OPENMFM PATIENT EDUCATION

Hypoplastic Left Heart Syndrome

A patient-friendly guide for pregnancy and newborn planning

HLHS is a serious heart condition that starts before birth. Knowing about it early helps your team plan a safer delivery, immediate newborn care, and the next steps after birth.

Also called HLHS

What is HLHS? BIG PICTURE

In **hypoplastic left heart syndrome**, the **left side of the baby's heart is too small or not fully formed**.

- The left side of the heart normally pumps blood to the body.
- In HLHS, it cannot do that job well.
- The right side of the heart has to do extra work to send blood to both the lungs and the body.
- HLHS is a **critical congenital heart defect**, so special care is needed soon after birth.

What parts of the heart are affected?

- **Left ventricle:** the main pumping chamber on the left side
- **Mitral valve:** the valve that lets blood enter the left ventricle
- **Aortic valve:** the valve that lets blood leave the left ventricle
- **Aorta:** the large blood vessel that carries blood to the body

These structures may be very small, very tight, or fully closed. The exact pattern can differ from one baby to another.

How was this found?

- HLHS is often first suspected on a **prenatal ultrasound**.
- It is usually confirmed with a **fetal echocardiogram**, which is a detailed ultrasound of the baby's heart.
- Doctors look for:
    - a very small left ventricle
    - a larger right ventricle
    - a small aorta
    - little or no blood flow leaving the left side of the heart

Doctors also look closely at the **opening between the upper heart chambers**. If it is very small, a baby may become sicker more quickly after birth.

Did I cause this? REASSURANCE

Most of the time, parents did **nothing** to cause HLHS.

- The exact cause is often unknown.
- Sometimes HLHS is linked with gene or chromosome changes.
- Your team may discuss genetic counseling or genetic testing.

This is not your fault.

What happens during pregnancy?

- Your care team may include maternal-fetal medicine, pediatric cardiology, pediatric heart surgery, neonatology, and genetic counseling.
- You may need repeat fetal heart ultrasounds during pregnancy.
- These follow heart growth, blood flow, and whether the opening between the upper chambers stays wide enough.

Serial fetal echocardiograms help the team watch for changing findings and plan ahead.

Where should my baby be delivered?

- A baby with HLHS should usually be delivered at or very near a hospital with **pediatric heart surgery** and a **newborn intensive care unit**.
- In many stable pregnancies, delivery is planned near the due date, often around **39 to 40 weeks**.
- A cesarean delivery is **not usually needed just because of HLHS**.
- The delivery method is usually based on regular obstetric reasons.

The goal is to have the right people, medicines, and newborn support ready at birth.

What happens after birth?

- Many babies with HLHS need **prostaglandin** soon after birth.
- This medicine keeps an important blood vessel open so blood can still reach the body.
- The baby will have an **echocardiogram after birth** to confirm the heart anatomy.
- Some babies may need an urgent **heart catheter procedure** if the opening between the upper chambers is too small.

Prenatal diagnosis helps because it gives the team time to prepare for the first minutes and hours after delivery.

Will my baby need surgery?

HLHS usually requires a **series of heart surgeries**. These surgeries do not make the heart normal, but they help the right side of the heart pump blood to the body.

Norwood procedure

Usually in the first week of life

The first major surgery after birth.

Glenn procedure

Often around 4 to 6 months

Reduces some of the workload on the heart.

Fontan procedure

Often around 2 to 4 years

Completes the usual staged pathway.

What is the long-term outlook?

- HLHS is a serious, lifelong heart condition.
- Children who have surgery need regular follow-up with a heart doctor.
- Some children may have challenges with growth, feeding, learning, exercise tolerance, heart rhythm, or heart function.

Every baby is different. Your pediatric cardiology and heart surgery team can give the most accurate guidance for your baby's specific heart findings.

Questions to ask your care team

- What type of HLHS does my baby have?
- Is the opening between the upper heart chambers wide enough?
- Are there any other heart or body findings?
- Should we consider genetic testing?
- Where should I deliver?
- What will happen in the first hour after birth?
- When would the first surgery be expected?
- What support is available for our family?

Key message

HLHS is a serious heart condition, but finding it before birth gives your team time to plan the safest delivery and newborn care.

- The goal is to have the right doctors ready.
- The goal is to have the right medicine ready.
- The goal is to have the right testing and heart surgery team ready.
- Early planning helps families move from shock to preparation.

Sources: [CDC](https://www.cdc.gov/heart-defects/about/hypoplastic-left-heart-syndrome.html), [OpenMFM fetal echo teaching deck](https://openmfm.org/decks/hlhs-sonography/hlhs_sonography.html), and [Children's Hospital of Philadelphia](https://www.chop.edu/conditions-diseases/hypoplastic-left-heart-syndrome-hlhs).