402A ABSTRACTS - Pediatric Cardiology

1097 Pediatric Pulmonary Hypertension and Transplantation

Monday, March 18, 2002, 9:00 a.m.-11:00 a.m.
Georgia World Congress Center, Hall G
Presentation Hour: 10:00 a.m.-11:00 a.m.

1097-97 Treatment of Pulmonary Arterial Hypertension Associated With Congenital Heart Disease With Intravenous Epoprostenol: Impact of Therapy on Timing of Transplantation


Introduction: Intravenous epoprostenol (EPO) improves exercise tolerance, hemodynamics, and survival in primary pulmonary hypertension (PPH). Patients with PPH and congenital heart disease (CHD) demonstrate improved hemodynamics with EPO over the short term. The objective of this study was to determine the long-term effects of EPO in CHD and implications for timing of lung or heart-lung transplantation.

Methods: The Rush database of 264 patients with PPH treated with EPO since 1991 was queried for patients with CHD. The 16 patients with CHD were matched in a 2:1 fashion with 32 PPH patients for age, gender, and functional class (FC) at the time of EPO Initiation. Hemodynamic and exercise tests at the time of EPO initiation and at one year were compared using students paired t-test. Survival over 5 years was analyzed using Kaplan-Meier analysis.

Results: EPO resulted in similar improvements in hemodynamics and tracheal时限s in both PPH and CHD at 1 year. In CHD the PVR declined by 30% from 17 to 12 Wood units (p=0.001), the systemic arterial O2 saturation improved from 86 to 90% (p=0.002), and the treadmill time improved 118% from 226 to 492 seconds (p=0.003). The 1, 3, and 5 year survivals were 93%, 88%, and 81% for CHD versus 84%, 66%, and 62% for PPH respectively.

Conclusion: The favorable response of CHD to EPO is similar to that of PPH. The survival of CHD with EPO exceed that of PPH and that of lung or heart-lung transplantation.

1097-98 Sildenafil (Viagra) in Childhood and Neonatal Pulmonary Hypertension


Background: Cyclic guanine monophosphate (cGMP) mediates vasodilation induced by nitric oxide (NO), inhibition of cGMP phosphodiesterase (PDE) promotes vasodilation. Therefore, we investigated the effect of oral sildenafil in pulmonary hypertension. Methods: We administered oral sildenafil (0.25 - 0.5 mg/kg) to 16 patients (median age 6 years, range 3 days to 18 years, gender 7M:9F). All patients had pulmonary hypertension defined by echocardiography or catheter measurement. During cardiac catheterisation (n=9) we measured hemodynamics before and 30 minutes after administration of sildenafil. We administered sildenafil to 6 patients with refractory suprasystemic pulmonary hypertension after gradual withdrawal of inhaled NO, without alapealisation. (diagnoses included repair of congenital diaphragmatic hernia, n=3, and pulmonary vein stenosis n=2). Five patients received inhaled nitric oxide therapy, given 4 times a day (primary pulmonary hypertension n=3, secondary pulmonary hypertension n=2). The following data were collected: mean pulmonary artery pressure, pulmonary vascular resistance, indexed pulmonary vascular resistance, mean systemic arterial pressure, and systemic vascular resistance. Results: The sildenafil dose range was 0.1 to 0.5 mg/kg, with a mean dose of 0.35 mg/kg. Pulmonary vascular resistance decreased from 10.5±4.9 to 7.6±4.6 Wood Units (p<0.05). There were no changes in mean systemic pressure, pulmonary vascular resistance, and systemic arterial pressure. Conclusion: Sildenafil is safe and may be effective in children with refractory suprasystemic pulmonary hypertension. Sildenafil is currently being evaluated in a multicenter, placebo-controlled trial.

1097-100 Presenting Features and Clinical Outcomes for Children With Metabolic Cardiomyopathies

Pete E. Daubney, Alan Nugent, Patty Chondros, Stephen Kohen, John Carlin, Robert O. Weintraub. National Australian Childhood Cardiomyopathy Study, Royal Children's Hospital, Melbourne, Australia.

Background: Metabolic diseases are an important cause of childhood cardiomyopathy. This review examines the presenting features and clinical outcomes for children with these conditions enrolled in the National Australian Childhood Cardiomyopathy Study (NACC).

Methods: NACC is a population-based study including all children in Australia with CM presenting <10 years of age from 1987-97. Cases were collected from all pediatric cardiologists and pediatric cardiologists' centres, as well as from adult cardiologists, regional arrhythmologists, cardiac transplant centres and congenital heart registries. Cases were excluded according to accepted WHO guidelines. Metabolic diseases were defined as those with a biochemical abnormality aetiologically linked to the cardiomyopathy. Children with progressive neuromuscular diseases and those with dominant systemic or neurological symptoms were excluded.

Results: There were 28 children with metabolic conditions (6.9% of study population) including 6.5% of patients with DCM, 25% with HCM and 32.5% of patients with unclassified cardiomyopathy. Congestive heart failure at presentation occurred in 21/28 (75%). Common presenting diagnoses were respiratory chain enzyme deficiencies (10), Barth syndrome (8), enzyme deficiency syndromes (4) and fatty acid oxidation defects (4). Children with respiratory chain enzyme deficiencies had variable cardiomyopathies including DCM, HCM and mixed hypertrophy with systolic dysfunction. 7/8 (87.5%) children with Barth syndrome had LV non-compaction (LVNC) and the other had DCM. Barth syndrome was present in 7/29 (24%) patients with LVNC. 6 of 8 with unclassified cardiomyopathy (increased LV wall thickness with systolic dysfunction) had metabolic disease. Overall mortality was 17/28 (60.7%). In 6/10 the diagnosis of a respiratory chain defect was not made until after death.

Conclusions: There is considerable clinical heterogeneity among children with metabolic cardiomyopathies. They should be suspected in children with atypical cardiac features. Barth syndrome should be excluded in males with LVNC. Routine assay of respiratory chain enzymes on postmortem or explanted hearts may improve the diagnostic yield.

1097-101 Risk Factors Associated With Posttransplant Coronary Artery Disease in Pediatric Cardiac Transplant Recipients

Nahid Hassansy, Leonard Bailey, Steven Gundy, Anne Ruzzo, Richard Chinnock, Paul A. Cecchinia, Loma Linda University Children's Hospital, Loma Linda, California, Loma Linda, California.

Background: Post-transplant coronary artery disease (PTCAD) is associated with a high mortality and a significant risk of graft failure. A number of associated factors have been postulated including ischemic time, gender mismatch, cytomegalovirus (CMV) infection, and rejection history. However, to date, the cause of PTCAD is not currently known.

Purpose: The purpose of the current study is to examine possible factors associated with PTCAD in a pediatric transplant population.

Method: Pediatric patients who underwent cardiac transplant between 1985-2001 were retrospectively reviewed.

Results: A total of 358 infants and children underwent orthotopic cardiac transplantation. Overall survival is 70%. Forty-three patients (12%) developed PTCAD. The average time to PTCAD was 5.56 yrs (range 1.14 - 14.45). Mortality within the PTCAD group was significantly higher that the remaining transplant population (57% vs. 26%, p<0.001). Patients who developed PTCAD had a higher number of rejection episodes per year (1.06 vs 0.43, p<0.001). There was a lower incidence of CMV found in the PTCAD group (82% vs 95%, p<0.001). Patients who developed PTCAD as had a shorter overall dopamine requirement in the post-operative period (0.37 vs 5.10 days, p<0.005). Factors not associated with PTCAD included: ischemic time of the graft, gender, gender mismatch, donor to recipient weight ratio, and left ventricular end-diastolic pressure, days on mechanical ventilation in the post-operative period, CMV status of the recipients, or HLA tissue typing mismatch.

Conclusion: PTCAD among pediatric cardiac transplant recipients sigificantly increases morbidity and mortality. PTCAD is associated with a higher number of rejection episodes. The role of CMV infection remains unclear. PTCAD is not associated with ischemic times, which has been previously suggested. Early and frequent coronary artery evaluation in children with frequent rejection episodes may be warranted.