

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.





Stage 1 - Norwood Operation using Sano Modification

Stage 1

Norwood Procedure using Sono Modification

The large main pulmonary artery and small aorta are fashioned together to make a new, larger aorta.

The wall that separates the top two heart chambers is removed (atrial septectomy). The PDA is removed.

A connection is made between the bottom right heart chamber and the branch pulmonary arteries going to the lungs. The connection is made with a tube (conduit). The conduit is sewn into the pulmonary artery branches at one end. The other end is sewn into the right ventricle (lower right chamber). This conduit allows blood to be pumped from the right ventricle to the lungs. Blood will also be pumped from the right ventricle into the aorta and out to the body.

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



