

Hypoplastic Left Heart Syndrome (HLHS)

What is it?

Hypoplastic left heart syndrome (HLHS) is a birth defect that affects the blood flow through the heart. Normally, the left side of the heart pumps blood to the body and the right side pumps blood to the lungs. In HLHS, the left side of the heart does not form correctly. HLHS affects multiple parts of the left side of the heart including the **left ventricle, mitral valve, aortic valve**, and part of **the aorta** that aren't developed or are very small. The left side of the heart cannot pump oxygen-rich blood to the body like normal. Babies with HLHS may also have a hole between the atria of the heart, called an **atrial septal defect**. A baby with HLHS also might have problems breathing, a pounding heart, a weak pulse, or blue skin color.

How common is it?

About 1 in every 4,300 babies is born in the United States each year with HLHS. That's about 960 babies a year.

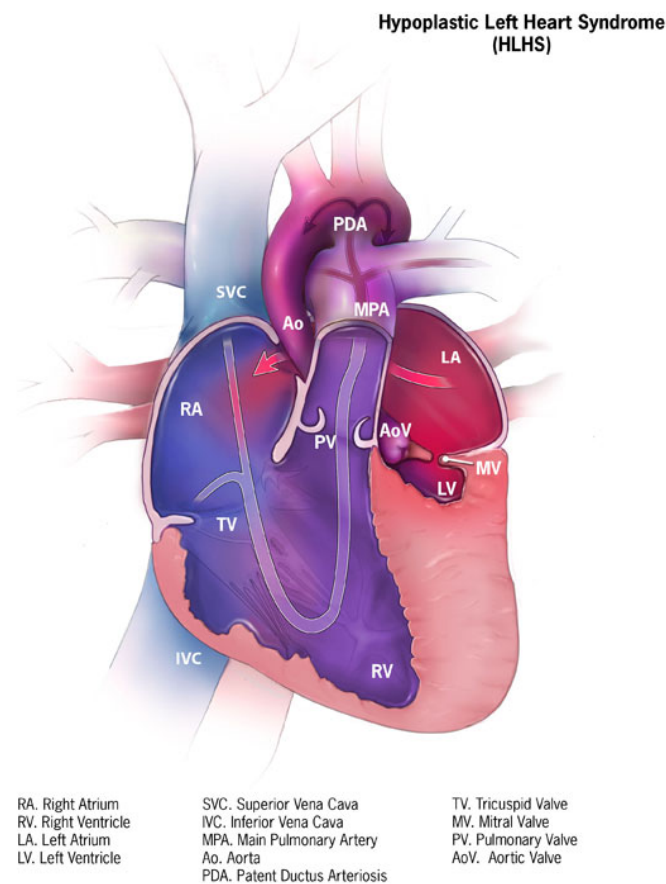


Image courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities

What causes it?

The cause of HLHS in most babies is unknown. Many factors may cause HLHS. More research is needed to understand the exact cause of it.

How is it diagnosed?

HLHS may be diagnosed during pregnancy or after. During pregnancy screenings are done to check for birth defects. After birth a doctor will do a physical examination. The doctor will see if a baby has blue-colored skin and lips, called **cyanosis**. This may be a sign of low levels of oxygen in the blood. The doctor also will listen to a baby's heart. If the doctor hears a heart murmur, or a "whooshing" sound, that may be a sign of a heart defect. A doctor may also see that a baby is having trouble breathing, a pounding heart, and poor feeding, all of which could be signs of a heart defect. Symptoms often are seen soon after birth in a baby with HLHS. A screening test is done shortly after birth to check for **critical congenital heart defects**, which can cause serious health problems or even death if left untreated. The screening test is called the **pulse oximetry screen**. A pulse oximeter is a tool that detects oxygen levels in blood. Low levels of oxygen in the blood could mean there is a heart defect. If a baby fails the screening, then the doctor should perform a diagnostic test called an **echocardiogram** to check for defects in the heart.

How is it treated?

HLHS is treated by multiple surgeries done in a certain order. This will increase the blood flow to the body by avoiding the poorly functioning left side of the heart. Surgery will help bring back the correct function of the heart. The three surgeries usually done are:

- **Norwood Procedure:** Usually done within the first 2 weeks of life
- **Bi-directional Glenn Shunt Procedure:** Usually done between 4 and 6 months of age
- **Fontan Procedure:** Usually done between 18 months and 3 years of age

Infants who have the surgeries can have lifelong complications. Medicine might be needed to help strengthen the heart muscle, lower the child's blood pressure, and help get rid of extra fluid in the body. Some babies may need high-calorie formula or even a feeding tube. Your child's doctor should discuss treatment options with you. Infants with HLHS will need regular follow-up visits to a **cardiologist**, a doctor who specializes in the heart.

For more information:

American Heart Association

http://www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/About-Congenital-Heart-Defects_UCM_001217_Article.jsp#.Wv2YtPnwbc

Centers for Disease Control and Prevention

<https://www.cdc.gov/ncbddd/heartdefects/hlhs.html>

