UNDERSTANDING HYPOPLASTIC LEFT HEART SYNDROME

CARMEN AND JOHN THAIN CENTER FOR PRENATAL PEDIATRICS

What is hypoplastic left heart syndrome?

Hypoplastic left heart syndrome (HLHS) is a condition in which the structures of the left side of the heart are underdeveloped and unable to function properly at birth. Specifically, the left ventricle, the aorta, the aortic and mitral valves, and the aortic arch are too small to be able to support circulation to the body, with varying degrees of severity.

In the normal heart, unoxygenated blood returns from the body to the right atrium and then the right ventricle, where it is pumped through the pulmonary artery to the lungs. After the blood is oxygenated in the lungs, it returns to the left atrium of the heart and is passed to the left ventricle. From the left ventricle it is pumped out to the body through the aorta. In HLHS the left ventricle is too small and weak to provide the necessary blood to the body. (See the diagram for an illustration of this). HLHS is fatal if left untreated.

How common is HLHS and what causes it?

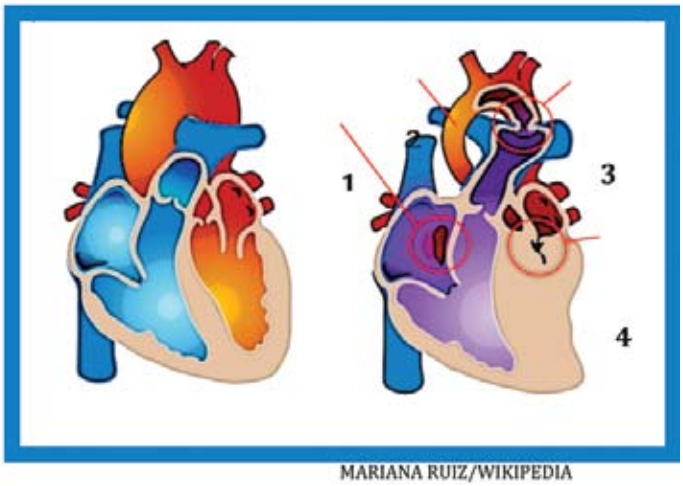
HLHS is one of the most common serious heart defects in newborn babies, seen in 1-4 out of every 10,000 live births. It is believed to make up 9% of all cases of heart defects present at birth, and is twice as common in boys as girls.

No one knows for sure what causes HLHS, but it likely happens very early in pregnancy when the heart is developing, around 5-8 weeks of embryonic life.

How is HLHS detected during pregnancy?

HLHS is usually detected during pregnancy through the ultrasound (sonogram) performed to check fetal development in the second trimester. In looking at the four chambers of the fetal heart, the left ventricle appears much smaller than the right ventricle, and the other structures of the left side of the heart will appear different as well.

The diagnosis of HLHS is usually made following a fetal echocardiogram, which is an ultrasound of the developing heart. At NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center,



MARIANA RUIZ/WIKIPEDIA

Illustration represents symptoms of Hypoplastic Left Heart Syndrome: 1) Opening between Atria; 2) Very Small Aorta; 3) Vessel Connecting Aorta; 4) Underdeveloped Left Ventricle

all fetal echocardiograms for HLHS are performed by Pediatric Cardiologists with expertise in the technique. Unfortunately, some cases of HLHS evolve over the course of the pregnancy and can not be detected until late in the third trimester. Of the cases diagnosed prenatally, almost all are confirmed after birth.

How will my pregnancy be managed now that HLHS has been detected?

An ultrasound will carefully examine the fetal development and exclude any other birth defects that are more frequently seen with HLHS, such as other types of heart defects, abnormalities in the digestive system, and brain malformations. Prenatal care should be managed by a Maternal-Fetal Medicine specialist (MFM), an obstetrician with special training and expertise in high-risk pregnancy. A fetal echocardiogram will be performed by a Pediatric Cardiologist to confirm the diagnosis and exclude other heart problems. Furthermore, a procedure called amniocentesis may be offered to you to examine the baby's chromosomes, since HLHS babies are more likely to have a chromosomal abnormality.

As your delivery date approaches, induction of labor or a cesarean section may be planned to optimize the delivery and ensure that the necessary specialists are available to care for the baby immediately after birth. However, labor management does not need to be changed because your baby has HLHS.

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How will the HLHS be treated after birth?

Until recently, HLHS was considered so serious that hope for survival was grim, and most HLHS babies were given comfort care and allowed to pass away on their own a few days after birth. HLHS remains a serious condition, but the surgical options have improved the outlook for these babies, and more parents are opting for surgery. In some cases, where the condition is so severe it can not be improved with surgery, heart transplantation may be the only option for survival.

The three surgeries needed to treat HLHS are complex but basically involve making the right side of the heart do the work of the left side as well. The details and timing of each stage are individualized for each case, so be sure to discuss them in detail with your medical team. If you decide that you would like your baby to have surgery, you might expect the following:

1. The first stage is called the **Norwood operation**, and takes place soon after birth. The heart is reconstructed to allow the right ventricle to pump blood to the body (instead of the left ventricle, which is too small), and something called a BT Shunt is performed which allows the right ventricle to maintain blood flow to the lungs.
2. At around three to eight months of age, the lungs will be mature enough for the second surgery, known as the **bi-directional Glenn**. This surgery reduces the workload of the right ventricle so it only pumps blood to the body (instead of both the body and lungs) and allows most of the blood from the body to flow directly into the lungs.
3. Between ages two and five, the final surgery is performed, known as the **Fontan operation**. This surgery allows all of the blood from the body to flow directly to the lungs by creating a channel through the heart to direct it. The result is a complete separation of the unoxygenated blood (from the body) and the oxygenated blood (from the lungs), such that all the blood from the body flows into the lungs directly, and the right ventricle pumps the blood from the lungs to the body.

What is the long-term outlook for babies with HLHS?

Long-term outcome for babies with HLHS has improved over the years as surgery has become better and heart transplantation improved. At NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center, we have the most experienced surgical team in the tri-state area, and almost all cases of HLHS detected at birth at other hospitals come to us.

Because surgery for HLHS is so new, the long-term effects of the condition or multiple surgeries are unknown. More studies are needed to determine the impact of HLHS and its treatment on the intellectual development of affected children and on the quality of life they can expect.

What are the chances I could have another baby with HLHS?

In most cases, HLHS occurs due to chance alone, so the chances for a second affected pregnancy are generally only 2-3%, though some studies suggest a higher chance of up to 13.5%. Speak with your Genetic Counselor and Geneticist to determine your specific chances for another affected pregnancy.

What can I expect from the specialists at Morgan Stanley Children's Hospital?

The specialists at Morgan Stanley Children's Hospital are among the most experienced with HLHS in the United States. The Center approach includes a whole team in your prenatal care to optimize the chances for your baby's well-being after birth. You might expect to meet the following specialists:

- **Maternal-Fetal Medicine** (MFM)—you will see one of a team of MFMs throughout your pregnancy, and every effort will be made for that doctor to deliver your baby.
- **Pediatric Cardiology**—Pediatric cardiology performs fetal echocardiograms in the prenatal setting to diagnose HLHS; all cases are confirmed after birth.
- **Pediatric Cardiothoracic Surgery** (CT Surgery)—A recently released report by the New York State Department of Health has shown that NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center and the NewYork-Presbyterian Komansky Center for Children's Health/Weill Cornell Medical Center are leaders in the state for best outcomes for surgeries performed on children with congenital heart defects.
- **Genetics**—While you may have already met with Genetics during pregnancy, you will likely speak with them again after the baby is born.
- **Neonatology**—since the Center's inception, our top-ranked 50-bed NICU regularly cares for babies with HLHS. Our staff has been continuously recognized for their excellence.

You should discuss with your Care Coordinator how these specialists will be involved in your care during pregnancy. The well-being of you and your baby are extremely important to everyone involved in your care. Together we are all dedicated to giving you the best pregnancy and healthiest outlook for your child.

About the Carmen and John Thain Center for Prenatal Pediatrics

Complex pregnancies receive better care when specialists collaborate. The Carmen and John Thain Center for Prenatal Pediatrics is dedicated to helping pregnant women and their families when a birth defect or genetic syndrome is detected before the baby is born. The Center offers sensitive, complete, up-to-date information and testing, and an integrated approach to care that begins in the prenatal period and continues after birth with pediatric follow-up. A collaborative, coordinated program of care is created among specialists in perinatology, neonatology, genetics, pediatric cardiology, pediatric surgery and all pediatric subspecialties.