1097-98 Treatment of Pulmonary Arterial Hypertension Associated With Congenital Heart Disease With Intravenous Epoprostenol: Impact of Therapy on Timing of Transplantation

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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 (NACCS). Methods: NACCS 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 paediatric cardiologists and paediatric cardiac sentres, as well as from adult cardiologists, regional cardiology, cardiac transplant and congenital heart centres. Cases were reviewed 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 (8.9% of study population) including 6.5% of patients with DCM, 25.5% with HCM and 32.6% of patients with unclassified cardiomyopathy. Congestive heart failure at presentation occurred in 21/28 (75%). Commonest diagnoses were respiratory chain enzyme deficiencies (10), Barth syndrome (8), carnitine 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-competition (LVMC) and the other had DCM. Barth syndrome was present in 7/29 (24%) patients with LVMC. LV 8% with unclassified cardiomyopathy (increased LV wall thickness with systolic dysfunction) had metabolic disease. Overall mortality was 17/28 (60.7%). In 6/10 a diagnosis of a respiratory chain defect was not made at initial presentation.

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 at postmortem or explanted hearts may improve the diagnostic yield.

1097-99 Risk Factors Associated With Posttransplant Coronary Artery Disease in Pediatric Cardiac Transplant Recipients

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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.

Methods: Pediatric patients for whom post-transplant coronary artery disease has been postulated were identified. Age, gender, race, number of ischemic events, surgical intervention, rejection and anti-rejection therapy were reviewed. The data were analysed to identify any significant risk factors. The database was prospectively reviewed.

Results: A total of 338 infants and children underwent orthotopic cardiac transplantation. Overall survival is 70%. Forty-three patients (12.1%) developed PTCAD. The average time to PTCAD was 5.66 years (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 (1.00 vs. 0.43, p<0.001). There was a lower incidence of CMV found in the patients who went on to develop PTCAD (40% vs. 60%, p=0.006). Patients who developed PTCAD had a shorter overall dopamine requirement in the post-operative period (2.87 vs 5.10 days, p<0.05). Factors not associated with PTCAD included: ischemic time before and after 20 minutes, gender, mismatch, donor type, donor to recipient weight ratio, right 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.

Conclusions: PTCAD in pediatric cardiac transplant recipients signifi cantly increases mortality. PTCAD is associated with the number of rejection episodes. The role of CMV infection remains unclear. PTCAD is not associated with ischemic times, as has been previously suggested. Early and frequent coronary artery evaluation in patients with frequent rejection episodes may be warranted.

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

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Conclusions: PTCAD in pediatric cardiac transplant recipients signifi cantly increases mortality. PTCAD is associated with the number of rejection episodes. The role of CMV infection remains unclear. PTCAD is not associated with ischemic times, as has been previously suggested. Early and frequent coronary artery evaluation in patients with frequent rejection episodes may be warranted.