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

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

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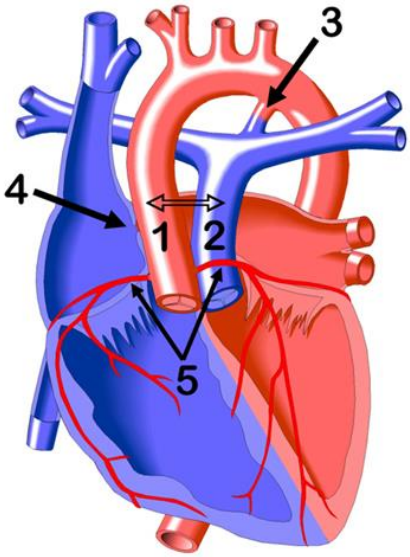
Embryology

- 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)

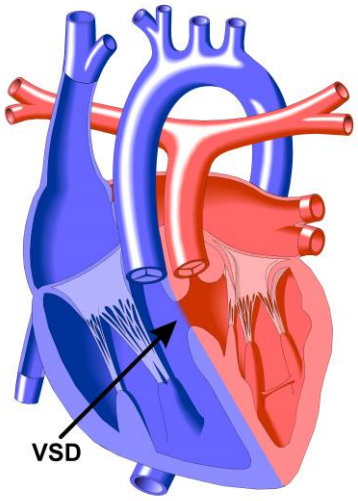
- Abnormal septation
  - Spiral failure in septation of great arteries
    - Pulmonary artery exits from morphologic left ventricle
      (As indicated by #2 in illustration below)
    - Aorta exits from morphologic right ventricle
      (As indicated by #1 in illustration below)
    - Abnormal relationship
      - Usually parallel
      - Aorta anterior and to right of the pulmonary artery
  - Results in parallel circuits
- 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)



Transposition of the Great Arteries (d-TGA)

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



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)

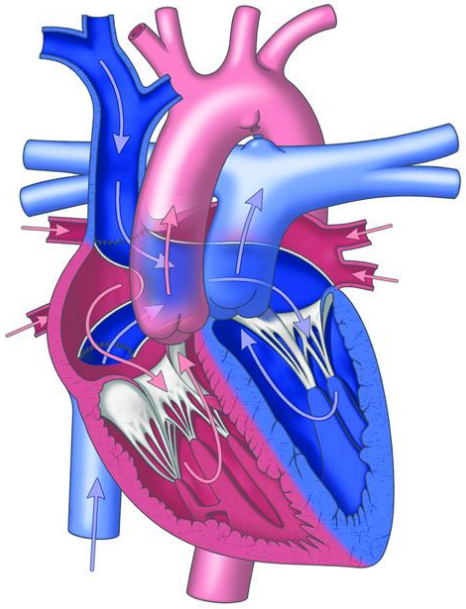
- Parallel circulation
    - Right side
        - Desaturated blood from body returns to right atrium (RA)
        - Right sided ventricle pumps desaturated blood through aorta back to body
    - Left side
        - Oxygenated blood from lungs returns to left atrium (LA)
        - Left sided ventricle 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 E_1 (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)

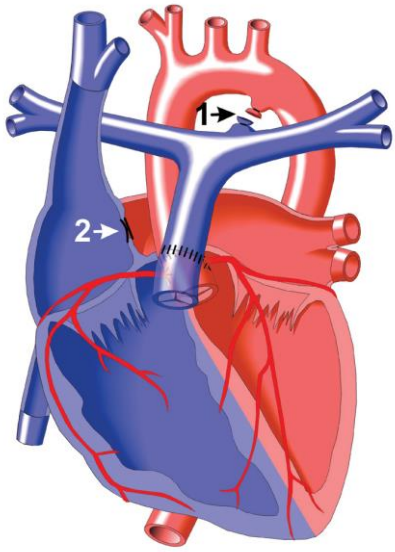


Mustard Procedure

- 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)

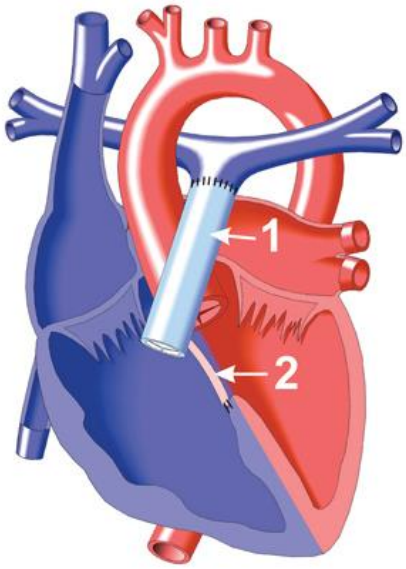


Arterial Switch Repair

- 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
Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

Specific Neonatal Considerations

- Early medical management
  - Maintain patent ductus arteriosus with intravenous prostaglandin E_1
    - 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 (PaO_2<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:

Alonso-Gonzales, R., Dimopoulos, K., Ho, S. Y., Oliver, J. M., Gatozoulis, M. A. (2010). The right heart in adults with congenital heart disease. *Rev Esp Carsio, 63(9),* 1070-86.

Marino, B. S., et al. (2012). Neurodevelopmental outcomes in children with congenital heart disease: Evaluation and management. A Scientific Statement from the American Heart Association. *Circulation,* 126, 1143-1172.

Curley, M. A. Q., & Moloney-Harmon, P. A. (2001). *Critical Care Nursing of Infants and Children*, (2nd ed.). Philadelphia, PA: W.B. Saunders Company.

Evertt, A. D. & Lim, D. S. (2010). *Illustrated Field Guide to Congenital Heart Disease and Repair*, (3rd ed.). Charlottesville, VA: Scientific Software Solutions, Inc.

Love, B. L., Mehta, D., & Fuster, V. F. (2008). Evaluation and management of the adult patient with transposition of the great arteries following atrial level (Senning or Mustard) repair. Available at www.medscape.org/viewarticle/576767 . Accessed online 3/30/2011.

Mavroudis, C., & Backer, C. L. (Eds.). (2003). *Pediatric Cardiac Surgery*, (3rd ed.). St. Louis, MO: Mosby.

Meijboom, F., & Webb, G. (2009). Transposition of the great arteries after a Mustard atrial switch procedure. In C. Warnes, (Ed.). *Adult Congenital Heart Disease*. Oxford, GB: Wiley-Blackwell.

Moore, K. L., & Persaud, T. V. N. (2008). *The Developing Human. Clinically Oriented Embryology*, (8th ed.). Philadelphia, PA: WB Saunders/Elsevier.

Nieves, J. A. (2013). Transposition of the great arteries. Chapter 8: Cardiovascular Disorders, Specific Diseases. In M. F. Hazinski, (Ed.). *Nursing Care of the Critically Ill Child,* (3rd ed.). St. Louis, MO: Elsevier.

Park, M. K. (2008). *Pediatric Cardiology for Practitioner,* (5th ed.). Philadelphia, PA: Elsevier.

Pillutla, P., Shetty, K. D., & Foster, E. (2009). Mortality associated with adult congenital heart disease: Trends in the US population from 1979 to 2005. *American Heart Journal,* 158(5), 874-879.

Skinner, J., Hormung, T., & Rumball, E. (2008). Transposition of the great arteries: from fetus to adult. *Heart,* 94, 1227-1235.

Slota, M. C. (Editor). (2006). *Core Curriculum for Pediatric Critical Care Nursing*. *American Association of Critical Care Nurses,* (2nd ed.). Philadelphia, PA: WB Saunders.

Schwerzmann, M., Salehian, O., Harris, L., et. al. (2009). Ventricular arrhythmias and sudden death in adults after a Mustard operation for transposition of the great arteries. *European Heart Journal, 20*09.

Warnes, C. A. (2006). Transposition of the great arteries. *Circulation,* 114, 2699-2709. Available at http://circ.ahajournals.org/content/114/24/2699 Accessed 4/2011.

Warnes, C. A., & Somerville, J. (1987). Transposition of the great arteries: Late results in adolescents and adults after the Mustard procedure. *British Heart Journal,* 58, 148-55.

Wernovsky, G. (2001). Transposition of the great arteries. In H. D. Allen, E. B. Clark, H. P. Gutgesell, D. J. Driscoll. *Moss and Adams' Heart Disease in Infants, Children' and Adolescents,* (6th ed.). Philadelphia, PA: Lippincott Williams & Wilkins.

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