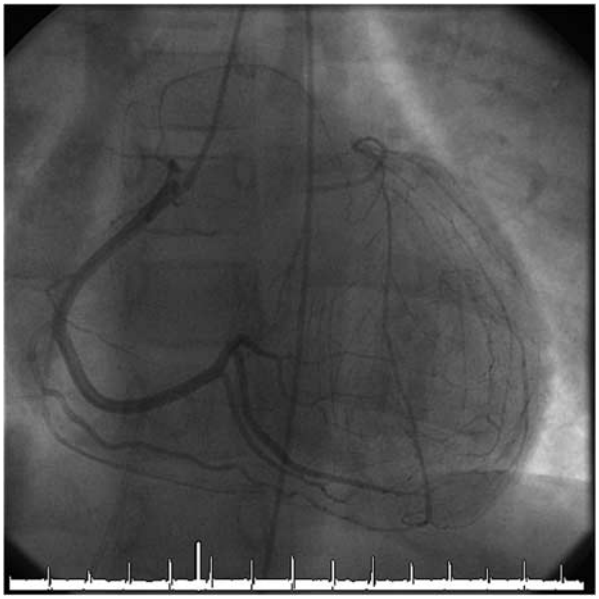

Figure: selective right CA angiography.

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P-130

Significance of Tenascin-C in Diagnosis of Chronic Rheumatic Heart Disease

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Introduction: The differential diagnosis of valve insufficiency, either rheumatic or congenital, may not be allways possible. Besides silent carditis, patients with congenital valve disease have also risk of developing chronic rheumatic heart disease (CRHD). Tenascin-C (TnC), an extracellular matrix glycoprotein, increases in inflammatory diseases. The balance between the oxidant and antioxidant systems is found to be impaired in many inflammatory diseases and also CRHD is a chronic inflammatory disease. The aim of this study was to assess the role of TnC in the differential diagnosis of rheumatic and congenital valve diseases, and also to evaluate the oxidant-antioxidant system in childhood.
Methods: The study groups, aged 3-17 years, consisted of 25 children with CRHD, 25 children with congenital valve disease and 20 healthy age-matched control subjects. Total antioxidant status (TAS), total oxidant status (TOS), oxidative stress index (OSI) and TnC levels were compared among the groups. Chi-square, ANOVA and Mann whitney-U tests were used for statistical analysis.
Results: In children with CRHD, with a mean age of 13.52 ± 2.71 years, the percentages of mitral and aortic regurgitations were found 88% and 40% respectively. Mitral valve prolapse ($n = 14$) and bicuspid aorta ($n = 11$) were present in patients with congenital valve disease ($n = 25$; mean age = 10.91 ± 4.19 years). Mean TnC level of the patients with CRHD was significantly higher than congenital ($p < 0.01$) and control groups ($p < 0.05$). However, there was no statistically significant difference between the congenital and control groups in terms of TnC. The values of TAS, TOS and OSI were found to be statistically similar ($p > 0.05$) in all groups.
Conclusions: Tenascin-C can be used as a biochemical marker in the differential diagnosis of CRHD from congenital valve diseases. As the oxidant and antioxidant systems were found to be in equilibrium in chronic rheumatic and congenital valve diseases, oxidative stress can be thought not to have a marked role in the etiopathogenesis of CRHD.
Key words: Children, chronic rheumatic heart disease, congenital valve disease, tenascin-C, total antioxidant status, total oxidant status.

P-131

A New Biochemical Marker in Diagnosis of Acute Rheumatic Fever: Ischemia Modified Albumin

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Introduction: Ischemia modified albumin (IMA), as measured using the albumin cobalt binding test, is currently the most promising biomarker for early detection of cardiac ischemia. The purpose of this study was to assess the role of IMA in diagnosis of acute rheumatic fever (ARF) and also to evaluate the IMA levels in children with chronic rheumatic and congenital valve diseases.
Methods: The study groups, aged 5-18 years, consisted of 40 children with ARF, 35 children with congenital valve disease, 33 children with chronic rheumatic heart disease (CRHD) and 20 healthy age-matched control subjects. The group of ARF was divided in three groups according to modified Jones Criterias. The IMA levels were compared among groups and also subgroups, and the association to acute phase reactants were investigated.
Results: The patients with carditis ($n = 18$), isolated arthritis ($n = 12$) and chorea ($n = 10$) were the subgroups of ARF group. Mitral and aortic regurgitations were present in children with CRHD in the percentages of 93.9 and 39.4, respectively. Mitral valve prolapse and bicuspid aorta were present in children with congenital valve diseases in the percentages of 54.3 and 45.7, respectively. The mean IMA level of ARF group was significantly higher than chronic rheumatic, congenital and control groups, seperately ($p < 0.001$). The mean levels of IMA in both carditis and isolated arthritis subgroups of children with ARF were significantly higher than control group ($p < 0.001$, $p < 0.01$, respectively), however, there was no statistically significant difference was found between the chorea subgroup and control subjects. Additionally, statistically significant correlations were detected between the mean IMA level of children with ARF and acute phase reactants ($p < 0.001$ for WBC, $p < 0.01$ for ESR and CRP). No statistically significant differences were found among the mean IMA levels of rheumatic, congenital and control subjects.
Conclusions: As the IMA levels of children with ARF were found to be increased, IMA could be used as a biomarker in diagnosis of ARF and also can be thought to be assumed as an acute phase reactant.
Key words: Acute rheumatic fever, children, chronic rheumatic heart disease, congenital valve disease, ischemia modified albumin.

P-132

Evaluation of Serum Ischemia Modified Albumin Levels in Children with Acute Rheumatic Fever Before and After Therapy

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Objective: The aim of the study was to investigate ischemia modified albumin (IMA) levels in children with acute rheumatic fever (ARF) before and after therapy and compare them with those of controls.
Methods: 27 patients with ARF and 18 healthy children, age and sex matched were included in our study. The diagnosis of ARF was established according to the modified Jones Criteria. Follow-up studies were made at the time of acute phase reactans levels returned normally. IMA levels were measured using the rapid and colorimetric method with the albumin cobalt binding test. CRP, erythrocyte sedimentation rate (ESR) and white blood cell (WBC) were estimated by routine methods.
Results: Children with ARF had significantly higher serum ESR, CRP and IMA levels than same parameters of the controls at the time on admission. IMA (ABSU) was measured as 0.41 ± 0.10 in the control group, 0.57 ± 0.9 in the study group before treatment and 0.48 ± 0.12 in the study group after treatment. After treatment, statistically important decrements were determined in the levels of ESR ($p < 0.001$), CRP ($p < 0.001$) and IMA ($p < 0.001$). There was no significant difference for IMA levels between after treatment and control group ($p > 0.05$). IMA levels at the time on admission were correlated positively with ESR ($r = 0.605$, $p < 0.01$) and CRP ($r = 0.543$, $p < 0.01$).
Conclusion: We concluded that increased IMA levels in patients with ARF at the time of diagnosis could be considered as a sign

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of increased inflammation. We also suggested that IMA levels could be used as a follow-up marker such as CRP and ESR for to evaluate the efficacy of treatment in ARF.
Key words: Acute rheumatic fever, ischemia modified albumin, inflamation, treatment.

P-133

Anatomopathological analysis of the valvar dysplasia and aortic arch abnormalities in common arterial trunk

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Background: Some of the associated abnormalities in common arterial trunk (CAT) like aortic arch obstruction and truncal valvar dysfunction may impact on the operative mortality and morbidity.
Objective: We sought to evaluate the gross features of the aortic arch and the histo-morphometric morphology of the truncal valve, looking for a possible relation between these anatomical abnormalities.
Methods: Twenty three heart specimens with CAT were analyzed. Gross features, such as aortic arch route and obstruction, and the presence of an arterial duct, were annotated. The thickest truncal semilunar leaflet was sampled for histological analysis. Linear measurements of thirteen of the leaflets were obtained: the thickness, length and total area. The collagen area fraction was also determined for the sampled leaflet using color detection in sirius-red stained histological sections.
Results: Five (26.31%) specimens had some abnormality of the aortic arch: four retroesophageal subclavian arteries and one coarctation. In 6 (31.57%) of the available cases, the arterial duct was present and 1 (5.26%) had an arterial ligament. It was possible to establish a significant association between the presence of a patent arterial duct and a retroesophageal subclavian artery ($p = 0.038$). The thickest valvar segments were the medial and distal ones. The valvar thickness correlated to the total leaflet area ($R2 = 0.805$; $p < 0.001$ and $R2 = 0.843$; $p < 0.001$, respectively for the medial and distal leaflet segments). The group with an arterial duct presented a trend toward greater total area of the semilunar leaflet ($p = 0.053$). Patients with values of collagen area fraction larger than the group average (44,2%) showed a tendency to be older ($p = 0.095$).
Conclusion: The association between an aberrant subclavian artery and the presence of an arterial duct in CAT can serve as a guide to the echocardiographer in the diagnosis of associated lesions. Dysplasia of the valvar leaflets is characterized by thickening of its medial and distal segments, and may be considered a progressive process, taking into account the trend toward increased collagen area in older patients. Finally, the association of a greater leaflet thickness and the presence of an arterial duct could suggest a common causal factor leading to both abnormalities in CAT.

P-134

P wave duration and dispersion in children with uncomplicated FMF

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Introduction: Familial Mediterranean fever (FMF) is an autoimmune disease inherited as an autosomal recessive trait and is characterized by recurrent attacks of fever and sterile polyserositis. Cardiovascular involvement is one of the leading cause of morbidity and mortality among FMF patients. A recent study found that FMF patients had an abnormally high P wave duration (Pdu) and P wave dispersion (Pdi), markers for supraventricular arrhythmogenicity.
The aim of our study was to further evaluate atrial dispersion and its relationship with systolic and diastolic ventricular functions in children with uncomplicated FMF.
Methods: The study group consisted of 25 children with uncomplicated FMF and age-and sex-matched 25 healthy controls. We performed electrocardiography with Doppler echocardiography on patients and controls. All participants underwent 12-lead electrocardiography under strict standards. The P-wave dispersion was calculated as the difference between maximum and minimum P-wave durations.
Results: There were no significant differences between the groups regarding age, weight, height, systolic and diastolic blood pressures, heart rates, C-reactive protein.
While systolic functions were in normal range, some of the diastolic function parameters were impaired in patients with familial Mediterranean fever during childhood. The P-wave dispersion of the patients with FMF was significantly greater than that of the controls group($p < 0,05$)
Conclusions: P-wave dispersion was higher in the children with FMF than in the healthy control subjects.Increased Pdi and Pdu in our uncomplicated FMF children might be related to depression of intra-atrial conduction due to atrial dilatation and increased sympathetic activity. These children should be closely followed up for risk of life-threatening arrhythmias.

P-135

Congenital heart disease in an infant with pseudohypoaldosteronism type 2 (Gordon syndrome)

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Pseudohypoaldosteronism type 2 (PHA2), also called Gordon's syndrome, is a rare autosomal dominant disease characterized by familial hypertension and hyperkalaemia; distal renal tubular acidosis despite normal glomerular filtration; normal or low normal serum aldosteron and plasma renin activity; and hypercalciuria and the increased risk of urolithiasis. All findings may not be determined in the infancy period. The congenital heart disease in PHA2 has not been reported previously. We, here, report an infant with PHA2 associated with cyanotic congenital heart disease.
A one-month-old boy, ninth child of a non-consanguineous marriage, was referred to our pediatric clinic with a history of cyanosis. He was born at term after unremarkable pregnancy. There was no family history of renal or other medical diseases. His weight and height were 4890 g and 57 cm (both 25-50th percentile), respectively. Physical examination was unremarkable apart from cyanosis and a soft systolic ejection murmur at the upper left sternal border. Vital signs were normal. Serum electrolytes, hepatic and renal function tests were normal. Echocardiography revealed complete atrioventricular septal defect; pulmonary atresia; and hypoplasic left ventricle and pulmonary arteries. Angiography confirmed the diagnosis. After 15th day of the admission he developed persistent hyperpotassemia. He was not receiving any drug leading to hyperkalaemia. The laboratory findings were as follows: serum sodium 139 mmol/l (reference range 135-145), potassium 6.4 mmol/l (3.5–5.5), chloride 103 mmol/l (98-106), creatinine 0.3 mg/dl

(0.3-0.7), blood urea nitrogen 4.2 mg/dl (5-18), calcium 10 mg/dl (8.8-10.8), phosphorous 4.4 mg/dl (3.7-5.6), aldosteron 74 ng/dl (5-90), plasma renin activity 9.5 ng/ml/h (<16.6), trans-tubular potassium gradient 4.2 (>8), and spot urine calcium/creatinine 0.2 (<0.8). Venous blood gases were as follows: pH: 7.41 (7.35-7.45), PCO2 33 mmHg (35-45) and HCO3 21 mmol/l (22-29). Renal ultrasound was normal. He diagnosed as having PHA2. Hydrochlorothiazide (1 mg/kg/d) was given. On third day of hydrochlorothiazide, serum potassium level returned to normal (4.2 mmol/l).

PHA2 should be considered in patients with an isolated hyperkalaemia. This case suggests PHA2 may be associated with congenital heart diseases. This is an unreported constellation.

P-136
Growth-differentiation factor-15 and Tissue Doppler imaging in Detection of Asymptomatic Late-Onset Anthracycline Cardiomyopathy in Childhood Cancer Survivors

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Introduction: Anthracyclines (ANT) are potent drugs in the treatment of cancer. Anthracycline cardiotoxicity can manifest anytime, from the start of chemotherapy to decades after its completion.

Both Doppler tissue imaging and speckle-tracking echocardiography results correlate well and appear to be more sensitive than standard echocardiography in detecting subclinical anthracycline cardiotoxicity. Growth differentiation factor-(GDF-15)15 is a distant member of the transforming growth factor-β cytokine superfamily. Serum GDF15 level is an independent risk factor for development of cardiovascular events and are predictive of recurrent myocardial infarction and death from pulmonary embolism.

The aim of this study was to evaluate the value of plasma levels of GDF-15 and Tissue Doppler imaging (TDI) in in detection of late myocardial dysfunction in survivors of childhood cancer treated with anthracyclines.

Methods: Thirty-eight children who received ANT chemotherapy (ANT dose: 100-375 mg/m2) and were followed up for avarage 34 months (11-82 months) were enrolled. 2DE with aspects of conventional indexes and tissue Doppler imaging (TDI) were performed. Serum levels of GDF-15, troponin I were measured. Twenty-six healthy children served as the control group.

Results: There were no significant differences in conventional indexes of 2DE and troponin I levels between the ANT and the control groups. Among diastolic parameters showed in significant differences between the patient and the control group. Growth differentiation factor 15 was significantly elevated in patient group as compared with control group ($p < 0,05$).

Conclusions: The heart function of patients who received ANT chemotherapy needs to be monitored for a long term. TDI and GDF-15 level can be used as early indexes for monitoring the heart function.

P-137
Reduced left ventricular end-diastolic volume in children with hypertrophic cardiomyopathy can be reproducibly measured with 3-D echocardiography and correlates with abnormal physiology

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Background: Hypertrophic cardiomyopathy (HCM) is associated with hypertrophy that reduces left-ventricular end-diastolic volume, often with an asymmetrical cavity that renders volume calculations by M-mode measurements invalid. The aim of this study was to investigate whether 3-D volume measurements of left ventricular (LV) volume can be used for assessing disease severity in children with HCM.

Methods: Ultrasound examinations were performed with Philips IE33 and analysed with 3-D QLab software (version 7). 80 healthy children and teenagers age 0.2 – 20 years were used to study the factors influencing LV volume, and 22 young HCM-patients (age 0.03 – 21 years; mean 10.4 years) who had had LV-volume measured before commencing therapy comprised the study group.

Results: LV volume in normal children correlated positively independently with both body surface area (BSA), and age. We also found a sizable gender difference in the 10-18 year olds, with males having 13% higher end-diastolic volumes (49.8 ± 5.3 versus 43.9 ± 4.5 ml/m2 BSA; $p = 0.0008$), and 17.6% higher stroke volumes (31.4 ± 3.2 versus 26.7 ± 3.1 ml/m2 BSA; $p = 0.00005$) than females. The smaller 0-9 year group showed a similar but non-significant trend. Each HCM-patient was given an age- and gender-matched control for statistical comparisons. The normal age-matched controls (average age 10.6 years) had a mean end-diastolic volume (EDV) of 44.9 ml/m2 BSA [95% CI 41.2-48.6]; median 45.4 ml /m2 BSA. HCM-patients had a mean EDV of 35.0 ml/m2 BSA [30.3-39.7]; median 35.7 ml /m2 BSA ($p = 0.00019$ signed rank). Ejection fraction was higher in HCM-patients (median 69.2% versus 66.6% in controls; $p = 0.0028$), but stroke volume at rest nevertheless remained lower in the HCM-patients (median 24.3 versus 28.3 ml/m2 BSA; $p = 0.00019$). Percent reduction in EDV correlated with diastolic dysfunction expressed as E:A ratio, and E:e ratio (correlation coeffients 0.44 and -0.47) and there was an inverse relationship with ejection fraction (-0.54). The reproducibility of EDV in HCM-patients between two independent observers was good with a mean difference of 0.17 ml/m2 BSA [95% CI -0.7 till 1.0 ml/m2 BSA].

Conclusions: Reduced end-diastolic LV volume and stroke volume in children with hypertrophic cardiomyopathy correlates with abnormal physiology and may be a useful tool to assess disease progression.

P-138
The Use of Artesunate in a child with acute myocarditis caused by HHV 6 virus

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Artesunate is an antimalarial drug derived from a plant, Artemisia annua and in vitro and sporadic human subjects shown to have antiviral activity. The drug has been used for multidrug resistant human cytomegalovirus. We report the use of artesunate in a one year old child in heart failure due to an acute viral myocarditis caused by Human Herpes Virus-6 (HHV 6) which did not respond to standard therapy.

The child developed signs and symptoms of an HHV-6 infection and was in severe heart failure one week later. IgM and IgG were positive for HHV-6. The child was treated with inoptropics, diuretics, immunoglobulin and gangcyclovir without signs of improvement after four weeks of therapy.

A left ventricular assist device (LVAD) was implanted for unloading the left ventricle. The myocardial biopsy showed fresh myocyte necrosis with inflammation and CD3 positive lymphocytes and CD 68 positive macrophages expressing enhanced amounts of MHC class II molecules. The presence of HHV-6 was found in the

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myocytes. Artesunate was given orally 10 mg/kg/day for 10 days. The control biopsy showed no signs of active inflammation and some myocardial recovery. The treatment had no observed side-effects. The heart recovery permitted successful explantation of the LVAD and discharge.
Conclusion: Artesunate was used in a child with HHV-6 myocarditis and severe heart failure. The biopsy and clinical status improved. No side effects of artesunate were noted.

P-139
Evaluation of the cardiovascular changes in the children with bicuspid aortic valves
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Introduction: Bicuspid aortic valve is the most commonly seen malformation but its cardiovascular effects during childhood have not been well studied. In this study, markers predicting cardiovascular changes and the relationship between progression of the disease and valvular dysfunction were evaluated in children with isolated bicuspid aortic valve and compared with the evaluations of children with tricuspid aortic valve.
Methods: The study was a prospective study. 41 children with bicuspid aortic valve was study group and 25 with tricuspid aortic valve was control group. The age was ranging between 5-15y. Blood lipids, endothelin, homocysteine, hsC-reactive protein, myeloperoxidase activity, glutathione peroxidase, matrix metalloproteinase 9, tissue metalloproteinase inhibitor levels were studied to evaluate cardiovascular risk and changes. By transthoracic echocardiography, aortic valve structure, left ventricle, and ascending aorta were evaluated. The carotid artery intima-media thickness were measured. The data were compared with the control group's data.
Results: There was no relation between the type of bicuspid aortic valve and aortic insufficiency and stenosis. Left ventricular hypertrophy could be seen in patients with minimally dysfunctioning bicuspid aortic valve so they should be followed very closely for the occurence of diastolic dysfunction. Biochemical markers showing endothelial dysfunction and carotid artery intima-media thickness were not changed in patients with bicuspid aortic valve. Tei index was significantly high in the study group. Patients with normal or mildly affected cardiac function had influences of global functions of left ventricule that can not be detected with M-mode echocardiography. Aortic diameters of the patient group were increased significantly at the level of aortic sinotubular junction and ascending aorta. The increase in the aortic valve insufficiency was due to the increase in the diameter of the aorta. Contrary to adults with bicuspid aorta, increased aortic flexibility was associated with the non-occurence of atherosclerosis yet. There was no relation between aortic elasticity and aortic stenosis and insufficieny level.
Conclusions: This study points out that atherosclerosis do not develop in early ages of patients with bicuspid aorta with mild aortic stenosis and insufficiency. The detected pathological effects of bicuspid aortic valve were left ventricular dysfunction, aortic diameter and aortic elasticity increase.

P-140
The importance of intraoperative transesophageal echocardiography in the evaluation of congenital heart diseases: 2 years experience of our center
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Introduction (or Basis or Objectives): Preoperative transesophageal echocardiography (TEE) provides a review of the preoperative diagnoses and decisions. Postoperative TEE gives ideas for further initiatives through the evaluation of surgical repair surgical repair.
Methods: 553 patients underwent surgery due to congenital heart disease between November 2009 and December 2011 in our center. 186 patients who were operated underwent TEE in the operating room. The application rate of TEE among all procedures was found to be 34%. TEE could not be performed in patients under 6 kg, who had contraindication for TEE, due to technical reasons. In addition, TEE could not be performed in cases that do not require intracardiac repairs (PDA closure, shunts, coarctation repair, repair of vascular rings, pulmonary band surgery) and in patients with ASD.

Pre-operative diagnosis	Preoperative diagnosis
C-TGA Rastelli	Significant tricuspid regurgitation
TAPVD	Stenosis in the connection point of pulmonary vein-left atrium
Supravalvular aortic stenosis	Stenosis in the supravalvular region
c-TGA +VSD+PS	Significant PS
Subaortic ridge	Subaortic stenosis
VSD	Residual ventricular septal defect
DORV	LVOT obstruction
Additional surgical procedures	**Postoperative control**
Conduit banding	Minimal TR
Expansion of anastomosis line	No obstruction
Expansion of stenosis	No supravalvular stenosis
Conduit application	No stenosis in conduit
Expansion of ridge resection	No stenosis
No residual VSD.	No residual VSD
Elimination of LVOT obstruction	No LVOT obs. AV Block

TTE	Decision
VSD (perimembranous, wide)	VSD closure
Tetralogy of Fallot	VSDclosure,transanuler patch
Tetralogy of Fallot	VSD closure, Transanuler patch
VSD(perimembranous, wide)	VSD closure
TEE	**Per-op decision**
Multiple apical muscular defects	Pulmoner banding
Aortic valve obstructing RVOT	VSD closure and aortic valve repair
Aortic valve obstructing RVOT	VSD closure and aortic valve repair
Additional subpulmonic defect	The closure of both VSD

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The preoperative TEE was compared with preoperative transthoracic echocardiography. The surgical procedure was assessed by postoperative TEE and the consideration of whether there was a need for a re-intervention was established.
Results: The surgical plan was changed according the preoperative TEE in four patients scheduled for surgery after TTE. The rate of patients in whom the surgical plan was changed among all the patients was found to be 2%. The status of 4 patients in whom surgical strategy was changed was summarized in Table 1.There were 7 patients for whom heart-lung pump was needed again with perioperative TEE in operations scheduled by preoperative TEE and the decision of the council. (Table-2) This ratio was determined as 3.7% among all patients.
*Conclusions:*TEE is a method providing important information that can change the surgical strategy in patients in whom TEE must be applied in pediatric cardiac surgery centers.

P-141
Clinical profile of hypertrophic cardiomyopathy in children – own experience
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Introduction: The clinical presentation in children with HCM is heterogeneous, ranging from asymptomatic forms to malignant expressions that may result in sudden cardiac (SCD) or heart failure-related death.
Aim of study: Retrospective analysis of clinical profile in children with HCM hospitalized in the years 1991-2011. Mean follow-up was 7.4 yrs (ranged from 1month to 20.5 yrs).
Material and methods: We analyzed 92 patients, mean age 10.4 ± 5.1 yrs. Patients demographics, clinical symptoms, family history of HCM and SCD as well as the results of echocardiography, 12-leads ECG, 24-h Holter ECG were analyzed.
Results: The mean age at diagnosis of HCM was 5.7 yrs. In 32(35%) children HCM was diagnosed in infancy. A family history of HCM was observed in 42(46%)pts. Family SCD due to HCM occurred in 25(27%)children. Patients most frequently reported fatigue, impaired exercise tolerance (66/72%), chest pain (29/32%), recurrent syncope (12/13%). In 60 (65%)pts asymmetric hypertrophy, in 30(33%) concentric hypertrophy, whereas in 2(2.2%) apical hypertrophy of the LV were found. The averaged septal thickness was 16.5 mm, which in relation to BSA was 266% of the mean standard. The massive LV hypertrophy ⩾ 30 mm was observed in 9(9.8%)pts. Abnormal resting ECG recording was found in 95%pts. In 24-h Holter ECG in 13(14%)children nsVT, whereas in none VT were observed. Cardiac arrest with successful resuscitation occurred in 4(4.3%)pts. In 14(15%) children ICD was implanted, in 4(4.3%)pts for secondary prevention and in 10(11%) as primary prevention of SCD. For HTx were enrolled 5 children (in 1pt HTx was performed, 3pts died while waiting for HTx, 1 child is still waiting). In follow-up died 10(11%)pts, average annual mortality was 1.47. In 4(4.3%) children SCD occurred (mean age 11.2 yrs), 6(6.5%)pts died due to progressive heart failure (mean age 8.9 yrs).
Conclusion: (1) In our group of children with HCM a high incidence of familial form of HCM and sudden cardiac death in the family history were observed. (2) The most common clinical symptom was fatigue, a progressive impaired in exercise tolerance, recurrent syncope and chest pain. (3) The frequency of implantable cardioverter-defibrillator for primary prevention of SCD and annual mortality rate was comparable with the data of the literature.

P-142
Risk stratification and prevention of sudden cardiac death in children with hypertrophic cardiomyopathy
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Background: Hypertrophic cardiomyopathy is the commonest cause of sudden cardiac death (SCD) in children. The aim of study was to evaluate the risk factors for SCD and the possibility of primary and secondary prevention of SCD in children with HCM .
Methods: Retrospective analysis of 92pts, mean age 10.4 ± 5.1 yrs with HCM diagnosed from 1991 to 2011. Mean follow-up was 7.4 yrs. Patients demographics, clinical symptoms, family history of HCM and SCD as well as the results of echocardiography, ECG, 24-hour ECG, exercise test were examined. Data from all patients have been analyzed regarding the presence of major risk factors for SCD according to ACC/AHA/ESC 2006 recommendations.
Results: The mean age at diagnosis of HCM was 5.7 ± 5.6 yrs. Screening for familial HCM revealed 42(46%) positive cases, of whom 79% had a first degree affected family member. The major risk factors for SCD were present in 51(55%) pts: cardiac arrest in 4(4.3%)pts, sustained VT in none of pts, family history of SCD (FSCD) in 25(27%), syncope in 12(13%), LV thickness ⩾30 mm in 9(9.8%), abnormal exercise BP (ABPR) in 20(21%) and nsVT in 13(14%)pts. ICD was implanted in 14(15%)pts, in all 4 pts after cardiac arrest as secondary prevention, in 10(11%) children as primary prevention. Among the 10 patients with ICD implanted as primary prevention, 4 pts had one (3pts recurrent syncope, 1pt nsVT), 5 children had two (FSCD+nsVT; syncope+IVS⩾30 mm; nsVT+IVS⩾30 mm; FSCD+syncope; FSCD+ IVS⩾30 mm) and 1pt had three major risk factors (FSCD+IVS⩾30 mm+ABPR). The appropriate shock was diagnosed in 2pts, inappropriate interventions were observed in 1pt with ICD implanted as secondary prevention. In children in whom the ICD was implanted as primary prevention, there was no adequate or inadequate discharge.
Conclusions: (1) The major risk factors for SCD was found in up to 51% of children, of whom 22 (24%) had ⩾ two risk factors. (2) Most common indication for ICD implantation as primary prevention of SCD were the major risk factors such as family history of SCD, recurrent syncope and massive LV hypertrophy. (3) High-risk patients with HCM ought to be prospectively identified and ICD implantation should be considered.

P-143
Outcome of Congenital Heart Defects Associated with 22q11.2 Deletion
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Introduction: 22q11.2 deletion is the most common of the chromosomal microdeletions. This study looks at the association of congenital heart defects with 22q11.2 deletion.
Method: The data used is from the Congenital Anomaly Register and Information Service for Wales (CARIS) from 2001-2010, irrespective of pregnancy outcome.
Results: There were 40 cases of 22q11.2 deletion reported in Wales, giving a gross prevalence of 1.2 per 10,000 total births (1 in 8335 total births). Of these, 30 cases had both 22q11.2 deletion and at least one congenital heart defect (75% of all

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22q11.2 deletions reported). In 26 of these 30 cases, pregnancy resulted in a live birth (87%), 3 cases were terminated (10%) and the remaining case was a still birth (3%). Of those with no heart anomaly, 8 were live births (80%), and the remaining 2 cases were terminated (20%). No other anomaly was reported in 3 cases of 22q11.2 deletion. Apart from congenital heart defects, the commonest structural anomalies present were cleft soft palate (4 cases) and absent thymus (3 cases).

Table 1. Types of heart defects associated with 22q11.2 deletion

Heart Defect	Frequency	Number resulting in live birth
Ventricular septal defect	15	13
Tetralogy of fallot	6	5
Interruption of aorta	6	6
Truncus arteriosus	4	1
Atrial septal defect	4	4
Double outlet right ventricle	3	3
Pulmonary valve atresia	3	3
Pulmonary artery atresia	3	2
Major aortopulmonary collateral arteries	3	2
Patent foramen ovale (in term babies)	3	3
Pulmonary valve stenosis	2	2
Overiding aorta	2	2
Persistent right aortic arch	2	2
Pulmonary artery stenosis	2	2
Enlarged right atrium and/or ventricle	2	2
Patent ductus arteriosus (in term babies)	2	2
Bicuspid aortic valve	2	2
Aortic valve stenosis	1	1
Subaortic stenosis	1	1
Hypoplasia of aorta	1	1
Dysplastic pulmonary valve	1	1
Persistent left superior vena cava	1	1

Table 2: Comparison of q2211.2 deletion with/without heart defects

	CDH without heart anomalies	CDH with heart anomalies
Antenatal detection of q2211.2	20% (2/10)	23% (7/30)
Survival to 3 years (1998-2008 live births)	75% (6/8)	78% (21/27)

Conclusion: 22q11.2 deletion commonly presents with congenital heart anomalies. Karyotyping for the deletion should be offered to babies presenting with heart disease. Antenatal detection of the deletion and survival is similar for all cases irrespective of the presence of congenital heart anomalies.

P-144

Pulmonary hypertension in preterm infants admitted to a neonatal intensive care unit

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Aim of study is to evaluate incidence of Bronchopulmonary dysplasia (BPD) and identify possible risk factors for developing BPD and Pulmonary Hypertension(PH) in infants with BPD.
Methods: Medical records of premature infants admitted in the period 2002-2010 were rewieved. 402 infants identified with a birth weight (BW)<1500 g. Eightytwo had BPD (need of oxygen at 28 postnatal days). Eight were excluded. The remaining 74 infants with BPD were evaluated. Differences in infants with and without BPD, with and without PH, with mild BPD and with moderate/severe BPD were evaluated with a student t–test, chi-squared test and multivariate analysis.
Results: Incidence of BPD was 18%. BPD infants had significantly lower gestational age(GA) and BW than infants without BPD. Infants with BPD compared to infants without BPD had more often; intubation at birth, mechanical ventilation within 24 hours of birth, treatment of patent ductus arteriosus and laser treatment of retinopathy of prematurity (ROP), lower apgar score, days of CPAP treatment and days of mechanical ventilation. Fifty percent of infants with BPD developed moderate/severe BPD. Infants with moderate/severe BPD differed significantly from infants with mild BPD with respect to mechanical ventilation within 24 hours of birth, treatment with diuretics and laser treatment for ROP. Thirtyeight percent of infants with moderate/severe BPD developed PH versus 8% of infants with mild BPD. The incidence of PH was 4% amongst premature infants with BW<1500 gram and 23% amongst infants with BPD. Days of CPAP treatment, days of oxygen therapy, number of infants receiving diuretic therapy, intubation at birth and postnatal infection was higher among infants with PH and BPD, compared to infants with only BPD. Surfactant therapy and mechanical ventilation within 24 hours of birth were significantly associated with developing PH in BPD infants.
Conclusions: Invasive care strategies with excessive oxygen, mechanical ventilation and episodes of postnatal infections seem to aggravate lung function in premature infants and may lead to pulmonary hypertension in infants with BPD. More research in development and treatment of BPD complicated with PH is warranted. Regular screening with echocardiography for PH in infants with BPD is required.

P-145

Usefulness of telemedicated echocardiograms for neonates admitted to NICU

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Introduction: Recent advances in telecommunication technology have made it possible to transmit echocardiographic images over long distances. This technology may be beneficial to newborns with heart defects who are born in primary care centers located far from pediatric subspecialty centers. The purpose of this study was to investigate the usefulness of telemedical cardiography in the diagnosis of congenital heart disease of neonates.
Methods: We reviewed all telemedical diagnosis/consultation obtained from neonates in NICU between January 2007 and December 2011 at Juntendo Shizuoka Hospital retrospectively. A telemedical link was created using internet line (iChat from Mac), between Shizuoka Children's Hospital (pediatric cardiology site), and Juntendo University Shizuoka Hospital (primary care site). Neonates with possible cardiac disorders were identified either by genecologists or pediatricians at primary care settings, then later requested telemedical cardiogram.
Results: Telemedical echocardiograms were performed for 23 neonates. 21 neonates (91%) were transferred to the pediatric cardiology site. Eventually all of them were diagnosed properly:1 diagnosed before birth; 14 done by primary pediatricians' cardiograms; 5 remotely done; 2 done at subspecialty centers. Immediate changes in management were required in 15 (65%) neonates including emergency transportations: 6 transferred by newborn ambulance; 4 transferred by helicopter. 10 neonates

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(48%) were transfered in shock. Out of those 10 neonates, 9 neonates were in the need for prostaglandin E1, to obviate or for treatment of ductal shock. One neonate had to be treated with catecholamine to maintain adequate blood pressure. All neonates were safely sent to the pediatric cardiology site, and were able to receive further special treatments and follow up.
Conclusions: Telemedical echocardiogram provided accurate diagnostic information in neonates. It is suggested that an adequate initial management can be performed for sick neonates with congenital heart failure in primary care setting without cardiology specialists by using this technology.

P-146

Prognostic role and diagnostic accuracy of Urinary Neutrophil Gelatinase-Associated Lipocalin in the Earl Diagnosis of Acute Renal Damage After Pediatric Cardiac Surgery

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Background: Neutrophil gelatinase-associated lipocalin (NGAL) is gaining consensus in the early diagnosis of acute renal injury (AKI) in various pathological settings in adult patients, including cardiac surgery. Conversely, the diagnostic accuracy of urinary NGAL related to AKI syndrome after pediatric cardiac surgery is still controversial.
Methods: We prospectively enrolled 148 children (0-18 years, median 36,4 ± 55,33 months) undergoing surgery at our Institution for correction/palliation of congenital heart defect (CHD). Urinary samples for NGAL measurement (with ARCHITECT platform, Abbott Diagnostics) were collected pre-operatively, at 2, 6 and 12-hours post cardiopulmonary bypass. Brain natriuretic peptide (BNP) was measured basally and at 12-36 hours (with Triage reagents, Access Immunoassay Systems, Beckman Coulter, Inc.). The presence of AKI was defined in accordance with both RIFLE and AKIN guidelines.
Results: The peak of urinary NGAL values (mean 183.0□ 432.4 ng/L, range 0.30-3888 ng/L) occurred within 6 hours, being most common at 2 hours after surgery. The peak of NGAL occurred significantly earlier than that of serum creatinine () ($P < 0.001$). NGAL peak values positively correlated with Aristotle score ($P = 0.003$), body surface area ($P = 0.002$), time of intubation ($P < 0.001$), intensive care unit stay ($P < 0.001$) and cardiopulmonary bypass ($P < 0.001$). However, after multiple regression analysis only cardiopulmonary bypass time ($R = 0.607$, $P < 0.001$) and BNP at 12-36 hours values ($R = 0.312$, $P < 0.001$) were significantly related to NGAL peak values. Finally, the diagnostic accuracy of urinary NGAL peak values for AKI showed an AUC ROC curve of 0.82 with an optimal cut-off value of 80 ng/L (sensitivity 0.71, specificity 0.85, predictive positive value (PPV) 0.76, predictive negative value (NPV) 0.81). NGAL peak values tended to have better diagnostic accuracy than the NGAL values of other samples collected at 2, 6 and 12 hours after surgery, respectively (Table-1).

Table-1: ROC Curve for the diagnosis of AKI of U-NGAL measurements at various time intervals.

SAMPLE	AUC	Specificity	Sensitivity	PPV	NPV
NGAL peak	0,82	0,85	0,61	0,76	0,81
NGAL 2 hours	0,81	0,83	0,71	0,75	0,81
NGAL 6 hours	0,78	0,68	0,81	0,64	0,84
NGAL 12 hours	0,76	0,81	0,61	0,68	0,76

Conclusions: Our data suggest that urinary NGAL provides significant diagnostic and prognostic information in children undergoing surgery for CHD.

P-147

Extra-cardiac malformations, chromosomal abnormalities and clinical syndromes in neonates with congenital heart disease

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Introduction: Congenital heart disease (CHD) is frequently associated with extra-cardiac malformations, chromosomal abnormalities and recognised clinical syndromes. The management of the heart lesion may be influenced by the treatment needed for the associated anomalies and vice versa. A multidisciplinary approach may be warranted and counselling of the family regarding long-term outcome should be modified accordingly. The aim of this study is to assess the incidence of extra-cardiac malformations, chromosomal abnormalities and clinical syndromes in neonates with congenital heart disease.
Methods: Data on all neonates referred for echocardiography to our institution were collected prospectively. Infants with abnormal echocardiograms were followed up in clinic and were managed appropriately. Information regarding the presence of chromosome abnormalities, recognised syndromes and extra-cardiac defects was recorded. The classification of birth defects was performed using the EUROCAT (European Surveillance of congenital anomalies) Registry Guide 1.3.
Results: Over a period of 5.5 years (6/2006-12/2011) 866 neonates (aged 1-28 days) were referred for echocardiography, of which 305 had CHD (35.2%). Two hundred fifty nine patients (84.9%) had isolated CHD. Nineteen (6.2%) had chromosome abnormalities, 5/305 (1.6%) had clinically recognised syndromes and 22/305 (7.2%) had extra-cardiac anomalies not associated with genetic abnormalities or clinical syndromes Among those infants the indication for echocardiography was the identification of the extra-cardiac lesion or suspected syndrome in 21/46 (45.6%). The most common genetic or clinical syndrome associated with CHD was Trisomy 21 (13/24, 51.2%). In this group the most prevalent cardiac anomaly was atrioventricular septal defect (7/24, 21.2%), whereas in infants with extra-cardiac anomalies not associated with genetic or clinical syndromes the most common diagnosis was interventricular septal defect (11/22, 50%). Anomalies of the gastrointestinal system were the most prevalent in the latter group (7/22, 31.8%).
Conclusion: About 15% of neonates with CHD have extra-cardiac anomalies, chromosomal abnormalities or recognised syndromes. Neonatologists and paediatric surgeons have a low threshold for echocardiographic referral of patients with extra-cardiac anomalies or syndromes associated with increased risk of CHD.

P-148

Results of total correction of complete atrio-ventricular canal (CAVC) depending on coexistence of Down syndrome in children.

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CAVC is predominantly seen in Down syndrome (DS) patients (70%), with correction results in DS and non-DS patients

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ambiguous due to differences in anatomy and clinical course. The objective was assessment of CAVC correction results and prognosis depending on concomitant DS. Clinical material: 131 patients, with 108 DS–G1 and 23 normal karyotype subjects -G2, operated on from 1990 to 2003.
Methods: All patients were subjected to physical examination, ECG, CXR and ECHO. Testing was done postoperatively with the mean follow-up of 9.2 years. Statistical analysis: descriptive statistics, t-Student test, Yates's χ^2 test and Kaplan-Meier survival curve were analyzed.
Results: Type A was more common in G2 (69.6% vs. 48.6%, $p = 0.1$), type C in G1 (32.1% vs. 17.4%, $p = 0.2$). Qp/Qs in G2 was significantly higher than in G1 (3.5 vs. 2.4, $p < 0.05$), as well as significant common A-V valve regurgitation (AVVR) (34.8% vs. 25%). Age at surgery was $x = 8.8$ months. Postoperatively, significant left AVVR was more common in G2 (17.6% vs 34.8%). Significant residual VSD was equally common (4.6% vs. 3.8%), as was LVOTO (2.8% vs. 4.3%). Eighteen patients (13.7%) died: nine–within 30 days postoperatively and nine–at the mean time of 10.1 months. Mortality rates in G1 were non-significantly higher (14.8% vs. 8.7%, $p = 0.6$), due to infections, CHF and pulmonary hypertension crisis. Early mortality risk factors were younger age at surgery ($x = 5.4$ vs. $x = 9.0$ months, $p < 0.05$), lower body mass ($x = 4.0$ kg vs. $x = 5.6$ kg, $p < 0.05$) and concomitant PDA (77.8% vs. 27.6%, $p < 0.05$). Reoperations included 15 patients (G1: 11.1%, G2: 13%) due to left AVVR, LVOTO and/or residual VSD. The 10-year survival for was 87.3%, being higher in G2 (85.2% vs. 91.3%). Survival without reoperation was seen in 88.5% children, being comparable in both groups (87% vs. 88.9%).
Conclusions: CAVC morphology is more favorable for repair in DS children, however higher susceptibility for infections and risk of pulmonary hypertension crisis may affect clinical course and early mortality. 10-year survival and freedom from reoperation are similar in both groups but incidence of significant left AVVR may result in more frequent future re-operations in non-DS patients.

P-149

Outcome Of Critical Chd: Comparison Between Prenatal And Post-Natal Diagnosis In 548 Neonates

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Introduction: Critical congenital heart diseases (CHD) account for 2–3 /1000 of birth malformations and about 50% of CHD. Data from literature report that prenatal diagnosis may improve outcome of neonates with transposition of great arteries (TGA) and left-heart side obstructions. Report the outcome of patients with critical CHD admitted in our Division (Pediatric Cardiology/ Cardiosurgery) in eleven years of activity (Jan 2000-2011), comparing when they were diagnosed in utero and in post-natal life.
Methods: We reviewed 548 neonates with CHD (TGA: 112/548, 20.4%, AoCo: 128/548, 23.3%, PAIVS 85/548, 15.5%, HLHS 54/548, 9.8%). Mean age at admission was 3.0 + 2.1 days (range 0-30 days): Among them, 240/548 (43.8%) received prenatal diagnosis of critical CHD: 48.2% systemic ductal-dependent CHD, 44.8% pulmonary ductal-dependent CHD and 42.8% of transposition of great arteries.
Results: As first treatment, 323/548 neonates (59.0%) were submitted to a percutaneous procedure, 225/548 neonates (41.0%) were submitted to cardiothoracic intervention. Overall mortality was: 100/548 (18.0%). If we exclude neonates with HLHS, this data drop to 12,5% (64/512). At a mean follow-up of 5,8 + 1,4 y (range 0-11 y), we recorded a mean overall survival of 74.2%. We divided neonates which received prenatal diagnosis (G1) from those which didn't receive it (G2). G1 pts were amitted in 1st day of life ($P = 0.03, <0.05$). We report a pre-surgical mortality of 2.0% in G1 and 9.0% in G2, $P = 0.02$ (<0.05). We report early survival of 90.0% in G1 and 80.0% in G2, P 0.04 (< 0.05)
Conclusions: Prenatal diagnosis of critical CHD allows to plan the delivery, the 3rd level centre admission, early-starting PGE2 infusion, to prevent metabolic and respiratory acidosis, so improving clinical pre-surgical conditions. We report an overall survival of 75% at 5 yrs (included complex CHD) of patient with critical CHD, if diagnosed and treated in a 3rd level pediatric cardiology and cardiosurgery centre. Early survival of neonates with prenatal diagnosis in our series is about 90%. In conclusions, prenatal diagnosis of critical CHD statistically improve the outcome of affected neonates.

P-150

Patent Ductus Arteriosus and Pulmonary Hypertension in a Child with 49, XXXXY Syndrome

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Introduction (or Basis or Objectives): 49, XXXXY syndrome is a rare sex chromosome aneuploidy disorder. It was first reported in 1960 by Fraccaro et al. and has an approximate incidence of 1 in 85000 male births. 49, XXXXY syndrome have emphasized the "classic triad" of mental retardation, radioulnar synostosis and hypogonadism.
Case Report: A 5-month-old boy was admitted for poor feeding, heart failure and convulsion. Patient was treated by oxygen with hood, dopamine, and dobutamine for heart failure, antibiotics for suspected bronchopneumonia and phenytoin for repeated convulsions. He was born as a 19 year old mother's first child at term via caesarean section, with birth weight of 2250 gr, body length of 43 cm, head circumference 32,5 cm. He had dysmorphic facies including hypertelorism, short palpebral fissures, low nasal bridge, broad nose, dysplastic ears, cleft palate, micrognathia, short neck, micropenis, unilateral cryptorchidism. Cardiac auscultation reveals a third degree heart murmur on lower left sternal border. His thyroid function tests were normal. Cranial MRI showed both lateral dilated ventricles. His karyotype analyses was 49, XXXXY (Fig).
Echocardiography revealed a large patent ductus arteriosus (PDA). Angiography demonstrated PDA, measuring 2.5 mm at its narrowest diameter, and shunting left to right. Hemodynamic parameters were as follows: right atrium mean 11 mmHg, pulmonary artery mean pressure (MPA) 57 mmHg, aorta mean 63 mmHg. 5x 4 mm Amplatzer Ductal Occluder-I was successfully deployed through retrograde way. The MPA mean pressure decreased slightly to 40 mmHg immediately after the intervention.
Conclusions: The first case reported by Fraccaro et al. was a child with PDA. The prevalence of congenital heart disease among patients with the 49, XXXXY syndrome is 14%. PDA is the most common congenital heart defect in 49, XXXXY. Although our case was 5 month-old and had a medium PDA, severe pulmonary hypertension was established. There was not a risk factor for

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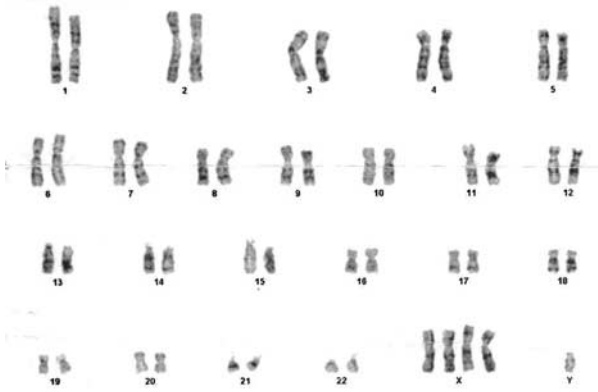
primary pulmonary hypertension. In fact, unless there is a very large PDA, pulmonary hypertension will not develop in a non-syndromic 5 month-infant. We think that, pulmonary hypertension develops faster in 49, XXXXY syndrome than PDA with normal population. Consequently, PDA closure should be made in early period of life for these patients.



P-151

Central Blood Pressure and Augmentation Index in the Pediatric Age-Group

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Objectives: Non-invasive approach in the measurement of arterial compliance has made this method applicable for the examination of larger populations. In adults arterial compliance already emerged as an important marker of vascular health. However, there is a lack of information in children. Therefore, this study aimed to determine Augmentation Index (AIx) and aortic systolic blood pressure (aSBP) in the pediatric age-group.
Patients and methods: From February 2011 to November 2011 we examined 249 healthy children with a median age of 12.7 years (128 girls, Inter quartile range (IQR): 11.8–14.1 years) and with a slightly increased BMI-SDS of 0.78 (-0.37 –2.08). Measures of aterial compliance were obtained after 5 minutes rest in supine position based on an oscillometric method (Mobil-O-Graph, IEM, Stolberg Germany) using circumference depending cuffs at the upper arm.
Results: Median aSBP was 101 mmHg (95–108 mmHG). Aortic systolic blood pressure was associated with higher BMI-SDS ($r = .415$; $p < .001$) and was also higher in older subjects ($r = .330$; $p < .001$). Augmentation index in the whole study group was 12% (9–20%). No associations to BMI-SDS ($p = .891$) or age ($p = .865$) could be found.
Conclusions: Children with higher BMI-SDS seem to be at early risk to manifest higher aSBP. Cut-off values for aSBP and the role of the augmentation index has to be clarified in further research.

P-152

First results of "Catch the (pulse) wave"–Is pulse wave velocity in children feasible to measure with an oscillometric device?

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Objective: Measuring arterial stiffness parameter by pulse wave velocity (PWV) is recommended by the European Society for Hypertension and European Society of Cardiology. PWV is already well established for risk stratification in adults. However, there is a lack of information in children. Fast and easy measurement devices are especially attractive for risk screening in pediatric populations. Therefore the aim of our study was to assess the feasibility of an oscillometric device (Mobil-O-Graph, IEM, Stolberg, Germany) in evaluating vascular stiffness parameters in children.
Patients and methods: In November 2011, 95 school children with a median age of 14.0 years (42 girls, range 8.1 – 19.7 years) without severe diseases were included into the study. The measurements took place in a pediatrician practice. Cuffs were chosen in dependence of upper-arm circumference (5 different cuffs were used). The measurements start after 5 minutes of rest in supine position on the left upper arm.
Results: Valid measurements were obtainable in 91 of the 95 children (96%). There was a significant increase of the PWV with age ($r = .215$, $p = .041$) and BMI-SDS ($r = .281$, $p = .007$).
Conclusions: The measurement of pulse wave velocity using the Mobil-O-Graph device is also feasible in children. Since our data indicates that PWV increases with age and BMI-SDS the aim of future research needs to be establishment of references values for children and adolescents.

P-153

The prevalence of persistent Left Superior Vena Cava in patients with and without congenital heart disease

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Background: The persistent left superior vena cava (LSVC) is one of the common anomalies of the systemic veins. It usually drains to the coronary sinus. The prevalence of LSVC in the general population is between 0.1–0.5%. It is more common in patients with congenital heart disease (CHD) (3-5%).
The importance of detecting LSVC prior to cardiac surgery is paramount for the management of the systemic veins canulations. Failure to detect its presence before surgery may lead to some difficulties and prolongation of the intervention.
Objective: To estimate the prevalence of persistent LSVC in patients with or without congenital heart disease in our patient population.
Methods: All echocardiographic studies and data of patients seen in our hospital between December 2010 and November 2011 were reviewed for presence of persistent LSVC. The demographic data, CHD diagnosis, presence of LSVC and associated diseases or syndromes were documented. The incomplete studies were excluded from analysis.
Results: The total no of screened patients were 1802, there was no gender predominance. Patients with no CHD made 53% of our population, while those with CHD were 47%. The prevalence of LSVC in the healthy children was 0.9% and in those with CHD was 7%. Down syndrome was one of the most commonly associated syndromes. The most common cardiac lesion associated with LSVC was complete atrio-ventricular-septal- defect. In all but one patient, the LSVC drained to the coronary sinus.
Conclusion: The prevalence of LSVC in the current population is higher than reported in the literature. Down syndrome have higher incidence of LSVC, especially patients with complete atrioventricular septal defect.

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P-154

The Role of N-Terminal Pro-Brain Natriuretic Peptide in the Diagnosis and Management of Congestive Heart Failure due to Dilated Cardiomyopathy in Children

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Introduction (or Basis or Objectives): N-Terminal ProBrain Natriuretic Peptide (NTProBNP) is a neurohormone which is stored mainly in myocytes of the ventricles and released as a result of pressure and volume overload or myocardial damage. Recently natriuretic peptides were revealed to be useful in the diagnosis, follow-up and in the predicting the degrees of heart failure in children and adults.
Methods: 59 patients; 37 in control (aged 1months-14 years) and 22 patients (aged 2 months-14 years) with acute congestive heart failure with dilated cardiomyopathy (DCMP) were enrolled. Patients with renal and hepatic diseases were excluded. Ross scoring was used in grading heart failure. Scoring was done and NTProBNP levels were measured in admission and 7th day of treatment.
Results: Patients were classified into 4 groups according to Ross score before and after treatment.
Cut-off level of NT-ProBNP was determined after the treatment for study group. Strong relationship was found between degree of heart faiulure and NTProBNP levels (Fig), but not with echocardiographic findings (EF, FS, LVMI, LVEDD) and cardio-thoracic index on telecardiography. There wasn't significant difference between NT-ProBNP levels and age-gender.

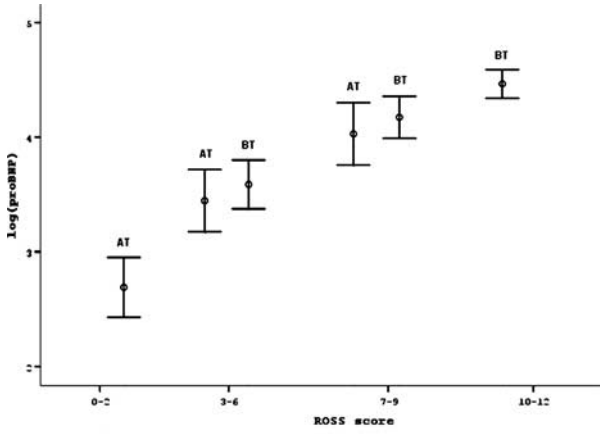

Fig: Ross score and NT-ProBNP levels before and after the treatment for study group

Conclusions: NT-ProBNP was sensitive in grading of heart failure due to DCMP. It could be an alternative method for diagnosis and treatment of heart failure due to DCMP.

P-155

The Role of the Leptin-to-Adiponectin Ratio in Cardiovascular Prevention in Childhood.
First results from the "Get Fit – Stay Healthy" Project

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Objective: The development of cardiovascular disease begins in childhood [1], with structural and functional changes of the endothelium already existing in obese children [2]. But it is incompletely understood which factors are involved in the pathogenesis of atherosclerosis and further the protective mechanisms to obtain endothelial health are still unclear.
However the improvement of prevention strategies relies on a better understanding of which risk factors are linked to early subclinical atherosclerotic changes and thereby of allowing for the identification of children at high cardiovascular risk.
Patients and Methods: Therefore we investigated 117 healthy school children (52 female) aged in the median 12.3 years (interquartile range: 11.9 – 12.9 years) with a median body mass index standard deviation score (BMI-SDS) [3] of 0.06 (interquartile range -0.93–1.18). The children took part in the "Get-fit – Stay healthy" project, starting off with baseline measurements from April 2011 to October 2011. Thirty children (25.6%) with a BMI-SDS >1 were defined to be at cardiovascular risk. We examined waist circumference, waist-to-height ratio, blood pressure and serum lipids. All children had ultrasound sonography (ProSound alpha7, Hitachi/Aloka) on carotid intima media thickness (IMT), carotid artery compliance and stiffness.
Results: Univariate analysis showed that children at risk had higher leptin-to-adiponectin ratio ($p < 0.001$), a lower high-density lipoprotein ($p < 0.001$) and higher triglycerides ($p = 0.001$). There was no difference in low-density lipoprotein ($p = 0.751$), total cholesterol ($p = 0.284$) and systolic blood pressure ($p = 0.268$). Carotid IMT ($p = 0.845$) and stiffness ($p = 0.191$) as well as compliance ($p = 0.374$) did not differ. Leptin-to-adiponectin ratio remains as only independent risk factor ($p < 0.001$) in the multivariable model.
Conclusions: Leptin-to-adiponectin ratio is a strong independent marker of cardiovascular risk already in this age group. Its impact exceeds that of other blood lipid markers and systolic blood pressure. Early negative structural or functional changes towards a reduction in stiffness and compliance as well as an increase of the intima-media complex could not be shown.

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P-156

Cardiac rhabdomyomas associated with tuberous sclerosis complex in childhood in 11 cases

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Introduction: Cardiac rhabdomyomas (CRs) are the most common pediatric heart tumor and closely associated with tuberous sclerosis complex (TSC).
Methods: We reviewed our patients with CRs who received a diagnosis of TSC previously or during follow-up between June 1996 and January 2012. We evaluated the presentation type, clinical course and outcome of all the cases diagnosed both prenatally and postnatally. All the medical data of the patients obtained from their medical records and computerized database in our hospital, retrospectively.

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Results: 11 (% 34) of 32 patients with TSC had a total of 29 CRs. The median follow-up period was 2 years (range: 15 days-15 years). Five patients (% 45) had multiple tumors. Clinical presentation was cyanosis in two patients and cardiac murmur in three patients. Six patients were asymptomatic and CRs detected while cardiac evaluation for TSC. Prenatal screening revealed intracardiac tumors in two patients who received diagnosis of TSC in the follow-up. One of them had single cardiac tumor, epilepsy and supraventricular tachycardia due to Wolff-Parkinson-White syndrome and rhabdomyoma regressed spontaneously in two years. The other patient who had prenatally diagnosed had multiple CRs with significant intracardiac obstruction and ventricular tachycardia. Because he was accepted inoperable, he managed medically (mTOR inhibitor) and obtained rapid response to the treatment initially. Two patients (% 18) underwent cardiac surgery in the infancy period because of hemodynamically significant obstruction and their tumors removed totally. Six of the 29 CRs (% 21) showed complete regression spontaneously in three patients.
Conclusions: CRs are common in the patients with TSC and although they are pathologically benign tumors, sometimes they can cause life threatening events in this population. Thus, cardiac screening of the patients with TSC and fetuses or children of the parents with TSC can reveal significant pathologies in these patient groups.

P-157
What's the outcome of carditis in Children with Sydenham's Chorea

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Introduction: Acute rheumatic fever (ARF) is a common cause of heart disease in our country. The carditis is most common major findings accompany Sydenham chorea (SC). However there are very limited studies which evaluating outcome of carditis in patients with chorea.
Methods: We screened all patients diagnosed with ARF, between 1999 and 2011 years, retrospectively. We evaluated only children diagnosed with SC at the first attack of ARF. To determine frequency and outcome of carditis in patients with SC, we evaluated their clinical and echocardiographic features at the initial and follow-up examinations.
Results: A total of 551 cases were diagnosed with ARF within the study period and SC was established in 69 cases (12.5%). SC was remarkably more common in pre-adolesants girls. Half of patients had bilateral chorea at presentation. At the initial examination; 20 patients (28.9%) had a normal echocardiogram. The diagnosis of silent carditis was established in 20 (28.9%) of cases and clinical carditis was established in 29 (42%) of patients. Pure mitral regurgitation was detected in 36 (73.4%) of patients with carditis. Carditis was mild in 41 patients (83%), only one case had severe carditis with third degree mitral and aortic regurgitation. 63 cases were followed from one to ten years. Improvement rate of valvulitis in cases with silent carditis (29.4%) was not different than cases with clinical carditis (18.5%) ($p > 0.05$). Persistence of valvular pathologies occurred in 72.2% of cases with carditis in the long-term follow-up (>2 years). Most patients (88.8%) complied with secondary prophylaxis, so relapse of carditis was exclusively prevented in our patients. Recurrence of chorea was identified in 20.6% of cases and not associated with clinical or laboratory evidence for streptococcal reinfection.
Conclusion: Patients with chorea usually had mild carditis and it showed resolution during follow-up. Relapse of carditis was exclusively prevented with secondary prophylaxis in our population. Recurrence of chorea was not rare, despite regular treatment with benzathine penicillin.

P-158
Controversies in Prophylaxis and Treatment of Post Surgical Pericardial Syndromes: a Critical Review with a Special Attention to the Paediatric Age

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Background: post-surgical pericardial syndromes (PSPS) are common complications after cardiac surgery, however their pharmacological treatment remains controversial. We reviewed the accuracy and limits of clinical trials over prophylaxis and treatment of PSPS with the aim to identify (if feasible) a therapeutical flow-chart.
Methods: A computerized literature search in the National Library of Medicine using the keywords pericardial effusion +/- cardiac surgery +/- paediatric/congenital.
We redefined the research adding separately the keywords *post-pericardiotomy syndrome (PPS), anti-inflammatory-drugs, non-steroidal anti-inflammatory-drugs (NAIDs), steroids, and colchichine.* Case reports, and studies not entirely constituted by post-surgical patients were excluded.
Results: We identified 12 clinical trials for PSPS prophylaxis (8 works) and treatment (4 works). Three majors classes of agents have been tested: NSAIDs, corticosteroids and colchicine.
Studies regarding PSPS treatment present many limitations: end-points were not homogeneus (not allowing a metanalysis), only a few agents (NAIDS and corticosteroids) have been tested in randomized controlled trials (RCT), some studies considered only specific age/disease sub-groups, and different agents have never been compared in the same protocol.
NSAIDs have been employed in 3 RCT in adults with contrasting results (2 studies proved benefits, 1 did not), while no studies were conducted in children.
Despite a widespread use of steroids in cardiac surgery, especially in children, only one small paediatric study instead proved prednisone effectiveness in PPS treatment.
As far as PSPS prophylaxis is concerned, robust evidences from two wide RCT support and a recent metanalysis support the role of colchicine in adults.
On the contrary prophylaxis with NAIDs and corticosteroids failed to prove significant advantage in children, while a few data are available for adults.
Conclusions: Evidences for the treatment of PSPS are fragmentary and incomplete. As a result, on the basis of actual knowledge, it results difficult to understand when to treat and which agent to employ, especially in children. Treatment is usually based on personal experience and institutional practice with NAIDS generally employed first and steroids reserved for more severe forms of PSPS or recurrences.
Further wider and multi-agents studies are advised in order to establish a therapeutical flow-chart for the treatment of PSPS.

P-159
Risk factors for cardiovascular disease in school age children and teenagers

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Objectives: To detect and to prevent cardiovascular risk factors as: overweight, obesity and hypertension in school age children and teenagers.
Methods: We investigated 3747 pupils, 6-18 yo., from different schools from Timisoara, measuring: weight, height, BMI and blood pressure. A questionnaire regarding nutritional behavior and physical activity was completed.
Results: There were 2150 girls and 1532 boys in the study, 2744 (73%) from the city and 1003 (27%) from the village. Overweight was found in 20.66% boys and 17.6% girls. Obesity was found in 16.7% boys and 9.16% girls. Overweight and obese boys were equal in both areas, but obese girls were less in town girls. Hypertension was detected in 399 urban children (10.65%) and only in 40 (3.98%) of rural. From the urban area, 47.6% were girls and 52.4% were boys with hypertension. From the village area, 32.5% were girls and 67.5% were boys with hypertension. Increased BMI was found in 172 hypertensive children from town, and in only 17 hypertensive children from village. Overweight in hypertensive children was found in 61 boys and 35 urban girls, in comparison with only 8 boys and 2 village girls. Obesity was present in 51 hypertensive boys and 25 town girls, in comparison with only 1 hypertensive boy and 6 hypertensive girls from the village. The values of hypertension were analyzed with charts. Fast food eating, sandwiches, snacks and sweet drinks were preferred by children from town. Playing outside the house was more frequent in the village group. PC and static play was more frequent in the town group.
Conclusions: Obese boys were dominant in urban boys group. Girls were more interested in better look than boys. Hypertension was detected especially in urban children. Children from village eat healthier and do more physical activity. Risk factors for cardiovascular disease are dramatically high in urban children, especially boys, because of fast food eating, snacks, sweet drinks and sedentariness. After this study, a web site for informing and instructing the children and their families, regarding the cardiovascular risk prevention was promoted by our hospital in the schools from Timisoara.

P-160
Neurodevelopmental outcome at 2 years corrected age following highly conservative treatment of the patent ductus arteriosus in extremely low gestational age neonates (ELGANs).

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Objectives: There is no consensus on the optimal management of the patent ductus arteriosus (PDA) in ELGANs. Although no single intervention has clear demonstrable clinical benefit, treatment for PDA is widespread. There is a lack of long-term outcome data when PDA management is highly conservative. We describe the clinical and developmental outcomes at 2 years corrected age in a population of ELGANs in which echocardiographic assessment and treatment of PDA were extremely rare.
Methods: The outcomes of all infants born at less than 28 complete weeks gestation (range 23 + 1 to 27 + 6 weeks) between January 2006 to June 2009 and admitted on their day of birth to a single tertiary NICU were ascertained (Simpson Centre for Reproductive Health, Edinburgh). PDA treatment was noted. Clinical and developmental assessment outcomes at 2 years corrected age were obtained using a standardised proforma based on the Health Status Questionnaire and reported according to BAPM 2008 classification. Where cognitive outcomes are described, assessment was with Bayley-III scales for infant development.
Results: 115 ELGANs were admitted. 21 died before discharge and 1 died in infancy. Of the surviving 93 infants, none were treated with COX inhibitors, one had primary surgical PDA ligation and another had PDA ligation in infancy. Both children's two-year outcomes are known. Two-year follow up data are not available for 15/93 children. At discharge from NICU in those 15, two had PVL (one of whom also had laser treatment for ROP), one more had laser for ROP, none had IVH grade >2 and one received home oxygen. Two-year outcomes for the remaining 78 are as follows:

	No significant disability			Neurodevelopmental impairment		
Impairment	None	Mild	%	Moderate	Severe	%
Motor ($n = 78$)	70	2	92.3	3	3	7.7
Hearing ($n = 78$)	74	1	96.2	2	1	3.8
Vision ($n = 78$)	75	0	96.2	3	0	3.8
Language ($n = 76$)	61	4	85.5	7	4	14.5
Cognition ($n = 40$)	37	0	92.5	3	0	7.5

Conclusions: Survival and neurodevelopmental outcome at 2 years corrected age in a population of ELGANs in which diagnosis and treatment of PDA were highly selective are favourable. Randomised controlled trials of PDA treatment are still required.

P-161
Treatment strategies for Pulmonary Sequestration in Childhood: resection, embolization, observation?

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Objective: The ideal treatment strategy for pulmonary sequestration in the pediatric population, whether resection or embolization, is not clearly defined. In our institution both therapies were performed depending on point of entry. The aim of this study was to assess the outcome of these different management strategies.
Methods: Retrospective, single centre review. Local pediatric and cardiology databases of our tertiary referral center were used to identify patients. The main inclusion criterion was the established diagnosis of a pulmonary sequestration. Patients (n = 48) were divided into three groups based on treatment received: conservative management (n = 5), surgery (n = 22) and embolization (n = 21).
Results: Diagnosis was made by fetal ultrasound (n = 9), routine ultrasonography (n = 20), computerized tomography (CT), mostly CT-angiography, (n = 18) or arteriography (n = 1). Respiratory symptoms, i.e. recurrent chest infection, were the most common clinical presentation in the surgical group, whilst cardiac failure was the most frequent symptom in the embolization group ($p < 0.01$;

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95% CI: 0.3 to 0.9). The median age at treatment was 8.0 months (range: 1.2–166.0) in the surgical group, 4.0 months (range: 0.2 - 166) in the embolization group and 8 mo (range: 0.3 – 197.0) in the conservatively managed group. There was no significant difference between the ages of the former two groups ($p = 0.9$). There were six complications in the surgical group and one in the treatment group ($p = 0.1$). Outcomes in both groups were comparable with good results on follow-up. Failure of embolization, due to the sharp angle of take-off of the vessel precluded safe coil placement in one patient. In this case a vascular clip was placed surgically on the anomalous vessel during cardiac surgery for the underlying cardiac lesion.
Conclusion: Both surgery and endovascular embolization are equally effective and safe treatments for pulmonary sequestration. The presenting symptoms dictates therapy: surgery in case of infection and embolization if a shunt needs to be abolished. Our institutional policy remains unchanged.

P-162
Combination of Bosentan and Sildenafil therapy in patients with congenital heart defects (CHD): A case series
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Background: Data on the effects of an oral combined pulmonary vasodilator therapy in patients with CHD are scarce. Our aim was to evaluate a possible benefit of a combined therapy compared to a monotherapy with either Bosentan or Sildenafil in different forms of operated CHD.
Methods: Seven patients (median age 13.5 (5.2–27.9) years, 3 f) treated with either Sildenafil or Bosentan were retrospectively analyzed before and during a combination therapy with both Bosentan and Sildenafil. Underlying diagnosis were protein losing enteropathy following Fontan operation ($n = 2$) and pulmonary hypertension secondary to CHD (PAH-CHD) ($n = 5$), patients with Eisenmenger syndrome were excluded : atrioventricular septal defect ($n = 1$), atrial septal defect ($n = 2$), ventricular septal defect ($n = 2$). Invasive pulmonary artery pressures (PAP) and PA resistance, oxygen saturation at rest, NYHA functional class, 6 minute walking distance (6MWD), NT-pro BNP were investigated before start of combination therapy and at latest follow-up. Mean doses of Bosentan were 2.8 ± 1.5 mg/kg/d and Sildenafil 1.7 ± 0.7 mg/kg/d. Mean follow up time was 2.5 years (0.4–5.2 years).
Results: Mean PAP in PAH-CHD patients decreased from 62 ± 23 to 48 ± 22 mmHg, PA resistance decreased from 18 ± 11 to 14 ± 11 U$\star$m^2 (n.s.). Protein losing enteropathy improved with discontinuation of Albumin replacement. NYHA functional class of all patients improved from 2.4 ± 0.5 to 1.7 ± 0.7 ($p = 0.003$). Oxygen saturation increased from $91 \pm 9\%$ to $94 \pm 3\%$ (n.s.). 6MWD did not improve significantly over time (534 ± 53m vs. 545 ± 37m). NT-pro BNP levels also did not change. In one patient the Bosentan dose had to be decreased because of elevated liver enzymes, no other major side effects were noted.
Conclusion: The combination of an oral therapy with Bosentan and Sildenafil was safe in our patient group without major side effects. NYHA functional class significantly improved. However other functional parameters like 6MWD, oxygen saturation and NT-pro BNP as well as invasive hemodynamic parameters in PAH-CHD patients did not change significantly.

P-163
Clinical experience with recombinant tissue plasminogen activator in the management of thrombosis in children
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Thrombotic events may complicate the clinical course of many neonatal and pediatric pathologic processes. The primary treatment goal for this condition is the restoration of vascular flow through surgical revascularization or pharmacological dissolution of the thrombus. There are a number of practical advantages to the routine use of recombinant tissue plasminogen activator (rTPA) for non-invasive thrombolysis. Experiences about rTPA use in children are less than the adults but getting increased. We conducted a retrospective review of 11 patients who received rTPA at our institution (pediatric cardiology, pediatric oncology and neonatal intensive care unit; intracardiac thrombus ($n = 5$), prosthetic heart valve thrombus ($n = 1$), vascular complication ($n = 5$)), six of whom showed full recovery. One patient needed surgical intervention because of minor bleeding complication. Thrombi have shrunken but have not dissolved completely in two cases, and one newborn patient died from sepsis without observing the fibrinolytic response. Thrombolytic therapy represents an alternative to surgery in children. It is effective, safe and can be easily administered.

P-164
Careers and employment in children with congenital heart disease
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Introduction: Advances in diagnosis and interventional/surgical management of congenital heart malformations had as a result a considerable number of patients reaching the adult life. The aim was to investigate the careers and employment rate of children with heart problems.
Methods: Data from the archives of 114 patients followed up for 28 years.
Results: Seventy two (63,2%) patients were males, 42(36,8%) females. Males age 20-37 mean 28,5 years, females 20-33 mean 26,5, 74(64,9%) living in Athens. Socioeconomic level of parents was 14(12,2%) low, 82(72%) medium, 18(15,8%) high. As far as their occupation was concerned, 61(26,8%) private business, 62(27,2%) clerks in private sector, 39(17,1%) civil servants, 11(4,8%) farmers, 11(4,8) military/navy, retired, 42(18,4%) mothers were housewives.
The 78(68,4%) had simple malformations, 36(31,6%) complex, 56(41,1%) followed up, 5(4,4%) had interventional management, 52(45,6%) operated on, 1(0,9%) interventional and surgical management, 10(19,2%) more than one operations. Three (2,6%) were primary school graduates, 59(51,8%) high school, 43(37,7%) university, 9(7,9%) technical education. Choice for studies 96(84,2) personal, 8(7%) due to introductory exams, 10(8,8%) at random. Forty nine (43%) were working as clerks in private sector, 19(16,7%) their own business, 9(7,9%) school teachers, 9(7,9%) civil servants, 2(1,75%) sailors, 15(13,2) in other jobs: psychologist, painter, plumper, cook. Choice was personal 19(16,7%), related to studies 22(19,3%), 38(33,3%) at random, 22(19,3%) family business, 1(0,9%) talent. Between 11(9,6%) jobless, 3(2,6%) believed that they couldn't work because of the cardiac problem, 1(0,9%) didn't want.

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Conclusions: Heart malformations doesn't interfere in employment and careers. On the contrary the vast majority of the patients 98(86%) were employed.
Key Words : Congenital heart disease, employment, careers.

P-165

Syncope in children and adolescents: a four-year experience at the department of paediatrics in parma–italy

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Syncope is defined as a transient loss of consciousness (T-LOC) due to transient global cerebral hypoperfusion, characterized by rapid onset, short duration, and spontaneous complete recovery. It is a common event in childhood and adolescence; usually of benign significance, it may be the first manifestation of a serious disease.
The aim of this study was to review all patients admitted in our Emergency Unit for a T-LOC in a 4 years period of time, in order to determine the incidence in the general paediatric population, and the aetiology.
From January 2007 to January 2011 307 children (aged 1 to 18 years old, median age 9 years and 8 months) were admitted in our General Paediatric Emergency Unit for a non – traumatic T-LOC. They were classified according to the 2009 ESC guidelines. A clear aetiological diagnosis was possible in 293/307 cases (95.4%). 68 cases of 293 were excluded because of a diagnosis not matching the syncope criteria.
Two hundred twenty five patients were classified as having experienced a syncope (73.2% of the totality LOC); reflex syncope were 210 (93.3% of syncope, 198 vasovagal, 12 situational); ortostatic hypotension caused the LOC in 7 (3.1% of syncope), cardiac syncope was diagnosed in 6 patients (2.6%) with the following arrhythmias or proarrhythmic conditions: one sudden complete AV block in acute rheumatic fever, one atrial flutter in post-operative complex congenital heart disease, one paroxysmal supraventricular tachycardia, one recurrent ventricular tachycardia in cardiac fibroma and two long QT syndromes. The incidence of syncope in the paediatric population of Parma during a 4-year period resulted in 86.5/100.000 per year.
Our experience presents the data of a General Paediatric Emergency Unit, the unique in our area, with a non-selected population of patients and may represent the real incidence in our area. Our study confirmed that syncope in paediatric patients, usually benign in the majority of cases, in a small percentage of patients may be the first manifestation of a serious underlying condition. In our opinion the ECG is mandatory in every case.

P-166

Incidence of congenital coronary artery anomalies in young patients with a structurally normal heart

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Background: Congenital coronary arteries anomalies (CCAA) have been described with an incidence of 1.3% in the adult population, and may result in sudden cardiac death. There is lack of data concerning prevalence of CCAA in young patients with an otherwise structurally normal heart.
Patients and Methods: Between 2003 and 2010, we retrospectively analysed selective coronary artery angiographies of 264 consecutive patients less than 21 years of age with an echocardiographically structurally normal heart. All patients underwent selective coronary angiographies within a protocol of our institution prior to catheter ablation for supraventricular reentrant tachycardia. Special attention was given to abnormal origin, course or number of coronary arteries as well as stenosis or abnormal connections.
Results: Median age was 13.4 (2.1-21.3) years, median weight 48.8 (14.6-124) kg. CCAA were found in 9 of 264 (ca 3.4%) of patients. These included coronary artery fistula in 4 patients, anomalous origin of the circumflex artery from the right coronary artery in 3 patients, absent circumflex artery in 1 patient, and a solitary coronary artery in 1 patient, respectively. None of the CCAA required an intervention. No complications after coronary angiography were observed.
Conclusion: Isolated CCAA were more prevalent in our young patients than reported before.

P-167

The Role of N-Terminal Pro-Brain Natriuretic Peptide Levels to Predict Clinically Significant Ductus Arteriosus in Preterm Infants

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Aim: This prospective study aimed to investigate the diagnostic utility of N-terminal pro-brain natriuretic peptide (NT-proBNP) measurements to predict clinically significant patent ductus arteriosus (PDA).
Methods: In this study, 26 preterm infants (mean gestation 31.2 $\pm$ 2.5 week) were evaluated. Infants were categorized into three groups: without PDA, small PDA ($<$1.5 mm) and significant PDA ($\geq$ 1.5 mm). Plasma NT-proBNP levels were measured on the first, third and seventh days of the patients. Echocardiography was performed on the same periods of time, blinded to NT-proBNP concentration.
Results: As the distribution of 26 patients, 8 of the patients had small PDA (30,8%), 9 of the patients had significant PDA (34,6%) and other 9 of the patients did not have PDA (34,6%). As the PDA diameters increase, a significant raise in levels of NT-proBNP ($p = 0.001$) was observed (Fig 1). This effect was particularly demonstrated in the significant PDA group ($p = 0.001$). There were no significant differences according to the echocardiographic parameters among the groups.

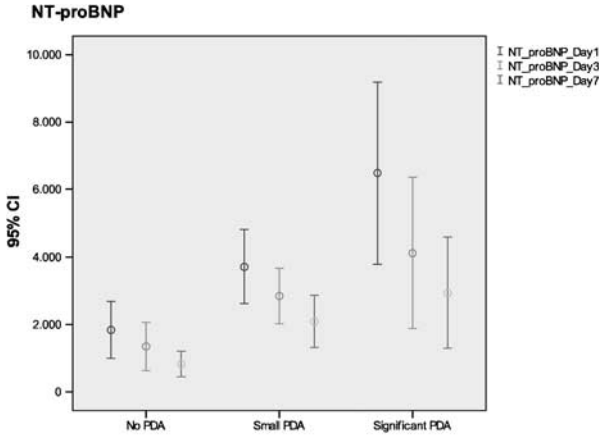

Figure 1. NT-proBNP levels of the patients according to the patent ductus arteriosus.

Fig. 1. NT-proBNP levels of the patients according to the patent ductus arteriosus.

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Conclusion: The NT-proBNP levels rise with increasing diameter of the PDA and due to NT-proBNP levels being high especially in hemodynamically significant PDA group suggests that NT-proBNP levels might be used in differentiation of significant PDAs.

P-168

Clinical characteristics of pulmonary arterial hypertension associated with congenital heart disease: baseline results from the Turkish Congenital Heart Disease-associated Pulmonary Arterial Hypertension (THALES) Registry

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Introduction: Turkish Congenital Heart Disease-associated Pulmonary Arterial Hypertension Study (THALES) is a national multicenter prospective observational registry which aims to provide information regarding demographic and clinical characteristics, laboratory data, prognosis and treatment patterns in pulmonary arterial hypertension associated with congenital heart disease (APAH-CHD). The registry is designed to record data at baseline and follow-up at 1-year intervals thereafter; the baseline clinical characteristics of patients enrolled to date are presented in this study.
Methods: Pediatric and adult patients (>3 months of age) with APAH-CHD (mean pulmonary artery pressure [mPAP] >25 mmHg, pulmonary capillary wedge pressure ≤15 mmHg, and pulmonary vascular resistance index [PVRI] >3 Wood units.m^2), were enrolled at 61 centers over the country.
Results: Between May 2009 and October 2011, 1034 patients (female: male 1.38:1) aged 3 months-79 years (mean 16.91 ± 17.91, median 11 years) were enrolled. 249 patients (24.1%) were <2, 416 (40.2%) were 2-18, and 369 (35.7%) were >18 years of age. Mean mPAP was 54,67 ± 22,15 mmHg, PVRI 10,23 ± 10,84 WU.m^2. 55.4% of patients had Eisenmenger's syndrome due to unrepaired or partially repaired CHD with significant residual defects (Group I), 34.5% had PAH associated with systemic-to-pulmonary shunts due to unrepaired or partially repaired CHD with significant residual defects (Group II), 1.2% had PAH with small defects (Group III), 7.3% had PAH after repair of CHD in the absence of significant residual defects (Group IV), and 1.6% had PAH associated with unrepaired or partially repaired complex cyanotic CHD and increased pulmonary blood flow (Group V). Dyspnoea was the most frequent symptom (89.8%), followed by chest pain (10.4%), and haemoptysis (6.9%). Syncope/pre-syncope was reported in 5.9%, the highest percentage recorded in group IV (16.2%).
Functional class was predominantly III in Group I (48.2%), and II in groups II-V (75%, 83.3%, 57.5%, 66.7%, respectively). 40.1% received targeted PAH therapies (monotherapy in 80.7%, combination in 19.3%).
Conclusions: This study seeks to define characteristics of APAH-CHD according to age groups and type of underlying CHD. In due course, we hope to gain additional information regarding the course, prognosis, impact of surgical/interventional procedures on outcome, planning and timing of targeted therapies in this group of patients with PAH.

P-169

Left Atrial and Pulmonary Capillary Wedge Pressure Relationship is Valuable for Patients with Pulmonary Hypertension

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Aim: The present study aims to determine whether pulmonary capillary wedge pressure can be used to replace left atrium pressure in case the latter cannot be measured for patients with pulmonary hypertension.
Materials and Methods: The present study reviews a total of 1040 patients with pulmonary hypertension caused by left to right cardiac shunt that were recorded throughout the Turkish Congenital Heart Disease Pulmonary Hypertension Study (THALES) held between 2009 and 2011. Approximately 996 patients of the study group underwent cardiac catheterization. The mean left atrial pressure and pulmonary capillary wedge pressure values of these patients were 10.09 ± 0.209 mmHg (range: 4-26) and 10.97 ± 0.107 mmHg (range: 1-26) respectively.
Results: The correlation between mean left atrial pressure and mean pulmonary capillary wedge pressure was found to be statistically significant for both pediatric and adult patient groups ($r = 0.728$ $p < 0.001$ and $r = 0.534$ $p < 0.001$ respectively). There is a statistically significant correlation between mean pulmonary capillary wedge pressure and each of three parameters including mean pulmonary artery pressure, systolic pulmonary artery pressure and diastolic pulmonary artery pressure ($p < 0.001$ for each)
Conclusion: There are a number of similar studies in literature. However, these studies have been done on heterogeneous patient groups that consist of adults with congenital cardiac defects and coronary cardiac diseases leading to a left to right shunt. Being conducted over a relatively larger study cohort, the present study shows that left atrium pressure significantly correlates with pulmonary capillary wedge pressure in patients with pulmonary hypertension caused by left to right cardiac shunt. Another result is that pulmonary capillary wedge pressure can be safely utilized to compute transpulmonary gradient whenever left atrium pressure cannot be measured.

P-170

Matrix Metalloproteinase-8 Activity is Increased in Type 1 Diabetes Children with High-Risk Diabetes HLA and Systemic Inflammation

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Background: Matrix metalloproteinases (MMPs) and myeloperoxidase (MPO) are colocalized to lipid-laden macrophages, and play a central role in initiation and propagation of chronic vascular diseases including atherosclerosis. Prior cross-sectional

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studies from our centre on children and adolescents with type 1 diabetes suggested possible propensity conferred by diabetes-risk HLA DQ2/8, particularly in an inflammatory milieu, to peripheral vascular dysfunction, an important precursor of atherosclerosis. In the same population, we aimed to assess whether this putative interplay between DQ2/8 and inflammation also reflects into increased activity of MMP and MPO.
Methods: Blood pressure, inflammatory, lipid, HbA1c, cyclic guanilate monophospate (cGMP), along with degree of exposure to secondhand tobacco smoke (STS) were determined in 74 children and adolescents with type 1 diabetes at baseline and 1 year later. MMP-8 and MPO levels were measured only at the 2nd time-point.
Results: In univariate regression, baseline BMI, HbA1c, CRP(log), and TC/HDL were all predictors of 1-year MMP-2 ($p < .05$ for all), while exposure to STS, BMI, cGMP, and TC/HDL predicted levels of MPO ($p < .05$ for all). The rise in serum MMP-8 was most increased in those with both DQ2/8 and CRP >1 mg/l ($p = 0.01$), but no such difference was noted with regard to MPO.
Conclusion: In young patients with type 1 diabetes, increased activities of MMP and MPO appear to relate mainly to dyslipidemia, but inflammation, particularly in those with diabetes-risk HLA, and exposure to tobacco smoke could be important stimuli as well.

P-171
Males with 45, X/46, XY have similar cardiovascular problems as females with Turner syndrome

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Cardiovascular pathology in Turner syndrome is well described and afflicts 25 to 45% of patients. We hypothesised that males with mixed gonadal dysgenesis and 45, X/46, XY (MGD) have similar cardiovascular problems as female Turner patients with 45, X/46, XY (TS).
Patients and methods: in a multicenter Belgian study, we collected 16 patients with mosaicism 45, X/46, XY who were followed at an endocrinology department: 6 females with Turner syndrome and 10 males with mixed gonadal dysgenesis. Age ranged from 1 to 38 y with a median of 13 y. Data on sexual phenotype and growth were collected from the files. Patients underwent blood pressure measurement, ECG, echocardiography and MRI (in 10/16).
Results: Turner patients with 45, X/46, XY (age 13-38 y) had normal female genitals, short height and delayed pubertal induction. All received growth hormone and hormone replacement therapy. Males with 45, X/46, XY (age 1-24y) had abnormal genitals ranging from minor abnormality (EMS12) to ambiguous genitals (EMS5). Seven received growth hormone, the other 3 were still too young.
All patients had normal blood pressure. Prolonged QTc was found in 5 (3/10 MGD, 2/6 TS). Structural heart abnormalities were equally found in TS (3/6) and MGD (5/10). Males with MGD had a high incidence of bicuspid aortic valve (50%) and dilation of the ascending aorta (20%). Both cases of dilated ascending aorta were in young boys (age 10 y) who were not followed at the cardiology department. Dilation was important (28 and 30 mm). Both boys also had a bicuspid aortic valve and one had an impaired ventricular function.

Structural cardiac abnormality	MGD (n = 10)	TS (N = 6)
VSD	1	0
Bicuspid aortic valve	5	2
Dilation of the ascending aorta	2	0
Coarctation	0	1
Right arteria lusoria	0	1

Conclusion: males with 45, X/46, XY have the same cardiovascular defects as females with Turner syndrome. Dilation of the ascending aorta can be important and might become life threatening. We advise a cardiac screening in all males with 45, X/46, XY. As in Turner syndrome, life-long follow up may be recommended.

P-172
Physical activity in children following cardiovascular interventions

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Introduction: Physical activity taken up at a very early age has a positive influence on both motor and psychosocial development. In a group of children after cardiovascular surgery a certain controlled level of physical activity is not only possible, but necessary. There is limited bibliography available with information about the development, health, lifestyle, body efficiency, and the level of physical activity among those people several years after the surgery.
The aim: To evaluate the level of physical activity among children after cardiovascular surgery.
Materials and methods: 98 children at a school age after cardiovascular surgery, and 98 parents/legal guardians of these children took part in the survey. Mean age for children with HLHS was 10 ± 3 and for other CHD 12 ± 4. There were 17 children with HLHS after BDG or Fontan operation, 17 children after TGA or DORV/TGA arterial switch procedure, 7 after tetralogy of Fallot correction and 57 after different other operations. The questionnaire for parents consisted of 29 questions, 11 general and 18 about physical activity. The questionnaire for children had 9 questions concerning physical activity.
Results: Over 50% of the children surveyed participate in PE lessons, but as many as 79% of them get tired faster than their peers. Only 13% of the parents/legal guardians claim that their children's participation in everyday activities is lower than the one represented by their peers. Most of the parents/legal guardians think that physical activity is beneficial as far as proper development is concerned, and let their children undertake it. The children from the group interviewed often take up different forms of physical activity. Most of them leave their homes during holidays, but only 26% of them have ever been to a camp. The statistical analysis did not show any differences between children after Fontan operation in comparison to other group of CHD concerning the necessity for physical activity.
Conclusions: Parents and children saw the necessity for physical activity in everyday life. Organized forms of physical activity for children after CHD operations are needed, especially during holidays.

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P-173

Echocardiographic evaluation of left ventricular function in normotansive obese children: A comparative analysis according to body mass index

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Objective: The purpose of our study was to identify the impact of the body mass index (BMI) overweight and obesity in normotansive children on structural and functional changes on the left ventricular function (LV).
Metaryal-Methods: Normotansive 30 children with overweight (group 2) (mean age: 13.2 ± 2.1 years, BMI: 25-30 kg/m2), and 30 children obesity (group 3) (mean age: 13.3 ± 2.0 years, BMI ⩾30 kg/m2), and 30 healthy controls (BMI: 18-24.9 kg/m2) were included in this study. Continuous ambulatory pressure monitoring in obese groups, standard and pulse wave (PW) Doppler echocardiographic examination have been evaluated in all study groups.
Results: In overweight and obese children left atrial volume (ml), left atrial/aortic root diameter ratio, LV interventricular septum and posterior wall thickness (mm), LV end-diastolic diameter (mm) and volume (ml), LV mass (g) were significantly higher compared to the control group ($p < 0.01$). Transmitral E/A and pulmonary vein (PV) systolic/diastolic velocities (S/D) ratio were decreased, but E-wave deceleration time (msec) and by the end-diastolic distance from the mitral annulus to the LV apex (cm) were increased in both obese groups ($p < 0.05$). BMI was a significant correlated with duration of obesity and LV mass(g) ($r = 0.527$, $r = 0.506$, $p < 0.01$, respectively). Significantly negative correlation was found between BMI and Mitral E/A, PV S/D ratio ($r = -0.230$, $r = -0.577$, $p < 0.01$, respectively). PVA velocities are increased in obese groups.
Conclusion: In our study subclinical left ventricular myocardial dysfunction was noted in obese groups which correlates with BMI. Determination of diastolic dysfunction by PV PW Doppler can be usefull an pre-obese period.

P-174

Thrombotic profile in Fontan patients assessed by whole-blood assays and endothelial biomarkers

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Introduction: After a Fontan procedure the patient face an increased risk for thromboembolic complications. The etiology for this increased risk is not well defined. This study aimed to evaluate whether global whole-blood assays of coagulation as well markers of endothelial activation/damage and glycocalyx degradation can detect hypercoagulability in Fontan patients.
Methods: We performed kaolin-activated thrombelastography (TEG) and functional fibrinogen determined by TEG on citrated whole blood, and multiple electrode platelet aggregometry (MEA) on heparinized blood from Fontan patients. Furthermore, plasma was analyzed for biomarkers reflecting tissue/endothelial cell/glycocalyx damage (histone-complexed DNA fragments, Protein C, soluble CD40 ligand, soluble thrombomodulin, syndecan-1, tissue-type plasminogen activator). Variables from whole-blood assays and the endothelial biomarkers were compared in patient groups, stratified according to age, antithrombotic therapy, post-Fontan thromboembolic event, and degree of glycocalyx degradation (concentration of syndecan-1). Correlations between endothelial biomarkers and demographic-, anatomical-, clinical- and biochemical parameters were investigated.
Results: Whole-blood assays were performed in 118 Fontan patients (median age: 14.0 years; IQR: 8.1-19.5) and analyses of endothelial biomarkers were performed in 79 Fontan patients (median age: 14.0; IQR: 8.2-19.5). Of the 118 patients, 79 (67%) received antiplatelet therapy, 27 patients (23%) vitamin K antagonist, three patients (3%) unfractionated heparin, and nine patients (8%) no antithrombotic therapy. Six patients (5%) had a known post-Fontan thromboembolic event. None of the patient groups demonstrated evidence of hypercoagulability in the whole-blood assays compared to reference data and no statistically significant differences were found between groups in the stratified analyses, except for those expected from type of antithrombotic therapy. A strong correlation between the endothelial biomarkers was found in all age- and antithrombotic therapy groups. Concentration of all the endothelial biomarkers was above the median in six patients (8%) (chi-square test, $p = 0.11$). No other general correlation was found.
Conclusions: Eight percent of Fontan patients show evidence of considerable endothelial activation/damage including glycocalyx degradation assessed by endothelial biomarkers. This subset of patients is potentially more thrombogenic than other Fontan patients, however no characteristics of this group were discovered. Whole-blood assays did not show evidence of hypercoagulability in Fontan patients and thrombotic profile was identical between different Fontan groups.

P-175

The Frequency of Aspirin Hepatotoxicity in Children with Acute Rheumatic Fever

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Introduction: Aspirin is the most commonly used anti-inflammatory drug in treatment of acute rheumatic fever (ARF). During therapy aspirin hepatotoxicity may develop. However present studies have been based on symptomatic patients. This study was planned to establish the frequency of hepatotoxicity in children with ARF treated with aspirin.
Methods: One hundred and thirty six patients who were treated with aspirin for ARF (first attack, rebound, reactivation) were included in the study. Aspirin (maximum dose is 3,5 g/day) was started at a dose of 100 mg/kg in patients with carditis and 70 mg/kg in patients with arthritis. Transaminase levels were checked before and 3th, 7th, 14th and 30th days of treatment and also at the days that patients showed symptoms of hepatotoxicity. Patients have been questioned at all visits for hepatotoxicity findings.
Results: The number of patients and the treatments in the study are 136 and 144, respectively. Hepatotoxicity occurred in 39,5% (57) of treatments, and 31% (18) of those the patients were symptomatic. In patients with hepatotoxicity, mean alanine and aspartate aminotransferase levels were 248 ± 347 U/L and 229 ± 309 U/L, respectively. At 15 treatments, transaminase levels decreased to normal values in an average of 21 ± 15 days by reducing the dose to 65 mg/kg. At 42 treatments medicine was changed because the transaminase levels kept rising despite dose reduction (nonsteroid anti-inflammatory drug = 8, prednisolone = 34). At 40 treatments transaminase levels returned to normal values in a mean of 24 ± 14 days. In 2 patients whose transaminase levels have continued rising, hepatitis B and cytomegalovirus infections were detected.
Conclusion: The incidence of aspirin hepatotoxicity in ARF is considerably high and it is necessary to check transaminase levels

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periodically, even if patients treated with aspirin are asymptomatic. In case of toxicity, follow up after dose reduction and a change in therapy is the suitable option if elevation in transaminase levels persists despite dose reduction. Nonsteroid anti-inflammatory drugs seemed to be an effective alternative for patients with aspirin hepatotoxicity. In patients whose transaminase levels elevation persists, attention must be paid to infectious hepatitis.

P-176

Cardiovascular complications of obesity at children in North-Eastern Region of Romania

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Introduction: Obesity is the most prevalent nutritional disorder among children and adolescents. Childhood obesity predisposes to hypertension, insulin resistance and type 2 diabetes, hyperlipidemia, liver and renal disease, and reproductive dysfunction. The purpose of this study is to evaluate the cardiovascular complications of obesity in children and adolescents.
Methods: Were taken in the study 273 children aged 6-17 years hospitalized for a period of five years (January 2007 – January 2012) in Pediatric Cardiology Department, Children's Emergency Hospital "Sfanta Maria", Iasi, Romania.
They followed the age, sex, body mass index, blood pressure, biological VSR, serum fibrinogen values, total cholesterol and results of echocardiography and ophthalmological examination.
Prospective echocardiographic measurements were performed in 273 obese children. Two-dimensional, M-mode and color M-mode ultrasound, conventional pulse wave Doppler imaging were used to assess cardiac function
Results: The cases of obesity were most common in female subjects (55%), with preponderance in the age range 13-15 years in both sexes. We found increased blood pressure values in 18.85% of cases. Measurements of LV mass, LV wall thickness and LV end-diastolic diameter and volume were significantly elevated in 34 obese children (12.45% of cases).
Fibrinogen level and total cholesterol elevated recorded average higher among female subjects (54% of cases hypercholesterolemia, that 59% of cases with increased plasma fibrinogen).
Conclusion: In this study it was found the occurrence of cardiovascular complications of obesity secondary-school period adolescent. The echocardiographic exam confirms the elevated LV mass in obese children.
Is necessary to follow the periodic height and weight, as well as evaluating blood pressure, total cholesterol, fibrinogen among children with obesity for preventing cardiovascular complications and framing of these patients in a cardiovascular risk group.

P-177

Cardiovascular outcome in children born very preterm after intrauterine growth restriction with severely abnormal umbilical artery blood flow

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Introduction: Low birth weight (BW) and preterm birth have been linked to cardiovascular disease in adulthood. The combined effects of intrauterine growth restriction (IUGR) and preterm birth on cardiovascular outcome are not clarified. Objective: To evaluate cardiovascular function at 7 years of age in children born very preterm after IUGR with abnormal blood flow.
Methods: Blood pressure (BP), carotid artery intima media thickness (IMT) and cardiac size, diastolic and systolic function were assessed with a digital BP monitor and ultrasonography, respectively, in thirty 6-8 year old children born very preterm (PT) after IUGR (PT-IUGR) with a median (range) BW 650 (395-976) g and median (range) gestational age (GA) 27 (24-29) weeks. Additional 30 children born PT with BW 1010 (660-1790) g matched for GA at birth (PT-AGA) and 30 children born at term (T-AGA) with BW 3530 (3000-4390) g were studied. At the time of study, both the PT-AGA and T-AGA groups were matched for gender and age with the PT-IUGR group.
Results: Systolic and mean BP were elevated in both the PT-IUGR group mean (SE)(106 and 77 mm Hg) and the PT-AGA group (106 and 76 mm Hg) compared to the T-AGA group (100 and 72 mm Hg) ($p = 0.018$ and 0.014, respectively). The PT-IUGR group had lower IMT compared to the T-AGA group ($p < 0.05$). Cardiac size and function did not differ between groups ($p > 0.1$).
Conclusion: IUGR appears to be associated with structural abnormalities in the vessel wall, whereas elevated BP during childhood is primarily related to preterm birth irrespective of fetal growth impairment.

P-178

Anticoagulation therapy for preventing thromboembolism in children after extracardiac conduit Fontan procedure: does it make a difference?

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Objective: The risk of thromboembolic complication for Fontan patients is well-documented despite of empiric use of antithrombotic therapy.
Aim: The study aimed to estimate the incidence of thromboembolism among patients underwent extracardiac conduit (ECC) Fontan procedure receiving anticoagulation therapy alone, or as adjunctive to antiplatelet therapy.
Method: 85 patients were evaluated [male-to-female ratio: 50:35, median age:10.3 years (IQR:7.3-14.5), median follow-up: 4.6 years (IQR:2.8-7.6)] after ECC Fontan palliation performed between 1999.01.01-2010.12.31 at our centre. All patients received iv. heparin in the very early postoperative days; followed by either oral vitamin K antagonist (VKA) lifelong, or VKA and aspirin sequentially: VKA for 6 months after non-fenestrated ECC Fontan palliation or postoperative interventional closure of fenestration, continued with aspirin lifelong. Effect of different therapeutic strategies on the occurrence of thromboembolic events was analyzed. Peripheral vein thromboses were excluded. Factors predisposing to thromboembolism in Fontan circulation were also explored.
Results: Thirtyfour patients (40%) received VKA monotherapy (5 patients with and 21 patients without fenestration, respectively, and 8 patients with fenestration closed interventionally). Fifty patients (59%) received sequential antithrombotic therapy: 25 patients without fenestration and 25 patients with interventionally closed fenestration. 1 patient died unrelated to thromboembolism on the 1st postoperative day.
The overall clinically symptomatic thromboembolism rate was 4.7% (4/85 patients). Thromboembolism was recognized as seizures or hemiparesis after a median of 1.03 years (IQR:0.1-2.9) postoperatively, confirmed by MRI (3 cases) or CT (1 case). Three patients were on VKA therapy (INR 2 ± 0.2) at the time of thromboembolism regardless of the final type of antithrombotic regimen, and 1 patient on iv. heparin (PTI 54). Fenestration was open

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in 2 patients. Factors predisposing to thromboembolism in Fontan circulation, such as arrhythmia, ventricular dysfunction, severe valvular regurgitation, protein-loosing enteropathy, polycytaemia or hereditary thrombophylia, could not be demostrated simultaneously.
Conclusion:
1. Thromboembolism after ECC Fontan procedure was relatively rare among our patients.
2. Applied antithrombotic regimens showed no difference, since almost all events occured on VKA therapy.
3. Thromboembolism was not associated with failing Fontan circulation or factors predisposing to thromboembolism.
4. Thrombembolism rate was not higher in patients with open fenestration than in rest of the cohort.

P-179
Stiffness of the abdominal aorta in children with β thalassemia major and thalassemia trait related with ferritin levels

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Objective: Increased iron stores have been implicated in the association with increased risk of cardiovascular events. The aim of this study was to investigate the impact of iron overload on abdominal aortic stiffness in patients with β TM and compared to the children with thalassemia trait (TT) and healthy controls.
Methods: This prospective study included three groups; Group I: 29 patients with β TM, Group II: 28 patients with TT, Group III: 29 healthy controls. In all subjects, hemoglobin, cholesterol, high-density lipoprotein-cholesterol, and low-density lipoprotein-cholesterol levels were measured. Blood pressure was measured in all subjects. The average serum ferritin level was assessed in β TM patients. All children were noninvasively evaluated with transthoracic echocardiography. Abdominal aorta diameters were measured. Aortic strain (S), pressure strain elastic module (Ep), pressure strain normalized by diastolic pressure (Ep*), aortic stiffness β index (βSI) and, aortic distensibility (DIS) were calculated using the measured data.
Results: There was no statistically significant difference between the groups in sex, mean age, body mass index, heart rate, and blood pressure ($P > 0.05$). In β TM patients S (0.23 ± 0.058 /. $0.29 \pm 0.06/0.33 \pm 0.07$ $P < 0.0001$) and DIS ($1.23 \pm 0.42/1.5 \pm 0.51/1.81 \pm 0.53$, $P < 0.0001$) were significantly lower compared with the other groups. However, Ep ($181.1 \pm 58.7/147.1 \pm 52/121 \pm 44$, $P < 0.0001$), Ep* ($2.98 \pm 0.98/2.36 \pm 0.92/2.06 \pm 0.92$, $P < 0.0001$), and βSI ($2.26 \pm 0.67/1.79 \pm 0.63/1.56 \pm 0.59$, $P < 0.0001$) were significantly higher in β TM patients and TT subjects than controls. There was a statistically significant negative correlation between ferritin and S, DIS ($p < 0.05$ $r = -0.564, -0.411$). However, there was a statistically significant positive correlation between ferritin and βSI, Ep ($p < 0.05$, $r = 0.027, 0.375$).
Conclusions: Increased abdominal aortic stiffness was detected in β TM patients and this increase in arterial stiffness correlated with ferritin levels.

P-180
The risk factors of recurrency of vasovagal syncope in children and adolescents and the value of head-up tilt table test

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Introduction: Syncope is one of major complaint for 1% of all emergency admissions. It is usually a bening problem, but it can be recurrent. It remains unclear who are at high risk of recurrent syncope. This study was performed to determine the long-term follow-up of children and adolescents with syncope or pre-syncope, and to analyse risk factors for recurrent syncope.
Methods: This study included 150 patients (50 male,100 female) aged between 8-18 years, undergoing a tilt test. They were subsequently followed-up in clinic visits with a final interview at the clinic or by telephone at the end of the follow-up period.
Results: Tilt table test was positive in 97 and negative in 53 patients respectively. The types of the syncope in tilt table positive group were mixed in 48, vazodeppresor in 34 and cardioinhibitor in 15 patients. Sixty-five patients had syncope in passive phase and 32 had in provacative phase of the test .The avarage age, the number of the episodoes and the presence of the trauma were not statistically different between tilt positive and negative patients. During a mean follow-up of 3-78 month, recurrent syncope was observed in 27 of 100 female and 13 of 50 male patients. When comparing syncope-free children at follow-up, children with recurrent syncope had a greater number of historical syncopal spells (6,3 v.s. 2,6, $P = 0000$). The recurrency rate was higher in patients who had more than 4 episodes initially and who had syncope in the first 20 minutes of the tilt table test. The sex distribution was found similiar in recurrent and non-recurrent group. The number of the episodes decreased after tilt table test ($p = 0.000$; $3,86 \pm 4,75$ v.s $0,73 \pm 0,44$). The recurrence rate was similar between positive and negative tilt test groups (24,7% vs 30,1%, respectively; $P = 0,47$).
Conclusion: Our findings demonstrated that the predictors of longterm outcome in adolescents are the total number of spells documented before admission and the occuring time of syncope during the test. The risk of syncope recurrence was found independent from head-up tilt test results.

P-181
Factors affecting the short term prognosis of patients with Acute Rheumatic Carditis

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Objective: The aim of the study is to evaluate the factors contributing the short term prognosis of patients that have acute rheumatic carditis with laboratory findings, electrocardiography, echocardiography and flow cytometric findings at the initial phase of the diagnosis.
Methods: Twenty-two patients with acute rheumatic carditis were enrolled in this study. Patients matched with 20 apparently healthy child, there were no significant differences between groups ($p > 0,05$). All children were evaluated with echocardiography according to ASE guidelines including Tissue Doppler Imaging, electrocardiography and flow cytometric study from peripheric blood sample conducted at the initial phase of the diagnosis.
Results: Echocardiographic finding revealed that 68.2% of patients have combined valvular regurgitation, 22.7% have isolated mitral valve regurgitation and 9.1% have isolated aortic regurgitation. Echocardiographic measurements; LVEDd, LA volume index, LA/Ao, mitral valve E, E/Ea were increased in patient group ($p < 0,05$). TDI measurements Mitral valve lateral Sa and Ea were decreased significantly ($p < 0,05$). Flowcytometric measurements revealed lympopenia in 7/22 (31.8%) patients. CD3 and CD4 lymphocytes were decreased simultaneously in 9/22 (27.3%) patients. CD3, CD4 and CD8 levels

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were decreased in 3/ 22 (13.6%) patients. Echocardiographic re-examination after 6 months of diagnosis revealed that patients without healing valvular regurgitation had increased CD4/CD8 ratio at the initial phase of the diagnosis but this result was statistically insignificant. Serum albumin levels were significantly decreased at non -healing group (2,6 ± 0,5 gr/dl vs 3,3 ± 0,5gr/dl).
Conclusions: Our findings showed us that increased LA volum index and mitral valve E/Ea ratios is obivious in acute rheumatic carditis. Flow parameters from TDI, initial ASO titration, elevated ESR, decreasing at Hb level or lymphopenia at flowcytometry and CD3, CD4, CD8 and NK levels have no prognostic value. Patients with initial hypoalbuminemia showed no regression of cardiac findings after following for six months. Although patients with high CD4/CD8 ratio have a poorer prognosis, this finding wasn't statistically significant. This finding was may be due to a smaller size of our patient group. Consequently, we suggest initial serum albumin levels and high CD4/CD8 levels as predictors of severity of cardiac involvement in the short term follow-up period of acute rheumatic carditis.

P-182
Assessment of cardiac function in children with Friedreich's Ataxia: A single centre experience
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Introduction: Friedreich's ataxia (FA) is an autosomal recessive neurodegenerative disorder, caused by unstable GAA expansions in the FA gene encoding for the 210-amino-acid protein frataxin. Cardiac associations with FA are hypertrophic (HCM) and dilated (DCM) cardiomyopathies, ventricular dysfunction and arrhythmias. Presentation and Cardiac abnormalities in the paediatric population with FA are not well described. We aim to look at these.
Material and Methods: 12 year retrospective analysis of data in a single centre; clinical features, electrocardiographs (ECG) and echocardiograms of all patients with FA analysed looking for patterns of presentation and progression of cardiac disease.
Results: 21 patients with confirmed diagnosis of FA were seen (8 male and 13 female). 19 patients analysed (insufficient data in 2 patients). Median age of presentation to cardiology was 11 years (5-15 years). Referrals were from paediatricians/neurologists, 10 with confirmed diagnosis at referral, 7 with neurological signs and two with heart murmurs. All had progressive neurological signs, five were wheel chair bound. Three had kyphoscoliosis with two needing spinal surgery. ECG abnormalities at presentation were noted in all with isolated T changes (100%); Left ventricular hypertrophy (LVH) in 13 (66%). None had palpitations/syncope or arrhythmias. Echocardiograms assessed for left ventricular mass, global and regional function. At presentation, 16 (84%) had concentric HCM; of these 11 had normal global systolic function, with additional diastolic dysfunction in three. Two (10%) had DCM, and one (5%) had hypertrophic obstructive cardiomyopathy (HOCM). Regional wall motion abnormalities were seen in all patients. 16 patients had serial scans, six (31%) had some improvement of cardiac lesion; these were also on Idebenone treatment. Vitamin E and Coenzyme q10 was used in three; and one patient with HOCM on beta-blocker. There was progression of LVH and or dilatation in 3 (16%). All are alive; nine have ongoing reviews and 10 transferred to adults.
Conclusions: Concentric hypertrophied cardiomyopathy is the most common cardiac abnormality with specific ECG changes seen in all of them. Regional wall motion abnormalities were seen even where global function was normal. This highlights the importance of undertaking detailed ventricular analysis for assessment of both global and regional function.

P-183
Acquired von Willebrand disease due to severe pulmonary valve stenosis: A relevant cause of bleeding in children with Noonan syndrome ?
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Noonan syndrome (NS) is a congenital disorder characterized by dysmorphic facies, short stature and congenital heart defects. Various haemostatic disorders have been described, however, not all were related to a relevant bleeding diathesis, present in up to 65% of patients. Several subgroups of NS patients–especially those with PTPN11 mutation–are associated with pulmonary stenosis. It is known, that heart defects with a large blood flow through a restrictive opening are prone to shear stress related destruction of the VWF, resulting in acquired von Willebrand disease (aVWD). Our aim was to find out whether the pulmonary stenosis is responsible for the bleeding tendency in some of the NS patients.
We investigated the haemostatic system in 15 children with genetically proven NS (14 with PTPN 11, one with SOS1 mutation, responsible for 50% and 10-15% of NS gene mutations, respectively). All of them were asked for signs and symptoms of bleeding, and all underwent a full echocardiographic study.
Platelet count, global coagulation parameters, fibrinogen and antithrombin were normal in all patients. None of the patients had a relevant reduction of a coagulation factor activity. Five patients showed a pulmonary stenosis with systolic gradients >60 mmHg. In three of them a deficiency of the high molecular weight multimers (HMWM) and a pathologic collagen-binding capacity was detected, suggesting aVWD. 9/15 (60%) of patients–two with a deficiency of HMWM–indicated a relevant bleeding diathesis and complained about easy bruising, 3/15 reporting spontaneous gum bleeding.
The mechanism responsible for the destruction of the HMWM in other heart defects could also explain bleeding in some of the NS patients: A high velocity jet across the (here: pulmonary) stenosis appears to cause chronic shear-induced platelet-VWF interaction and clearance. It was first described in aortic stenosis by Warkentin in 1992.
Our discovery of an aVWD in 3/15 children with NS suggests a considerable prevalence of this disorder at least in patients with clinically relevant pulmonary stenosis. This is of importance as in most of them cardiac catheterization or open heart surgery have to be performed and might be compromised by the bleeding tendency.

P-184
How to describe complex congenital heart defects? A hierarchical approach for diagnosis, using the Anatomic and Clinical Classification of Congenital Heart Defects.
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Background: To describe complex congenital heart defects (CHD) in their diversity requires a common language, in order to unify the diagnostic process. As a first step, we designed the Anatomic and Clinical Classification of CHD (ACC-CHD) based on a multi-dimensional approach encompassing anatomic, diagnostic and therapeutic criteria. The second step aims to establish a hierarchy among the different components of a complex CHD, which is fundamental since it places the main lesion into one, and only one, of the 10 groups of the ACC-CHD.
Methods: The hierarchy within the 10 groups (G) and 23 subgroups of the ACC-CHD was based on the clinical aggregation process reflecting the routine practice of paediatric cardiology, but also on anatomy and criteria used for medical and surgical management. For example, transposition of the great arteries is the main defect (D1) when associated with a VSD but not when associated with double-inlet ventricle, because of the different surgical management: arterial switch versus univentricular approach. We applied this method to data acquired from a population-based cohort of patients with CHD in France (the Epicard study) made up of 2867 cases (82% live births, 1.8% stillbirths and 16.2% pregnancy terminations).
Results: Among the 583 cases with more than 2 CHD, G1 was D1 in 100% of cases, G5 in 88%. G2 and G10 were almost never D1(Table).

ACC-CHD Groups	D1(%)	D2(%)	D3(%)	D4(%)
1. Heterotaxy, including isomerism and mirror-imagery	100	0	0	0
2.Anomalies of the venous return	3	50	30	17
3. Anomalies of the atria and interatrial communications	17	78	5	0
4.Anomalies of the atrioventricular junctions and valves	42	37	16	5
5.Complex anomalies of the atrioventricular connections	88	13	0	0
6.Functionally univentricular hearts	44	34	17	5
7.Ventricular septal defects	52	43	3	1
8. Anomalies of the outflow tracts and VA connections	44	35	15	5
9. Anomalies of the extrapericardial trunks	46	33	16	6
10. Congenital anomalies of the coronary arteries	0	70	30	0

Conclusion: This hierarchical approach using ACC-CHD has proved its utility for clinical and epidemiologic studies, and can provide a structure for various databases.

P-185
A new anatomic approach to the ventricular septal defect in interruption of the aortic arch

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Objectives: To analyze the anatomy of the ventricular septal defect (VSD) in heart specimens with interruption of the aortic arch (IAA), in order to confirm the hypothesis of different embryologic mechanisms for the different anatomic types of IAA.

Material and methods: We examined 27 hearts from the anatomic collection of the French Reference Center for Complex Congenital Heart Defects with IAA, concordant atrioventricular and ventriculoarterial connections, and 2 distinct great arteries. Hearts were classified according to Celoria and Patton: type A, interruption distal to the left subclavian artery (A), type B, between the left subclavian and the left carotid artery (B), type C, between the 2 carotid arteries (C). We focused on the anatomy of the VSD viewed from the right ventricular side.
Results: There were 10 A, 17 B, no C. One A (with aortopulmonary window) and 1 B had no VSD. The VSD was conoventricular, located between the 2 limbs of the septal band, in 4/9 A and 16/16 B ($p = 0.005$), with posterior deviation of the outlet septum. In A, the VSD was conoventricular in 4, with muscular borders in 2 and fibrous extension of the posterior limb of the septal band in 2, muscular in 3, membranous in 2. In B, the conoventricular VSD had entirely muscular borders in 4, fibrous extension of the posterior limb of the septal band in 9, and was juxta-arterial in 3; there was no fibrous continuity between the tricuspid and aortic valve.
Conclusion: The VSD in IAA type B is always conoventricular, with posterior deviation of the outlet septum, but without any fibrous tricuspid-aortic continuity. The VSD in IAA type A can be of any type. This reinforces the hypothesis of different pathogenic mechanisms responsible for the 2 types of IAA, and the inclusion of IAA type B in the group of conotruncal defects. The absence of fibrous tricuspid-aortic continuity indicates that the fibrous extension of the posterior limb of the septal band found in some hearts may be due to the posterior deviation of the outlet septum rather than to a perimembranous extension of the VSD.

P-186
Conotruncal (outflow tract) defects: is the ventricular septal defect always the same?

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Background: The ventricular septal defect (VSD) in conotruncal (outflow tract) defects opens into the right ventricle between the limbs of the septal band. However, the terminology of the VSD is ambiguous, particularly in tetralogy of Fallot (TOF): the conoventricular VSD, due to the malalignment between the outlet septum and the muscular septum, is also described as perimembranous because of the presence of a fibrous continuity between the aortic and tricuspid valvar leaflets.
Aim of the study: To analyze the anatomy of the VSD in hearts with conotruncal (outflow tract) defects, in order to solve this nomenclatural ambiguity.
Material and methods: We reviewed 196 heart specimens from the anatomic collection of the French Center of Reference for Complex Congenital Heart Defects: 63 TOF, 54 TOF with pulmonary atresia (TOF-PA), 54 common arterial trunk (CAT), 16 interrupted aortic arch type B (IAA-B) and 19 malalignment VSD without outflow tract obstruction (MVSD). Special attention was paid to the borders of the VSD, viewed from the right ventricular side.
Results: The VSD was conoventricular, located between the 2 limbs of the septal band, in all hearts. There was a fibrous continuity between the tricuspid and aortic valve in 74% of MVSD, 67% of TOF, 24% of TOF-PA, 2% of CAT, 0% of IAA-B ($p < 0.005$). The aortic valve was continuous with the anterior tricuspid leaflet, but not with the septal leaflet, contrary to perimembranous VSD.

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Conclusion: All conotruncal (outflow tract) defects share the same VSD, conoventricular in type. However, there are some differences between these defects regarding the inferior rim of the VSD. The continuity of the aortic valve with the anterior tricuspid leaflet (and not the septal leaflet) indicates that this continuity may be a consequence of the deviation of the ventriculo-infundibular fold, along with its outlet septal component, rather than a perimembranous extension of the VSD. Finally, these differences suggest that conotruncal (outflow tract) defects should be considered as an anatomic continuum rather than distinct physiological phenotypes.

P-187

Efficacy and safety of home INR testing during Warfarin treatment for children and adults with congenital heart disease

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Objective: To evaluate the efficacy and safety of home INR (international normalised ratio) testing in children and adults with congenital heart disease (CHD) prescribed warfarin therapy.
Methods: Prospective data collection with retrospective analysis in consecutive patients with CHD discharged from University Hospital Leicester (UHL) following initiation of warfarin therapy. Home INR testing in all patients was performed using the CoaguCheK XS device (Roche Diagnostics GmbH, Mannheim, Germany). Results were telephoned to UHL and warfarin prescribed by the attending cardiologist according to internal protocols.
Results: Complete data was available in 60 of 89 patients (20 female, 45 children, mean age ± SD 13.7 ± 5.9, Fontan circulation = 37, mechanical heart valve = 22, other = 1). Median follow-up was 3.5 years (interquartile range 1.9 years); total cohort follow-up was 193.5 person-years. Overall mean test frequency was 4.0 ± 3.8 tests per month (4.3 ± 4.2 in children and 3.2 ± 2.1 in adults; 3.4 ± 4.1 in Fontan patients and 4.4 ± 1.3 in mechanical valve patients, both P = NS. 61.7 ± 19.5% of INR results were within target range (adults vs children and Fontan vs valve both P = NS), 16.9 ± 10.3% were above range and 21.4 ± 13.9% were below range. Major anticoagulation-related complications occurred in two patients (intracranial haemorrhage in a child and thromboembolic stroke in an adult).
Conclusions: Home INR testing by adults and children with congenital heart disease provides effective anticoagulation control with acceptable test frequency and low major complication rate.

P-188

Chest pain in children and adolescents : a frequent complaint

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Introduction: Chest pain is a frequent presenting complaint in children and adolescents. It is frequently perceived by both children and parents as heart related, therefore causing a lot of worry and emotional upset. It frequently leads to referral to a pediatric cardiology outpatient clinic for more specific evaluation. In the pediatric population, chest pain is very rarely due to a heart problem. Noncardiac chest pain is by far the most common cause of chest pain in the pediatric population.
Methods: Retrospective review of all cases of chest pain referred to our pediatric cardiology outpatient clinic over a one year period (2011). Charts, ECG, Holter, echocardiography studies and exercise test results were reviewed.
Results: From January 1st to December 31th 2011, 82 patients presented with chest pain. The median age at presentation was 11 years old (range 4-17 years old). There were 36 boys and 46 girls. In 36 cases chest pain was the only complaint. In 60% of cases symptoms occurred with exercise. Patients were evaluated with ECG (n = 82, 100%), Holter (n = 16, 20%), R-test (n = 6, 7%), echocardiography (n = 81, 99%) and exercise stress test (n = 35, 43%). In 17 patients chest pain was triggered and patients could be examined while being symptomatic. In 99% of patients chest pain was due to a noncardiac cause, of which 78% presented with musculo-skeletal or chest wall pain. Other noncardiac causes included hyperventilation and vasovagal malaise. In only 1% (8 patients) chest pain was due to a cardiac cause, of which 7 presented with an arrhythmia and 1 patient with signs of myocardial ischemia in the context of a severe aortic stenosis.
Conclusion: All patients presenting with chest pain warrant a thorough evaluation. In the vast majority of cases chest pain has a musculoskeletal origin. Reassuring both parents and patients about the benign nature of chest wall pain is of great importance. Although rare, a cardiac cause for chest pain should be sought for. It is most likely to be associated with abnormal cardiac findings and to occur upon exertion.

P-189

Aminoterminal pro B-type natriuretic peptide concentrations in newborns: congenital heart disease or respiratory failure due to neonatal lung disease?

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Background: NT-pro-BNP has been shown to differentiate between heart and lung disease in adult and pediatric patients with respiratory distress. The aim of this study was to evaluate whether NT-pro-BNP could differentiate between congenital heart disease (CHD) and respiratory failure due to lung disease or perinatal asphyxia in newborns admitted to our NICU.
Methods: From Feb 2009 to Oct 2011 ninety-five neonates were recruited for this single-center prospective study. Inclusion criteria were CHD or acute respiratory failure with need of mechanical ventilation or nasal-CPAP. At admission all neonates underwent physical examination, chest X-ray, and echocardiography. Plasma NT-pro-BNP levels were evaluated on day of life (DOL) 1,2,3, and 5. Exclusion criteria were <37 weeks of gestation (n = 1), syndrome or other major extracardiac malformations (n = 2), coincidence of CHD and asphyxia (n = 2), missing parental consent (n = 4), missing NT-pro-BNP levels on DOL3 (n = 6). Finally, 80 patients could be included in the statistical analysis, 40 were diagnosed with CHD (fetally diagnosed:31, postnatally diagnosed:9), 25 with lung disease and 15 with asphyxia.
Results: Median NT-pro-BNP concentrations were 2607, 5154, 10012 pg/ml on DOL1, 13636, 5604.5, 4119 pg/ml on DOL2, 18469, 3955, 3139 pg/ml on DOL3, and 16946, 2838, 2985 on DOL5 in the CHD, lung disease, asphyxia group, respectively. Compared to the lung disease group NT-pro-BNP concentrations were significantly higher in the CHD group on DOL 2,3, and 5. On day DOL1 the difference did not reach significance. The CHD group compared to the asphyxia group revealed on DOL1 significantly higher NT-pro-BNP concentrations in the asphyxia group. However, on DOL2 and on DOL3 NT-pro-BNP concentrations were significantly higher in the CHD group. On DOL5 NT-pro-BNP concentrations did not show a significant difference between the CHD and asphyxia group.

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Conclusions: NT-pro-BNP differentiates between congenital heart disease (CHD) and lung disease in neonates older than 24 hours. In presence of perinatal asphyxia NT-pro-BNP concentrations are elevated secondary to asphyxia and differentiation between heart and lung disease is not possible any more.

P-190

Dear Surgeon, Have I got it right? Does it matter at all?

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Introduction: Coronary artery anatomy in simple TGA is well-studied area. It still however, poses a challenge to many Paediatric cardiologists to accurately define and diagnose unusual coronary arrangement preoperatively. Intraoperative findings are gold standard although intramural coronaries can be challenging.
Aims: To study whether accurately defining coronary anatomy preoperatively in simple TGA influence outcome of surgery and to ascertain value of echocardiogram in accurately diagnosing difficult coronary anatomy (intramural coronaries).
Methods: Retrospective analysis of data from 35 patients with simple d-TGA between 2008 and 2011 and comparison of their pre-operative coronary arrangement on echocardiography with intraoperative findings. We then looked at patients with discrepant anatomy and compared them with patients who had consistent anatomy to identify any difference in postoperative outcome until first attendance to clinic post discharge.
Results: Total 35 patients, 19 had discrepant coronary arrangement (54.3%). 25 male infants (71.4%), median weight 3.68 kg ± 0.44 (2.5 to 4.3), median age at surgery 8.5 days (3 to 33), 9 day discrepant vs. 8 consistent). Preoperative status was comparable in both groups, although antenatal diagnosis ratio was reversed (12 discrepant vs. 2 consistent). There was no statistical difference in duration of hospital stay (7 to 34 days discrepant group vs. 7 – 21 days consistent group, p-value 0.36). There was no statistical difference in duration of cardiopulmonary bypass time and cross clamp time (p-values 0.43 and 0.12 respectively). Lactate level on admission to intensive care from theatre was not significantly different (p-value of 0.10). Primary chest closure rates 11 discrepant vs. 10 consistent. The intraoperative and postoperative echocardiograms for cardiac function on both groups were comparable. Two patients in discrepant group had intramural coronaries. One of them required reoperation and extracorporeal life support in first 48 hours post-operatively. Apart from this patient in discrepant group, cardiac assessment with electrocardiogram and echocardiogram at 7 to 10 days post op and during first clinic visit were satisfactory in both groups. There was no significant arrhythmia or mortality.
Conclusion: Defining coronary anatomy preoperatively does not influence outcome in simple TGA however defining intramural coronaries help surgeon to plan their approach pre operatively.

P-191

Prevalence of hypertension in children after early repair of coarctation of the aorta

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Introduction: Aortic coarctation is associated with an increased risk of hypertension even when corrected successfully at an early age. Previous studies have associated this risk to increased arterial stiffness and dysfunction of the baroreceptor reflex.

Purpose and Methods: Retrospective descriptive study analysing hypertension prevalence, in a group of 37 patients born between 1994 and 2006, with coarctation of the aorta who underwent surgical repair in the first year of life, with a minimum follow-up of five years. Hypertension was defined as blood arterial pressure (BP) greater than the 95th percentile for age and sex, measured by 24 hour ambulatory blood pressure monitoring (ABPM) placed in the right upper limb. Transtoracic echocardiography was performed in all patients to exclude recoarctation.
Results: Patients under five or over eighteen years, with recoarctation, deceased, with complex congenital heart disease, which aren't followed by our department or impossible to contact and refused to participate were excluded. In 27% there were no associated cardiac malformations and in the other group the following heart lesions were found: bicuspid aortic valve in 49%, ventricular septal defect in 22% and mitral valve disease in 16%. Age at surgery ranged from three days to eleven months with a median of 44 days. The end to end anastomosis surgical technique was adopted in 57% of the cases, construction of a subclavian flap in 30% and mixed technique in 13%. None of the patients performed a second surgery. The median age at the time of placement of the ABPM was 9 years, with a prevalence of arterial hypertension in 46%, 24% nondipper type. In our hypertensive group, there were no significant association with the surgical technique, with 47% corrected with end to end anastomosis, 47% with aortoplasty with construction of a subclavian flap and 6% with mixed technique.
Conclusions: In our population, we verified the prevalence of arterial hypertension in about half of the patients. This is consistent with the literature, that considers hypertension as a major concern at a long term follow-up, even in early corrected aortic coarctation patients.

P-192

Continued Surgical Review: a Multidisciplinary Clinical Model for Quality Control and Mentoring

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Objective: To present a new modality for continuous monitoring and improvement of surgical performance, based on clinical review of cases by a multidisciplinary team. To evaluate a novel, stage-based mentoring system for young consultants in congenital cardiac surgery.
Methods: A new activity was instituted where all procedures performed by three congenital cardiac surgeons were retrospectively reviewed every fortnight. Patient characteristics, diagnosis, surgical procedure and all Adverse Events (AEs) including near misses were presented and discussed for each procedure in a slide-show format. All entries were categorized in an ad hoc dataset as "mentored", when surgeries were performed by the junior consultant, and "non-mentored" when performed by two senior surgeons. Mentoring consisted in case-selection, surgical assistance or both. Results were compared with multiple ANOVA analysis.
Results: Between Jan 1st 2010 and June 31st 2011, 38 meetings were held, with 100% coverage of the surgical activity. In 18 months, 222, 239 and 268 procedures (total 729) in the first,

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second and third semester, respectively, were reviewed. These included Central Cardiac Audit Database (CCAD)-eligible consecutive congenital cardiac operations (197, 215 and 213 operations, respectively) and related subsequent interventions (during same hospitalization). In the non-mentored activity the averaged incidence of AEs was 0.76/Op, 0.32/Op and 0.27/Op, respectively. In the mentored activity incidence of AEs was 0.39/Op, 0.25/Op and 1.15/Op, respectively. The incidence of near misses was 0.05/Op, 0.06/Op and 0.06/Op respectively, whereas mortality rates across the board were 1%, 2.8% and 2.3% respectively. The mentored activity had a significantly lower incidence of AEs than the non-mentored one in the first semester of 2010 (0.39/Op vs. 0.76/Op, $p = 0.001$) but a significant higher incidence in the first semester of 2011 (1.15/Op vs. 0.27/Op, $p = 0.001$). The activity has led to the preparation of two new sets of institutional guidelines, initiated two new clinical activities, and led to the adoption of different surgical techniques.
Conclusions: Quality monitoring in a clinical setting allows high degree of scrutiny of surgical performance and immediate interventions when required. This mentoring system allows a controlled integration of surgeons still in a learning curve with no significant impact to the unit's level of performance.

P-193
Mild hepatic disorder occurs early after Fontan procedure and worsens at time progresses

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Background: Hepatic impairment may occur early after Fontan procedure. But hepatic damage is indistinct because of its mild change. In Fontan patients elevation of central venous pressure causes hepatic fibrosis for a long time. In patients with hepatitis thrombocytopenia is index of liver fibrosis. We attempted to detect the process of hepatic lesion from early-stage to middle-stage in Fontan patients.
Methods: The medical records of 122 patients after Fontan surgery were reviewed. Blood tests ranged 1 month to 15 years after procedure. We divided the term after Fontan four groups: from 1month to 2 years; from 2 to 5 years; from 5 to 10 year; from 10 to 15 years. Labolatory data of healthy person were used as control who were matched sex and age. First, indexes of liver abnormality in Fontan patients were compared with those of controls. Second, we defined thrombocytopenia as platlet count (Plt) 19.0×104/ml under, which was the lower (2.5th percentile) reference limit of Plt in control. We compared cardiac functions between patients with thrombocytopenia and patients with non-thrombocytopenia.
Results: Hepatopacific markers, such as alanine aminotransferase, aspartate aminotransferase, lactate dehydrogenase, and ganma-glutmyl transpeptidase were significantly higher than those of matched controls in all periods after Fontan procedure. The levels of total bilirubin became significantly higher after 2 years postoperatively and its disparity continued. Similarly Plt decreased after 5 years and total protein after 10 years. Plt lessened more highly in later stage. The number of patients with thrombopenia was 46 in Fontan group. The levels of brain natriuretic peptide and ejection fraction of major ventricle were not different in patients with thrombopenia and in patients with non-thrombopenia.
Conclusion: Our study showed liver abnormality arose early after Fontan procedure. Each variation of marker appeared in different period. Thrombocytopenia, which reflected hepatic fibrosis, occurred in Fontan patients certainly and developed with time. But hepatic impairment in Fontan patients was not introduced by viral hepatitis. We don't know whether thrombocytopenia is proper to assess hepatic fibrosis in Fontan patients. We should establish assessment method of liver damage by routine blood tests in patients after Fontan procedure.

P-194
Neuropsychiatric development and health in children with and without congenital heart block born to mothers with Ro/SSA autoantibodies – a retrospective follow-up

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Introduction: Women with Ro/SSA autoantibodies have an increased risk of having a child with neonatal lupus erythematosus (NLE) which includes manifestations such as congenital heart block (CHB). Some studies indicate that the CNS may be affected in children with NLE, and other studies have shown that children of women with SLE have an increased risk for learning disabilities. We therefore investigated neuropsychiatric development in children with and without CHB born to mothers with Ro/SSA autoantibodies.
Methods: Individuals born between 1980 and 2010 were selected from a population-based cohort of CHB patients. Medical records for siblings with and without CHB were retrieved from children healthcare centers and school health services and data on neuropsychiatric development (locomotor skills, hearing, speech, attention, learning, behavior, anxiety and depression) was extracted. Records from 109 individuals, 58 with CHB and 51 of their siblings without CHB were collected. A questionnaire was sent to the mothers to gather information on maternal diagnosis and treatment during pregnancy.
Results: The median time of follow-up was 12.7 years (25th-75th percentile: 8.1-17.5 years). Neuropsychiatric symptoms or disease were reported in 22 (20%) of the 109 children, 15 of which had CHB and 7 without CHB. Among the mothers of these 22 children, only one was steroid-treated during pregnancy. The most commonly reported problems were speech (9%), locomotor (7%), learning (7%), and hearing impairment (7%). Two categories observed reached a statistical difference between the groups, attention deficit; 10% in the CHB group and 0% in siblings ($p < 0.02$) and learning impairment; 12% in the CHB group and 2% in siblings ($p < 0.05$). Among the 9 mothers of children with attention deficit and/or learning impairment, 7 mothers (78%) were diagnosed with SLE ($p < 0.01$). None of the siblings had any reported neuropsychiatric diagnosis, whereas 4 children with CHB had a reported neuropsychiatric diagnosis. One female had dyslexia, one male had autism and two males had ADHD.
Conclusions: Our data suggest that impairment in neuropsychiatric development in terms of attention deficit and learning impairment is more frequent in children with CHB than in their siblings. However, this risk appears predominantly confined to children of mothers with SLE.

P-195
B-type natriuretic peptide according to magnetic resonance imaging findings in surgically repaired tetralogy of Fallot

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Objective: B-type natriuretic peptide (BNP) is an established diagnostic marker in congestive heart failure and left ventricular dysfunction. Recent reports suggest a role for BNP to detect right ventricular dysfunction, too. In patients with surgically repaired tetralogy of Fallot (TOF) we found a correlation between BNP and echocardiographic parameters reflecting right ventricular volume load (Int J Cardiol 2010;143:130-4). The aim of this study was to evaluate the association of plasma BNP and magnetic resonance imaging (MRI) findings in this patient group.
Methods: Plasma BNP concentration was measured (Triage BNP assay, Alere®) in all patients with repaired TOF who underwent cardiac MRI evaluation in our hospital. BNP levels were compared with age and gender-specific normal values, and additionally with evaluated MRI parameters.
Results: 27 Patients (16 males, 11 females; aged 8.2 to 39.7 years) were evaluated at a median age of 15.6 years (interquartile range [IQR] 12.5–19.0 years) and 13.5 (median; IQR 10.0–15.0) years after surgically repair of TOF. Plasma BNP levels were between 5 and 57pg/ml (median 16pg/ml; IQR 8-26pg/ml). According to age and gender, BNP was normal in 14/27 and slightly increased in 13/27 patients (BNP standard deviation score median 2.0; IQR 0.6-3.6). There was no correlation between BNP and age at corrective surgery, but BNP increases with space of time from surgical repair to MRI assessment. BNP was significantly correlated ($r = 0.47$, $p = 0.01$) with right ventricular end diastolic volume (144 ml/m^2; IQR 121-166 ml/m^2), but neither with right ventricular ejection fraction nor pulmonary regurgitation fraction.
Conclusion: In the half of patients with surgically repaired TOF BNP plasma concentration was slightly increased. There was a significant correlation between BNP and end diastolic right ventricular volume assessed by MRI. Therefore, MRI data support our previous echocardiographic data that elevated or increasing plasma BNP levels can indicate right ventricular volume load. However, a normal BNP level does not exclude right ventricular dilatation.

P-196
Central Hemodynamics in Pediatric Patients after Successful Aortic Arch Repair

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Introduction: In patients after aortic arch repair, future hypertension is one of the most important problems. It is reported that the prevalence of hypertension increases with age. Recently, the importance of central blood pressure evaluation in the management of hypertension has been recognized. Therefore, we examined the central hemodynamics in patients after aortic arch repair.
Methods: This study enrolled 20 patients aged under 15 year (7.7 ± 3.3 years old) with aortic coarctation (14) or interrupted aortic arch (6) after a successful aortic arch repair (i.e. no pressure gradient in aortic arch). The methods of aortic arch repair were extended to end-to-end anastomosis in 14, subclavian flap in 5 and Blalock-Park operation in 1. The period after aortic arch repair was 7.6 ± 3.3 years. The patients diagnosed as hypertension judging from Japanese guideline were excluded. None of them were taking any medication and none were diagnosed to be suffering from Turner syndrome. The aortic pressure waveform was recorded using a pressure sensor mounted catheter, and central hemodynamical parameters were compared with those in normal aortic circulation patients.
Results: There were no significant differences in age, height and body weight between the two groups. Central systolic blood pressure (100.4 ± 10.1 vs. 90.5 ± 10.7 mmHg, $p = 0.0013$) and pulse pressure (38.6 ± 8.0 vs. 32.3 ± 4.8 mmHg, $p = 0.0011$) were significantly high in patients after aortic arch repair. Augmentation index (25.9 ± 14.2 vs. $6.0 \pm 16.5\%$, $p < 0.0001$) and heart rate corrected augmentation index (30.4 ± 14.6 vs. $13.4 \pm 19.1\%$, $p = 0.0012$) were also high in the patients.
Conclusions: Although the brachial blood pressure is not elevated, central hemodynamics demonstrated abnormal profile in patients after aortic arch repair even in childhood. One of the reasons of the elevated central aortic pressure is the enhanced aortic pressure wave reflection. It is important for close observation of these patients, although they do not demonstrate hypertension in childhood.

P-197
Sense of Coherence, Rather Than Exercise Capacity, is the Stronger Mediator to Obtain Health Related Quality of Life in Adults with Congenital

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Objective: Sense of coherence (SOC) is a psychological approach that focuses on factors that support human health and well-being, rather than on factors that cause disease. It assumes its three components comprehensibility, manageability and meaningfulness are resources to enhance quality of life directly. This study aimed to evaluate the relationship between SOC and quality of life in adults with congenital heart disease (CHD) and to compare it with the relationship of exercise capacity and quality of life.
Patients and methods: From April 2010 to Mai 2011 we consecutively included 546 young adults (236 female, 26.9 years, range 16-71 years) with various CHD in the study. Patients completed the SOC-13 questionnaire and the health-related quality of life questionnaire SF-36. Afterwards they performed a cardiopulmonary exercise test.
Results: In adults with CHD, SOC was enhanced compared to reference values (CHD: 74,0 [63.8 ; 81.0 vs. reference value: 69.7 [68.5 ; 69.7] ; $p < .001$) corresponding to 106.1% [91.8 ; 116.7%] of predicted reference value. SOC was not associated with the underlying heart defect ($p = .565$) or heart defect severity ($r = .044$; $p = .301$). It was moderately related to all dimensions of quality of life ($r = .260$ to $r = .686$, $p < .001$) except to health transition. It was only poorly associated with exercise capacity ($r = .098$; $p = .023$) and age ($r = -.097$; $p = .023$).
Conclusions: Adults with CHD have an enhanced SOC. SOC is moderately correlated with quality of life, and seems to be a stronger mediator of health related quality of life than exercise capacity. SOC might explain the rather good quality of life in patients with CHD despite their reduction in exercise capacity. Both must be focused on in the management of patients with CHD.

P-198
Long-term Survival, Exercise Performance and Quality of Life in patients after Fontan Surgery – Results From a Single Centre

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Objective: Several modifications of the Fontan procedure evolved throughout the last decades. This single-center study evaluates survival, as well as functional outcome on long-term follow-up in the context of the different surgical modifications.
Patients and Methods: All of the 308 patients born before the year 2000 and having undergone a Fontan procedure in our institution were included into the study. From October 2006 to February 2011 we prospectively studied 146 of the 215 eligible survivors with a median age of 20.7 years (13.8; 28.0 years, 58 female). Patients completed the health-related quality of life questionnaire SF-36 and afterwards they performed a cardiopulmonary exercise test.
Results: After a median follow-up of 11.7 years (6.2; 20.1) 91 (29.5%) patients had died and 5 patients (1.6%) had undergone heart transplantation. Survival free from transplantation after 30 day and 5, 10 and 20 years was 90.2%, 84.1%, 78.4% and 63.6%, respectively. The recent techniques improved survival ($p = .007$). Peak oxygen uptake was reduced to 23.8 ml/min/kg (18.9; 28.9 ml/min/kg) corresponding to 64.7% of predicted. There were no differences in-between the different Fontan types ($p = .975$). Quality of life in the fields of physical functioning, general health and vitality were reduced in Fontan patients.
Conclusions: This study shows that patients profited from the experience and modification of the Fontan procedure in terms of short- and long-term survival due to a decrease in perioperative mortality. Further profits with regard to exercise performance and quality of life could not be detected.

P-199

Partial anomalous pulmonary venous connection to superior vena cava surgery in Adults: Our experience

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Objectives: Partial anomalous pulmonary venous connection (PAPVC) to superior vena cava (SVC) occurs in 10-15% patients with an atrial septal defect (ASD). PAPVC surgery aims to ensure a good drainage of the pulmonary veins (PPVV) into the left atrium, without residual shunt, neither residual SVC/PPVV stenosis, nor changes in heart rhythm. We present our experience with patients older than 14 years of age.
Material & Methods: Retrospective analysis of 26 patients with PAPVC to SVC operated in our Grown up Congenital Heart (GUCH) Unit during the last decade. Diagnosis made with echocardiography 100% and cardio-resonance 85%. Catheterization performed only in cases of doubtful operability and/or presence of coronary risk factors. Surgery is indicated if QP/QS >1,5 and there are clinical data of hemodynamic overload.
Results: Age: 37 ± 18 years, 65% males. Preoperative studies show ASD (88%), Arrhythmia (19%), ⩾ moderate tricuspid insufficiency (TI) (11%), double VCS (19%), QP/QS $2,3 \pm 0,7$ and systolic pulmonary pressure (PSP) 39 ± 10 mmHg. Functional NYHA class was I (54%), II (38%), III (8%). PAPVC location was low (atrio-caval junction – 31%), half (below azygos – 50%), and high (same azygos level – 19%). Median sternotomy was performed with cardiopulmonary bypass and moderate hypothermia. We used three surgical techniques: VCS septation with venotomy (65%), Warden (8%), and septation from atriotomy (27%). Tricuspid annuloplasty was associated in 11%. There was no early or late mortality. Two patients (8%) had arrhythmia in the postoperative hospitalization. Mean postoperative intubation time was 9 ± 9 hours and hospital stay $8,3 \pm 3$ days. Mean follow up is 41 ± 35 months. During this period, 1 patient (4%) has developed SVC stenosis that was stented percutaneously. All others had normal drainage of the systemic and pulmonary veins. Currently 88% patients are in NYHA class I and 12% in class II. PSP values are normal and TI has improved (88% ≤ trivial, 12% mild)
Conclusions: PAPVC surgery in our GUCH unit has good results (no mortality, minimal morbidity). In the postoperative follow up, the functional class improves, TI diminishes and PSP values reach normal values.

P-200

Percutaneous versus surgical closure of atrial septal defect; still a battle?

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Introduction: To compare percutaneous as opposed to surgical closure of isolated secundum atrial septal defects in terms of hospital stay, efficacy and complications.Results: There were no postoperative deaths. Hospital stay was shorter in group A (2,691,06 days versus 8,772,94 days $p < 0,001$). The rate of early postoperative complications was higher in group S (71,4% versus 25% in group A). On the contrary there were no statistical significant differences between the two groups in terms of late (one year) complications. The incidence of post defect correction arrhythmias was significantly higher in group S compared to group A. There was significant decrease and in some cases normalization of postclosure right ventricular size by both methods (8,57,3 mm, 25% in group A, versus 13,796,59 mm, 35% in group S). The regression though occured early (six months) in group A, while in group S regression was observed after six months of follow up (late remodelling).
Conclusions: Percutaneous ASD closure is a rather safe technique that provides, in expert hands and in highly specialized centers, shorter hospital stay, lower postoperative complication rate, and earlier cardiac remodelling compared to surgical closure.

P-201

Vitamin D kinetics and parathyroid function in patients with congenital heart disease

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Introduction: Recently, it is reported that vitamin D deficiency may contribute to the systemic illness that accompanies chronic heart failure. The reports also suggest the serum Parathyroid Hormone (PTH) level that activates vitamin D in the liver is an useful marker of heart failure. This study was designed to examine vitamin D and PTH levels in patients with congenital heart disease and chronic heart failure.
Methods: We measured the serum 25-hydroxyvitamin D (25-OH D), 1,25-dihydroxyvitamin D and PTH levels in 49 patients with congenital heart disease (age ranging 20-69 years old). Out of 49, 32 patients were in NYHA functional class II or III. Their clinical data such as cardiothoracic ratio (CTR), fraction

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shortening of systemic ventricle, B-type natriuretic peptide (BNP) and percutaneous oxygen saturation (SpO2) were also evaluated. According to previous report, we defined vitamin D deficiency as serum 25-OH D <10ng/ml and hyperparathyroidism as serum PTH >100pg/ml.
Results: Seventeen patients without heart failure did not show vitamin D deficiency and/or elevation of serum PTH level. Out of 32 patients with chronic heart failure, 20 patients (63%) had vitamin D deficiency and/or elevation of serum PTH. These two parameters were inversely correlated each other. In particular, Serum PTH significantly correlated with NYHA (P = 0.003), BNP (P = 0.003) and CTR (P = 0.001).
Conclusions: We showed that vitamin D deficiency and secondary hyperparathyroidism are common in patients with congenital heart disease and heart failure. Serum PTH and 25-OH D levels correlated with several clinical markers of heart failure, suggesting that vitamin D deficiency may deteriorate heart failure in patients with congenital heart disease.

P-202
Microvolt T-wave alternans in adults with the chosen forms of congenital heart diseases
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Introduction: Sudden cardiac death (SCD) in adults with congenital heart diseases (CHD) is usually caused by ventricular arrhythmia (VA). The problem of primary prevention of SCD in this group remains unsolved. Among the noninvasive methods of risk stratification is microvolt T-wave alternans (MTWA), reflecting repolarization inhomogeneity. Aim of the study was to evaluate the incidence of spectral MTWA among adults with chosen forms of CHD; to assess the coincidence between MTWA and malignant VA, as well as other clinical findings presumably associated with an increased risk of malignant VA and SCD.
Methods: The study group:102P (47M), mean age 34,2 ± 13,6y with CHD characterized by pathology within right ventricle or single ventricle physiology (TGA, UVH, Ebstein's anomaly, ccTGA, Eisenmenger syndrome, DORV, CAT, unoperated ToF). Controls: 45 volunteers age and sex-matched. All subjects underwent spectral MTWA test, ambulatory ecg monitoring, cardiopulmonary test, BNP assessement. MTWA positive and indeterminate were labeled 'abnormal'.
Results: Abnormal MTWA was present more often in the study group, compared to controls (39,2% vs 2,3%, p = 0,00001). The highest ratio of abnormal MTWA was in: Eisenmenger syndrome (87,5%), unoperated ToF (66,7%), TGA (50%) and UVH (44,4%). Among subjects with abnormal MTWA sustained ventricular tachycardia (sVT) was observed more often compared to MTWA(-): 19,4% vs 3,6%, p = 0,026. The patients with abnormal MTWA had a lower blood saturation (94,5% vs 97%, p = 0,047), more often were males (58,3% vs 33,9%, p = 0,031), had higher NYHA grade [2,0(min-max 1,0-3,0) vs 1,0(min-max 1,0-3,0), p = 0,04)], worse cardiopulmonary parameters: %PeakVO2 (58,7 ± 15,4% vs 66,4 ± 17,1%, p = 0,034), %HRmax (81,4 ± 11,7% vs 88,8 ± 11,0%, p = 0,003). Factors associated with abnormal MTWA: sVT (OR = 20,7 p = 0,037), male gender (OR = 15,9 p = 0,001) in multivariate regression; in univariate analysis: male gender (OR = 2,7, p = 0,021), presence of VA (OR = 2,6, p = 0,049), NYHA > I (OR = 2,06, p = 0,033), %HRmax, %PeakVO2, VE/VCO2 slope (OR = 0,94, p = 0,005; OR = 0,97, p = 0,042; OR = 1,05, p = 0,037).
Conclusions: The abnormal MTWA occurs significantly more often in adults with CHD characterized by pathology within right ventricle or single ventricle physiology than among healthy subjects. The probability of abnormal MTWA increases in patients with malignant VA, in males and among subjects with heart failure and cyanosis. MTWA might be of potential role in risk stratification for SCD.

P-203
Microvolt t-wave alternans in adult patients with repaired tetralogy of Fallot
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Introduction: Sudden cardiac death (SCD) occurs in 6% patients (P) with repaired tetralogy of Fallot (TOF). Indications for SCD primary prevention in TOF are unknown. Microvolt t-wave alternans (MTWA) is applied for SCD risk stratification.
Study aim: evaluation of spectral MTWA occurrence in adults after TOF repair and determination its association with ventricular arrhythmia (VA) and potential risk factors of VA and SCD.
Methods: Study group:102 adults after TOF repair (46 men) in sinus rhythm aged mean 31,2 ± 8,3years operated at mean 6,2 ± 4,3years. Surgery: palliative Blalock-Taussig shunt- 27P (26,5%), reoperation- 13P (12,7%). 89P (87,3%) NYHA I. Control group: 45 adults (23 men) aged mean 31,0 ± 9,2years. MTWA classification: positive(+), indeterminate(ind), negative(-), non-negative(non-) (including MTWA(+) and MTWA(ind) due to similar prognostic significance). VA classification: malignant-sustained ventricular tachycardia (sVT), potentially malignant-nonsustained ventricular tachycardia (nsVT) and >10 premature ventricular complexes per hour (PVC/h).
Results: 13P-MTWA(+), 25P-MTWA(ind) and 64P-MTWA(-). 15P excluded due to excessive noise. MTWA(+) significantly more frequent in TOF vs. control group (1P (2,2%), P = 0,0001). Male gender domination in MTWA(+) vs. MTWA(-) (P = 0,005). No difference in sVT occurrence, heart rate variability and BNP between analyzed subgroups. Enddiastolic right ventricle diameter (RVEDD) smaller in MTWA(-) vs. MTWA(+) and MTWA(non-) (P = 0,004; P = 0,005, respectively). Pulmonary insufficiency (IP) more frequent in both MTWA(+) and MTWA(non-) vs. MTWA(-) (P = 0,045; P = 0,015, respectively). Greater QT dispersion (QTd) in both MTWA(+) and MTWA(non-) vs. MTWA(-) (P = 0,004; P = 0,04, respectively). Maximal oxygen consumption (peakVO2) greater in MTWA(-) vs. MTWA(non-) (P = 0,012). VE/VCO2 slope smaller in MTWA(-) vs. MTWA(-non-) (P = 0,04). Factors increasing MTWA(+) occurrence in univariate logistic regression: male sex (OR = 8,04), IP (OR = 3,5), RVEDD (OR = 1,14), QTd (OR = 1,03) and in multivariate logistic regression: male sex (OR = 10,5), QTd (OR = 1,06). Factors increasing MTWA(non-) occurence in univariate logistic regression: IP (OR = 3,57), >10PVC/h and/or nsVT and/or sVT (OR = 3,26), RVEDD (OR = 1,11), QTd (OR = 1,03), VE/VCO2 (OR = 1,08) and peakVO2 fall (OR = 0,91).
Conclusions: Abnormal MTWA occurs more often in adults with repaired TOF, especially men, compared to healthy population. Presence of malignant VA does not increase probability of MTWA. Occurrence of abnormal MTWA rises with SCD risk factors like IP, RVEDD resulting in heart failure. MTWA

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potential significance in SCD risk assessment in TOF population needs further observation.

P-204

Late complications after Senning or Mustard type of repair in transposition of the great arteries

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Aim: Patients after Senning or Mustard repair for transposition of the great arteries during late follow-up may develop systemic right ventricular failure and decrease in exercise tolerance. Our aim was to describe the outcome of our population.
Materials and Methods: Patients with TGA from our adult congenital heart disease database were selected for analysis. Clinical, echocardiographic and/or CMR and cardioexercise testing were documented.
Results: There were 47 patients with median age 29 years (). At the last follow-up evaluation 59% of patients were asymptomatic (New York Heart Association class I.). At cardioexercise testing only 27% of patients achieved a predicted peak VO2 >70% of their normal. Echocardiography and/ or CMR showed ⩾ moderate systemic right ventricular dysfunction in 33% and ⩾ moderate tricuspid regurgitation 34% of patients. History of atrial arrhythmia requiring treatment was present in 21% of patients; 41% of patients (5 of 12 patients) were over 30 years of age. Heart failure was present in 15% of patients; 4 of 6 patients over 35 had severe symptoms. After 35 years of age only 50% of patients were alive.
Conclusion: High morbidity and mortality is observed in patients with TGA and Senning or Mustard repair after 30 years of age.

P-205

Late complications after tetralogy of Fallot repair

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Aim: Patients tetralogy of Fallot (TOF) repair during childhood are prone to develop late complications, as arrhythmias and right heart failure. Our aim was to describe the outcome of our cohort of adults patients.
Materials and Methods: Data from TOF patients seen at our centre's adult congenital heart disease clinic between 2010-2011 was analyzed. Clinical, echocardiographic and/or CMR and cardioexercise testing were documented.
Results: There were 128 patients with median age 29 years (18-62). The mean age at first operation was 6.0 +/- 7 years and the mean postoperative follow-up duration was 23 +/- 6 years. All patients underwent TOF reconstruction, 17% with pulmonary valve homograft implant. At the last follow-up evaluation 60% of patients were asymptomatic (New York Heart Association class I.). One third of patients had cardioexercise testing, and the mean peak VO2 was 23 ml/kg/min. Echocardiography and/ or CMR showed ⩾ moderate right ventricular dysfunction in 25% and ⩾ moderate pulmonary regurgitation in 74% of patients. History of arrhythmia requiring treatment was present in 20% of patients; patients over 40 were more likely to develop atrial arrhythmia. Fifteen percent of patients had PM/ICD; one patient had biventricular pacing. Four percent of all cohort was treated for heart failure.
Conclusion: Late complications after TOF repair increase with aging. More commonly patients develop arrhythmia. Regular follow-up in congenital cardiac clinics is mandatory in this cohort of patients.

P-206

24 year-old pregnant woman with ventricular septal defect and pulmonary hypertension

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Introduction: Pregnancy in patients with pulmonary hypertension (PAH) and congenital heart defects (CHD) is associated with a high maternal mortality. Experiences with therapeutic options during and after pregnancy are rare.
Case: A 24 year-old woman presents to our adult congenital heart clinic in the 22nd week of gestation due to a heart murmur. She is acyanotic (SaO_2 94%), a 3/6 systolic murmur is audible and she is NYHA II. Echocardiogram shows a large VSD with bidirectional shunt. During the course of pregnancy no complications occur, Caesarean section is performed in the 37th week of gestation, she gives birth to a healthy daughter. Postpartum she receives prostacycline i.v. for 5 days, heparin as well as oxygen supplementation. 3 months later exertional dyspnoea occurs (NYHA III), cardiac catheterization shows the PA pressure systemic (PAP/SAP: 0.72) with high pulmonary vascular resistance index (PVRI) at 15 $U^{\star}m^2$. O_2 and NO testing results in an increased left to right shunt and a significant reduction of PVRI (3.3 $U^{\star}m^2$). Bosentan therapy is started, after 12 months the 6-minute walking distance (6MWD) improves from 503 m (collapse) to 560 m, PVRI decreases at rest (5.9 $U^{\star}m^2$) and shows sigificant vasoreagibility with O_2 testing (1 $U^{\star}m^2$), PAP is unchanged. Partial closure with a 6 mm fenestrated VSD patch is performed, followed by an unproblematic post-operative period. 12 months after surgery (24 months of Bosentan therapy) the exercise capacity further improves (NYHA II) and 6MWD increases to 682 m. Cardiac catheterization shows a further decrease in PVRI at rest (4.4 $U^{\star}m^2$) as well as a lowered PAP (PAP/SAP: 0.4). Bosentan therapy is stopped, 6 months later the 6MWD improves to 721 m.
Discussion: Adult patients with PAH due to a large VSD without Eisenmenger physiology may show an adequate vasoreagibility of their pulmonary vasculature. Thus an uncomplicated course of pregnancy and postpartal period is possible. Bosentan may support partial VSD closure in due course and can result in an improvement of NYHA functional class and 6MWD.

P-207

Long-term outcome of pulmonary atresia with ventricular septal defect

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Introduction: We reviewed data of all patients with pulmonary atresia and ventricular septal defect examined and treated at our institution between January 1970 and December 2006. The aim was to evaluate anatomical substrates and outcome in this patient group.
Methods: Patient records of 106 patients (48 females) with median follow-up of 9.4 years (range 0-36.5 years) were studied. Angiographic data was available and analyzed in 76 cases.
Results: Confluent true pulmonary arteries (CTPA) were present in 93 (88%) patients. 44 (42%) patients had on average 2.6 major aortopulmonary collateral arteries (MAPCAs). 19 patients (18%) had completely MAPCA dependent pulmonary blood circulation (cMDPC), and of them 8 (42%) had CTPA. 104 patients had altogether 258 operations. Mean age at first operation decreased

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from 19.6 months (95% CI 8.0; 31.1) between 1970-1997 to 1.4 months (95% CI 0.4; 2.4) between 1998-2007. Full repair was possible in 73 patients (69%) at median age of 3.1 years, of whom 66 (90%) patients had CTPA and 13 (18%) patients had cMDPC. Reintervention rate after full repair was 49%. Overall freedom from reintervention at 2 and 5 years postoperatively was 79% and 74%, respectively.
Native pulmonary artery size was significantly bigger in patients who survived ($n = 60$) compared to those who died ($n = 46$). Median McGoon index was 1.4 vs. 1.0 ($p < 0.02$) and median Nakata index was 110 vs. 62 ($p < 0.01$). In patients surviving full repair, CTPA were significantly bigger compared to those who died after repair (Nakata 114 vs. 74, $p < 0.04$). The origin of the pulmonary blood circulation whether from native pulmonary arteries or from MAPCAs, didn't significantly affect the survival. Extremely small or absent intrapericardial pulmonary arteries negatively affected 10-year survival. Estimated 10-year survival was 60%, and 78% for patients who achieved full repair. Additional extracardiac anomalies were present in 36% patients.
Conclusions: In our material of 106 patients with PA+VSD, complete repair has been possible in 73 patients (67%) of which 56 (76%) are alive. Extracardiac anomalies were present in 36% underscoring importance of their recognition. These observations are important for pre- and postnatal counseling.

P-208

Preliminary neuropsychological assessment of adolescents after congenital heart defect repair

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Congenital heart defects (CHD) in children are a risk factor in normal cognitive development. A consequence of a growing number of adolescents after CDH repair is increasing incidence of neurodevelopmental abnormalities and a growing interest in the problem. The objective of the study was neuropsychological assessment of adolescent patients with various surgical and/or interventional procedures due to CHD (GUCH) performed in childhood.
Material and methods: The assessment included 18 patients aged x- 17.6 ± 19.1 years; the group consisted of 13 with past simple and 5 complex CHD, who because of age had to be transferred to adult cardiological care. The tests included the Wechsler Intelligence Scale (WAIS-R (PL)/WISC-R) and neuropsychological tests evaluating memory, attention, praxis, abstract thinking and visuospatial functions (Verbal Fluency Test, Rey-Osterrieth Complex Figure Test, Digit Cancellation Trial, Lucki books- a set of memory and executive function trials, DUM- a visual memory test, 10 words verbal learning trial, Wisconsin Card Sorting Test).
Results: Only four patients (2 after CoAo, 1 after ASD2 and 1 after PVS repair) achieved results not indicative of organic type cognitive dysfunctions. The remaining adolescents demonstrated various degrees of cognitive difficulties typical of CNS dysfunction or damage with organic background. One patient after PDA closure showed impairment of involuntary visual memory only. Seventeen individuals revealed disturbances characteristic for frontal region dysfunctions (decreased verbal fluency in the letter category, attention and praxis disorders, as well as learning impairment independent of material modality), while 16 patients additionally showed visual memory impairment (characteristic of both right hemisphere temporal and frontal region dysfunctions). In 40% of patients after complex CHD repairs (TOF+PA, SV post Fontan) and 1 with AVS/AVR, visuospatial skills impairment was noted, typical for temporo-parieto-occipital region lesions.
Conclusions: 1. Of most commonly demonstrated neurodevelopmental abnormalities, adolescent GUCH patients show executive function impairment. 2. Precise assessment of the association between the type and intensity of cognitive disturbances and cardiological characteristics of the patients requires continued investigations in a more numerous group of patients.

P-209

Increased Transition to Adult Care After Interrupted Aortic Arch Repair in a Multi-institutional Cohort

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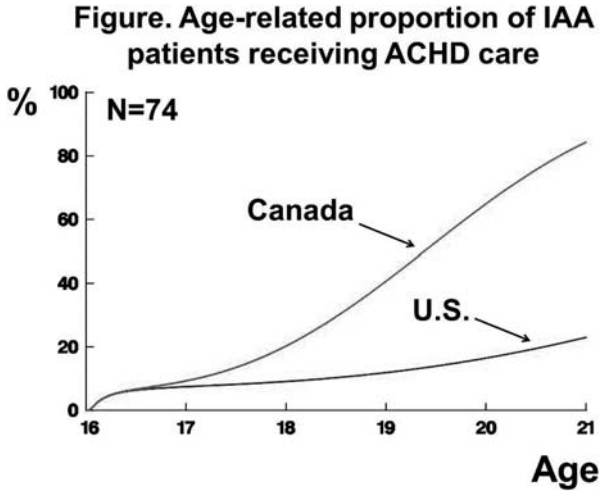
Objectives: Improved survival after congenital heart disease (CHD) surgery has led to the need for deliberate transition to adult congenital heart disease (ACHD) care. We examined this transition in young adults after repair of interrupted aortic arch (IAA), focusing on the differences between Canadian and other American residents.



Methods: Survivors followed into adulthood from a Congenital Heart Surgeons Society (CHSS) inception cohort of neonates (1987-1997) with IAA completed a CHSS developed questionnaire

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on transition to ACHD care (n = 75, age 18-24 years; 12% (9/75) Canadian, no difference in age by Canadian vs. U.S., p = 0.1). Details regarding current care, first ACHD care experience, psychosocial factors, and parental involvement were collected. Features related to demographics, cardiac morphology, index IAA repair, subsequent procedures (interventional and surgical), country of residence and institution were explored for association with time related transition to ACHD care (TRTTAC).

Results: The study population included 65 U.S., 1 Brazilian, and 9 Canadian (8/9 from a single institution) patients. Of the patients who responded, 53/73 (73%, question not answered (QNA) = 2) still received their primary cardiac care from a pediatric cardiologist, and 43/66 (65%, QNA = 9) have never had any form of adult cardiac care. Of the 21/75 (28%) patients who transitioned to adult care, 12 (57%) reported referral from a pediatric hospital. Of these 12, 5 (42%) were from Canada. Of the same 21 patients, 18 (86%) were referred on a non-emergent basis. Of these 18, 8 (44%) were from Canada. TRTTAC was completed in 10% of the total cohort of patients by age 18, 23% by age 20, and 32% by 21. The only factor associated with TRTTAC was living in Canada (p = 0.003) (Figure), with 84% of Canadian patients and 23% of U.S. patients transitioning by age 21.

Conclusions: The system of care at a particular institution may account for the differences seen between countries given the preponderance of Canadian patients from a single institution. While transition to ACHD care is evolving and can be accomplished effectively within many clinical models, the high proportion of TRTTAC in Canada highlights a model warranting further investigation.

P-210

Factors associated with self-reported functional health status in a multi-institutional cohort of young adults with Interrupted Aortic Arch

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Objectives: Improved survival after congenital heart surgery has led to increasing interest in functional health status (FHS) of patients as they transition to adulthood. We sought to identify factors associated with self-reported FHS of young adults with repaired IAA.

Methods: Follow-up in 2010 of survivors (aged 13-24 years) from a 1987-1997 inception cohort of neonates with IAA included completion of FHS questionnaires [Child Health Questionnaire-CF87 (age <18, n = 51) or the Short Form-36 (age > = 18, n = 66)], and a questionnaire about features of 22q11 deletion syndrome (DS) (n = 141 survivors), as few subjects had had genetic testing. Patient characteristics (including 22q11DS features), medical history, and psychosocial factors associated with FHS domains were sought using multivariable linear regression analyses.

Table. Comparison of IAA Patients to Normative Data

CHQ-CF87 Domain	IAA patients <18y Mean (SD)	Normal 10-15 year olds Mean (SD)	P
Global Health (GGH)	77.2 (17.2)	-	-
Physical Functioning (PF)	86.6 (15.6)	88.8 (14.0)	.3
Role/Social Limitations – Emotional (RE)	85.6 (20.7)	85.9 (21.0)	.9
Role/Social Limitations – Behavioral (RB)	88.0 (25.3)	86.5 (21.5)	.7
Role/Social Limitations – Physical (RP)	92.3 (16.7)	88.3 (21.0)	.1
Freedom from Bodily Pain (BP)	87.8 (19.3)	74.4 (23.1)	<.0001
Behavior (BE)	73.0 (16.5)	76.6 (14.6)	.1
Global Behavior (GBE)	71.0 (27.6)	-	-
Mental Health (MH)	77.6 (15.2)	72.7 (16.0)	.03
Self Esteem (SE)	79.3 (15.6)	81.8 (15.8)	.3
General Health Perceptions (GH)	64.5 (14.8)	66.4 (14.6)	.4
Family Activities (FA)	80.0 (23.8)	-	-
Family Cohesion (FC)	73.9 (22.6)	-	-
SF-36 Domain	**IAA patients ≥18y Mean (SD)**	**Normal 18-24 year olds Mean (SD)**	**P**
Physical Component Summary (PF/RP/BP/GH)	52.4 (7.5)	53.5 (9.2)	.3
Mental Component Summary (VT/SF/RE/MH)	49.3 (12.0)	46.1 (13.3)	.04
Physical Functioning (PF)	50.7 (8.0)	53.2 (9.7)	.02
Role-Physical (RP)	50.9 (8.6)	52.8 (9.6)	.09
Freedom from Bodily Pain (BP)	55.9 (8.0)	52.0 (10.6)	.0002
General Health (GH)	49.2 (10.9)	49.7 (11.8)	.7
Vitality (VT)	52.6 (11.3)	47.0 (11.7)	.0002
Social Functioning (SF)	49.1 (10.0)	49.2 (12.3)	.9
Role-Emotional (RE)	49.0 (11.8)	49.8 (12.5)	.6
Mental Health (MH)	50.7 (11.8)	46.9 (13.0)	.01

* Note that a higher score for any given domain denotes better functional health status as related to that category.

Results: Domain scores were significantly higher than norms in 2/9 CHQ-CF87 and 4/10 SF-36 domains, and only lower in the physical functioning domain of the SF-36 (table). Factors most commonly associated with lower scores were those suggestive of features of 22q11DS (low calcium levels, recurrent childhood infections, genetic testing/diagnosis, abnormal facial features, hearing deficits), the presence of behavioral and mental health problems, and a higher number of procedures. Poor FHS scores were less commonly associated with specific anatomy, higher number of medications, lower family income, lower weight and age at the index repair, shorter procedure free interval, and having other medical problems. Depending on the FHS domain, factors explained from 10% up to 70% of score variability (R2 = 0.10-0.70, adj-R2 = 0.09-0.66). Of note, FHS was minimally related to IAA morphology and repair type.

Conclusions: Morbidities related to 22q11DS, psychosocial and recurrent medical problems affect FHS in IAA survivors, and dominate over cardiac history. Nonetheless, these survivors generally perceive themselves to have higher FHS than their peers – a seemingly paradoxical association that may reflect known phenomena (response shift, disability paradox, sense of coherence). Evaluation and surveillance/strategies aimed at definitive surgical treatment, mental health, and genetic issues might be an important program component of cardiac care in the transition from adolescence through early adulthood. Ongoing assessment of FHS in this cohort will be required to detect deteriorations related to increasing complexity and stress associated with mature adult roles.

P-211

Could subclavian flap repair be superior to end-to-end anastomosis for coarctation of the aorta?

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Aim: To evaluate the long-term outcome in patients who had subclavian flap repair (ScF) or end-to-end anastomosis (EEA) of native coarctation of the aorta in childhood. We hypothesised that since subclavian flap repair does not leave a circumferential scar, it would be associated with greater aortic diameter at the repair site and lower flow velocity compared to end-to-end repair as assessed by cardiac magnetic resonance imaging (cMRI).
Methods: From a specialist adult congenital cardiology clinic, we identified all patients older than 18 years of age who underwent repair of coarctation of the aorta in childhood and who had undergone cardiac magnetic resonance imaging prior to any reintervention. Operation type, minimal aortic diameter at the repair site, velocity at repair site and Coarctation Index (ratio of diameter at repair site to diameter at diaphragm) were recorded and an index of <0.75 defined re-coarcation. Chi square and unpaired Student's t-test were performed and a P value <0.05 was considered statistically significant.
Results: Data was available for 47 patients (ScF = 22, EEA = 25). median age 28.2years (17.8 – 76.8), median age at repair 3 years (< 1 to 31), 24 (51%) female. The Coarctation Index was significantly better in the ScF group vs the EEA group (0.89 ± 0.28 vs 0.74 ± 0.19, $P = 0.046$) and statistically fewer patients in the ScF group had a Coarctation Index <0.75 (Chi squared 4.9, $P = 0.03$). Flow velocity at the repair site was lower in the ScF group than in the EEA group ($1.57(\pm 0.44)$ vs $1.74(\pm 0.57)$m/s) but this did not reach statistical significance.
Conclusion: There is a lower incidence of re-coarctation of the aorta in adults who underwent repair using the subclavian flap technique versus those who underwent end-end anastomosis. Further studies are needed to determine whether this translates into reduced need for reintervention, and superior clinical outcomes in these patients.

P-212
Long term follow-up of repaired tetralogy of Fallot: impact of shunt palliation.

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Background: Palliation of Tetralogy of Fallot (TOF) with systemic-to-pulmonary shunts has been the standard for symptomatic neonates for decades. Currently many centers promote all neonates and infants for primary total correction; however such strategy remains controversial.
Aim: To evaluate the impact of previous palliation in operated-TOF on adverse clinical events, functional capacity, biventricular volume and function, pulmonary regurgitation (PRF) and pulmonary anatomy (PA) and flow pattern.
Method: From Sept. 2003 to Dec. 2011, 280 operated-TOF have been evaluated. After excluding patients <15 years and/or already re-operated, the study population was constituted by 148 patients (mean 28 ± 11 yrs, range 15-67); 59% of them had a transannular patch, 36% an infundibular patch and 5% a RV-PA conduit/homograft. Median age at correction was 2.8 yrs, range 0.2-33 and mean follow-up at the last MR study was $23,5 \pm 8$ yrs. Sixty-six patients (44%) had at least one previous shunt palliation before surgery (Shunt Group, mean age 28.5 ± 10.5 yrs vs. No-Shunt Group mean age 27 ± 11 yrs). In both groups bi-ventricular volumes, EF, pulmonary regurgitation fraction (PRF), RVOT and PA anatomy and flow, functional capacity and adverse outcome were evaluated.
Results: Patients with previous shunt underwent intra-cardiac correction later (3,9 years, range: 0,2-25 vs. 2 years, range: 0,3-33 $P = 0,008$) and present more pulmonary branches stenosis (39% vs. 13%, $p = 0,004$). Comparing the two groups, functional capacity, LV End-systolic, End-diastolic, EF and adverse clinical events were not significantly different. RV End-Diastolic Volume (RVEDV) was slightly higher in No-shunt Group (147 ± 49 ml/m2 vs. 138 ± 44 ml/m2) but without reaching significativity. However if we take into account only pts operated by means transannular patch (87 pts), RVEDV and PRF were significantly different between the No-shunt and shunt group (respectively 167 ± 44 ml/m2 vs. 149 ± 38 ml/m2 $p = 0.04$; $43,3 \pm 11\%$ vs. $35 \pm 12\%$, $p = 0,001$).
Conclusion: our data confirmed that previous shunt in TOF leeds to a significant incidence of pulmonary branches distortion but is not associated to adverse clinical events or impaired functional capacity. Moreover in patients operated by means transannular patch, previous shunt leeds to a less PRF and less RV dilatation.

P-213
Management of pulmonary hypertension in Down syndrome

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The incidence of congenital heart disease CHD in Down Syndrome (DS) is about 40%. Many will have large septal defects. Previously these were often unrecognised and untreated leading to Eisenmenger syndrome. In addition, there are many other reasons for pulmonary hypertension PH in DS.
The pulmonary hypertension clinic at the Bristol Heart Institute is responsible for a population of around 6.5million. A total of 208 patients have been seen in this service since its inception in 2005, 59 with DS. Transition arrangements cover the transfer from our paediatric clinic to the adult clinic. Shared care arrangements with the Hammersmith hospital have led to the clinic being performed jointly in Bristol.
Before 2005, many patients with DS were unable to access therapy due to the excessive journey times; the furthest location being 222 miles away, representing a 14 hour round trip or journey by fixed wing aircraft. Such a journey to travel up to London in addition was often prohibitive for a patient with learning difficulties and their carers.
Of the 59 patients with DS, with mean age 28 years, 17 were male. All had CHD, 8 having had palliative cardiac surgery, the rest being untreated. At start of therapy, mean oxygen saturations were 65% and mean six minute walk distance (6MWD) was 120m.Treatment was with oral medication in 28 patients for an average of 1.7 years. Intravenous therapy was not deemed to be appropriate in any patient and one received inhaled Iloprost. Oral sildenafil was avoided in male patients with DS due to the side effects. 17 received Bosentan, 13 received sildenafil (4 having dual therapy) and one received Ambrisentan. Others were either too well (NYHA class II) or too stable to want therapy.
There were no complications of therapy. None had abnormal liver function tests, 6MWD increased after therapy by a mean of 45m and quality of life was significantly improved on questionnaire (CAMPHOR). Only 7 patients died in the 6 years of the clinic.
This early data represents benefits of therapy for a specific group of patients previously denied access because of their special needs.

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P-214

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

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Background: Septic shock is one of the major causes of death in children and is characterized by a massive inflammatory response. To present date, no studies have been performed to assess the impact of such an 'inflammatory hit' on aortic wall structure and myocardial performance. Given the strong similarities in inflammatory pathways between septic shock and atherosclerosis, aortic wall abnormalities and associated ventricular sequelae may be expected.The objectives of the current study were therefore to prospectively assess aortic elasticity and biventricular function in a group of pediatric meningococcal septic shock (MSS) survivors by using MRI.
Methods: Eighteen MSS survivors (8 male; mean age ± standard deviation 14.5years ± 3.9; imaging performed 8.2years ± 2.4 after MSS) treated with at least 2 inotropic and vasoconstrictive agents for 48 hours or longer and 18 for age and gender matched controls were studied. Routine MRI was used to assess aortic pulse wave velocity (PWV) and systolic and diastolic biventricular function. Independent-samples-t-test and Pearson-correlation-coefficient were used for statistical analysis.
Results: MSS patients showed reduced aortic elasticity vs. controls (PWV in aortic arch: 4.1m/s ± 0.3 vs. 3.3m/s ± 0.5, $P < 0.01$; PWV in descending aorta: 3.9m/s ± 0.9 vs. 3.2m/s ± 0.4, $P < 0.01$). Systolic biventricular function was preserved (LV ejection fraction 57% ± 8 vs. 56% ± 6, $P = 0.74$; RV ejection fraction 56% ± 8 vs. 52% ± 6, $P < 0.01$), whereas biventricular mass was increased (LV 52.1gram/m2 ± 8.4 vs. 36.0gram/m2 ± 9.9, $P < 0.01$; RV 26.8gram/m2 ± 6.5 vs. 10.4gram/m2 ± 5.0, $P < 0.01$). Also, delayed biventricular relaxation was found after MSS: peak filling rates corrected for end-diastolic-volume (PFREDV) across the mitral and tricuspid valve were significantly reduced (mitral: PFREDV of E wave 2.54 ± 0.56 vs. 3.08 ± 0.63, $P = 0.01$; PFREDV of A wave 1.10 ± 0.26 vs. 1.31 ± 0.30, $P = 0.03$; tricuspid: PFREDV of E wave 1.81 ± 0.44 vs. 2.09 ± 0.29, $P = 0.04$; PFREDV of A wave 1.11 ± 0.22 vs. 1.42 ± 0.39, $P < 0.01$). Increased PWV in aortic arch and descending aorta were associated with increased LV mass ($r = 0.62$, $P < 0.01$, and $r = 0.51$, $P < 0.01$, respectively) and delayed LV relaxation parameters.
Conclusions: Despite adequately preserved systolic biventricular function, reduced aortic elasticity in pediatric patients after MSS may indicate aortic wall pathology, being associated with biventricular hypertrophy and concomitant delayed biventricular relaxation. Long-term prognosis after MSS may therefore be adversely affected considering the cumulative effects of cardiovascular disease during a lifetime.

P-215

Cardiopulmonary Exercise Test Responses in Children with Obstructive Sleep Apnea Syndrome

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Introduction: There is growing evidence linking obstructive sleep apnea syndrome (OSAS) with multiple cardiovascular and metabolic diseases. Studies in adults found that OSAS patients have also reduced exercise capacity, but there are limited data about cardiopulmonary exercise test (CET) responses in children with OSAS. The aim of this study is to evaluate cardiopulmonary responses to exercise in children with OSAS.
Methods: Twenty seven subjects, without any systematic disease, aged 7 to 14 years (mean age 10,5 ± 1,8 years), referring for evaluation of systematic snoring (⩾4 nights/week), underwent overnight polysomnography and CET on a cycle ergometer (Protocol: maximal incremental 10 Watt/min). According to the Apnea Hypopnea Index (AHI) subjects were divided into two groups: A. mild OSAS (AHI = 1-5, n = 15), B. moderate-severe OSAS (AHI >5, n = 12). The results were compared to those of 13 healthy control subjects matched for age, sex, and body size.
Results: There were no differences in age, sex, BMI, systolic blood pressure (SBP) at rest, SBP and diastolic blood pressure (DBP) at exercise among the groups. Diastolic BP at rest was significantly higher in children with OSAS (63,7 ± 7,5 mmHg vs 56,9 ± 2,9 mmHg, $p = 0,02$), as well as work rate on CET (101 ± 30Watt vs 79 ± 19Watt, $p = 0,15$). Significantly lower were VO2max (40,3 ± 8,4 vs 47,55 ± 7,9 ml/kg/min, $p = 0,13$) and VO2max (%) (77,7 ± 15 vs 93 ± 10,5, $p = 0,02$).
Conclusion: The present study demonstrates that young patients with OSAS have early in life a distinctive response to graded exercise, characterized by higher work rate, and lower VO2max. Early identification of OSAS using CET shows promise for selecting patients at risk for this disorder in the clinical setting.

P-216

Obstructive Sleep Apnea Syndrome and Cardiovascular System in Children

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Introduction: Obstructive Sleep Apnea Syndrome (OSAS) has been shown to be an independent risk factor for cardiovascular disease in adults. However, a few data are known about the effect of OSAS on cardiovascular system in children. The aim of this study is to determine changes on cardiovascular system in children with OSAS.
Methods: Twenty seven subjects, without any systematic disease, aged 7 to 14 years (mean age 10,5 ± 1,8 years), referring for evaluation of systematic snoring (⩾4 nights/week), underwent overnight polysomnography and complete echocardiographic assessment. According to the Apnea Hypopnea Index (AHI) subjects were divided into two groups: A. mild OSAS (AHI = 1-5, n = 15), B. moderate-severe OSAS (AHI >5, n = 12). Blood pressure (BP), lipidaemic profile and CRP were measured. The results were compared to those of 13 healthy control subjects matched for age, sex, and body size.
Results: There were no significantly differences in age, sex, BMI, lipidaemic profileand systolic BP at resr. Children with OSAS had significantly higher diastolic BP (63,7 ± 7,5 mm Hg vs

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56,4 ± 3 mm Hg, p = 0,02) at rest, CRP levels (0,46 ± 0,88 mg/dl vs 0,1 ± 0,3 mg/dl, p = 0,036) and Right Ventricular end-Diastolic dimension (RVDd) (16,3 ± 3,1 mm vs 13,1 ± 3,5 mm, p = 0,05). Left ventricular dimensions (Left Ventricular end-Diastolic dimension – LVDd, Left Ventricular diastolic mass – LVdmass, Left Ventricular Posterior Wall diastolic –LVPWd, IntraVenticular Septum diastolic – IVSd)) were not statistically significant different between the two groups and were within normal limits.
Conclusion: The present study demonstrates that young patients with OSAS have early in life changes on cardiovascular system. Plasma CRP levels and diastolic BP were increased among children with OSAS and right ventricular dysfunction was apparent in early life.

P-217
Pulmonary functions before and after cardiac surgery in infancy

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Objectives: To assess pulmonary functions before and after cardiac surgery in infants with congenital heart diseases and pulmonary overflow, and to clarify which echocardiographic parameter correlates best with lung mechanics.
Methods: Thirty infants with left to right shunt congenital acyanotic heart diseases and indicated for reparative surgery of these lesions between 2008 and 2009 were assessed by echocardiography and pulmonary functions before the operation and at 6 months postoperatively. The following infant pulmonary function tests were performed: tidal volume (VT), respiratory rate (RR), compliance (Crs) and resistance (Rrs). Functional residual capacity (FRC) and airway resistance was assessed by baby body plethysmography.
Results: The mean age of patients was 10.47 ± 3.38 months and their mean weight was 6.81 ± 1.67Kg. VSD and combined lesions were the main cardiac diseases (26.7%). Comparison of the pulmonary function tests revealed a statistically highly significant improvement of all parameters between the pre-operative and at 6-months post-operative visits ($p < 0.0001$). Systolic pulmonary artery pressure had a statistically significant negative correlation with Crs ($r = -0.493$, $p = 0.006$) and positive correlation with FRC ($r = 0.450$, $p = 0.013$). PA and LA sizes had a statistically negative correlation with Crs/Kg ($r = -0.398$, $r = -0.395$, $p = 0.029$, $p = 0.031$, respectively). While PA size had statistically positive correlation with Reff and sReff ($r = 0.416$ and 0.604, $p = 0.022$, $P = 0.0001$, respectively).
Conclusion: Surgical correction of congenital heart disease of left to right shunt has positive impact on lung compliance and airway resistance and these parameters are closely related to left atrial size, pulmonary artery pressure and its size.

P-218
Tissue Doppler, strain and strain rate echocardiography in healthy newborns and infants

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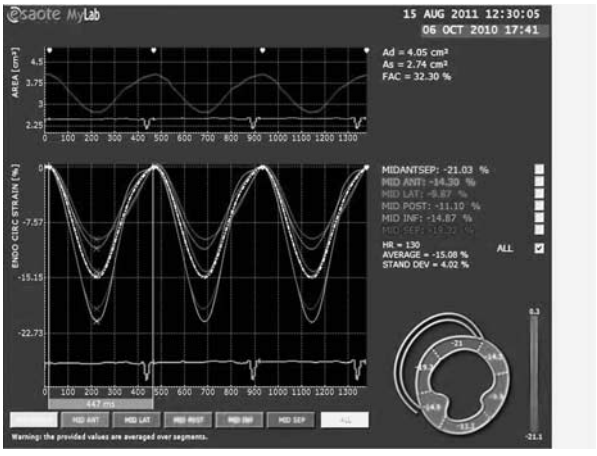
Objective: To evaluate the value of tissue Doppler, strain and strain rate echocardiography in healthy infants.
Materials and Methods: 149 healthy babies aging between 1 day and 3 months were included in this study. Subjects were separated to four groups: thirty-two healthy preterm infants (36-37 weeks of gestational age, group 1), thirty-two healthy term infants (≥ 38 weeks of gestational age, group 2), forty-seven healthy infants (1 month of postnatal age, group 3) and, thirty-eight healthy infants (3 month of postnatal age, group 4). Standard echocardiographic evaluations, pulsed wave Doppler, tissue Doppler, strain and strain rate study for the left ventricle were applied by same person using a MyLab50 echo machine (Esaote, Florence).
Results: Mitral E and mitral A velocities determined by pulsed wave Doppler echocardiography were increased from group 1 to group 4. But this increasing was not statistically significant between group 3 and group 4. Mitral A' velocity determined by tissue Doppler echocardiography was increased from group 1 to group 4. This increasing was found significant statistically between group 2 and group 3, group 2 and group 4, group 3 and group 4, group 1 and 3, and group 1 and group 4 ($p = 0.001$). Mitral E' velocity determined by tissue Doppler echocardiography was increased from group 2 to group 3 ($p = 0.005$), from group 1 to 3 ($p = 0.001$) and, from group 1 to 4 ($p = 0.001$). The longitudinal and circumferential velocity, strain and strain rate values decreased from base to apex in all subjects ($p < 0.001$), (Table, Figure).

Table: Values of longitudinal left ventricular systolic strain

	Segment	Group 1	Group 2	Group 3	Group 4
Apical 4 Chambers	Septal Basal	-13.54 ± 2.67 $^{\beta}$	-13.89 ± 2.54$^{\&}$	-14.47 ± 1.91$^{£}$	-15.72 ± 1.49
	Septal Mid	-10.41 ± 2.48 $^{\beta}$	-10.58 ± 2.60$^{\&}$	-11.40 ± 2.01	-12.56 ± 1.98
	Septal Apical	-5.65 ± 2.05 $^{\beta}$	-6.28 ± 2.33$^{\&}$	-6.58 ± 1.71	-8.15 ± 2.11
	Lateral Basal	-14.34 ± 2.56	-14.69 ± 3.04	-15.21 ± 1.87	-14.89 ± 2.23
	Lateral Mid	-10.81 ± 2.34	-11.66 ± 3.03	-11.58 ± 1.63	-11.34 ± 2.24
	Lateral Apical	-6.29 ± 1.72	-6.76 ± 2.28	-7.06 ± 1.43	-7.24 ± 1.68

$^{\beta}$ = Between Group 1 and Group 4 $p < 0.05$, $^{\&}$ = Between Group 2 and Group 4 $p < 0.05$, $^{£}$ = Between Group 3 and Group 4 $p < 0.05$



Conclusion: Significant cardiac hemodynamic alterations have been occurred during the newborn and early infancy period.

P-219
Decreased systolic function measured by Speckle tracking echocardiography in children with ESRD

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Introduction: Cardiovascular disease is the main cause of death in patients with End-Stage Renal Disease (ESRD) since childhood. In young adults with ESRD abnormal left ventricular geometry and function are correlated with a poor cardiovascular prognosis. Speckle tracking echocardiography (STE) is able to reveal early abnormalities in LV systolic function prior to manifestation of hypertrophy. Therefore, we aimed to assess LV systolic function using STE in children with ESRD and healthy controls.
Methods: 26 children with ESRD and 24 healthy control subjects, matched for body surface area (BSA), were assessed with STE. Parameters related to LV systolic function (global longitudinal strain and global radial strain) were compared in the ESRD and control groups using linear regression analysis. Hypertension was defined as either a systolic or diastolic blood pressure $>$ p95 of the Task Force Report normal values corrected for age and gender.
Results: Children with ESRD were older than their healthy controls, matched for BSA; mean age (SD) 13.4 (4.5) vs 10.5 (4.4) years, mean difference [95% Confidence Interval (CI)] 2.9 [0.4-5.5] years, ($p = 0.025$). At time of the echocardiogram, 15 children were treated with dialysis and 11 children had a kidney transplant. The median (range) time of renal replacement therapy (RRT) was 48 (1-208) months. Twelve of the 26 children (46%) with ESRD had hypertension for more than 3 months after start RRT. At time of the echocardiogram, 16 children (62%) used anti hypertensive drugs at time of the echocardiogram. After adjustment for age, the ESRD patients had a significant lower global longitudinal strain than the control subjects (mean (SD) 18.2% (2.9) vs. 20.5% (2.2), mean difference [95%CI] 1.8 [0.3-3.3], $p = 0.021$). There were no significant differences found for the global radial strain between these two groups. In this small sample size, we found no relation between systolic dysfunction and the duration of RRT, use of antihypertensive drugs or hypertension
Conclusion: Children with ESRD have significantly decreased LV systolic function, measured by global longitudinal strain, compared to healthy matched controls. Early identification of systolic dysfunction might improve the clinical management of these patients.

P-220

Assessment of right ventricular function with tissue Doppler imaging in children with asthma

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Objectives: In the patients with asthma, pulmonary hypertension develops due to recurrent hypoxia and chronic inflammation, leading to right heart enlargement with ventricular hypertrophy. Patients with severe asthma can experience cor pulmonale later in life, but little is known about right ventricular function early in the disease. This study aimed to investigate the right ventricular function in asymptomatic children with asthma as detected by conventional echocardiography and tissue Doppler imaging (TDI).
Methods: Fifty-one paediatric patients (mean age: 10.4 ± 2.2 years) with asthma and 46 age- and sex-matched healthy children (mean age: 10.9 ± 2.4 years) were studied. All subjects were examined on conventional echocardiography and TDI, and 51 patients with asthma had pulmonary function tests on spirometry.
Results: The right ventricle wall was statistically ($p = 0.01$) thicker among the asthmatic patients (4.7 ± 1.5 mm) than among the healthy children (3.6 ± 0.4 mm). But the conventional pulsed Doppler indices of right ventricle did not differ significantly between the asthmatic patients and the control subjects ($p > 0.05$). The results of TDI examining the right ventricular diastolic function showed that mean early diastolic flow velocity (E′) and late diastolic flow velocity (A′) (13.2 ± 2.3 cm/s and 5.1 ± 1.4 cm/s, respectively), E′/A′ ratio (2.7 ± 0.7) and isovolumetric relaxation time (67.7 ± 10.2 msn) of the lateral tricuspid annulus among the asthmatic patients significantly differed ($p = 0.01$) from those among the healthy children (16.4 ± 1.8 cm/s, 8.2 ± 2.0 cm/s, 2.1 ± 0.5, and 46.2 ± 8.7 ms, respectively). From the pulmonary function tests, only peak expiratory flow rate was positively correlated with the E′/A′ ratio of the tricuspid annulus ($r = 0.38$, $p = 0.01$).
Conclusions: This study showed that although the findings of clinic and conventional echocardiography of were apparently normal in children with asthma, TDI demonstrated subclinical dysfunction of right ventricle, which is positively correlated with the peak expiratory flow. These findings signify the diagnostic value of TDI in the early detection and monitoring of such deleterious effects among asthmatic patients.

P-221

Pharmacological Assessment of Anti-Platelet Drugs by Whole-Blood Aggregation and Serum Thromboxane B2 in Kawasaki Disease Patients

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Introduction: Anti-platelet agents are key to prevention of cardiovascular events in Kawasaki disease (KD) patients with coronary lesions (CALs). However, there exist side effects, including bleeding, hepatic damage, gastrointestinal dysfunction, aspirin resistance, and poor compliance with anti-platelet therapy. The latter is usually difficult to evaluate—at least by interview only—because the maintenance of compliance greatly depends on the patient's recognition of necessity for anti-platelet therapy. To assess the efficacy of anti-platelet drugs and simultaneously develop a method for evaluating compliance, we measured platelet aggregation and serum thromboxane B2 (TXB2) levels in KD patients.
Methods: Twenty out of 37 KD patients received anti-platelet therapy (mainly aspirin); the remaining did not receive any medication. Whole-blood aggregation was analyzed using collagen as the stimulus, and evaluated on the basis of platelet aggregation threshold index (PATI). Serum TXB2 was measured with a specific enzyme-linked immunosorbent assay.
Results: The PATI was high in subjects who underwent the anti-platelet therapy, with the exception of 1 case of non-compliance, and the level of serum TXB2 was lower than in non-treated patients. On the other hand, in the non-compliant patient, PATI and serum TXB2 levels were similar to those of the non-treated patients. After 2 months, PATI was increased, and serum TXB2 levels were decreased.
Conclusions: Whole-blood aggregation and serum TXB2 levels were useful for not only the pharmacological evaluation of anti-platelet drugs but also for the objective assessment of compliance.

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P-222

Can Simple Echocardiographic Measures Reduce the Number of Cardiac Magnetic Resonance Imaging Studies to Diagnose Right Ventricular Enlargement in Congenital Heart Disease?

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Background: Right ventricular (RV) enlargement is used as a criterion for treatment of RV outflow tract dysfunction in patients with congenital heart disease (CHD). Although RV volumes are most accurately measured by cardiac magnetic resonance (CMR), CMR is a limited resource. Our objective was to investigate whether simple echocardiography measurements can adequately predict RV volumes below clinical thresholds, thereby reducing the need for CMR in some patients.
Methods: Children with repaired tetralogy of Fallot (TOF), double outlet right ventricle (DORV) or truncus arteriosus (TA) who underwent CMR and echocardiography within a 4-week interval were retrospectively studied. From the 4-chamber view, indexed RV lateral wall length (RVLWLi), end-diastolic perimeter length (RVEDPi) and end-diastolic area (RVEDAi), were measured. Results were compared to CMR indexed RV volume (RVEDVi). Sensitivity and specificity of echocardiography threshold values predicting RV volumes <170 ml/m2 were determined.
Results: 51 children (age 12.7 ± 3.5; M:F 25:26) were reviewed. RVEDAi correlated with CMR RVEDVi ($r = 0.6$, $p < 0.0001$). RVEDPi and RVLWLi did not correlate with CMR. RVEDAi <20 cm2/m2 had 100% specificity to predict RVEDVi $\leq$ 170 ml/m2 (AUC 0.79); reducing the need for CMR in 15/51 patients (29%). A threshold RVEDAi of 22 cm2/m2 would reduce CMR in 21/51 patients (41%) at the expense of 1 false negative result. The coefficient of variation was 14.7% for intra-observer and 9.6% for inter-observer variability.
Conclusion: Specificity of echocardiography measured RVEDAi can be set to predict RV volumes below a 170 ml/ m2 threshold in 100% of cases. This may reduce the need for CMR to determine RV volumes in $\geq$25% of CHD patients, potentially reducing patient burden and costs.

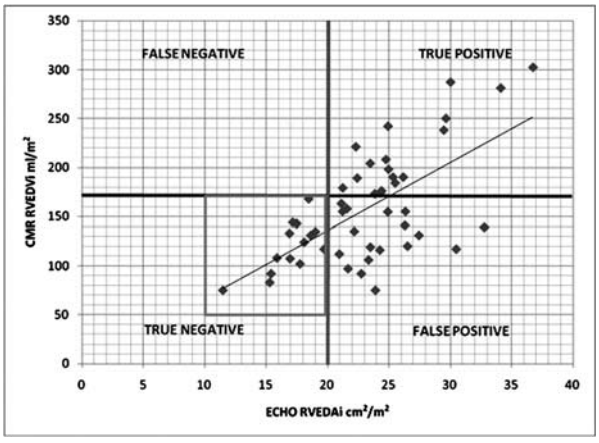

Fig. 1: Correlation between echo RVEDAi and CMR RVEDVI

P-223

Strain and Strain Rate Echocardiography Findings in Children With Congenital Left Ventricular Outflow Tract Obstruction

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Introduction: Obstruction to left ventricular outflow impose increased aferload, and if severe and untreated, result in hypertrophy and eventual dilatation and failure of the left ventricle. However, various studies have reported that subclinic alterations occur in systolic and diastolic myocardial functions in patients with aortic tenosis and preserved ejection fraction. The aim of our study was to evaluate the myocardial functions with strain/strain rate echocardiography in LVOTO patients with normal cardiac functions determined by conventional echocardiographic techniques and comparing them with healthy controls.
Materials and Methods: A total of 58 patients with various degrees of isolated congenital left ventricular outflow tract obstruction and 73 healthy controls were enrolled in this study. Two-dimensional cine-loop recordings of apical 4-chamber and basal short-axis views were digitally stored for off-line analysis. Conventrional echocardiography parameters, longitudinal, circumferential and radial peak systolic strain and strain rate values were determined.
Results: According to the peak systolic pressure gradient patients were divided into two groups; mild (68.9%) and moderate-severe aortic stenosis (31.9%). Global longitudiunal strain (LS)(-23.12 ± 3.6 and -24.2 ± 3.4), and strain rate (LSR) (-1.49 ± 0.32 and -1.6 ± 0.32) were lower whereas circumferential strain (CS) (-25.9 ± 4.7 and -22.4 ± 6) and strain rate (CSR) (-1.82 ± 0.46 and -1.66 ± 0.51) were higher in the patient group than in control subjects. Diffrence was significant for global LSR and CS (<0.05). Regional analysis revealed lower LS values in the basal part of the left ventricular free wall and lower LSR in the basal parts of both of the septum and free wall in the patient group than in control group ($p < 0.05$). Regional CSR, radial strain (RS) and strain rate (RSR) values were not statistically different between patient and control groups.
Conclusion: In conclusion: compatible with the previous studies, impairment of the left ventricular long axis function occurred earlier and was more prominent in basal parts of the interventricular septum and free wall of left ventricle. According to these findings, S/SR echocardiography in addition to conventional methods in evaluation of the left ventricular functions and determining the subtle alterations in LVOTO patients, will be helpful in management and timing of the treatment.

P-224

Dynamic Changes in the Vaso-Vasorum as an Inducing Factor for Vasculitis in Kawasaki Disease

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Introduction: Kawasaki Disease (KD) involves a diffuse and systemic vasculitis of unknown etiology that mainly affects infants and children. Although a lot of analyses had already been done clinically, histopathologically or molecular-biologically about the mechanism of coronary arterial lesions, it is still not well elucidated. The objective of this study is to analyse the process of the formation of aneurysms or the change of coronary arteries themselves using animal model.We investigated the involvement of the invasion from the adventitia in the mechanism of vascular involvement and the development of disease state by scanning electron microscope (SEM), micro CT, and sequential histopathology using murine model of vasculitis induced with Candida albicans water-soluble fraction

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(CAWS), because KD is associated with a very low mortality recently, and we seldom have a chance to get autopsied heart.
Materials and meshods: To prepare the animal model for KD, CAWS was intraperitoneally injected to C57BL/6N mice for 5 days as reported by Ohno et al.
We observed the changes of vasa vasorum at aorta and the orifice of coronary arteries by SEM and micro CT, and also compared neovascularization and distribution at the media and the adventitia quantatively by immunohistochemical analysis.
Results: As previously reported, obvious inflammation were detected 2 weeks after the injection of CAWS, and also subsequent aneurysmal formation, and intimal thickening 4 weeks after it. In model mice, we found micro vessels verging on the adventitia of Aorta (vasa vasorum), and they increased in model mice. We observed each 1w, 2w, 3w and 4w model mice, and found they started increasing 1w after the injection of CAWS, before obvious vasculitis was microscopically detected.
Conclusion: This result indicates that the vasculitis starts by the disorder of vasa vasorum in Kawasaki disease.

P-225
Assessing of vascular Rings, slings and their potentially life-threatening complications using MRI, CT, and Tracheobronchoscopy from newborns to seniors
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Objective: Vascular rings and slings (R+S) are rare congenital cardiovascular malformations, but frequently cause tracheomalacia, which in turn is of prognostic relevance. Aim of this study was to explore the diagnostic accuracy of MRI and CT-angiography for the assessment of cardiovascular malformation, tracheal pathology, and their topographic relationship.
Method: In the last 10 years 106 patients (median [range] 1.5 y [1 week to 56y]) got further examination by MRI (n = 76) or CT (30; only the smallest and critical ill patients) due to suspected R+S. The extension and the type of R+S as well as their relationship to esophagus, trachea or bronchi was assessed and correlated to conventional catherization (if available due to associated heart defects; n = 10), tracheobronchoscopy and intraoperative findings (if R+S was confirmed by MRI or CT).
Results: In 61 out of 104 patients, R+S was found (double aortic arch (AoA), n = 14; right AoA+aberrant left subclavian artery+left ligament arteriosus, 30; pulmonary sling, 6; others, 11). In all patients, MRI/CT confirmed the topographic relation of R+S to trachea corresponding to bronchoscopic and intraoperative findings. Although MRI and CT had a weakness in detecting atretic segments and ligaments directly, there was no false positive case. CT had a better spatial resolution (0.5 mm) and air/soft-tissue contrast, therefore virtual bronchoscopies were possible.
Conclusions: MRI and CT allow a non-invasive assessment of rings and slings including potentially tracheal, bronchial or esophageal compression even in the smallest patients. Furthermore, 3D post-processing improves the preoperative planning. MRI and CT may be considered the first-line imaging modalities in these patients.

P-226
Cardiac Magnetic Resonance in Children with Acute Myocarditis
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Background: Diagnosis of acute myocarditis (AM) is challenging because its clinical presentation may overlap with that of common infectious diseases or be diagnosed as idiopathic dilated cardiomyopathy (DCM). While Cardiac Magnetic Resonance (CMR) imaging has emerged as an important non-invasive tool in the diagnostic procedure of AM in adults, data on CMR in children remain scarce.
Aim: To describe feasibility of CMR and its contribution for the diagnosis and follow-up of AM or for the etiology left ventricular (LV) dysfunction of unknown origin in children.
Methods: From November 2008 to April 2011, 43 children underwent CMR for clinical suspicion of AM with or without LV dysfunction of unknown origin. CMR sequences included unenhanced cine-steady state free precession (SSFP), black-blood-prepared T1-weighted images and T2-weighted images and T1-weighted images (EGE) and 3D late gadolinium-enhanced after injection of gadolinium chelate (LGE). The diagnosis of myocarditis was based on the recently consensus criteria. CMR was repeated during follow-up in children with confirmed diagnosis of AM.
Results: AM was diagnosed by CMR in 30/43 children: 22/30 had LV dysfunction, 8/30 had normal LV function but elevated blood levels of troponin I. T2 hyper-signal was present in 21 cases, EGE and LGE were present in 28/44 cases, the 3 patterns were simultaneously present in 11 cases. Two children died during hospitalization. All survivors with LV dysfunction had normal echocardiography after a median follow-up of 10 months. 24/30 patients had control CMR that revealed in 4 cases the persistence of inflammation in T2-weighted images and in 6 case persisting LGE. No children with AM without LV dysfunction developed dilated cardiomyopathy. The remaining 13/43 children without AM on CMR were diagnosed with DCM: 2/13 normalized after 4 and 30 months of follow-up respectively, and 11/13 are still followed for dilated cardiomyopathy.
Conclusion: CMR in children with clinical suspicion of AM or with LV dysfunction of unknown origin is feasible and useful in the diagnostic work-up. It may help to adapt medical targeted therapy and to be more precise in prognosis assessment in infants recently diagnosed with DCM of unknown origin.

P-227
A Stress Echocardiography study of Exercise Capacity and Cardiac Function following Arterial Switch Operation for simple Transposition of the Great Arteries
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Introduction: The arterial switch operation (ASO) for repair of transposition of the Great Arteries (TGA) was designed to restore the morphological left ventricle as the systemic ventricle and improve cardiac function and longevity. The aim of this study was to assess cardiac and hemodynamic responses to exercise and exercise capacity in a cohort of children who had undergone an ASO.
Methods: We retrospectively reviewed stress echocardiograms performed in 19 children with ASO for simple TGA (Patients) during semi-supine cycle ergometry (SSCE) and compared them to healthy controls (Controls) (n = 29). Subjects exercised on a semi recumbent cycle ergometer to volitional fatigue. Workload was progressively increased every three minutes, by 20 to 40 Watts. Echocardiography, Doppler, heart rate (HR) systolic (SBP) and diastolic (DBP) blood pressures were taken at rest, 1.5 minutes into each stage, immediately, and 3 minutes post-exercise. LVEDI,

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LVESI, shortening fraction (SF), rate corrected mean velocity of circumferential fibre shortening (MVCFc), wall stress at peak systole (sPS), stroke volume index (SVI), and cardiac index (CI) were obtained. Segmental wall motion was assessed.
Results: At rest, SBP and DBP was lower in patients (108 vs 118 bpm, $p = 0.05$; 60 vs 76 bpm, $p < 0.001$, respectively). LVEDI and LVESI were larger in the patients ($p = 0.03$), while SF, MVCFc, and sPS were similar. Resting HR was lower in patients (66 vs 80 bpm; $p = 0.001$), SVI was higher (46 vs 39 ml/m2; $p = 0.02$), and CI was similar. Patients performed less cumulative work (941 vs 1228 J/kg; $p = 0.02$). At peak exercise, DBP (64 vs 76 mmHg; $p = <0.001$) was lower, LVEDI and LVESI were higher in the patients (3.3 vs 2.9 cm/m2; $p = 0.009$; 1.7 vs 1.4 cm/m2; $p = 0.001$, respectively). SF was also lower in the patients (46 vs 50%; $p = 0.04$) and wall stress was higher (73 vs 52; $p < 0.001$). HR was lower in the patients (141 vs 182 bpm; $p < 0.001$), while SVI and CI were similar between groups. Segmental wall motion was normal.
Conclusions: Despite a lower cumulative workload, HR and SF, the exercise capacity and functional responses were normal in many of the patients. Assessing wall motion during SSCE may help screen for coronary artery insufficiency.

P-228
Evaluation of Ventricular Function in HIV-infected pediatric and adolescent patients: the use Speckle Tracking

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Backround: Since introduction of highly active antiretroviral treatment (HAART), HIV has become a chronic disease with decreasing mortality. Multiple studies in adults have shown an early evolution of atherosclerosis and ventricular dysfunction due to chronic inflammatory activation and to the altered lipidic status, for which the prolonged medication is partially responsible.
Objective: To study the ventricular torsion in HIV-infected pediatric and adolescent patients with Speckle Tracking compared to a control group.
Methods: We designed a multicentric study to evaluate ventricular torsion by echocardiography in HIV-infected pediatric and adolescent population, compared to healthy controls matched by sex, age and body mass index. A portable echo-device (Phillips CX50) with a 5 Hz Speckle Tracking transducer was used. Clinical and anthropometric variables, lipid profile, exposure to antiretroviral drugs were recorded. Ventricular function was evaluated with 2D Echocardiography, Mode M, valvular Doppler, tissue Doppler and Speckle Tracking.
Results: 77 cases and 68 controls were included. 59,8% females, mean age of 14.9 years. 96,6% were vertically HIV-infected. 78,2% had undetectable viral load, all but 4 patients were on HAART.
The following left ventricular function parameters were obtained: E/A ratio of 1,6 in the HIV group versus 1,8 in the control group ($p = 0,15$), E/E′ ratio of 5,2 in the cases versus 4,9 ($p = 0,282$), shortening fraction of 36% in the HIV group versus 41,4% ($p = 0,03$), and eyection fraction of 65,8% in the cases versus 72,3% ($p = 0,02$).
25% of the images were analyzed by two technicians, the correlation between the main technician and the expert was good obtaining a coefficient of 0.8.The mean global torsion in the HIV-infected group was 5,4° versus 5,1° in the controls ($p = 0,000$).
Conclusion: Speckle Tracking is an easily reproducible technique, which can be performed by different trained technicians without significant differences. No clinical differences were found between cases and controls. Ventricular torsion measured by Speckle Tracking was significantly different in healthy controls and HIV-infected patients.

P-229
Surgical simulation of congenital heart disease by using flexible biomodels made by streolithography and vacuum casting

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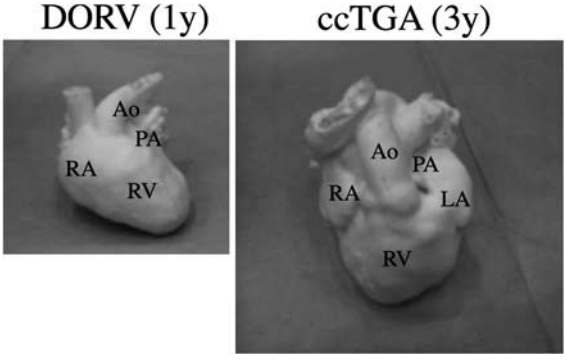
Introduction: Stereolithography is a rapid prototype technology whereby an ultraviolet laser beam selectively polymerizes and solidifies photosensitive and polymeric liquid plastic. By using this technique, 3D volumetric image data of multi-slice CT (MSCT) can be converted into plastic models that enhance our spatial perception of real-life anatomy and pathology. However, the materials of the stereolithography are restricted to specific photosensitive plastic or urethane, which are not satisfactory for idealistic simulation surgery. Recently, a vacuum casting method has been developed in rapid prototype industries, where more detailed prototypes with different stiff materials can be manufactured.
Methods: Six biomodels of various congenital heart diseases were manufactured in this study. The patients include single right ventricle with right isomerism (4m), double outlet right ventricle (1y), congenitally corrected transposition of the great arteries (3y), atrial septal defect (9y), and tetralogy of Fallot (56y). Three-dimensional volumetric data sets of MSCT angiography were converted into standard triangulated language (STL) files to guide the laser beam of stereolithography. Plastic replicas representing the both outer and inner surface of the heart tissues were initially made with stereolithography. Then, urethane materials with appropriate stiffness representing the real heart tissue were injected into the space between the inner and the outer casts by using vacuum casing method. After solidification of the urethane materials, the casts were carefully removed and the final products of flexible heart replicas were obtained (PCT-International Patent Application submitted: PCT/JP2010/061249)).
Results: The vacuum casting in association with stereolithography technique enabled us to manufacture precise replicas with similar texture of the real individual heart in all the 6 cases. This technique also allowed the surgeon to cut and suture, facilitating the simulation of the surgical operation.



Conclusions: The vacuum casting method in association with stereolithography is a promising technique for medical education,

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preoperative practice, simulation of individual surgery, and planning of novel and innovative surgical procedures of congenital heart disease.

P-230

Evaluation of optimal voltage for reduce radiation exposure of area detector computed tomography in children after Kawasaki disease

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Background: It is well known that low exposure voltage of computed tomography (CT) could reduce radiation exposure, but also lowered resolution of the image.
Objective: The aim of this study was to evaluate radiation exposure and image quality of coronary arterial lesions after Kawasaki disease (KD) using area detector CT.
Methods: Consecutive 36 patients who are less than 15 years old after KD (mean age: 8.9 +/- 3.9 years old) were enrolled in this study, all of the patients underwent 320 area detector CT (Acquilion ONE, Toshiba) for assessment of coronary artery lesions; 14 patients with severe coronary arterial lesions, 16 patients with regression of coronary dilatation, 5 patients with myocarditis due to KD, and a patient with mild abnormality of myocardial perfusion image. We compared age, image quality, radiation dose length product and effective radiation dose (mSv). These patients were divided in 2 groups with a voltage of 120 kV (group-H) and 80 or 100 kV (group-L). Image quality was ranked into these 3 categories; 3 points: all coronary arteries were well visualized, 2 points: images were adequate even if the distal arteries were not visualized, and 1 point: the image were inadequate.
Results: Inadequate image is completed in an only patient. The mean age of these groups were 11.0 +/- 2.8 years old in group-H and 7.0 +/- 3.8 years old in group-L ($p < 0.0001$). Average image quality was 2.5 +/- 0.6 points, 2.6 +/- 0.5 points in group-H and 2.4 +/- 0.6 points in group-L ($p = 0.07$). Average radiation dose length product was 206.7 +/- 182.5 mGycm, 338.0 +/- 178.9 mGycm in group-H and 89.3 +/- 73.9 mGycm in group-L ($p < 0.0001$). Average effective radiation dose was 3.40 +/- 3.14 mSv, 5.75 +/- 3.04 mSv in group-H and 1.30 +/- 1.04 mSv in group-L ($p < 0.0001$).
Conclusion: Low tube voltage including 80 or 100 kV could achieve low radiation exposure with average effective radiation dose of 1.3 mSv for children with mean age of 7.0 years old without lowering the image quality of area detector CT after KD.

P-231

Extendable Multimodal Cardiac Analysis Framework

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Background and Objectives: Diagnostic MR imaging in congenital heart diseases is utilizing increasingly multi-modalities in multi dimensions for assessment of anatomy, blood flow and tissue characterization. We present a novel cardiovascular imaging postprecessing framework (CAIPI) that integrates data into one comprehensive framework and provides open-access through an universially available web-front.
Methods and results: For CAIPI we created a novel work-flow concept that uses strict 4D alignment and synchronization of image data. This does not only offer a synchronized visualisation of spatio-temporal relations of different datasets but also the combined analysis of different image types.
CAIPI provides a flexible biventricular volumetry tool that is fully independent of orientation and supports arbitrarily oriented views for interaction. Furthermore a flexible analysis tool for the myocardium is integrated, to segment and compare regions of interest in LE, T1, T2 and T2* maps. By the integrated workflow the results of multiple modalities can be combined such as perfusion MRI, late-gadolinium enhanced MRI, angiographies and 4D blood flow.
To provide easy access to the clinical research community CAIPI integrates the well known open-access web frontend Osirix. Complex image processing and visualisation are based on MeVisLab. Through plugin mechanism the software environment is easily extendible by image processing applications.
Conclusion: We expect that the presented software platform will facilitate multi-modality multi-dimensional image processing in congenital heart dieses. The open-access architecture will make high quality software environment available to the user community. The plugin structure allows rapid integration of software prototypes from other research groups.

P-232

Cardiac functions, intima-media thickness and epicardial adipose tissue in obese children with nonalcoholic fatty liver disease: Echocardiographic and tissue Doppler imaging study

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Aim: Nonalcoholic fatty liver disease (NAFLD) encompasses a range of liver histology severity and outcomes in the absence of chronic alcohol use. The recent rise in the prevalence rates of overweight and obesity likely explains the NAFLD epidemic worldwide. The aim of recent study was to evaluate the effect of obesity on systolic and diastolic cardiac functions, intima-media thickness (IMT) and epicardial adipose tissue in obese children with NAFLD.
Materials and Methods: Ninety three of 400 obese patients with NAFLD were compared with age and sex matched 150 controls. Serum lipids', cholesterol, glucose, insulin, and liver enzymes' levels were measured during the fasting state. A complete echocardiographic study including M-mode, Doppler and tissue Doppler imaging (TDI) was performed. Also, IMT was measured from common carotid artery and epicardial adipose tissue thickness was measured during end-diastole from the parasternal long axis views. The patients with NAFLD were classified into three subgroups according to ultrasonographic visualizing (Stage 0, 1and 2).
Results: The patients with NAFLD had a significantly higher body mass index (29,53 ± 3,81 vs. 18,53 ± 2,70 kg/m2, $P < 0.001$), total adipose mass (40,33 ± 6,40 vs. 18,08 ± 5,01% $P < 0.001$) higher insulin (15,19 ± 15,96 vs. 6,53 ± 2,83 mcIU/ml, $P < 0.001$), and total cholesterol levels (163,20 ± 36,71 vs. 150,71 ± 24,02 mg/dL, $P = 0.002$). Increased thickness of the intraventricular septum, posterior wall (0,749 ± 0,168 vs. 0,630 ± 0,129, $P < 0.001$; 0,768 ± 0,153 vs. 0,671 ± 0,152 mm, $P < 0.001$), and larger LV mass and LV mass index (129,387 ± 43,695 vs. 93,861 ± 32,016 g, $P < 0.001$ and 41,572 ± 9,286 vs. 30,856 ± 8,216 g/m2.7, $P < 0.001$, respectively) were found in

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NAFLD group. NAFLD group had higher Tei index values on both ventricles (LV 0,397 ± 0,062 vs. 0,372 ± 0,060 P < 0.001, RV 0,416 ± 0,061 vs. 0,385 ± 0,052 P < 0.001 and septum 0,384 ± 0,052 vs. 0,368 ± 0,042 P < 0.001, respectively). Also, IMT of common carotid artery and epicardial adipose tissue thickness were significantly higher in NAFLD group. Only total cholesterol level, total adipose mass and mitral lateral annulus systolic motion velocity were statistically different in two stages of NAFLD.
Conclusion: In conclusion patients with NAFLD had mildly altered LV and RV functions and increased IMT and epicardial adipose tissue thickness. Also, liver steotosis was positively associated with total adipose mass percentage and interventricular septum systolic thickness.

P-233
Obstructive left heart disease in neonates with a "borderline" left ventricle: diagnostic challenges for choosing the best outcome
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Background: In most newborns with left heart obstruction, the choice between a single ventricle or biventricular management pathway is clear. However, in some neonates with a "borderline" left ventricle, this decision is difficult. Existing criteria do not reliably identify neonates who will have a good long term outlook after biventricular repair (BVR).
Objectives: Prospective assessment of the outcome after BVR of newborns in whom the left ventricle (LV) was considered "borderline" by an expert group.
Methods: Prospective follow up of neonates with obstructive left heart disease associated with a "borderline" LV who underwent biventricular management between January 2005 and April 2011.
Results: Thirteen of 154 (7.8%) neonates who required intervention for left heart obstruction met echocardiographic (ECHO) inclusion criteria. At first and last ECHO, mitral valve Z score was -1.76 (±1.37) and -0.66 (±1.47) (p = 0.013) respectively, aortic valve -1.02 (±1.57) and -0.23 (±1.78) (p = 0.056), and LV end-diastolic volume 13.77 (±5.8) and 20.85 (±8.9) ml/m^2 (p = 0.006). All 12 survivors are clinically well. However, LV diastolic dysfunction and pulmonary artery hypertension was present in 5/12 (36%). Endocardial fibroelastosis (EFE) was detected in 5 patients at last echo follow up, but only in 2 preoperatively. Cardiac MRI did not confirm EFE in any of assessed patients.
Conclusions: We were unable to reliably predict outcome after BVR of neonates with left heart obstruction and a "borderline" LV. The presence of EFE with consequent diastolic dysfunction is more important than LV volume in determining the outcome. Prospective identification of EFE remains challenging.

P-234
Certain ECHO and advanced ECG indexes are associated with family history for major adverse events in children and adolescents with HCM, and can be distinguished from athlete's heart
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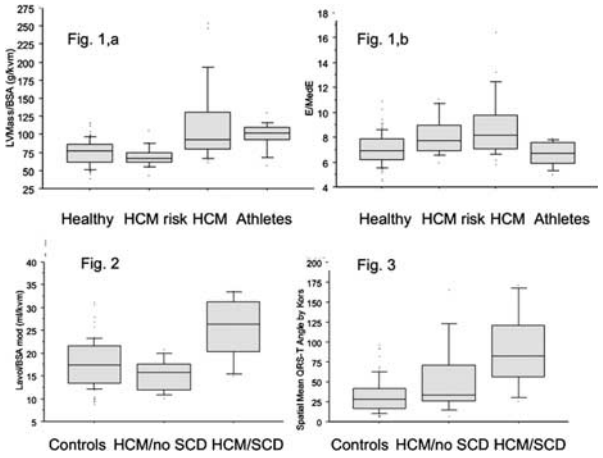
Introduction: Hypertrophic cardiomyopathy (HCM) remains a common cause of significant morbidity and mortality in the young. Sudden cardiac death (SCD) may sometimes occur in spite of lack of significant hypertrophy, making thus the early diagnosis HCM of utmost importance. Furthermore to distinguish HCM from "athlete's heart" is still an important clinical problem.
Methods: The study population, part of a cohort from an ongoing prospective study, consisted of children and adolescents (age range 0,1-30 years, median 14,7 years) with HCM heredity, either without (HCM-risk; n = 17) or with LV hypertrophy (Z-score for IVS and/or PW >2,5; n = 21), healthy controls (n = 81) and athletes (n = 13, endurance physical exercise >10 hours/week, Z score for IVS and/or LPW >2). Left atrial (LA) and ventricular (LV) size, mass (both adjusted for BSA), and LV thickness and function were assessed by echocardiography with Tissue Doppler (TD). Spatial mean QRS-T angle by Kors was measured via advanced 12-lead ECG analysis using Cardiax® (IMED Co Ltd, Budapest, Hungary and Houston, USA).
Results: Compared to controls, septal E/Ea ratio by TD and LA volume were significantly increased in both the HCM (p < 0,001) and HCM-risk (p = 0,01) groups, while QRS-T angle was significant increased in HCM group (p < 0,001) versus controls and HCM-risk. Athletes had increased LV mass (p < 0,01) versus controls and HCM-risk, comparable to HCM group (p = 0,2), but similar septal E/Ea ratio and QRS-T angle as controls (Figure 1, p > 0.5). Family history of major adverse events (SCD, cardiac arrest, ICD, heart transplantation) was associated with further increase of LA volume among HCM-risk individuals (Figure 2, p < 0,001), and with further increase of QRS-T angle among HCM patients (p < 0.01, Figure 3).
Conclusion: In addition to TD, advanced ECG provides a valuable diagnostic tool to distinguish HCM from athlete's heart. In HCM patients, family history for major adverse cardiac events appears to be associated with more profound electrical abnormalities, as suggested by its association with increased QRS-T angle. This finding might explain in part the earlier suggested further increase in SCD risk in HCM patients with family history for major cardiac events.



P-235
Effects of Remote Preconditioning on Skeletal Muscle Metabolism During Exercise, a Pilot Study
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Introduction: Remote ischemic preconditioning (RIPC) induced by transient limb ischemia releases a dialyzable, circulating, protective factor that reduces ischemia-reperfusion (IR) injury.

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We have recently shown that RIPC improves competitive, but not submaximal, performance in elite athletes, presumably by modifying skeletal muscle resistance to 'relative ischemia' at maximal effort.
In this study we examine potential mechanisms for the effect of RIPC using magnetic resonance spectroscopy (MRS) to study indices of muscle metabolism and bioenergetics, including measurements of mitochondrial oxidative phosphorylation and anaerobic glycolytic metabolism.
Methods: 10 healthy subjects, 20 to 27 years of age, were randomised to RIPC (4 cycles of 5 minutes arm ischemia) or sham preconditioning with cross-over. On each occasion, supine exercise was performed on a calibrated non-magnetic up-down ergometer (Lode AEI Technologies). Each subject performed a 90 second (s) submaximal exercise test at 85% of a previously tested maximal work rate followed by ten 30s bouts at 70% of the maximal work rate each separated by 15s of rest. Magnetic resonance imaging (MRI) and 31P-MRS data were obtained.
Results and Discussion: We observed trends towards an increased in ATP production from the anaerobic glycolytic pathway (0.69 ± 0.35 (sham) vs. 1.10 ± 1.27, $p = 0.21$), and towards a larger change in cellular pH (-0.28 ± 0.17 (sham) vs. -0.59 ± 0.93, $p = 0.16$) during exercise, after RIPC. However none of the 24 resting or exercise 31P-MRS parameters reached statistical significance. The lack of effect of RIPC on submaximal performance reflects our previous data on submaximal swimming performance.

P-236

An analysis of left ventricular strain in premature infants and full-term infants during the early postnatal period using Velocity Vector Imaging-An investigation into the development of cardiac function in newborns-

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Background: The cardiac function in newborns and, in particular, premature infants, is considered to be immature. However, few reports exist in which a detailed investigation was carried out into the development of cardiac function based on myocardial deformation data.
Purpose: To evaluate myocardial strain of the left ventricle and the rate of change in the cross-section of the left ventricle regarding premature infants and full-term infants using velocity vector imaging (VVI) and investigate the mode of development of the myocardial function in the perinatal period.
Subjects and methods: The subjects comprised 30 premature infants and full-term infants with no congenital heart disease or lung disease (gestational age 29 weeks and 3 days to 41 weeks and 1 day, 1 to 60 days old). The circumferential (CS) and longitudinal (LS) strain in the left ventricle was obtained using the left ventricular short-axis view (base and apex) and four-chamber view of the apex using VVI. Moreover, the left ventricular short-axis area of the base and the apex was measured by tracing the lining membrane, and the rate of change from the telediastolic phase to the peak value was respectively investigated. A correlation between the abovementioned data and the number of weeks of fixed gestation was identified.
Results: The CS of the apex and the systolic phase CSR showed a positive correlation with the number of weeks of fixed gestation ($r = 0.60$, $P < 0.001$ and $r = 0.54$, $P < 0.001$, respectively) and the diastolic CSR showed a negative correlation ($r = -0.48$, $P = 0.003$). Regarding the rate of changes in the cross-section, only the apex showed a positive correlation with the number of weeks of fixed gestation ($r = 0.58$, $P < 0.001$), with the base not exhibiting any correlation.
Conclusion: It is believed that such development of a deformation pattern of the apex may be a cause for the development of cardiac function in premature infants and full-term infants during the perinatal period and is thus considered to be important new knowledge regarding the cardiac function of newborns.

P-237

Impact of aortic elastance on right ventricular function in Hypoplastic Left Heart Syndrome after Fontan palliation

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Introduction: Systemic right ventricular (RV) function is the most important determinant of long term outcome in patients with palliated hypoplastic left heart syndrome (HLHS). Abnormal elastic properties of the reconstructed aorta, may negatively impact on ventricular function. We therefore aimed to assess systemic RV function and aortic arterial elastance in palliated HLHS patients to describe the relationship between the two.
Methods: Fifty-six HLHS patients (median age 5.4 (range 2.9-14.2) years) were studied at a median of 2.6 (range 0.8-12.7) years after completion of the Fontan circulation with the pressure-volume conductance system.
Results: Arterial elastance of the reconstructed aorta (Ea) was higher in palliated HLHS patients compared to normal values from the literature [Chen et al., JACC 1998] for the unoperated aorta in a biventricular circulation (3.1 ± 1.1 vs. 2.2 ± 0.8 mmHg/ml; $p < 0.01$). Ea correlated negatively with RV ejection fraction as an afterload dependent measure of systolic ventricular function ($r = -0.31$, $p = 0.02$). However, load independent systolic RV function measured as end systolic elastance (Ees) increased with increasing Ea ($r = 0.29$, $p = 0.02$). Therefore, ventriculo-arterial coupling (Ea/Ees) remained within the physiological range. End diastolic stiffness (Eed) of the systemic RV showed a positive correlation with Ea ($r = 0.6$, $p < 0.001$).
Patients who needed an intervention for significant coarctation after aortic reconstruction showed higher Ea and Eed even four years after successful coarctation treatment (Ea: 3.4 ± 1.2 vs. 2.8 ± 1.0 mmHg/ml, $p = 0.04$ and Eed: 0.67 ± 0.44 vs. 0.45 ± 0.3 mmHg/ml, $p = 0.04$).
Conclusion: The elastance of the reconstructed aorta of palliated HLHS patients is abnormally high. Aortic elastance, a determinant of afterload, negatively impacts on ejection fraction but not on load independent systolic myocardial function. Diastolic stiffness is increased when aortic elastance is high.
The effects of abnormal aortic elastance on intrinsic systolic function and diastolic stiffness were more pronounced in patients who underwent successful treatment for aortic coarctation.

P-238

Patients after Arterial Switch Operation Have Impaired Global and Regional Systolic Right Ventricular Function: A Speckle Tracking Echocardiography Study

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Introduction: Current standard treatment of D-Transposition of the great arteries is the neonatal arterial switch operation (ASO). Functional assessment of the right ventricle (RV) after this procedure remains problematic due to its complex ventricular geometry. Myocardial deformation imaging by 2D strain echocardiography is a novel method representing a diagnostic possibility for assessing the RV function. This study aims to investigate the global and regional systolic function of the RV.
Methods: We compared echocardiographic measurements – from standard and tissue Doppler imaging (TDI), between 58 patients (mean age 5.7 ± 4 years) after neonatal ASO and 13 healthy controls (mean age 7.6 ± 4 years). Additionally apical four-chamber images (frame rate 74 ± 6 frames/s) were analyzed offline and the global and regional peak systolic strain (PSS) of the RV was derived. These data were compared between both groups with the Student's t-test. A p-value of less than 0.05 was considered significant.
Results: Age and body surface area were comparable between patients after ASO and those from the control group. Compared to the control group, the ASO group had greater RV wall thickness (3.67 ± 0.67 mm vs. 2.9 ± 0.29 mm, $p < 0.0001$) and greater RV diameter (15.1 ± 4.3 mm vs. 12.9 ± 2.17 mm, $p = 0.01$). The peak systolic TDI velocity of the anterior RV wall was lower in the ASO group than in the control group (7.0 ± 1.1 cm/s vs. 11.54 ± 1.4 cm/s; $p < 0.001$). The ASO patients had lower TAPSE (13.7 ± 2.5 mm vs. 20.5 ± 4.5 mm; $p < 0.001$). Right ventricular global PSS was diminished in the ASO patients compared to the control group (-10.03 ± 4.5 vs. -15.5 ± 2.3; $p < 0.01$). Regarding the regional RV function: PSS was lower in the ASO group than in the control group in the middle (-16.38 ± 5.47% vs. -22.07 ± 5.22%; $p < 0.001$) and in the apical (-11.83 ± 5.31% vs. -24.24 ± 6.15%; $p < 0.001$) RV free wall segments. There were no significant differences in the basal PSS between both groups (-20.16 ± 7.34% vs. -21.13 ± 7.03%, $p = 0.68$).
Conclusion: Patients after ASO have diminished global systolic RV function. There is regional systolic dysfunction in the middle and apical segments of the RV which can be demonstrated by speckle tracking echocardiography.

P-239

Atria volume ratio as a simple echocardiographic parameter that might be useful in corrected Tetralogy of Fallot evaluating the timing for re-intervention in this population

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Introduction: ToF is one of the most frequent complex congenital heart defect that we encounter in our every-day-life ambulatory activity. MRI is the unquestioned diagnostic tool to evaluate the right timing for percutaneus valve replacement but it costs, and is not always easy available.
Aim: to find out simple parameters that might be useful to better predict a significative RV dilatation and dysfunction in this population.
Methods: Each patient undergone physical examination, timing of radical correction and history of previous BT shunt palliation, ECG, Echocardiogram and Cardiopulmonary test. About ECG findings it was possible to compare prospectively a "basal" one (the first after surgery), ECG1, and the most recent one, (ECG2). QRS duration (QRSd) was one of the main indipendent variable. In this analysis patients in natural history and with pulmonary atresia+interventricular septum defect were excluded.
Results: Total population was 76 patients. 42 females and 34 males with a mean age of 19 ± 14 years. 59 patients had classic type of ToF, a half received a radical correction before 6 months of age. ECG mean follow-up was 8,2 ± 4 y in 45 patients
Only 7 patients received a BT shunt palliation before: two in the group of early correction and 5 in the group of late correction. The statistically interesting findings of this work were: QRS widening strongly correlates with time of correction, age, pulmonary insufficiency, peak VO2 and indexed RV telediastolic volume (in our population, an indexed TDV of 70 ml/mq, at echo calculation, correlates significantly with a > 140 msec QRS widening). A strong correlation between the atrial volume ratio (RA/LA)>1.7 a more reproducible parameter than ventricle volumes, and QRSd was found ($p = 0.007$).
Conclusion: ToF patients have a good life expectancy, however, they experiments soon or later arrhythmic problems and right heart failure appear. The criteria for re-intervention based on MRI are the gold standard but is not always easily available. Together with what already stated such as QRS widening >140 msec, RA/LA volume ratio and RV TDV >70 ml/mq echo measured might be an added parameter for follow-up and timing for valve replacement.

P-240

Norwood I type procedure for Hypoplastic Left Heart Syndrome at time of Fontan completion: does it make any difference?

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Background: Stage one reconstruction (Norwood operation) for Hypoplastic Left Heart Syndrome (HLHS) can be performed with either a modified Blalock-Taussig shunt (mBTS) or a right ventricle-pulmonary artery (RV-AP) conduit. Hemodynamic and physiological differences between the 2 surgical palliations have been described before stage 2 surgical reconstruction. However, few data are available about differences at time of Fontan.
Aim: To evaluate hemodynamic and physiological differences between the 2 surgical palliations for HLHS at time of Fontan procedure.
Methods: The clinical records of consecutive 17 patients with HLHS who completed Fontan between 2007 and 2011 were retrospectively reviewed. Following first and second stage palliation, serial clinical evaluations and echocardiograms were performed in all patients, followed by cardiac catheterization before Fontan. Pre-catheterization data included the degree of right ventricular dysfunction and atrioventricular valve regurgitation (AVVR). Significant ventricular dysfunction was defined "a priori" by pre-catheterization echocardiographic examination or evidence of hemodynamic compromise (ventricular end-diastolic pressure >14 mmHg).
Results: Of 17 patients undergoing Fontan at a mean age of 50.5 months (range 38.2-57.5 months), 16 had an extracardiac conduit and 1 had a fenestrated lateral tunnel. At stage 1 palliation, 6 patients had a mBTS (group 1) and 11 had a RV-AP conduit (group 2). Follow-up echocardiograms and cardiac

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catheterization data were available in all patients. Significant right ventricular dysfunction was present only in 1 patient, in group 2. Moderate AVVR was present in 2 patients (11,7%), in group 2. The hemodynamic results did not showed statistically significant differences. For mBTS versus RV-AP shunt there was a pulmonary artery pressure (PAP) of 10,8 ± 1,3 versus 11,8 ± 1,4 (p = 0.21), a Qp/Qs ratio of 0,66 ± 0,09 versus 0,65 ± 0,06 (p = 0.83), a right ventricular end-diastolic pressure (RVEDp) of 8 ± 1,58 versus 8,9 ± 1,64 (p = 0.31), a coronary perfusion pressure of 64,2 ± 9,73 versus 60,6 ± 8,39 (p = 0.47).
Conclusions: At time of Fontan, the degree of right ventricular dysfunction, AVVR and hemodynamic data were not statistically different between the two groups. Status of patients with HLHS at time Fontan completion is not influenced by Norwood I type of initial surgical palliation.

P-241
Accuracy of gadolinium-enhancemend 3-dimensional magnetic resonance angiography to detect venovenous collaterals in patients with total cavopulmonary connection (TCPC)

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Introduction: In patients with total cavopulmonary connection (TCPC), venovenous collaterals may cause cyanosis and haemodynamic impairment. The aim of the study was to evaluate the accuracy of gadolinium-enhancemend 3-dimensional magnetic resonance angiography (GE 3D MRA) to detect these abnormal vessels.
Methods: From January 2010 to December 2011, 13 patients (median age 16 years, range 12 to 28) with TCPC underwent both GE 3D MRA and cardiac catheterization within a median of 6 months. Combined findings were compared.
Results: GE 3D MRA detected venovenous collaterals in 6/13 (46%) of the patients. Mean oxygen saturation was 93% in these patients. All the 8 major collaterals diagnosed in these patients by catheterization were correctly diagnosed by MRA. Compared to catheterization, GE 3D MRA had 100% sensitivity and specificity for the diagnosis. All the collaterals were closed using coils, plugs and covered stents and mean oxygen saturation increased to 97%.
Conclusions: GE 3D MRA is fully accurate to detect venovenous collaterals after TCPC. This technique might reduce the irradiation exposure in the cath lab, might reduce the total amount of the contrast injected and might orient the procedure of percutaneous closure.

P-242
Echocardiographic assessment of diastolic dysfunction in children with congenital heart disease – correlations with invasive data

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Introduction (or Basis or Objectives): Diastolic dysfunction is an important cause of heart failure in adult population. Studies regarding diastolic dysfunction in children are scarce and with small numbers of patients.
Methods: Fifty patients scheduled for the left heart catheterization were prospectively enrolled in the study. After induction of general anesthesia a short functional echocardiography was performed including all standard diastolic parameters measured according to current guidelines. Once arterial access was gained a catheter was advanced into the left ventricle (LV) and high-fidelity pressure measurements were performed using a Millar catheter. Invasive diastolic parameters (Tau, min dP/dT, max –dP/dT and end diastolic pressure (EDP)) were measured on the pressure tracings.
Results: Study population consisted in 4 groups of patients: aortic coarctation (CoA, n = 16), aortic stenosis (AS, n = 8), Kawasaki disease (Kw, n = 15) and heart transplant patients (HTx, n = 11). Mean age at procedure was 10 ± 5 years with no differences between groups. Patients with outflow tract obstruction (OTO, including CoA and AS) had significantly higher EDP, Tau values and LV mass index than those without (HTx and Kw) (see table). Also, they had longer deceleration time and IVRT, higher pulmonary S/D ratio but similar E/A ratio; septal and lateral tissue velocities (e') were lower and E/e' ratio increased compared to patients without OTO (see table). Only 7 patients had prolonged Tau (5 CoA, 2 AS), of which 3 had completely normal echo data, while 4 had abnormal e' lateral and E/e' ratio, 2 pts also with increased E/Vp ratio, all other parameters being within normal range. Nineteen patients (14 with OTO) had EDP > 12 mmHg, associated with prolonged Tau in 5 pts. There were no significant correlations between echo parameters and LV EDP, while Tau correlated with IVRT (R = 0.4) and E/Vp (R = 0.35), p < 0.05. E/e' ratio >2SD for age was a good predictor of EDP higher that 12 mmHg (OR: 12.8, 95% CI: 2.4-78, sensitivity 58%, specificity 90%).
Conclusions: Elevated filling pressures are relatively common in patients with outflow tract obstruction but are not necessarily associated with early relaxation abnormalities. Echocardiographic parameters correlate poorly with invasive measurements of diastolic function. Increased E/e' ratio is a predictor of elevated filling pressures with relatively low sensitivity.

	Outflow tract obstruction* (n=24)	No outflow tract obstruction (n=26)	p
End diastolic pressure (mmHg)	16.3±4.1	9±4.8	p<0.001
Max –dP/dT (mmHg/sec)	1037.3±474.1	1002.4±345.7	p=0.38
Min dP/dt (mmHg/sec)	-1185.5±350.8	-1104.7±382.9	p=0.22
Tau (msec)	42.8±12.8	33±7.6	p<0.001
Left ventricular mass index (g/m$^{2.7}$)	43.6±18.5	30.1±7.5	p<0.001
Mitral E/A ratio	110.8±20.4	97.9±22.9	p=0.45
Mitral deceleration time (msec)	156.4±37.9	132.5±33.6	p=0.016
IVRT (msec)	65.67±13.1	59.3±12.7	p=0.047
Pulmonary S/D ratio	1.1±0.2	0.76±0.2	p<0.001
Mitral A – PV A reversal duration (msec)	20±22	13±15	p=0.1
e' lateral (cm/sec)	11.7±3	14.1±4	p=0.013
e' septal (cm/sec)	10.8±1.9	13±3.1	p=0.002
E/mean e' ratio	10.9±3.8	8±2	p<0.001
Mitral flow propagation velocity (cm/sec)	61±16.2	62.5±14.5	p=0.37
Mitral E/ flow propagation velocity ratio	2.1±0.7	1.7±0.4	p=0.023
Left atrial volume index (ml/m^2)	29.7±8	27.3±5	p=0.1

*3 patients had blended mitral inflow

P-243
Evaluation of Cardiovascular Changes in Children with Aortic Coarctation Treated with Endovascular Stents

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Introduction (or Basis or Objectives): Early and late effects of treatment of aortic coarctation with endovascular stents are not well known. In this study we aimed to investigate early effects of endovascular stent application on coarctation by evaluating blood pressure, vascular wall elasticity, carotid intima media thickness, serum NT-proBNP levels, left ventricular functions in children.
Methods: Fifteen patients who were diagnosed between February 2009-May 2010 and 30 healthy controls were included in

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this study. Left ventricular structure and functions were evaluated echocardiographically before, first and sixth months after stent placement. Elastic functions of aorta were assessed by systolic and diastolic diameters of aorta. In order to evaluate end organ damage carotid intima-media thickness was examined ultrasonographically. Left ventricular dysfunction was determined by serum biochemical plasma NT-proBNP levels. Blood pressures were recorded before and after stent application

Results: Mean age of patient group was 10.5 ± 3.7 and control group was 9.6 ± 3.4. Systolic blood pressure before stent application was 134.4 ± 16.3 mmHg, one month after stent was 114.5 ± 12.1 mmHg and six months after was 115.5 ± 9.5 mmHg. Systolic blood pressures were significantly elevated before and after treatment. Mean pressure difference at coarctation side was 29.5 ± 9.9 mmHg before the procedure and 5.7 ± 5.8 mmHg after the procedure. There was a significant decrease in left ventricle indexes at first and six months after procedure. Elevated carotid intima-media thickness and aortic wall stiffness didn't decrease at first and sixth months after the procedure. Decreased aortic elasticity before the procedure didn't alter after the procedure. A significant negative correlation was detected between pressure difference at coarctation and aortic elasticity. High NT-proBNP levels before the procedure decreased significantly at sixth month.

Conclusions: In children with aortic coarctation, increase in left ventricular mass index, carotid intima media thickness, aortic wall stiffness and decrease in aortic elasticity were interpreted as signs of the presence of cardiac and vascular involvement. Six months after the repair of coarctation and control of hypertension, incomplete recovery of cardiovascular function suggests that patients should also be followed after the procedure.

P-244
Cardiac MRI stress testing in children with myocardial ischaemia

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Introduction: Cardiac MRI is a non-invasive, high-resolution imaging modality widely used in patients with congenital heart disease. The role of imaging of inducible myocardial ischaemia and scarring, clinically established in adults, is rarely used in children with acquired or congenital heart disease.

Methods: We performed 64 sans on 57 children, mean age 12 ± 3 years (range 6-18) with Kawasaki disease ($n = 10$), switch procedure for TGA ($n = 21$), PAIVS ($n = 10$), ALCAPA repair ($n = 4$), anomalous course of coronary artery ($n = 4$) and other pathologies ($n = 8$). The patient's symptoms, heart rate and BP were monitored throughout the procedure. Sedation was not required and patients were familiarised with the scanner/procedure beforehand.

A complete stack of short-axis cine images was acquired to assess LV/RV function. Adenosine was infused intravenously at 140 mcg/kg/min over 3 minutes. Perfusion images were acquired during injection of gadolinium. Breath-hold/non-breath-hold coronary and late enhancement images were acquired to detect the presence of coronary aneurysms and myocardial scar/fibrosis respectively.

Results: The completed scan was achieved in 57/64 (89%) lasting ~ 1 hour. No adverse reactions to adenosine or gadolinium were recorded. During adenosine the heart rate increased from 75 ± 15 to 110 ± 10bpm with no significant change of BP. Inducible perfusion defects were identified in 10/64 (16%), myocardial infarction in 7/64 (11%), all corresponding to a coronary territory. Non ischaemic myocardial fibrosis was identified in 3 patients. Coronary MRA identified coronary dilatation/aneurysm in 6/10 (60%), all with Kawasaki disease.

Conclusions: Perfusion and scar imaging in children is feasible and uncomplicated. It has the potential to identify myocardial abnormalities non-invasively as seen in the adult population, and it is potentially valuable for guiding clinical management.

P-245
The role of preoperative catheterization in TCPC candidates

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Introduction: Assessment of patients before operation with Total CavoPulmonary Connection (TCPC) traditionally includes a cardiac catheterization with angiography to evaluate the pulmonary arteries and measure pulmonary artery pressure (PAP). Magnetic resonance imaging and computed tomography are evolving as reliable techniques for non-invasive evaluation and the necessity of routine catheterization is questioned.

Objectives: To evaluate the necessity of a routine cardiac catheterization before TCPC.

Methods: We reviewed 50 consecutive patients undergoing TCPC. Risk factors known prior to preoperative assessment ($\geqslant$ moderate AV insufficiency, $\geqslant$ moderate ventricular dysfunction, pulmonary artery branch stenosis, postoperative complications after Glenn operation) were reviewed. Hemodynamic data from bidirectional Glenn operation, catheterization and TCPC were collected, as well as assessment methods and outcome. Adverse outcome was defined as death within 30 days, transplantation, take down, ICU time $>$10days, pleural drainage $>$15 days or catheterization within 30 days.

Results: 30-days-survival was 100%. A total of 16 patients (31%) had adverse outcome, mainly prolonged time with pleural effusions. In total, 44 catheterizations, 5 MRI and 4 CT examinations were performed.

Comparison of hemodynamic data between patients with adverse and normal outcome revealed that patients with adverse outcome had significantly higher PAP at Glenn operation, but not at catheterization. Pre- and postoperative PAP at TCPC was also significantly higher in the group with adverse outcome. The transpulmonary gradient was significantly higher postoperative at TCPC in the group with adverse outcome, but not preoperative or at Glenn operation.

Conclusions: Invasive hemodynamic data from a catheterization did not add determinant information in any patient. As pressure measurements done at the time of the Glenn operation correlated well with intraoperative measurements before TCPC, the preoperative catheterization is not indicated for measurement of PAP. Hemodynamic data from previous Glenn operation may be used as a reliable measurement of PAP and as a predictor of adverse outcome after TCPC.

A catheterization should be performed as part of preoperative assessment only if an intervention is needed or there is a suspicion of deteriorated hemodynamics after the Glenn operation, in other cases noninvasive methods are sufficient.

P-246
Reproducibility of a steady-state submaximal CMR-guided exercise test

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Background: We have designed an exercise test to assess cardiac function and hemodynamic changes by cardiovascular MR (CMR) during steady-state submaximal physical stress at 25% of maximum oxygen consumption. The aim of this study was to prove the reproducibility of this exercise test.
Method: We twice examined ten healthy volunteers (24 ± 3 years old) with the CMR steady-state submaximal stress test. To do this, we fixed a pulley to a special frame, which we mounted on the MR-table. The volunteers' legs were connected by a rope passing over the pulley. Steady-state exercise was defined as 144 up and down strokes of the extended legs per minute, directed by an electronic metronome. We measured Heart Rate (HR), Blood Pressure (BP), enddiastolic volume of the left ventricle (LV-EDV) and Cardiac Index (CI) first under rest and then under stress and compared the stress results of both tests; we also compared the change from rest to stress in both examinations.
Results: HR increased by 36 ± 26% during the first, and by 31 ± 18% during the second examination (Δ5%). During the first stress test, stress HR was 13 ± 8% higher than it was at the second time. Systolic and diastolic BP increased by 12 ± 6% and 24 ± 17% at the first, and by 8 ± 5% and 16 ± 15% at the second test, respectively (Δ4% and Δ8%); during the first test, BP was 14 ± 8% and 21 ± 12% higher than at the second one, respectively. LV-EDV decreased by 5 ± 4% during the first, and by 13 ± 9% during the second examination (Δ8%). The first stress LV-EDV was 8 ± 7% higher than the second one. Stress CI first increased by 33 ± 19% and then by 41 ± 20% (Δ8%), there was no difference in stress CI between both tests.
Conclusion: HR and BP were higher at the first test compared to the second one. This may be explained by a learning process of the volunteers, but has to be studied further. However, the changes of LV-EDV and CI were very similar. The stress test may give new insights into hemodynamics of different underlying congenital heart defects.

P-247
Cardiac functional assessment in mucopolysaccharidosis

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Introduction: In mucopolysaccharidosis (MPS) cardiovascular involvement includes valvular dysfunction, coronary artery disease and conduction abnormalities, with significant morbi-mortality. The impact of enzyme replacement therapy (ERT) on cardiac lesions is yet not well defined and, additionally, the decision to undergo surgical procedures may be challenging. There are no reliable parameters to monitor the clinical and therapeutic approach in MPS, nor has the role of tissue Doppler (TD) evaluation in this clinical setting been described. Our aim was to characterize the morphological and functional echocardiographic abnormalities in patients with different types of MPS undertaking ERT.
Methods: Cardiovascular studies were performed in eight patients with MPS, including biochemical markers of ventricular dysfunction (BNP, serum sodium, C reactive protein), EKG and morphological and functional echocardiographic cardiac evaluation.
Results: Patients aged ranged between four and 23 years old (five males). Six had MPS type VI and two type II, all undergoing ERT. The assessed biochemical parameters were all normal and EKG evaluation showed first-degree atrioventricular block in two patients. Valvular thickening with dysfunction was present in all cases, affecting mainly left heart structures (mitral regurgitation: 8; mitral stenosis: 7; aortic regurgitation: 7). Although only one patient had systolic dysfunction by M-mode evaluation (median LVEF 34%; median LVSF 34.5%), the systolic velocities of the lateral and septal walls of the mitral annulus were reduced (four and five cases, respectively). Regarding LV diastolic function, both Doppler velocities (LV inflow) and TD evaluation (early diastolic velocity of the lateral and septal mitral valve annulus) were suggestive of diastolic dysfunction in six cases. Those with normal function corresponded to the youngest patients, with an earlier beginning of the ERT. With respect to the RV function, all cases had normal systolic function (evaluated by TAPSE and TD systolic velocities of the lateral tricuspid valve annulus). In three cases the RV inflow velocities profile was suggestive of RV diastolic dysfunction and TD abnormalities were found in six patients.
Conclusions: MPS patients show a significant rate of cardiac involvement and TD abnormalities seem to be an early finding. Taking this into account, a regular follow up of the affected patients is mandatory.

P-248
Diagnosis and quantification of patent foramen ovale shunt: the role of transcranial Doppler sonography

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Introduction: Patent foramen ovale (PFO) occurs in about 30% of general population. Its role in cryptogenic stroke is still debated, but from recent follow-up studies it seems that the amount of right-to-left shunt (RLS) is a major determinant of stroke recurrence. A contrast transesophageal echocardiography (TEE) is currently regarded as gold standard for RLS detection, but its invasive nature has fostered the search for new diagnostic techniques. In recent years, transcranial Doppler (TCD) has proved to be a valid alternative to TEE for its high sensitivity and specificity, relative ease of execution and little discomfort to the patients, but its use in the pediatric population is scarce.
Objective: To report our experience with TCD for the detection of RLS in pediatric patients.
Methods: Twenty patients (16.3 ± 4.2years, 57% female) were included in this study. All had ASD/PFO, but not RLS, documented by transthoracic echocardiography and were referred for percutaneous closure. TCD was performed according to a standard protocol: while blood flow in middle cerebral artery (MCA) was monitored through TCD, an agitated contrast agent (9 ml isotonic saline solution and 1 ml air) was injected into an antecubital vein, in two conditions: during normal breathing and after Valsalva maneuvre (VM). The number and pattern of microbubbles of air in the MCA was recorded. A four-level categorization, according to microbubbles count, was applied: (1) 0 microbubbles (negative result); (2) 1-10 microbubbles; (3) >10 microbubbles and no curtain, and (4) curtain. All patients with 5 or more microbubbles documented by TCD were considered to have tested positive for the presence of RLS. On the day of the percutaneous intervention, under sedation, TEE was performed to detect or rule out an intracardiac RLS.
Results: RLS was documented by TEE and TCD in 9 patients. Two patients had RLS documented by TCD, but not by TEE.

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No shunt was documented in 7 patients. In no case was a cardiac cause for RLS documented by TEE and not by TCD.
Conclusions: TCD is a valid alternative for RLS detection. Our experience, albeit short-numbered, has demonstrated its feasibility, safety and efficacy, obviating the need for sedation/anesthesia in pediatric patients.

P-249
Long-term outcome of small residual shunts in pts with multiperforated interatrial aneurysms following device implantation
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Introduction: Multiperforated interatrial aneurysms are associated with residual shunts following device closure. We sought to find the outcome of these small residual shunts over time.
Methods: We studied 29 pts who underwent device closure of atrial septal defects associated with multiperforated aneurysms. All pts had multiple left to right shunts by transesophageal echocardiography (TEE). An Amplatzer septal occluder device was placed under general anesthesia and TEE guidance. Transthoracic echocardiograms (TTE) were done at 1, 3, 6 and 12 months following the procedure to assess for device positioning, residual shunts or possible thrombi. Saline contrast echocardiography was performed with and without the Valsalva maneuver after one, two or more years to assess for right to left shunts.
Results: All pts had good quality TEE images, depicting 1-4 small residual atrial communications in the periphery of their interatrial aneurysm that were not included within the discs of the device. TTE images did not depict definite left to right shunts at 1, 3, 6, 12 months in all 29 pts, but in 7 pts shunting by TTE was equivocal.
Saline contrast echocardiography with the Valsalva maneuver revealed right to left shunting in 14 out of 29 pts, one year following device placement (48%). Two years following device placement the repeat saline contrast echocardiography with the Valsalva maneuver showed no residual right to left shunting in 10 out of the 14 pts with positive contrast echo at one-year follow-up (76%).
Conclusion: Spontaneous closure of small residual atrial communications may occur even after the usual one-year follow-up, after device closure. Endothelization of the device results in elimination of shunting across the atrial septum and may expand beyond the actual borders of the device, causing spontaneous closure of small residual communications in the adjacent aneurysmal tissues at various periods of time after the initial procedure. Placement of an additional device should probably be postponed for at least another year, until repeat saline contrast echocardiography is performed.

P-250
Right ventricular volume in Tetralogy of Fallot: a 2D echocardiographic method validation in comparison with cardiac magnetic resonance
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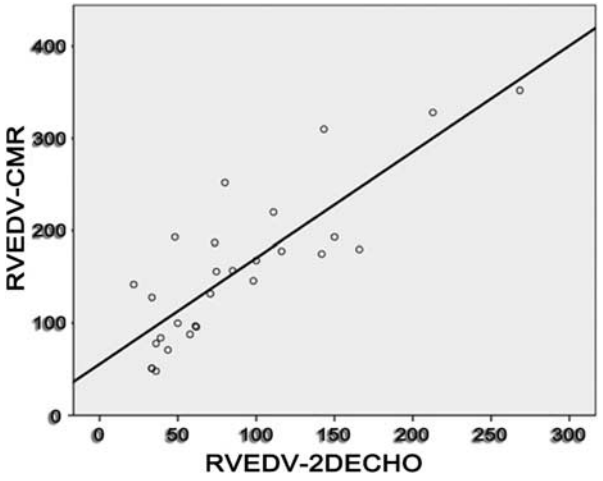
Introduction: Quantification of right ventricular end-diastolic volume (RVEDV) is an essential parameter in the long term follow up of repaired Tetralogy of Fallot (TOF) patients. Currently Cardiac Magnetic Resonance (CMR) is the gold standard, with limitations including anaesthetic procedures in children. Previous 2D-Echocardiography (2DE) quantification has given poor results. The aim of this study is to validate a new echocardiographic method, assuming that RVEDV is composed of two geometrical elements: an ellipsoid shape (from inlet to apex) and a truncated cone shape (outlet).
Methods: Cross-sectional study of TOF repaired patients. Analysis of the relationship between RVEDV obtained by CMR (RVEDV-CMR) and calculated by 2DE (RVEDV-2DE). Total RVEDV-2DE was obtained by the sum of the two volumes: in apical four chamber view using Simpson formula for an ellipsoid and parasternal short axis view of the right ventricular outflow tract using truncated cone formula. Statistical analysis was performed applying Pearson coefficient method and logistic regression.
Results: Twenty eight repaired TOF patients, mean age 11 years (ranging from 5 to 19) were included. Mean RVEDV-2DE versus RVEDV-CMR were 87.7 versus 156 ml (ranging from 22 versus 48 ml–268 versus 352 ml). RVEDV-2DE had significant correlation ($p < 0.0001$) with the RVEDV-CMR, Pearson Correlation Coefficient of 0.834. Linear regression model resulting in the following: RVEDV-CMR = 55,6+(1.15xRVEDV-2DE).



Conclusions: Quantitative assessment of RVEDV-2DE using this 2D-Echocardiographic method is feasible and correlates with RVEDV-CMR in a wide group of children and adolescents with repaired TOF. This information could be useful in patients with contraindications for Cardiac Magnetic Resonance.

P-251
Stents in Aortic Coarctation: Beyond Gradients and Pressures
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Percutaneous treatment of aortic coarctation is usually evaluated by residual pressure gradient and persistence of systemic hypertension. Evaluation of other parameters may provide valuable data to determine the effect of treatment on ventricular and arterial function. Longitudinal strain study can be used to assess improvement in left ventricular (LV) myocardial deformation and function. Valvular and arterial factors that oppose LV ejection can be assessed by valvulo arterial impedance (VAI).
Eight patients (5 with native aortic coarctation, 3 with recoarctation), submitted to aortic stenting from July to December 2011, were studied. Invasive pressures and gradients and dP/dt assessed by pressure wire in LV, ascending and

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descending aorta, were calculated before and immediately after stenting. Conventional echocardiographic evaluation, pulsed tissue Doppler, speckle tracking longitudinal strain and myocardial velocities and evaluation of VAI were performed before intervention, 24 hours after, and at medium term follow up (3 to 6 months). Statistical analysis was performed with SPSS 17.0.

Median (range) age was 25 years (6 to 57), 4 were female. Mean (SD) invasive pressure gradient varied significantly from 48,4 mmHg (28,3) before to 7,8 mmHg (11,8) after treatment ($p = 0,01$). Mean (SD) of global longitudinal strain varied significantly from -18,9% (2,79) before intervention to -21,9% (1,71) 24 hours after ($p = 0,02$), and this was maintained at follow up ($p = 0,02$ compared to pre cath). This trend was also seen in patients who remained hypertensive. Mean (SD) VAI varied significantly from 5,2 mmHg/ml.m2 (1,21) before intervention to 4,6 mmHg/ml.m2 (0,98) at follow-up ($p = 0,02$). Changes in longitudinal strain achieved with intervention did not correlate with initial or final gradient nor with VAI changes. E/E' did not change. Mean dP/dt increased from 1548 mmHg/s to 1831 mmHg/s in LV and from 305 mmHg/s to 581 mmHg/s in descending aorta. Median number of antihypertensive drugs in patients under medication reduced from 3 to 1.

Stenting provided excellent immediate hemodynamic results, with low residual gradients and statistically significant systemic pressure reduction. Longitudinal strain and VAI improved significantly after percutaneous treatment. This multiparameter evaluation may provide a better knowledge of cardiovascular physiology after stenting and a more accurate evaluation of the benefits of percutaneous treatment.

P-252
Assessment of Ventricular Septal Defects by Live Three-dimensional Echocardiography

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Objective: This study aimed to investigate the usefulness and feasibility of real-time three-dimensional transthoracic echocardiography (RT3D-TTE) in assessing ventricular septal defects (VSD) and compare the findings with those obtained by two-dimensional echocardiography (2D-TTE).

Methods: 37 patients (19 girls, 18 boys) aged 1 month-16.5 years (mean 3.27 ± 3.87, median 1. 50 years) with VSD diagnosed by 2D-TTE were prospectively examined by RT3D-TTE. The morphology, shape, number, size (maximum area, long and short axis diameters at end-diastole) of the defect, its relation and distance to the tricuspid and aortic valves, presence and degree of aortic overriding were analyzed by RT3D-TTE displaying the VSD in facing views from the right ventricular aspect; the results were compared to 2D-TTE findings in all and intraoperative findings in 20 patients. Standard transthoracic windows were used for 3D image acquisition; off-line measurements were used for quantitative analysis. VSD was isolated in 22 patients, existed as a component of congenital heart disease in 18, associated defects were present in 12 patients.

Results: Optimal RT3D-TTE images were obtained in 36 of 37 patients. Mean time required for 3D data acquisition was 4.09 ± 1.49 minutes, and for image processing 20.13 ± 10.46 minutes, declining significantly with the learning curve (r:-0.7, $p < 0,001$). The VSD was perimembranous in 20, perimembranous-outlet in 9, perimembranous-inlet in 5, muscular in 2 patients; the defect was single in all. There was complete agreement on defect morphology between 3D-TTE, 2D-TTE and intraoperative findings. Although mean end-diastolic VSD diameter measured by 3D-TTE from the right ventricular facing view was significantly larger than the corresponding 2D-TTE diameter (16.3 ± 8.7 vs 9.9 ± 4.35 mm, $p < 0.05$), there was a good correlation between them ($r = 0.79$, $p < 0.001$). 3D-TTE was superior to 2D-TTE in visualizing the shape of VSD, measuring tissue rims from the aortic and tricuspid valves, and in defects hidden by ventricular septal aneurysm or tricuspid septal leaflet which can be erased digitally to reveal the VSD underneath.

Conclusions: RT3D-TTE provides reliable qualitative and quantitative information regarding the morphology, shape, size, tissue rims of VSDs from unique facing views, and may serve as a feasible and valuable tool in the assessment of these defects prior to transcatheter or surgical closure.

P-253
Right ventricle systolic and diastolic function in juvenile-onset systemic lupus erythematosus patients

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Background: Cardiovascular events are a major cause of mortality and morbidity in patients with systemic lupus erythematosus (SLE). Several studies have described impaired systolic and diastolic function in adults with SLE, but very few reports have been published in children.

Aim: The aim of our study was to describe right (RV) ventricular systolic and diastolic function in a population of adolescents and children with SLE .

Methods: Cross-sectional and retrospective study of juvenile-SLE patients' medical records. All patients underwent two-dimensional, M-mode, conventional Doppler and tissue Doppler imaging (TDI) to evaluate systolic and diastolic right ventricular function.

Results: 13 Juvenile-SLE female patients were included, with disease onset between 6 and 16 years of age (mean 11,8 years), and mean disease duration of 6,6 years (2-15 years). One patient had moderate to severe aortic regurgitation (AoR), one had pulmonary hypertension (HTP), 5 patients had systemic hypertension (HTA); RV systolic function was normal in all patients except for one, with AoR (TDI S velocity 9,3 cm/s, tricuspid annular plane systolic excursion (TAPSE) 14,8 mm).

RV diastolic function markers were altered in some patients: decreased peak velocity of E wave in 4 patients, with increased A' wave velocity in 1 patient (HTP case). E/E'and E/A ratios were normal in all cases. RV outflow tract acceleration time was decreased (implying increased mean pulmonary artery pressure) in the patient with HTP.

Due to the small sample size, no correlations with other clinical or laboratorial markers were statistically significant. BNP was increased only in the AoR patient.

Conclusion: SLE patients can have subclinical cardiac dysfunction, and diastolic changes can be an early sign. Subtle abnormalities of the RV diastolic function were found even in the presence of a preserved systolic function. Preclinical detection of ventricular dysfunction may identify a pediatric population at risk requiring early and aggressive interventions for the prevention of cardiovascular events.

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P-254

Critical Neonatal Aortic Valve Stenosis, Immediate and Intermediate Outcome of Percutaneous Balloon Valvuloplasty

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Objective: To evaluate immediate and midterm results after balloon valvuloplasty in an infant population with critical aortic stenosis, giving special consideration to relief of aortic stenosis, degree of aortic regurgitation (AR), left ventricular function, and duration of freedom from reintervention.
Method: A retrospective follow up study was performed in 25 neonates who underwent aortic valve balloon dilatation from July 2006 to July 2010. We assessed the clinical and echocardiographic outcome for degree of restenosis, LV function, AR and need for reintervention.
Results: The median age at dilatation was 45 days (range from 6 days – 120 days). The median weight was 3.9 kg (1.9 kg-6.4 kg). The balloon valvuloplasty was performed with manual inflation of balloon through the femoral artery. The mean systolic pressure gradient across the aortic valve decreased from 70 ± 7.23 mmHg to 40 mmHg ± 4.3. Mild aortic regurgitation developed in 12 patients while in one patient non- coronary cusp was perforated, leading to severe AR, needing surgery. There was one death during the procedure.
At mean follow up of 25 months (3.0–48 months), there was no progression of AR. No patient had needed re-intervention for re-stenosis. LV function improved in 9 out of 11 patients (82%) while there was one death 2 months post procedure due to persistent LV dysfunction.
Conclusion: Balloon valvuloplasty in infants with critical AS is a safe and effective therapeutic procedure. The mortality is 8%. Mild aortic regurgitation does not progress in short and intermediate term. There is no restenosis in short term and LV function improves in majority of the patients.
Key Words: Aortic stenosis, Balloon valvuloplasty, Left Ventricle (LV) dysfunction, Interventions, Aortic regurgitation.

P-255

Influence of balloon size on aortic regurgitation in neonates undergoing Balloon Aortic Valvuloplasty–a retrospective study over an 11-year period

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Background: Transcatheter balloon aortic valvuloplasty has become the first-line treatment for critical or severe aortic stenosis in neonates in many centres. Aortic regurgitation following balloon aortic valvuloplasty remains a major concern. An optimal balloon size to aortic annulus ratio in order to minimise aortic regurgitation post-procedure, whilst relieving the obstruction, has not yet been identified.
Methods and Results: In this retrospective study, data regarding 29 neonates with critical or severe congenital aortic valve stenosis, who underwent balloon aortic valvuloplasty in the first 28 days of life, over an 11 year period, was evaluated. The balloon size used, ranged from 71 to 160% of the annulus size, with an average of 89%, based on the aortic annulus size as measured on angiography. The aortic regurgitation immediately following the procedure was trivial in 8 (27.6%), mild in 13 (44.8%), moderate in 7 (24.1%) and severe in 1 (3.4%) patient. The balloon to annulus ratio had no statistically significant effect on the degree of aortic regurgitation immediately after the procedure (p value of 0.259), at first follow up within 6 weeks of the procedure (p value of 0.961) or at follow up at 1 year (p value of 0.92).
Conclusion: This study did not show any significant relationship between the balloon to annulus ratio during interventional dilatation and the degree of aortic regurgitation following the procedure. A large multicentre prospective study, concentrating on the effect of the balloon size used, would be ideal.

P-256

Balloon Atrial Septostomy performed 'out-of-hours': Effects on the outcome

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Background: Balloon atrial septostomy is a common palliative procedure in neonates with cyanotic congenital heart disease with restricted interatrial blood flow. Despite its advantages, balloon atrial septostomy is not a risk-free procedure and can be associated with numerous complications. The objective of this study is to determine whether the performance of this procedure out-of-hours has a significant impact on the incidence of adverse outcome measures.
Methods and Results: 106 neonates who underwent balloon atrial septostomy between 2004 and 2010 were studied retrospectively. 64 infants had the procedure done within routine hours (9.00-18.00), while 42 neonates underwent the procedure out-of-hours (18.01-8.59). Procedure-related complications occurred in thirty-two infants (30.2%), which included (12/64 (18.8%) in routine hours and 20/42 (47.6%) in the out-of-hours group). During further follow-up after surgery and including both major and minor adverse events, seven more infants (10.9%) suffered complications after balloon atrial septostomy in the routine hours group while four more infants (9.5%) suffered complications in the out-of-hours group. This totalled the complication rate in the routine hours group to nineteen infants (29.7%) and twenty-four infants (57.1%) in the out-of-hours group ($P = 0.001$). A higher overall mortality rate was also noted in the out-of-hours group.
Conclusions: Balloon atrial septostomy performed out-of-hours produced higher complication rates as opposed to balloon atrial septostomy performed during routine hours. Only essential cases should be undertaken in the night, while all other cases should be deferred to the daytime to limit unnecessary adverse complication.

P-257

Interventional closure of muscular VSDs at a young age

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Purpose: The presence of muscular ventricular septal defects (mVSD) may pose an high circulatory burden in young patients. Surgical closure is difficult because of the multilayer structure of these malformations. We present the course of 13 patients (age 10 days – 7.3 years, median 9 months; bodyweight 2.2 and 18 kg (median 8.7 kg) undergoing transcatheter closure of such defects.
Method: All closures were assisted by transesophageal echo guidance. Entrance sites and routes of delivery were chosen according to the anatomic presentation of the defect. For transvascular closure of 5 singular and 8 multiple mVSD 5 Amplatzer PDA II occluder, 6 Amplatzer VSD occluder and

4 Amplatzer vascular plugs IV were used. Follow-up time ranges from 23 days to 2.8 years (mean 1.2 years).
Results: 3 defects were observed closed immediately. 9 mVSD showed a residual shunt immediately after the implantation procedure, which further reduced during follow-up in all patients. In one patient a second defect opened up after closure of one defect and was closed in a second attempt. Two intra-procedural complications occurred. A sudden complete AV-Block led to transcatheter explantation of the device in one patient. This patient with a restrictive cardiomyopathy died during follow-up. In one patient on ECMO a left ventricular perforation with the device already in place had to be over sewn in an emergency operation.
Conclusion: With some of the newer devices available, which pass through smaller delivery sheaths of 4 to 6 F, interventional closure of mVSD has become a feasible option in the treatment of patents of all ages and a bodyweight from 2.2 kg on, who present with elevated right ventricular pressures and high shunt volume. The procedure is challenging in newborns and infants and severe complications may occur.

P-258
Coronary Interventions in infants with congenital heart diseases
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Purpose: During the last decade the surgical and interventional techniques for the treatment of congenital heart diseases were constantly refined. Today children, who where considered to be inoperable some years ago undergo surgical or combined surgical-interventional treatment on a regular basis. This study focuses on coronary interventional procedures in children with CHD.
Population: During the last years we performed 16 coronary interventions in 14 patients. The Age ranged from 9 days to 26 years, mean 6 years, the bodyweight from 1.7 kg to 65 kg, mean 18 kg.
Results: During 10/16 procedures closure of coronary fistulas was attempted and successfully performed with a variety of implantable devices (coils, plugs). Coronary to right ventricular fistulae in pulmonary atresia/intact ventricular septum were occluded in 3/14 patients. In 3/14 patients acutely obstructed coronary arteries were successfully treated in 2/3 cases with balloon dilation and by stent implantation (1/3). All procedures were successfully performed without any mayor complications. One patient with a bodyweight of 1.7 kg and balloon dilation of the left main coronary artery died 2 weeks after the intervention because of haemostatic complications.
Conclusion: Balloon dilation or stent implantation is a realistic option in postsurgical coronary stenoses. It helps to improve myocardial perfusion and function without re-operation in severely depressed patients. Closure of coronary arterial fistulae with some of the newer low profile devices is possible even in newborns. Fistulae with a short channel to the right ventricular cavum remain critical for interventional closure if distal coronary perfusion has to be maintained.

P-259
Has biventricular repair become an option in patients with pulmonary atresia, intact ventricular septum and coronary to right ventricular fistulae?
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Purpose: Patients with pulmonary atresia and intact ventricular septum (PAIVS) are usually planed for uni-ventricular palliation as, in the presence of coronary to right ventricular (RV) fistulae (CAF), decompression of the right ventricle caused severe problems for the coronary perfusion and myocardial function. In patients with PAIVS and a well developed RV a bi-ventricular approach is possible if the CAF can be closed.
Methods: In a male newborn with PAIVS, reasonably sized RV, hypoplastic pulmonary arteries, multiple aorto-pulmonary collateral arteries (MAPCA) and multiple CAF a coronary stent was placed in the arterial duct to secure pulmonary perfusion. At the age of 3 months and with a bodyweight of 4 kg several CAF from the right and left coronary artery were occluded using retrograde and antegrade approaches. All, but one small, CAF could be closed by coils and vascular plugs. In a second step the duct was surgically ligated and the RV connected to the small pulmonary artery by patch insertion. Due to poor biventricular performance two big aorto pulmonary collaterals were closed in the cathlab the other day. The RV contractility improved. Patient was discharged 4 months ago and is awaiting bi-ventricular repair with homograft implantation in RV to PA position.
Results: Despite high pressures in the RV the global myocardial function improved during follow-up and the antegrade pulmonary perfusion secures oxygen saturations above 80% with only two aorto-pulmonary collaterals and one CAF left.
Conclusions: Highly selective placement of coils and vascular plugs can effectively reduce coronary arterial run-off in patients with PAIVS after opening of a RV to PA connection. The RV can regain its contractility and biventricular repair seems to become a realistic option in this patient. The further course of this patient and the evaluation of comparable other patients have to prove the reliability of this approach.
Selective and supra selective coronary angiographies are necessary for the decision whether closure of CAF would be possible, thus, making patients with diagnosis of PAIVS and CAF candidates for biventricular repair.

P-260
Immediate and Intermediate Results of Patent Ductus Artriosus Transcatheter Closure in Kurdistan (Multicenter experience)
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Background: A retrospective study to review our experience and determine the efficacy and safety of transcatheter closure of patent ductus Arteriosus in all three cardiac centers in Kurdistan, using the Amplatzer Duct Occluder type I and II (ADO) for the first time in Kurdistan.
*Patients & Methods:*A total of three hundred and nine patients which include (228 females & 81 males) were diagnosed to have PDA, between March 2008 to February 2011 in all three cardiac centers in Kurdistan namely (Hawler, Sulaimany and Duhok cardiac centers), where they underwent Transcatheter closure of the PDA with ADO type I and II. The common ages on closure was 1-5 years (range 5.5 months to 32 years), and weight ranged from 6 kg to 58 kg.We retrospectively analyses medical records, echocardiography, angiographic and hemodynamic data and follow up results of the patients.patients included in the study was according to standard recommendation for transcatheter closure of PDA, in which ages was above 6 kgs with no pulmonary hypertension, interrupted IVC and the anatomy feasible for closure.
Results: Among 309 cases did cath procedures, the ratio of female to male ratio was 2.6:1 and the age of doing PDA closure at time of procedure was between 5.5months to 33 years, the mean age is

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(6.3 +/- 6.5 years) and the weight range between 6 kg and 58 kg, mean of (11.5 +/- 7.5 kg). The most commonly used device is Amplatzer I PDA device with high closure success rate of 97.1% after 6 and 12 months follow up and the first cases of Amplatzer II PDA Device closure done in Iraq and Kurdistan. There was immediate & complete closure in 271 on angiography. Complication was noted as embolized PDA device to right pulmonary artery (RPA) in two patients, one of them retrieved by cath and other one by surgery.
Conclusion: Transcatheter Amplatzer duct occluder is safe and effective in closing PDA in most patients with PDA even large as much as 10 mm and new Amplatzer PDA II device allows small delivery sheath in which can be done for children with less than 6 kg with less complications.

P-261
Paediatric Ventricular Assist Devices: A Successful Nurse Training Programme and a Review of the Litterature
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Introduction: Ventricular Assist Devices (VADs) have been shown to play a successful role in 'Bridge to Recovery,' and 'Bridge to Transplantation' in paediatric cohorts. Where incidence of paediatric heart failure is rising and enlisted heart transplantation children face the highest rates of mortality amongst those awaiting solid organ donations, this technology offers great potential. Developing effective VAD programmes has never been so important. Nurses play an instrumental role in the VAD MDT as day-to-day providers in post-implantation care. The present study outlines a successful nurse training programme and curriculum.
Rationale: Where physician and perfusionist availability is often limited, nurses are able to provide VAD patients' 24-hour care. Their regular bedside manner evokes a role at the family interface, easily passing their training onto parents and thus, decreasing patient length of the stay. In this way, and in many others, an effective nurse training programme can help in the cost containment of a VAD programme, making it a viable treatment option for a hospital to provide.
Programme Description: All Stollery Children's Hospital nursing staff directly involved in VAD patient care undergo complete VAD training consisting of 2-phases providing a basis of general physiology, care, and monitoring of patients. Unlike many programmes, VAD training is provided to a wide cohort of staff to offer patients a continuum of care throughout their patient journey.
The VAD training course is a 12-hour training day led by a physician and nurse coordinator with VAD-specific expertise. Broken up into 8 modules, the course overviews indications of VAD therapy, device architecture, post-operative patient haemodynamics, anticoagulation, monitoring, emergency response and complications, change in patient condition, dressing protocols, and equipment management. Each aspect is evaluated using simulated clinical scenarios to judge critical thinking and appropriate response strategies. A 'hands-on' assessment is completed by each nurse and a written exam is issued. Each VAD-trained nurse attends a 6-hour recertification training day every 12 months.
Conclusion: An efficient training programme curriculum for nurses is important for the success of a hospital's VAD programme. The Stollery's successful training programme provides a basis from which other VAD programmes can devise their training technique.

P-262
Transcatheter closure a huge left atrial aneurysm with the right pulmonary artery to left atrium connection using with Amplatzer Ventricular Septal Occluder
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Right pulmonary artery to left atrium connection is a very rare cyanotic congenital heart defects. Although its main therapy should be surgery, herein, we describe the case of a 5-year-old girl who successfully closed with Amplatzer muscular ventricular septal defect occluder. The patient was asymptomatic at last follow-up.

P-263
An immediate imperfect result of balloon dilatation for aortic coarctation in infants and young children engenders an adequate late result.
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Objective: To study the early and late effects of balloon angioplasty (BA) of aortic coarctation and recoarctation in infants.
Background: BA, although considered inferior to stent implantation concerning the long term outcome is still the technique of choice to treat aortic coarctation in young children.
Methods: 50 consecutive infants with native aortic coarctation or recoarctation, underwent BA. Patients were divided into two groups according with an immediate residual pressure gradient <15 mmHg (group A) or >15 mmHg (group B) after BA.
Results: Peak to peak gradient diminished from $32.9 + 13.2$ to $17.0 + 10.5$ mmHg ($p < 0.0001$), but the majority of patient ($n = 31$, 62%) pertained to group B. In this group there were 1 procedural death and 6 early reinterventions. Thus 43 patients were immediately treated with BA only. At a median follow-up of 4.5 years (range 1-10) 2 patients of group A and 9 of group B needed a further BA or surgery ($p = 0.69$). Cox analysis showed that a residual gradient >15 mm Hg was the only predictor of reintervention. The event-free survival at 10 years in patients of group A and B was 87% and 55%, respectively ($p = 0.014$). At log rank test group B had a risk of undergo a second angioplasty or surgery 5-times higher compared with group A patients.
Despite these expected results, we observed that in 58% of group B patients treated with BA only no reintervention was necessary on account of the normalization of aortic flow and systemic blood pressure.
Conclusions: BA for aortic coarctation in infants offers suboptimal early results in 62% of patients. Although the immediate residual pressure gradient is the only predictor of reintervention in 47% of children with an immediate imperfect result the aortic obstruction disappears and no further interventions are necessary.

P-264
Closure of the patent ductus arteriosus with the new Duct Occluder II additional sizes device
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Objective: The objective of this study was to evaluate the technical feasibility, safety and efficacy of the new device Amplatzer duct

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occluder II additional sizes (ADO II AS) for closure of patent ductus arteriosus (PDA).
Background: Trancatheter device closure is the standard care for PDA. Currently available technology is not designed for closure of small PDA in young children.
Methods: From April to September 2011, 11 children (7 females, median age 2 years, median weight 11.4 Kg) underwent PDA closure with the ADO II AS. Ten had isolated PDA, 1 had PDA associated with preductal coarctation. There were 4 type A, 3 type C, 1 type E and 3 type D PDAs. We evaluated early and short-term results.
Results: All but two PDAs were closed via an antegrade approach. Mean fluoroscopy and procedural times were 8.0 + 3.9 and 49.8 + 27.9 minutes. Mean radiation dose was 5038 + 944.9 Gy/cm2. No complications occurred. Immediate trivial residual shunt was present in 1 patient. In all devices the retention discs laid flat against the walls of the pulmonary artery and aorta, without protrusion into the vessel lumen. The echocardiography performed after 24 hours did not show any residual shunt. At a median follow-up of 2 months the PDAs were completely occluded without obstruction of the pulmonary arteries or aorta.
Conclusions: The new device ADO II AS was safely deployed with complete resolution of the PDA shunt. The lower profile and symmetry of this device allows for venous or arterial approach and smaller delivery catheter size. The ADO II AS proved to be suitable to several anatomical types and might be a preferable alternative for closure of small-moderate PDAs.

P-265
Experience with percutaneous perforation of the pulmonary valve in neonates with pulmonary atresia and intact interventricular septum
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Pulmonary valvuloplasty may promote RV growth in neonates with pulmonary atresia and intact ventricular septum (PA). Percutaneous valve perforation and dilatation represents an alternative to surgery.
The aim of this study was to assess results and outcomes after percutaneous pulmonary valvuloplasty in neonates with PA.
Material and Methods: All patients with PA selected for percutaneous procedure (perforation and dilatation) were included in the study. Procedures were performed under general anesthesia, mechanical ventilation, prostaglandins infusion, under echocardiographic guidance. Anatomic lesions, type of procedure, short and mid-term outcomes were recorded.
Results: From 2003 to 2011, 13 patients with PA and intact septum underwent 15 percutaneous procedures for perforation and subsequent balloon dilatation of the pulmonary valve, at a mean age of 3.6 days and mean weight of 3.1 kg (2.1 to 3.9 kg). RV was tripartite in 6, bipartite in 8 and unipartite in 1. Four procedures failed to perforate the valve, 2 of them underwent a second and successful procedure. Eleven procedures were successfully performed, with a 5 to 8 mm balloon size (mean 6.5, median 6 mm). Pericardial effusion occurred in 2 cases, requiring emergency surgery in 1. No death occurred during procedure. Five patients (38.5%) underwent subsequent aorto-pulmonary shunt and 2 had ductus arteriosus stenting (15.4%), 2 to 35days after procedure (mean 12.4days), because of persistent significant hypoxemia. Four iterative percutaneous pulmonary valve dilations were performed (30.7%), 6 days to 11 months after first procedure. Two patients died at 4 days and 8 months of age. All 11 survivors (85%) have biventricular anatomy with no (2 cases), mild (8 cases) or moderate (1 case) RV hypoplasia, at 1 month to 8 years follow up (mean 3.5years).
Conclusion: Percutaneous pulmonary valve perforation is an effective procedure in neonates with pulmonary atresia and intact septum. Although short-term aorto-pulmonary shunt is needed in half of the cases, biventricular heart anatomy may be expected in long-term follow-up.

P-266
GORE® SEPTAL OCCLUDER: early clinical results.
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Background: Transcatheter closure of patent foramen ovale (PFO) is a widespread procedure. However the "quest" for the ideal device is still ongoing. Here we present the procedural and early results of transcatheter closure of PFO with the GORE® Septal Occluder.
Methods: Four Italian centers participated in a multicentric registry and collected data from 25 patients undergoing PFO closure by using GSO device. Indication for closure was previous stroke in 15 and previous TIA in 10 subjects.
Results: The procedure was successful in all of the patients. The procedure was performed with local anaesthesia, fluoroscopic and intracardiac echocardiographic imaging in 12 subjects while it was performed under general anaesthesia, fluoroscopic and trans-esophageal echocardiographic imaging in 13 subjects. Twenty-three patients received a 25 mm device while 2 received a 30 mm device. Procedure and fluoroscopy times were 47 ± 12 and 6 ± 2 minutes, respectively. Residual shunting at procedure was absent in all subjects when performed at the basal status while a mild shunt was present in 3 subjects at Valsalva Manoeuvre. Two subjects (8%) experienced vascular complications. At one month follow-up one subject experienced atrial fibrillation. All subjects showed a well positioned device with no signs of complications. No neurological recurrences occurred. No significant residual shunting was found.
Conclusion: GORE® Septal Occluder in our experience is an easy, safe and effective device in closing PFO.

P-267
Stent implantation in patients with native and recurrent coarctation of the aorta. Is application of bare metal better then covered stents?
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Introduction: The aim of the study was to analyze the results of bare metal (BMS) or covered stents (CS) implantation in patients (pts) with native (CoA) and postoperative (ReCoA) coarctation of the aorta.
Material: Between 1999 and 2011, 85 pts underwent stent implantation (75 BMS and 12 CS). Indication for CS use were aortic aneurysm or dissection (native or jatrogenic) in 6 pts, critical stenosis in 4 pts or tubular CoA in 2 pts.
Results: Presented in the table below.
In 53 y old woman with CoA during implantation of CS stroke with hemiparesis occurred. In 40 y old man with ReCoA during

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	Stents In CoA	Stents In ReCoA
No of pts.	65	20
Age (y)	27,4 (6,5–57)	27,4 (10–54)
Grad. before (mmHg)	48 (18–89)	43 (26 -111)
Grad. After (mmHg)	8 (0-35)	13,2 (0-37)
F-up (y)	4,1 (0,2-9,5)	5,2 (0,1-12,6)
Successful (%)	94	80
Migration Stiff leasion (pts)	4 0	0 4

predilatation with balloon acute aortic wall dissection occurred treated successfully with CS implantation. In another 2 pts (12 and 13 y old girls ReCoA & CoA was dilated with BMS stent and 5e and 2 years later during planned redilatation of the stent small aneurysm of aorta was seen in the middle portion of BMS treated with CS. No other complications were observed.
Conclusions: BMS implantation in CoA and ReCoA has good acute, intermediate, and long-term outcome. In selected patients CS implantation can be the treatment of choice or life seving procedure in case of acute complication. Continuous follow-up of patients after stent treatment of aortic coarctation is required due to associated long-term morbidity related to aortic wall complications, systemic hypertension, recurrent obstruction as well as potential need for another interventions.

P-268

For small babies a new Amplatzer Duct Occluder II Additional size: Initial experience of our clinics

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Background: Using any PDA occluder for smaller than 6 kg babies may be harmful. A new Amplatzer Duct Occluder II Additional Sizes (ADO II AS) gives us some opportunity about this subject. We report our initial single-center experience with the new ADO II AS at the small infants.
Material: From August until December 2011, 6 consecutive small babies with PDA were enrolled for intention-to-treat with ADO II AS. The median age was 4 months (2 to 7 months); the median weight was 4.4 kg (4–9.5 kg).
Results: ADO II AS implantation was attempted in all 6 patients. The mean PDA diameter was 2.12 ± 0.57 mm, and the mean PDA length was 4.41 ± 0.92 mm. In one patient, the ADO II AS could not close the defect; hence it was changed to an ADO II. We achieved complete occlusion of the duct in all of the patients on the day of insertion. No major or minor complications occurred.
Conclusion: Transcatheter occlusion of PDA at the small babies with the ADO II AS device is safe and effective, with a high rate of complete occlusion. The device can be deployed completely in the ductal body without any descending aortic obstruction or left pulmonary artery obstruction because of it's specially configuration. Also, the advantages of using it are the smaller sheath sizes and softer shape.

P-269

Non-routine using of the Amplatzer Occluders and the Amplatzer Plugs in children

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Background: Occasionally, some of medical devices used to treat congenital cardiac lesions in the cardiac catheterization laboratory are used on an off-label and non-routine basis. This article discusses about non-routine practice of the Amplatzer atrial and ventricular septal occluders anda plugs.
Methods: In a prospective study, from November 2006 to December 2011, used Amplatzer occluders and plugs in a pediatric cardiology ward were reviewed. Data were collected and analyzed.
Results: The children (n = 442) were catheterized to close their defects by using the Amplatzer devices. The mean age and body weight of the patients was 5.87 ± 4.2 years (range 1 month-17 years) and 20.94 ± 13.73 (range 2-87) kg. Routine using devices counts were 428 patients (96.8%) and non-routine ones were 14 (3.2%). Non-routine, off-label using indications and used devices were the Amplatzer Vasculer Plug 1 and 4 (coronary fistula occlusion in three patients, the left ventricle pseudoaneurysma occlusion in one patient, atypical PDA occlusion in one patient, anomalous pulmonary venous drainage occlusion in one patient), the Amplatzer Ductal Occluder I and II (perimembraneous VSD occlusion in one patient and muscular VSD occlusion in three patients), the Amplatzer Muscular VSD Occluder (very large PDA occlusion in two pulmonary hypertensive patients, the RPA to LA fistulous connection closure in one patient), the Amplatzer Septal Occluder (very large aortopulmonary window closure in one patient). The devices were successfully implanted in all of extra-ordinary or difficult, extreme cases.
Conclusions: The use of a medical device outside of its approved label is commonly referred to as "off-label use." This study showed that the problem of off label, non-routine using the devices exists in pediatric cardiology. Percutaneous closure of defects with the non-routine Amplatzer devices is safe and effective with high success rate and high follow-up term outcome.

P-270

Transcatheter closure of ruptured sinus of Valsalva aneurysm with nitinol mesh occluders

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Introduction: Ruptured sinus of Valsalva aneurysm (RSOVA) is rare shunt lesion with scant data about its transcatheter closure (TCC).
Methods: From March 2007 to September 2011, 12 patients (pts) –mean age 37,2 years were selected for TCC. Two pts had acquired RSOVA after previous cardiac surgery. Another pt after previous surgical closure of RSOVA had 2 recanalizations. Echocardiography revealed the rupture of right or noncoronary sinus into right atrium in 9 pts and into right ventricle in 2 another pts. In 1 pt RSOVA was open from left coronary sinus to pulmonary artery (PA).The defect diameter was from 3,8 to 10 (mean 6,9) mm. Different nitinol mesh occluders (ductal, atrial or muscular VSD) were applied by antegrade venous approach.In all cases arterio-venous loop was created
Results: All defects were successfully closed (in 1 pt during 2 sesions). In one pt embolization of 2 mm larger than defect Amplatzer Duct occluder occurred. The device was retrieved with lasso and bigger occluder (Cardio-O-Fix PDA) was sucesfully aplied. In another pt (after tetralogy of Fallot repair) with left RSOVA to PA ST-segment depression was observed on the ECG and the procedure was abandoned. All treated pts had com-plete closure at discharge. In 72 y old woman with renal failure, aortic dissection and after aortic valve replacement Cardio-O-Fix PDA occluder was applied in proximal enterance

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to the ruptured SOVA. Because of the presence of important residual leak on the edge of the implant the procedure had to be supplemented by closing of the distal RV orifice of SOVA with 10 mm Amplatzer Muscular VSD Occuder .Trivial aortic regurgi-tation occurred in 1 pt without progression in follow-up. In one pt 3 y after the procedure new shunt close to occluded RSOVA was found. The patient is scheduled for another attempt of TCC of RSOVA.
Conclusion: In appropriately selected pts with RSOVA, trans-catheter closure is a feasible and effective method of treatment.

P-271
Short- and Mid-term Results of Balloon Angioplasty for Coarctation of the Aorta in Neonates
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Background: To review our clinical experience with short- and mid-term results of balloon angioplasty for coarctation of aorta in neonates.
Methods: The data of 51 neonates who underwent balloon angioplasty for aortic coarctation between December 2004 and March 2010 were retrospectively studied.
Results: Balloon angioplasty was performed in 51 neonates (mean age 13 ± 9 days). Of these 41 (80.4%) were male and 10 (19.6%) female. Isolated coarctation was found in 13 patients (25%) and complex coarctation, in 38 (75%). The mean systolic pressure gradient across the coarctation site fell from 36 ± 20 mm Hg before dilatation to 8.6 ± 7.0 mm Hg following the intervention. The mean follow up period was 8.7 ± 9.6 months (range 1-46, median 6 months). Recoarctation developed in 20 (39.2%) patients after an average 3.2 ± 3.1 months. Of these, 9 (45%) underwent redilatation and 11 (55%) surgical repair. On follow up, seven (13.7%) patients died with a follow-up of 1-7 months.
Conclusion: According to our short- and mid-term results, balloon angioplasty has a higher recoarctation rate than surgery. For this reason, balloon angioplasty should be done to ensure survival until full corrective surgery can be performed in patients with complex cardiac disease and poor general condition
Key words: aortic coarctation, balloon angioplasty, neonates.

P-272
Transcatheter Closure of Atrial Septal Defect by Amplatzer Occluder Devices [ASO] In Benghazi Cardiac Center
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Objective: Our study reports the clinical experience and outcome of transcatheter closure of atrial septal defect done in Benghazi Cardiac Center. Significant atrial septal defects are closed surgically or through a transcatheter device, in order to avoid pulmonary hypertension in late life.
Methods: During a 5 years period between December 2004 and December 2009, 68 ASD patients were referred to Benghazi cardiac center for possible transcatheter closure with ASO .17 patient were excluded for TCC, 10/17 patients after TOE evaluation [in which 4 with small defect, 4 with large ASD, 2 with deficient inferior rim], 7/17 after angiographic, balloon sizing and TOE. TCC with ASO done in 51 patients [35 female and 16 male] All procedure carried out under general anesthesia & fluoroscopy guide and TOE control .The selected device was 1 to 2 mm larger than the maximum defect size .The physical examination and TTE were performed prior to procedure and follow up [0,1,3,6,12 months and yearly after].
Result: Patients age between 3.5–61 years, and weight between 15–108 kg, diameter of ASD 6 – 3.2 mm.42 patients with single defect, 4 with 2 ASD [, 5 with fenestrated ASD. The size of device range 8–34 mm [ASO, PFO and cribriform device].
Conclusion: The Amplatzer septal occluder is an effective ASD transcatheter treatment device Careful and detailed patient evaluation and selection of an ASO of appropriate size are important factors for success and avoidance of complication .The atrial septal aneurysm which is frequently associated with multiple fenestrated defects is not problem for transcatheter closure.

P-273
Percutaneous dilation of inferior caval vein and closure of left superior caval vein in the patient after Fontan operation
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Introduction: The purpose of this abstract is presentation of percutaneous treatment of patient after Fontan operation with sudden development of cyanosis.
Material: The 3-year-old girl was born with double inlet right ventricle. The banding of pulmonary artery was performed at 3 month of life, and later a hemi-Fontan operation was performed at 8 month of life. A Fontan completion was done at 10 month of life. She had a catheterization study prior to the Fontan operation which revealed good haemodynamic and morphology. The left superior caval vein was not present. She was doing well with saturation = 90%. When she was 2,5 year-old the saturation decreased to 72-74%. The angiographies revealed stenosis of inferior caval vein and the main flow from lower parts of body was through the hemiazygos and hemiazygos accesoria veins to patent left superior caval vein and to pulmonary veins atrium. The left superior caval vein was connected to right superior caval vein.
Results: After balloon occlusion test of left superior caval vein the increase of arterial saturation from 75 to 94%, without negative consequences, was observed. In these circumstances the successful balloon angioplasty of inferior caval vein was performed. A 8/6 Amplatzer Duct Occluder was deployed in left superior caval vein below the entrance of hemiazygos accessory vein. Control angiographies revealed right flow from lower parts of body with total occlusion of left superior caval vein without restriction of flow. There was no increase of pulmonary artery pressure.
On FU the girl is doing well. The saturation is about 94- 95%.
Conclusion: The active investigation of causes of desaturation in the Fontan patients is necessary in all cases. The best method is diagnostic catheterization with angiographies. The most complications can be treated during the same procedures.

P-274
Cerebral Microemboli Detection during Transcatheter ASD Closure
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Introduction: The aim of this prospective study was to determine the frequency and composition of cerebral microemboli in a paediatric population during transcatheter closure of atrial septal defects.
Methods: A middle cerebral artery in 23 patients (median age 3 years 1 month) was monitored for microembolic signals (MES) using multifrequency transcranial Doppler. MES were automatically identified and differentiated according to their composition; gaseous or solid. The procedure was divided into five periods: right cardiac catheterization, left cardiac catheterization, pulmonary angiography, sizing & sheath placement and device placement. Timing of all catheter manipulations and MES were registered and compared.
Results: MES were detected in all patients. The median number of signals was 63, (range 242 to 9). Over 96% of all MES were gaseous. The total number of signals detected during two periods, sizing & sheath placement, and device placement, was not significantly different (median: 18 and 25) but was significantly higher than each of the other three periods ($p < 0.001$). 37.4% of all emboli were detected during sizing & sheath placement, 47% during device placement. In eight patients the device was opened more than once and the number of embolic signals decreased each time. There was no correlation between number of signals and fluoroscopic time, duration of procedure, age or device size.
Conclusion: This is the first study to investigate the timing and composition of cerebral microemboli in a paediatric population during cardiac catheterization. Microemboli were related to specific catheter manipulations but were not associated with fluoroscopic time or duration of procedure.

P-275
Effect of unfractionated heparin during pediatric cardiac catheterization

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Introduction: Unfractionated heparin (UFH) reduces thrombotic risk related to catheterization but the effects of UFH on the coagulation system in children, especially on thrombin generation, and the proper monitoring of UFH remain unclear. The purpose of our study was to assess the strength of thrombin formation and to determine the effects of UFH in pediatric patients during cardiac catheterization. We also wished to assess the extent of heparinization using a new method, prothrombinase-induced clotting time (PiCT).
Methods: We studied 42 patients (aged 3-12 years) undergoing cardiac catheterization. Twenty-seven patients undergoing percutaneous closure of atrial septal defect or patent ductus arteriosus received a UFH bolus of 100 IU/kg (group A), and 15 patients undergoing venous catheterization did not receive UFH (group B) during the procedure. Thrombin formation was assessed by measuring plasma prothrombin fragment F1 + 2, thrombin-antithrombin complexes (TAT), and D-dimer. UFH was monitored by activated partial thromboplastin time (APTT), anti-FXa, and PiCT.
Results: Markers for thrombin generation remained low during catheterization in group A. In group B, both F1 + 2 and TAT increased significantly ($p < 0.05$) by the end of the procedure when compared with the respective preoperative levels or with the respective levels in group A. D-dimer levels remained low in both groups. In both groups, F1 + 2, TAT and D-dimer increased by the first postoperative day as compared to the respective baseline levels. In group A, 15 minutes after heparinization, APTT was over 180 s, anti-FXa in median 1.5 U/ml (range,1.1-2.4 U/ml), and PiCT in median 1.6 U/ml (range, 1.3-2.4 U/ml), and there was a correlation between anti-FXa and PiCT ($R = 0.84$, $p < 0.0001$). No thrombotic or bleeding complications were observed in either group.
Conclusions: Thrombin generation was enhanced in patients who did not receive UFH, which may increase the risk for thrombotic complications. In patients who received UFH, routine heparinization seemed excessive by all monitoring methods, greatly exceeding recommended therapeutic levels of heparinization. PiCT seems to be a viable method of monitoring heparanization in pediatric patients. Further studies are needed to clarify the adequate heparin dosing for children during cardiac catheterization to prevent thrombotic complications without predisposing the patient to bleeding complications.

P-276
Creation of Fontan Fenestration and Atrial Septal Defect with NRG RF Powered Transseptal Needle

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Introduction: In this study, we present the results of NRG Transseptal Needle application for the transseptal puncture (TSP) in 2 patients and in one patient to create Fontan fenestration.
Case 1: The results of cardiac catheterization of a 14 years-old child diagnosed with PH and glukogen storage disease type 1a were as pulmonary arterial pressure (PAP):118/48 mmHg, mean:69 mmHg, Rp:38 U/m2 and Rp/Rs:92%. Because of syncope development TSP was applied. 10W (2 second) radiofrequency energy was given with NRG Baylis Transseptal Needle (NRG-71-C1). After the application of TSP, septostomy dilation was performed with increasing balloon diameters. Balloon dilation was terminated after observation of a 10% drop of arterial oxygen saturation (SaO2).
Case 2: Cardiac catheterization was performed in a 12 years-old girl diagnosed with transcatheter closed patent ductus arteriosus and Eisenmanger syndrome. PAP was 106/51 mm Hg, mean 77 mmHg, Rp 21.5 U/m2, Rp/Rs %69. Despite combined treatment, symptoms were increased thus TSP was applied similarly to the first patient. However, after the radiofrequency energy was given 2 times to cross into the left atrium. SaO2 fell from 98% to 90%.
Case 3: A 27 years-old male patient with Fontan operation due to tricuspid atresia and 4 years ago lateral tunnel Fontan conversions was made and fenestration was opened surgically. Due to closure of fenestration, protein losing enteropathy, and syncope transcatheter Fontan fenestration was created with NRG Transseptal Needle. SaO2 fell from 89% to 80%.
Discussion: Radiofrequency has been used to create an ASD and in the Fontan palliation to create a fenestration. The NRG Transseptal Needle (NRG, Baylis Medical Company Inc.) has potential advantages over conventional needle. It requires little mechanical pressure, which might reduce the trauma of sudden advancement of the posterior left atrium through the interatrial septum as the needle crosses.
Conclusions: In few studies this device has been used to create the Fontan fenestration and no adequate data are available using this device for purpose of the TSP in patients with PH. However, the use of the device to provide control over the puncture and the decreasing the risk of arrhythmia, may be useful in these patients.

P-277
Initial results of PDA stenting in newborn with duct dependent pulmonary circulation

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Introduction (or Basis or Objectives): Surgical palliations of the congenital heart diseases which have duct dependent pulmonary circulation have high mortality and morbidity in the newborn period. Stenting of the Patent Ductus arteriosus as an alternative percutaneous approach is used by some cardiologist however not widely accepted. The aim of this study is to evaluate the early results of percutaneous PDA stent placement.
Methods: Nineteen patients who had had a PDA stent placed percutaneously between April 2010 and December 2011 were included in the study. Three patients had pulmonary atresia with ventricular septal defect (PA-VSD), eleven PA with intact ventricular septum (PA-IVS), four critical pulmonary stenosis (CPS) and one a PA with single ventricle physiology (CAVSD-PA). The seven patients with IVS-PA were also subjected to valve perforation by radiofrequency (RF). Before the intervention, all patients were diagnosed by standard echocardiography; their PDA anatomy was examined in detail.
Results: The mean age and weight during intervention were 14.2 ± 13.3 days and 3.1 ± 0.5 kg, respectively. The mean of procedure and scopy time, time of stay in intensive care, total out-of-hospital and total follow-up time were 116.31 ± 42.99 min; 31.14 ± 16.78 min; 4.88 ± 6.07 days; 11.00 ± 6.89 days and 215.62 ± 118.90 days, respectively. The mean of the radiation amount was 761.07 ± 792.10 cGy/cm2. The mean of saturation before and after intervention were 67.36 ± 5.83; 85.76 ± 6.87%, respectively. Procedure-related deaths were observed in two patients. The causes of death were pulmonary haemorrhage (n = 1) and retroperitoneal hemorrhage (n = 1). Two patients also died after discharge before surgery due to sepsis (n = 2,) and aspiration pneumonia (n = 1). Eight of 13 patients achieved stent patency during 6 months of follow up and re-stenosis developed in one patient (1/8; 12.5%) who had undergone a Glenn operation at 4.5 months of age.
Conclusions: The placement of a ductal stent has results comparable to those of palliative or reparative surgery, without sharing the risks of thoracotomy surgery. In the newborn with duct-dependent pulmonary circulation, the placement of a PDA stent by the percutaneous approach is a good alternative to surgery.

P-278

Percutaneous Extraction of Cardiac Leads With Evolution Mechanical Dilator Sheath in Young Patients

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Introduction: Endocardial lead infections and lead malfunctions in children is an important issue. This growing problem has increased the interest in percutaneous lead removal technology. In this report, we present our initial experience in percutaneous lead extraction with a novel hand-powered mechanical dilator sheath.
Methods: During 25 months between December 2009 and January 2012, 10 leads in 9 patients were removed. All of the extracted leads were older than 48 months. The leads were removed by using the Evolution mechanical dilator sheath (Cook Medical) with the only rotational cutting force and without laser or radiofrequency energy.
Results: Patients mean age was 15,7 (range 11-23) years old. Indications for lead removal included cardiac device infection in 3 (%33.3) cases, lead malfunction in the remaining 6 (66.7%) cases. The extracted device was a pacemaker in 7 (%77,7) cases and implantable cardioverter defibrillators (ICD) in remaining 2 (%22,3) of them. Among 10 leads, 8 (%80) were right ventricular and 2 (%20) were atrial electrode. The mean time from the preceding procedure was 6,3 years (4–10 years). Device was 9 fr in 6 of 9 cases and it was 11 fr in remainig 2 cases. Complete procedural success with Evolution system alone was achieved in 5 (%55,5) patients (6 leads) and one of them was completely removed with snaring. In 2 leads, partial procedural success was achieved with a remaining ventricular tip smaller than 4 cm (1 and 3 cm). In one patient procedure was failed with a remaining ventricular tip about 8 cm.This patient was reguired surgical intervention for remaining tip. In 8 patients of 9, clinical success was achieved (%88,9), and all of this patients discharged uneventfully without a major complication.
Conclusions: There are not sufficient data about mechanical technique with Evolution system for lead extraction in children. In an adult studies shows that, it is an effective method for chronically implanted pacemaker/ICD leads. So our experience in pediatric patients confirms that too. Further investigation is required to compare success and complication rates with other techniques.

P-279

Percutaneous closure of patent ductus arteriosus under 6 Kg: Is it Safe?

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Background: Percutaneous closure of patent ductus arteriosus is nowadays widely accepted as the treatment of choice of patent ductus arteriosus (PDA). This procedure is considered feasible and safe in patient over 6 kg.Conversely, in infants weighting ≤ 6 Kg, patent ductus arteriosus may be responsable of significant overload of left ventricular and heart failure. In this case surgery a correction is necessary and percutaneous closure has an off-label indications. The aim of this study is to evaluate the feasibility and safety of percutaneous closure of patent ductus arteriosus in children weighting ≤ 6 Kg.
Methods: From April 2000 to April 2011, 15 of 420 patients (3.5%) submitted to transcatheter PDA closure at our Institution were ≤ 6 kg (age 5,5 ± 1.1 months, range 3,5-5,9 mth; weight 5,5 kg ± 0.5 range 4.9- 6 kg).The morphology of ductus arteriosus was conical (n = 7), tubular (n = 6); and window type (n = 2). All pts showed cardiac volume overload at echocardiography and were on anti-congestive pharmacological therapy. In all cases the closure was performed by using the Amplatzer Duct Occluder (AGA Medical Corporation, Golden Valley, Minnesota, USA). In 1 patients, PDA closure was associated to a second interventional procedure (pulmonary sequestration embolization). All patients were included in the follow up program.
Results: Mean PDA diameter was 3.3 + 0.5 mm (range 2.5-4.5). Mean Qp/Qs was 4.2 + 4.3 (2.5-5-1). Overall feasibility of the procedure was 100% without early mayor complication. There was 1 minor complication represented by anemia post procedure. During the follow up (mean 38 ± 23.7 months, range 1-84 month) absence of residual shunt. Immediate occlusion rate was 25.0%, rising to 87.0% at last follow-up control. At last follow-up control, left ventricular diameters were normal in 13/15 (87%) patients. None of the pts is on anti-congestive pharmacological therapy.
Conclusion: Percutaneous closure of large, symptomatic PDA might be considered effective and safe also in very young infants with weight ≤ 6 Kg. Left ventricular overload is always evident in this patients, but at the follow-up, after percutaneous closure a reduction/normalizations of the left chamber diameters is the rule.

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P-280

Arterial duct redilatation/restenting: a single center experience

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Background: Maintaining arterial duct (AD) patency is widely considered an effective alternative to surgical systemic-to-pulmonary artery shunt in neonates with congenital heart disease and duct-dependent pulmonary circulation (CHD-DDPC). Re-dilatation/re-stenting is sometimes needed due to progressive flow reduction secondary to patients growth and/or intra-stent re-stenosis.
Methods: Aim of the study is to evaluate safety and feasibility of re-dilatation/re-stenting procedure compared to primary procedure. Between April 2003 and October 2011, 91 neonates underwent attempt of AD stenting as palliation of CHD-DDPC at our Institution (Group I). Among them, 15 patients needed a second procedure of re-dilatation/re-stenting (Group II).
Results: Procedure was successfully completed in all patients in Group II (100%) versus 85/91 patients of the Group I (93.5%). Twelve patients underwent ductal re-stenting, 2 patients ductal re-dilatation, after 4.8 + 2.6 months from the first procedure. After re-stenting/re-dilatation the duct size increased from 2.2 + 1.4 to 3.3 + 0.5 mm ($p < 0.01$, p = NS vs Group I) and percutaneous O2 saturation increased from 75 + 4 to 93 + 5% ($p < 0.0001$; p = NS vs Group I). Life-threatening complications and need for emergency surgical shunt was recorded in 2/14 group II pts (13% vs 1.1% in Group I) due to acute stent thrombosis after deployment in both patients. In-hospital procedural-related mortality was 6.6% in group II vs 1.1% in group I.
Conclusions: AD re-stenting/re-dilataion is a feasible mini-invasive palliation of CDH-DDPC supporting clinical improvement. Acute thrombosis is a life-threatening major complications most frequentely recorded in the re-restenting procedure. Early and clinical symptomatic stent occlusion seems to be a risk factor. Probably a more aggressive anti-platelet therapy could be necessary in this subgroup of patients.

P-281

Percutaneous Pulmonary Valve Implantation Implantation: Initial Results of Turkish Experience

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Introduction (or Basis or Objectives): Some of complex congenital heart diseases with right ventricular outflow lesions should be treated with conduits. Longevity of all conduits is not lifetime and they become dysfunctional because of calcification or valve degeneration. Therefore, patients who have a conduit need to undergo open heart surgery in several times in their lifespan. The conduit replacement surgery has still high mortality and morbidity rates in many countries all around the world including our country. Percutaneous pulmonary valve implantation (PPVI) has been performed in European countries and was approved by FDA in 2010. We have started to perform implantation soon after this approval. In this paper, we present our cases implanted with pulmonary valve by percutaneous way.
Methods: The patients who underwent PPVI between October 2010 and November 2011 were included in the study. The demographic, echocardiographic and hemodynamic data as well as clinical status of patients were assessed before and after implantation.

No	Age	Sex	Weight	Diagnosis
1	16	E	54	C-TGA
2	17	E	58	VSD-PA
3	39	K	51	VSD-PA
4	18	E	49	DORV-PS
5	21	K	73	TGA-VSD -PS
6	18	E	76	TOF- Cor. Ab

	Op.score	Conduit Age	Conduit Type	Conduit size
	1	13	Freestyle	19
	2	12	Freestyle	21
	1	20	Gore-Tex	19
	2	14	Gore-Tex	19
	2	10	Contegra	22
	1	16	Hema shield	14

	PRE-INTERVENTIONAL			
No	RV pressure mmHg	Ao. Pressure mmHg	RV/Ao. ratio	PR
1	90	120	0,7	0
2	78	110	0,7	0
3	48	108	0,4	4
4	42	114	0,3	4
5	104	116	0,9	3
6	54	128	0,4	4
	POST-INTERVENTIONAL			
	RV Pressure mmHg	Ao. pressure	RV/Ao ratio	PR
	35	110	0,3	0
	45	105	0,4	0
	44	105	0,4	0
	24	110	0,2	0
	70	114	0,6	0
	45	120	0,3	0

Results: Six pulmonary valve implantation procedures were performed. Edwards Saphien valve was implanted in five cases and Melody valve was used in one case. Indications were stenosis in two patients, severe regurgitation in two patients, and mixed lesion for two patients. No procedural mortality was observed. Right ventricular pressure and gradient of conduit were successfully reduced in patients with stenosis and mixed lesions. Pulmonary regurgitation disappeared in all patients after implantation. All patients were discharged 48 hours after procedure. No major complication occurred. Patients were followed up for one month to 1 year and there was no case of valve dysfunction, stent fracture or restenosis.
Conclusions: Procedural results and short-term outcomes of the PPVI were very promising in our patients. PPVI is a good alternative of surgical conduit replacement. Any patient with conduit dysfunction on pulmonary position should be assessed as a candidate for PPVI.

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P-282

Early and Mid-Term Outcomes of Transcatheter Management in Pulmonary Atresia with Intact Ventricular Septum

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Introduction (or Basis or Objectives): Pulmonary atresia with Intact ventricular septum (PAIVS) is revealed with broad spectrum of heterogeneous morphology. Perforation of the atretic valve, balloon dilatation and stenting of the patent ductus arteriosus are the percutaneous techniques which are used with increasing frequency also in our clinic.They have some advantages over surgery including short hospital stay, short ICU stay etc. The main goal of the primary interventional approach is to avoid surgery. However, a group of patient with PAIVS still need to surgery because of the poor right ventricular growth. Therefore, the final achievement of the initial percutaneous treatment strategies is still debatable. In this paper, we present the early and mid-term results of the percutaneous approach in our clinic in order to investigate the final effect of interventional therapy according to initial morphology.

Patient Data	
Age (day)	12.7 ± 12.1
Gender(M/F)	9/8
Weight (kg)	3.2 ± 0.9 (2.2-4.7)
RV Size	
Monopartiate	3
Bipartiate	11
Tripartiate	3
Coronary sinusoid (absent/present)	7/10
Ebstein malformation (absent/present)	15/2

Patient Data	Median (range)
Days of Intensive Care stay	2.54 ± 3.3 (1-11)
Days of hospital stay	7.2 ± 4.2 (3-23)
Balloon dilatation+PDA stenting	5
RF assisted Valvotomy	3
RF+PDA stenting	6
PDA Stenting	3
Pulse oxymetry saturation	
Before procedure	70 ± 6.14 (60-80)
After procedure	89.27 ± 4.61(80-96)
Early /Late death	2 (%12)/3(%18)

Methods: Between May 2010 and November 2011, seventeen neonates underwent transcatheter intervention. Detailed echocardiographic examination based on RV size, tricuspid valve morphology and coronary sinusoids were applied to all patients before the intervention. Nine of the patients were boys and 8 were girls. The mean age was 12.7 ± 12.1 days and weight was 3.2 ± 0.9 kg.Table 1 summarizes the patients' characteristics and preinterventional echocardiographic findings.

Results: Two procedure-related mortalities occurred. The mean follow-up period was 6.9 ± 5.2 months (2-19 months).The mean duration of intensive care was 2 ± 1, 8 days. One of the patients with PAIVS achieved biventricular physiology after pulmonary valve perforation. Three patients have been followed without any reintervention or surgery. Fourteen out of 17 patients achieved stent patency during 6 months of follow-up, while re-stenosis developed in one patient (1/13; %7.7) who had underwent Glenn operation at 5 months of age. Seven patients are still waiting for Glenn anastomosis without complication and reintervention (Table 2).
Conclusions: As a primary treatment, transcatheter management for PAIVS is a feasible, safe, and effective palliation in newborns. Right ventricular size determines the type of the intervention. The early outcomes can be comparable with surgical palliation. However, a group of PAIVS particularly with severe right ventricular hypoplasia cannot achieve a surgery free life even after successful primary percutaneous intervention.

P-283

Six years results of endovascular stenting of coarctation of the aorta in children – single center experience

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Objective: To describe the experience and outcomes of stent implantation in patients with native (CoA) and postoperative (rCoA) coarctation of the aorta performed in single Paediatric Cardiology Center.
Methods: Retrospective data collection was analyzed. Primary endpoints were peak systolic catheter gradient reduction, stented segment diameter increase. Early and late complications, changes in antihypertensive medication after stenting were recorded.
Material: Between 1998 and 2010, 70 pts with arterial hypertension (AH) underwent stent implantation (CoA-52pts, rCoA–18pts). For primary treatment 71 stents were used (9-Palmaz, 38-Cheatham-Platinum, 18-covered Cheatham-Platinum, 4-Intrastent, 2-Advanta). Covered stents were implanted due to small aneurysms after balloon angioplasty (3pts), coexistence of PDA (5pts), severe coarctation (12pts). Median patients age was 13.4 +/- 3,4 yrs (3-18).
Results: There was significant improvement ($p < 0.001$) in pre versus post stent coarctation diameters (5.45 +/- 2.42 mm (1-11.5, med.4) vs (15.59 +/- 2.69 mm (6.5-22, med.16) and systolic gradient (34.55 +/- 10.35 mmHg (16-67, med.32) vs. 2.77 +/- 4.6 mmHg (0-18, med.0). During follow-up period 6.05 +/- 4.02 yrs (0.2-14,5 med.6) 58%pts did not need antihypertensive treatment and all the others have better control of AH on lower doses of medications. About 93% (65/70) of the procedures were followed up by CT and confirmed good post-procedural anatomy in the first year after stent implantation. Stents fracture (5) and neointimal hyperplasia (6) were confirmed in CT during later follow-up. Additional procedures were performed in 46/189pts (24,33%) – stent redilation—9, covered stent implantation due to small aneurysms – 2, aortic arch narrowing – 2, stent fracture with in-stent stenosis – 5.
Conclusions: 1.Stent implantation in native and postoperative coarctation of the aorta has good acute, intermediate, and long-term outcome 2. Continuous follow-up of patients after stent treatment of aortic coarctation is required due to associated long-term morbidity related to aortic wall complications, systemic hypertension, recurrent obstruction as well and need for additional interventions.

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P-284

Immediate and short-term outcome of the Gore Septal Occluder® (GSO) in patent foramen ovale and atrial septal defect closure – Early clinical experience

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Background: A new GORE® septal occluder (GSO) was granted CE mark in Europe in June 2011 for the treatment of patent foramen ovale (PFO) and atrial septal defect (ASD). Major changes have been made to the device and delivery system compared to the HELEX® device. The new delivery system has simplified the implantation procedure and the retrievability of the device after deployment if needed. The design of GSO has improved the device apposition ability and tissue response whilst keeping its atraumatic design, low septal profile with minimal septal distortion and long-term biocompatibility. We report the immediate and short-term outcome of patients treated with the device.
Methods: Consecutive patients treated with GSO for PFO or ASD were evaluated within six months following their procedures.
Results: Ten patients (6 women) with a mean age 53 ± 10 (range 40 – 67) years were reviewed 76 ± 30 (range 45 – 120) days following their procedure. Nine patients had PFO closure due to at least one episode of paradoxical cerebral or peripheral arterial embolism and one patient had ASD closure.
All the procedures were performed under local anaesthetic using intracardiac echocardiograhic and fluoroscopic guidance. All the devices were successfully deployed upon the first attempt without the need for retrieval. There was no device embolisation or vascular complications. Only one patient had minor residual shunt on agitated saline contrast and valsalva manoeuvre immediately after the procedure.
During follow-up, one patient had developed paroxysmal atrial fibrillation. Two patients reported transient or intermittent palpitation without any arrhythmia documented. In all the patients, there was no evidence of thrombosis associated with the device and no residual shunt was noted on echocardiogram with agitated saline contrast and valsalva manoeuvre. There was also no evidence of wire fracture in the five patients who had follow-up fluoroscopy examination.
Conclusion: In a small number of predominantly patients with PFO, our initial experience indicates that GSO is associated with an excellent immediate and early complete closure rate without associated device thrombosis or wire fracture. Larger and longer-term study is needed to determine its efficacy and durability.

P-285

Percutaneous device closure of large patent ductus arteriosus in the very young

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The practice of percutaneous interruption of ductal shunt continues to evolve with the advent of novel occlusive materials. These evolutions have advanced the frontiers for interventional paediatric cardiologists, allowing them to undertake percutaneous ductal shunt interruption in progressively younger paediatric patients.
We report the case of a 3 month old baby symptomatic and failing to thrive with a large patent ductus arteriosus (PDA), measuring 4 mm in diameter. The birth weight of the baby was 3 kg and weight at 3 months was 2.9 kg.Elective percutaneous device closure of the large PDA was successfully undertaken with complete abolition of the ductal shunt using a 4/4 mm Amplatzer Duct Occluder II -ADO II. (AGA Medical Corporation, Nathan lane, North Plymouth, USA). The device was deployed across the ductus from the retrograde approach through the 4Fr ADO II device delivery system. (AGA Medical Corporation, Nathan lane, North Plymouth, USA). There was no flow obstruction in the descending aorta or left pulmonary artery. The baby exhibited immediate clinical improvement allowing withdrawal of anti-failure medications.
The device design of ADO II lends itself favourably for successful percutaneous closure of large PDA in small babies weighing <3 kg without any vascular complications.

P-286

Sternotomy As a Safe Alternative 'Access' for Transluminal Interventions In Low Birth Weight infants

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Background: transluminal interventions in low birth weight infants with CHD remain a therapeutic challenge: injury of the accessed vessels or risk for cardiac tear. We used sternotomy for hybrid direct cardiovascular access in 4 low birth weight infants as bail out stenting procedure.
Patients and methods: 4 patients: hybrid suite; sternotomy; A/ 3 patients (1620, 2190 & 2630 g) with Fallot with PA and hypoplastic pulmonary trunk (2-3 mm). Procedure : purse string on RV; 2 vascular clips as radio-opaque markers: one at the pulmonary valve, one at the puncture site. Double needle technique: one 21G needle as a reference for depth adjacent on the surface of the RVOT; 2nd 21 G needle used to puncture. Punction in two motions: first perpendicular to the surface into the RV, second angulation of the needle towards and through the atretic outflow tract. 0.0 14" wire into the pulmonary arterial branch; needle exchanged for a 4 Fr short sheath; angio by mini 1cc injections through side-arm. A 5/16 mm coronary stent deployed into the RVOT to obtain an "intracardiac Sano shunt". B/ 1 patient (970 g) with critical aortic coarctation and open duct. Procedure: purse-string on the ascending aorta with vascular clip; 0,014" wire into the descending aorta; needle exchanged for a 4 Fr short sheath; angiograms through side-arm. A 3/8 mm coronary stent deployed into the aortic isthmus; arterial duct clipped.
Results: Sternotomy well tolerated by all patients remaining hemodynamically very stable throughout procedure.
A. Fallot: adequate palliation with symmetrical anterograde flow (sats> 92%). After median 3 months an additional stent was necessary transvenously in all patients due to muscular in-fundibular stenosis. Two patients went to full repair at 5 months; one patient with multiple hilar stenoses requires additional transluminal procedures.
B. Coarctation: good aortic flow, stent resection and arch repair at 4 months. No associated morbidity.
Conclusion: Medial sternotomy can be a safe alternative access for bail out transluminal cardiac interventions in low birth weight infants, allowing conventional repair at bigger weight. The technique with 2 identical needles and radio-opaque markers simplifies the hybrid procedures.

P-287

Aorta-Right Atrial Tunnel Closure with Transcatheter Technique: A Case of 3 Years Old Child

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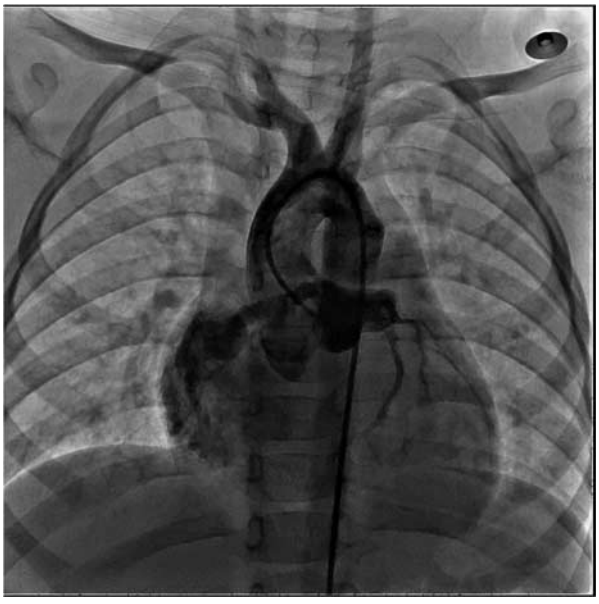
Introduction (or Basis or Objectives): Aorta-atrial tunnel is a rare and severe congenital disorder which can lead to death if untreated. A girl with persistent murmur in her left upper sternal border (8 months after PDA closure) was catheterized due to continuous flow from left coronary sinus to right atrium in echocardiography. An aneurysmatic tunnel sized 5,5 mm in diameter and 4 cm in length was closed by Amplatzer vascular plug 4 device. This case is important of being the first that aorta atrial tunnel was closed by vascular plug 4 device successfully.
Methods: 3 year old girl was referred to our center for continuing murmur 8 months after PDA closure. In her physical examination continuous murmur is heard in her right upper sternal border. Continuous flow from wide left coronary sinus to right atrium thru a tunnel was seen in transthoracic echocardiography. 30 mmHg pressure gradient was measured by continuous wave Doppler. Also Amplatzer ductal occluder –1device in ductus arteriosus and small apical muscular VSD were noticed. In angiography oxygen saturation in middle part of right atrium: was measured as 99%, in upper part: 83%, right ventricle: 90%. An aneurysmatic tunnel sized 4 cm in length and, 5,5 mm in diameter was seen (Picture 1). Tunnel was closed by vascular plug 4 device (picture 2). By aortic root angiography device remained stable in tunnel orifice and minimal shunt was seen through the device.
Results: Continuous murmur was disappeared in control examination of patient. Residual shunt wasn't seen in tunnel by transthoracic echocardiography. She was still followed by regular visits in our clinic.
Conclusions: In this study, we aimed to report the closure of extremely rare defect: aorta-RA tunnel, by Amplatzer vascular plug 4.



P-288

Interventional VSD closure using the Amplatzer Duct Occluder II™ in a small infant with concomitant Aortic Coarctation

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Percutaneous VSD closure has become an established alternative to surgery. Sheath size and bulkiness of the occluders limitate this method for smaller children. The Amplatzer Duct-occluder-II™ is smooth, flexible and implantable through small sheaths. We report a case of VSD closure with this occluder.
Patient: A 5 month old girl with 4,5 kg was referred with signs of heart failure and pulmonary edema. Echocardiography revealed a singular 5 mm midmuscular VSD with pulmonary hypertension. No additional intracardiac defects, the isthmic region was 2,5 mm, with an instantanous gradient of 50 mmHg and a small open duct. Surgical management of midmuscular VSDs may be difficult, especially when attempted without ventriculotomy. Interventional closure therefore seemed to be a promising alternative all the more coarctation was planned to be corrected by lateral access.
Procedure: The VSD was passed from the LV and a typical loop was formed. The 5F delivery system (TorqueVue low profile, AGA™) was introduced transvenously and advanced through the VSD into the left ventricle. An Amplatzer Duct occluder II with a length of 5 mm and a the middle portion of 5 mm was constituted and positioned under permanent transthoracic echo guidance. The soft appearance of the umbrella allowed exact positioning of the different parts, although the maneuvres had to be done very gently, not to pull the system out of the defect. Echo control showed an excellent alignment of both retention discs on the septum. Procedure time was 78 minutes, fluroscopy time 18 minutes. No procedural complication occured, no changes in ECG as well. Coarctation repair followed next day without complications.
Result: Two months after implantation the patient has still a mild to moderate slit like residual shunt at the inferior edge of the device. Nevertheless pulmonary pressure was normal, LV dimensions returned to upper limit of normal.
Discussion: VSD closure with flexible devices and reasonable sheath size can be an alternative to surgical closure even in small patients below 5 kg.Procedure and fluro time will decrease along the learning curve, oversizing of the device may help in avoiding residual shunts.

P-289

Trancatheter atrial septal defcts closure without balloon sizing.

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Introduction: transcatheter closure of secundum type atrial septal defects (ASD) is a valid alternative to surgery. Balloon sizing of ASD allows a careful choice of the device, but has also been associated with overstretching of the defect. Recent studies show that transcatheter closure without balloon sizing is safe and feasible and several equations to define device size on the basis of transthoracic (TTE) or transoesophageal echocardiography (TOE) have been proposed. The objective of our study is to report our experience with ASD closure with and without sizing.
Methods: between May 2006 and December 2011, 59 patients (aged 4 to 81 years) affected by ASD underwent transcatheter closure. Patients with patent foramen ovale were not included. All patients underwent TTE and TOE. 43 patients (group A) underwent to closure of ASD with balloon sizing and the last 16 (group B) without sizing.
Results: In group A mean maximal ASD diameter estimated by TTE was 15 mm (6-30 mm), TOE 14 mm (4-31 mm) and stretched diameter was 19 mm (6-30 mm). The mean diameter of devices was 20 mm (7-34 mm). In group B mean TTE diameter was 13 mm (4-31 mm), 13 mm (range 4-31 mm) with TOE, the mean diameter of device was 14 mm (7-28 mm). In both groups there were no adverse effects secondary to device implantation, and there were no residual defects.
Conclusions: the closure of ASD without balloon sizing is feasible, safe, allows the use of smaller devices, and finally is cheaper.

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P-290

Palliative pulmonary balloon valvuloplasty for symptomatic infants with tetralogy of Fallot and unfavourable for total surgical correction

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Objective: We aimed to evaluate the results, efficacy and utility of palliative pulmonary balloon valvuloplasty (PPBV) procedure performed on symptomatic infants with tetralogy of Fallot (TOF) unfavourable for total surgical correction, which is generally preferred about or after one-year old age in our center
Methods: Study population included 40 TOF patients all under one year of age underwent PPBV between July 2005 and November 2010. The indications for PPBV are hypoxia (oxygen saturation<70%) and/or McGoon index <1.7.
Results: PPBV procedure was successfully performed in all forty patients. Mean oxygen saturation increased from 66.1% to 83.5% after the procedure which was statistically significant. No mortality and significant complication has occurred due to procedure. Low saturation temporarily occurred in two patients immediately after the procedure. During follow-up, 11 patients had total repair directly without any other procedure and 19 patients had a second cardiac catheterization. In patients who had a second cardiac catheterization, statistically significant increase was detected between the McGoon indexes, the pulmonary annulus diameters Z-scores, and the right and left pulmonary artery diameters Z-scores. After the second cardiac catheterization, 17 more patients had total repair. When we consider all the patients, 28 of them (73.6%) were able to have total repair without any additional palliation, but only five patients required a BT shunt operation in spite of successful PPBV. The total repair time was delayed median 12.5 months (range: 2-23) for these 28 patients.
Conclusion: PPBV procedure could be considered an effective, beneficial and safe procedure in TOF patients that eliminates hypoxia, ensures the development of the pulmonary vascular bed and postpones the point at which total repair is done.

P-291

Percutaneous treatment of complex aortic coarctation and aortic arch hypoplasies with Cheatham Platinum stents

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Background: Transcatheter stent implantation (SI) is widely considered as the first-choice treatment of aortic coarctation in adolescents, adults and children over a certain weight. However the procedure is still challenging in complex anatomic variations such as sub-atretic obstruction or isthmus atresia, arcus aorta lesion, long-segment aortic hypoplasia or in the case of coexistence of aneurysm or in Turner syndrome.
Methods: Between August 2007 and December 2011 totally 79 stent implantation was performed in 74 patients with aortic pathologies. Safety and efficacy of the procedure was retrospectively evaluated in 25 patients with coarctation having different features than standard coarctation. In 9 patients coarctation was associated with PDA. The lesion was in transverse aortic arch in 3 cases. In 2 patients with isthmus atresia Sİ was performed after guide wire perforation and balloon dilatation. In 6 patients with sub-atretic coarctation predilation had to be done before SI since the severe obstruction did not let the passage of long sheath or even the catheter. The lesion was aneurysmatic in three cases. A patient had a very long coarctation segment ant the other had clinical features of Turner syndrome.
Results: Stent implantation was successfully completed in all cases. 29 Cheatham platinum stents (NuMED, Hopkinton, NY) were used in 25 cases. Mean age 15.82 ± 7.9 years (4.2- 38 years) and mean weight was 50.8 ± 21.6. Covered stents were used in all cases except in three patients with aortic arcus lesion. Mean pressure gradient dropped from 43.5 ± 23 to 2.7 ± 3.2 mmHg. There was no residual leak in patients with PDA. Aneursmatic lesion was completely covered in 3 patients. The mean follow up duration was 23.5 ± 12.3 (2-46), median 25 months (2-46 months). There was no procedure related death.
Conclusion: Our results showed that transcatheter SI is safe and effective in "complex" form of aortic coarctation. Since each case may require different technical approaches it is recommended to display detailed anatomical features before procedures.

P-292

Stenting of the Right Ventricular Outflow Tract in Patients with Tetralogy of Fallot

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Introduction: In a few patients with Tetralogy of Fallot (TOF) a palliative treatment has to be considered prior to corrective surgery, because of severe hypoxia. Beside surgical implantation of an aortopulmonary shunt and stenting of the ductus arteriosus, stent implantation into the right ventricular outflow tract (RVOT) is a third option, especially in patients with a small RVOT and a duct already closed.
Methods: We investigated safety and efficacy of RVOT stenting in patients with TOF retrospectively. For this we analyzed the intervention, as well as pre- and postinterventional data of n = 7 patients with TOF, in whom we intended to place a stent into the RVOT.
Results: The median age was 20 (1-167) days. The procedure was effective in 6/7 patients. In three of them we implanted one stent and in the other three we needed two stents to cover the whole obstructed segment. The diameter of the stents was 5 mm in four, and 6 mm in the other two patients. The median hospital stay after the intervention was 5 (1-8) days. The patient in whom we couldn't implant the stent had an atrio-ventricular-septal defect with TOF and an azygos continuation. This child got an aorto-pulmonary shunt, but died later on in multi-organ failure.
Currently n = 4 got surgical correction 44, 55, 86, and 144 days after RVOT stenting. The stents could be explanted easily despite adhesions, except one patient, in whom a larger incision of the right ventricle was needed to remove the two stents 55 days after implantation. One patient is waiting for surgical correction. One patient is lost follow-up,
Conclusions: RVOT stenting is an effective procedure to stabilise pulmonary perfusion and to postpone surgical correction in cyanotic patients with TOF. Corrective surgery is feasible as usual also after RVOT stenting.

P-293

3-Dimmentional rotational angiography to assess the pulmonary circulation in patients with single ventricle after Bidirectional Glenn operation

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Background: Rotational angiography with three-dimensional reconstruction (3DRA) is an emerging technology that has been successfully used in neuroradiology, electrophysiology, coronary angiography and more often in visualizing congenital heart defects. It could be a beneficial adjunct to fix plane angiography and could enhance diagnostic capabilities in patients with single ventricle after various stages of palliation. We report our experience using 3DRA to visualize the pulmonary circulation in patients with single ventricle after Bidirectional Glenn operation (BDG).
Methods: A retrospective analysis of all patients after BDG who underwent 3DRA was performed. Philips Allura system was used to acquire non-gated, breath-held images. During a 240 degrees, 4.1 seconds isocentric rotation, 122 angiographic images were acquired and automatically reconstructed in real time.
Results: Between 05/2010 and 12/2011, we performed 80 3DRA's in 68 patients after BDG. All patients underwent diagnostic catheterization, which in 32 (47%) was fallowed by 38 interventions. Median age and weight was 3.8 yrs (1.5-7) and 16 kg (8.5-58 kg), respectively. Median contrast dose for 3DRA acquisition and for total study was 2 ml/kg (0.7-3.3) and 4.8 ml/kg (2.0-15.5), respectively. Median area dose for the whole study, time of fluoroscopy and total time of study was 132,8 cGycm2 (25.9-1056.8), 7,7 min (0,7-80) and 52.5 min (15-180), respectively. There were no acute complications related to 3DRA. Overall quality of 3DRA images was graded by the primary operator as good in 64 (80%) studies and satisfactory in 9 (11%). Seven (9%) studies were graded as bad due to: angiographic catheter pushed too far into the proximal pulmonary artery making visualization of superior vena cava impossible in 5 (6%), wrong localization of isocenter in further 2. In the remaining 3DRA's vena cava superior, Glenn connection, right and left pulmonary arteries were visualized. In all 38 interventions 3DRA images were judged by the operator to be superior to fix plane angiography in making decision concerning the interventions or in assessing the result.
Conclusions: In patients after BDG operation 3DRA allowed for good visualization of superior vana cava, BDG shunt and course of pulmonary arteries. It was superior to fix plane angiography in planning and assessing results of percutaneous interventions.

P-294
Doppler and Radionuclide Pulmonary Blood Flow Patterns After Transcatheter Closure of Patent Ductus Arteriosus

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Objective: Impaired left lung perfusion (LLP) has been described after transcatheter closure of the patent ductus arteriosus (PDA). This study was conducted to evaluate impaired LLP following occlusion of persistent arterial duct with both echocardiography and radionuclide study.
Methods: Between November 2008 and December 2011, a total of 60 patients (mean age 16.5 ± 16.7 years) underwent successful transcatheter PDA occlusion. Cook detachable coil was used in 31 patients, Amplatzer duct occluder (ADO) was used in 29 patients. Echocardiography were performed to all patients in order to calculation of the Doppler velocity index (DVI) and lung scintigraphy. The DVI was calculated by the difference between the left pulmonary artery (LPA) and right pulmonary artery (RPA) peak flow velocities relative to the pulmonary trunk (PT), and expressed in percentage, terms.
Results: Decreased LLP was found in 8 patients, 2 with Cook detachable coil (6.5%) and 6 with ADO (20.7%). These patients displayed greater DVI values compared with the others [median DVI = 26 (10-50) versus 15 (8-50)] ($p = 0.043$). When DVI ⩾%30 is taken as the cut-off value, it is possible to estimate unimproved patients with 92.2% sensitivity and 85% specificity.
Conclusion: Impaired LLP may appear following transcatheter closure of PDA with various devices and DVI has high sensitivity and specificity in predicting patients with LLP deficiency.

P-295
Percutaneous revalvulation of the RVOT in Belgium

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Introduction: Percutaneous valve implantation of the RVOT is a feasible treatment option for important valvular dysfunction. A mandatory database has been requested for reimbursement in Belgium; the scientific data are available for analysis.
Patients and methods: Nationwide multicentre interim analysis of percutaneous valve implantation in the RVOT; prospective and ongoing study. Since 2006 percutaneous Melody® valve implantations are performed in Belgium. Until 12/2011 a total of 77 valves have been implanted in 4 centres.
Results: Median age at implantation of 20.2 years (range 6.0 – 81.6). Male predominance (ratio 2:1). Primary lesions : Tetralogy of Fallot (37), pulmonary atresia with VSD (5), common arterial trunk (6), TGA with VSD and PS (13), aortic valve (Ross) (12) and critical pulmonary stenosis (4). The RVOT characteristics before revalvulation : native or patch extended RVOT (7) requiring open cell stents to anchor, homograft (51), contegra conduit (3), bioprosthesis (16). The mean diameter of the conduit was 19 mm (range 10 – 27). Pulmonary stenosis was the main reason for revalvulation (41), pulmonary regurgitation (23) and mixed PS/PR (13). In 70 patients (90%) prestenting was done, with a clear trend to go for 100% prestenting. In 19 patients more than one stent was necessary (max 3). A total of 36 covered and 54 bare stents were used. RV systolic pressure dropped from mean 56 mmHg (range 28 – 104) to 42 mmHg (range 23–91) after valve implantation. Complications early : (2) pulmonary artery bleeding solved by covered stent implantation, (1) ischemia due to LAD compression solved by surgical removal of the stent. Complications late : stent fractures (3). No deaths.
Conclusion: Percutaneous valve replacement in conduits and homografts in the RVOT is a safe and grateful treatment with futile morbidity. Adequate prestenting to create a nice landing zone is a key feature avoiding many complications such as embolisation, recompression and fracture. Covered stents allow to expand any breackable – dilatable conduit beyond the nominal value at implantation or later during growth. Open cell stents allow to hook on the dilated "native" outflow tracts.

P-296
A new method for percutaneous treatment of coronary artery fistulae in young children

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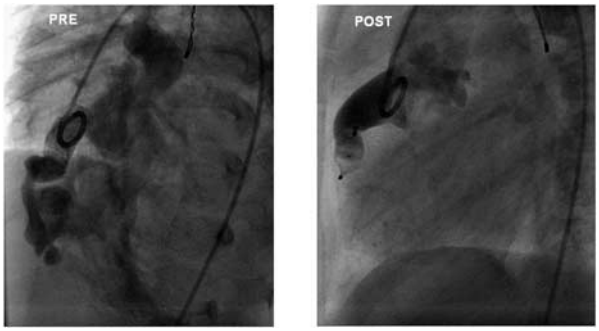
Background: Percutaneous occlusion is nowadays the treatment of choice for coronary artery fistulae (CAF). However, the arterial approach for different devices needs large introducers and catheters during a long procedure. Its use in very young children with large fistulae was hypothetical.
Method: A new Plug (Plug "4", AGA-SJM)can be introduced in a 4F catheter. This Plug in Nitinol is retrievable and very easily expanded.
Patients: Since 01/01/2010, 3 patients (10 months, 23 months, 28 months) with large CAF presented with clinical heart failure. Two CAF were between the right coronary artery and the right ventricle with multiple sites of drainage, one between the left coronary artery and the right atrium. Only one Plug 4 for each child was necessary with a diameter of 6 mm in the 3 cases. Complete occlusion was achieved immediately in 2, and within a few minutes after a temporary occlusion with a 4F balloon wedge pressure catheter.



Conclusion: Large coronary artery fistulae can now be treated percutaneously using small catheters during a relatively simple procedure. This new device brings an elegant solution for large CAF in young children, avoiding open heart surgery.

P-297
Balloon dilation of pulmonary vein stenosis using PAXLITAXEL eluting balloons: midterm result in an infant
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Introduction: Pulmonary vein stenosis has an unfavorable outcome because neither surgical nor interventional therapy prevents restenosis. According to promising results in preclinical studies, single infants with pulmonary vein stenosis have been treated by balloon dilatation using balloons coated with paclitaxel, an antimitotic agent from cancer therapy. First results were encouraging, however, follow-up was cut off early in the two patients published so far, because both died within a few weeks.
Case report: A girl with univentricular heart, increased pulmonary perfusion, and mesocardia was treated by pulmonary banding at 3 weeks. Within the next weeks an increasing stenosis of the left sided pulmonary veins was suspected by echocardiography and confirmed by cardiac catheterization. Subsequently a Damus-Kaye-Stansel anastomosis, an aortopulmonary shunt, and a sutureless repair of the left sided pulmonary venous obstruction were performed at the age of 4 months. At the age of 6 months, a stenosis of the aortopulmonary shunt caused implantation of a 4 mm coronary stent. Concurrently severe restenosis of the left pulmonary veins was diagnosed (Fig. 1) and treated by balloon dilatation. 6 weeks later, re-evaluation in the cath lab revealed severe restenosis, and again dilatation of the left pulmonary veins was performed now using paclitaxel coated balloons (5 and 6 mm diameter). This procedure was repeated at the age of 10, 13, and 16 months. 2 weeks after the last intervention (Fig. 2), surgical treatment with right sided Glenn anastomosis and left sided aortopulmonary shunt was performed. 8 days after surgery the girl went home. Out-patient follow-up after 6 weeks revealed the girl in a proper clinical condition with accelerated left-sided pulmonary venous return (Doppler Vmax 2.3 m/s).
Conclusion: Repeated balloon dilatation of pulmonary venous obstruction using paclitaxel eluting balloons may be useful in the interventional treatment of this frequently fatal condition. Although restenosis occurred also in our patient after the use of paclitaxel eluting balloons, the diameter of the treated vessel showed a reasonable increase, and the patient was able to undergo the next surgical step.

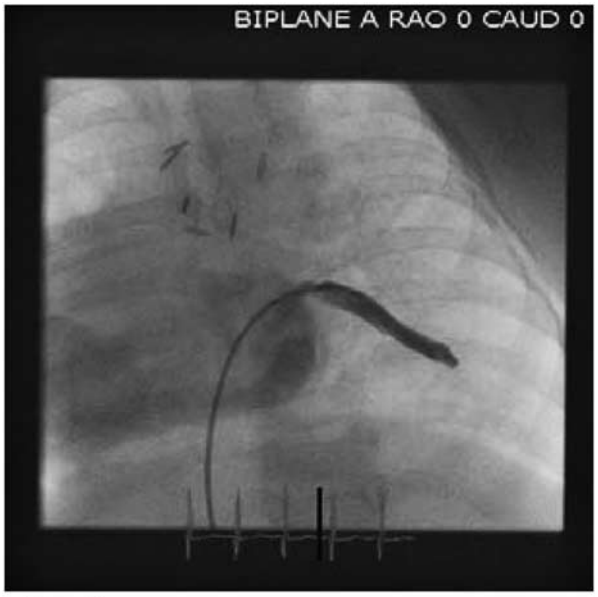

Fig. 1

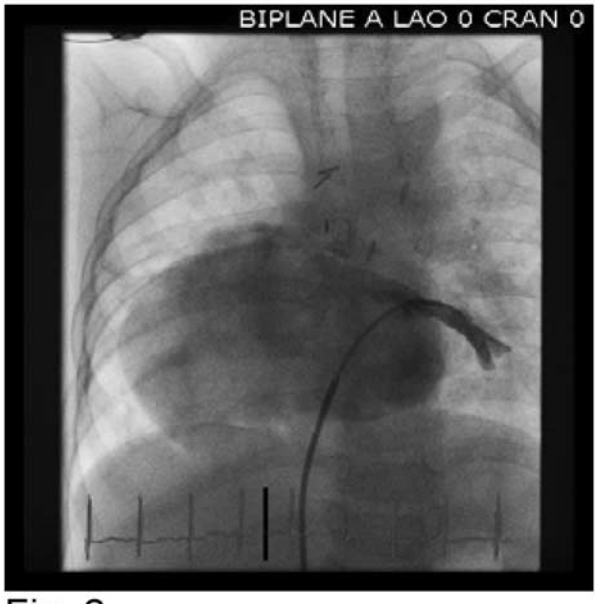

Fig. 2

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P-298

Interventional closure of perimembranous ventricular septal defects: Experience using the Amplatzer Duct Occluder II.

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Introduction: Interventional closure of perimembranous ventricular septal defects (pmVSDs) which are partially closed by an aneurysm remains controversial concerning AV-block. We report our experience using the soft Amplatzer Duct Occluder (ADO) II for pmVSD closure.
Methods: In five patients a pmVSD was closed between 4/2010 and 12/2011; age: 6 months – 8.6 years, pmVSD size 4-10 mm, formation of an aneurysm left an effective opening of 2.4 – 5.5 mm.Distance of pmVSD to aortic valve 3-5 mm.No patient had aortic insufficiency.
Results: Hemodynamic measurements: left to right shunt Qp/Qs 1.4-1.6/1; normal pulmonary artery pressures. Technical procedure of VSD closure was done via the classically described way forming a loop from the left ventricular side to the right side. Final closure was performed from the right ventricular side. The following devices were placed:
ADO II 4-4 mm in 1 patient, 5-6 mm in 2 patients, 6-6 mm in 1 patient and ADO II AS 5-4 mm in 1 patient.
24 hours post closure we visualised no residual shunts in all patients. None of the patients had aortic insufficiency or a remarkable AV valve insufficiency. No AV-block occurred. In 1 patient the right disk was deployed at the right atrial side. No tricuspid valve dysfunction occurred, device was left in place. One patient who was known with an intermittent AV block I pre catheter presented with a transient AV dissociation post VSD closure and a left anterior hemiblock and right bundle branch block 1 month after closure without any clinical symptoms. Total FU time was 1 – 20 months.
Conclusion: In our experience of 5 patients, pmVSDs with aneurysm formation can be closed with a good result using the Amplatzer Duct occluder II. A total closure rate of the pmVSDs could be reached. To avoid interference of the device with the tricuspid valve or a position of the right disc towards the right atrium, we recommend choosing the minimal device length needed. We might speculate that the ADO II device seems softer and less traumatic to the His bundle. Long term follow-up has to verify patency of AV-conduction.

P-299

Awareness about transcatheter removal of intravascular foreign bodies among pediatric health care providers

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Background: Central line insertion is a routine for invasive hemodynamic monitoring, total parenteral nutrition, infusion chemotherapy and medications. Fragmentation and dislodgment of lines into the vascular system is a rare complication. From our experience with the mode of referral of four patients, we had the impression that not all the paediatricians are aware of transcatheter approach to solve this problem.
Aim: To study the awareness of paediatric health care providers about the non surgical methods of retrieval of intravascular/cardiac foreign bodies.
Methods: A questionnaire was developed with 3 questions and given to all paediatric healthcare providers in two large tertiary care centres. The three questions were about number of patients seen with central line, experience with complications and which speciality would they refer patients with dislodged central lines. We included in the questionnaire residents beyond the 3rd year of training, fellows, consultants, of all paediatric subspecialities, including surgery.
Results: The questionnaire was given to 128 professionals. A total of 101 persons (79%) answered the questionnaire. Out of these 101, 14 were excluded due to incomplete answers. Total 87 questionnaires were included for analysis. 18% were consultants, 38% were assistant consultants and senior registrars, 43% were senior residents. Results showed that of those who answered the questionnaire, 39% saw more than 30 central lines per year, 39% experienced this complication. 81.5% opted for surgical method of removal, mostly to paediatric surgery (61%). Only 18.5% chose for nonsurgical methods. At our centre we successfully removed 4 lines over last 2 years.
Conclusions: Most paediatric health care providers in the two tertiary care hospitals evaluated are not aware that patients with such complications should be referred to paediatric cardiologists as percutaneous retrieval of intravascular foreign bodies is the standard method of treatment that can be performed easily, safely and successfully. It is important to raise the knowledge of percutaneous approach in paediatric services. Successful retrieval can obviate the need for major surgeries

P-300

Transcatheter retrieval of cardiovascular foreign bodies–A 15-year single centre experience.

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Introduction: Transcatheter retrieval of cardiovascular foreign bodies is well established but there are no large paediatric studies in the literature. We aimed to review our 15-years experience of transcatheter retrieval of foreign bodies such as occlusion devices, line fragments and wires from the cardiovascular system.
Methods: Retrospective record review of all children with transcatheter foreign body retrieval. Cases of retrieval of malpositioned patent ductus arteriosus (PDA) coils were also included.
Results: Transcatheter retrieval of foreign bodies from the cardiovascular system was attempted in 64 patients. Retrieved foreign bodies include embolized devices (n = 37), central venous and arterial line tips (n = 14), guide wires (n = 4), pulmonary artery stents (n = 4), ruptured balloon tip (n = 3), fractured ventriculo atrial shunt (n = 1) and fractured sheath introducer (n = 1). The incidence of device embolization for ASD, VSD and PDA devices was 1.4% (7/470), 2.8% (4/140) and 2.4% (26/1066) respectively. Retrieval sites included pulmonary arteries (n = 28), PDA (n = 8), descending aorta (n = 7), central veins (n = 11), right atrium (n = 5), right ventricular outflow tract (n = 3) and left ventricular outflow tract (n = 2). Median patient age and weight were 3.1 (0.1-16) years and 13.1 (1.7-74) kg respectively. Transcatheter retrieval was successful in 61/64 (95%) and had to be performed surgically in 3 patients. In 55 the retrieval was from venous and 9 were from arterial side. Mean sheath size was 8 (4-16) French. Gooseneck snare was the most commonly used retrieval device. Mean procedure time was 100 (15-316) minutes and fluoroscopy dose was 40 (0.4-320) Gy/cm2. The procedural complications included death in 1 patient within 24 hours of procedure with a retroperitoneal bleed, had transient loss of foot pulses in 4 and excess blood loss requiring transfusion in 2.
Conclusion: Transcatheter retrieval of cardiovascular foreign bodies can be performed safely in the majority of children including infants thus obviating need for the surgery. Retrieval of foreign bodies is an essential skill required by all interventionists not only for device implantation but also for other indications. It is essential to have a comprehensive inventory of equipment with staff conversant with their use.

P-301

Transcatheter Management of Failed Melody Valves After Successful Placement

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Background: Transcatheter replacement of pulmonary valve with Melody valve is an accepted therapy in select patients with failed right ventricular – pulmonary artery (RV-PA) homograft or pulmonary prosthetic valve. Short term result is excellent but mid & late outcome is unknown. Evaluation of the durability and longevity of the Melody valve is ongoing. This study examined the Melody valve failure after successful placement. Failure is defined as symptomatic recurrence of significant stenosis ± regurgitation.
Methods: Since July 2006, 62 of 63 Melody valves had been successfully implanted in 55 patients. The valves have failed in 6 patients 0.4-4.5 (mean 2.6, median 2.3) years after successful implant. Three of the six were pre-stented before valve replacement. One failed Melody valve was replaced surgically and one await cardiac catheterization. The remaining 4 patients underwent transcatheter placement of another Melody valve after evaluation by CT-angiography. All were pre-stented before replacing successfully with another Melody valve.
Results: There was no mortality, vascular or myocardial injury. The valves narrowed from a mean diameter of 19.7 (range 18-22, median 19.0) mm in diameter after the first Melody valve, to a mean of 11.4 (range 9-15, median 10.8) mm. All valve stents were fractured, spontaneous in two, and after high impact chest contact activity in the others. One was inadequately expanded at first Melody valve implant, and two were pre-stented. The ensuing significant stenosis was reduced from 46-64 (mean 56, median 57) mmHg to 15-34 (mean 22, median 17) mmHg after the second Melody valve implant. All new Melody valves were competent.
Conclusions: We conclude that it is feasible to manage failed Melody valves by transcatheter placement of a second Melody valve, thereby prolonging the lifespan of the original RV-PA conduit and avoiding another open heart procedure.

P-302

Stenting of Systemic to Pulmonary Artery Gore-Tex Shunts

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Introduction: Creation of systemic to pulmonary artery shunts (SPAS) remains the initial palliative procedure in the management of a wide range of infants with complex cyanotic CHD. There are risks of in-shunt and anastomotic stenoses, ongoing shunt occlusion, and somatic outgrowth of shunts. Transcatheter stenting of stenosed shunts holds promise to address these problems.
Methods: Eighteen stenting procedures of stenosed SPAS were performed over an 8-year period. Median age was 10.8(1.5–152) months. Median weight was 8.1(3.2-30.3)kg. All had placement of a pre-mounted coronary stent which from 2007 onwards was routinely chosen to be larger than the original shunt size.
Results: Twenty-four stents were placed to address occlusion, stenosis or uniformly small SPAS at a median interval of 7.9(0.3-81) months post surgery. Stent diameter chosen was a median of 114(100-133)% of original shunt size. Procedure time, including diagnostic study, was 96(66-239) min and dose exposure 13(1.2-77.0) Gycm2. Arterial saturations increased from a median 70(55-79)% to 82(75-85)% post-stenting [$p < 0.001$]. Minimal shunt diameter increased from a median of 2.3(1.0-2.7)mm to 4.3(4.0-6.0)mm [$p < 0.001$]. There was one procedural death due to shunt thrombosis. One patient died 36 hours post-procedure from cardio-respiratory arrest, with a patent shunt on ultrasound. There was one neurological insult following stent re-canalization of an occluded shunt but there was no neurological deficit seen during follow up.
Conclusion: Stenting of stenosed or small SPASs is a versatile and effective technique to address in-stent and anastomotic stenosis and to increase stent diameter and resultant pulmonary blood flow, thereby delaying further surgery.

P-303

Significant aortic obstruction with the Amplatzer Duct Occluder II

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Objective: Describe the cases of significant aortic protrusion of the ADO II.
Methods: Since its introduction in June 2008, the ADO II was favored for all types PDA<5.5 mm. Patients in whom we encountered a severe aortic obstruction of this device were described.
Results: Between 2008 and 2011 the ADO II was used in 61 patients. In three of them, the aortic disc severely obstructed the aorta. N1: (6.6 Kg; type A; 3.2 mm large; 7.4 mm long). The aortic disc of a 6-6 ADO II kinked 10 days later leading to severe coarctation. It was surgically resolved. N2: (6.9 kg; type C; 2 mm large; 8.4 mm long). The aortic disc of a 3-6 ADO II, anterogradely delivered, immediately bulged in the aorta after detachment. This was recovered by aortic balloon inflation. N 3: (7Kg; type A; 3 mm large; 8 mm long). The aortic disc of a 4-4 ADO II, arterially delivered, immediately kinked in the aorta after detachment. It was snared, retrieved and successfully replaced by a 5-4 ADO I.
Conclusions: the ADO II is not the device of choice in type A PDA. Discs are bulky and highly articulated. Immediate or even late aortic protrusion may occur leading to severe obstruction especially in small infants.

P-304

Eliminating Stent Slippage Using a Combined Sheath-Balloon Catheter

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Introduction: Percutaneous stent placement has become a mainstay of transcatheter treatment of vascular stenosis & coarctation. Pre-mounted stents may not be available in the sizes needed. The stents are usually hand crimped onto the balloon in vitro, and advanced into the stenosis through long sheaths previously placed. Stents can rarely slip off the balloon during its introduction through the hemostatic valve of the sheath, but more likely through the heart or vessels that have tight corners that distort or kink the sheath. The stent has to be retrieved, repositioned or remounted prolonging the procedure. Surgical removal may be necessary if the stent slips distally. Despite advances in balloon and catheter technology and delivery techniques, this complication persists. We describe a novel method of stent delivery that eliminates stent slippage using a front loaded stent-in-sheath delivery system.
Methods: The stents ($n = 8$) were delivered under GA to relieve pulmonary artery stenoses or prior to pulmonary valve replacements ($n = 6$). Initially, using a conventional balloon-in-balloon catheter, a Palmaz P4010 stent slipped off the balloon completely as it was advanced through the sheath in the right ventricular. The stent was retrieved and remounted on the balloon of an all-in-one balloon sheath catheter (NuMed Inc., Hopkinton, NY). The stent, front loaded on the balloon (10-18 mm), covered by the sheath,

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permitted movement of the stent-in-sheath as one system. The procedure was repeated in others with pulmonary artery stenosis.
Results: Multiple arrhythmias occurred during the passage of the stent-in-sheath through the RV, but there was no myocardial or vascular injury. Significant friction was encountered during the stents' passage through the RV and stenotic pulmonary valve. Despite significant force used to advance the stent-in-sheath through the RV and into the pulmonary artery, the stents remained on the balloon, and was deployed uneventfully
Conclusion: Using the front-loaded all-in-one balloon sheath catheter, stent slippage off the balloon is eliminated. The procedural time is shortened, and morbidity reduced. This should be the device of choice in stenting stenoses.

P-305
Implantation of the new Nit-Occlud PDA-R device in children below 10 kilograms
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Introduction: Interventional closure of a patent arterial duct (PDA) has become a common and safe procedure in most pediatric cath labs. However, despite the modern devices available, it still remains a challenge in those children with low body weight and a large PDA. Several new PDA occluder systems have been developed in the last years. One of them is the Nit-Occlud PDA-R device which was especially designed for large PDAs. The clinical experience and initial trial with this occluder published so far accepted only children with a body weight greater than 10 kilograms.
Methods: We report our most recent experience in five children (age 6-33, median 13.4 months) with a body weight from 5.4 to 13 kg (median 9 kg) with large PDAs: ductal length was 14,6 mm (median), there was a large ampulla (median 12.8 mm) which exceeded the diameter of the aorta and large diameter of the duct (mid-PDA median 6.8 mm, narrowest median 3.6 mm).
The occluder size is determined by the minimum diameter of the PDA, the occluder stent must be at least 1.5 times, better 2 times greater: in four cases, the Nit-Occlud PDA-R with an aortic disc of 12 mm, a stent of 7 mm and a length of 8.5 mm was selected and in the fifth case one with an aortic disc of 14 mm, a stent of 8.5 mm and a length of 9.5 mm.All devices were implanted using the femoral venous access with a 6F sheath.
Results: All five devices were successfully implanted under sedation, without general anesthesia and without complications, e.g. dislocation with pulmonary or aortic obstruction. A sufficient occlusion of the PDA was documented by angiography and echocardiography in all cases. The patients were discharged from hospital two days after implantation.
Discussion: The new Nit-Occlud PDA-R device is suitable even in children with a body weight below 10 kilograms, when a relative large PDA is present. The re-inforced retention disc allows an optimal positioning in the aortic ampulla without obstruction and the flexible cylindrical plug helps to adapt this device to various duct anatomies.

P-306
Treatment of extremely tortuous and hypoplastic aortic arches by implantation of JOTEC™ E-XL Aortic Stents
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Introduction: Extremely tortuous aortic arches combined with arch hypoplasia and stenosis is a rare finding. Even after successful stenting of the transverse arch, the blood pressure may stay high because of the anatomical course of the aortic arch. Therefore in many centers a surgical approach is preferred. The E-XL Aortic Stent (JOTEC GmbH, Hechingen, Germany) was initially manufactured for aortic lesions e.g. dissections. It has an open cell design in the middle section and a closed design at its ends. This retrospective study describes the immediate effectiveness of these stents in this specific patient group.
Methods: We report on three patients (9, 11 and 23 years) with the described anatomy who were treated in our center during the past 3 months. Despite successful stent-implantion in the transverse arch region, a relevant brachiocephalic hypertension and resting blood pressure gradient (20-40 mmHg) remained. After adequate angiographic documentation and measurements of the anatomy, the optimal stent size was selected. In two patients (9 and 11 yrs.), 18×70 mm E-XL Aortic Stents were implanted using a 12F delivery system, in the GUCH-patient a 24×100 mm stent was used via a 14F sheath by a transfemoral approach.
Results: In all cases, stent implantation was successfully performed without complications. Due to the length and size of the stents implanted, the aortic arches were straightened up and their diameter adequately extended in all patients. Only minimal residual pressure gradients (< 10 mmHg) were documented immediately after implantation. No long time effects are seen so far due to the short follow-up time.
*Discussion:*Extremely tortuous aortic arches with hypoplasia and stenosis usually cause brachiocephalic arterial hypertension and interventional treatment may be a therapeutic challenge. The combination of the closed cell design with a high radial force at its ends and the open cell design in the middle section makes the E-XL Aortic Stent an interesting alternative to common stent implantation in these patients. Kinking seems to be avoided and the tortuous anatomy can be straightened up. This combination makes these stents useful offer these challenging patients an interventional treatment modality.

P-307
Percutaneous pulmonary valve implantation in small children: a single center experience
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Introduction: The advent of percutaneous pulmonary valve implantation (PPVI) may avoid risks of serial surgical reinterventions in patients with dysfunctional right ventricle outflow tract (RVOT). As experience with PPVI is gained, new challenges arise from complicated anatomy and patient characteristics. A patient body weight over 35 Kg is the traditional recommendation to achieve successful Melody valve implantation. We report our experience in such a technique in smaller patients.
Methods and Results: From January 2009 through December 2011, 10 patients weighing ≤ 30 Kg underwent transcatheter Melody valve implantation in our institution; median age was 6 ± 2,5 years and median weight was 21,8 Kg (range 16-29,5 Kg). Primary implantation indication was Pulmonary Regurgitation in 7, and mixed disease in 3. Type of RVOT was bioprosthetic valve or conduit in 8 cases (7 Contegra Conduits), and native tract in 2.

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The valve was delivered from a femoral venous approach in 7, and from the right internal jugular vein in 3. Pullback gradient across RVOT decreased from 25 ± 8,95 mmHg to 10 ± 5,07 mmHg (p = 0,004). The Right Ventricle/aortic pressure ratio decrease from 0,65 ± 0,17 to 0,45 ± 0,10 (p = 0,004). Intraprocedure complications included: pulmonary hemorrhage (n = 1); complex tachycardia (n = 1); elevation of Left Ventricle filling pressure treated acutely with milrinone and non invasive ventilation (n = 1). No patient had more than mild pulmonary regurgitation early after implantation or during follow up (median time 11 months). Only 1 patient presented significant stenosis associated with stent fracture.
Conclusions: We consider percutaneous Melody valve implantation as a safe and effective therapy in infants with dysfunctional RVOT weighing less than 30 Kg (even as small as 15,8 kg).
Jugular vein rout may suppose a technical advantage for Melody delivery in these patients. Suitable anatomical substrate can also include native RVOT.

P-308
Interventional therapy of stenotic pulmonary arteries in adult patients with mechanical pulmonary valve prosthesis
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Introduction: There is a growing population of patients with congenital heart defects who require repetitive surgery for right sided cardiac lesions including pulmonary valve replacement. In some, mechanical valve replacement is performed. Stenosis of peripheral pulmonary vessels may occur in these patients too and interventional therapy may be indicated.
Methods: We report our experience in 3 patients with a mechanical valve in pulmonary position and a supravalvular pulmonary pathology in whom we have performed the interventions across the mechanical valve without complications. We continuously used a long sheath, which holds the mechanical valve open during the procedure, as a leaflet protection. All balloons were retracted into the long sheath and distal to the mechanical valve.
Results: 23 year old female patient who underwent mechanical aortic and pulmonary valve replacement for repair of a common arterial truncus showed a significant paravalvular aortic leak and subatretic stenosis of the right pulmonary artery just behind the ascending aorta. The paravalvular leak was closed and the RPA-stenosis was treated by primary stenting using a 34 mm CP stent with a Mullins balloon 14 mm x 40 mm.A 25 year old male with Tetralogy of Fallot (TOF) had a 21 mm SJM mechanical valve placed in pulmonary position after multiple previous surgeries including three conduit exchanges. A significant stenosis of the right pulmonary artery was detected and dilated several times using a VACS III(18 mm x 40 mm) balloon.The 21 year old male with pulmonary atresia and ventricular septal defect had undergone multiple operations, finally resulting in the implantation of a 21 mm SJM mechanical valve in pulmonary position. There was a gradient of 50 mm Hg between the RV and distal MPA due to supravalvular stenosis closely after the mechanical valve. The supravalvular stenosis was then successfully dilated by using a (18 mm x 30 mm) Mullins balloon.
Angiographic and echocardiographic function of the pulmonary valve was unchanged and excellent without malfunction after the procedure.
Conclusions: Interventional therapy of pulmonary vessels is feasible in patients with mechanical valves in pulmonary position. The procedure can be performed safely by using the technique described.

P-309
Deliberate dilatation of the meshwork of CP-Stents– creation of adequate flow to overstented vessels
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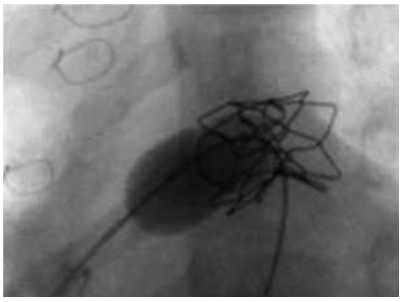
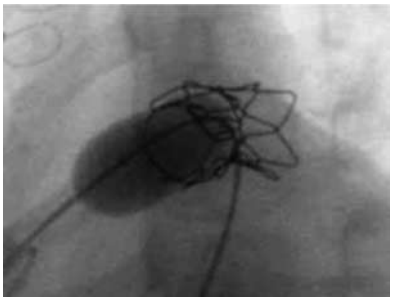
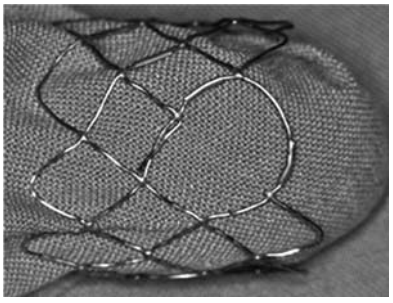
Introduction: The implantation of stents has become a routine interventional procedure for stenotic lesions in pediatric cardiology. However, the area of branching blood vessels is particularly difficult and "overstenting" may impair blood flow. We report our initial experience in deliberate dilatation of the meshwork of CP-stents to overcome this problem.







Patients: Two patients had a CP-stent plaved to treat left sided pulmonary artery stenosis and the stent length necessary for secure placement necessitated overstenting of the right pulmonary artery. In the third patient treatment of aortic recoarctation required overstenting of the left subclavian artery. In all patients flow impairment occurred, partially due to the underlying anatomy and vessel distortion. In all patients, the meshwork was crossed and sequential balloon dilatation using high pressure balloons was performed. Fracture of the welding points of the CP-stent was achieved and flow obstruction was resolved thereafter.
Discussion: The use of the CP-stent may offer specific possibilities when treating stenotic vessel areas with important side branches. Deliberate dilatation of the meshwork is possible with the use of high pressure balloons.

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P-310

Stenting of the right ventricular outflow tract below the pulmonary valve as a palliative measure in symptomatic neonates with TOF

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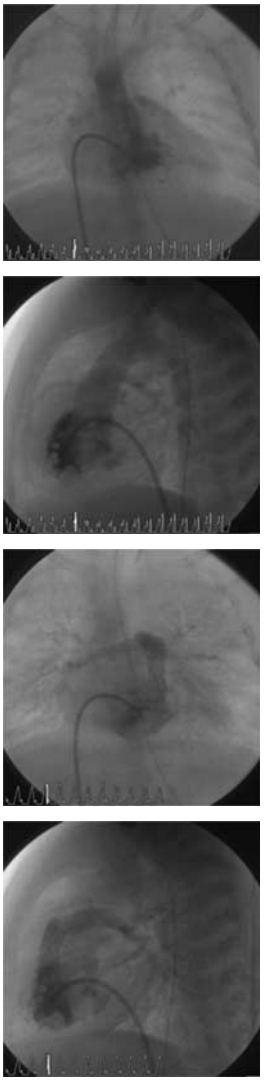
Introduction: Symptomatic neonates with tetralogy of Fallot (TOF) may require early treatment to improve oxygenation. Cyanosis in these patients is usually caused by severe infundibular stenosis, high grade valvular pulmonary stenosis, hypoplastic pulmonary arteries or a combination thereof. Conventional treatment may include balloon dilatation of the pulmonary valve, surgical shunt insertion or early repair.



Patients: We report of two symptomatic neonates with hypercyanotic spells in whom balloon dilatation was unsuccessful and severe cyanosis was caused by massive infundibular hypertrophy. Stent implantation into the RVOT was performed using premounted Palmaz-Genesis Stents (7 mm x 18 mm) and without overstenting the pulmonary valve. Immediate and persistent normalization of oxygenation occurred followed by complete relief of clinical symptoms. Elective repair could be performed thereafter.
Discussion: Stent implantation in the right ventricular outflow is a challenging procedure in neonates; however excellent relief of RVOTO may be achieved without impairing the pulmonary valve. Therefore this procedure offers an attractive alternative to palliate symptomatic newborns with TOF.

P-311

How many stents are enough to stabilize the RVOT before percutaneous pulmonary valve implantation

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Introduction: The percutaneous implantation of pulmonary valves (PPVI) has become standard in many centres. Initial results have shown that pre-stenting of the RVOT is necessary to guarantee good long-term results of the valve. Stenting should aim at a complete stabilization of the RVOT without recoil to prevent stent fracture.
Patient: We report on a 11 year old boy with d-TGA, pulmonary atresia and VSD who underwent a Rastelli procedure as initial treatment. Due to the anatomy, the implanted RV-PA Contegra®-conduit (12 mm) was entrapped between the heart and the sternum and severely compressed, leading to suprasystemic RV-Pressures. Before PPVI sequential stenting on the complete RVOT was necessary to stabilize the RVOT and then implant an 18 mm Melody® valve. We used a total of 6 stents to stabilize the RVOT and avoid repeat external compression and recoil. The residual gradient was 10 mmHg, the RV-Pressure 40/0-10. On follow-up there is no recoil now 8 months after the procedure and no signs of valve malfunction.
Discussion: Stabilization of the RVOT may require several stents to overcome the special anatomy in patients with congenital heart defects. This approach however should be followed to achieve long lasting function of the valve.

P-312

Transcatheter closure of patent Ductus Arteriosus in children weighing 8 Kg or less: a single center experience

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Introduction: Transcatheter occlusion has become an effective therapy in most patients with patent ductus arteriosus (PDA). Procedure complicactions and technical difficulties have been related to lower body weight patients, especially with large shunts. We reviewed clinical outcomes of transcatheter closure of PDA in children weighing less than or equal to 8 kg in a single center.
Methods: Between January, 2004 to December, 2011, 135 patients with PDA underwent transcatheter closure in our institute. Among them, 37 weighed less than or equal to 8 kg, All these patients, underwent transcatheter closure of PDA using either Amplatzer duct Occluder (ADO) Type I or II, or Amplatzer Septal Occluder (ASO). A retrospective review of the treatment results and complications was performed.
Results: The mean age of patients was 8,8 ± 5.9 months (median, 9 months) and the mean weight was 6 ± 1.7Kg (median 6.3 Kg), with 12 of 37 (30%) weighing less than or equal to 5 kg.The average minimum PDA diameter was 3 ± 1.2 mm (median 3 mm). Devices implanted were ADO I In 31 patients (83.33%), ADO II in 6 patients(16.67%), and ASO in one. Immediate complete occlusion occurred in 26 patients (70%), with no more than minimal residual shunt in the rest. None required surgery. 3 children (8%) had procedure related complications (mean weight 4 ± 0.6 Kg); mild narrowing of descending aorta was produced in 2 cases (immediate gradients of 10 mmHg and 15 mmHg); and mild narrowing of left

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pulmonary artery (LPA) in one (maintaining peak systolic velocity resulting in 20 mmHg in the 2 years echo control).
Conclusions: We consider transcatheter closure of PDA as a safe and effective therapy in infants weighing less than or equal to 8 kg. With sufficient experience, transcatheter closure of PDA can also be accepted as the gold standard of treatment for this group of patients.

P-314
Seven Years Libyan Experience in Structure Heart Disease Interventions
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Benghazi Cardiac Center and Benghazi Hospital for Sick Children, affiliated to (formerly) Al-Arab Medical University. 320-bedded tertiary referral teaching hospital, accredited by Board of Paediatrics
Benghazi cardiac center started in 2003, this tertiary referral centre for the Eastern region of Libya. The Heart Centre (BCC) serves the whole of North, West and the South referral with a population of 5.6 million people. The service includes diagnoses, the newborn, Infant, Paediatric and Adult Congenital heart disease and Cardiac Surgery. The Hospitals is designated a supra regional centre for infant cardiac surgery, we started diagnostic catheterization with Libyan team. Interventional cath was done as workshops with international Congenital Cardiologist visitors; we started interventional catheterization alone on 2007-2008. During these 7 years we perform around 239 structural heart disease interventional cases. 70 cases were ASDs transcatheter closure, 66 was PDA transctheter closure, 63 was pulmonary balloon valvuloplasty, 7 was aortic balloon valvuloplasty, 17 rashkind atrial septostomy, 5 balloon angioplasty for COA, 4 stent implantation in CoA and 2 in pulmonary branch stenosis, one case trascatheter closure of coronary artery fistula with PDA Amplatzer device, and 4 VSDs trascatheter closure.

P-315
Endovascular stent implantation for coarctation/recoarctation of the aorta
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Objectives: Endovascular stents have been used recently in coarctation (CoA) /recoarctation as an alternativ method to surgery or balloon angioplasty (BA). The aim of our study was to evaluate our results of endovascular stent implantation.
Methods: We studied retrospectively the datas between 2006 and 2012 regarding the types of previous invasive procedures, the types of endovascular stents, the change of invasive/noninvasive peak systolic gradient and antihypertension therapy after stent implantation.
Results: Twenty-one patients with CoA (11 native and 10 recoarctation after surgery or BA) with a mean age of 17 yrs (range 12-54) underwent stent implantation. The types of previous surgical repairs were patch aortoplasty of 7, subclavian flap of 2 and end-to-end anastomosis of 1. The ratio of bare/covered stent were 6/15. Peak systolic pressure gradient decreased from 42,20 $\pm$ 20 (18-88) mmHg to 5 $\pm$ 6 (0-22) mmHg ($p < 0.0001$). The peak velocity of descending aorta measured by echocardiography decreased from 3,7 $\pm$ 0,6 to 2,5 $\pm$ 0,5 m/s ($p < 0,0001$). Three patients developed complications, including two patient after bare stent implantation with small aorta rupture. One of them required surgical treatment, the other one healed without any procerudes. Another patient developed a small femoral arteriovenous fistula which resolved after conservative therapy. Follow-up for 20 $\pm$ 16 months the noninvasive peak systolic pressure gradient decreased from 52 $\pm$ 21 (20-90) mmHg to 16 $\pm$ 18 mmHg ($p < 0,0001$). At their last follow-up visit, 19 from the 20 patients with hypertension remained on antihypertensive medications.
Conclusions: The blood flow dynamics in the coarctation of the aorta could be successfully managed with endovascular stents, although the patients remained on antihypertensive therapy in lower doses. Major complications occurred only with bare stent implantation. The ratio of patch aortoplasty among recoarctation were high.

P-316
Single centre experience of Balloon Angioplasty of Branch Pulmonary Artery Stenosis after Arterial Switch Operation
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Introduction: Branch pulmonary artery (PA) stenosis is a well recognised complication after arterial switch procedure. Balloon angioplasty of the residual lesions is well established. We aim to evaluate the safety and efficacy of the procedure in our centre, from 2004 to 2011.
Methods: We have retrospectively identified all patients who had branch PA dilatation after arterial switch operation. Seventeen procedures were performed in fourteen patients. Two patients were excluded due to associated supravalvar pulmonary stenosis. Patient characteristics and haemodynamic data, including right ventricular (RV) and systemic arterial pressure, RV to systemic pressure ratio, pullback gradient across the branch PA stenosis and angiographic measurements, were analysed. Student's paired t test was used and a P value of <0.05 was considered statistically significant.
Results: Fourteen patients underwent a total of 15 procedures. The median age was 4 years (5 months- 16 years) and median weight 16.4 kg (6-61.1). In 11 out of 15 interventions the RV to systemic pressure ratio remained unchanged. In the remaining 4 the change was not statistically significant (p 0.10). There was no change in the pullback gradient in 9 procedures. In the remaining 6, the gradient was reduced from a mean of 26 to 21 mmHg (p 0.0211). Mild angiographic improvement was noted in 4 patients but there was no statistical significance (p 0.13). Two of the failed dilatations underwent successful stent placements during the same procedure and showed good immediate results. There was more than 50% reduction in pullback gradient and the RV to systemic pressure ratio dropped from 66% to less than 50%. There were no complications.
Conclusion: We have demonstrated that even though balloon angioplasty of branch PAs post arterial switch procedure is safe, it is not effective in the majority of patients. We are of the opinion that PA stenting should be the primary procedure in this group of patients, especially with the development of the latest stents which can be expanded to adult size.

P-317
Transcatheter Closure of Multiple Ventricular Septal Defects
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Introduction: The feasibility of transcatheter closure for single muscular ventricular septal defect (VSD) is well-demonstrated through several studies. Although several cases with multiple

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VSDs in the same patient are often included in those studies, specific report on multiple VSDs closure is lacking. Our goal was to compare feasibility, effectiveness and safety between cases with single and multiple congenital VSDs.
Method: From January 2007 to January 2012, transcatheter congenital VSD closure was performed in 30 patients, including 21 with a single muscular VSD and 9 with multiple VSDs. The mean age and weight was 2,4 (11 months to 7) years and 12 (7,3 to 28) Kg, respectively. All procedures were performed under general anesthesia and and transesophageal echocardiography guidance.
Results: Forty-three Amplatzer devices were implanted during 34 procedures, including 22 (1 to 6) for patients with multiple VSDs. Thirty Muscular VSD Occluder, 3 ADO II, 8 Amplatzer Septal Occluder and 2 Amplatzer Cribriform (35 mm) were used. In 3 patients with multiple VSDs, complete or partial closure of the defect with mild residual shunt was accomplished with a single device (Amplatzer Septal Occluder in 2 and Muscular VSD Occluder in one). There were 3 complications with single VSD closure and 4 in multiple VSD closure (p NS). One patient with single apical VSD experienced cardiac arrest during the procedure, necessitating circulatory support. He recovered but suffered from neurological sequel and his VSD was successfully closed perventriculary during subsequent surgical repair. Satisfactory device implantation was achieved in all the other patients. Other complications included in patients with multiple VSDs closure a tricuspid valve lesion with moderate regurgitation that was repaired at the time of subsequent surgery (pulmonary artery banding removal), a cerebral ischemic stroke with favorable outcome and the need for blood transfusion in 2 cases. In patients with single VSD closure, blood transfusion was necessary in 2 cases. There was no mortality.
Conclusion: Transcatheter closure of multiple congenital VSDs is feasible and effective, without significant additional morbidity as compared to single VSD closure. Various devices other than Muscular VSD Occluder may be used appropriately to close multiple defects.

P-318

The only venous access of transcatheter duct closure in children below 10 kilos.

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Introduction: The least invasive method of PDA closure is transcatheter method. However especially in the youngest group of patients (below 10 kg year of life) there is a significant probability of vascular complications, mainly arterial damage. The aim of study is preliminary presentation of our experience with PDA closure solely with venous access.
Materials and methods: In our center 98 ducts were closed with ADO in 2003-2012. In the last year 5 patients aged 9-14 months were enrolled in to interventional PDA closure with only venous access. The diameter of the ducts were 2,6-3,5 mm.From the venous access after location of the catheter in the way that part of the holes stayed in pulmonary artery and the second part in aorta angiography was performed. Later the proper sizes of implants were chosen basing on the measurements from that injection of contrast. The device was deployed in a standard manner but the procedure and final results were evaluated solely with echocardiography.
Results: Among all 5 patients enrolled into the PDA closure there were 4 patients who had PDA closed with ADO 6/4. There was one case of the complete duct constriction after introduction of PIG into the duct which resulted with inability of the precise PDA measurement and its morphology assessment. Because of that complication we had to add the arterial access and after the removal of the catheter from the duct there was an aortography performed. After 10 minutes the duct has dilated and then it was closed with PDA ADO occluder 6/4. The short and mid- term results are good – after 7 months of observation all patients are in good condition with PDA closed with echo assessment. In all the patients we observe complete normalization of the left chambers.
Conclusions: Transcatheter occlusion of the persistent arterial duct without an arterial access is the effective and safe method of treatment allowing to reduce the complications connected with the artery puncture. The possible complication of this method may be the constriction of the duct as the result of multiple maniof the catheters within the duct during the procedure.

P-319

Blalock-Taussig shunt angioplasty.

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Introduction: Blalock Taussig (BT) shunt occlusion or stenosis is uncommon but potentially life threatening and the current therapeutical options include surgical placement of another shunt or percutaneous angioplasty; only scarce data is currently available. The objective of this study was to evaluate results of BT shunt angioplasty.
Methods: Single centre retrospective study of the total number of patients that underwent catheterization with the intention of BT shunt angioplasty.
Results: Twelve procedures were performed in 11 patients between 1997 and 2011. Six patients were less than 24 months (median [range] 9.1 months [0.2 to 20.3]); the remaining had a median age of 15.7 years [10.2 to 30.1]. One patient was in critical condition before the procedure, the remaining were stable. BT shunts had been implanted in median 21.3 months [0.1 to 156.6] earlier; median diameter of 5 mm [3 to 5]. Five (42%) significant stenosis (with impaired contrast flow) and 7 occlusions (58%) were found. Heparin was administered and arterial approach was used in all patients. Balloon predilation followed by stent implantation (median diameter of 5 mm [4 to 5]) was used in 8 (77%) of procedures; plain balloon angioplasty (POBA) was performed in the remaining 4 cases. Normal contrast flow was obtained in 10 out of 12 shunts (83%). Three patients had a significant complication: one patient who had been operated 48h before the procedure had contrast extravasation (coil occlusion of the shunt was performed with success), another had pulmonary artery stenosis (later successfully managed by kissing-balloon technique) and a third patient died due to acute shunt thrombosis after balloon dilation. At a median follow-up of 60 months [3 to 248]: 7 out of 10 patients maintained shunt patency; one shunt restenosis occurred two years after POBA and underwent successful stent implantation; one stent thrombosis occurred three months after stenting and the patient was referred for new BT shunt surgery; one patient died two years later during the course of unplanned pregnancy.
Conclusions: BT shunt angioplasty is successful in the majority of patients, although complications are not uncommon. It is a valid alternative to reoperation, particularly in higher risk patients.

P-320

Perioperative assessment of patients with repaired tetralogy of Fallot undergoing pulmonary valve replacement

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Purpose: Pulmonary valve replacement (PVR) is commonly performed in patients with repaired Tetralogy of Fallot (TOF) to avoid late complications related to severe pulmonary regurgitation or residual RVOT obstruction. However, few data are available concerning perioperative complications. The aim of this study was to evaluate the perioperative complications and to determine predictive factors of the Low Cardiac Output Syndrome (LCOS) in patients undergoing PVR.
Methods and Results: 30 consecutive patients with TOF who underwent PVR between 2008 and 2009 were retrospectively enrolled. LCOS was defined according to arterial lactate level >3 mmol/l, inotropic support over 24 hours, or renal dysfunction. Mean age at PVR was 29.5 years. Surgical indications for PVR were RVOT stenosis (n = 4), severe pulmonary regurgitation (n = 25), and mixed lesion (n = 1). A stentless porcine aortic root (freestyle, Medtronic) was used in 26 patients (85%) with a range size from 21 to 29 mm.Carpentier-Edwards valve (n = 2), Hancock conduit (n = 1), and Contegra conduit (n = 1) were used for the other patients: PVR was conducted with beating heart using a normothermic cardiopulmonary bypass (CPB) (mean time 77 +/- 25 min) in 16 patients. Aortic cross clamp was achieved in 14 patients due to the need of additional surgery or surgeons' habit : CPB mean time was 113 +/- 21 min.Overall survival rate was 97% at 90 days. Postoperative complications were uncommon (Ventricular Tachycardia in 6%, Mechanical Ventilation over 24 hours in 6%, renal dysfunction in 10%) except for the LCOS (46%). CPB time over 80 min ($p < 0.01$) and aortic cross-clamp ($p = 0.03$) increased LCOS (OR = 33 (CI = 3.18-342.2, $p < 0.01$) and OR = 6 (CI = 1.15-31.3, $p < 0.01$), respectively. Surprisingly, age, ventricular volumes and function, and preoperative additional lesion (tricuspid regurgitation, residual pulmonary artery stenosis) were not predictive of perioperative complications.
Conclusion: these data underline the major role of myocardial protection during PVR in TOF patients. Short CPB time without aortic cross clamp decreases LCOS. Additional surgical repair requiring aortic cross-clamp and long CPB time should be well balanced with the perioperative complication risk, as tricuspid regurgitation can be improved with the reduction of the right ventricular volume after PVR, and pulmonary artery branch stenosis might be suitable for interventional catheterization procedure.

P-321
Infants with coarctation of the Aorta ± Hipoplastic aortic Arch: Median or lateral approach?

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Objectives: Surgical approach of infants with Coarctation ± hypoplastic aortic arch is a challenge. Choosing between Sternotomy or Thoracotomy is associated respectively with the need of extracorporeal circulation or not. We present our surgical results and follow-up.
Material & Methods: Retrospective analysis of 90 infants (age <1 year) with biventricular heart and surgical reconstruction of aortic arch during the period 2004-11: Sternotomy in 45 and Thoracotomy in 45. In Sternotomy group we use selective cerebral perfusion instead of circulatory arrest in order to eliminate the cerebral injury associated to the arrest. Cerebral monitorization is performed with invasive radial artery pressure and near infra-red spectroscopy. Statistical study with SPSS-15.0
Results: Preoperative characteristics of the groups are different: Sternotomy group has more associated cardiopathy, more hypoplastic arch, more dependence of intravenous prostaglandin, and more complexity in RASCHS-1 score. Referring to surgical techniques, Sternotomy patients received a termino-lateral anastomosis (78%), and Thoracotomy patients received a termino-terminal anastomosis (80%). Hospital mortality is 9% in both groups. Hospital morbidity analysis shows that there are no differences in neurological complications or early re-coarctation, but Sternotomy group presents more recurrent laryngeal nerve lesion and longer postoperative stay than Thoracotomies ($p < 0,05$). Mean follow up is 23 ± 19 months. One patient died in the follow-up (Thoracotomy group). Late re-coarctation is more frecuent in Thoracotomy (32%) than in Sternotomy (12%) (p:0,03), and is generally treated with percutaneous angioplasty. We reoperated 9% patients of the Sternotomy group and 15% of Thoracotomy group (pns).
Conclusions: The use of selective cerebral perfusion in median sternotomy approach of arch surgery lets the surgeon do the anastomosis in a bloodless field and preserves neurological function. Sternotomy in comparison with Thoracotomy has similar mortality, worse Hospital morbidity (recurrent nerve lesion, prolonged stay), but better patency of the arch anastomosis in the follow-up (low incidence of recoarctation). We recommend median sternotomy with selective cerebral perfusion if there is hypoplastic aortic arch and/or associated cardiopathy needing surgery.

P-323
Pulmonary artery branches stenosis in patients with congenital heart disease: the result of combined surgical and interventional approaches

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Introduction: In this study we sought to evaluate our recent experience with surgical plasty of pulmonary artery (PA) branches both for native and acquired stenosis.
Methods: We review data relative to the postoperative course of patients who underwent surgical PA plasty between January 2004 and October 2011. Primary outcomes included the need for further surgical procedures or interventional maneuvers on the PA branches for residual stenosis.
Results: Thirty-three patients were included in the study. Median age at surgery was 0.99 years (range 0.02-3.45 years), with a median weight of 6.1 kg (range 3.72-13 kg).
There were 12 native and 21 acquired PA stenosis. The location of the PA stenosis was mainly at the PA origin (n = 13, 38%) and at PA bifurcation (n = 11, 33%). The PA plasty was defined as simple in 13 patients (38%) and as complex in the remaining 20 patients (62%), including multiple maneuvers of the PA branches. Median Follow-up time after surgery was 4 years (range 0.17-7.75 years). One patient died 3 days after bidirectional cavo-pulmonary anastomosis and complex PA plasty for low output syndrome and one patient died 22 months after bidirectional cavo-pulmonary anastomosis, tricuspid valve plasty and simple PA plasty for congestive heart failure.
Nineteen patients (19/32 patients, 59%) underwent 36 hemodynamic interventions of the PA branches for residual stenosis. A stent implantation of the PA branches was deemed necessary in 6 patients (6/32 patients, 19%) who underwent previous unsuccessful post-operative balloon dilation. Three patients, who required postoperative

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interventions, underwent additional surgical maneuvers on the PA branches 6 months, 3.6 years and 4.4 years after initial PA surgical plasty. Twelve of the 19 patients (63%) who required postoperative balloon dilation have been operated before the age of 6 months; the majority of the PA stenosis were acquired (13/19, 68%) and underwent a complex plasty (12/19, 63%).
Conclusions: Pulmonary artery branches stenosis represent a life-threatening situation always jeopardizing patient's clinical status and outcome which often requires a prompt surgical or interventional resolution. A combined collaborative surgical followed by trans-catheter approaches is always needed particulary in cases of acquired PA stenosis who required complex surgical repair.

P-324
Long-term outcoume after repair of complete atrio-ventricular septal defect : A 30-year experience in a single institution

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Objectives: Atrio-ventricular septal defect (AVSD) can be repaired successfully during infancy. Cardiac event-free survival depends on the durability of left AV valve repair. In this retrospective study, we analyzed patients and operative characteristics as prognostic factors for long-term cardiac-related morbidity.
Methods: We identified 152 patients operated on between 1976 and 2008 for isolated complete AVSD. Eighty patients (54%) underwent surgical repair using a double-patch technique. From 1976 to 1994, deep hypothermia was routinely used whereas since 1995, moderate hypothermia/normothermia was used. Study endpoints were overall survival, freedom from left AV valve reoperation, freedom from MR >2, and freedom from any reoperation. Univariate Cox regression analysis was used to identify prognostic factors. Mean follow-up time was 83 months [0.2-356].
Results: Median age and weight at surgery were 6.3 months and 5.2 kg. Associated cardiac malformations (arch hypoplasia/coarctation, PDA) were diagnosed in 49 patients (32%). In-hospital mortality was 18.4% (28/152), 24.4% for children operated before 1995, 9.7% for children operated since 1995. Actuarial 15-yr survival was 76,2%. There were five early and nine late re-operations (six mitral re-repair, two LVOT and one RVOT obstructions), accounting for a reoperation-free survival of 90.2% at 15 years. At 15-years, freedom from MR greater than 2 was 82.6%. The use of deep hypothermia (HR 4.8), the presence of postoperative pulmonary hypertensive crisis (HR 2.1) and associated PDA (HR 2.1) were independent risk factors affecting survival. Residual MR greater than 2 also significantly impacted on survival (HR 4.2) and reoperation-free survival (HR 9.3). Repair before 6 months of age was predictive of left AV valve reoperation (HR 3.6).
Conclusions: This study confirms that AVSDs can be durably repaired in infancy whereas cardiac-related events are not unusual 15 years after repair. Most efforts should be pursued to optimize mitral repair at initial surgery. In addition, this study shows that most repair failures can be adequately addressed using valve-preserving strategies

P-325
An Expanded use of injectable pulmonic valve for Primary Repair

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Objective: Pulmonic valve insertion is needed for total correction of some type right ventricle outflow problems. Injectable pulmonic valve is designed for treatment of chronic pulmonic regurgitation in operated tetralogy of Fallot patients. We suggest that its advantages justify to expand its indication in primary correction of pathologies and we used this valve for primary repair in 3 patients.
Material and method: In 2 patients with pulmonary atresia and VSD and one patient with abscent pulmonary valve syndrome injectable pulmonic valve was used in primary repair. Their ages were 18 months, 6 and 11 years respectively. The patients with pulmonary atresia had pulmonary branch arterioplasty with patch concomittantly. In the patient with absent pulmonary valve syndrome injectable valve was inserted after annular enlargement with pericardial patch. At the end of the procedure all 3 patients had 0,5 right ventricle to aortic pressure ratio with direct measurement.
Results: All patient had uneventfull postoperative period and discharged from hospital at 2 nd week postoperatively. At discharge repeat echocardiography demonstrated that pulmonic valve function was normal without any gradient at valvular level.They were followed for 24,22 and 20 months and none of them had valve related problems during this period.
Conclusion: Injectable pulmonic valve is designed to have larger annular diameter than the actual pulmonary artery due to its expandable stent. In addition to that no react pulmonic valve has been proven to have advantages over other alternatives with its resistance to calcification, infection, degeneration and so, good longterm durability. We believe that using a biological valve instead of valved conduit can be good alternative to enlarge outflow tract liberally and to delay reoperation in the future.

P-326
Outcome and related parameters associated at maintenance of CI > 3 l/m/m2 following surgery for congenital heart disease

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Objective: Describe our experience in cardiac output monitoring by transpulmonay thermodilution in postoperative congenital heart disease and the relationship of the data obtained with the evolution of patients
Methods: Prospective analytical study in children. Measurements by TPTD (PiCCO^R) were made at 1, 4, 8, 16 and 24 hours after cardiac surgery in PICU and results were registered and hidden from medical team on charge of patients. Measurement data are expressed as mean$^+$SD. and patient data as median (range). Mann-Whitney test was used for comparison of patient's evolution.
Results: From 35 patients of 18 (3-144) months and 13 (3, 8-57) kg weigh, 17 maintained Cardiac Index (CI) > 3 l/m/m^2 in every control and had a better outcome with a shorter duration of

	CI > 3 18 patients 85 measurements	CI<3 17 patients 78 measurements	P
CI l/min m^2	4,23$^+$0,92	3,28$^+$0,82	0,000
SVI ml/m^2	35,14$^+$7,90	25,75$^+$7,93	0,000
GEDI ml/m^2	487$^+$167	421 $^+$176	0,016
ELWI ml/Kg	17,33$^+$9,76	21,09$^+$12,35	0,034
PVPI	3,36$^+$1,68	4,42 $^+$2,05	0,001
SVRI dyn*s*cm-5 *m^2	1353$^+$326	1623$^+$441	0,000
CPV mmHg	11,34$^+$3,12	13,72$^+$ 4,21	0,000
SvO_2%	78,14$^+$10,34	71,51$^+$10,45	0,000

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mechanical ventilation 12 (3-48) vs 25 (6- 432) hours ($p = 0.048$) and hospitalization in PICU 3 (2-7) vs 6 (2-34) ($p = 0.004$). Parameters obtained in TPTD measurements in both groups are in the table.
Conclusions: Maintenance of CI $>$ 3 l/min m^2 is related with a better outcome. We obtained a pattern of preload and afterload associated with CI maintenance. Classic parameters (CVP, SvO_2) held their value. Larger studies are needed to confirm these findings

P-327

Closure of Patent Ductus Arteriosus in the first year of life; does closure rate vary with clinician or patient factors?

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Introduction: We have had the impression that there are differences in numbers of infants referred for PDA closure from different areas of the East Midlands. We have therefore examined numbers of PDAs closed in the first year of life, as a proportion of other cases requiring intervention, by main postcode groups across the East Midlands. Potential reasons for any variation can therefore be explored.
Methods: Retrospective case note review. 6 Main post code areas identified; mainly (although not exactly) corresponding to catchment areas for referring obstetric/ neonatal centres. Procedures from 'non-main' postcode areas were excluded.
Results: Areas 1, 3 & 4 share the same level 3 neonatal service which does not have an on-site cardiac surgical unit. Area 2 has its own level 3 neonatal unit but also houses the cardiac surgical service. Area 5 looks to Area 2 for level 3 neonates and Area 6 has access to 2 different surgical units.

Post-Code Area	2009 PDA (total n)	2010 PDA (total n)	2011 PDA (total n)
1	1 (11)	1 (15)	3 (26)
2	13 (50)	9 (52)	12 (56)
3	1 (5)	0 (16)	0 (9)
4	3 (52)	5 (36)	1 (36)
5	2 (5)	2 (19)	5 (21)
6	1 (14)	0 (13)	0 (11)
Total	21 (69)	17 (151)	21 (159)

Conclusions: There are clear differences in referral rates for PDA closure across the East Midlands, which are not 'discounted' when the area are grouped by level 3 centre. It is not yet clear whether this relates to differences in demographics (eg area 2 has an very different ethnic mix and level of deprivation from the other areas) or to internal practice within the neonatal units themselves. If ease of access to surgical intervention is a barrier, this should also be explored, along with any potential effects on outcome.

P-328

Frequency of Complications and Outcomes Following Fontan Operation

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Introduction: Fontan operation is the ultimate palliation for CHDs which are not amenable to total correction. This retrospective study investigates the medium and long term complications and outcome of Fontan palliation.
Methods: 90 patients, who underwent Fontan operation between 1987 and 2010 in South Wales, were included in the study.
Results: 52 patients had extra cardiac repair. 46% had fenestration in Fontan circuit. Mean age at operation was 6.1 years (range 2 to 33). Mean follow-up was 7.7 years (range 0 to 26 years). There were a total of 7 deaths with 3 within 30 days of operation, one from PLE, one from arrhythmia, and 2 sudden deaths. The 1-, 5-, 10-, 15, 20 and 25-year survival was 94.9%, 94.9%, 92.5%, 92.5%, 79.3% and 79.3% respectively. In the medium to long term follow-up, 26% had arrhythmias; mostly occurring in the atriopulmonary group 73% versus 13% in the extracardiac group. 7 patients required permanent pacemaker. 29% had congestive cardiac failure, 7.8% developed protein losing enteropathy (PLE) and 2% developed plastic bronchitis. Two patients with plastic bronchitis made full recovery with PDE5 inhibitor in addition to removal of casts. However only 2/5 patients with PLE responded to medical and catheter/surgical treatment. PLE and plastic bronchitis more often occurred in non-fenestrated group. Congestive cardiac failure occurred more often in the morphological right ventricle group; 73% in the right ventricle group compared to 38% in the left ventricle group were on antifailure medication. There were no thromboembolic complications in any patients who have been on Warfarin.
Conclusions: Arrhythmias and cardiac failure are the most common long-term complications following Fontan procedure. PLE and plastic bronchitis constitute the most challenging clinical problems. Warfarin seems to be effective in preventing thromboembolic events. With growing population of Fontan survivors, efforts to improve Fontan dynamics by supporting cardiac function, or reducing pulmonary vascular resistance should be intensified to minimize the high morbidity and mortality from these complications in the long term.

P-329

Renal near infrared spectroscopy as realtime indicator for developing acute kidney injury in infants undergoing cardiac surgery with cardiopulmonary bypass

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Introduction: Acute kidney injury (AKI) is a frequent complication after cardiac surgery with cardiopulmonary bypass in infants. There is currently no standard method to monitor kidney function for early detection of AKI. An improved diagnostic algorithm would possibly allow to develop a strategy for prevention of AKI. Renal NIRS (near infrared spectroscopy) evaluates regional oximetry in a non-invasive continuous realtime fashion and reflects tissue perfusion. The aim of this study was to evaluate the relation between renal oximetry and development of AKI in the operative and postoperative setting in infants undergoing cardiopulmonary surgery.
Methods: In this prospective study we enrolled 59 infants (age median 88 ± 82 days, weight median 4316 ± 1622 g) undergoing

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cardiopulmonary bypass surgery for congenital heart disease (n = 37 cyanotic and n = 22 non-cyanotic) for uni- or biventricular repair. Renal NIRS was continuously measured intraoperatively and at least 24 hours postoperatively. The renal oximetry values were correlated with the pediatric Renal-Injury-Failure-Loss-End (pRIFLE) classification for AKI, additional kidney function parameters and the postoperative course.
Results: 27 (46%) infants developed AKI based on pRIFLE classification. No difference could be shown in the intraoperative renal oximetry of infants with or without AKI. However the NIRS demonstrated significantly lower renal oximetry periods in the first 24 and 48 hours postoperatively in AKI patients as compared to patients with normal renal function ($p < 0.05$). 3 (11%) of infants with AKI needed a renal replacement therapy and 2 (7%) died. In the non AKI group no fatal course was reported during the study period. The infants with decreased renal oximetry values developed significantly higher lactate levels 12 and 24 hours after the operation.
Conclusion: Our results suggest that prolonged lower renal oximetry values indicate impaired renal function and decreased systemic oxygen delivery followed by AKI. Renal NIRS might be a promising non-invasive method for the early detection of AKI in infants undergoing cardiac surgery with cardiopulmonary bypass.

P-330
Long-term effects of residual right ventricular (RV) outflow tract obstruction on RV dimension and function in patients after repair of tetralogy of Fallot

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Objectives: Preservation of the pulmonary valve, even at the expense of a mild residual stenosis, is the current surgical policy in the management of patients with tetralogy of Fallot (TOF). The purpose of this study was to assess the long-term effect of a residual right ventricular outflow tract obstruction (RVOTO) on RV dimension and function.
Methods: From April 2007 to December 2011, fifty-three children (mean age 13.4 ± 6.4 years) after repair of TOF (mean age at surgery 1.3 ± 1.3 years) were prospectively assessed by cardiovascular magnetic resonance imaging (3-T MR scanner, Siemens, Erlangen, Germany). Residual RVOTO on echocardiography was defined as a peak systolic RVOT gradient ⩾ 25 mmHg.
Results: Patients with RVOTO (n = 29; mean gradient 39.5 ± 11.9 mmHg) had significant less pulmonary regurgitation (PR) (25.2 ± 10.6% vs. 30.8 ± 9.3%; $p < 0.05$) compared to patients without RVOTO (n = 24; mean gradient 15.4 ± 7.0 mmHg). Children with RVOTO had significant smaller RV enddiastolic (94.0 ± 2.6 ml/m2 vs. 104.0 ± 20.7 ml/m2; $p < 0.05$) and endsystolic (42.9 ± 20.0 ml/m2 vs. 48.9 ± 13.2 ml/m2; $p < 0.05$) volumes compared to patients without RVOTO, while RV ejection fraction did not differ between the two groups (EF 55.5 ± 8.4% vs. 54.0 ± 6.6%; p = n.s.). Restrictive physiology, assessed by late diastolic forward flow in the main pulmonary artery, was equally distributed within both groups (31% vs. 25%; p = n.s.).
Conclusions: According to our data, residual RVOTO after repair of TOF does not affect RV function, while RV dimensions and the degree of PR are more favourable in the long-term follow-up of those patients. These results confirm the beneficial effects of the current strategy in repair of TOF.

P-331
ECMO as a Bridge to Recovery in Patients with Intractable Arrhythmias

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Introduction: Pharmacological treatment and cardioversion are the mainstay of antiarrhythmic treatment but few arrhythmias remain refractory to maximal conventional treatment.
ECMO facilitates myocardial recovery by establishing a period of haemodynamic stability, during which antiarrhythmic medication can be optimized. However in rare cases the patient can be stabilized on VA-ECMO and then taken to catheter lab for intra cardiac ablation.
The benefits have to be balanced against potential ECMO related complications.
Methods: Review of all paediatric patients cannulated for ECMO during the last 7 years at the Heart Link ECMO Centre with the primary diagnosis of an intractable arrhythmia. Patients with arrhythmias secondary to an other underlying cardiac condition such as cardiomyopathy, myocarditis or post cardiac surgery were excluded.
Results: Seven patients were cannulated for VA-ECMO for intractable arrhythmias (5 male), Median age 4.5 months (13 days – 14 months), Median weight (3.4-12.9 kg). Four had SVT (supraventricular tachycardia) and 3 had VT (ventricular tachycardia). Median maximal HR 272 bpm (220-350 bpm). Six out out seven patients required cardio pulmonary resuscitation prior to ECMO cannulation. Median hours on ECMO 108 hours (53-195hrs). Median ECMO flow 4 hours post cannulation 93 ml/kg/h (63-148 ml/kg/h). Three patients were taken to the catheter lab on ECMO and underwent successful radiofrequency ablation. One patient underwent balloon atrial septostomy on ECMO to off load the left heart. All patients came off ECMO in Sinus Rhythm/Junctional Rhythm. There were no relevant ECMO related complications. On follow up three months post de-cannulation all patients had normal systolic function parameters and normal chamber sizes on echocardiography.
Conclusion: VA-ECMO in patients with intractable arrhythmias provides sufficient cardiac output until adequate levels of antiarrhythmic drugs are achieved and allows to wean inotropic support which is potentially pro arrhythmic. VA-ECMO can be used to stabilize a patient to perform curative intracardiac ablation.
The prognosis and recovery of cardiac function for patients who went onto VA-ECMO for arrhythmias is excellent once control of the underlying arrhythmia is achieved.In this patient group we had no relevant ECMO related complication.

P-332
Total cavopulmonary connection in patients with the Down syndrome

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Objectives: Total cavopulmonary connection (TCPC) operation is rarely performed for a functional single ventricle in a child with Down syndrome. Therefore, the postsurgical outcomes are not well known. Patients with Down syndrome are at a risk of

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developing persistent pulmonary hypertension and airway obstruction, which may affect the outcome of univentricular repair. We evaluated mortality and related factors after TCPC in Down syndrome.
Methods: Between January 2004 and March 2010, we identified 8 patients with Down syndrome among 235 patients who had undergone TCPC. The course before TCPC, preoperative data, and postoperative course were evaluated. In addition, clinical parameters and the course after TCPC were compared between Down syndrome (N = 8, 6 boys and 2 girls) and Non-Down syndrome (N = 227) groups.
Results: The median age at the time of TCPC was 4.1 years (3.4–5.5 years). The preoperative mean pulmonary artery pressure was 13.9 ± 1.81 mmHg. We observed respiratory complications in 2 patients, surgical site infection in 3 patients, and chylothorax in 2 patients. No significant difference was observed between the Down and Non-Down syndrome groups in preoperative data and mortality rate: Down syndrome group, 1 of 8 patients (12.5%); Non-Down syndrome group, 5 of 227 patients (2.2%). However, when the clinical course after TCPC was examined, the duration of mechanical ventilation (P = 0.039), length of ICU stay (P = 0.009), duration of pleural drainage (P = 0.027), and length of hospital stay (P = 0.007) were found to be significantly prolonged in the Down syndrome group.
Conclusions: Patients with Down syndrome tended to show prolonged recovery after TCPC, and suspected etiologies include respiratory tract disease, prolonged chylothorax, and surgical site infection.

P-333
Inhaled Nitric Oxide after Superior Cavopulmonary Anastomosis – Effects on Cavopulmonary Pressure and Oxygen Saturation

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Introduction: Possible clinical problems in patients early after superior cavopulmonary anastomosis include low arterial oxygen saturation and/or high cavopulmonary pressure, which results in upper body congestion. Mild hypercapnea in order to lower cerebral vascular resistance was proposed for managing hypoxemia in such patients. The role of selective pulmonary vasodilation after superior cavopulmonary anastomosis remains unclear. This study aims to investigate the hemodynamic effects of inhaled nitric oxide (iNO) in children early after superior cavopulmonary anastomosis.
Methods: The medical files of 18 patients treated with iNO early after superior cavopulmonary anastomosis were retrospectively studied. The cavopulmonary pressure and the arterial oxygen saturation were serially measured before iNO treatment and in the first 24 hours after initiating it. Their changes were investigated with the paired samples T-test. A potential correlation between the arterial oxygen saturation and the arterial partial CO2 pressure (pCO2) was also searched. A value of $p \leq 0.05$ was considered significant.
Results: The age of the patients ranged from 3 months to 2.5 years, median – 9 months. Median dose of iNO was 20ppm (range 10-20ppm). Initial cavopulmonary pressure was 17.5 ± 2.96 mm Hg and it changed to: 16.9 ± 3.1 mm Hg at the first hour (p = 0.35); 16.2 ± 2.6 mm Hg at the 6th hour (p = 0.045); 15.8 ± 2.8 mm Hg at the 12th hour (p = 0.008) and 14.9 ± 2.5 mm Hg at the 24th hour (p = 0.010). Initial arterial oxygen saturation was 78.2 ± 10.1% and it changed to: 79.2 ± 10.4% at the first hour (p = 0.255); 80.1 ± 9.4% at the 6th hour (p = 0.051); 81.3 ± 9.2% at the 12th hour (p = 0.068) and 80.5 ± 9.7% at the 24th hour (p = 0.126). The pCO2 did not change significantly during the 24 hours of treatment and there was an insignificant slight negative correlation between pCO2 and arterial oxygen saturation (Pearson coefficient = -0.21, p = 0.054).
Conclusions: Patients after superior cavopulmonary anastomoses with cavopulmonary pressures above 15 mm Hg and/or arterial oxygen saturation below 80% may benefit from iNO inhalation. We observed significant reduction of the cavopulmonary pressure for 24 hours after initiating the treatment. There was also a tendency for elevation of the arterial oxygen saturation, which was unrelated to changes in pCO2.

P-334
LATE ARTERIAL SWITCH FOR TRANSPOSITION OF THE GREAT ARTERIES

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Objectives: Evaluation of pulmonary artery banding with adaptable pulmonary artery band (PAB) versus conventional PAB in patients requiring a late arterial switch operation for transposition of the great arteries (TGA)

Background: In late diagnosis of TGA the LV involutes as it pumps against low resistance. The LV needs retraining in preparation for an arterial switch. We report our results with different techniques.
Methods: 10 patients underwent retraining of the LV, 6 pts had a traditional PAB placed (group 1) with additional creation of an ASD (5), aortopulmonary shunt placement (3) and PDA ligation (1). 4 pts had a telemetrically adaptable PAB placed without associated procedures (group 2).
Results: Mean weight and age at PAB was 5.8 kg and 7.4 mths for group 1, 5.8 kg and 11.7 mths for group 2. Time between palliation and switch was 4.2 mths in both groups.
Group 1 showed an increase in banding gradient from 49 to 68 mmHg at the time of the switch. 4 pts required reoperations before switch, 2 of these had 1 re-operation and 2 had 2 re-operations. 2 pts died, 1 after PAB and 1 after switch. Group 2 showed an initial banding gradient of 26 mmHg at 5% closing of the PAB. Progressive closure of the PAB led to a mean gradient of 64 mmHg at the time of switch. There were no re-interventions or deaths.
Conclusions: Retraining of the LV by the adaptable PAB allows precise control of the tightening, avoids repetitive surgery and minimises morbidity.

P-335
Inferior mini-sternotomy provides excellent cosmetic outcomes in children with congenital heart disease.

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Introduction and objectives: Minimally invasive surgery (MIS) has been previously reported to improve cosmetic outcomes in children with congenital heart disease (CHD). However, the scar has been rarely objectively assessed. We report evaluation of cosmetic outcomes in children with CHD undergoing cardiac surgery through inferior mini-sternotomy.
Methods: From January through December 2010, we operated on 15 children (female = 8) with septal defects –ASD (n = 13),

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partial AV canal (n = 1) and VSD (n = 1)- through inferior mini-sternotomy. Mean age at surgery was 60 months (range from 8 to 151 months). Follow-up data collection was carried out and validated by two different observers at least 6 months after surgery. Objective and subjective evaluation of the cosmetic results were performed with The Vancouver Scar Scale and The Patient Scar Assessment Scale (PSAS), respectively.
Results: All patients underwent surgery with no early or late complications. Two patients were lost to follow-up. For a mean follow-up was 17 months (range from 8 to 24 months), the mean dimensions of the scar was 6.7 ± 1.9 cm.The objective score in the Vancouver were ≤ 2 in 11 cases (near the normal skin). With respect to PSAS evaluation, parents qualified cosmetic outcomes as 8/10 (being 10, maximum satisfaction possible).
Conclusions: MIS through inferior mini-sternotomy provides excellent cosmetic outcomes in children with CHD. This approach could be considered of choice in children with septal defects.

P-336
Paediatric cardio-pulmonary assist via central ECMO with an integrated left ventricular vent
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Introduction: For children with low cardiac output, veno-arterial (VA) extracorporeal membrane oxygenation (ECMO) remains the mainstay of mechanical circulatory support. In experimental study using canine heart failure model, additional decompression of the left ventricle (LV) advances myocardial recovery. In spite of this experimental result, the utilization of an integrated LV vent on ECMO is not a common clinical practice. In this study, we reviewed our experience in temporary circulatory support via a central ECMO with integrated LV vent in children with cardiac failure refractory to medical management.

Case Nr.	Age	Diagnosis
1	12 y	Dilatative Cardiomyopathy
2	13 y	Acute myocarditis
3	1 mo	Pulmonary Artery Atresia with MAPCAs
4	4 mo	Bland-White-Garland-Syndrom
5	3 mo	Complete Atrioventricular Septal Defect Typ C

Performed surgery	ECMO duration (d)	Short term outcome
ECMO impl.	10	Berlin Heart Excor
ECMO impl.	6	Discharged home
Unifocalisation and ECMO impl.	5	Discharged home
Left coronary artery trans-location and ECMO impl.	5	Discharged home
cAVSD repair and ECMO impl.	>1	still on ECMO

Methods and Results: In year 2011 and 2012, five children acquired temporary circulatory support via ECMO with integrated LV vent. All cases are approached through median sternotomy. Arterial cannula was placed in ascending aorta. The right atrium was cannulated for venous return in all cases except in case 4, in which the bicaval cannulation was used. LV vent was inserted through right superior pulmonary vein and was connected to the venous line on ECMO, so that complete left heart decompression was achieved.
Conclusions: ECMO represents a feasible and effective method to support critically ill patients with existed low cardiac output. Cautious selection of patients, accurate timing of ECMO implantation as well as postoperative management remain challenging. In the presence of low cardiac output and insufficient interatrial shunt, additional LV decompression via LV vent could avoid the left heart distension and might promote myocardial recovery. It helps to prevent pulmonary congestion and the associated pulmonary hemorrhage. According our experience, we recommend the use of ECMO with an integrated LV vent in children with intractable cardiac failure.

P-337
Biventricular correction for complex congenital heart defects associated with aortic arch anomalies after initial Norwood-type operation. Case series report.
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Introduction: Aortic arch anomalies are associated with the various type of heart defects ranging from ventricular septal defect to the left heart hypoplasia. Decision about single- or two-stage biventricular repair in children with good left ventricle depends on child's weight, type of associated heart defect and local conditions during surgery.
THE AIM OF THIS STUDY was to present series of four cases with complex heart defects associated with interrupted or hypoplastic aortic arch and aortic valve atresia or hypoplasia, in whom anatomical conditions did not allow for singe stage biventricular correction and the two stage repair with initial Norwood procedure was performed.
Case reports: Two patients has a hypolpastic aortic arch associated with aortic valve atresia and ventricular septal defect, one has interrupted aortic arch with ventricular septal defect and one has interrupted aortic arch with atrial and ventricular septal defects. All patients had initial Norwood type operations with right ventricle to pulmonary artery shunts and various modifications depending on heart anatomy. One child required angioplasty of RV-PA shunt with stent implantation 10 months after initial operation, in one case B-T shunt was performed 19 months after Norwood procedure. Biventricular correction was performed in all four cases an average of 18,25 months after Norwood procedure. In two of the cases pulmonary homograft, in one contegra conduit and in one matrix valved graft was used for right ventricle outflow tract reconstruction. All children are in good condition with laminar flow through the neoaorta and pulmonary graft and after average 20(SD ± 6,5) months of follow up with no need of reinterventions and reoperations. One child require pharmacotherapy (Salbutamol) because of advance atrioventricular block (II/III degree).
Conclusions: Double-stage biventricular repair in cases of interrupted or hypoplastic aortic arch associated with aortic valve atresia or hypoplasia and ventricular septal defect, with

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initial Norwood-type operation ensures good postoperative effect and is an interesting alternative for single-stage correction in case of significant left ventricle outflow tract obstruction and low body weight neonates with poor local condition unsuitable for arch reconstruction.

P-338

Unusual case of a traumatic VSD and ventricular aneurysm in a child secondary to a blunt chest trauma.

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Introduction: Cardiac injury from a blunt chest trauma is a very rare complication during childhood and can range from a simple contusion to myocardial rupture. Patients can present with symptoms immediatly thereafter or not be diagnosed till several years later.
Case report: Secondary to a fall from a horse a 12 year old girl was injured on the left side of her chest by the animal resulting in a brief loss of consciousness. After being transported by EMS to the closest ER where she arrived in shock and was stabilized with several bolus of volume; PRBCs and FFP. An initial CT revealed a grade 4 liver laceration. On the transport to Sickkids she had short runs of ventricular tachycardia which were successful treated with Amiodarone. Due to profound hypotension she required further volume boluses and was later started on Norepinephrine. Additionally, a repeated CT showed a lung contusion and an echocardiography revealed a moderate sized muscular apical VSD and a left ventricular aneurysm. As a result of further respiratory deterioration, she was intubated. Troponin was significantly elevated (almost 3700 ng/L) and an ECG showed ST-elevation in the inferior leads. In the following days she recovered and the lung and liver injury healed. Despite her intracardiac left-to-right-shunt, due to her newly aquired VSD, she did not develop any related symptoms. The ventricular function remained good with only dyskinesia of the apex. On the day 19 after her accident she underwent a successful surgical repair and was discharged 3 days later.
Conclusion: Cardiac trauma, especially a ventricular aneurysm can be challenging to manage in the presence of liver injury with respect to anticoagulation. An early diagnosis and subsequent adequate management are important factors to minimize complications and to obtain a good outcome.

P-339

Custom made external aortic root support, an alternative for aortic root replacement in patients with Marfan

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Introduction: Classical repair for aortic root dilatation in patients with Marfan disease involves major surgery. Anticoagulation is needed when the valve is replaced and if the valve is preserved a substantial risk of valve failure exists.
Methods: We report on two successful implants with a custom made external aortic root (AR) support device (Exstent, Ltd) thereby avoiding these substantial disadvantages.
Results: Our first patient is a 19 year old girl with Marfan's syndrome. Her AR dimensions on echo-cardiography increased from 31 mm in 2000 to 45 mm in 2010 (CT: 42 mm). In our second Marfan patient (male, 22 yrs), echocardiographic AR dimensions increased from 35 mm in 2006 to 40 mm in 2010 (CT: 45 mm). By computer-aided design, a model of the individual patient's aorta was created from cardiac CT images and an external AR support device was manufactured.
Both patients were operated in March 2011 using a midline sternotomy. The AR was dissected towards the annulus, thereby freeing the ostia of the coronaries and this without using cardiopulmonary bypass. The custom made woven graft was placed around the AR and secured. Both procedures were finished within two hours with minimal blood loss. Patients were discharged after 7 days and 8 days respectively. A control MRI after six months revealed unchanged dimensions of the AR (42 mm and 46 mm respectively).
Conclusion: External AR support is a possible alternative for AR replacement in Marfan patients. The procedure involves a sternotomy but avoids the use of cardio-pulmonary bypass. The aortic valve and the endothelium remain intact thereby avoiding the need for anticoagulation therapy. MRI after 6 months revealed unchanged dimensions of the AR.

P-340

Correlation between echocardiographic estimation of the left atrial pressure and its invasive measurement in paediatric patients following cardiac surgery

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Introduction: Correlation between hemodynamic capillary pulmonary pressure and echocardiography E/E' ratio has been widely reported in adults. Our main objective was to assess whether there was a correlation between direct left atrial pressure measurement (by catheter) and the echocardiographic E/E' ratio in children with congenital heart disease after cardiac surgery. It is worth to highlight that there are no medical studies where such a correlation has been investigated.
Methods: Forty-eight echocardiographic postoperative studies in 27 patients were included, prospectively, between March of 2009 and March of 2011 incorporating variables such as thorax condition (open or closed), mechanical ventilation, nitric oxide or inotropic support, which we thought could modify our results. The left atrial catheter was introduced through a pulmonary vein when finishing the operation. For statistical analysis we used the SPSS (Statistical Package for the Social Sciences, Version 15.0) program.
Results: There were 14 male and 13 female, median age 2.7 years old. Transposition of great arteries (7/27), Tetralogy of Fallot (5/27) and atrio-ventricular septal defect (3/27) were the most frequent congenital heart diseases. Overall, the medium left atrial pressure and E/E' ratio were 12,0 and 10,6 mmHg respectively, with a positive correlation of +0,37. However when selecting only those studies were performed 72h after surgical intervention (n = 16), the correlation increased to +0,86. We hypothesize that during the first three days left atrial pressure is high likely because there is systo-diastolic dysfunction (related to the surgical aggression, myocardial ischemia and extracorporeal circulation), positive fluid balance due to post-bypass inflammatory syndrome, and vasopressor support. In contrast, E/E' ratio was lower during the first days due to low cardiac output and generates slow blood flow, and hence, smaller E waves. After 72h, systolic and diastolic functions improve, cardiac output increases, less vasoactive support is required, and negative fluid balance is generally attained. All these facts make possible a fall in left atrial pressure and an E wave rise.

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Conclusion: Therefore, it is our contention that three days after heart surgery, echocardiographic E/E' ratio is a useful tool to estimate left atrial pressure, which could assist in making more appropriate therapeutical decisions.

P-341

Expanding treatment options for the patent arterial duct (PDA)–Amplatzer Ductal Occluder-II Additional Sizes (ADO-II AS) Early clinical experience

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Background: Since 1998, the Amplatzer generation of duct occluders have been used with very good success. The latest addition to the ADO family—the ADO II AS possesses a lobe and two discs which extrude only 1 mm to 1.5 mm beyond the lobe delivered through a 4Fr catheter. There are 9 devices—3, 4, 5 mm waist sizes and 2, 4, 6 mm device lengths. The ADO-II AS can be used to close ducts in infants under 6 kg.
Objectives: To evaluate the feasibility, clinical efficacy and duct morphology for the new ADO-II-AS. procedure approach, complications and duct closure success.
Methods: Prospective single centre study from January 2011 to date. 20 patients were electively selected for implantation of ADO-II AS 15 patients had volume overload with heart failure in 5.

	RANGE	MEDIAN
AGE -months	2-89	10.5
WEIGHT-kg	2-19	10
PDA NARROWEST POINT- mm	1.5-4	2.5
PDA LENGTH-mm	3.5-6.6	5.5
PDA TYPE	A-C	A

Results: 17 devices were successfully implanted and all by arterial delivery. Of the other 3, 1 had surgical ligation and 2 had ADO devices. There were 2 venous attempts which were unsuccessful; 1 was delivered arterially and the other had an ADO. Follow up echocardiography within 24 hours post implantation and at 6 weeks showed no residual shunts in all 17 cases.
Complications: There was 1 acute but no late embolization.
Conclusions: ADO-II AS is a welcome added device for PDA closure, it can be delivered by the arterial or venous route; more experience is required to identify the types of ducts that will benefit most from its use. However the device lends itself to use in smaller infants.

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