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Ductal Closure in the Premature Infant with Paracetamol

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We discuss our experience in the Neonatal Intensive Care Unit with closure of hemodynamically significant patent ductus arteriosus with oral Paracetamol. We have treated a series of five under 1250 gram and under 30 week premature infants with either failure to respond to Ibuprofen (during a period when Indomethacin was unavailable for clinical treatment) or contraindications to its use after experiencing a surprising and unexpected closure in the first of the series treated with Paracetamol (for an unrelated indication.) All the infants responded definitively. We discuss the theoretical mechanism and advantages of paracetamol over conventional treatments in the treatment of PDA and describe a prospective double blind study developed to conclusively evaluate its clinical role.

The Expression of NKX2.5 Gene in Congenital Heart Disease

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Background: Congenital heart diseases (CHD) are the most common heart developmental anomaly disease.Nevertheless, the aetiology of CHD in the majority of cases remains unknown. Abnormal cardiac development appears to occur through a process that is heterogeneous and complex, with both environmental and genetic risk factors. Heart formation is a complex process regulated by many transcription factors. Although most of CHD cases are sporadic, there are several genes which were found to be associated with CHD, including NKX2.5, TBX5, GATA4, etc.

Objective: To analyze the changes of NKX2.5 mRNA in human myocardium of congenital heart diseases (CHD) and to explore the relationship between NKX2.5 and congenital heart disease.

Method: The total mRNA were extracted from 10 cases with atrial septal defect (ASD) myocardium and 10 cases with ventricular septal defect (VSD) and 8 cases with non-CHD myocardium. 10 ASD cases are l2.71¡À8.12 (1.6~25) years old, 10 VSD cases are l2.44¡À4.98 (2¡«18) years old, 8 non-CHD cases are 22.92¡À12.33 (5-38) years old. The levels of NKX2.5 gene mRNA expression in the myocardium of CHD and the control group myocardium were detected using the RT-PCR (Reverse Transcription-Polymerase Chain Reaction) technique.

Results: The NKX2.5 gene was expressed in both the non-CHD and CHD myocardium. The levels of NKX2.5 gene mRNA expression were (73.72¡À6.21)£¥in the non-CHD myocardium, and (49.39¡À4.95)£¥in the ASD myocardium, and (29.69¡À3.48)£¥in the VSD myocardium. The levels of NKX2.5 gene mRNA expression in the ASD and VSD myocardium were significantly less than those in the non-CHD myocardium (t=9.26, 19.06, respectively; P <0.01)£®

Conclusion: The levels of NKX2.5 gene mRNA expression in the ASD and VSD myocardium were significantly less than those in the non-CHD myocardium. The results suggest that the abnormal expression of NKX2.5 may be involved in the pathogenesis of ASD and VSD.

After Tetralogy Repair Children Have Right Ventricular Dilation Disproportionate to Body Growth

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Background: After repair of tetralogy of Fallot (TOF), patients frequently exhibit significant pulmonary regurgitation (PR) that results in right ventricular (RV) dilation and ventricular septal dyskinesia predisposing to RV and left ventricular (LV) dysfunction. The rate of RV dilation and development of RV and LV dysfunction in the face of PR in growing children are unknown.
Hypothesis: After TOF repair, RV and LV growth remain proportionate to body growth, and RV and LV systolic performance parameters remain stable during childhood.
Methods: We retrospectively reviewed 35 consecutive repaired TOF patients who underwent ≥ 2 clinical cardiac magnetic resonance studies between 2005 and 2011. The first and last studies were compared. All patients were < 18 yrs and had moderate PR (Regurgitant Fraction > 20%) at both studies. Volumetric data was indexed for body surface area (BSA). Patients who underwent catheter or surgical intervention in the interim were excluded. Significance was $P < 0.05$.
Results: For the entire study population, the mean age and BSA were 9.8 ± 4.0 yrs (range 1.4 - 16.1 yrs) and 1.1 ± 0.3 m2 initially, with a mean follow up of 2.7 ± 1.1 yrs. The BSA increased to 1.3 ± 0.3 m2 at the follow-up study. Patients with at least moderate PR demonstrate increases in RV end-diastolic and end-systolic volumes (see Table). The RV ejection fraction mildly decreased (60.1 ± 7.0% vs 57.3 ± 6.0%, p=0.054). There was no significant difference in pulmonary regurgitant fraction, fractional net branch pulmonary blood flow, or LV performance parameters.
Conclusions: During childhood, patients with at least moderate PR exhibit RV dilation disproportionate to body growth. These data are important when considering TOF patients for surveillance studies and pulmonary valve replacement.

	Right Ventricle		Left Ventricle	
	I-EDV (cc/m^2)	I-ESV(cc/m^2)	I-EDV(cc/m^2)	I-ESV(cc/m^2)
First Visit	122.2 ± 29.8	49.7 ± 16.2	62.1 ± 11.8	18.9 ± 5.8
Final Visit	132.8 ± 31.9	57.8 ± 19.0	61.6 ± 14.1	19.3 ± 5.4
Average Δ per year	4.4 ± 9.5	3.2 ± 8.9	0.3 ± 5.4	0.3 ± 2.6
P-Value	0.002	0.007	0.845	0.723

*I-EDV = Indexed End-Diastolic Volume; I-ESV = Indexed End-Systolic Volume

Anomalous Left Coronary Artery from Pulmonary Artery (ALPACA) - Is It Really so Rare?

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Background: Abnormal left coronary artery from pulmonary artery (ALCAPA) is described in the literature as a rare congenital cardiovascular defect that occurs in approximately 1/300000 live births. The mortality of untreated ALPACA has been estimated to range from 35 % -85% in the first year of life. Our data and experience shows a much higher incidence of this entity.
Methods: Hospital data bases were retrospectively searched for cases of ALPACA diagnosed at our center over the past 10 years.
Results: During a period of 10 years, 2001-2011, we diagnosed and treated 9 patients with ALPACA. Seven patients came to medical attention because of respiratory distress and asthma like symptoms that developed during the first month of life. Two patients were referred for cardiac evaluation because of failure to thrive and systolic murmur. Age at clinical presentation ranged from 1-7 months. Diagnosis was made at 2-48 months, average 15 months. The main initial diagnostic tool was echocardiography and diagnosis was confirmed by cardiac catheterization. During the last period of study, CT and MRI were used in order to confirm the diagnosis. All 9 patients had successful surgical repair by re implantation of the left coronary artery to the aorta.
Conclusion: Relying on the published incidence and based on the annual birth rate in our hospital we should have diagnosed and treated only one tenth of the actual diagnosed patients over a period of 10 years. Our rate is tenfold higher than expected. We speculate that ALCAPA is either more frequent in certain geographic areas or simply under diagnosed worldwide. A high index of suspicion and new diagnostic modalities like MRI and CT angiography may show that the current reported incidence is actually too low.

Short QT Syndrome in the Pediatric Age Group - Rare and Challenging Diagnosis

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Background: Short QT syndrome, is a fairly novel entity in the medical literature, but with very high morbidity and mortality. At this time, there is scarcity of information regarding all aspects of the disease, especially management in children.

Aim: We present a case of the new diagnosis of Short QT syndrome in a 13 months old baby that came to our attention prior to starting beta blockers for facial hemangeoma, due to a family history of aborted cardiac arrest.

Results: The child's father at age 31 years, prior to patient's birth, experienced sudden cardiac arrest and underwent resuscitation following which he had an intracardiac defibrillator implanted. At the time of presentation the child was asymptomatic and during the clinic visit had a normal cardiac physical exam and echocardiogram. On the EKG the patient was found to have a QTc interval of 0.294, no ST segment, and no peaked T-waves. It was decided to start Quinidine, to defer ICD implantation and refer the family for genetic testing.

Conclusions: The definition of the Short QT interval is not clear at this time, but appears to be QTc less than 320ms while QTc less than 360ms is highly suggestive. Most patients are male, with >50% of the cases having a significant family history of cardiac arrest. Present genetic studies have identified 3 genes; gain of function mutation of a potassium channel and 2 loss of function mutations in a calcium channel. Current treatment at this time is uncertain. It does appear that patients who carry the potassium channel mutation benefit from treatment with Quinidine. Among adults who experienced a sudden cardiac arrest an ICD is recommended, however in pediatrics it is a more complex decision, given the rapid growth of a child, device sizes and the overall higher complications rate. Further studies are needed and probably an establishment of international database could benefit gathering of information about this rare condition.

Vascular Ring and Sling (and other things) Imaging-Two Year Experience

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Background: Airways obstruction and stridor are common in children, a portion of these are caused by abnormalities in the embryologic development of the aorta and pulmonary arteries, known as vascular rings and slings. In most centers Computerized Tomographic Angiography (CTA) is used to evaluate these abnormalities. Recently we have begun to use MRI/A instead of CTA in order to avoid ionizing radiation exposure and injection of iodinated contrast agents administration.

Methods: A retrospective analysis of imaging data from December 2009 to November 2011 was performed. The study group included 17 patients with suspected aortic arch or pulmonic arterial abnormalities by either physical examination and/or echocardiography, including fetal echocardiography(age range: 2 days-36 years, mean age: 63 months; weight of 3.1 kg-57 kg) Seven patients were scanned using CTA and ten patients using MRI/A . All the studies were supervised and interpreted by a dedicated team of a cardiologist and radiologist for detecting cardiovascular and extravascular abnormalities.

Results: Among the CTA (N=7) patients, 4 patients had a vascular ring associated with right aortic arch, 1 patient had a vascular sling, 2 patients had tracheo-bronchial compression by an enlarged pulmonary artery. Among the MRI/A patients 7 patients had a vascular ring associated with right aortic arch, and 3 patients had aortic arch abnormalities without vascular ring. Complication: one infant had apnea easily managed by bag-mask before entering the MRI scanner ventilation and was then scanned. Both CTA and MRI/A findings were confirmed by the operative findings. No difference was found in the accuracy of pre-operative diagnosis between CTA or MRI/A findings.

Conclusion: Cardiac MRI and CTA are comparable modalities to evaluate vascular rings and slings. In our experience MRI/A, lacking of risks of ionizing radiation and iodinated contrast agents, has become a useful alternative in stable patients.

Gerbode Type Defect in Adults after Repair of Tetralogy of Fallot

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Background: Left ventricular (LV) to right atrial (RA) communications can be congenial or acquired. They are well known in patients with repaired atrio-ventricular septal defect and are increasingly recognized in adults with acquired heart disease. LV to RA communications or Gerbode-type defects (GD) are poorly recognized in adults with tetralogy of Fallot (TOF).
Objective: The objective was to assess the prevalence and the clinical features of the GD in a contemporary cohort of adults with repaired TOF.
Methods: Adults (¡Ýage 18 years) with repaired TOF, actively followed at the Toronto Congenital Cardiac Center for Adults (n=502) and at the Congenital Cardiac Clinic, Rambam Medical Centre Haifa, Israel (n=24) were included. Their charts were reviewed for the diagnosis of a GD and acquisition of the anatomic features and surgical details.
Results: A GD was identified in 9 of 526 (1.7%) patients. The GD was adjacent to the ventricular septal defect (VSD) patch and was not hemodynamically relevant in all patients. Though present for many years or since surgical repair, the GD was diagnosed late after TOF repair and misinterpreted as high velocity jet of tricuspid regurgitation, which lead to the incorrect diagnosis of an elevated right ventricular systolic pressure.
Conclusions: The presence of a Gerbode-type defect is a rare complication in adults with repaired TOF. Detection of a GD is a diagnostic challenge and a potential source of misinterpretation, which results in misdiagnoses.