

HYPOPLASTIC LEFT HEART SYNDROME

What is HLHS?

HLHS is the one of the most challenging and complex form of congenital heart diseases. Without treatment, this defect is usually fatal within the first weeks of life. The Children's Heart Program has expertise in diagnosing and surgically repairing the hearts of babies born with HLHS so that most can live full lives.

Hypoplastic left heart syndrome is a congenital heart defect in which the left side of the heart is small, or hypoplastic. The left ventricle, mitral valve, aortic valve and aorta are all under-developed, and the left side of the heart is unable to send enough blood to the body. This results in the left side of the heart being unable to support the circulation needed by the body's organs.

What causes HLHS?

There is no known cause of HLHS. 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 like aortic stenosis or coarctation of the aorta.

How is HLHS diagnosed? What are signs and symptoms?

HLHS is often diagnosed by fetal ultrasound before the baby is born. Sometimes HLHS is not detected before birth. Those babies may initially appear healthy, although their oxygen levels will be slightly decreased. They may develop difficulty breathing, fast breathing, poor feeding, or 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, an electrocardiogram (ECG) and an echocardiogram (ultrasound of the heart). The echocardiogram would show the abnormal structure of the heart.

Why does HLHS make babies sick?

The left side of the heart is normally responsible for receiving blood rich in oxygen from the lungs and pumping that blood out to the body. In HLHS, the blood that returns from the lungs comes into the left atrium (or collecting chamber), but little or no blood can get into the left ventricle and out to the aorta. This means that there is no way to deliver oxygen and nutrients to the body. Instead, the blood from the left atrium crosses through a hole in the wall separating the left and right atrium (called a PFO) and mixes with the oxygen poor blood that is returning from the body. That mixture of blood will get pumped out to the lungs by the right ventricle.

Before birth and for a short time after birth, babies are able to use other routes to get blood to the body. A connection between the pulmonary artery (artery from the right ventricle to the lungs) and the aorta allows the right ventricle to pump some of the blood out to the aorta. That connection, called a patent ductus arteriosus (PDA), is present in everyone before birth and closes within the first week of life. It is when that PDA closes that babies with HLHS get very sick, as they no longer have a way to get blood to their bodies.

What can I expect after my baby is born?

Babies with these types of heart defects will need evaluation and stabilization in the neonatal intensive care unit after birth. The diagnosis will be confirmed with an echocardiogram, and babies may get large IV's placed. Some babies need a breathing tube placed to help manage their condition. The immediate treatment for newborns with HLHS is to ensure that the blood returning from the lungs has a way to get to the body. Babies are started on an IV medication called prostaglandins to keep the PDA open. This allows the right ventricle to be the pump to both the lungs and the body. Some babies may also need a procedure to enlarge the PFO, to make sure that the blood returning from the lungs to the left atrium can easily pass into the right side of the heart.

Because the care for children with congenital heart disease is complex, your baby will be cared for by a team of skilled clinicians. This starts before birth with maternal-fetal medicine and fetal cardiologists and continues with pediatric cardiologists and nurse practitioners, a pediatric cardiac surgeon, neonatal and pediatric intensive care physicians, pediatric anesthesiologists, pediatric cardiac operating room staff, pediatric nurses and many others.

What is the treatment/surgery for HLHS?

Newborn surgery

These treatments help to stabilize the baby for surgery, but don't provide a long term solution. We don't yet have a way to make the left side of the heart normal, but there is a series of surgeries that allow us to use one ventricle as a pump and still get blood to both the body and lungs. The first surgery happens in the first week or so of life. The procedure is called the Norwood operation. Babies typically need about a month in the hospital to recover from this procedure.

The Norwood operation is an open heart surgery that accomplishes 3 goals: getting blood to the lungs, getting blood to the body and making sure that the blood returning from the lungs can get to the right side of the heart. The surgeon attaches the pulmonary artery and the aorta to create 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 is placed that connects either the right ventricle or the aorta to the pulmonary artery branches and creates a stable way to get blood to the lungs. Finally, the wall between the right and left atria is cut out to allow all the blood coming back from the lungs to get to the right side of the heart.

The Norwood procedure accomplishes the 3 goals, but is not a final solution. The right ventricle has to do extra work, by pumping blood to both the lungs and the body, and the body gets blood that is a mixture of oxygen rich and oxygen poor blood. Also, the tube that gets blood to the lungs will not grow with the baby.

Later surgeries

When babies are 4 to 6 months of age, a second procedure is done. This is called a Glenn shunt, and it is the first step of 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 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 pump the blood to the body, so it doesn't have to work as hard. However, all of the oxygen poor blood from the lower body is still coming into the right atrium by another large vein (called the inferior vena cava, or IVC) and mixing 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 final procedure, called the Fontan operation, takes care of this problem and is usually done around 2 to 4 years of age. In this surgery, the IVC is removed from the heart and connected to the pulmonary arteries. At this point, all of the oxygen poor blood will drain directly into the lungs, and the right ventricle will pump oxygen rich blood to the body. Children typically stay in the hospital for about 7-10 days after surgery.

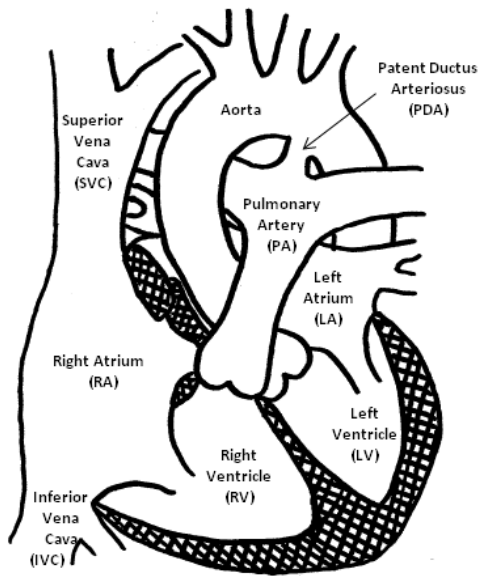
What other procedures or follow up will my baby need?

We refer to the time between the Norwood surgery and the Glenn procedure as the interstage period. Although most babies are able to be discharged home, it is still a high risk time for babies with HLHS, and we monitor them very closely. We instruct parents on what to look for and give you a machine to check their oxygen levels daily and a scale to weigh them daily. This allows us to pick up problems early. Some babies have difficulty feeding after the Norwood surgery and may not be able to take all their feeds by mouth. Some may require placement of a tube directly into their stomach to make it easier to get feeds.

Children with HLHS will need lifelong follow up with a cardiologist. In addition to the 3 surgeries that we expect, they may need other procedures. Most children undergo a cardiac catheterization procedure before the Glenn and Fontan surgeries. Although follow up visits are frequent when they are young, after completing the series of surgeries they will be seen every 6 to 12 months. At follow up visits, children 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.

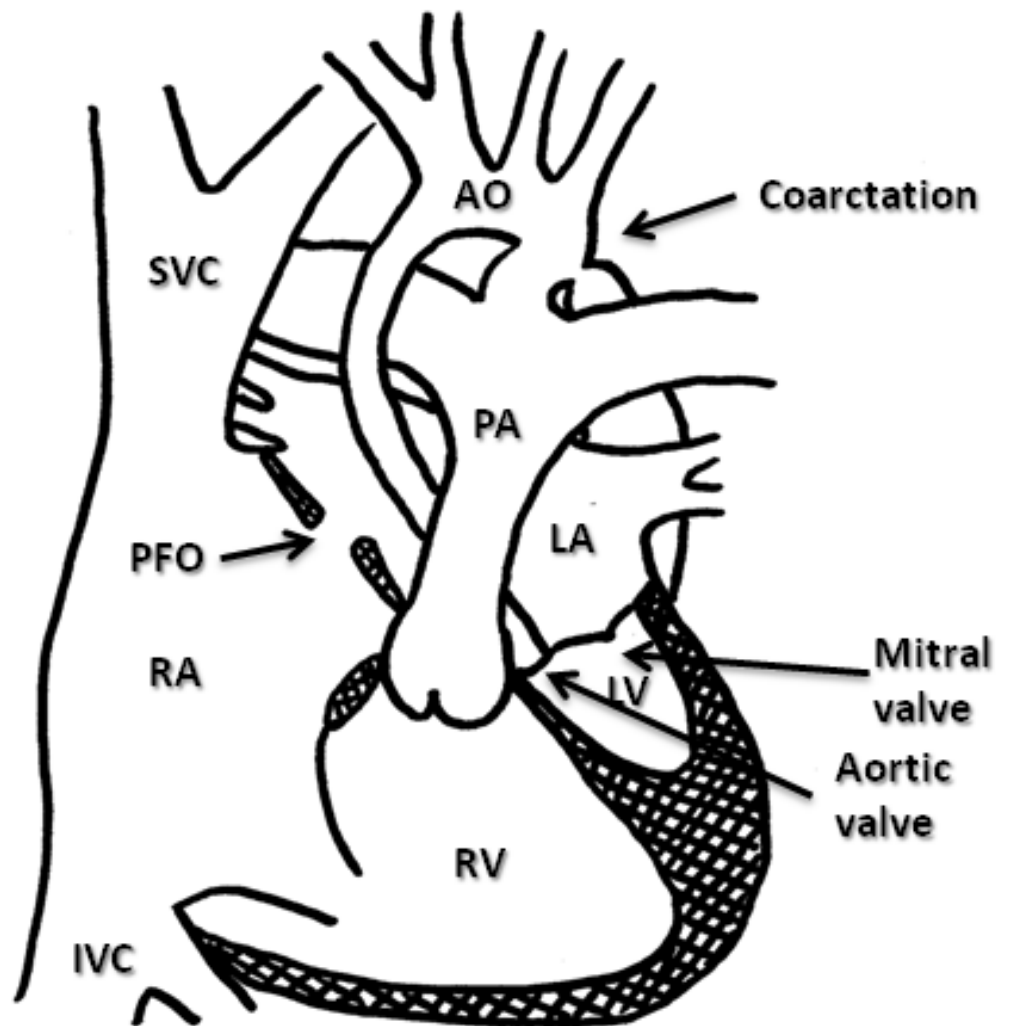
What is the long term outcome for children with HLHS?

Survival for infants with HLHS has been improving as we have perfected our technique and management of this complicated disease. Surgery for HLHS was only first started a few decades ago. Initially there was a high mortality rate; now we expect that over 75 % of infants born with HLHS will survive past five years, and that most of those survivors will also survive into adulthood. Patients with HLHS may need to take medications and may have some limitations in their exercise ability. Some children may have learning difficulties or ADHD. Despite this, they are typically able to have happy and productive lives. As we develop new technologies and ways of approaching this disease, we expect that infants born today with HLHS will have even better outcomes.



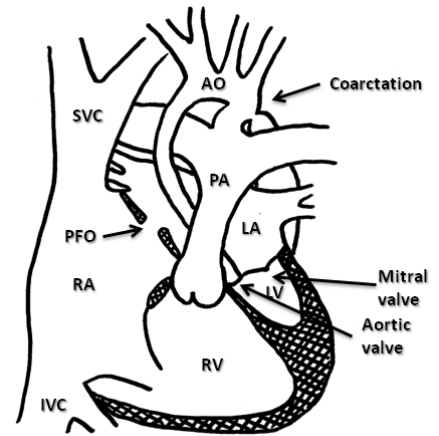
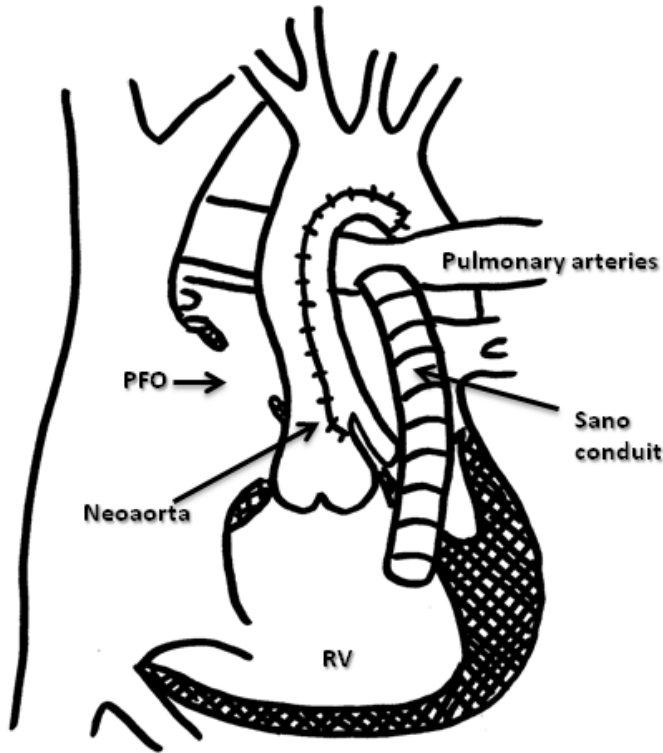
**Normal
Newborn
Heart**

**Hypoplastic
Left Heart
Syndrome
(HLHS)**

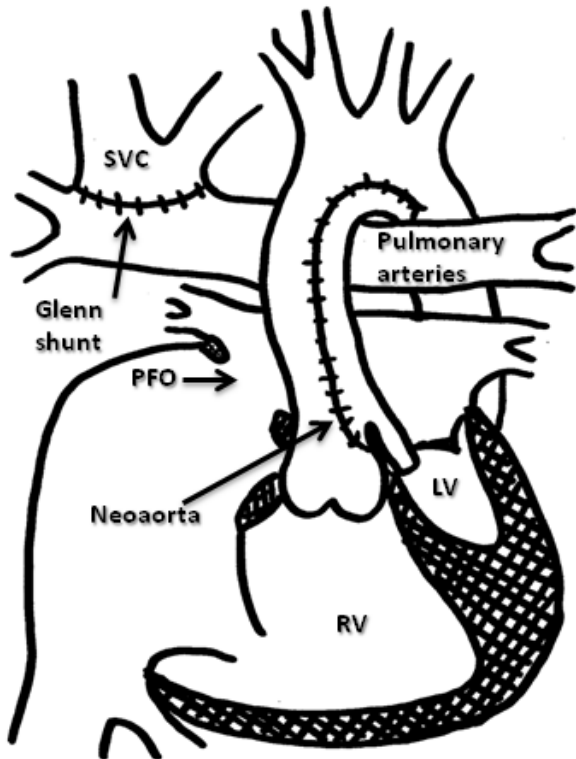


After surgeries:

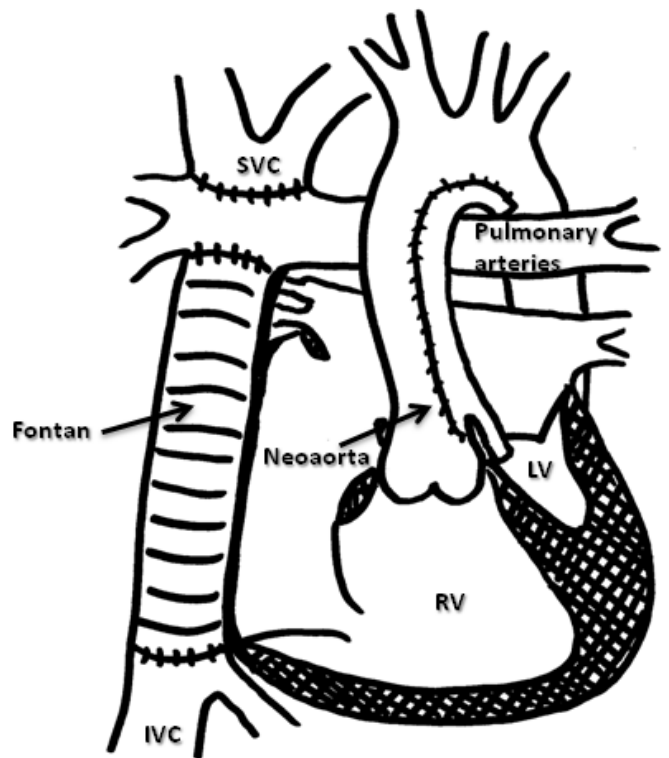
Before surgery



1. Norwood



2. Glenn



3. Fontan