CONDITION

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome is a birth condition in which the heart's left side cannot pump enough blood to the body.

WHAT IT IS
Severe congenital underdevelopment of the heart's left side

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Blue-gray color, fast breathing, poor feeding, unusual sleepiness

WHAT CAUSES IT
Usually unknown; genetic and environmental factors may contribute

HOW IT IS DIAGNOSED
Fetal or newborn echocardiogram

MAIN TREATMENT
Staged heart surgery or transplant, plus lifelong specialist care

CAN IT BE CURED?
No - surgery does not normalize circulation

IS IT AN EMERGENCY?
Yes - newborn circulation can collapse without treatment

LONG-TERM OUTLOOK
Improved survival, with lifelong risks and specialist follow-up

Overview

Hypoplastic left heart syndrome, or HLHS, is a serious heart condition present at birth. The structures on the left side of the heart are too small or poorly formed to pump blood effectively to the body.

The affected structures can include the left ventricle, which is the main pumping chamber for the body, and the mitral and aortic valves. The first part of the aorta, the body's largest artery, is also often very narrow.

Before birth, the placenta supplies oxygen and temporary fetal blood vessels allow blood to bypass parts of the heart and lungs. A vessel called the ductus arteriosus helps blood reach the body. After birth, this vessel normally begins to close. In a baby with HLHS, closure can sharply reduce blood flow to the organs and cause the circulation to fail.

HLHS therefore needs specialist care immediately after birth. Treatment does not make the heart anatomically normal. Instead, surgery rearranges blood flow so the right side of the heart can support the body. Heart transplantation is another option for some children.

Symptoms

Babies with HLHS may appear stable immediately after birth while the ductus arteriosus remains open. Symptoms often develop as it starts to close, usually during the first hours or days of life.

Possible signs include:

A blue or gray color is called cyanosis. It happens when the blood reaching the body contains less oxygen than usual.

HLHS is sometimes found before symptoms develop, either during pregnancy or through newborn heart screening. Screening does not identify every case, so symptoms still need urgent assessment even if screening results were normal.

- Blue, gray, or unusually pale skin, lips, or tongue
- Fast, difficult, or labored breathing
- Poor feeding or tiring during feeds
- Unusual sleepiness, weakness, or difficulty waking
- Cool hands and feet
- A weak pulse or poor circulation
- Less urine than expected

Causes and risk factors

HLHS develops while the baby's heart is forming early in pregnancy. In most cases, the exact reason is unknown. It is not usually caused by anything a parent did or did not do.

Changes in genes involved in heart development may contribute. HLHS can occasionally occur with a chromosome condition or another genetic syndrome, but many babies have no identified genetic condition.

The chance may be higher when a close family member has HLHS or another congenital heart condition. "Congenital" means present at birth. Even with a family history, it is usually not possible to predict with certainty whether another pregnancy will be affected.

Parents may be offered genetic counseling and testing. A genetics professional can review the baby's other features, test results, and family history. Testing may find an explanation, but a normal result does not rule out every genetic contribution.

Diagnosis

HLHS may be suspected during a routine pregnancy ultrasound. A fetal echocardiogram can then examine the baby's heart in detail. This is a specialized ultrasound that shows the heart's chambers, valves, major blood vessels, and blood flow.

Finding HLHS before birth allows delivery and immediate care to be planned at a hospital with newborn heart specialists and pediatric cardiac surgery.

After birth, an echocardiogram usually confirms the diagnosis. This painless ultrasound shows how small the left-sided structures are and whether blood can move between the heart's chambers and major vessels.

Other assessments may include:

During cardiac catheterization, a thin tube is passed through a blood vessel to the heart. It may be used for diagnosis or to perform a procedure.

- Pulse oximetry to measure oxygen levels
- An electrocardiogram to record the heart's electrical activity
- Chest imaging and blood tests
- Genetic testing when appropriate
- Cardiac catheterization in selected cases to measure pressures or clarify anatomy

Treatment and management

A newborn with HLHS is cared for in a specialist cardiac center. Medicine called prostaglandin is usually given through a vein to keep the ductus arteriosus open until a longer-term plan is in place. The baby may also need support with breathing, circulation, feeding, and body temperature.

If blood cannot move freely between the upper chambers of the heart, an urgent catheter procedure or operation may be needed to enlarge that opening.

For many children, treatment involves three planned operations. Timing and surgical techniques are tailored to the baby's anatomy and health.

The first operation, commonly called the Norwood procedure, is usually performed in the newborn period. It creates a new route for the right ventricle to pump blood to the body and provides a separate path for blood to reach the lungs. Some centers use a combined surgical and catheter-based approach called a hybrid procedure.

The second operation, often called the Glenn procedure, is usually performed several months later. It connects a major vein from the upper body directly to the lung arteries. This reduces the workload on the right ventricle.

The third operation, the Fontan procedure, is often performed in early childhood. It directs blood returning from the lower body toward the lungs without first passing through a pumping chamber.

These operations are called staged palliation. "Palliation" means they improve circulation without correcting the heart's original structure. After the Fontan procedure, blood returns to the lungs mainly through pressure in the veins, while the right ventricle pumps oxygen-rich blood to the body.

Heart transplantation may be considered instead of staged surgery or later if the circulation or heart muscle is failing. Transplantation is limited by donor availability and requires lifelong medicines that reduce immune rejection.

Some families may also consider care focused on comfort rather than surgery. The specialist team should explain all appropriate options, expected burdens, and uncertainties without pressure.

Complications

HLHS and its treatment can affect many parts of the body. Risks are different at each stage and do not occur in every child.

Possible early complications include poor blood flow to the brain, kidneys, or other organs; bleeding; infection; blood clots; abnormal heart rhythms; or problems with a surgical connection. The right ventricle may weaken, or the tricuspid valve may leak.

The period between the first and second operations can be especially vulnerable. Many heart centers use home-monitoring programs that may include tracking weight, feeding, oxygen levels, and specific symptoms. Families receive their own instructions about which changes require an immediate call.

Long-term concerns after Fontan surgery can include:

Regular assessment helps identify these problems before they cause major symptoms.

- Reduced pumping function or heart failure
- Abnormal heart rhythms
- Blood clots
- Liver scarring related to raised venous pressure
- Fluid around the lungs or in the abdomen
- Loss of protein through the bowel
- Abnormal material forming in the airways
- Slower growth, feeding difficulties, or reduced exercise capacity
- Learning, movement, attention, or emotional difficulties

Prevention

HLHS usually cannot be prevented. Its exact cause is often unknown, and it begins early in fetal development.

If one pregnancy has been affected, genetic counseling can help explain what is known about the chance in a future pregnancy. A fetal echocardiogram may be offered in a later pregnancy to look closely at the developing heart. This does not prevent HLHS, but it can support earlier diagnosis and delivery planning.

Living with it and what to expect

HLHS is a lifelong condition, even after all planned operations. Survival has improved with modern surgery and intensive care, but outcomes remain different from one child to another. The heart's pumping ability, valve function, other medical conditions, and complications all influence the course.

During infancy, much of family life may center on feeding, medicines, monitoring, appointments, and recovery from surgery. Some babies need high-calorie nutrition, feeding-tube support, or help from feeding specialists.

Over the following years, many children attend school, form friendships, and take part in family and community life. Some have limits on endurance or need breaks during physical activity. Regular movement is often valuable, but suitable activities should be based on the child's circulation, heart rhythm, and specialist advice.

Developmental checks are important because children with HLHS have a higher risk of

difficulties with movement, speech, learning, attention, or behavior. Early physical, occupational, speech, educational, or psychological support can help when a need is identified.

Follow-up continues throughout life with a congenital heart team. Monitoring may include heart imaging, rhythm tests, exercise assessment, blood tests, and checks of liver and kidney health. Adults need care from specialists in adult congenital heart conditions rather than routine cardiology alone.

Pregnancy can place major strain on a Fontan circulation. Anyone considering pregnancy needs assessment before conception from an adult congenital heart specialist and a high-risk pregnancy team.

Some people eventually need further catheter procedures, surgery, or a transplant. Others remain stable for long periods. Lifelong follow-up is needed even when someone feels well.

When to get medical help

HLHS in an undiagnosed or untreated newborn is an emergency. Call emergency services if a baby:

For a child already receiving HLHS care, follow the cardiac team's emergency plan. Seek urgent help for a clear change from the child's usual color, breathing, oxygen level, alertness, feeding, urine output, or circulation. Do not delay emergency care while waiting for a routine appointment.

Contact the cardiac team promptly for less sudden changes such as worsening feeding, repeated vomiting, slower weight gain, new swelling, increasing tiredness, palpitations, reduced activity, or fever around the time of a procedure.

Routine follow-up remains important when the child appears well. Scheduled visits are used to monitor heart function, surgical pathways, growth, development, rhythm, liver health, and medicines.

- Becomes blue, gray, very pale, limp, or difficult to wake
- Has severe breathing difficulty, pauses in breathing, or grunting
- Cannot feed because of breathlessness or weakness
- Has cold, mottled skin with a weak response
- Collapses or becomes unresponsive

Common questions

Can a baby survive hypoplastic left heart syndrome?

Yes. Staged surgery and intensive care have allowed many children to survive into adulthood. HLHS remains a serious condition, and some babies do not survive surgery or later complications. The individual outlook is shaped by heart anatomy, other health conditions, surgical results, and how the circulation functions over time.

Is HLHS curable?

No current treatment makes the left side of the heart develop normally. Staged operations reroute blood flow, while transplantation replaces the heart. Both approaches require lifelong specialist care.

How many operations does a child with HLHS need?

Many children have three main operations: the Norwood, Glenn, and Fontan procedures. Additional catheter procedures or operations may be needed if a connection narrows, a valve leaks, the heart rhythm changes, or heart function declines.

Can HLHS be detected before birth?

It often can. A pregnancy ultrasound may raise concern, and a fetal echocardiogram can confirm the heart anatomy. Some cases are first recognized after birth.

Is hypoplastic left heart syndrome inherited?

HLHS is not usually inherited in a simple, predictable pattern. Genetic factors may contribute, and congenital heart conditions can occur more often in some families. Genetic counseling can provide information based on the child's findings and family history.

Can a child with HLHS have a normal life?

Many children attend school and take part in everyday activities, but their circulation is not normal. They may have reduced stamina, developmental needs, repeated procedures, or later complications. Ongoing congenital heart care helps activities, education, and support evolve with the child's needs.

Sources

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