Hypoplastic Left Heart Syndrome (HLHS)

Several abnormalities of development occur on the left side of the heart:

- The valve from the top left heart chamber to the bottom left heart chamber is poorly developed (mitral stenosis) to not formed (mitral atresia).
- The bottom left heart chamber is poorly developed (left ventricular hypoplasia), the valve from the bottom left heart chamber to the main artery is poorly developed (aortic stenosis) to not formed (aortic atresia).
- The main artery leaving the left side of the heart (aorta) is underdeveloped. The aorta is narrowed as well (coarctation of the aorta).
- There is an open connection between the aorta and the pulmonary artery (patent ductus arteriosus).
- There is also a small hole between the top two chambers (atrial septal defect).

The surgical options for HLHS include cardiac transplantation and the Norwood Procedure. The Norwood Procedure is a series of operations designed to have the blue (unoxygenated) blood flow directly to the lungs and then use the existing right heart to pump the red (oxygenated) blood to the body.

Norwood Stage I - The large main pulmonary artery and small aorta are fashioned together to make a new, larger aorta. There is a small tube (shunt) put in to connect the lung artery to the aorta. This shunt gives blood flow to the lungs. The wall that separates the top two heart chambers is removed (atrial septectomy). The narrowing of the aorta is opened. The connecting blood vessel is removed.

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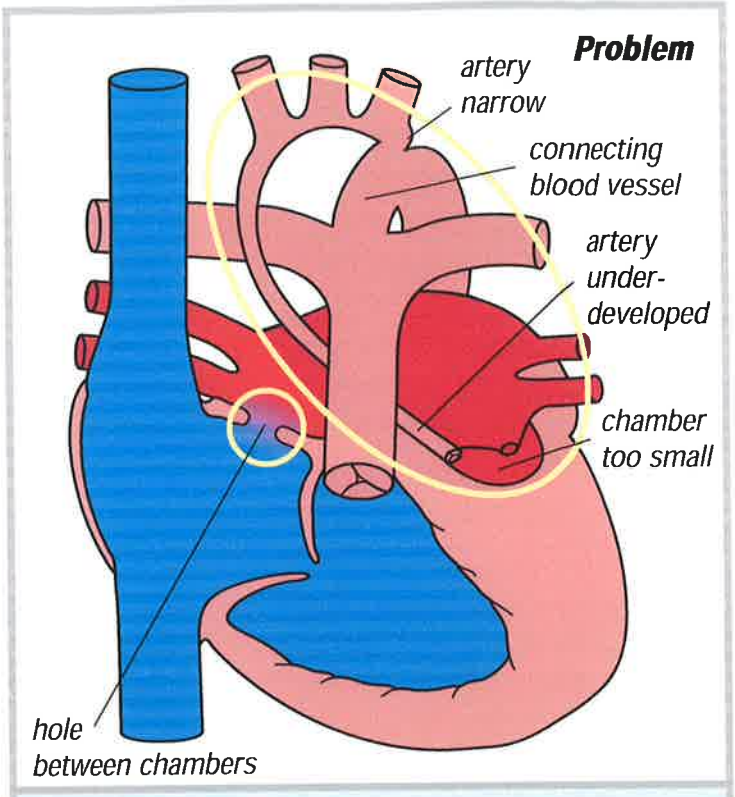

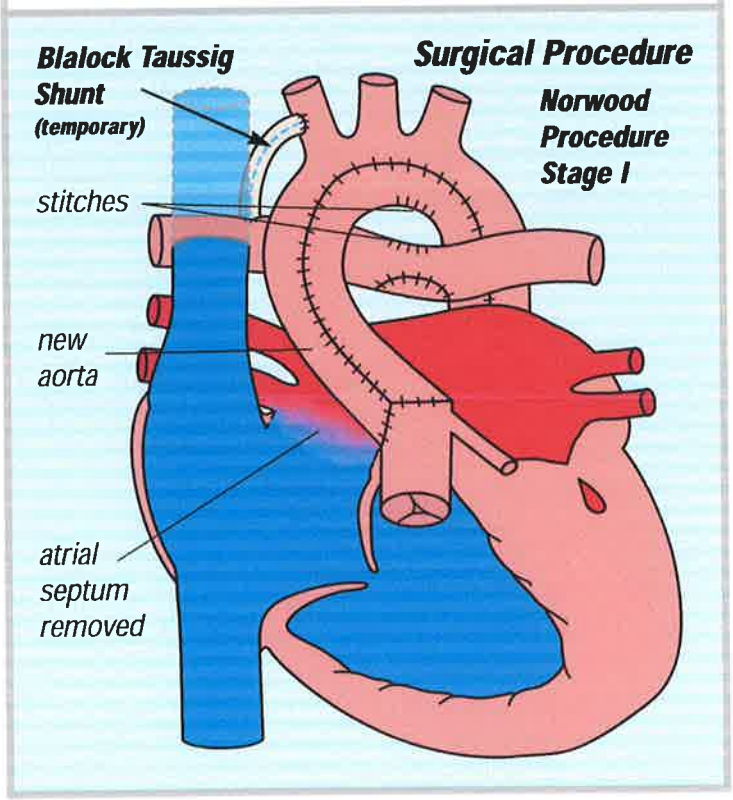

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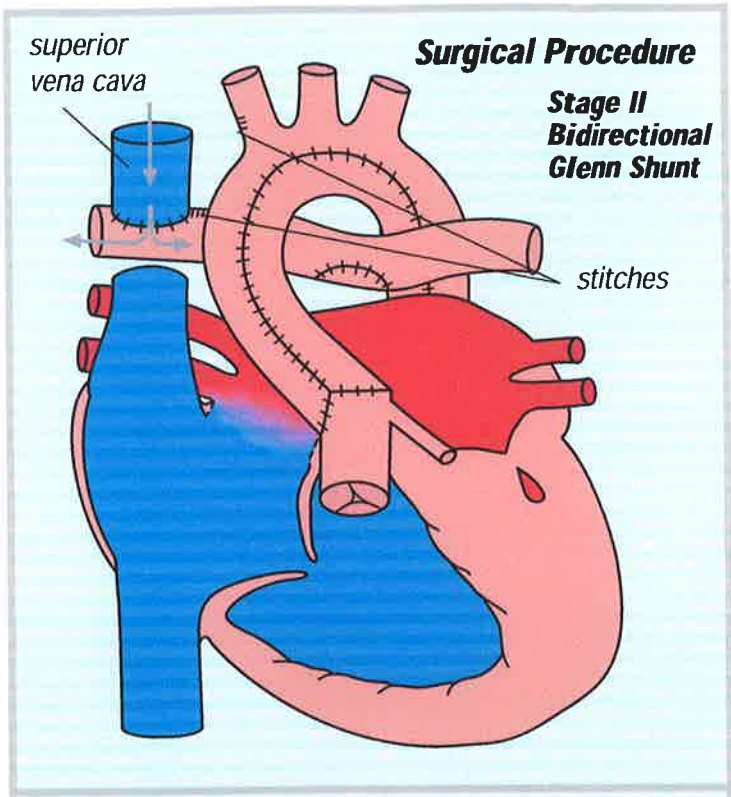
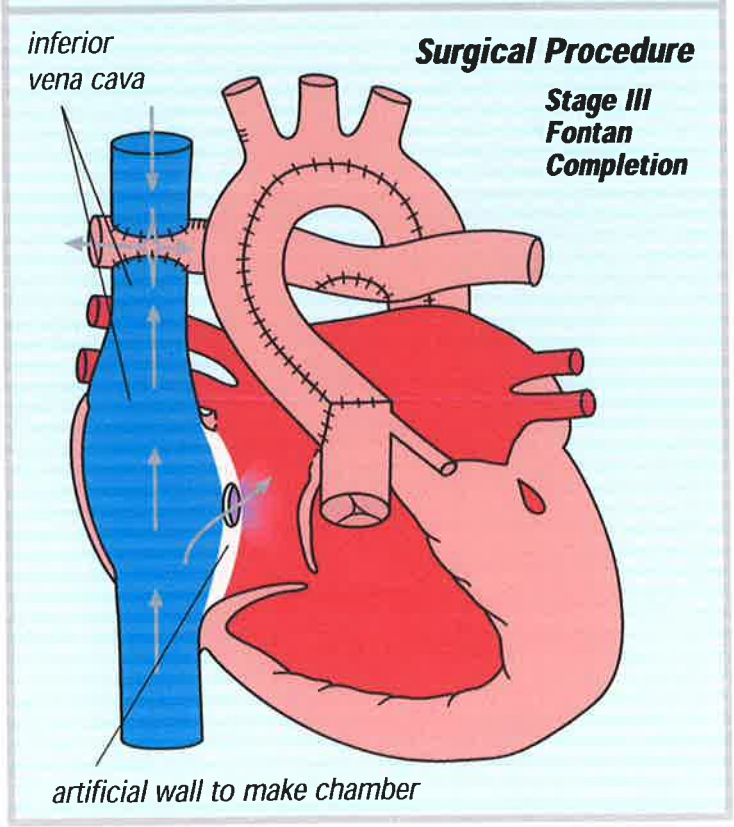
Stage II - Bidirectional Glenn Shunt - The blue blood from the head, neck and upper body is directed to the right lung artery through the superior vena cava. This allows the blood to flow into the lungs for oxygen. The shunt from the Stage I operation is removed. The Stage II procedure reduces the work load of the heart.



Stage III - Fontan Completion - The blue blood from the lower part of the body is sent to the lungs. This is done using the inferior vena cava, the right atrial wall and/or artificial material (Gortex®). A tube is placed through the top right part of the heart and connected to the right pulmonary artery branch. This rerouting of the blue blood allows it 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. Some small holes are placed in the tube to allow the heart and body to adjust gradually to the new blood flow system. These small holes (fenestrations) close off on their own a few months after surgery. The surgery goal is to:

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



Norwood Stages I, II and **III** are done through a median sternotomy (chest) incision.