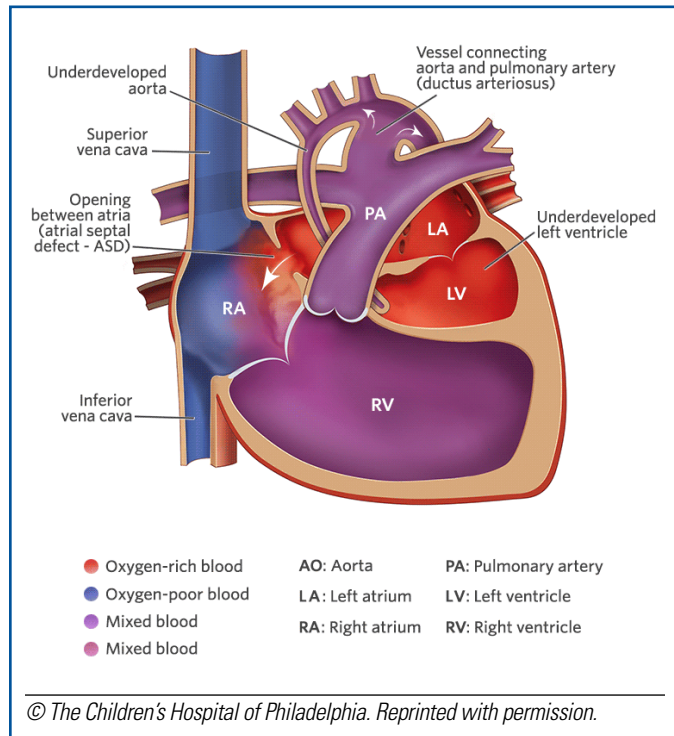




Cardiac Defects: Hypoplastic Left Heart Syndrome



Hypoplastic left heart syndrome (HLHS) is a severe congenital heart defect in which the left side of the heart is underdeveloped.

The heart's left side has the job of pumping oxygenated blood into the aorta, the large artery that carries blood to the body. In a child with HLHS

- The mitral valve, which separates the two left chambers of the heart, may be too small or completely closed (atretic).
- The left ventricle (the lower, pumping chamber) may be very small.
- The aortic valve, which separates the left ventricle and the aorta, may be too small or completely closed (atretic).

In addition to the most common form of HLHS, there are a number of complex cardiac conditions with variations in the structures as described. In these children, where one ventricle also is small (sometimes called "HLHS variants"), the treatment strategy is similar to those with the more typical HLHS.

What are the symptoms of HLHS?

The following symptoms of HLHS may be present at birth or several days later:

- blue or purple tint to lips, skin, and nails (cyanosis)
- difficulty breathing
- difficulty feeding
- lethargy (sleepy or unresponsive).

How is HLHS diagnosed?

Often, HLHS is diagnosed before birth, with fetal echocardiogram (ultrasound). Your baby's provider will prepare a plan for delivery and care immediately after birth.

Sometimes HLHS is diagnosed hours or days after birth and the baby will need immediate therapy. Diagnosis of HLHS may require some or all of these tests:

- echocardiogram (also called echo or ultrasound)—sound waves create an image of the heart
- electrocardiogram (ECG)—a record of the electrical activity of the heart
- chest X ray
- pulse oximetry—a noninvasive way to monitor the oxygen content of the blood
- cardiac catheterization—a thin tube is inserted into the heart through a vein and/or artery in either the leg or through the umbilicus ("belly button")
- cardiac magnetic resonance imaging—a three-dimensional image shows the heart's abnormalities.

Your baby will need intravenous medicines and, possibly, a ventilator for help with breathing. Cardiologists and cardiac nurses will immediately begin procedures to help stabilize your baby.

What are the treatment options?

HLHS is most often fatal without early intervention. It will typically require open heart surgery to redirect the oxygen-rich ("red") blood and oxygen-poor ("blue") blood in a series of three reconstructive operations known as staged reconstruction.



Stage I—Norwood Procedure

Stage I, known as the Norwood procedure, occurs within a few days of birth. In Stage I of reconstruction of a heart with HLHS, the shunt used is called a Blalock-Taussig shunt. Alternative types of shunts may be used based upon a child's individual anatomy.

Alternative approaches to the Stage I Norwood procedure may be recommended for neonates who are in more critical condition, such as heart transplantation or a combination of surgery and catheter-based treatment called a hybrid procedure. The hybrid treatment has been shown to be a feasible alternative to the Norwood procedure. Hybrid treatment avoids major surgery for your child while still providing adequate survival rates and potential neurodevelopmental improvement (Yerebakan et al., 2016). Compared with 25 years ago, there are now many different options for treatment of this complex heart condition; an individualized approach is taken for each and every child. Your baby's provider will explain each individual option, and why one particular approach might be recommended for your child.

Stage II—Glenn Procedure

Stage II, known as the *bidirectional Glenn* or the *hemi-Fontan*, typically occurs within 4 to 6 months of birth.

Stage III—Fontan Procedure

Stage III, known as the *Fontan procedure*, typically occurs between 1 ½ to 4 years of age. In Stage III of reconstruction of a heart with HLHS, a technique called an extracardiac Fontan is used. The small hole intentionally placed to connect the conduit to the right atrium is called a *fenestration*. In some children, a different modification, termed a *lateral tunnel fenestrated Fontan* is used. Your baby's provider will explain the differences and why one might be recommended for your child.

Frequent surveillance in infancy and early childhood is important to minimize risk factors for the eventual Fontan operation. Your child also will need a customized series of diagnostic tests between the planned stages of surgery, and throughout childhood. Additional surgical or catheter therapies, or in rare cases heart transplantation, also may be recommended.

After these operations

- The right side of the heart will do what is usually the job of the left side—pumping oxygenated blood to the body.
- The deoxygenated blood will flow from the veins to the lungs without passing through the heart.

What is the follow-up care for HLHS?

Between the Norwood and Glenn Procedures

Though early outcomes for patients with single ventricle heart defects after staged reconstruction have improved dramatically, the period between the Norwood procedure and the Glenn procedure remains a very vulnerable time for infants. Your baby's provider will focus on the care and monitoring of your baby between the first and second reconstructive surgeries.

Through Age 18

Children who have had surgical reconstruction for HLHS require lifelong care by a cardiologist experienced in congenital heart disease. Sometimes they experience serious health problems. Many remain on medication, and additional surgeries may be required.

Patients with Fontan circulation are referred to as single ventricle patients. As these patients get older, doctors are recognizing that, although some do fine, many experience complications, including lung, liver, and gastrointestinal diseases.

In addition, as a group, children with complex congenital heart defects who have had open-heart surgery as infants are at a higher risk for neurodevelopmental issues when compared with children without congenital heart defects.

Your baby's cardiologist will follow your baby until he or she is a young adult, coordinating care with the primary care providers.

References

- Yerebakan, C., Valeske, K., Elmontaser, H., Yööröker, U., Mueller, M., Thul, J., . . . Akintuerk, H. (2016). Hybrid therapy for hypoplastic left heart syndrome: Myth, alternative, or standard? *The Journal of Thoracic and Cardiovascular Surgery*, 151(4), 1112–1123.

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