# Congenitally Corrected Transposition of the Great Arteries (ccTGA or l-loop TGA)

What the Nurse Caring for a Patient with CHD Needs to Know

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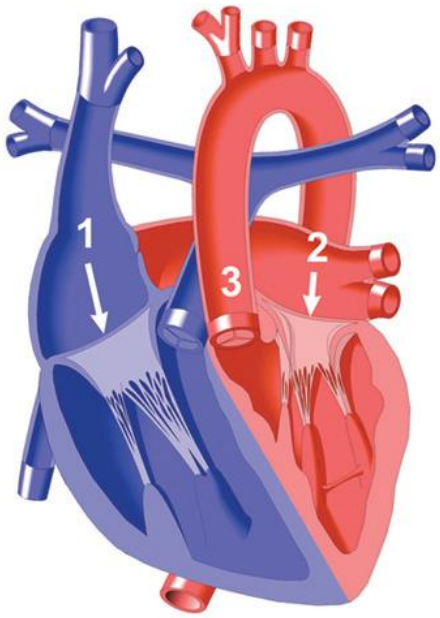
**Embryology**

- Rare congenital heart defect -< 1% of patients 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 (RV) on right side of heart
      - Places eventual morphologic left ventricle (LV) on left side of heart
      - Brings atrium to right and posterior of ventricles
  - Normal development of the great arteries (Moore, 2008)
    - 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 [congenitally corrected transposition of the great arteries (ccTGA) or (l-TGA)] results from looping of the primordial heart tube to the left instead of the right

**Anatomy** (Alonso-Gonzales, 2010; Warnes, 2006)

- Atria in normal anatomic position
- Atrioventricular (AV) and ventriculoarterial (VA) discordance
  - Right heart (As indicated by #1 in illustration below)
    - Right atrium connects to morphologic left ventricle (LV)
    - Pulmonary artery exits from morphologic left ventricle (Pulmonic ventricle)
    - Atrioventricular valve has two leaflets (mitral valve).
  - Left heart (As indicated by #2 in illustration below)
    - Left atrium connects to morphologic right ventricle (RV)
    - Aorta exits from morphologic right ventricle (Systemic ventricle)
    - Atrioventricular valve has three leaflets (tricuspid valve)
- Great arteries abnormally positioned
  - Side by side
  - Aorta may be anterior and to the left (As indicated by #3 in illustration below)




Congenitally Corrected Transposition of the Great Arteries (ccTGA)



- Associated lesions (Thorne, 2009)
  - Present in 95% of patients
  - Include
    - Ebstein-like anomaly of the tricuspid valve (90%)
    - Ventricular septal defects (VSD) - 70%
    - Pulmonary outflow tract obstruction – 40%
    - Complete heart block (2% with 2% increased risk/year)
      - Unusual position of AV node and His bundle
      - May be precipitated by tricuspid valve or VSD surgery
- Situs abnormalities
  - Common – Dextrocardia
  - Suspect ccTGA if abdominal situs solitus

**Physiology** (Alonso-Gonzalez, 2010)

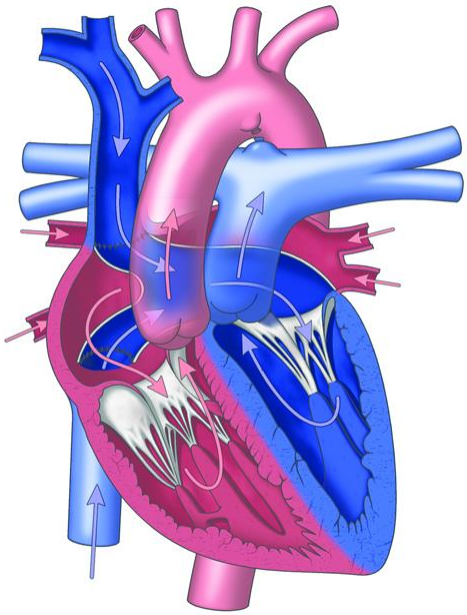
- Blood flow as in normal heart
  - Ventricular morphology transposed
    - Morphologic LV = pulmonic ventricle
    - Morphologic RV = systemic ventricle
  - AV valves correspond to morphology of ventricles
    - Mitral valve
      - Right sided
      - Pulmonic ventricle
    - Tricuspid valve

                - Left sided
                - Systemic ventricle
- Asymptomatic
    - No associated lesions
    - May not be diagnoses until 7th to 8th decade of life, or found on autopsy
    - Symptoms may develop in 3rd or 4th decade of life
        - Usually due to arrhythmias
        - Systemic AV valve regurgitation
        - May include
            - Dyspnea on exertion
            - Syncope secondary to:
                - Atrial arrhythmias
            - Complete heart block
            - Heart failure
                - Diminished ventricular contractility
                    - Ventricular failure
                    - Systemic ventricle morphologically a right ventricle
                - Dilation from AV valve (tricuspid valve) regurgitation
- Symptoms related to associated lesions
    - VSD
        - Usually large, perimembranous
        - L-to-R shunt with normal development of pulmonary vascular bed and decreasing pulmonary vascular resistance
        - Congestive heart failure signs/symptoms
    - Conduction system abnormalities
        - Sinus node normal
        - AV conduction tissue markedly abnormal
        - Complete heart block – 30%
            - May be present at birth
            - Develops at rate of 2%/year
        - Other arrhythmias
            - Sick sinus syndrome
            - Atrial arrhythmias
            - Reentrant AV tachycardia
            - Ventricular tachycardia
    - Left-sided ventricular outflow tract obstruction – 30-50%
        - Decrease cardiac output
        - Decreased ventricular function

**Procedures/Surgical Interventions**

- Cardiac catheterization
    - Indicated for:
        - Hemodynamic assessment
            - Ventricular function
            - Measurement of pulmonary pressures
        - Define coronary artery anatomy
        - Assessment/management of pulmonary stenosis (PS)
        - Device closure of VSD
    - Increased intra- and post-procedure risk of temporary or complete heart block
        - Abnormal conduction pathways

        - Abnormal pulmonary outflow tract
- Surgical Interventions
    - Repair of associated lesions
        - Physiological approach
        - Timing depends upon symptoms
        - AV valve replacement/repair (See Defect Guideline on Mitral and Tricuspid Valve Replacement/Repair)
        - VSD repair (See Defect Guideline for VSD)
    - Double switch (Nieves, 2013; Warnes, 2006)
        - Anatomical restoration
            - Attempt to avoid progressive, long-term problems
            - Prevent systemic ventricular dilation and decrease in function
        - Timing
            - Complex
            - Depends on associated lesions that would impact either atrial or arterial switch
        - Includes both an atrial switch and an arterial switch
            - Atrial Switch - Senning or Mustard Procedure
                - Creation of baffle within the atrium 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

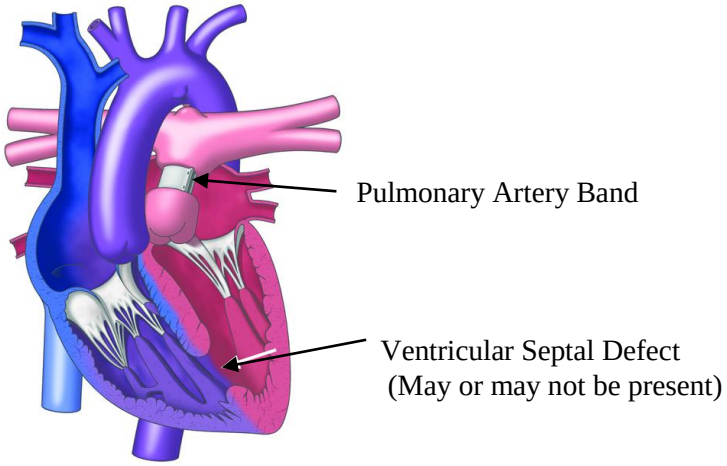


Mustard Procedure



            - Arterial Switch

- Great arteries transected and moved to associated ventricle
    - Aorta arises from left (now systemic) ventricle
    - Pulmonary artery arises from right (now pulmonic) ventricle
- Coronary arteries moved to neo-aorta
- Requires conditioning of morphologic left ventricle to become systemic ventricle
    - May not be necessary with presence of equal ventricular pressures with a VSD and pulmonary stenosis (Illustration below shows pulmonary artery (PA) band and a VSD)
    - Usually done with pulmonary artery band

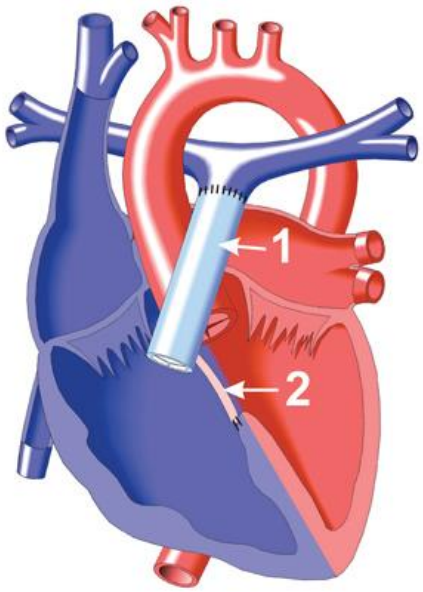



Pulmonary Artery Band prior to Double Switch
Illustration does NOT have ventricles in ccTGA relationship,
Ventricles are in normal anatomic position
Illustration HAS great arteries in transposed position.



- Rastelli Procedure
    - Indication: ccTGA associated with a large subaortic VSD and pulmonary valve stenosis
    - Timing depends on pulmonary blood flow and ventricular function of patient
    - Procedure
        - Patch placed to direct blood through the VSD to the aorta
        - Pulmonary artery connected to the RV with a valved conduit
        - Morphologic left ventricle pumps to systemic circulation
    - Outcomes

- Requires conditioning of LV to become systemic ventricle
- Requires re-operation for conduit replacement due to conduit stenosis, calcification, degeneration




Rastelli Procedure



- Transplantation
  - May occur in both children and adults
  - Indications
    - Systemic ventricular failure
    - Associated defects not amenable to surgical intervention
    - Heart failure not controlled by medical management

**Specific Considerations**

- Periodic follow-up from time of diagnosis
  - Required with or without associated lesions
  - Will determine timing of interventions
- Timing of interventions depends:
  - Presence/severity of associated lesions
  - Medical management of heart failure
  - Right ventricular (systemic ventricle) failure
  - Arrhythmias
    - Complete heart block

        - Atrial arrhythmias
- Increased surgical risk
    - Double switch
    - Ventricular failure
    - Incompetent AV valve
    - Heart block
- Post-operative care
    - Determined by procedure
    - See Peds/Neo Guidelines for post-operative management

**Long Term Complications/Interventions** (Alonso-Gonsales et. al, 2010; Warnes, 2006) (Refer to Ped/Neo and Adult Problem Guidelines for further discussion and management for problems listed below)

- Primary arrhythmia = Heart Block
    - Due to unusual position of AV node and His Bundle
    - Progressive incidence of complete AV block
    - May be precipitated by tricuspid valve or VSD surgery
- Additional arrhythmias
    - Sick sinus syndrome
    - Atrial flutter
    - Re-entrant AV tachycardia
    - Ventricular tachycardia
- Ventricle failure
    - Systemic ventricle has RV morphology
        - Different physiology for contractility
        - Different anatomy for inflow and outflow
            - Ventricular dilation increases AV valve regurgitation
            - Infundibulum increases sub-aortic stenosis
    - Atrioventricular valve regurgitation increases with RV dysfunction
    - Perfusion to RV by single coronary artery

**Routine cardiology care** (Baas, 2014; Thorne, 2009)

- Periodic clinical evaluation
    - Timing depends on associated lesions and interventions
    - Requires assessment of rhythm and ventricular function
        - Electrocardiogram (EKG)
        - Annual Holter Monitor
        - Imaging study
            - Transthoracic echo and/or MRI
            - Cardiac MRI or radionuclide angiography (in patients with pacemakers)
        - Exercise testing
    - Pediatrics
        - Requires even if no associated lesions
        - By pediatric cardiologists
    - Adult
        - At least annually
        - By adult congenital heart disease specialist
- SBE prophylaxis (See 2015 AHA Guidelines for Pediatric and Adult Patients)

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

- Recommendation
    - 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
    - Right ventricular (RV) function
        - Long-term effect of pregnancy on RV function unclear
        - Increased risk of heart failure and arrhythmias with tricuspid valve regurgitation
    - 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

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