

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

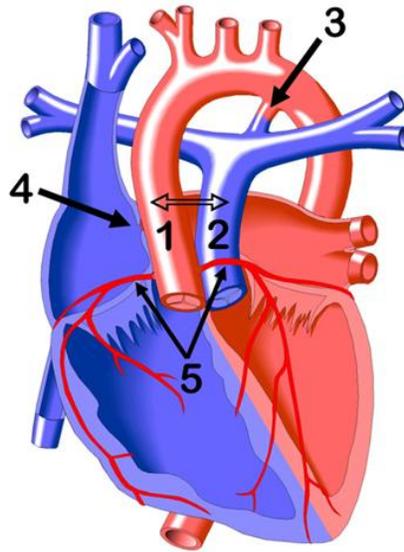
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## I. Embryology

- 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 5<sup>th</sup> 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 5<sup>th</sup>-6<sup>th</sup> 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)

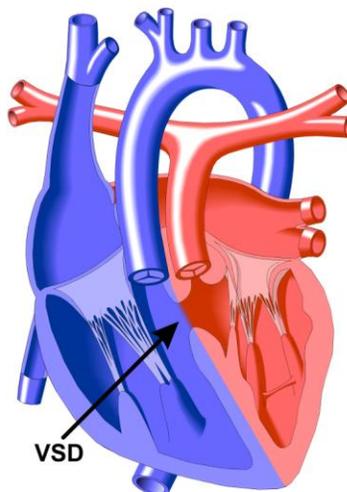
- A. Abnormal septation
  - 1. Spiral failure in septation of great arteries
    - a. Pulmonary artery exits from morphologic left ventricle  
(As indicated by #2 in illustration below)
    - b. Aorta exits from morphologic right ventricle  
(As indicated by #1 in illustration below)
    - c. Abnormal 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)

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



Transposition of the Great Arteries with a VSD

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### 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

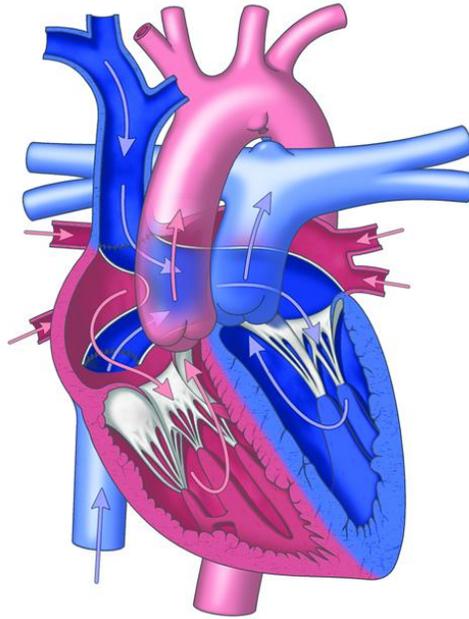
#### B. Survival depends on mixing of the systemic and pulmonary circulations

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)



Mustard Procedure

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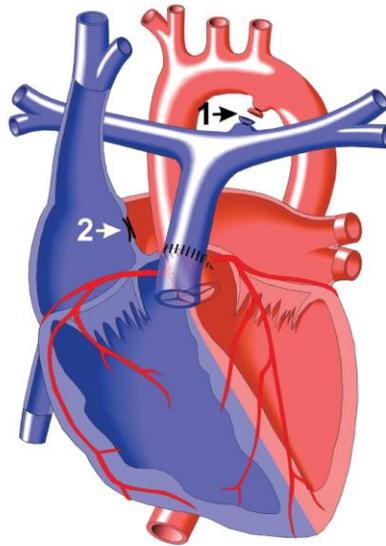
3. Ventricular function
  - a. Right ventricle becomes systemic ventricle
  - b. Left ventricle becomes pulmonic ventricle
  - c. Major influence on long-term outcomes
4. Outcomes (Meijboom, 2009)
  - a. Excellent midterm results
  - b. Significant long term issues
    - (1) Right ventricular failure
    - (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

2. Procedure - Jatene in 1976 with improvements in operative technique by Lecompte in 1981

- 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

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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

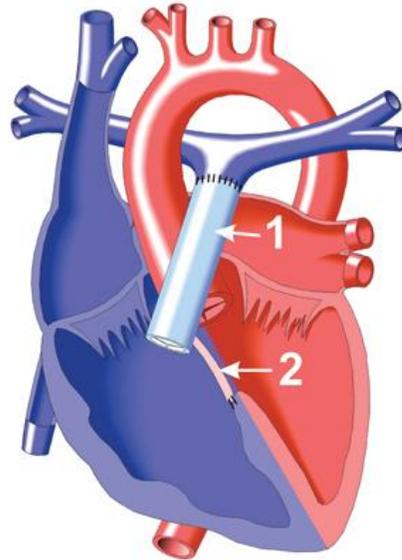
1. Timing

- 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

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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

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9/2011