

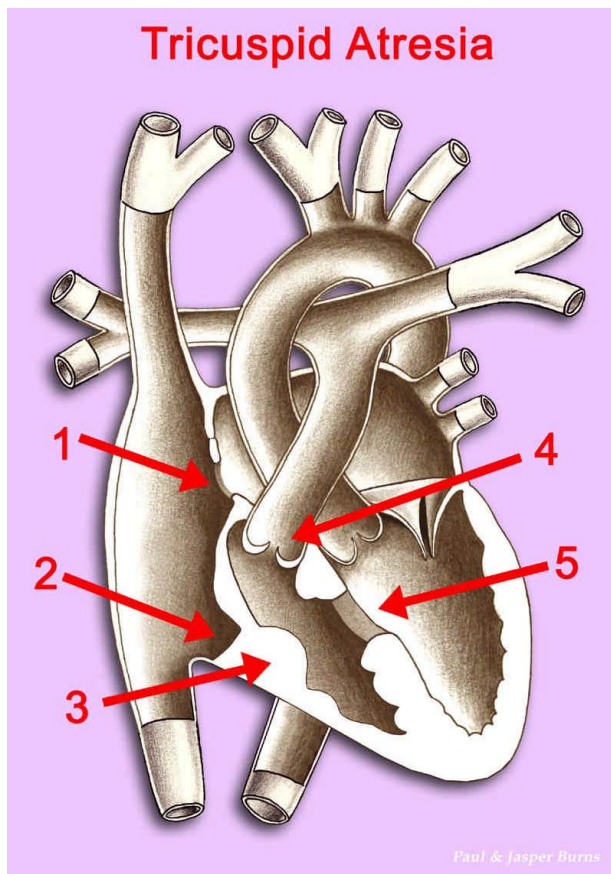
Tricuspid Atresia



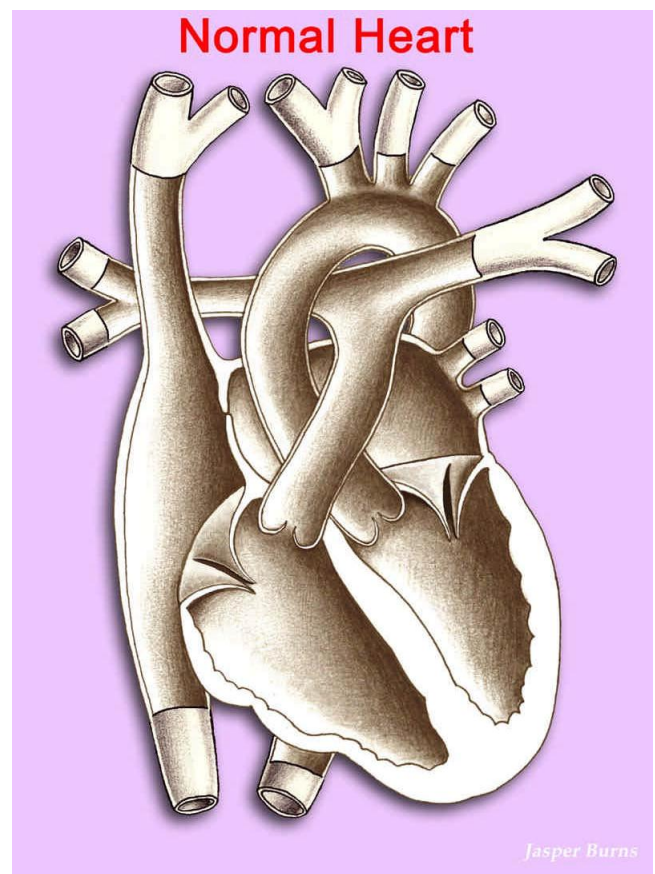
What Is It?

In Tricuspid Atresia, the heart has three valves rather than four. The tricuspid valve, which connects the right atrium (collecting chamber) and right ventricle (pumping chamber) in the normal heart, is abnormal and does not open. In addition, the atrial septum, or muscle wall, which divides the left and right atria, has an opening in it, called an atrial septal defect (ASD). This allows the mixing of blood from the right and left atria.

The muscle wall, which separates the right and left ventricles, is known as the ventricular septum. In Tricuspid Atresia, this septum generally has a hole in it, called a Ventricular Septal Defect, or VSD. In addition, the right ventricle is usually quite small and the pulmonary valve, through which blood is pumped to the lungs, can be narrowed. This narrowing of the pulmonary valve is known as a stenosis.



Tricuspid Atresia

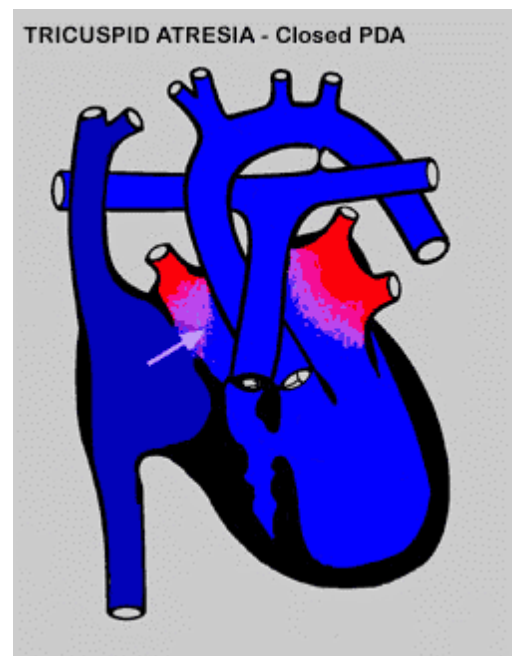
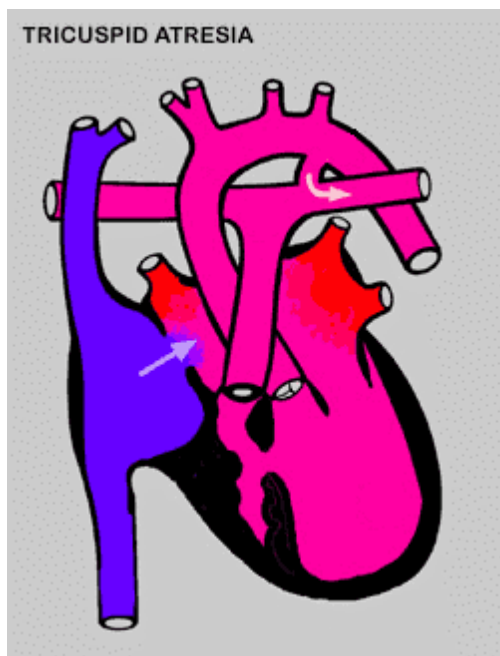


Normal Heart

- 1) Atrial Septal Defect
- 2) Missing Tricuspid Valve
- 3) Hypoplastic (very small) Right Ventricle
- 4) Pulmonary Stenosis (narrowing of pulmonary valve)
- 5) Ventricular Septal Defect

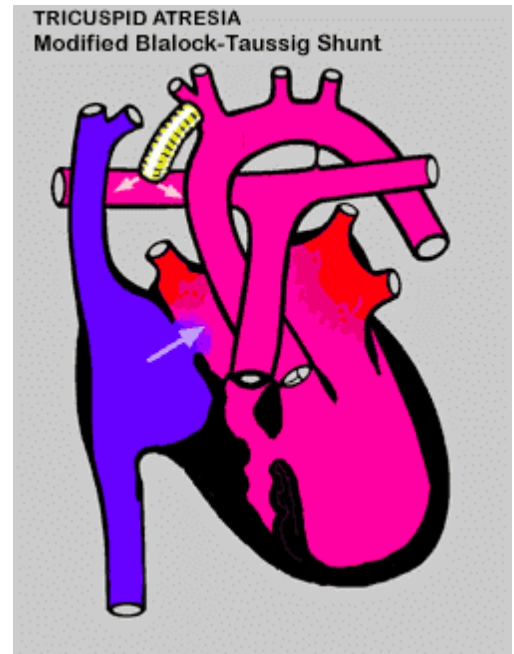
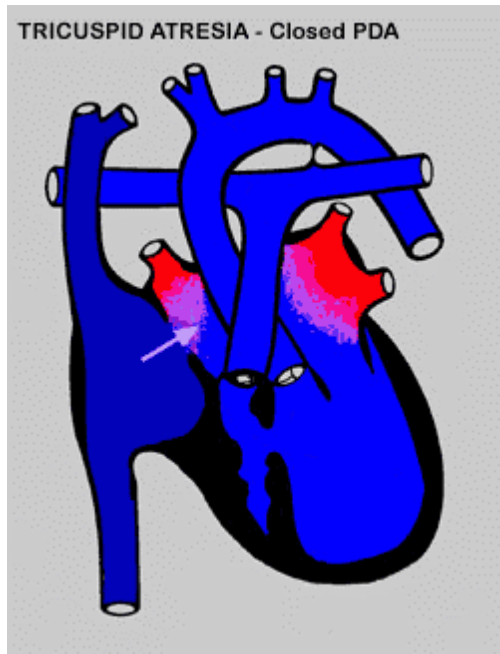
What Are Its Effects?

Many children with Tricuspid Atresia need blood flow across the Patent Ductus Arteriosis (PDA) to maintain adequate pulmonary (lung) blood flow. When the PDA closes soon after birth (see diagram at lower right), the body becomes starved for oxygenrich (red) blood and the patient will show worsening cyanosis, or blueness. These children may become very ill and require surgery to increase the blood flow to the lungs.



How Is It Treated?

After the Patent Ductus Arteriosus (PDA) closes, red blood may be directed to the body through the insertion of a Modified Blalock-Taussig Shunt, which is a small tube made of Gore-Tex® (yellow in the diagram, below right) that connects the aorta and pulmonary artery. Though in a different position, the "B-T Shunt" has much the same function as the Patent Ductus Arteriosus.



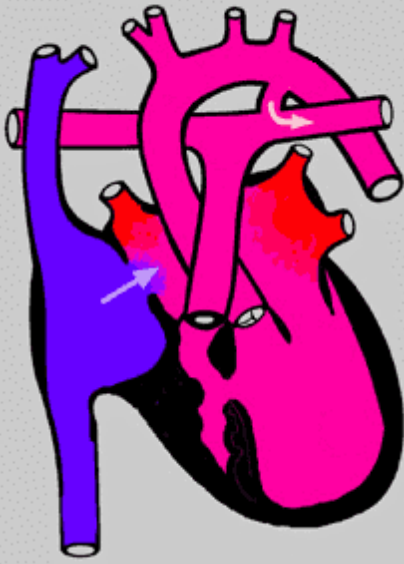
A child with Tricuspid Atresia may ultimately undergo an operation known as the Fontan Procedure. This directs the red, oxygenated blood to the body and the blue, oxygen-poor blood to the lungs.

There are different forms of the Fontan Procedure. In the one shown in the diagram below, blood flow from the lower half of the body is channeled directly into the right pulmonary artery by fashioning a tunnel connecting the inferior vena cava to the pulmonary artery (notice the yellow wall with a small opening to release pressure). After entering the right pulmonary artery, the blood can go both to the left and the right lung.

The stump of the superior vena cava is attached to the right pulmonary artery in this procedure, allowing all systemic venous blood (shown in blue) to flow directly into the pulmonary arteries, bypassing the heart.

In many heart centers, this operation is done in stages. Usually the superior vena cava is attached first. This is referred to as a bi-directional Glenn operation or a hemi-Fontan. When the blood from the lower half of the body is channeled up to the lungs, this completes the Fontan.

TRICUSPID ATRESIA



TRICUSPID ATRESIA
Fontan Procedure

