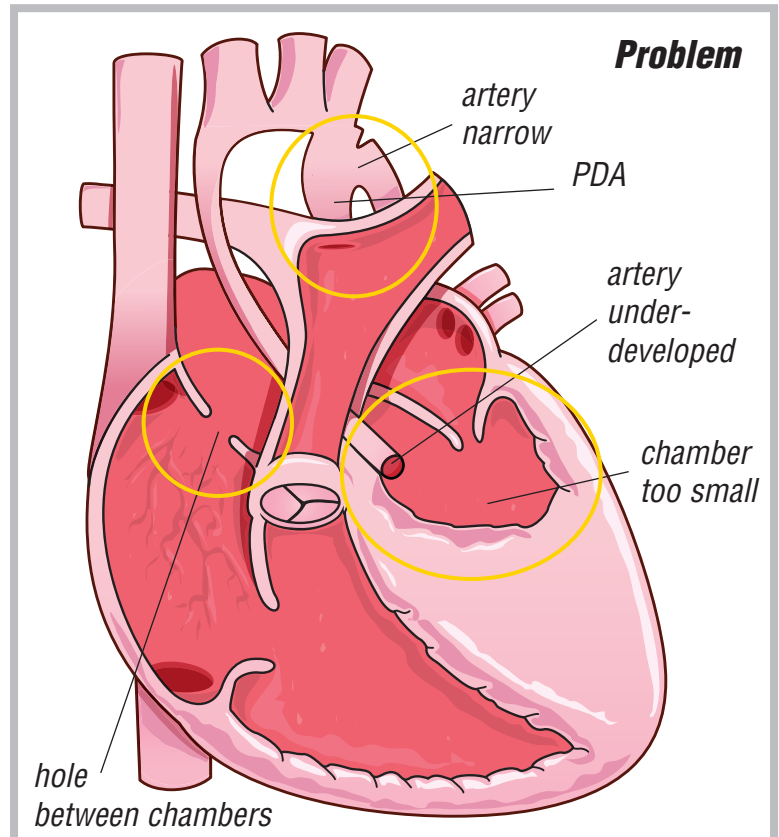


Hypoplastic Left Heart Syndrome (HLHS)

Hypoplastic left heart syndrome (HLHS) is a serious heart defect. With HLHS, the left side of the heart never fully develops during pregnancy. So the right side of the heart must pump blood to both the lungs and the rest of the body. This reduces blood flow to organs and if not treated, can lead to shock or even death. Thanks to advances in care, the outlook for babies with HLHS is better now than in the past.

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) or 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) or 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 or PDA).
- There is also a small hole between the top two chambers (atrial septal defect).



The surgical options for HLHS include cardiac transplantation (replacing the child's entire heart with a donated heart), or a series of 3 operations. These operations will not correct the defect, but will rebuild the heart so that the single pumping chamber on the right side has only the job of pumping blood out to the body.

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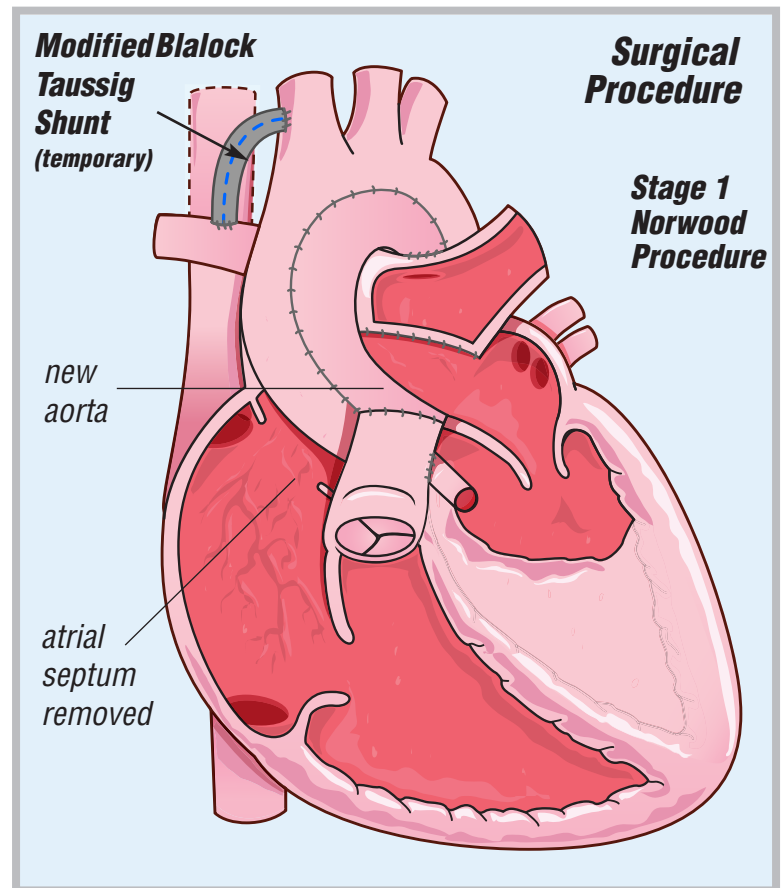
Stage 1 - Norwood Operation using Blalock Taussig shunt

Stage 1

Norwood Procedure - The Norwood Procedure is the first of a series of 3 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.

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 PDA is removed.

All three stages are done through a median sternotomy (chest) incision.



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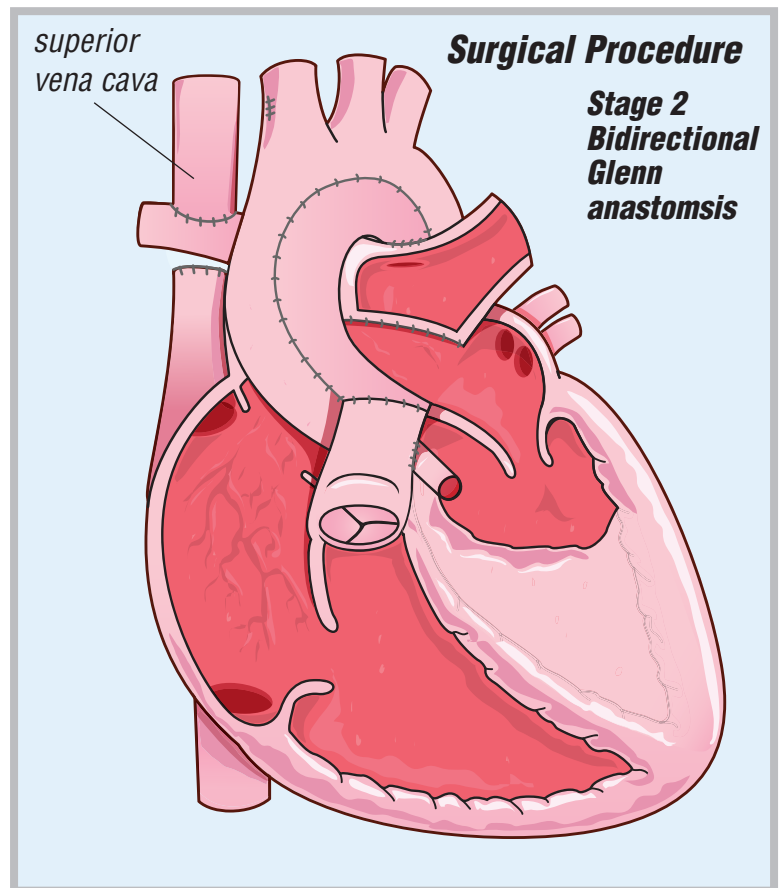
Stage 2 - Bi-directional Glenn Anastomosis

Stage 2

Bidirectional Glenn anastomosis -

The blue blood from the head, neck and upper body is directed to the right lung artery through the superior vena cava. This is done by removing the superior vena cava (SVC) from the top right chamber of the heart and re-attaching it to the blood vessel that supplies blood to the lungs. This allows the blood to flow into the lungs for oxygen. The temporary shunt from the Stage I operation is removed. The Stage II procedure reduces the work load of the heart.

All three stages are done through a median sternotomy (chest) incision.



Hypoplastic Left Heart Syndrome (HLHS)

Stage 3 - Intracardiac Fontan Procedure

Stage 3

Intracardiac 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, the right atrial wall and a baffle (artificial wall). An artificial material is fashioned into a wall through the top right part of the heart and is connected to the pulmonary artery. This 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 so the single right pumping chamber has only one job to perform
- have the blue blood enter the lungs directly (without having to be pumped through the heart)
- have the heart itself as the pump of oxygen-rich blood to the body as its only job or function

All three stages are done through a median sternotomy (chest) incision.

