

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

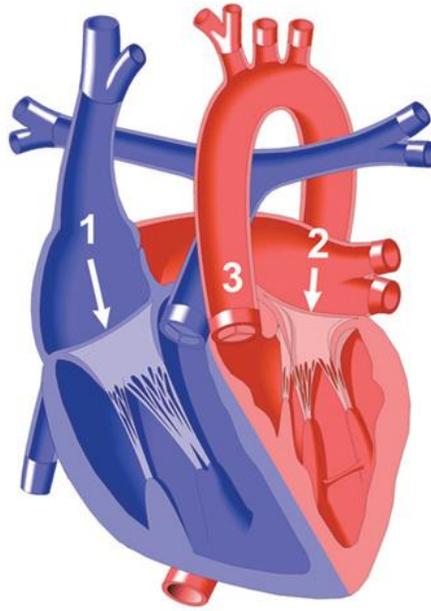
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I. Embryology

- A. Rare congenital heart defect -< 1% of patients 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 (RV) on right side of heart
 - b. Places eventual morphologic left ventricle (LV) on left side of heart
 - c. Brings atrium to right and posterior of ventricles
- C. Normal development of the great arteries (Moore, 2008)
 - 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 [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

II. Anatomy (Alonso-Gonzales, 2010; Warnes, 2006)

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



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C. Associated lesions (Thorne, 2009)

1. Present in 95% of patients.
2. Include
 - a. Ebstein-like anomaly of the tricuspid valve (90%)
 - a. Ventricular septal defects (VSD) - 70%
 - b. Pulmonary outflow tract obstruction – 40%
 - c. Complete heart block (2% with 2% increased risk/year)
 - (1) Unusual position of AV node and His bundle
 - (2) May be precipitated by tricuspid valve or VSD surgery

D. Situs abnormalities

1. Common – Dextrocardia
2. Suspect ccTGA if abdominal situs solitus

III. Physiology (Alonso-Gonzalez, 2010)

A. Asymptomatic if no associated lesions

B. Symptoms

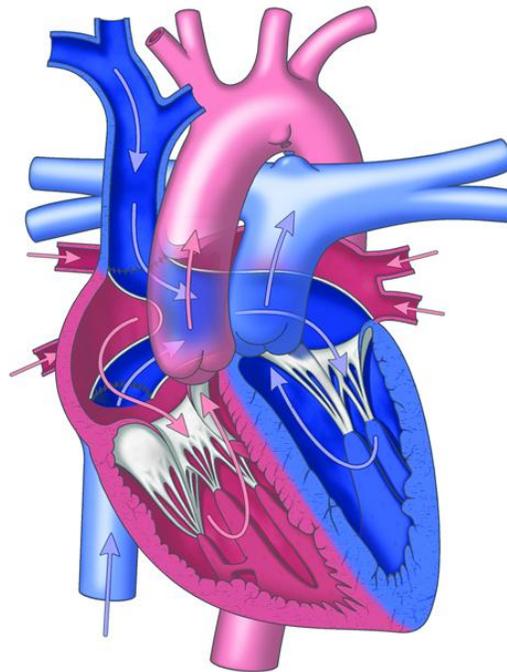
1. Develop in 3rd or 4th decade of life
2. Dyspnea on exertion
3. Syncope secondary to:
 - a. Atrial arrhythmias
 - b. Complete heart block
4. Heart failure
 - a. Ventricular failure - Systemic ventricle is morphologic right ventricle
 - b. AV valve (tricuspid valve) regurgitation

IV. Long-term outcomes/problems (Refer to Problem Section for further discussion and management for problems listed below)

- A. Depends on presence/severity of associated lesions
- B. Right ventricular (systemic ventricle) failure
- C. Arrhythmias
 - 1. Complete heart block
 - 2. Atrial arrhythmias

IV. Type of Repair

- A. Repair of associated lesions
 - 1. AV valve replacement/repair (Refer to section on Valve Replacement/Repair)
 - 2. VSD repair
- B. Double switch (Warnes, 2006)
 - 1. Includes both an atrial switch and an arterial switch
 - a. Atrial Switch - Senning or Mustard Procedure
 - (1) Creation of baffle within the atrium 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



Mustard Procedure

Illustrated with d-TGA where the morphologic right ventricle with the tricuspid valve is on the right and the morphologic left ventricle with the mitral valve is on the left. In ccTGA, when the Mustard Procedure is accompanied by an arterial switch operation, the aorta is moved from the anatomic right ventricle on the left to the anatomic left ventricle on the right. In the end the aorta exits the morphologic left ventricle which is on the right side of the heart, and the pulmonary artery arises from morphologic right ventricle on

the left side of the heart. The great arteries will appear as illustrated above, but will arise from the morphologically “correct” ventricle.

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b. Arterial Switch

(1) Great arteries transected and moved to associated ventricle

* Aorta arises from left (now systemic) ventricle

* Pulmonary artery arises from right (now pulmonic) ventricle

(2) Coronary arteries moved to neo-aorta

2. Requires conditioning of morphologic left ventricle to become systemic ventricle

a. 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)

b. Usually done with pulmonary artery band

C. Rastelli Procedure

1. Timing

a. Use: in ccTGA associated with a large subaortic VSD and pulmonary valve stenosis

b. Depends on pulmonary blood flow and ventricular function of patient

2. Procedure

a. Patch placed to direct blood through the VSD to the aorta

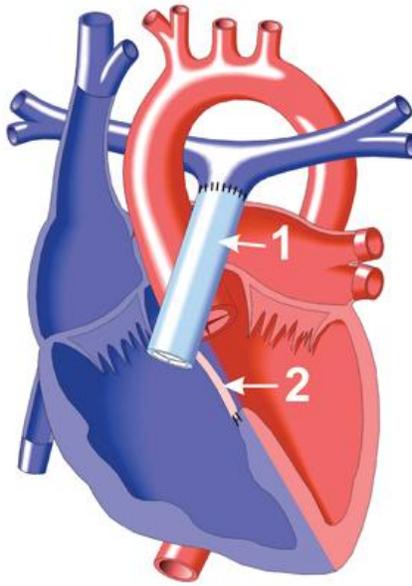
b. Pulmonary artery connected to the RV with a valved conduit

c. Morphologic left ventricle pumps to systemic circulation

3. Outcomes

a. Requires conditioning of LV to become systemic ventricle

b. Requires re-operation for conduit replacement due to conduit stenosis, calcification, degeneration



Rastelli Procedure

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D. Transplantation

V. Long Term Complications/Interventions (Alonso-Gonsales et. al, 2010; Warnes, 2006)

A. Primary Arrhythmia - Heart Block : (Refer to Problem Section on Arrhythmias for further discussion and management)

1. Due to unusual position of AV node and His Bundle
2. Progressive incidence of complete AV block
3. May be precipitated by tricuspid valve or VSD surgery

B. Ventricle failure (Refer to 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
3. Perfusion to RV by single coronary artery

C. Increased surgical risk

1. Double switch
2. Ventricular failure
3. Incompetent AV valve
4. Heart block

VI. Routine cardiology care includes periodic evaluation (Thorne, 2009)

- A. Annual clinical evaluation by a specialist in adult congenital heart disease
- B. Electrocardiogram
- C. Annual Holter Monitor
- D. Imaging study
 1. Transthoracic echo and/or MRI

2. Cardiac MRI or radionuclide angiography (in patients with pacemakers)
- E. Exercise testing

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

A. Recommendation

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. Right ventricular (RV) function
 - a. Long-term effect of pregnancy on RV function unclear
 - b. Increased risk of heart failure and arrhythmias with tricuspid valve regurgitation
2. Use of aspirin in patients with history of atrial arrhythmias
3. Antibiotic prophylaxis for infective endocarditis at the time of rupture of membranes for vaginal delivery

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