Hypoplastic Left Heart Syndrome (HLHS)

What is Hypoplastic Left Heart Syndrome?

Hypoplastic Left Heart Syndrome (HLHS) is a congenital heart defect. Congenital means present at birth. They occur early in pregnancy often before a mother even knows she is pregnant. These structural defects of the heart can vary in severity from simple problems to more serious problems.

HLHS describes a grouping of cardiac defects that involves the heart and it’s blood vessels. Most of the structures on the left side of the heart are small and underdeveloped. The degree of this underdevelopment can vary between children.

The parts of the heart that are usually affected by HLHS are:

- **Left ventricle** - It is one of two pumping chambers of the heart. After birth, it receives oxygen rich (red) blood from the left atrium and pumps it into the aorta. In babies with HLHS this is underdeveloped.
- **Aorta** – It is a big blood vessel that takes the blood to the body. In babies with HLHS the portion of this blood vessel that leaves the heart is smaller than normal.
- **Valves** - Mitral valve is the valve that controls blood flow between the left atrium and the left ventricle. Aortic valve is the valve that regulates blood flow from the heart into the aorta. In babies with HLHS these valves can be small, narrowed or closed.

In a healthy heart, the left ventricle is very muscular and strong so it can pump blood to the body. When this ventricle is small and weak, it is unable to pump the blood the body needs. HLHS is a severe life threatening illness.
What are the causes of HPHS?

For every 10,000 babies born, approximately 1-3 babies will have HLHS.

Some congenital heart defects may have a genetic link causing heart problems to occur more often in certain families. There is a chance that there are other heart defects and organ abnormalities. Some babies with HLHS have a chromosomal abnormality or other genetic problem.

But most of the time HLHS occurs by chance and it appears to be a random event that can happen to anyone.

What does this mean?

During pregnancy, HLHS is well tolerated by the baby as the baby does not need to use his/her lungs and the heart does not need to send blood to the lungs to pick up oxygen. The placenta and umbilical cord carries oxygen to the baby. However once the baby is born HLHS becomes a life threatening illness.

All babies born with HLHS need surgery or a heart transplant. The surgery aims to use one ventricle, the right ventricle, to do the work that is normally done by the right and left ventricle. Heart surgery for babies with HLHS occurs in 3 stages.

You will meet with a paediatric cardiovascular surgeon (specialist in heart surgery for children) who will be able to describe these surgeries in detail and outline the risks and expected outcomes. The surgeon will also be able to discuss heart transplantation.

What other tests should we consider?

Other tests include a detailed ultrasound that is used to assess the baby’s other organs. A fetal echocardiogram, which is a detailed ultrasound of your baby’s heart, will be done.

An amniocentesis may be offered. During an amniocentesis, a small amount of amniotic fluid is taken from the area around the baby and tested for chromosome problems.

What will happen around the time of the baby’s birth?

Babies with HLHS will need to be born at a hospital with a paediatric cardiovascular surgeon. Babies with HLHS are closely monitored in the intensive care unit and further testing after birth will help the paediatric cardiologist (heart specialist for children) plan your baby’s care.
This level of care may not be available in your hospital and you may need to deliver at another centre. Your doctor will be able to tell you where your baby needs to be delivered.

**What does this mean for my baby’s future?**

The paediatric cardiologist will be able to discuss with you in detail what this means for your baby’s future and how it may affect his/her quality of life.

**What do we do now?**

You will meet with a doctor that specializes in high-risk obstetrics. A referral to a paediatric cardiologist and a paediatric cardiovascular surgeon will be made. These doctors will discuss with you in detail your options for further testing, discuss with you test results and provide you with treatment options. They will be able to answer any questions you may have.

There is no right or wrong answer on what to do next. Your doctor will provide you with all available information and assist you in making an informed decision. We will support you in whatever decision you choose to make.

**Where can I get more information?**

Sick Kids Toronto  
Hypoplastic Left Heart Syndrome  
http://www.sickkids.ca/HLHS/

Children’s Hospital Boston  
Hypoplastic left heart syndrome (HLHS)  
http://www.childrenshospital.org/az/Site502/mainpageS502P0.html

The Children’s Hospital of Philadelphia  
Hypoplastic Left Heart Syndrome (HLHS)  
http://www.chop.edu/service/cardiac-center/heart-conditions/hypoplastic-left-heart-syndrome-hlhs.html