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

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Background: Cyclic guanosine monophosphate (cGMP) mediates vasodilation induced by arterial perfusion of eNOS-derived NO and promotes vasodilation, Therefore, we investigated the effect of oral sildenafil on pulmonary hypertension (PH) associated with congenital heart disease (CHD) demonstrate improved hemodynamics with EPO over the short term.

Methods: The dataset of 264 patients with PH treated with EPO since 1991 was queried for patients with PH. The 16 patients with PH who were matched in a 2:1 fashion with 32 PPH patients for age, gender, and functional class (FC) at the time of EPO initiation. Hemodynamics and exercise time before and 2 weeks after initiation of EPO at and at one year was compared using patients paired t-test. Survival over 5 years was analyzed using Kaplan-Meier analysis.

Results: EPO resulted in similar improvements in hemodynamics and at baseline with both PPH and CHD at 1 year. In CHD the PVR declined from 30% from 17 to 12 Wood units (p<0.001), the systemic arterial O2 saturation improved from 86 to 91% (p<0.004), and the event time improved 118% from 228 to 492 seconds (p=0.023). The 1, 3, and 5 year survivals were 93%, 88%, and 81% for CHD versus 64%, 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.

1097-99 Basal Pulmonary Vascular Resistance and Its Response to Exogenous Nitric Oxide Late After Fontan Operation

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Background: Baseline pulmonary vascular resistance (PVR) fails with exogenous nitric oxide late after Fontan-type operation, even in patients with some pulsatility in the pulmonary circulation. Therapeutic strategies to enhance pulmonary endothelial NO release may have a role in these patients.

Methods: PVR fails with exogenous nitric oxide late after Fontan-type operation. These data suggest that endogenous NO production may be reduced late after Fontan-type operation, even in patients with some pulsatility in the pulmonary circulation. Therapeutic strategies to enhance pulmonary endothelial NO release may have a role in these patients.

1097-10I 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, cyclosporin (CMX) 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 less than 18 years of age who had undergone cardiac transplantation at Loma Linda University Children’s Hospital during the years 1985-2001 were retrospectively reviewed.

Results: A total of 358 infants and children underwent orthotopic cardiac transplantation. Overall survival is 70%. Forty-three patients (12.1%) developed PTCAD. The average age at 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 (1.06 vs. 0.43, p<0.001). There was a lower incidence of CMX found in the PTCAD group than in those 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 (3.67 vs. 5.10 days, p=0.05). Factors not associated with PTCAD included: ischemic time below 180 minutes, gender, mismatch, CMX type, donor to recipient weight ratio, and left ventricle end-diastolic pressure, days on mechanical ventilation in the post-operative period, CMX status of the recipients, or HLA tissue typing mismatches.

Conclusion: PTCAD occurred in pediatric cardiac transplant recipients with significantly increased mortality. Further increases morbidity. PTCAD is associated with the number of rejection episodes. The role of CMX infection remains unclear. PTCAD is not associated with ischemic time, as has been previously suggested. Early and frequent coronary artery evaluation in children with frequent rejection episodes may be warranted.