**Transposition of the Great Arteries**  
*(TGA, d-loop)*

Mary Rummell, MN, RN, CPNP, CNS  
Clinical Nurse Specialist, Pediatric Cardiology/Cardiac Surgery  
Doernbecher Children’s Hospital, Oregon Health and Science University  
Portland, Oregon

I. Embryology
   A. Most common cyanotic congenital heart disease - 5% of all newborns with congenital heart disease
   B. Normal development of ventricular situs (Moore, 2008)
      1. Occurs during the 5th week of gestation
      2. Twisting of the primordial heart tube to right (d-looping)
         a. Places eventual morphologic right ventricle on right side of heart
         b. Places eventual morphologic left ventricle on left side of heart
         c. Brings atrium to right and posterior of ventricles
   C. Normal development of the great arteries
      1. Occurs during 5th-6th week of gestation
      2. Genetically influenced by neural crest cells
      3. Formed from common trunk at the top of the fetal heart
         a. Common trunk consists of bulbus cordis and truncus arteriosus (TA)
         b. Tissue growth and blood flow creates spiral septation
            (1) Blood flow streams from the ventricles
            (2) Tissue ridges grow within the trunk
         c. Spiral septation creates two arteries
            (1) Pulmonary artery exits from the morphologic right ventricle
            (2) Aorta exits from the morphologic left ventricle
   C. Abnormal development (d-TGA) results from failure in spiral septation of truncus arteriosus

II. Anatomy (Warnes, 2006)
   A. Abnormal septation
      1. Spiral failure in septation of great arteries
         a. Pulmonary artery exits from morphologic left ventricle
            (As indicated by #2 in illustration below)
         b. Aorta exits from morphologic right ventricle
            (As indicated by #1 in illustration below)
         c. Abnormal relationship
            (1) Usually parallel
            (2) Aorta anterior and to right of the pulmonary artery
      2. Results in parallel circuits
   B. Communication between the pulmonary and systemic circuits
      1. Required for survival
      2. Three levels
         a. Only one essential
         b. Great artery level with a patent ductus arteriosus (PDA)
            (As indicated by #3 in illustration below)
         c. Atrial level with an atrial septal defect (ASD)
            (As indicated by #4 in illustration below)
Transposition of the Great Arteries (d-TGA)


d. Ventricular level with a ventricular septal defect (VSD)  
   (As indicated in illustration below)

Transposition of the Great Arteries with a VSD

C. Associated lesions (Warnes, 2006)
   1. Present in one-third of patients.
   2. Include
      a. Ventricular septal defects (VSD) - 50%
      b. Pulmonary outflow tract obstruction - less than half
      b. Coarctation of the aorta - 5% of associated lesions

III. Physiology (Love, 2008)
   A. Parallel circulation
      1. Right side
         a. Desaturated blood from body returns to right atrium
         b. Right ventricle pumps desaturated blood through aorta back to body
      2. Left side
         a. Oxygenated blood from lungs returns to left atrium
         b. Left ventricle pumps oxygenated blood through pulmonary artery back to lungs
   B. Survival depends on mixing of the systemic and pulmonary circulations
      1. Through associated lesions
      2. Medical/surgical interventions
         a. Completed within first hours of life
         b. Medical - maintain open ductus arteriosus with intravenous prostaglandins
         c. Cardiac catheterization
            (1) Rashkind balloon atrial septostomy – 1966
            (2) Balloon catheter passed thru atrial septum via a patent foramen ovale (PFO), balloon inflated in left atrium, pulled through PFO to tear a hole in atrial septum
         d. Surgery
            (1) Blalock-Hanlon procedure – 1950
            (2) Excision of atrial septum

IV. Type of Repair
   A. Atrial Switch Surgery (two wrongs make a right)
      1. Usually completed by one year of age
      2. Procedures (Love, 2008; Warnes, 2006)
         a. Senning Procedure – 1954
            (1) Creation of baffle within the atrium from atrial tissue to direct venous return to the contralateral ventricle
            (2) Systemic venous blood directed through the tricuspid valve into the anatomic and morphologic right ventricle
            (3) Pulmonary venous blood directed through the mitral valve into the anatomic and morphologic left ventricle
         b. Mustard Procedure – 1964
            (1) Creation of baffle within the atrium with treated pericardium to direct venous return to the contralateral ventricle
            (See illustration below)
3. Ventricular function
   a. Right ventricle becomes systemic ventricle
   b. Left ventricle becomes pulmonic ventricle
   c. Major influence on long-term outcomes
4. Outcomes (Meijboom, 2009)
   a. Excellent midterm results
   b. Significant long term issues
      (1) Right ventricular failure
      (2) Significant rhythm disturbances
      (3) Sudden death
      (4) Baffle obstruction, leak, and calcification

B. Arterial Switch
1. Timing (Warnes, 2006; Mavroudis, 2003)
   a. Optimally within first two weeks of life
      (1) Before left ventricular muscle is deconditioned to maintain LV
          function and contractility
      (2) After pulmonary arterial pressure and resistance have decreased
          from fetal levels to decrease incidence of pulmonary hypertensive
          crises
      (3) After allowing for some maturation of the neonatal myocardium
   b. After two weeks of life
      (1) Evaluation of left ventricular function
      (2) Consider conditioning of left ventricle
   a. Restores normal anatomic relationship between great arteries and ventricles
   b. Great arteries transected and re-anastomosed above semilunar valves to appropriate ventricle (As indicated by suture line in illustration below)
   c. Coronary arteries moved to neo-aorta
   d. Ligation of ductus arteriosus (As indicated by #1 in illustration below)
   e. Closure of foramen ovale/enlarged by balloon septostomy (As indicated by #2 in illustration below)

Arterial Switch Repair


3. Outcomes
   a. Complex neonatal surgery
      (1) Depends on skill of surgeon
      (2) Procedure of choice since 1980s
   b. Long-term problems
      (1) Coronary artery stenosis
      (2) Distortion/stenosis of pulmonary arteries
      (3) Dilation of neo-aortic root
      (4) Aortic valve regurgitation

C. Rastelli Procedure
   1. Timing
      a. Use: d-TGA associated with a large subaortic VSD and pulmonary valve stenosis
      b. Depends on pulmonary blood flow and ventricular function of patient
   2. Procedure (Mavroudis, 2003; Warnes, 2006)
      a. Patch placed to direct blood through the VSD to the aorta (As indicated by #2 in illustration below)
b. Pulmonary artery divided, pulmonary valve over-sewn, RV connected to main PA with valved conduit ((As indicated by #1 in illustration below)
c. Morphologic left ventricle pumps to systemic circulation

3. Outcomes
   a. Requires re-operation for conduit replacement due to conduit stenosis, calcification, degeneration

![Rastelli Procedure](image)

Illustrations reprinted from PedHeart Resource, [www.HeartPassport.com](http://www.HeartPassport.com), © Scientific Software Solutions, 2010. All rights reserved

V. Long Term Complications/Interventions (Alonso-Gonzales et al, 2010; Love, 2001)
   A. Arrhythmias (Refer to Problem Section on Arrhythmias for further discussion and management)
      1. Supraventricular arrhythmias
         a. Atrial fibrillation
         b. Atrial flutter
         c. Other atrial tachycardias (Paroxysmal or persistent)
      2. Ventricular tachycardia
         a. Sustained or not sustained
         b. Related to progressive RV deterioration
      3. Associated with pacemaker placement
         a. Sick sinus syndrome
         b. Complete heart block
         c. Related to medical treatment for atrial tachyarrhythmias
         d. Bradycardia
      4. Sudden death – factors that increase risk
         a. Complex TGA involving ventricular septal defect
         b. Arrhythmias in operative period
         c. History of Supraventricular arrhythmias
d. Advanced New York Heart Association functional classification

e. RV dysfunction with broad QRS complex

B. Ventricle failure (See Problem Section on Systemic Ventricular Failure for further discussion and management)
1. Systemic ventricle is morphologic right ventricle
2. Atrioventricular valve regurgitation increases with RV dysfunction

C. Baffle leaks/obstruction
1. Neurological event
2. Superior vena cava (SVC) obstruction
3. Pulmonary venous obstruction/maybe cause of pulmonary artery hypertension

D. Pulmonary hypertension (See Problem Section on Pulmonary Hypertension for further discussion and management)

VI. Routine cardiology care (Love, 2008; Meijboom, 2009, Schwerzmann, 2009; Warnes, 2006)

A. Annual:
1. Clinical evaluation
2. Electrocardiogram
3. Transthoracic echo and/or MRI
4. Consideration of annual Holter Monitor
   a. Bradycardia (Marked or Symptomatic)
   b. Palpitations
   c. Presyncope/syncope

B. Every 2-4 years: Cardiac magnetic resonance (CMR) imaging or radionuclide angiography (in patients with pacemakers)

VII. Care during pregnancy (Refer to problem section on pregnancy in adults with CHD) (Canobbio, 2006; Warnes, 2006)

A. Recommendations
1. Consultation with cardiologist with adult congenital heart disease experience before pregnancy
2. Scheduled cardiology evaluation and follow-up during pregnancy
3. Multidisciplinary coordination for labor, delivery, and post-partum periods

B. Considerations during pregnancy
1. Risks increase with presence of significant hemodynamic lesions and functional capacity
2. Right ventricular (RV) function
   a. Long-term effect of pregnancy on RV function unclear
   b. Assess for early signs/symptoms of heart failure and arrhythmias
3. Use of aspirin in patients with history of atrial arrhythmias
4. Antibiotic prophylaxis for infective endocarditis at the time of rupture of membranes for vaginal delivery

References:


9/2011