



Hypoplastic Left Heart Syndrome

A GUIDE TO HELP UNDERSTAND YOUR BABY'S HEART



PROVIDED BY THE
CENTER *for* ADVANCED FETAL CARE
the FETAL HEART PROGRAM



UNIVERSITY *of* MARYLAND
MEDICAL CENTER

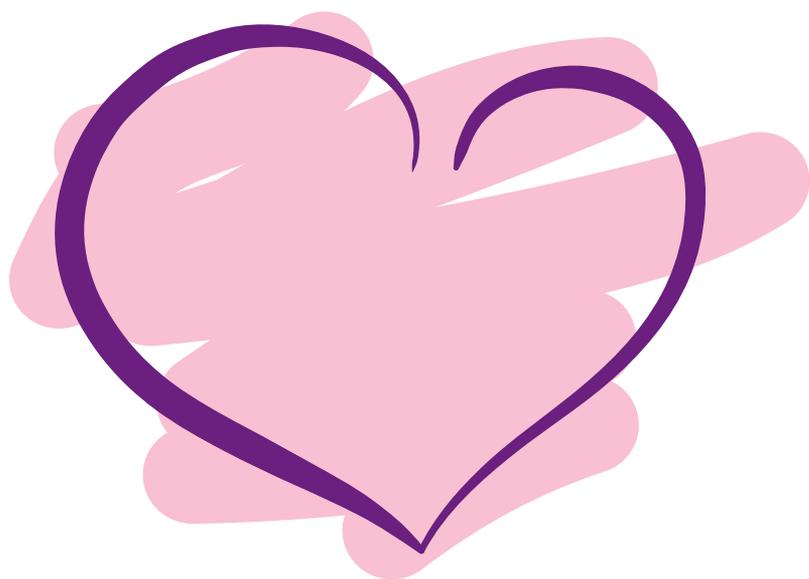


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This guide will help you understand your child's heart. It is not a diagnosis and should never be used instead of medical advice.

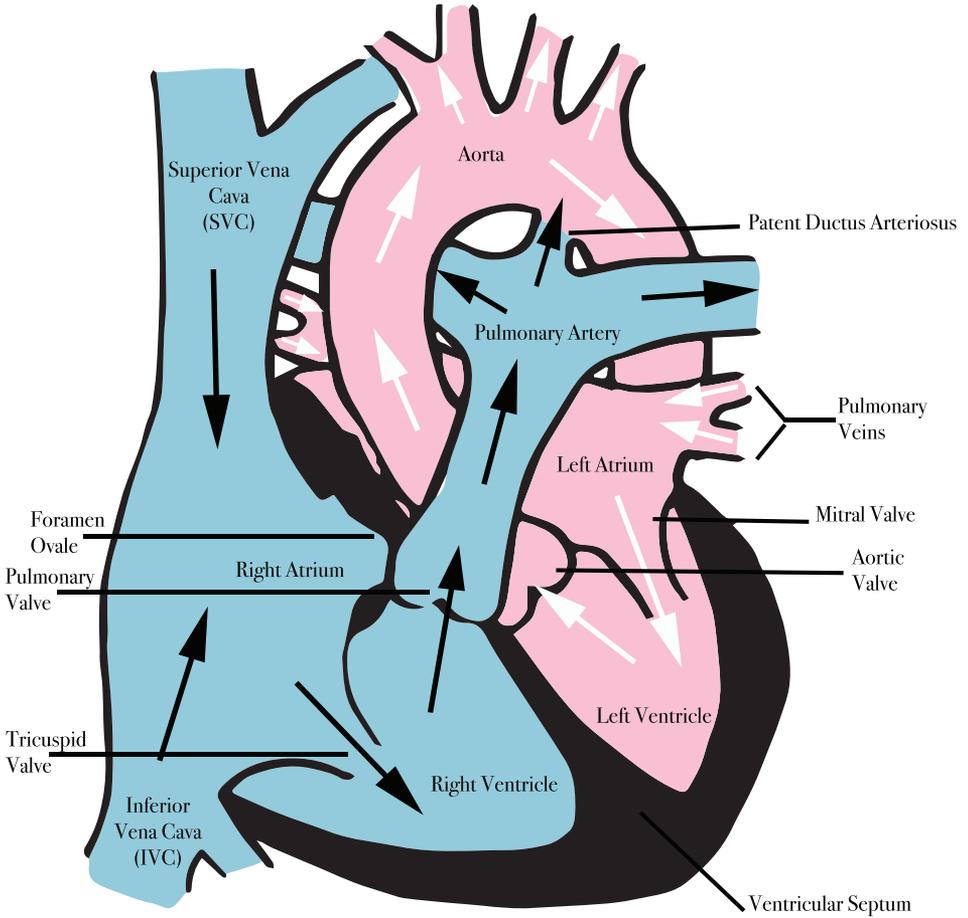
Our goal at the Center for Advanced Fetal Care is to assist you on the journey ahead and help educate you to better communicate with your team of physicians, friends and family.

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Illustrations by Dr. Alicia Chaves



The Normal Heart



The heart is a complex organ which pumps blood through the body. It drives the circulatory system, which carries oxygen and nutrients to the vital organs through a system of arteries and veins.

The heart has four chambers. The top two chambers are called the atria, which are separated by the atrial septum. The bottom two chambers are the ventricles, which are separated by the ventricular septum. As blood passes through each individual chamber, it exits through a valve. Each side of the heart works as its own pump.

Pump 1 - Right side pumps blood to the lungs. (blue on diagram) Blood travels from the right atrium through the tricuspid valve into the right ventricle. From the right ventricle, it travels through the pulmonary valve into the pulmonary artery to the lungs. Oxygenated blood from the lungs returns to the left atrium through the pulmonary veins.

Pump 2 - Left side pumps blood to the body. (pink on diagram) From the left atrium, oxygenated blood travels through the mitral valve to the left ventricle. Then, blood exits through the aortic valve to the aorta, the main artery, sending blood to the rest of the body. Once blood has supplied oxygen to the vital organs, it returns to the right atrium through the inferior vena cava (IVC) and the superior vena cava (SVC), the main veins of the body, to begin the process again.





What is a congenital heart defect?

A congenital heart defect is an abnormality of the structure and/or function of the heart. The defect typically develops during the early stages of pregnancy.

Why did this happen to my baby?

A congenital heart defect is the most common abnormality found in babies. Congenital heart disease may occur due to environmental factors, chromosome abnormalities, or genetic conditions; the majority of heart defects are multifactorial, which means that it occurs because of interactions between genes, chance, and the environment. In most cases, there is not an explanation for why a baby is born with a heart defect.

It is important to remember that any baby can have a congenital heart defect. Neither parent is to blame. Our hospital offers monthly support groups for families with children born with a congenital heart defect.

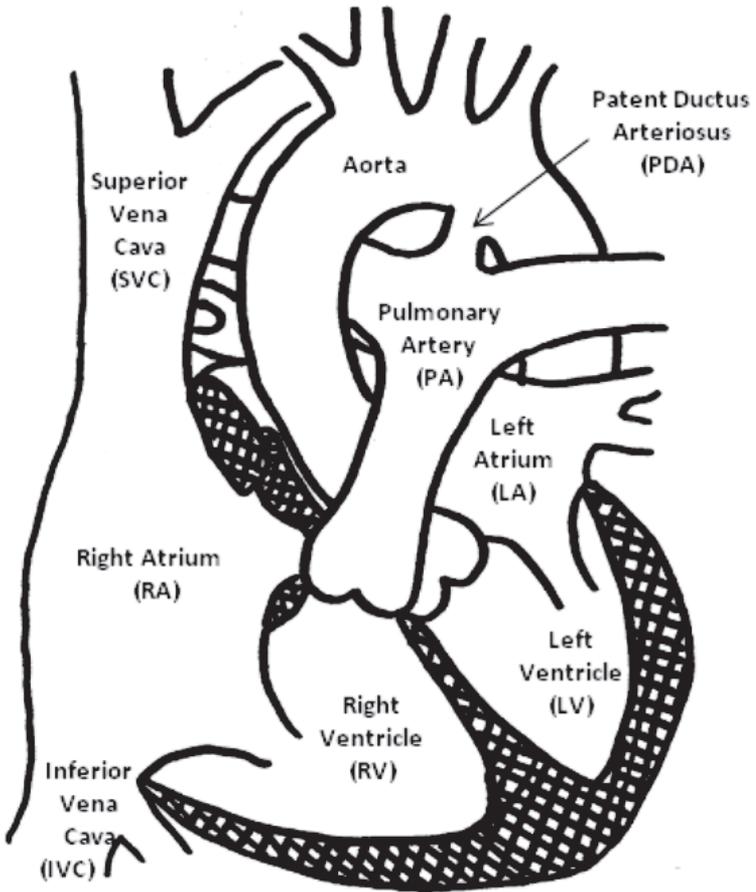
For more information, visit the fetal heart program online at: umm.edu/programs/fetalheart/patient-information/resources



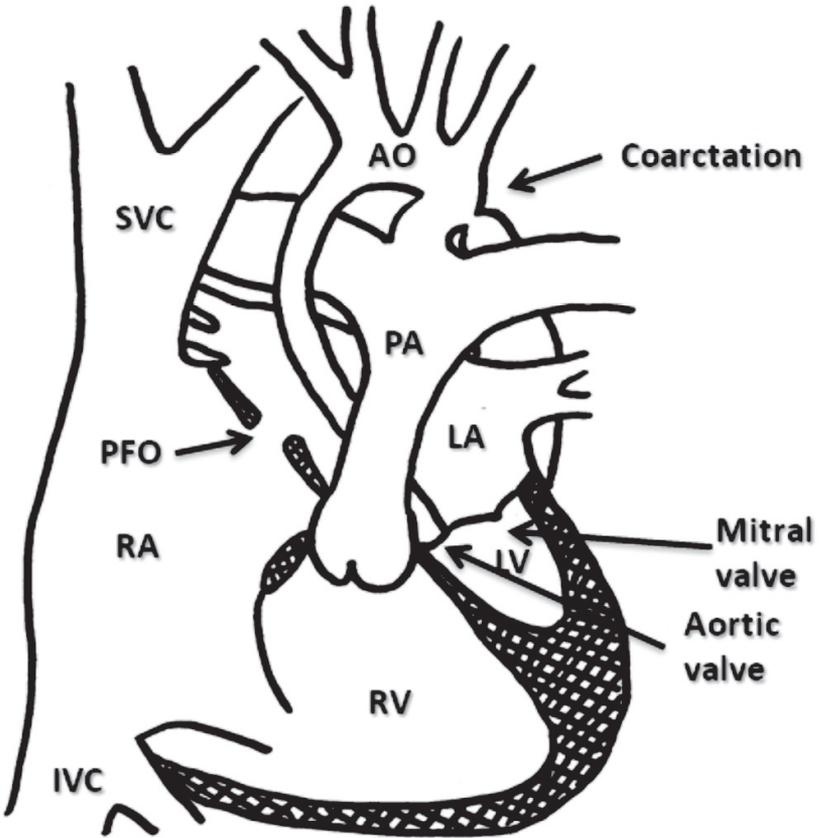
What is Hypoplastic Left Heart Syndrome?

Hypoplastic left heart syndrome (HLHS) is a severe congenital heart defect (CHD). The left side of the heart is small, or hypoplastic. The left ventricle, mitral valve, aortic valve and aorta are all underdeveloped, causing the left side of the heart to not send enough blood to the body.

Normal Heart



Hypoplastic Left Heart Syndrome



For more information visit the fetal heart program online at:
umm.edu/programs/fetalheart/health-information/services/hlhs

How is Hypoplastic Left Heart Syndrome Diagnosed?

HLHS is often diagnosed by fetal ultrasound before the baby is born. Sometimes, though, HLHS is not detected before birth. A baby may initially appear healthy, even though his/her oxygen level will be slightly decreased. The baby may develop difficulty with breathing, have fast breathing, poor feeding, or have cool and pale extremities. If HLHS or another heart defect is suspected, the baby will be evaluated by a pediatric cardiologist. That evaluation would include measuring the oxygen level, and having both an electrocardiogram (ECG) and an echocardiogram (ultrasound of the heart). The postnatal echocardiogram would show the abnormal structure of the heart.

What causes Hypoplastic Left Heart Syndrome?

The cause of HLHS is often unknown. Approximately 10% of patients with HLHS also have other birth defects, and some have genetic disorders. HLHS is more common in families where another family member has HLHS, or other congenital heart defects, such as aortic stenosis or coarctation of the aorta. Babies who have HLHS because of a chromosomal or genetic condition, usually have physical and developmental problems in addition to their heart defect. Many of these chromosomal conditions can be tested for during pregnancy using either a chorionic villus sampling (CVS) or an amniocentesis. These procedures carry a small risk of miscarriage; however, many providers and patients believe that the benefits outweigh the risks. If families choose not to have these tests during pregnancy, all babies with HLHS will be checked for genetic and chromosomal conditions in the newborn period.

What can I expect during my pregnancy?

If your baby is diagnosed before birth with HLHS, a team of specialists will care for you and your unborn baby. The team at the University of Maryland Medical Center includes a maternal fetal medicine specialist (MFM), cardiologists (fetal and pediatric), genetic counselor, neonatologist, and a pediatric cardiac surgeon. Your baby will be monitored closely by fetal ultrasounds and a delivery plan will be discussed among you, your obstetrician, and the various other specialists. Induction of labor may be scheduled for a pregnancy affected by HLHS to ensure that your care team is present at delivery. If there are no maternal or fetal issues other than the baby's HLHS, our goal is for you to have a normal delivery.

What can I expect after my baby is born?

After delivery, your baby will be taken to the neonatal intensive care unit (NICU) for evaluation and to begin cardiopulmonary stabilization. Because a baby with this condition can have sudden changes, the specialists in the NICU will closely monitor your baby's vitals. A postnatal echocardiogram will determine the severity of the HLHS. The immediate treatment for newborns with HLHS is to ensure that the blood from the lungs has a way to get to the body. Surgery is required for a baby with HLHS. Prior to surgery, cardiopulmonary stabilization begins with an echocardiogram to monitor the patent ductus arteriosus (PDA). A medication, prostaglandins, is given through an IV to maintain the PDA. This allows the right ventricle to pump blood to the lungs and the rest of the body. A baby may also need a procedure to enlarge the patent foramen ovale (PFO), to guarantee that the blood returning from the lungs to the left atrium can easily pass into the right side of the heart. Your baby may need the support of a ventilator. Without these treatments or surgery, hypoplastic left heart syndrome can be fatal.

How is Hypoplastic Left Heart Syndrome treated?

HLHS is the one of the most challenging and complex forms of congenital heart disease. Without treatment, this defect is usually fatal within the first weeks of life. The Fetal Heart Program and the Children's Heart Program at the University of Maryland Medical Center has the expertise to diagnose and surgically repair the heart of a baby born with HLHS. Currently, there is no way to make the left side of the heart normal, but there is a series of three surgeries that result in using one ventricle as a pump which successfully gets blood to both the body and lungs. The three surgeries are the Norwood operation, Glenn Shunt, and Fontan procedure, as shown on page 12.

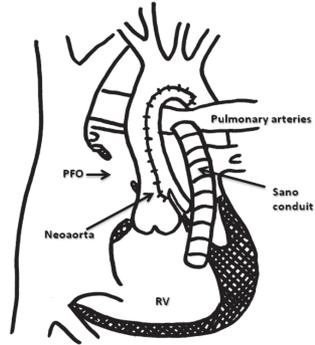
The Norwood operation is the first surgery to stabilize your baby's heart. This open heart surgery typically occurs in the first week of your baby's life. The Norwood accomplishes 3 goals: 1. getting blood to the lungs; 2. getting blood to the rest of the body; and, 3. making sure that blood returning from the lungs can get to the right side of the heart. The surgeon connects the pulmonary artery to the aorta, which creates one large blood vessel that goes from the right ventricle to the body. This means that the branches of the pulmonary artery that go out into the lungs are no longer attached to the heart. A tube connects either the right ventricle or the aorta to the pulmonary artery branches, creating a stable way to get blood to the lungs. Finally, the wall between the right and left atria is cut out, creating an atrial septal defect, which allows the blood returning from the lungs to get to the right side of the heart.

When your baby is 4 to 6 months of age, a Glenn shunt procedure is done. The Glenn shunt is the first step toward separating the lung and body circulations. The large vein that drains the oxygen poor blood from the head and upper body into the right atrium (called the superior vena cava or SVC) is detached from the heart and then attached directly to the pulmonary arteries. This lets the blood drain directly into the lungs. The tube connecting the right ventricle, or aorta, to the pulmonary artery is removed. After this procedure, the right ventricle only has to deliver blood to the body, so it doesn't have to pump as hard. All of the oxygen poor blood from the lower body still goes into the right atrium, but by another large vein, the inferior vena cava, (IVC), then mixes with the blood from the lungs. This means that the blood going to the body still has a decreased oxygen level. Babies typically stay in the hospital for about 7-10 days after surgery.

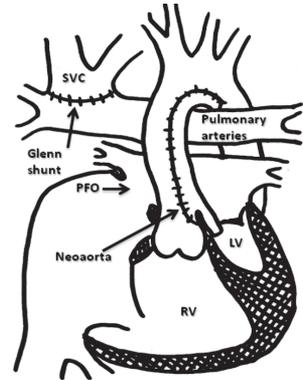
The time between the Norwood surgery and the Glenn procedure is the interstage period. Although most babies are able to be discharged home, it is still a high risk time for babies with HLHS, so we monitor them very closely. We instruct you on what to look for, and give you two machines - one to daily check your baby's oxygen levels, and another for daily weight. Your monitoring and reporting helps timely identify problems. Your baby may have difficulty feeding after the Norwood surgery, and may not be able to take all feedings by mouth. Your baby may require placement of a tube directly into the stomach for feedings.

The final procedure is the Fontan operation. It is generally performed at 2-4 years old. In this surgery, the IVC is removed from the heart and then connected to the pulmonary arteries. Therefore, all of the oxygen poor blood drains directly into the lungs, and the right ventricle pumps oxygen rich blood to the body. Your baby may stay in the hospital for approximately 7-10 days after this surgery.

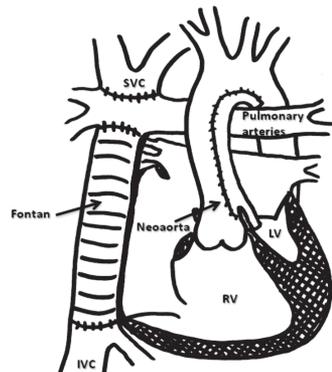
HLHS Norwood



HLHS Glenn



HLHS Fontan



Will my baby have a normal childhood?

Children with HLHS need lifelong follow up care by a cardiologist. In addition to the 3 surgeries that have been described above, there may be other procedures which are recommended by a cardiologist. Most children undergo a cardiac catheterization procedure before the Glenn and Fontan surgeries. Although follow up visits are frequent when young, after completing the series of surgeries as described above, your baby will be seen every 6 to 12 months. At follow up visits, your baby will be checked regularly with ECGs and echocardiograms. Other tests, such as cardiac MRIs, exercise stress tests, or Holter (24 hour ECG) monitors may be used.

Will we have another child with a congenital heart defect?

Studies suggest that if you have one child with a congenital heart defect, your risk of having another child with a heart defect is about 2%-3%. If your baby's congenital heart defect is associated with a chromosome abnormality or a genetic syndrome, a genetic counselor can discuss with you and your family the risk of having another baby with the same condition. In any future pregnancies, we highly recommend nuchal translucency screening with an early fetal echocardiogram during your first trimester. Then, we recommend a targeted anatomy ultrasound between 18-20 weeks, and a fetal echocardiogram between 22-24 weeks.

How can the Fetal Heart Program help?

The Fetal Heart Program at the University of Maryland Medical Center is dedicated to the care and support of you and your unborn child. Our world class program aims to diagnose congenital heart defects as early, and as accurately as possible. We strive to create personalized prenatal care and optimize your delivery plan. Our multidisciplinary team is devoted to you and your baby's needs before and after birth.



It has been
our privilege
to care for you and your child.



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