Transposition of the Great Arteries

Jacqui Benner, MD PGY-1
Physiology

• Formation of two parallel circuits $\rightarrow$ incompatible with life unless...
  • VSD or ASD or PFO or PDA or bronchopulmonary collateral circulation
• Interatrial shunting most important due to ability to have bidirectional flow
• Large VSD may have mild or inapparent cyanosis
• Reverse differential cyanosis between pre and post ductal sats
  • Higher post ductal sats due to right to left shunting from pulmonary artery into descending aorta
Pathophysiology

• Saturations at birth 30-50% $\rightarrow$ metabolic acidosis and hypoxemia $\rightarrow$ myocardial tissue damage

• Physiologic decrease in PVR $\rightarrow$ pulmonary overcirculation $\rightarrow$ left heart failure in 1$^{st}$ WOL
Diagnosis

• More difficult to diagnose on fetal ultrasound due to absence of size discrepancy between ventricles on four chamber view
  • More accurate with outflow tract evaluation to determine crossing

• Echocardiogram
  • Delineation of coronary arteries – may need angiography if unable to see

• CXR may or may not show cardiomegaly and increased pulmonary vascularity

• ECG may show biventricular hypertrophy or right ventricular hypertrophy (normal in first few days of life)
Associated Cardiac Anomalies

• Complex vs Simple D-TGA (w/ or w/out other anomalies)
• VSD – 50% of patients
  • Affects clinical presentation
• Left ventricular outflow tract obstruction – 25%
  • Pts with intact IVS – systemic right ventricular pressures → bowing of IVS causing obstruction of mitral valve – rare in neonates
  • Pts with VSD have increased risk of pulmonary stenosis or atresia and aortic arch obstruction (coarctation or interruption)
• Mitral and Tricuspid valve – important when considering surgery
• Coronary Artery Variations – important to delineate prior to surgery
Large VSD

- Minimal cyanosis
- Eventual left heart failure as PVR drops
- CXR with cardiomegaly and increased pulmonary vascular markings
- ECG may show biventricular hypertrophy
Pulmonary Stenosis

- Inadequate pulmonary blood flow
- Severe hypoxemia and acidosis
- No heart failure
- Normal chest xray
Initial Postnatal Management

• Focus on maintaining sufficient mixing between circuits
  • PGE1 – alprostadil – major side effect is apnea and hypotension
  • Balloon atrial septostomy – for severe hypoxemia - if there is adequate mixing, may be able to discontinue PGE1
Surgical Correction

- Mortality without surgical correction is 90%, with surgery <5%
- Usually performed in the first two weeks of life
  - One case series showed increased morbidity and healthcare costs when delayed past three days of life
  - One small study showed delay of correction > 2 weeks associated with reduced brain growth and lower language scores at 18 months (poor follow-up)
- Atrial Switch Procedure vs Arterial Switch Procedure (ASO) vs Rastelli procedure
Atrial Switch Procedure

• SVC and IVC rerouted to L atrium
• Pulmonary veins rerouted to R atrium
• Eventual heart failure
• Atrial arrhythmias
Arterial Switch Procedure

• First performed in 1975!! Preferred procedure since late 80s
• Transection of great arteries at root and reimplantation
• Requires reimplantation of coronary arteries
• LeCompte maneuver – pulm a. anterior to aorta – decreases risk of pulm stenosis
Rastelli procedure

• First described in 1969!!!
• Performed in pts with D-TGA and large VSD and LVOT obstruction
  • Based on LVOT obstruction and status of pulmonary (neo-aortic) valve

B. Aorta connected to LV via VSD
  • VSD may need to be enlarged → increased risk of complete heart block

C. Conduit from right ventricle to pulmonary artery
Complications

• 5-25% require reintervention

• Pulmonary artery stenosis – most common
  • Usually supravalvular, may be branch pulmonary a. stenosis from tension

• Coronary artery stenosis or insufficiency
  • 89% in first 3 months due to anatomic obstruction

• Neo-aortic root dilation or valvular regurgitation
  • Regurgitation usually not clinically significant

• Rastelli’s requires serially replacement of conduits as child grows
Outcome

• **Sports Participation**
  - Those who had ASO and have normal function and are asymptomatic can participate in all sports
  - Those who had atrial switch and are asymptomatic can participate in low-moderate activity sports
  - Exercise capacity with ASO at low-normal (91%) and Rastelli ~ 75%

• 20 year survival rate 95-97% with ASO and 80% with atrial switch, 80% 10 year survival with Rastelli

• **Neurodevelopmental**

• Study between 1983 and 1999 for ASO - 93 percent free from major cardiovascular adverse events (arrhythmia, cerebrovascular accident, heart failure-related hospitalization, or cardiovascular death) at 25 years outcomes
Resources


