

Tricuspid Atresia

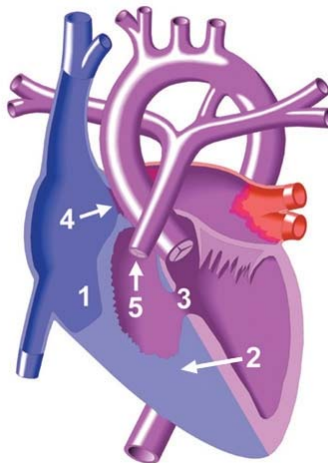
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I. Embryology

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

II. Anatomy

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



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- D. Associated defects—Type 1 (normally related great vessels) (As illustrated):
 - 1. Ia. Hypoplastic right ventricle (RV) (Number 2 in Illustration) and pulmonary atresia (Number 1 in Illustration)
 - 2. Ib. Hypoplastic right ventricle and pulmonary hypoplasia (Number 5 in Illustration), small ventricular septal defect (VSD) (Number 3 in Illustration)
 - 3. Ic. Normal RV size, no pulmonary hypoplasia and large VSD
- E. Associated defects—Type II d-transposition of the great arteries (d-TGA):
 - 1. IIa. Hypoplastic right ventricle, VSD and pulmonary atresia
 - 2. IIb. Hypoplastic right ventricle, VSD and pulmonary or subpulmonary stenosis
 - 3. IIc. Hypoplastic right ventricle, large VSD, no pulmonary stenosis
- F. Associated defects—Type III l-transposition of the great arteries (l-TGA):
 - 1. IIIa. Hypoplastic left ventricle, pulmonary or subpulmonary stenosis

2. IIIb. Hypoplastic RV, subaortic stenosis

III. Physiology

A. Tricuspid atresia with VSD

1. 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.
2. 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 pulmonary hypoplasia affect the amount of ventricular shunting and pulmonary blood flow which results in the level of desaturation.
3. 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.
4. If d-TGA is present the blood will enter the RA and cross the atrial shunt 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.

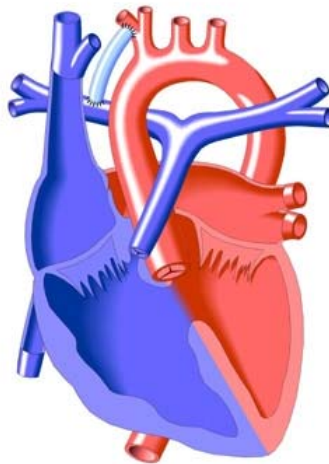
B. Tricuspid atresia without VSD

1. 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.
2. Blood perfuses the pulmonary bed through a left to right shunt through the PDA.

IV. Types of repairs

A. Palliation for decreased pulmonary blood flow

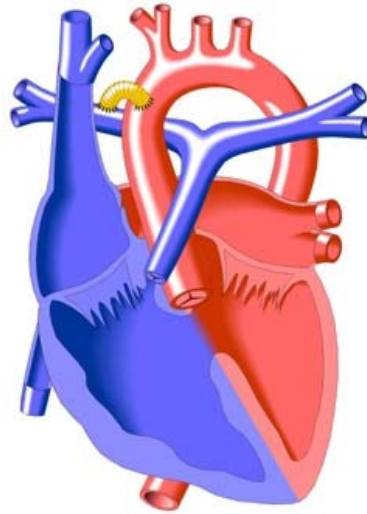
1. Systemic to pulmonary artery shunt: increases pulmonary blood flow through surgically created left to right shunt at the great vessel level
 - a. Classic Blalock-Taussig shunt: end to side anastomosis subclavian to right or left branch pulmonary artery shunt (rarely performed)
 - b. Modified Blalock-Taussig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



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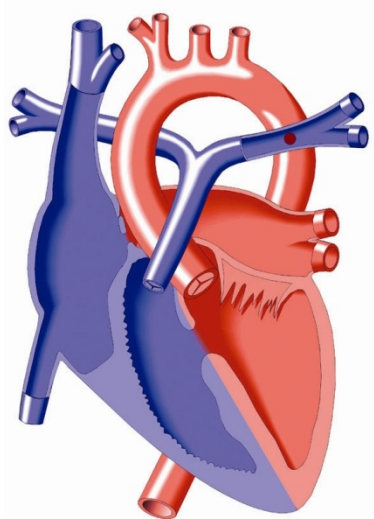
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- c. Central shunt (As Illustrated): Gortex interposition graft between aorta and main pulmonary artery



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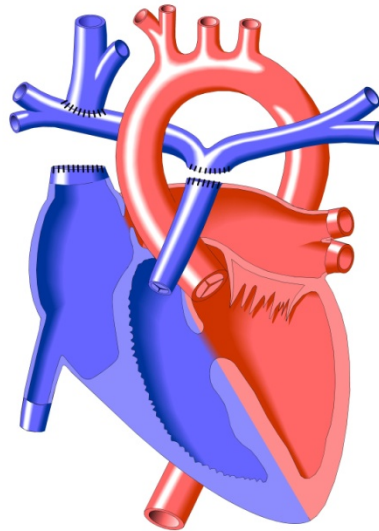
- d. Potts (As Illustrated): Direct anastomosis descending aorta to left pulmonary artery (LPA) (historical)



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- (1) Potential for development of pulmonary vascular obstructive disease (PVOD)
 - (2) Potential for distortion of LPA
- e. Waterston: Direct anastomosis ascending aorta to right pulmonary artery (RPA) (historical)

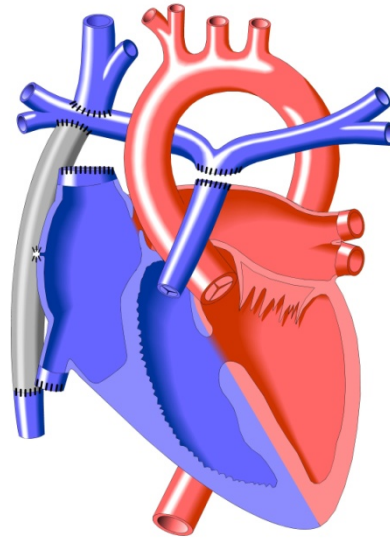
- (1) Potential for development of PVOD
- (2) Potential for distortion of RPA
- B. Palliation for increased pulmonary blood flow
 - 1. Control amount of pulmonary blood flow to prevent CHF and pulmonary vascular disease from pulmonary overcirculation
 - a. Pulmonary artery band
 - (1) Band placed on PA, adjusted to pressure in PA not blood flow
 - (2) May still result in pulmonary overcirculation
 - (3) May distort PA's at site of band placement
 - (4) May migrate to one branch PA creating overcirculation in one lung with associated pulmonary vascular changes
 - b. Ligation of main pulmonary artery and placement of systemic to pulmonary artery shunt
 - (1) Pulmonary blood flow controlled by size/location of shunt and vessel to which it is attached
- C. Palliation for tricuspid atresia and d-TGA
 - 1. Norwood or Damus Kaye Stansel
 - a. Provide systemic outflow and controlled pulmonary blood flow through shunt
- D. Hemifontan/Bidirectional Glenn (As Illustrated)



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- 1. Superior vena cava (SVC) to pulmonary artery (PA) connection
 - a. SVC flow directly to PA
 - b. Inferior vena cava (IVC) flow continues through right heart
 - c. Increases SVC pressure and pulmonary blood flow dependent on SVC-PA-LA gradient
- E. Fontan
 - 1. Physiologic correction for single ventricle lesion
 - 2. Pulmonary blood flow achieved through SVC/IVC /PA to LA pressure gradient (transpulmonary gradient)

3. Goal of surgical technique is to achieve systemic venous flow (IVC/SVC) directly into PA's bypassing ventricular contribution
4. A hole (Fenestration) between the systemic venous/PA connection and common atrium utilized to assist hemodynamic adjustment to acutely elevated venous pressures
5. Surgical options for Fontan operation
 - a. Lateral tunnel: Gortex graft placed inside RA to direct IVC flow through RA/SVC junction and into MPA
 - b. Extracardiac (As Illustrated): Gortex or Dacron circumferential tube graft from IVC to MPA



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

V. Long term complications: potential interventions (See Problem List included with these documents)

- A. Arrhythmia: ablation, pacemaker, ICD, medications, conversion to lateral tunnel or extracardiac Fontan connection with plication of RA
- B. Ventricular dysfunction: rhythm and transplant
- C. Atrioventricular valve regurgitation (AVVR): Valve repair/replacement
- D. Fontan pathway obstruction: reoperation for relief of conduit stenosis
- E. Protein losing enteropathy (PLE): lose of protein into abdomen, diarrhea, edema: no known cause or definitive treatment but may try conversion Fontan, create ASD or transplant
- F. Plastic bronchitis: casts that occlude bronchus, no treatment
- G. Thromboembolic events: anticoagulation varies from center to center but minimally life long aspirin (ASA)

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