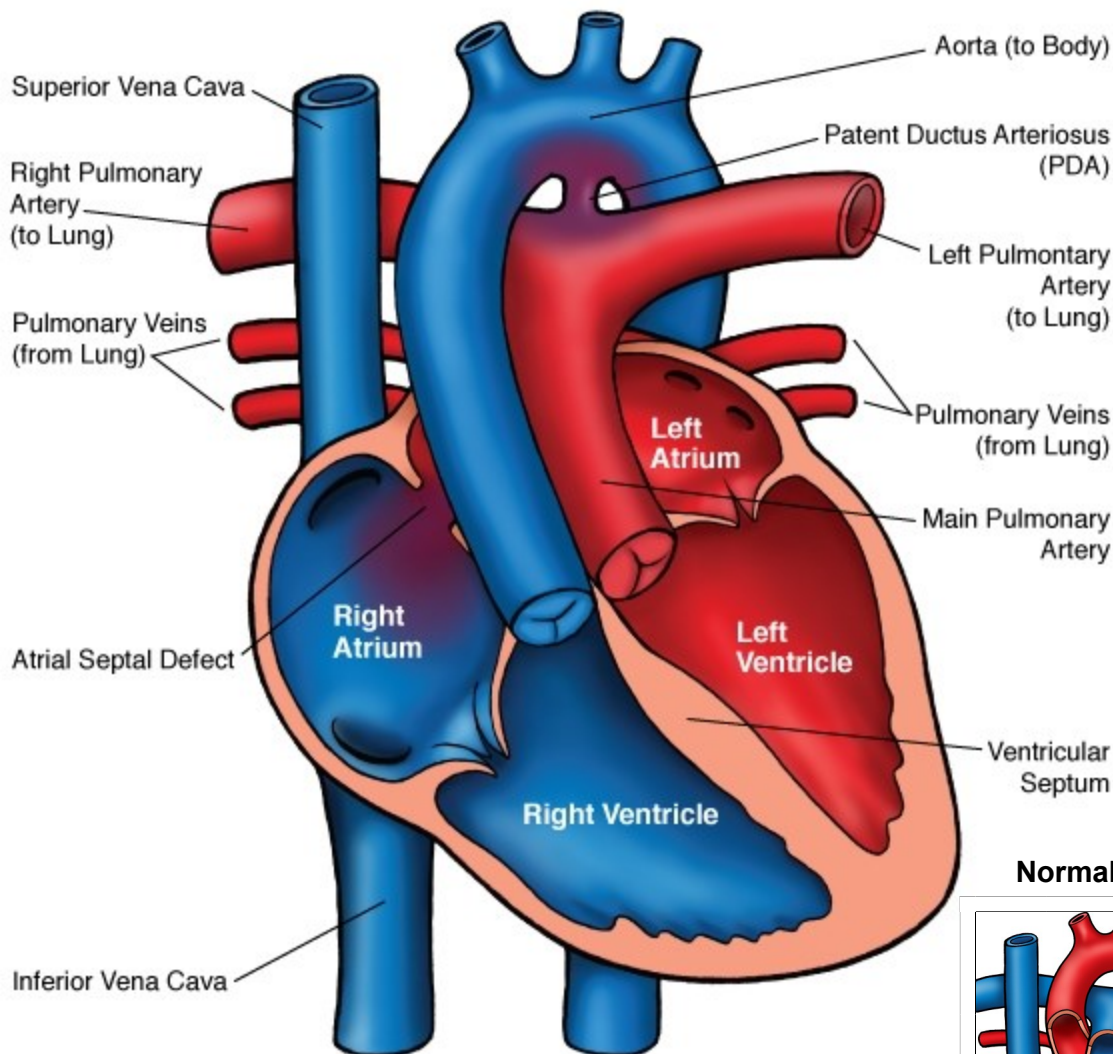


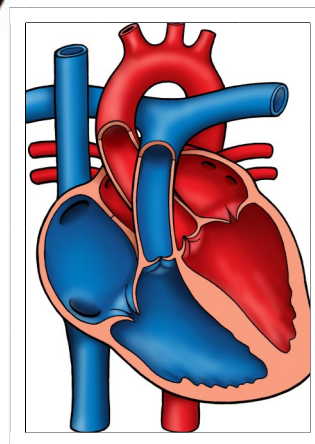
Transposition of the Great Arteries (TGA)



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Notes:

Normal Heart



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Transposition of the Great Arteries (D-TGA)

In a structurally normal heart, the aorta arises from the left ventricle and the pulmonary artery (PA) arises from the right ventricle. This allows deoxygenated blood from the body to be pumped through the right side of the heart, to the PA and out to the lungs for oxygenation. The oxygenated blood is then returned to the left side of the heart and pumped out to the body through the aorta.

In transposition of the great arteries (D-TGA), the aorta and PA are transposed or switched—meaning that the aorta arises from the right ventricle and the PA arises from the left ventricle. The coronary arteries arise normally from the aorta. The aorta is to the right of the PA, which is why the D-prefix is used (dextroposition, meaning rightward). With this arrangement, blood is returned to the body without receiving oxygen from the lungs, and oxygenated blood continues to circle from the left side of the heart back to the lungs. This results in lack of adequate oxygen to the vital organs of the body, including the heart muscle. Essentially, blood is moving in 2 separate circuits without adequate communication, unless a large atrial septal defect (ASD) or ventricular septal defects (VSD) is present. D-TGA in the absence of an adequate ASD or VSD is fatal without intervention as a neonate. D-TGA occurs in 5-7% of all congenital heart defects and is more common in males than females (3:1 ratio).

Physical Exam:

- Moderate to severe cyanosis (blueness beginning at birth).
- Single, loud S2 heart sound.
- A holosystolic murmur may be heard if a VSD is present.
- Signs of congestive heart failure (CHF): poor feeding, hepatomegaly (enlarged liver), and dyspnea (difficulty breathing) develop.

Diagnostics:

- Chest X-ray: Cardiomegaly (enlarged heart) and increased pulmonary vascular markings are typically present. The silhouette of the heart may appear egg-shaped, which is characteristic of D-TGA.
- EKG: The QRS axis is rightward. Right ventricular hypertrophy (RVH) is often present after the first few days of life. Left ventricular hypertrophy (LVH) as well as RVH may be present in infants who have a large VSD, patent ductus arteriosus (PDA), or obstructive pulmonary vascular disease.
- Echocardiogram: Diagnostic.
- Computerized Tomography Angiogram: Performed if needed to determine coronary artery origins.

Medical Management/Treatment:

- Prostaglandin E (PGE) should be started as soon as possible to keep the ductus arteriosus patent or reopen the ductus to improve oxygenation until catheter or surgical intervention can occur.
- Oxygen should be provided to decrease pulmonary vascular resistance (PVR) and increase pulmonary blood flow to improve oxygenation to the body and vital organs.
- Cardiac catheterization and balloon atrial septostomy (Rashkind procedure) occur in the first days of life for infants without adequate atrial communication to improve mixing of oxygenated and deoxygenated blood.

Transposition of the Great Arteries (D-TGA)

- Coronary anatomy is determined either by echocardiogram, computerized tomography angiogram (CTA), or in the catheterization lab prior to surgical intervention.
- Surgical options are determined based on individual anatomy. Please see Rastelli Procedure, Arterial Switch Operation, and Nikaidoh Procedure. Your child's cardiologist will discuss surgical options and timing with you.
- Life-long cardiology follow up is necessary.
- Following arterial switch operation, aspirin therapy is used for 3-6 months to prevent coronary thrombosis. This may be life-long for children with a single coronary artery.

Long-Term Outcomes:

- Without surgical intervention, 90% of children will die by the age of 6 months.
- Supravalvar pulmonary or aortic stenosis, arrhythmias or semi-lunar valve regurgitation may occur infrequently.
- Arrhythmias are more common after atrial baffle operation than Arterial Switch Operation.
- In general, children have normal growth and development in the absence of residual cardiac disease or other co-morbidities.