Transposition of the Great Arteries  
(TGA, d-loop)  
What the Nurse Caring for a Patient with CHD Needs to Know

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

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

![Atrial level with an atrial septal defect (ASD)](image1)

Transposition of the Great Arteries (d-TGA)

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- **Ventricular level** with a ventricular septal defect (VSD)
  (As indicated in illustration below)

![Ventricular level with a ventricular septal defect (VSD)](image2)

Transposition of the Great Arteries with a VSD

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

- **Physiology** (Nieves, 2013, Love, 2008)
  - Parallel circulation
    - Right side
      - Desaturated blood from body returns to right atrium (RA)
      - Right sided ventricle pumps desaturated blood through aorta back to body
    - Left side
      - Oxygenated blood from lungs returns to left atrium (LA)
      - Left sided ventricle pumps oxygenated blood through pulmonary artery back to lungs
  - Survival depends on mixing of the systemic and pulmonary circulations
    - Through associated lesions
      - PDA
      - ASD
      - If present, VSD
    - Medical/surgical interventions
      - May be indicated within the first few hours of life
      - Medical management to maintain an open ductus arteriosus
        - Intravenous prostaglandin E₁ (IVPGE)
      - Cardiac catheterization
        - Indicated in severely hypoxemia
          - Even with IVPGE
          - Inadequate atrial level communication and insufficient mixing
        - Rashkind balloon atrial septostomy – 1966
        - Balloon catheter passed thru atrial septum via a patent foramen ovalae (PFO), balloon inflated in left atrium, pulled through PFO to tear a hole in atrial septum
        - Monitor for post-procedure complications
          - Bleeding at catheter insertion site
          - Tamponade
          - Arrhythmias
          - Cerebrovascular accidents
    - Surgery
      - Blalock-Hanlon procedure – 1950
      - Excision of atrial septum

- **Type of Surgical Repair**
  - Atrial Switch Surgery (two wrongs make a right)
    - Usually completed by one year of age
    - Procedures (Love, 2008; Warnes, 2006)
      - Senning Procedure – 1954
- Creation of baffle within the atrium from atrial tissue to direct venous return to the contralateral ventricle
- Systemic venous blood directed through the tricuspid valve into the anatomic and morphologic right ventricle
- Pulmonary venous blood directed through the mitral valve into the anatomic and morphologic left ventricle

**Mustard Procedure – 1964**
- Creation of baffle within the atrium with treated pericardium to direct venous return to the contralateral ventricle
  (See illustration below)

**Ventricular function**
- Right ventricle becomes systemic ventricle
- Left ventricle becomes pulmonic ventricle
- Major influence on long-term outcomes

**Outcomes (Meijboom, 2009)**
- Excellent midterm results
- Significant long term issues
  - Right ventricular failure
  - Significant rhythm disturbances
  - Sudden death
  - Baffle obstruction, leak, and calcification

**Arterial Switch (Nieves, 2013; Warnes, 2006; Mavroudis, 2003)**
- Timing
  - Optimally within first two weeks of life

Before left ventricular muscle is deconditioned to maintain LV function and contractility
After pulmonary arterial pressure and resistance have decreased from fetal levels to decrease incidence of pulmonary hypertensive crises
After allowing for some maturation of the neonatal myocardium
  - After two weeks of life
  - Evaluation of left ventricular function
  - Consider conditioning of left ventricle

Procedure - Jatene in 1976 with improvements in operative technique by Lecompte in 1981
  - Restores normal anatomic relationship between great arteries and ventricles
  - Great arteries transected and re-anastomosed above semilunar valves to appropriate ventricle (As indicated by suture line in illustration below)
  - Coronary arteries moved to neo-aorta
  - Ligation of ductus arteriosus (As indicated by #1 in illustration below)
  - Closure of foramen ovalae/enlarged by balloon septostomy (As indicated by #2 in illustration below)

Arterial Switch Repair

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- Outcomes
  - Complex neonatal surgery
    - Depends on skill of surgeon
    - Procedure of choice since 1980s
  - Long-term problems
    - Coronary artery stenosis
    - Distortion/stenosis of pulmonary arteries
    - Dilation of neo-aortic root
    - Aortic valve regurgitation
- Developmental/intellectual delay

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

Rastelli Procedure

**Specific Neonatal Considerations**
- Early medical management
  - Maintain patent ductus arteriosus with intravenous prostaglandin E₁
    - Increases pulmonary blood flow to increase pulmonary venous return
    - Increase LA pressure and left-to-right shunting at atrial level
    - May or may not benefit simple d-TGA without VSD or LV outflow obstruction
  - Hypoxia (PaO₂<30mmHg)
    - Monitor for adequate ventilation
    - Monitor for anemia
    - May require supplemental oxygen
    - Acidosis
- Monitor/maintain pH
- Provide intravenous fluid replacement
- Administer sodium bicarbonate
  - Pulmonary edema
    - May develop with severe hypoxemia, inadequate atrial mixing
    - May require mechanical ventilation
  - Neurologic injury
    - Air embolus
      - Systemic venous return exits RV through aorta, cerebral arteries arise from aortic arch
      - Assure that IV tubing has no air
      - Use 2mm air filter on all IV lines
    - Thrombus or ischemia from hypoxia
  - Minimize oxygen consumption
    - Maintain Normothermia
    - Decrease environmental stimulation
    - Manage pain and anxiety
- Interventions – see above catheterization and surgical procedures
- Post-operative management
  - See Peds/Neo Guidelines for Post-operative Care
  - Specific monitoring for:
    - Coronary artery kinking, stretching, occlusion
      - Monitor EKG for ST segment
      - Monitor for arrhythmias
    - Ventricular function
      - Arrhythmias
      - Low cardiac output syndrome
    - Residual lesions
      - New murmur
      - cyanosis
    - Systemic hypertension
    - Pulmonary hypertension (See Peds/Neo Guidelines on Pulmonary Hypertension)

**Long Term Complications/Interventions** (Alonso-Gonzales et al, 2010; Love, 2001)

**Arterial Switch Procedures**
- Coronary artery obstruction
  - Stenosis, kinking, clots, occlusion/compression from pulmonary artery
  - May cause ventricular dysfunction
  - Requires intervention either interventional catheterization or surgical
  - May require transplant
- Supra-valvar stenosis
  - PA stenosis most frequent cause for re-intervention at any age
  - Usually related to suture lines at site of arterial anastomosis
  - Usually requires surgical intervention with patch angioplasty
- Dilation of aortic valve

**Atrial Switch Procedures**
- Arrhythmias (Refer to both Peds/Neo and Adult Guidelines on Arrhythmias for further discussion and management)
  - Supraventricular arrhythmias
- Atrial fibrillation
- Atrial flutter
- Other atrial tachycardias (Paroxysmal or persistent)
  - Ventricular tachycardia
    - Sustained or not sustained
    - Related to progressive RV deterioration
  - Associated with pacemaker placement
    - Sick sinus syndrome
    - Complete heart block
    - Related to medical treatment for atrial tachycardias
    - Bradycardia
  - Sudden death – factors that increase risk
    - Complex TGA involving ventricular septal defect
    - Arrhythmias in operative period
    - History of supraventricular arrhythmias
    - Advanced New York Heart Association functional classification
    - RV dysfunction with broad QRS complex
- Ventricle failure (See Adult Guidelines on Systemic Ventricular Failure for further discussion and management)
  - Systemic ventricle is morphologic right ventricle
  - Atrioventricular valve regurgitation increases with RV dysfunction
- Baffle leaks/obstruction
  - Neurological event
  - Superior vena cava (SVC) obstruction
  - Pulmonary venous obstruction/maybe cause of pulmonary artery hypertension
- Pulmonary hypertension (See Peds/Neo and Adult Guidelines on Pulmonary Hypertension for further discussion and management)

**Routine cardiology care** (Marino, 2012; Love, 2008; Meijboom, 2009, Schwerzmann, 2009; Warnes, 2006)
- Complex CHD
- Requires life-long follow-up by a cardiologist trained in congenital heart disease

**Neonatal/Pediatrics**
- Frequency depends on post-operative course/complications
- Early post-operative evaluation
  - Between 1-2 weeks following discharge
  - Should include clinical evaluation, electrocardiogram (EKG) and echocardiogram (ECHO)
- Periodic evaluations
  - Schedule depends on presence of complications
  - Usually every 6 months-1 year
  - Include clinical evaluation, EKG, ECHO
  - Neurodevelopmental assessment and screening

**Adults**
- Annually
  - Clinical evaluation
  - EKG
  - Transthoracic echo and/or MRI
  - Consideration of annual Holter Monitor
    - Bradycardia (Marked or Symptomatic)
• Palpitations
• Presyncope/syncope
• Every 2-4 years: Cardiac magnetic resonance (CMR) imaging or radionuclide angiography (in patients with pacemakers)

**Care during pregnancy** (See Adult guidelines on Pregnancy in adults with CHD) (Canobbio, 2006; Warnes, 2006)

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

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

**References:**


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