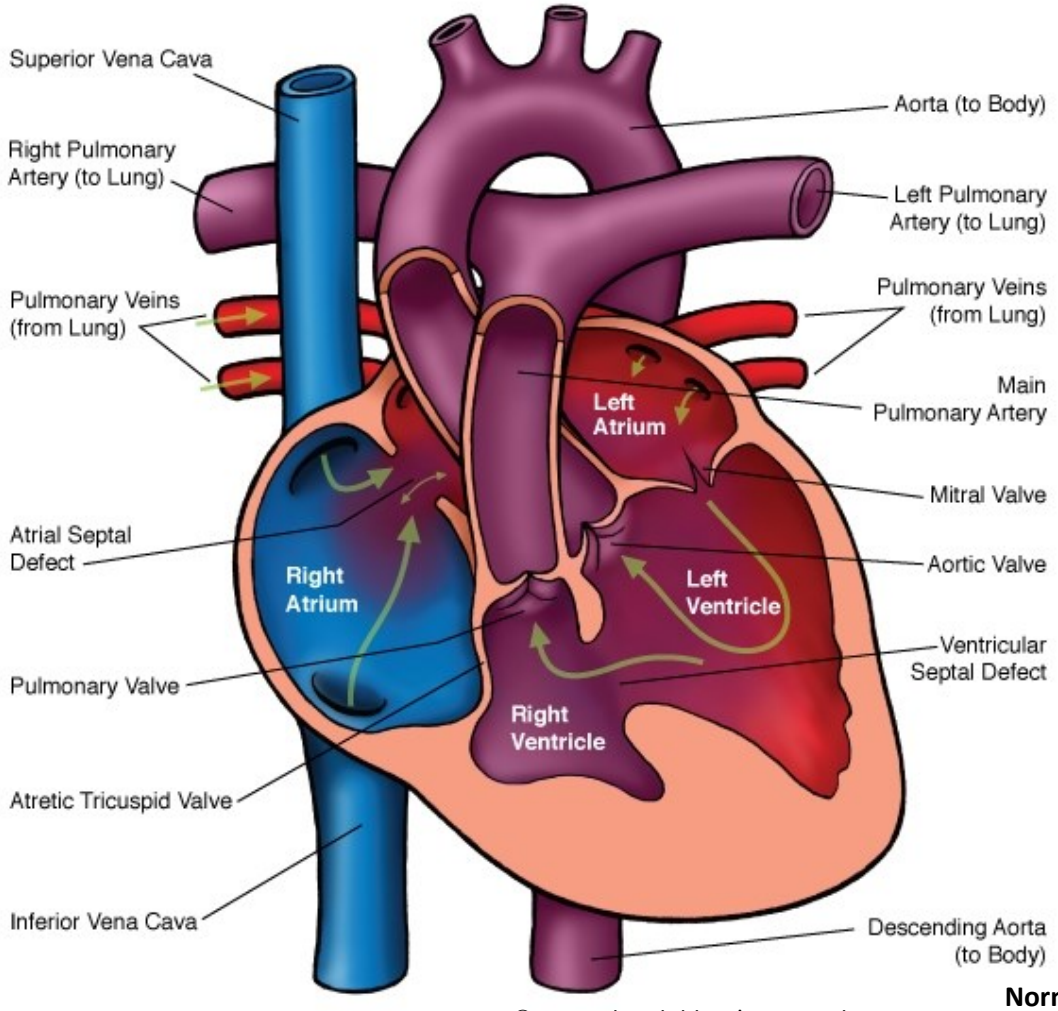
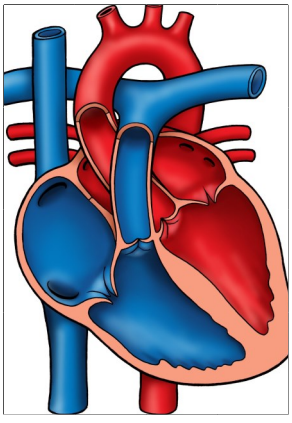
Tricuspid Atresia



Normal Heart



NOTES:

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Tricuspid Atresia
(TA)

Tricuspid atresia (TA) is the absence of a patent tricuspid valve. Due to lack of normal blood flow from the right atrium through the atretic (closed or underdeveloped) tricuspid valve, the right ventricle is hypoplastic (small and underdeveloped). Associated heart defects including atrial septal defect (ASD), ventricular septal defect (VSD), or patent ductus arteriosus (PDA) are necessary for survival. TA is classified by the presence or absence of pulmonary stenosis (PS) and transposition of the great arteries (TGA). In 50% of children with TA, normally related great arteries, a small VSD, and PS are present. Due to decreased pulmonary blood flow, the pulmonary arteries are hypoplastic. When TGA is present, the pulmonary valve is normal and pulmonary blood flow is increased in most cases. Occasionally, the pulmonary valve is stenotic (severely narrowed) or atretic with decreased pulmonary blood flow. Coarctation of the aorta or interrupted aortic arch is frequently associated with individuals who have TA and TGA. TA accounts for 1% of congenital heart disease.

Physical Exam/Symptoms:

- Severe cyanosis (blue color) from birth.
- Tachypnea (fast breathing) and poor feeding.
- Murmur: Grade II-III/VI holosystolic VSD murmur is heard at the left lower sternal border.
- Hepatomegaly (enlarged liver) may be present.

Diagnostics:

- Chest X-ray: Heart size is normal to slightly enlarged. Pulmonary vascular markings are usually decreased.
- EKG: Left ventricular hypertrophy (LVH) is usually present. A superior QRS axis is characteristic.
- Echocardiogram: Diagnostic.

Medical Management/Treatment:

- Prostaglandin E (PGE) therapy is started immediately after birth in neonates with severe cyanosis to keep the ductus arteriosus patent until cardiac catheterization or surgery.
- Balloon atrial septostomy in the catheterization lab for infants with inadequate interatrial communication (small ASD or PFO).
- Infants with VSDs allowing adequate blood flow do not require surgery as neonates, but are watched closely for decreasing oxygen saturations due to spontaneous reduction in VSD size.
- Surgical repair in multiple stages is necessary for survival. Operations may include modified Blalock-Taussig shunt (BTS), Damus-Kaye-Stansel (DKS), bidirectional Glenn, Fontan, or Kawashima procedure. Your cardiologist will discuss your surgical options and timing with you.
- Postoperative medical management includes diuretic therapy (Lasix), aspirin or coumadin to prevent thrombus (clot) formation, and ACE inhibitors (enalapril, captopril).
- Bacterial endocarditis prophylaxis should be observed prior to any dental procedure.
- Lifetime close cardiology follow-up is necessary to monitor for late complications.

Long-Term Outcomes:

- 5 year survival rate for children with TA is 80%; 10 year survival is 70% worldwide.
- Long-term survival and developmental outcomes vary widely based on postoperative outcomes and other co-morbidities.

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