Blalock-Tausig Shunt in a Neonate with Pulmonary Atresia and Intact Ventricular Septum and Right Aortocoronary Atresia with Right-Ventricular-Dependent Coronary Circulation

Jerman J, Spaeth J
Cincinnati Children’s Hospital Medical Center, Cincinnati, OH, USA

Introduction: Patients with pulmonary atresia and intact ventricular septum (PA/IVS) account for 3% of congenital heart disease and present a significant challenge to the congenital cardiac team. In 50% of patients with PA/IVS, RV sinusoids form and can lead to RV-to-coronary fistulas that allow deoxygenated blood to perfuse the myocardium. In 20%, anterograde coronary bloodstream may be absent. In 3-34%, aortocoronary connections are absent, causing complete RV-dependent coronary circulation (RVDCC). The overall mortality in untreated patients is high and ranges between 50-100%. The appropriate treatment pathway for these patients is debated. We present a patient with PA/IVS-RVDCC who underwent a BT shunt.

Case Report: Our patient was a term neonate who was found to have hypoxemia after birth despite 100% FiO2. A TTE demonstrated PA/IVS with a TV z-score of -3.3. The cath study revealed a hypoplastic TV, atretic RCA orifice, multiple coronary stenoses, and suprasystemic RV pressures. The decision was made to pursue single ventricle palliation. She was taken to the OR for a BT shunt and PDA ligation. Anesthesia was induced with fentanyl and pancuronium. The patient exhibited stable hemodynamics during the off pump procedure, requiring a low dose epinephrine infusion. After arrival to the CICU, the patient developed hypotension requiring vasopressin and calcium infusions. Repeat cardiac markers revealed an initial rise in the CK-MB and Trop I levels which returned to baseline POD 1.

Discussion: The determinants of the most appropriate treatment pathway depend on the degree of RV and TV hypoplasia, presence of RVDCC, and degree of TR. Many institutions will perform right ventricle decompression (RVD) unless RVDCC is noted on cath. However, Giglia et al presented a series of patients with RV-to-coronary fistulas where RVD was performed in 16. Seven had fistulas and no coronary stenosis and all survived. Six had stenosis of a single coronary artery and 4 survived. The remaining 3 had stenosis and/or occlusion of both the RCA and LAD. All died after RVD from acute LV failure. Guleserian et al described 32 patients with PA/IVS-RVDCC who all underwent BTS. Overall mortality at 5 yrs was 18% with all deaths occurring within 3 months of BTS. Coronary atresia was associated with 100% mortality, concluding that single-ventricle palliation yields excellent long-term survival except for those with coronary atresia. Reference 1. Our patient had complete RVDCC with RCA atresia. We performed a BT shunt given her RVDCC with the goal of pursuing Fontan vs transplantation in the future. Anesthetic considerations include maintaining RV pressure to assure adequate coronary perfusion and close monitoring for evidence of ischemia. Significant decreases in preload or afterload may be fatal.

References:
2. Andropoulos. Anesthesia for Congenital Heart Disease, ed 2.