Hypoplastic Left Heart Syndrome (HLHS)

Hypoplastic left heart syndrome (HLHS) is a severe congenital heart defect in which the left side of the heart (mitral valve, left ventricle, aortic valve, and aorta) is underdeveloped. The condition is present at birth. In a child with HLHS:

- The mitral valve is too small or completely closed (atretic).
- The left ventricle is very small.
- The aortic valve is too small or completely closed (atretic).

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

The left side of the heart is unable to send enough blood to the body. As a result, the right side of the heart must maintain the circulation for both the lungs and the body. The right ventricle can support the circulation to both the lungs and the body for a while, but this extra workload eventually causes the right side of the heart to fail.

The only possibility of survival is a connection between the right and the left side of the heart, or between the arteries and pulmonary arteries. Both the foramen ovale and ductus arteriosus normally close on their own a few days after birth. In babies with HLHS, blood from the right side of the heart travels through the ductus arteriosus. This is the only way for blood to get to the body. If the ductus arteriosus is allowed to close, the patient may quickly die because no blood will be pumped to the body. Babies with known HLHS are usually given Prostaglandin E to keep the ductus arteriosus open.

Because there is little or no flow out of the left heart, blood returning to the heart from the lungs needs to pass through the foramen ovale or an atrial septal defect back to the right side of the heart. If there is no foramen ovale, or if it is too small, the baby could die. Those with this problem have the foramen ovale opened, either with surgery or balloon septostomy during a heart catheterization.

Hypoplastic left heart syndrome is rare. It is more common in males than in females and there is no known cause. About 10% of patients with HLHS also have other birth defects. There is no known prevention of this defect. As with many congenital diseases, the causes of HLHS are uncertain and have not been linked to a mother's disease or behavior.
Symptoms
The following symptoms of HLHS may be present at birth or several days later:

- Abnormal heart sounds
- Peripheral and/or central cyanosis
- Difficulty breathing – tachypnea, shortness of breath
- Difficulty feeding
- Lethargy (sleepy or unresponsive)
- Bounding PMI
- Weak peripheral pulses
- Cold extremities
- Liver enlargement if heart failure is present

Diagnosis
Often, hypoplastic left heart syndrome is diagnosed before birth, with fetal echocardiogram. If diagnosed prenatally, the hospital 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
- Electrocardiogram (ECG)
- Chest X-ray
- Pulse oximetry
- Cardiac catheterization
- Cardiac MRI

Initial Care
Once the diagnosis of HLHS is made, the baby will be admitted to the neonatal intensive care unit. A ventilator may be needed. Prostaglandin E1 is given to keep blood circulating to the body by keeping the ductus arteriosus open. These measures do not solve the problem, but stabilizes the infant. The condition always requires surgery.

Surgical Treatment
Treatment of HLHS typically requires open-heart surgery to re-direct the oxygen-rich blood and oxygen-poor blood in a series of three reconstructive operations known as “Staged Reconstruction.” In some hospitals, heart transplantation is considered a better choice than the three-step surgery process. However, there are few donated hearts available for small infants.
Stage I, known as the Norwood procedure, occurs within a few days of birth. The illustration below shows a “Blalock-Taussig” shunt. Alternative types of shunts may be used based upon the individual anatomy. Stage I of the Norwood procedure consists of building a new aorta by:

- Using the pulmonary valve and artery
- Connecting the hypoplastic old aorta and coronary arteries to the new aorta
- Removing the atrial septum
- Making an artificial connection from either the right ventricle or a body-wide artery to the pulmonary artery to maintain blood flow to the lungs, known as a shunt

Afterwards, the baby usually goes home. The child will need to take daily medication and be closely followed by a pediatric cardiologist, who will determine when the second stage of surgery should be done.

Stage II, known as the bidirectional Glenn, typically occurs within four to six months of birth. An alternative technical modification called the “Hemi Fontan” may be used based on a child’s individual anatomy. This procedure connects the superior vena cava directly to the pulmonary arteries. During stages I and II, the child may still appear somewhat blue (cyanotic).

Stage III, the final step, is known as the Fontan procedure, and typically occurs between one-and-a-half to four years of age. The illustration below shows a technique called an “extracardiac Fontan”. In some children a modification, called a “lateral tunnel fenestrated Fontan”, is done. In this procedure, the rest of the veins that carry blood from the inferior vena cava are connected directly to the blood vessels to the lungs. The right ventricle now serves only as the pumping chamber for the body and no longer the lungs and the body. After this final step, the baby is no longer blue.

These staged procedure are the most common for HLHS, however, alternative approaches may be recommended for some children, including transplantation or “hybrid” procedures. An individualized approach is taken for each and every child. 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. Some patients may need more surgeries in their 20s or 30s if they develop hard to control arrhythmias or other complications of the Fontan procedure.
Normal Heart

Hypoplastic Left Heart Syndrome (HLHS)

Vessel Connecting Aorta and Pulmonary Artery (Ductus Arteriosus)

Underdeveloped Aorta

Superior Vena Cava

Aorta

Mitrail Valve

Pulmonary Valve

to Lungs

Pulmonary Venae

from Lungs

Atrial Septum

Tricuspid Valve

Interior Vena Cava

Normal Heart

Hypoplastic Left Heart Syndrome (HLHS)

Undeveloped Left Ventricle

Opening Between Atria (Atrial Septal Defect - ASD)

Superior Vena Cava

Aorta

Mitrail Valve

Pulmonary Valve

to Lungs

Pulmonary Venae

from Lungs

Atrial Septum

Tricuspid Valve

Interior Vena Cava

HLHS: Staged Reconstruction (three stages of surgical repair)

Hypoplastic Left Heart Syndrome (HLHS) Stage I - Norwood

Homograft Patch (To Reconstruct Aorta)

Blalock-Taussig Shunt

Surgically Enlarged Atrial Septal Defect

LA: Left Atrium
PA: Pulmonary Artery
LV: Left Ventricle
RV: Right Ventricle

Oxygen-rich Blood
Oxygen-poor Blood
Mixed Blood

Hypoplastic Left Heart Syndrome (HLHS) Stage II - Bidirectional Glenn

Divided Blalock-Taussig Shunt

Superior Vena Cava Attached to Right PA

Homograft Patch on PA

Left PA

LA: Left Atrium
RA: Right Atrium
LV: Left Ventricle
RV: Right Ventricle

Oxygen-rich Blood
Oxygen-poor Blood
Mixed Blood

Hypoplastic Left Heart Syndrome Stage III Extracardiac Fenestrated Fontan

Conduit to Connect IVC to Right PA

Fenestration Between conduit and RA

Left PA

LA: Left Atrium
RA: Right Atrium
LV: Left Ventricle
RV: Right Ventricle

Oxygen-rich Blood
Oxygen-poor Blood
Complications
Complications include:

- Blockage of the artificial shunt
- Chronic diarrhea from protein losing enteropathy
- Ascites
- Pleural effusion
- Heart failure
- Arrhythmias
- Strokes and other nervous system complications
- Sudden death

Prognosis
If left untreated, hypoplastic left heart syndrome is fatal. Survival rates for the staged repair continue to rise as surgery techniques and care after surgery improve. Survival after the first stage is more than 75%. The child's outcome after surgery depends on the size and function of the right ventricle.

Follow Up Care
Children who have had surgical reconstruction for HLHS require life-long 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, while 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 to children without congenital heart defects.

References