

Tricuspid Atresia

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

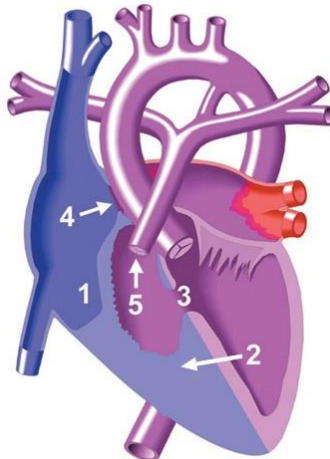
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Embryology

- Formation of cardiac septa: Days 27 to 45
 - Active fusion of the cushions or passive expansion of cardiac chambers occurs
 - Endocardial cushions divide the atrioventricular (AV) canal into the mitral and tricuspid valves
- Formation of the atrioventricular cardiac valves: Days 34 to 36
 - Formed from endocardial cushion tissue
 - Tricuspid valve, papillary muscles and chordate tendineae formed largely from the conus septum

Anatomy

- Agenesis of the tricuspid valve (TV)
- Atrial septal defect (ASD) (Number 4 in Illustration)
- Patent ductus arteriosus (PDA)



Tricuspid Atresia

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- Associated defects—Type 1 (normally related great vessels) (As illustrated):
 - Ia. Hypoplastic right ventricle (RV) (Number 2 in Illustration) and pulmonary atresia (Number 1 in Illustration)
 - Ib. Hypoplastic right ventricle and pulmonary hypoplasia (Number 5 in Illustration), small ventricular septal defect (VSD) (Number 3 in Illustration)
 - Ic. Normal RV size, no pulmonary hypoplasia and large VSD

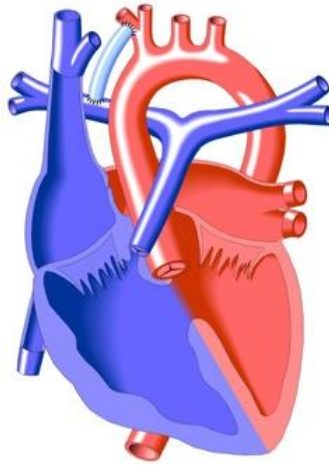
- Associated defects—Type II d-transposition of the great arteries (d-TGA):
 - IIa. Hypoplastic right ventricle, VSD and pulmonary atresia
 - IIb. Hypoplastic right ventricle, VSD and pulmonary or subpulmonary stenosis
 - IIc. Hypoplastic right ventricle, large VSD, no pulmonary stenosis
- Associated defects---Type III l-transposition of the great arteries (l-TGA):
 - IIIa. Hypoplastic left ventricle, pulmonary or subpulmonary stenosis
 - IIIb. Hypoplastic RV, subaortic stenosis

Physiology

- Tricuspid atresia with VSD
 - Blood enters the right atrium and cannot exit due to agenesis of the TV and crosses the atrial septal defect into the left atrium (LA) causing systemic desaturation.
 - Blood then crosses the mitral valve (MV) and enters the left ventricle (LV). Blood enters the right ventricle across the VSD. The size of the VSD and degree of pulmonary hypoplasia affect the amount of ventricular shunting and pulmonary blood flow which results in the level of desaturation.
 - In patients with little or no pulmonary hypoplasia the patient may not be cyanotic and actually present with congestive heart failure (CHF) from overcirculation of the pulmonary bed.
 - If d-TGA is present the blood will enter the RA and cross the atrial defect into the LA and cross the MV into the LV which gives rise to the PA. Blood crosses the VSD into the right ventricle and exits to the aorta and the balance of systemic and pulmonary blood flow is dependent on the degree of associated pulmonary stenosis.
- Tricuspid atresia without VSD
 - Blood enters the right atrium and cannot exit due to agenesis of the TV and crosses the atrial septal defect into the left atrium causing systemic desaturation and exits the heart via the LV and aorta.
 - Blood perfuses the pulmonary bed through a left to right shunt at the level of the PDA.

Types of repairs

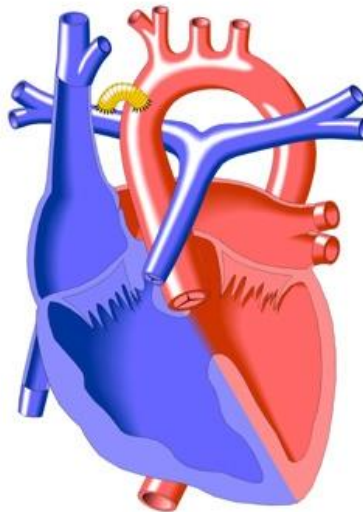
- Palliation for decreased pulmonary blood flow
 - Systemic to pulmonary artery shunt: increases pulmonary blood flow through surgically created left to right shunt at the great vessel level
 - Classic Blalock-Taussig shunt: end to side anastomosis subclavian to right or left branch pulmonary artery shunt (rarely performed)
 - Modified Blalock-Taussig shunt (As Illustrated): Gortex interposition graft between subclavian or innominate artery and right or left branch pulmonary artery



Modified Blalock-Taussig Shunt

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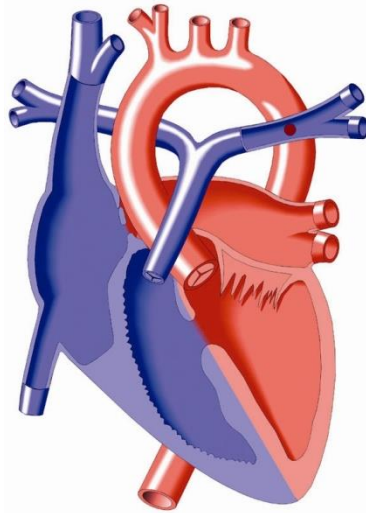
- Central shunt (As Illustrated): Gortex interposition graft between aorta and main pulmonary artery



Central Shunt

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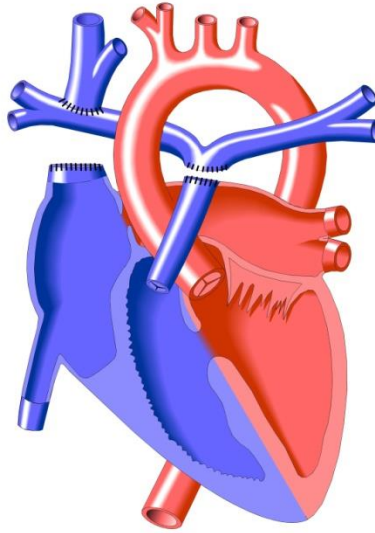
- Potts (As Illustrated): Direct anastomosis descending aorta to left pulmonary artery (LPA) (historical)



Pott's Shunt

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- Potential for development of pulmonary vascular obstructive disease (PVOD)
 - Potential for distortion of LPA
 - Waterston: Direct anastomosis ascending aorta to right pulmonary artery (RPA) (historical)
 - Potential for development of PVOD
 - Potential for distortion of RPA
 - Palliation for increased pulmonary blood flow
 - Control amount of pulmonary blood flow to prevent CHF and pulmonary vascular disease from pulmonary overcirculation
 - Pulmonary artery band
 - Band placed on PA, adjusted to pressure in PA not blood flow
 - May still result in pulmonary overcirculation
 - May distort PA's at site of band placement
 - May migrate to one branch PA creating overcirculation in one lung with associated pulmonary vascular changes
 - Ligation of main pulmonary artery and placement of systemic to pulmonary artery shunt
 - Pulmonary blood flow controlled by:
 - Size/location of shunt
 - Vessel to which it is attached
 - Palliation for tricuspid atresia and d-TGA
 - Norwood or Damus Kaye Stansel
 - Provide systemic outflow and controlled pulmonary blood flow through shunt
 - Hemifontan/Bidirectional Glenn (As Illustrated)

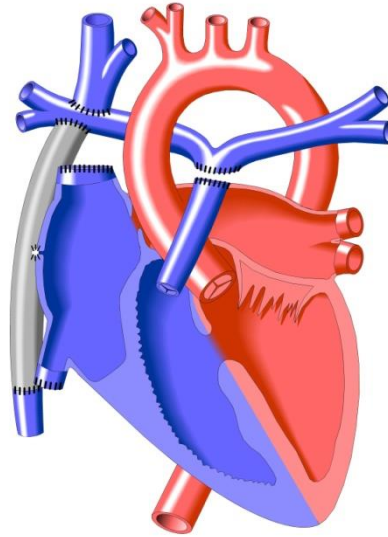


Bi-directional Glenn Shunt

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- Superior vena cava (SVC) to pulmonary artery (PA) connection
 - SVC flow directly to PA
 - Inferior vena cava (IVC) flow continues into right heart
 - Increases SVC pressure and pulmonary blood flow dependent on SVC-PA-LA gradient (Transpulmonary gradient)
- Fontan
 - Physiologic correction for single ventricle lesion
 - Pulmonary blood flow achieved through SVC/IVC /PA to LA pressure gradient (transpulmonary gradient)
 - Goal of surgical technique is to achieve systemic venous flow (IVC/SVC) directly into PA's bypassing ventricular contribution
 - A hole (Fenestration) between the systemic venous/PA connection and common atrium utilized to assist hemodynamic adjustment to acutely elevated venous pressures
 - Surgical options for Fontan operation
 - Lateral tunnel: Gortex graft placed inside RA to direct IVC flow through RA/SVC junction and into MPA
 - Extracardiac (As Illustrated): Gortex or Dacron circumferential tube graft from IVC to MPA



Lateral Tunnel Fontan

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- Direct RA to PA anastomosis: connection of right atrial appendage to PA (not preformed currently)

Specific considerations:

- Neonate
 - Elevated pulmonary vascular resistance initially therefore TA with VSD may not demonstrate signs of CHF
 - Elevated pulmonary vascular resistance initially may limit pulmonary blood flow through systemic to pulmonary shunt in TA without VSD
 - Requirement for establishment of systemic outflow in TA with dTGA may result in coronary artery flow abnormalities or imbalance in systemic/pulmonary blood flow based off relative resistance in each circuit
 - Pulmonary overcirculation resulting in systemic underperfusion, LCOS and possibly shock may occur with systemic to pulmonary shunt
 - Pulmonary undercirculation resulting in systemic hypoxemia may occur as result of hypotension, elevations PVR, distortion or kinking of the shunt or pulmonary artery or acute thrombosis of the shunt
- Post-operative management
 - Pulmonary overcirculation managed by decreasing systemic resistance with afterload reduction
 - Pulmonary undercirculation managed by optimizing pulmonary resistance and systemic blood pressure with early assessment of potential mechanical obstruction
 - Adequate control of pain, sedation, and temperature to optimize the balanced circulation

- Avoidance of pulmonary process (pleural effusion, pneumonia, pneumothorax, atelectasis, collapse) which alters pulmonary vascular resistance potentially resulting in imbalance of systemic-pulmonary circulation
- Support volume overloaded single ventricle with diuretics, inotrope as needed
- Support oxygen delivery with hematocrit greater than 40%
- Early consideration for reoperation for band tightening if CHF persists after initial band
- Follow upper and lower extremity blood pressure if DKS or Norwood reconstruction required
- Follow end organ function primarily bowel (risk of NEC), kidney function (risk of AKI from surgery/systemic underperfusion), neurologic insult (LCOS, hypoxemia, acidosis)

Long term complications: potential interventions (See Peds/Neo and Adult Guidelines for identified problems: Arrhythmia Management, Ventricular Dysfunction, Tricuspid Valve, Mitral Valve, Anticoagulation Management)

- Arrhythmias
 - Catheter or surgical ablation
 - Pacemaker and/or ICD placement
 - Medications
 - Conversion to lateral tunnel or extracardiac Fontan connection with plication of RA
 - Maze procedure if performing operation for Fontan conversion
- Ventricular dysfunction
 - Symptom management
 - Repair associated defects such as AVVR causing dysfunction
 - Transplant
 - Ventricular assist device as bridge or as destination therapy
 - Creation of fenestration if not previously present (risk cyanosis for increased CO)
- Atrioventricular valve regurgitation (AVVR): Valve repair/replacement
- Fontan pathway obstruction: reoperation for relief of conduit stenosis
- Protein losing enteropathy (PLE) (See Peds/Neo Guidelines for Nutrition)
 - Loss of protein into abdomen, diarrhea, edema
 - No known cause or definitive treatment
 - Potential solutions
 - Conversion Fontan
 - Create ASD
 - Transplant
- Plastic bronchitis: casts that occlude bronchus, no curative/definitive treatment
- Exercise intolerance
- Hepatic congestion/failure from long term elevation venous pressure
- Thromboembolic events
- Anticoagulation
 - Varies from center to center
 - Minimally life-long aspirin (ASA)

References:

Castaneda, A. R., Jonas, R. A., Mayer, J. E., Hanley, F. L, (Eds.). (1994). *Cardiac Surgery of the Neonate and Infant*, Philadelphia, PA: WB Sanders.

Park, M. K. (2008) *Pediatric Cardiology for Practitioners*, St. Louis, MO: Mosby.

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

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

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