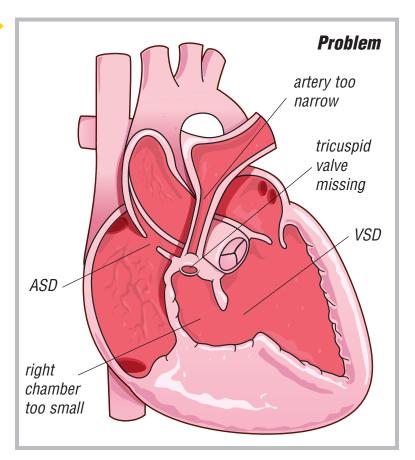
The tricuspid valve fails to develop. This is the valve that lets blood move from the top right heart chamber (atrium) to the bottom right heart pumping chamber (ventricle). The lower right chamber of the heart is either very small or missing. Often there is failure or incomplete development of the pulmonary valve and the main pulmonary artery. This restricts the blood flow to the lungs.

There is a hole between the top two chambers of the heart (atrial septal defect or ASD). This hole is necessary for the child to survive until the surgery to correct the tricuspid atresia is done. There may also be a hole between the two lower chambers of the heart (ventricular septal defect or VSD).

Surgical procedures are most often done in stages:



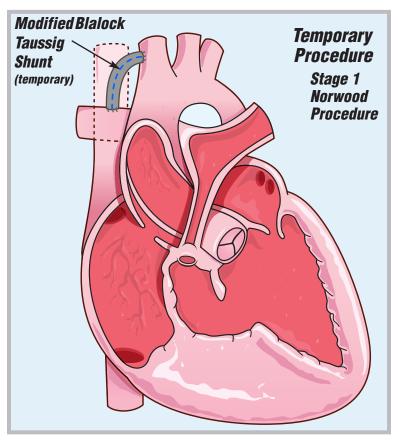


Stage 1 - Norwood Operation using Blalock Taussig shunt

Stage 1

Norwood Procedure - The first operation is most often the temporary placement of a small tube (shunt) from the aorta or one of its branches to the pulmonary artery (Modified Blalock Taussig Shunt). The shunt is put in to allow more blood to flow to the lungs. It may be placed on the left, right or both sides of the pulmonary artery branch. The shunt(s) will be removed when no longer needed.

The shunt is placed through a thoracotomy or a sternotomy incision.



Stage 2 - Bi-directional Glenn Anastomosis

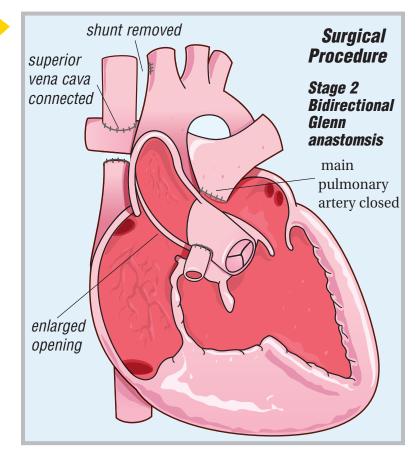
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Stage 2

Bidirectional Glenn anastomosis -

The Glenn Shunt reduces the workload on the heart. The main pulmonary artery is closed. The superior vena cava is connected with the right pulmonary artery. The blue blood from the head, neck and upper body is sent to the lungs without going through the heart. The hole between the top two chambers is enlarged (atrial septectomy). The temporary shunt placed earlier is removed.

The Glenn Shunt operation is done through a median sternotomy (chest) incision.



Stage 3 - Extracardiac Fontan Procedure

Stage 3

Extracardiac Fontan Procedure -

The blue blood from the lower part of the body is directed to the lungs. This is done using the inferior vena cava and a conduit (tube). Artificial material is fashioned into a tube. It is connected from the inferior vena cava to the pulmonary artery. This bypass or rerouting allows the blue blood to enter the lungs without being pumped by the heart. The heart remains available to receive the red blood from the lungs and then pump it to the body. Sometimes, a small hole (fenestration) is placed in the baffle (with a snare around it) to allow the heart and body to adjust gradually to the new blood flow system. The single hole is closed later through a small incision. (Another method is to place several small holes in the baffle to allow the heart and body to adjust gradually to the new blood flow system. These holes close off on their own a few months after surgery.)

The goals of the procedure:

- separate the red and blue blood
- have the blue blood enter the lungs directly
- have the heart itself as the pump of oxygen-rich blood to the body

The Extracardiac Fontan Procedure is done through a median sternotomy (chest) incision.

