

Cardiology in the Young

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ISSN 1047-9511

Abstracts of the 22nd Annual Meeting of the Mid-West Pediatric Cardiology Society, Indianapolis, Indiana, USA, 24–25 September 1998

ELECTROPHYSIOLOGY

Physiologic stressors inducing sudden death in the very-long-chain ACYL-CoA dehydrogenase deficient mice

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Objective: To investigate the hypothesis that physiologic stresses such as fasting and other environmental stressors may precipitate cardiomyopathy and sudden death in VLCAD deficiency.

Background: Mitochondrial fatty acid β -oxidation is the major source of energy of the heart, and VLCAD catalyzes the first step in the β -oxidation pathway. The most common phenotypes associated with human VLCAD deficiency are cardiomyopathies, metabolic crisis, and sudden death.

Method: To test our hypothesis, we generated VLCAD knockout mice that could be experimentally manipulated. These mice underwent different physiologic stressors to assess survival and the presence of cardiac dysfunction. Homozygous deleted and wild type animals, males, and females were randomized to 4 different groups: no intervention, fasting alone, cold room alone, and fasting in the cold room. Environmental stress was created by placing these animals in the cold room at 4°C for 6 and 12 hours. Blood glucose, rectal temperature, plasma and urine samples for metabolite analysis were obtained at scheduled intervals. Cardiac echocardiograms were also obtained. Tissues were studied by histologic assessment.

Results: The VLCAD knockout mice survive birth. The adult homozygous deleted mice are viable and asymptomatic. When fasted at 28 days of age, 50% of the VLCAD-deficient males die in the first 24 to 36 hours. When fasted adult males and females are placed in the cold room, the VLCAD homozygous mice males and females die within 12 hours. The wild type animals males and females survive these stresses. The mean blood glucose for the dying animals decreased sharply from 80 to 20 mg/dl, simultaneously their rectal temperature decreased from 36°C to 20°C. The echocardiograms of all the animals show good contractility and normal LV mass, dimension and thickness, although severe bradycardia when hypoglycemic and hypothermic. The histology revealed accumulation of fat in the cardiomyocytes and hepatocytes of the dead animals. **Conclusion:** Physiologic stresses induce metabolic crisis and sudden death in the VLCAD knockout mouse model.

Specific induction of p38 kinase activity in cardiac volume overload following aorto-caval shunt in rats.

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Earlier studies have shown rapid activation of mitogen activated protein (MAP) kinase family such as c-Jun NH₂-terminal kinase (JNK) and

extracellular signal regulated kinase (ERK) during the mechanical stress associated with pressure hypertrophy (PO), but not of p38. These kinases are regulated by different upstream MAP Kinase Kinase signaling cascade, and regulate cell growth through phosphorylation of specific transcription factors. The activity of MAP kinases in volume overload hypertrophy (VO) has not been characterized. Cardiac VO was induced in 30 adult Sprague-Dawley rats by aorto-caval shunt. Sham operated animals (n=10) served as controls. Heart/body wt ratio was higher in VO than in shams one week post-surgery ($46 \pm 3\%$ $p < .05$). Time-course of JNK, ERK and p38 kinase activities was analyzed after 2, 5, 10, 15, 30 min and 2, 4, 24 and 48 hr of VO using *in vitro* kinase assay with appropriate substrates. Results showed rapid induction of p38 kinase activity within 2 min which became significantly higher at 5 to 30 min with a peak at 10 min ($465 \pm 50\%$ increase, $p < .05$) and return to basal levels in 2 hr following the shunt. However, activity of the other two kinases remained unchanged. To characterize the phenotype of VO heart, steady-state levels of α - and β -myosin heavy chain (MHC) mRNA were analyzed after 24 and 48 hrs of VO. Results showed decrease ($60 \pm 5\%$, $p < .005$) in α -MHC mRNA at 24 and 48 hrs but no increase in β -MHC mRNA. These data differentiate VO phenotype from that reported for PO at these time points (i.e., β -MHC and no changes in α -MHC mRNA). Our findings are the first to implicate the role of p38 kinase in the early stages of VO and reveal decreased α -MHC mRNA in this form of hypertrophy. The molecular phenotypic differences in VO and PO may account for the disparate cardiac morphological (mass/volume relations) and functional properties observed in these two condition.

Large left to right shunt and congestive heart failure increase total but not resting energy expenditure

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Background: Patients with large left to right shunt have slow growth. We hypothesized that reduced growth with large left to right shunt and congestive heart failure (CHF) is due to increased resting and total energy expenditure.

Methods: Four month old infants with isolated ventricular septal defect (VSD) as a model of pure left:right shunt were compared to healthy controls (CTL). Qp:Qs was quantified by cardiac catheterization or echocardiography. Indirect calorimetry was used to measure resting energy expenditure (REE); doubly labeled water method was used to measure total energy expenditure (TEE), energy intake (EI) and total body water (TBW). (Table)

Conclusions: Large Qp:Qs and CHF increase TEE but not REE in VSD model of left to right shunt. The difference between TEE and REE reflects the energy cost of activity. This, rather than increased REE or decreased energy intake, leads to reduced growth. Estimate of energy needs to optimize intake or time of intervention should be based on TEE rather than REE.

Table 1.

| Results: TEE increased with shunt size. REE did not change (all values mean \pm SD, * $p \leq 0.05$, ** $p \leq 0.01$, † $p < 0.005$) | | | | | | |
|---|-----|-----------------|-----------------|-----------------|------------------|------------------|
| Group | No. | Weight (kg) | REE(kcal/kg/d) | EI (kcal/kg/d) | TEE (kcal/kg/d) | TBW (%) |
| CTL | 10 | 6.8 \pm 1.4† | 43.9 \pm 14.1 | 87.0 \pm 11.8 | 61.9 \pm 10.2† | 59.7 \pm 6.4** |
| VSD | 15 | 5.1 \pm 0.9† | 46.2 \pm 10.8 | 89.1 \pm 17.5 | 88.3 \pm 22.6† | 68.6 \pm 9.0** |
| NonCHF | 7 | 5.6 \pm 0.6** | 44.3 \pm 7.5 | 82.3 \pm 12.2 | 76.4 \pm 18.3 | 63.5 \pm 8.8* |
| CHF | 8 | 4.7 \pm 1.0** | 47.8 \pm 13.4 | 95.1 \pm 19.7 | 98.7 \pm 21.7 | 73.1 \pm 7.0* |
| Qp: Qs<2.5 | 9 | 5.4 \pm 1.0 | 49.5 \pm 11.2 | 79.4 \pm 9.3† | 73.9 \pm 18.8† | 66.3 \pm 9.9 |
| Qp: Qs \leq 2.5 | 6 | 4.8 \pm 0.7 | 41.2 \pm 8.9 | 103.7 \pm 17† | 106.8 \pm 14† | 72.1 \pm 6.9 |

Arterial oxygen saturation (SaO₂) does not accurately predict adequacy of systemic oxygen delivery in neonates following norwood palliation of hypoplastic left heart syndrome

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Introduction: In Norwood patients a SaO₂ of 70–80% has traditionally been thought to indicate balanced circulation ($0.5 < Q_p/Q_s < 2$), assuming a normal arteriovenous oxygen saturation difference of ~25%. The study goals were to develop a mathematical model of the circulation post Norwood I repair to test the adequacy of using SaO₂ to predict systemic oxygen delivery and, to provide clinical data to show the value of continuous SvO₂ monitoring (1,2).

Methods: We used a mathematical model (see figure below) derived from the Fick principle of $SvO_2 = SaO_2 - [(Q_p/Q_s) * (SaO_2 - 1)]$ assuming fully saturated pulmonary venous blood. Clinical data was obtained from the first 24 hrs. post-procedure. SvO₂ data were obtained via a 4 Fr. optical catheter placed in the SVC during surgery. Circulatory parameters and mechanical ventilation (including intentional hypercapnea) were manipulated to maintain adequate systemic oxygen delivery, defined as a SvO₂ >50%, and an arteriovenous oxygen difference (AVDO₂) of <5 vol. %. MAP, SaO₂, SvO₂, FiO₂, EtCO₂, and CVP were recorded with an PC-based data acquisition system.

Results: The mathematical model shows that SaO₂ in the target range of 70–80% can be achieved both by circulatory balance and by pulmonary over circulation and low SvO₂. Adequate systemic oxygen delivery depends upon both SaO₂ and Q_p/Q_s, which cannot be identified by SaO₂ alone. Data from continuous recordings in seven consecutive neonates revealed in the first 24 hrs postop, 66 episodes of SvO₂ <50% only 26 of which were identified by out of range SaO₂, for a 40% sensitivity, 75% specificity, and 61% total predictive value of the traditional method.

Conclusions: SaO₂ is a poor predictor of adequate circulatory balance in postoperative Norwood patients. Continuous SvO₂ monitoring identifies low systemic oxygen delivery before traditional techniques.

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High-intensity focused ultrasound effect on cardiac tissues: potential for clinical application

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High-intensity focused ultrasound (HIFU) is an evolving technology with the potential for a wide variety of therapeutic applications. Functioning in the frequency range of 500 kHz to 10 MHz, it can be focused to cause localized hyperthermia at a variety of depths without affecting intervening tissue. Clinical applications in neurosurgery, urology, oncology and, more recently, studies assessing the possibility of using HIFU for cardiac ablations have been promising. Pediatric and fetal cardiology are fields in which a non-invasive technique for causing localized tissue damage would be invaluable and may replace invasive and hazardous procedures. The purpose of this study was to investigate the effect of HIFU on cardiac tissues. Porcine valvular leaflet, canine pericardium, newborn atrial septum and newborn right atrial appendage were studied. All specimens were mounted and immersed in a water bath. Using a 1 MHz phased array transducer (Proprietary Technology), energy of 40 or 50 volts was applied for a range of 3 to 25 seconds to the specimen. Macroscopic and microscopic observations were performed before and after HIFU was applied. High-intensity focused ultrasound created a precise defect that ranged in size 1 to 1.2 millimeters in these cardiac tissues. No other damage was identified to the surrounding tissues.

Conclusion: High-intensity focused ultrasound can create precise defects in different cardiac tissue without damage to the surrounding tissue. Further investigation is needed in order to assess the potential use of this technology in clinical settings.

Catheter approach for left accessory pathway ablation: analysis of the pediatric ablation registry data

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The optimum catheter approach, i.e., retrograde (R); transatrial (T); coronary sinus (CS), for radiofrequency (RF) ablation of left accessory pathways (AP) is debatable. The aim of the study was to analyze multicenter (n=29) data to elucidate advantages and limitations of the approaches.

Among 1391 left AP ablations, T was used 966 times; R 462; CS 105 at some time during the RF procedure. Patients: mean age/wt, 12.3 yr/49.5 kg; underlying heart disease (HD), 7%. A single approach was used in 1258/1391 with success (S) in 96% T, 92% R, and 88% CS. When 1 or 2 more approaches were used after the first approach failed (F), S was 84%. When using only 1 approach, S correlated (multivariate analysis, $p < 0.05$) with experiences (E), lateral AP; failure (F) with HD, posterior septal (PS) AP. When T was tried anytime during the procedure, independent correlates of S/F were: E/HD, PS. Similarly for R, correlates of S/F were: none/PS, and for CS: PS/lateral. For fluoroscopy time (FT), T/CS added mean 10.3/9.2 min but R none. Regardless of approach, FT was longer with HD, higher weight, PS, and shorter with E. Major complications (MC) were less with E, greater with younger patients, but unaffected by catheter approach.

Conclusion: E is an important factor for high S, short FT and low MC regardless of approach. While S is demonstrated for each approach, greater S can be expected when more than 1 approach is tried. For greater chance of S for PS AP, the CS approach should be considered early in the procedure.

Autonomic changes in children with neurocardiac syncope

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Background: Neurocardiac syncope in children and adolescents is a common problem. However, there is limited information regarding the autonomic alterations associated with this condition. Previous studies have been unable to determine any noninvasive markers that correlate with a positive tilt table test.

Methods: Noninvasive autonomic testing (handgrip, Valsalva, deep breathing tests), 24 hour time – and frequency-domain analysis of heart rate variability (HRV) as well as tilt-table testing were performed in 8 children (13.2 \pm 3.2 yrs old, 3 M, 5 F) presenting with suspected neurocardiac syncope. Ten healthy children (13.0 \pm 2.9 yrs old, 6 M, 4 F) without any history of syncope or cardiac disease served as controls.

Results: (expressed as mean \pm standard deviation):

| Parameter | Syncope | Control | P-value |
|--|-----------------|------------------|---------|
| HRV: Mean NN (Mean RR Interval) | 720 \pm 79 | 807 \pm 106 | (0.071) |
| SDNN (St Dev RR Interval) | 74.9 \pm 16.1 | 89.3 \pm 22.9 | NS |
| pNN50 (% of adjacent intervals with >50 msec difference) | 19.3 \pm 5.7 | 28.2 \pm 11.8 | (0.054) |
| Tilt: RR Interval at 2 min (msec) | 581 \pm 49 | 682 \pm 125 | 0.037 |
| RR Interval at 5 min (msec) | 542 \pm 57 | 648 \pm 103 | 0.038 |
| SDRR at 5 min (msec) | 26.1 \pm 10.5 | 49.7 \pm 27.2 | 0.029 |
| RR Interval at 10 min (msec) | 493 \pm 67 | 623 \pm 92 | 0.026 |
| SDRR at 10 min (msec) | 25.8 \pm 10.3 | 52.6 \pm 22.8 | 0.025 |
| RR Int – 2 min pre-syncope (msec) | 517 \pm 80 | 648 \pm 103* | 0.013 |
| SDRR 2 min pre-syncope (msec) | 25.4 \pm 13.9 | 49.7 \pm 27.2* | 0.047 |
| SBP 2 min pre-syncope (msec) | 106 \pm 7 | 123 \pm 13* | 0.042 |

*Comparison with 5 minute values in control patients

Analysis of variance of tilt data revealed that, from 0 to 2 minutes, the RR interval in syncope patients decreases at a greater rate than in control patients. Baseline measurements as well as values from noninvasive tests

(i.e. Valsalva, handgrip and deep breathing) did not reveal significant differences between controls and those experiencing syncope.

Conclusions: Although exhibiting normal autonomic testing at baseline, children with neurocardiac syncope have faster heart rates and decreased heart rate variability throughout tilt-testing. Blood pressure and variability are diminished and heart rate is increased as early as two minutes prior to fainting. This suggests parasympathetic withdrawal and/or sympathetic increase may have an important role in precipitating neurocardiac syncope. Additionally, some time-domain measures of heart rate variability (i.e. Mean NN, pNN50) might constitute noninvasive parameters that correlate with a positive tilt table test. These measures suggest that even resting autonomic tone may be altered in patients who experience neurocardiac syncope.

Left ventricular dysfunction in chronic right ventricular-paced patients

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We hypothesize that persistence of the abnormal myocardial electrical activation sequence with chronic right ventricular (RV) apical pacing results in left ventricular (LV) dysfunction. The presence of paced regional wall motion abnormalities necessitates assessment of function with methods that incorporate global changes in ventricular area and derived volume. Automatic border detection echocardiography (ABD) allows instantaneous tracking of the LV endocardial border throughout the cardiac cycle with on-line derivation of multiple parameters of systolic and diastolic function.

Methods: ABD derived indexes of LV function were measured and analyzed in 22 chronically RV-paced patients with the prior diagnosis of congenitally complete AV block (CAVB) in 11, acquired CAVB in 8 and sinus node dysfunction in 3 patients. In order to assess LV function in relation to the duration of RV-apical pacing, patients were divided as follows: Grp I (mean f/u 4.3 ± 2.6 yr; age 12.2 ± 5.6 yr; n=11) vs. Grp II (mean f/u 12.3 ± 2.9 yr; age 24.9 ± 4.8 yr; n=11).

Results: Diminished LV systolic function and a trend towards reduced diastolic function were observed in Grp II patients.

| | SAX Fractional area of change (%) | Peak emptying rate (sec ⁻¹) | Peak filling rate | % Rapid Filling (sec ⁻¹) |
|---------|-----------------------------------|---|-------------------|--------------------------------------|
| Grp I | 53.8 (8.3) | 3.7 (0.6) | 4.6 (1.0) | 81.8 (6.3) |
| Grp II | 44.8 (10.8) | 3.2 (1.1) | 4.0 (1.5) | 78.6 (8.1) |
| p value | < 0.05 | NS | NS | NS |

Conclusion: Long-term apical RV pacing has a negative impact on global left ventricular contraction. Ventricular dysfunction with chronic asynchrony of RV and LV contraction may potentially be reversed by methods that simulate the normal myocardial electrical stimulation sequence such as high RV septal pacing.

CARDIAC CATHETERIZATION/INTERVENTIONAL

Early experience using intravascular stents in children with coarctation of the aorta: promising results ... but proceed with caution!

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Balloon expandable Palmaz stents (ST) are effective in the transcatheter treatment of pulmonary artery stenosis and other vascular obstructions. However, their use in coarctation of the aorta (CoA) is limited. Over 2 yrs, 19 ST (16 P-308, 3P-188) were implanted in 15 pts: 10 males, 5 females; ages 4.5–15.7, 11.8 yrs; wt 17.4–72.8, 46.5 kg. At cath balloon aortic angioplasty (BA) was initially attempted before ST in 10 pts: 6 with native CoA [Group 1A], and 4 with previous surgery (S) and/or BA [Group 1B]. Abdominal CoA was present in 2 pts in Group 1B. ST were primarily deployed in 5 pts [Group 11], all with previous S. Expanded balloon-ST diameters ranged from 10–18 mm, 14.2 mm. There was no difference in age or size between the 2 groups. Results: (Table 2)

Table 2.

| Grp | Systolic Gradient (mmHg) | | | | Area of Stenosis (mm) | | | |
|-----|--------------------------|---------|----------|---------|-----------------------|---------|---------|----------|
| | Pre BA | Post BA | Pre ST | Post ST | Pre BA | Post BA | Pre ST | Post ST |
| 1A | *51.7±10.3 | 19±15.2 | — | 1.5±2.3 | *3±0.8 | 8.8±2.7 | — | 14.7±2.7 |
| 1B | 30.5±4.2 | 20.8±6 | — | 2.5±3.8 | 5.8±2.6 | 6.8±2.6 | — | 10.7±1.8 |
| 11 | NA | NA | 29.4±3.8 | 0.6±0.9 | NA | NA | 7.8±3.1 | 14.1±2.5 |

p < .05 (Non-parametric testing)

Complications: Group 1A: Aortic aneurysm (AA) diagnosed by spiral CT in 2 pts with 1 requiring S; redilation of ST after ST-wall separation in 1 pt. Group 1B: 2nd ST placed for residual CoA in 2 pts. Group 11: ST-sheath entrapment with vascular injury requiring S in 1 pt. **Conclusions:** 1) ST provided excellent relief of CoA gradients, regardless of site or previous S/BA. 2) In native CoA in older pts, tight stenosis is common with significantly larger gradients. BA with full expansion of ST may lead to AA. Serial, graduated dilation may decrease vascular injury and incidence of AA. 3) Spiral CT can detect AA, avoiding ST artifact and should be used in these pts. 4) Cautious optimism and close f/u required!

Transcatheter closure of perimembranous ventricular septal defects with a new device

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Background: Transcatheter closure of Perimembranous ventricular septal defects (PVSD) is controversial because of excellent surgical results and unavailability of an ideal device. The purpose of this study was to evaluate a new device for closure of PVSD in pigs.

Methods: Yucatan micro pigs (n=11) with naturally occurring PVSD were the subject of this study. The median echocardiographic diameter of the PVSD was 5 mm. Six of these pigs had a small aneurysm of the ventricular septum. The device was constructed from 0.004" Nitinol wires with two retention buttons and a connecting waist filled with polyester fibers. The device was shaped to have either concentric left ventricular retention button with 3 mm round flange (n=3), or eccentric button with a 2mm flange towards the aorta and 5mm towards the muscular septum (n=8). A 7 Fr sheath was used to deliver the device from the right ventricular side.

Results: Implantation was successful in all animals. Complete closure was achieved in six (54.5%) immediately after placement and in all pigs after 1 week. Aneurysm of the membranous increased in size in 2 of 3 pigs using the concentric device, but none in pigs with eccentric device. Aortic regurgitation developed in 2 of 3 pigs with concentric device and 1 of 8 pigs with eccentric device. Three pigs developed tricuspid regurgitation. Mitral chordal tendon rupture occurred in one pig because of technical error during placement. The pigs were euthanized after 3 months. Pathologic examination showed the devices completely covered with smooth neoendocardium.

Conclusions: Effective closure of a PVSD is feasible using this new device. The eccentric device appears to minimize aortic regurgitation. More animal studies are needed before human trials can be conducted.

Is ductus arteriosus reopening or late closure following transcatheter coil placement a significant issue?

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Objective: This study was done to evaluate whether late spontaneous closure of an incompletely coil occluded patent ductus arteriosus (PDA) or late reopening of a successfully coil occluded PDA are significant issues.

Background: Late closure of a residual PDA after incomplete occlusion using various devices has been reported. Recent reports documented a high frequency of late PDA reopening following apparent successful coil occlusion.

Methods: All patients who underwent transcatheter PDA coil occlusion using Gianturco .038" coils were studied. For each patient, the post coil(s) placement angiogram, early post procedural (within 8 hours) echocardiogram, and follow up echocardiograms were reviewed.

Results: Forty-three patients with a median age of 3.3 years (range 0.6–76.9) and a median weight of 14.3 kg (range 6.5–72) underwent transcatheter PDA coil placement. The average PDA size was 2 mm. Thirty-four patients (79%) had complete angiographic occlusion. The remaining 9 patients had trivial angiographic leaks. Forty-one patients (95%) were felt to have complete occlusion on the early post procedural echo. The patients have had a median follow up of 9 months (average of 11 months). One of the two residual leaks seen on the early post procedural echocardiogram has completely resolved. Four patients (10%) who were felt to have complete

PDA occlusion on the early echocardiogram have been found to have echocardiographic leaks on later follow up. Three of these four had an angiographic leak. Only one patient has been found to have a new late leak after having had complete PDA occlusion documented by both angiography and early post procedural echocardiography. All of these leaks are silent.

Conclusions: If both angiography and echocardiography demonstrate complete ductus occlusion, late appearance of even a trivial silent leak is unlikely to occur. Early spontaneous closure of trivial angiographic leaks is not uncommon (78%). Late spontaneous closure of a small echocardiographic leak can occur. In the face of an angiographic leak, the finding of complete closure on the early post procedural echocardiogram could represent a transient clotting phenomena or inadequate imaging. We recommend attempting to achieve complete angiographic closure if possible.

Trans-umbilical artery balloon angioplasty of aortic coarctation in the neonate: feasibility and effectiveness

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Whereas there is a controversy with regard to the utility of balloon angioplasty (BA) in the management of aortic coarctation (AC), some have used it effectively. However, the problem of femoral artery compromise is of concern. Therefore, we have utilized transumbilical route to accomplish BA in native AC in the neonate. Fifteen consecutive symptomatic neonates, ages 1 to 30 (median 7) days, with isolated (N=8) or with associated complex congenital heart defects (N=7) underwent transumbilical artery catheterization and angiography followed by BA with 5, 6 or 7 mm diameter (2 cm long) balloons. The balloon diameter was between the diameters of aortic isthmus and descending aorta at the level of diaphragm. PGE₁ infusion was not started prior to the procedure (N=10) or, if the infant has been on PGE₁, it was discontinued six hours preceding BA (N=5). The procedure was successfully accomplished in 14 (93%) of 15 neonates. The peak-to-peak gradient across the aortic coarctation (30 ± 14 vs 8 ± 4 mmHg decreased; $p < 0.01$) and coarcted segment (2.3 ± 0.7 vs 5.5 ± 1.2 mm) increased ($p < 0.01$) following BA. Successful weaning off of the ventilator and subsequent discharge from hospital was accomplished in all isolated coarctations. Palliative (banding of pulmonary artery in complex heart defects) (N=3) or corrective (surgical closure of large perimembranous ventricular septal defect) (N=2) surgery or medical management (N=9) was accomplished successfully in all 14 infants. Transumbilical artery balloon angioplasty is feasible and effective in successful management of neonates with symptomatic aortic coarctation. Experience with a larger number of infants and examination of follow-up data is necessary to further confirm these observations.

Ductal dependent aortic valve stenosis in infants: comparison with non-ductal dependent aortic stenosis and predictors of outcome. The multi-center registry for balloon aortic valvotomy through a carotid cutdown in infants

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Balloon aortic valvotomy through a carotid cutdown was accomplished in 92 infants at 4 centers between 1988 and 1998. Fifty-one were ductal dependent. Results for the 2 sub-groups are listed in the table:

| | Non-ductal dependent | Ductal dependent | P |
|-------------------------|----------------------|------------------|--------|
| Age (days) | 33.5±43.2 | 4.7±6.9 | <.0001 |
| Wt (kg) | 3.7±1.0 | 3.2±0.4 | 0.006 |
| Aortic Diameter (mm) | 8.0±1.4 | 6.6±1.2 | <.0001 |
| LV pre BV (mmHg) | 137±33.1 | 112±31.1 | 0.001 |
| Gradient pre BV (mmHg) | 72.2±28.8 | 56.4±27.2 | 0.013 |
| LV post BV (mmHg) | 93.5±18.7 | 81.6±16.7 | 0.004 |
| Gradient post BV (mmHg) | 21.3±12.1 | 19.5±10.5 | 0.469 |
| Deaths | 2 | 15 | 0.0004 |
| Reintervention | 6 | 13 | 0.04 |

Uni and multivariate analysis shows that mitral stenosis ($p < .005$), a non apex forming LV ($p < .005$), and an aortic valve diameter < 6 mm ($p < .05$) were predictors of a poor outcome. Variables not predictive of a poor outcome included weight, LV diastolic diameter, mitral valve diameter, and aortic arch abnormalities. In conclusion, infants with aortic stenosis and ductal dependent systemic circulation have a worse prognosis than non-ductal dependent infants. They require intervention at an earlier age, have a higher mortality rate and reintervention rate. Mitral stenosis, a non apex

forming LV, and an aortic valve diameter < 6 mm are strong predictors of a poor outcome.

Snare assisted vascular access: a new technique

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Introduction: Access to the central circulation can be difficult in small infants, particularly in the presence of postoperative edema. We describe a new technique which utilizes any existing central catheter to establish additional vascular access sites.

Methods: Using existing arterial or venous access, a 4F endhole catheter is advanced to the target vessel. A 10 mm Amplatz® gooseneck snare is advanced through the catheter and opened within the desired site of entry. Under fluoroscopic guidance, a 21 gauge needle is introduced percutaneously through the snare loop into the soft tissues below the vessel. A 0.018 inch diameter mandril wire is then advanced through the needle into the tissues below the vessel. The needle is then partially withdrawn. The snare is pulled back, engaging the soft tip of the wire and pulling it into the vessel lumen. Once the snare is disengaged from the guidewire, the desired catheter can then be introduced over the wire by standard technique.

Results: Through April 1998, this technique was used in five patients (age 11 days – 2 mos., weight 2.7–3.9 kg). Access was successfully achieved in all patients: Femoral arterial and venous access from the contralateral vessels (using a JL 2.5 catheter), femoral venous access from the internal jugular vein (JR 2.5), a peripherally inserted central catheter (PICC) into the right basilic vein from the left subclavian vein (JR 2.5), a PICC in the left basilic vein from a transhepatic sheath (straight Glide® catheter), and an internal jugular catheter from the femoral vein (straight Glide®). Procedure time for PICC insertion averaged 80 minutes and total fluoroscopy time averaged 12 minutes. There were no complications from this procedure.

Conclusions: Snare assistance is a safe and effective technique which provides a reliable means of establishing additional secure vascular access.

Coil occlusion of patent ductus arteriosus with resultant coil contraction

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Aim and Methods: There are 25 pediatric patients at the Children's Hospital of Pittsburgh who have undergone coil occlusion of a patent ductus arteriosus (PDA) in the cardiac catheterization laboratory between July 1996 and May 1998. Coil occlusion was achieved with Gianturco coils using the snare technique. Using the snare technique to achieve proper placement of the coil within the duct, the coil becomes temporarily elongated. On follow-up chest roentgenograms we have observed contraction in the length of these coils. A retrospective review of chest roentgenograms and echocardiograms was performed to establish the incidence and outcome of this observation. A comparison of the chest roentgenograms were made between the immediate post catheterization chest film and follow-up film obtained 10 to 16 weeks following coil placement. Lateral chest roentgenograms were compared since this projection offered the least variability in technique and the best visualization of coil morphology.

Results: There were 25 patients ranging between 1 and 16 years (median 1 year) with weights ranging between 7 and 66kgs. Two patients had more than one coil placed and were excluded from analysis. Angiographic measurements of the ductal diameters ranged between 1.3mm and 5.5mm. Coil sizes used were: 6mm × 10cm (n=8), 5mm × 8cm (n=7), 4mm × 6cm (n=3), 3mm × 5cm (n=1), 5mm × 10cm (n=1), 6mm × 10cm (n=1), 6mm × 8cm (n=1), 8mm × 10cm (n=1) and 5mm × 5cm (n=1). All 23 patients had complete occlusion of their PDA with the placement of a single coil. Twelve of the 23 patients have had repeat chest films taken 10–16 weeks following the immediate post procedure film that were available for comparison. All twelve patients had a contraction in the length of the coil seen on their follow-up film. Nine of these twelve patients had an echocardiogram performed on the same day as their repeat chest film. All nine echocardiograms demonstrated no residual shunt and no obstruction to flow in the descending aorta or pulmonary arteries.

Conclusion: Contraction in the length of Gianturco coils is seen after placement in pediatric patients for coil occlusion of a patent ductus arteriosus. Possible explanations for coil contraction could be intrinsic properties of the coil to assume its original length after placement or extrinsic factors such as blood clot formation around or within the coil. Contraction in the length of the coil does not appear to be associated with any residual ductal shunting.

CLINICAL CARDIOLOGY/CARDIOVASCULAR SURGERY

Three-dimensional echocardiographic interrogation of the fetal heart: free-hand rapid acquisition utilizing a new technology

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The fetal heart is a three-dimensional (3D) structure in nature. Three-dimensional echocardiography of the fetal heart is a modality that has a number of potential advantages over standard two dimensional (2D) imaging. Until recently, acquisition and display technologies have been inadequate in overcoming the challenges of fetal 3D imaging because of: 1) small cardiac size, 2) respiratory and gross fetal movements, and 3) the need for electrocardiographic gating. The 3D FreeScan system (EchoTech, Munich, Germany) is a novel 3D modality that addresses these technical problems. This system utilizes and electromagnetic sensor device attached to the transducer of any commercially available echocardiographic system. A free-hand scan in a fan-like motion results in a set of polar coordinates that localize the probe in space. Data are digitally stored and processed, and images are displayed as 2D tomographic planes or various 3D rendered images.

Purpose: The purpose of this study was to assess the feasibility of this novel 3D technology in the imaging of the fetal heart.

Methods: Three-dimensional interrogation was performed on a 36 week estimated gestational age (EGA) fetus with a normal heart and a 34 week EGA fetus with hypoplastic left heart syndrome. Gating with the cardiac cycle was accomplished by measuring the fetal heart rate by Doppler interrogation and entering this value into the 3D FreeScan algorithm.

Results: Six acquisition scans were performed on each fetus, and 10 of the 12 were suitable for 3D rendering. Acquisition times ranged between 6 and 8 seconds and post-processing and display times were less than one minute, allowing the scanner to determine the adequacy of the images at the bedside. The reformatted 2D tomograms, surface-rendered 3D images, and volume-rendered 3D images clearly displayed cardiac chambers, outflow tracts, pulmonary veins and spatial relationships between them.

Conclusions: 1. The 3D FreeScan system is a new technology that enables fetal heart imaging in three dimensions. 2. Short acquisition and post-processing times allow analysis of the 3D cardiac image at the bedside. 3. These features make 3D imaging of the normal and abnormal fetal heart a reality, supplanting the traditional 2D imaging.

Experience with intraoperative transesophageal echocardiography during surgery for congenital heart defects: analysis of impact and indications for use

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Objective: To define the impact of intraoperative transesophageal echocardiography (ITEE) in congenital heart surgery and identify appropriate indications for ITEE.

Methods: Medical records of 795 patients having ITEE for congenital heart defects (CHD) were reviewed. ITEE had major impact when new information altered the planned procedure or led to a revision of the initial repair. Impact was defined by the cardiologist and surgeon involved in the operation. Frequency of impact was analyzed by comparison of subgroups of patients based on the primary planned surgical procedure.

Results: Median age was 10 years (range 1 day-85 years). ITEE had major impact in 15.0% (119/795). Impact was more frequent during reoperations than during a patient's first operation (21.2%, 81/382 vs. 9.7%, 40/413; $P < 0.001$). Impact was more frequent than average in 'Rastelli' procedures with an anterior aorta (52.1%, 12/23), LV outflow enlargements (24.4%, 12/49), repair of AV septal defects (23%, 11/47), aortic valve repair/replacement (19.5%, 9/46) and RV to PA conduit replacements (18.7%, 14/75). Three groups had significantly lower rates of impact than average: simple ASD (0%, 0/52; $P < 0.01$), Ebstein anomaly (6.9%, 6/87; $P = 0.04$) and complete repair of Truncus arteriosus or Tetralogy – with or without pulmonary atresia (6.5%, 6/92; $P = 0.03$). There were no significant complications caused by ITEE.

Conclusions: ITEE has become an integral part of congenital cardiac surgical practice. Observed rates of major impact and the absence of complications justify use of ITEE during most intracardiac repairs of CHD, particularly in patients requiring reoperation. Use of ITEE may not be justified in patients with simple ASD, Ebstein anomaly (if no valve repair is attempted) or during complete repair of uncomplicated Tetralogy or Truncus arteriosus.

Risk factors for graft vasculopathy in pediatric heart transplant recipients

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Intracoronary ultrasound (ICUS) is a sensitive method for the detection of graft vasculopathy (GV) in adult heart transplant (HT) recipients and has been used in risk factor analysis in this population. Risk factors for the development of GV in pediatric recipients, however, has been poorly characterized.

Methods: We analyzed ICUS images and correlated results with potential risk factors. Sixty-four ICUS studies were performed in 44 pts at a mean of 4

years (1–11 years) after HT. Median age at HT was 5.9 years (9 days–23.6 years), median age at study 12 years (1.3–24.6 years) and mean wt 37.8 kg (9–85 kg). Analyzed risk factors included donor and recipient age, race, and gender, donor-recipient gender or race mismatch, graft ischemic time, cholesterol, presence of hypertension, CMV infection, episodes of severe rejection (3A), mean first year biopsy score, left ventricular shortening fraction at the time of study, and time from transplant (TT). Eight to 10 random images were obtained from each ICUS study and analyzed for intimal thickening. The presence of any intimal thickening was considered GV. An intimal index (II) was obtained using total vessel area minus luminal area/total area. The point of maximal thickening (MIT) was measured. Vessel disease was graded 0–4 based on these results.

Results: Angiograms were normal in 43/44 pts. GV was present in 30/44 pts (68%) by ICUS with severe disease (3A) presents in 11 pts (25%). A weak positive correlation was present comparing TT with mean II ($p < .02$, $R.369$) and MIT ($p < .01$, $R.4$) in all pts. No factors other than TT were significant for the development of GV. Twenty pts had at least 1 follow-up study; 4 pts had 2 follow-up studies. Only MIT showed significant increase over time ($p, .002$, $R .5$) suggesting focal rather than diffuse progression of GV. Pts were then divided into 2 groups based on the presence or absence of severe GV and risk factors were compared between the 2 groups. No factor other than TT was found to be significant.

Conclusions: GV is common in pediatric HT recipients. There is significant progression of GV as TT increases, however no risk factors for GV other than TT were identified. Further studies are needed to confirm these results.

Late onset of pulmonary hypertension following successful mustard surgery for transposition of the great arteries

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Background: The development of pulmonary hypertension in adolescent and adult patients following successful Mustard surgery for the treatment of transposition of the great arteries is a concern not completely defined. The present study was designed to evaluate long term Mustard patients for the development of pulmonary hypertension, unrelated to previous pulmonary baffle obstruction or persistent left to right shunting lesions.

Methods: All patients followed at Riley Hospital for Children with d-transposition of the great arteries and operated > 7 years before were reviewed to find those with serial estimates of pulmonary pressures. 93 patients repaired between 1972 and 1989 were identified. These patients underwent Mustard repair at 0.5–61 months of age (mean 20.5 months). 9 patients died either postoperatively or within the first 4 years following surgery. 24 additional patients failed to have adequate long term evaluations for pulmonary hypertension to be included in the study.

Results: Serial data was adequate for analysis in 60 patients who had been operated between 7–25 years earlier. Echo and/or catheterization at follow up 7–25 years after surgery showed normal pulmonary pressures in 52/60 (87%), but identified 8 patients (13%) with significant pulmonary hypertension (pulmonary pressures >50% systemic pressures). Four of these had anatomic explanations for their pulmonary hypertension. The remaining 4 patients (7%) had pulmonary hypertension in the absence of pulmonary venous obstruction or left to right shunt lesions. Three of these patients had mildly elevated pulmonary pressures (>33% but less than 50% systemic pressures) at initial postoperative catheterization 9, 11, and 21 months after surgery. These 4 patients were repaired at 25, 25, 17, and 16 months of age and were diagnosed at 16, 15, 12, and 17 years of age as having pulmonary vascular disease.

Conclusions: This study demonstrates that patients with transposition of the great arteries who have undergone Mustard surgery are at increased risk of developing pulmonary hypertension even in the face of normal or slightly elevated pulmonary pressures early after surgery. Those operated on when older than 1 year of age are most suspect. It is important to follow these patients clinically and with 2-D echocardiography/Doppler since pulmonary hypertension can develop late after surgery even when early postoperative pressures are normal or near normal.

The relationship of routine cyclosporine level surveillance to mortality and morbidity after pediatric cardiac transplantation

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While cyclosporine (CSA) levels are often monitored after pediatric cardiac transplantation, the benefits of routine CSA level surveillance are unclear. We analyzed the variability of trough CSA levels in 49 pediatric heart transplant recipients who survived greater than one month, were on the same immunosuppression (CSA, azathioprine, steroid) regimen and CSA level surveillance protocol, and whose CSA dose and level had been stabilized in the therapeutic range (150–300 ng/ml) prior to hospital discharge. CSA level variability was defined as the percentage of CSA levels that were

considered sub-therapeutic (low: <100 ng/ml) and/or toxic (high: >450 ng/ml) out of the total number of CSA levels obtained for each patient. CSA levels drawn for potential drug interactions or possible poor CSA absorption from gastroenteritis were excluded from analysis. Median follow-up for the groups was 42 months (range 6–116 months). For the group, the median percentage of low levels was 3% (range 0–16%), high 5% (0–36%) and combined 10% (0–38%). High CSA variability, defined as a combined high + low level percentage $\geq 20\%$, occurred in 8/49 patients. High CSA variability was significantly associated with the following recipient variables: age >12 months at time of transplant ($p < 0.05$), single parent households ($p = 0.028$), black race ($p = 0.009$), hospital days per year of follow-up ($p = 0.036$), death >6 months after transplant ($p = 0.01$), and recurrent ≥ 2 episodes of ISHLT biopsy grade $\geq 3A$ rejection ($p = 0.0004$). This preliminary study indicates that high CSA variability, which is likely highly related to medical compliance, is a marker for pediatric transplant recipients at greater risk for recurrent rejection and death after transplant.

Use of peak echocardiographic doppler gradient across ventricular septal defects underestimates right-sided pressures in patients with 'sloped' doppler signals

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Background: Most flow velocity profiles across ventricular septal defects (VSDs) derived by Doppler echocardiography have a uniform, plateau shape to the VSD signal. In those patients (pts.), there is a high correlation between the instantaneous peak velocity of that signal and the catheterization (cath) measured peak-to-peak gradient between the left ventricle (LV) and right ventricle (RV). Some children with VSDs were noted to have Doppler velocity flow profiles where the signal peaks early or late in systole, giving the appearance of a 'sloped' signal. The Doppler derived instantaneous peak gradient in these pts. may not be a true estimate of the peak-to-peak gradient between the LV and RV.

Methods: We studied 4 patients with 'sloped' VSD signals. During cardiac catheterization, simultaneous recordings of LV and RV pressures were obtained using 2 Fr. Millar catheters. Simultaneous continuous wave Doppler measurements across the VSD were recorded. Offline beat-to-beat analysis of Doppler signals and pressure tracings was performed. Peak, mean systolic and end-systolic velocities of the Doppler signals were measured and compared to the peak-to-peak cath gradient between the LV and RV.

Results: The instantaneous peak echo gradient ($45.7 \text{ mmHg} \pm 22.9 \text{ mmHg}$) in the four pts. overestimated the peak-to-peak cath gradient ($22.7 \text{ mmHg} \pm 11.5 \text{ mmHg}$) by an average of 23.0 mmHg . The correlation between peak echo and peak cath gradients had an r^2 value of 0.66, (cath gradient = $1.61 \times$ peak echo gradient + 8.95). The end-systolic echo gradient ($18.4 \text{ mmHg} \pm 12.5 \text{ mmHg}$) correlated closely with the peak cath gradient, with an r^2 value of 0.99, (cath gradient = $1.08 \times$ end-systolic echo gradient + 6.03). The mean systolic echo gradient ($23.8 \text{ mmHg} \pm 14.9 \text{ mmHg}$) also correlated closely with the peak cath gradient, with an r^2 value of 0.86, (cath gradient = $1.20 \times$ mean systolic echo gradient + 3.45).

Conclusion: The data shows that the instantaneous peak echo gradient clearly overestimates the cath derived peak-to-peak gradient in pts. with 'sloped' VSD signals. Mean systolic and end-systolic echo gradients correlate better with the peak-to-peak gradient between LV and RV determined at cath. Therefore, the use of peak echo gradient in pts. with 'sloped' signals across the VSD leads to underestimation of the true RV and pulmonary artery systolic pressures.

A review of a single institutional experience with fetal echocardiography: implications for the future

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Fetal echocardiography has been shown to be a sensitive diagnostic tool for the *in utero* recognition of most major congenital heart diseases (CHD). As the number of referrals has greatly increased, issues of available manpower, time and cost have been raised, especially in today's cost containment environment. To evaluate the various aspects of our fetal medicine clinic, we retrospectively reviewed all patient encounters from January 1987 through December 1997. The records were analyzed for various factors including indication for referral, percent yield by indication, sensitivity, specificity and postnatal follow-up where available. A total of 2735 studies were performed on 2595 fetuses during this time period. There were 210 fetuses diagnosed with CHD or potentially life-threatening arrhythmias. The table shows the data for the 11 most common referral indications.

Although arrhythmias have a high yield, 64% of the abnormal findings were isolated premature atrial contractions with no structural heart disease or evidence of sustained arrhythmias. These data suggest that certain common 'high-risk' indications have a very low yield that may not be significantly higher than the average pregnancy. It is recognized that a negative study for parents of previous children with CHD can be as

| Indication | # of studies | # abnormal | % yield | 95% CI |
|-------------------------------------|--------------|------------|---------|-------------|
| Insulin dependent diabetes mellitus | 811 | 28 | 3.5 | (2.2–4.7) |
| Previous child w/CHD | 628 | 11 | 1.8 | (0.7–2.8) |
| Fetal arrhythmia | 246 | 114 | 46.3 | (40.1–52.6) |
| Abnormal 4 chamber view | 165 | 84 | 50.9 | (43.3–58.5) |
| Maternal CHD | 138 | 2 | 1.4 | (0.0–3.4) |
| Family Hx CHD | 16 | 0 | | |
| Extracardiac anomalies | 98 | 16 | 16.3 | (9.0–23.6) |
| Lithium exposure | 74 | 0 | | |
| Anticonvulsant exposure | 79 | 0 | | |
| Aneuploidy | 41 | 23 | 56.1 | (40.9–71.3) |
| Single umbilical artery | 27 | 6 | 22.2 | (6.5–37.9) |

important as an abnormal one due to high levels of anxiety. However, indications such as irregular heart rhythm and certain teratogen exposures in the setting of a normal 4 chamber view on a routine obstetrical ultrasound may need to be reconsidered.

Bicaval versus right atrial heart transplantation in children

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Between 1992 and 1997, 26 consecutive children underwent orthotopic heart transplantation for cardiomyopathy (11), complex congenital cardiac anomalies (11), adriamycin toxicity (3), and transplant coronary artery disease (1). Standard right atrial anastomosis (RA) was used for the first 13 (1992–1995) and bicaval anastomosis (BICAV) for the subsequent 13 (1995–1997). Patients were reviewed and compared retrospectively for early and late survival, donor ischemia time, requirement for pacing, degree of tricuspid regurgitation (TR), and need for tricuspid valve replacement (TVR). After BICAV patients were evaluated for IVC-to-RA and SVC-to-RA gradient. Mean age in the RA group was 8.0 ± 6.3 years, in the BICAV group 11.3 ± 5.7 years ($p = \text{NS}$). There was one early and one late death in the RA group; there were no deaths in the BICAV group ($p = \text{NS}$). Mean donor ischemia time in the RA group was 154 ± 33 minutes, in the BICAV group, 138 ± 51 minutes ($p = \text{NS}$). No patient required a permanent pacemaker. TR in the RA group as assessed by serial transthoracic color Doppler echocardiography was trivial (4), mild (3), moderate (4), and severe (2). In the BICAV group it was assessed as trivial (9) and mild (4) (RA versus BICAV, $P < 0.05$). Two patients in the RA group have required TVR, none in the BICAV group ($p = \text{NS}$). No patient had an SVC-RA or IVC-RA gradient after BICAV anastomosis. In children, the use of BICAV technique instead of RA significantly reduced postoperative TR, and did not affect early or late survival or prolong ischemia time.

A non-invasive index for global ventricular function: a predictor for rejection following pediatric cardiac transplantation?

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Numerous techniques have been used to assess ventricular function in patients following cardiac transplantation. A non-invasive index (Tei index) for combined systolic and diastolic ventricular function may better reflect global cardiac function. The Tei index is defined as the sum of the isovolumetric contraction and relaxation time divided by the ejection time. The purpose of this study was to determine if the Tei index may predict rejection in pediatric patients following cardiac transplantation.

Methods: Six transplant recipients were identified as having experienced both an episode of biopsy-proven rejection (grade 2 = focal, moderate) as well as an episode of non-rejection on biopsy. Echocardiographic data was blindly reviewed from studies coincident with their biopsies. The following parameters were measured: right and left ventricular isovolumetric contraction and relaxation times, ejection times, left ventricular shortening and ejection fractions. The parameters from periods of non-rejection and rejection were compared for each patient by paired *t* test.

Results: M/F = 5/1, age range = 3–19 years, mean 10.5 years. The mean LV Tei index during periods of non-rejection was 0.37 ± 0.08 . During rejection episodes, the mean LV Tei index increased to 0.47 ± 0.17 . There was a clear trend toward increased LV Tei index during periods of biopsy-proven rejection. Five of six patients (83%) demonstrated an increased LV Tei index during periods of rejection. RV Tei index, LV shortening fraction, and LV ejection fraction were not predictive of rejection.

Conclusion: When applied to the left ventricle, the non-invasive Tei index for global ventricular function may serve as a predictor for cardiac rejection in pediatric transplant patients. A large, prospective study is needed.

Modifications of Norwood surgery for improved long-term outcome

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Right ventricular dysfunction and diffuse distortion/hypoplasia of pulmonary arteries are common after Norwood I surgery (NS) for hypoplastic left heart syndrome (HLHS) or variants (HLHV). These affect outcome of subsequent surgeries including eventual Fontan operation. To minimize these problems, myocardial protection during NS was modified through frequent antegrade cardioplegia. In addition, systemic-pulmonary shunting was central, from proximal pulmonary artery and primarily directed towards left pulmonary artery (LPA). In the past 2 years, 43 babies with HLHS or HLHV had NS at age $8.9d \pm 8.2$ (SD). Operative mortality was 19%. Serial echo studies were analyzed in 18 patients with HLHS to evaluate RV function pre/post NS, using RV end diastolic area (EDA) and end systolic area (ESA), and expressed as percent area change (%AC = EDA-ESA/EDA). Tricuspid regurgitation was absent in 8 (42%), mild in 6 (33%), moderate in 4 (25%). Range of % AC for normal RV: 18–29%. In HLHS group, % AC pre NS was $36.3 \pm 9\%$ vs post NS (at mean age of 100d) of $33.8 \pm 9\%$, $p=NS$. Angiograms ($n=15$) obtained before Glenn surgery were analyzed. RV function was normal. Discrete mild narrowing of LPA or RPA was noted at shunt site 14; however, distal pulmonary arteries showed normal anatomy. Diameters of RPA and LPA were 7.0 ± 0.6 mm and 5.1 ± 0.5 mm, respectively ($p < 0.03$). When related to the diameter of descending aorta at level of diaphragm, RPA/DAo and LPA/DAo ratios were in normal range and were 0.66 and 0.50, respectively. Modification of myocardial protection techniques and of systemic-PA shunting during NS help preserve normal RV function and RPA/LPA anatomy, conducive to better long-term outcome in HLHS/HLHV.

Impaired exercise parameters in pediatric heart transplant recipients

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Background: Adult cardiac transplant patients have decreased endurance time and exercise performance. The endurance and chronotropic response in pediatric cardiac transplant recipients is unknown.

Materials and Methods: We performed treadmill studies by modified Bruce protocol in 18 recipients who were at least 1 year post transplantation and had no episodes of rejection for 1 month prior to the exercise test.

Results: The patients were 7.8–21 (mean = 16.4) years old, and 1.1–7.6 years post transplantation. There were 2 girls, 16 boys. Surgery was by either biatrial (6) or bicaval anastomosis (12). Immunosuppression was cyclosporine-based in all. Prednisone doses were 0–0.34 mg/kg/day (mean 0.18) and 17/18 were on antihypertensives. All tests were terminated due to fatigue. The endurance time was 6.03–12.5 minutes (mean 8.49), which was below the predicted 10th percentile for age and gender in 13/18 patients. The peak heart rate mean was 158 ± 18 bpm (range 131–202) which was only 75% of the predicted maximum for age. The mean resting and peak systolic

blood pressures were 127 ± 11.5 and 153 ± 23.4 mmHg. The maximal oxygen uptake (VO₂) was 17.3–42 ml/kg/min (mean 27.1, which was 54% of predicted). The method of surgical repair did not effect the endurance, VO₂ or peak heart rate. Three patients had arrhythmias during exercise (premature ventricular contractions 2, junctional rhythm 1).

Conclusion: Response to exercise in pediatric heart transplant recipients includes diminished chronotropic response, endurance and maximum oxygen uptake. Future studies are warranted to see if these patients can improve their performance with regular aerobic conditioning.

Who is at risk following the Norwood procedure? Identification of patients at risk for low systemic oxygen delivery following the Norwood procedure for hypoplastic left heart syndrome

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Early identification of low systemic oxygen delivery in patients with hypoplastic left heart syndrome (HLHS) following the Norwood procedure should improve outcome.

Methods: In this prospective study, superior vena cava saturation (SvO₂) and arteriovenous oxygen content difference (DAVO₂) were recorded hourly for the first 48 hours in 22 consecutive patients following the Norwood procedure from 6/96–2/98. Age, preoperative support, shunt size and anatomic subtype (aortic atresia v antegrade aortic flow) were evaluated using multiple logistic regression to determine their impact on early postoperative systemic oxygen delivery. Satisfactory oxygen delivery was defined as an $\Delta AVO_2 \leq 5$ vol% and $SVO_2 \leq 50\%$.

Results: Thirty-day survival was 100%, there was one hospital death, hospital survival 95% (21/22). Risk factors for decreased systemic oxygen delivery are summarized in the table below.

Conclusion: Aortic atresia, older age, larger shunt size and preoperative support are previously identified mortality risk factors. These same factors were predictive of decreased systemic oxygen delivery, which could be effectively managed using continuous SvO₂ monitoring.

| | Risk for SvO ₂ ≤ 50% | | ΔAVO ₂ ≤ 5 vol% | |
|--------------|---------------------------------|------------|----------------------------|------------|
| | p value | odds ratio | p value | odds ratio |
| Age > 8 days | P<0.001 | 1.75 | P=0.022 | 1.46 |
| !Shunt Size | P=0.004 | 1.52 | P<0.001 | 1.96 |
| Preop Vent | P<0.001 | 0.24 | P=0.35 | |
| Preop Ino | P=0.002 | 1.76 | P=0.7 | |
| Ao Atresia | P<0.001 | 1.86 | P<0.001 | 2.13 |

Preop = preoperative, Vent = mechanical ventilation, Ino = inotropes, Ao = aortic