The Earlier, The Better: The Role of ABO-Incompatible Heart Transplantation in the Treatment of Newborn Complex Congenital Heart Disease

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Background

With advances in neonatal cardiac surgical repairs and palliative procedures, the use of heart transplantation for the treatment of congenital heart defects in the newborn period is uncommon. There are a few congenital heart defects that are considered to be incompatible with surgical repair or palliation: pulmonary atresia with intact ventricular septum and right ventricular dependent coronary circulation (PA/IVS, RVDCC), transposition of the great arteries with a single ventricle and heart block, complex heterotaxy syndromes, severe atrioventricular canal or semilunar valve defects, and in some centers, hypoplastic left heart syndrome. Actuarial graft survival in newborn heart transplant recipients is approximately 59% at age 25.

We describe a case of ABO incompatible heart transplant in a newborn with PA/IVS, RVDCC. PA/IVS is a rare congenital cardiac malformation that comprises approximately 1 – 1.5% of all congenital heart defects. 50% of these patients may have RV sinusoids with concomitant coronary artery to RV fistulas. A smaller subset of patients have a severe form where there is absent anterograde aortocoronary flow, and instead have RVDCC. The myocardium in these patients is supplied by desaturated blood from the RV, often resulting in catastrophic myocardial ischemia. RVDCC is present in approximately 9-34% of patients with PA/IVS. Clinical management of these patients can be difficult, and due to their high early mortality rate after single ventricle palliative surgery, it has been recommended for these patients to proceed directly to heart transplantation.

Case Presentation

Our patient is an ex-38.4 week male born via repeat c-section with APGAR scores of 9 and 9. Around 8 minutes of life baby's color turned dusky with SpO2 80's on room air with poor response to oxygenation. Transthoracic echocardiogram showed PA/IVS, a patent foramen ovale with right to left flow, a hypertrophied and hypoplastic RV with moderate dysfunction and a hypoplastic tricuspid valve. The LV had normal size and function. RV coronary sinusoids were seen with severely dilated left main and left anterior descending arteries with fistulous connection to the RV outflow.

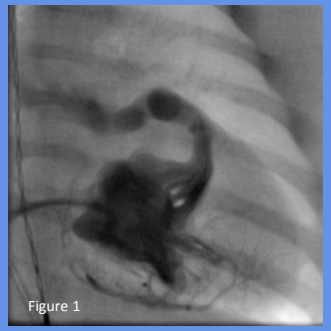
Figure 1

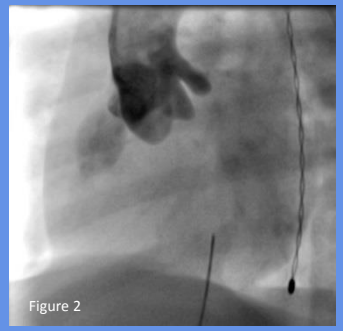
Figure 2

Management

Cardiac catheterization showed supra-systemic RV pressures and RVDCC. The right coronary artery was atretic with filling only from the RV, and there was a large coronary fistula between the left coronary artery (LCA) and right ventricular outflow tract (Figure 1). There were small branches of the LCA arising from the fistula, but the LV did not have robust coronary circulation likely due to steal from the fistula (Figure 2). On DOL 4 the patient became tachycardic with ST-segment depressions and rising troponins, with multiple episodes of bradycardia. Extracorporeal membrane oxygenation in this case was contraindicated due to obligatory ductal shunting and coronary steal. In the setting of myocardial ischemia related to coronary steal, the patient was immediately listed for heart transplantation as UNOS 1A across all blood types.

The patient underwent ABO-incompatible bi-atrial orthotopic heart transplant on DOL 12 and weighed 3.8 kg. Cardiopulmonary bypass (CPB) was complicated by the need for an exchange transfusion to achieve IgM anti -A antibody titers less than 1:4. Exchange transfusion was repeated prior to releasing the aortic cross-clamp. The anti-A titers were 1:1 at separation from CPB. CPB time was 136 minutes, cross-clamp time 63 minutes, organ ischemic time 219 minutes. The patient received packed red blood cells 300cc, platelets 100cc, cryoprecipitate 60cc. The patient was transported to PICU paced at 140, on epinephrine, norepinephrine, and milrinone infusions. The patient is currently 9 months post-transplant and growing well.

Discussion

Due to the limited availability of donor organs, the transplantation of ABO incompatible hearts has become an acceptable approach in infants less than 15 months of age. There have been several studies that show similar early survival for ABO incompatible transplants as in ABO compatible transplants. It is believed that newborn infants do not produce sufficient isohemagglutinins (serum anti-A or anti-B antibodies) to mount a hyperacute rejection following transplant until approximately 12 to 14 months of age.

Allowing ABO incompatible heart transplantation increases organ availability in young patients and allows for use of organs from donors who have less common blood types. In summary, heart transplantation in the neonatal patient population poses unique challenges but can be the only chance of survival for some neonates with congenital defects without an option for repair or palliation.

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