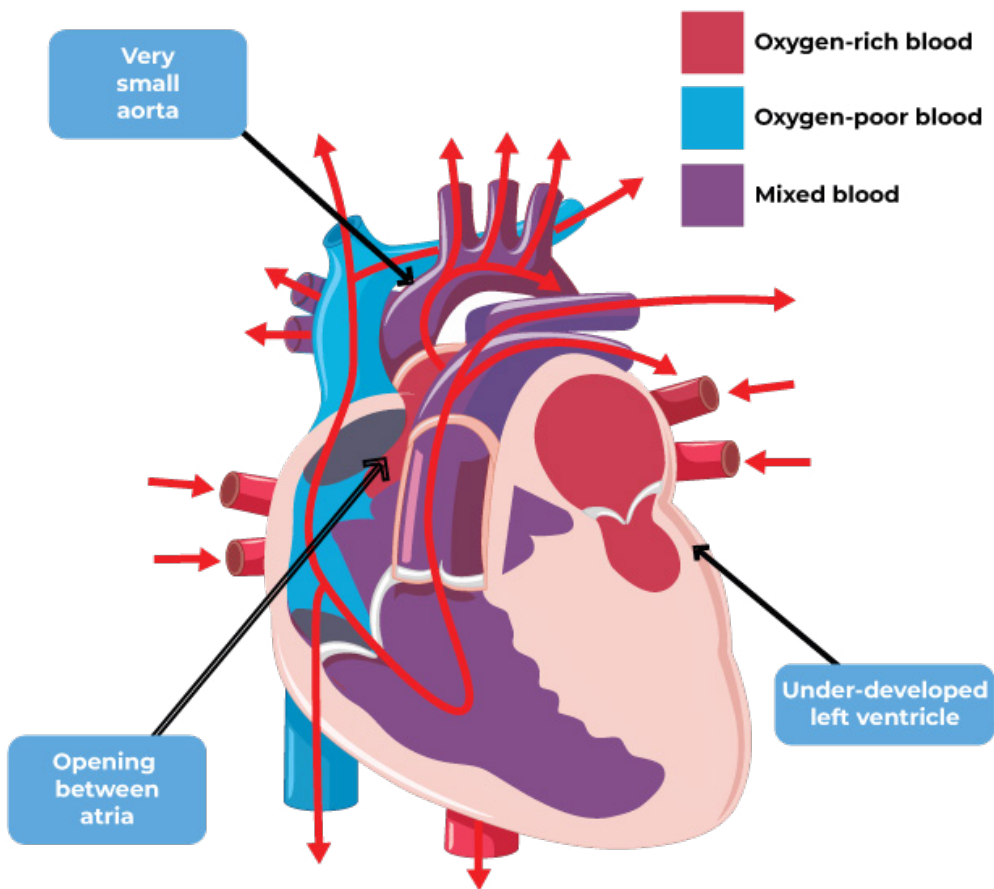


UNDERSTANDING HYPOPLASTIC LEFT HEART SYNDROME





WHAT IS HYPOPLASTIC LEFT HEART SYNDROME?

Hypoplastic Left Heart Syndrome (HLHS) is a birth defect that affects the normal blood flow through the heart. This is caused during pregnancy when the left side of the heart doesn't develop properly, meaning it is a congenital birth defect (CHD).

HOW HLHS EFFECTS THE LEFT SIDE OF THE HEART

The left ventricle is too small and underdeveloped

The mitral valve is not formed or very small

The aortic valve is not formed or very small

The mitral valve is not formed or very small

Often babies who are born with HLHS will also have an atrial septal defect, which is a hole that separates the two upper chambers of the heart.

CAN CHILDREN LIVE A NORMAL LIFE WITH HYPOPLASTIC LEFT HEART SYNDROME?

Hypoplastic Left Heart Syndrome is a sliding scale with some affected only marginally and others profoundly. It is a non life compatible condition at birth and each child must have surgery to make corrections; many children will have lifelong complications and may need follow up visits with a cardiologist.

HYPOPLASTIC LEFT HEART SYNDROME OCCURANCE IN CHILDREN

Approximately, HLHS occurs in **1 in 5,000 children** in the UK and Ireland.

It is estimated to be **200 diagnoses** made in the UK and Ireland **per year**.



WHAT CAUSES HYPOPLASTIC LEFT HEART SYNDROME?

Like many congenital septal defects, the cause of Hypoplastic Left Heart Syndrome is unknown.

Some babies may suffer from HLHS because of changes in their genes or chromosomes.

Environmental factors during pregnancy are sometimes thought to increase the risk of HLHS in babies, especially with mothers who have taken medication or undergone medical procedures.

Genetics are also thought to be a potential cause too.

HOW IS HYPOPLASTIC LEFT HEART SYNDROME DIAGNOSED IN CHILDREN?

DURING PREGNANCY

During pregnancy, prenatal tests are used to check birth defects and other conditions.

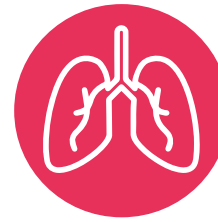
Hypoplastic Left Heart Syndrome can be diagnosed during pregnancy with an ultrasound scan and if it is suspected, an **echocardiogram** can be used to confirm the diagnosis.



AFTER BIRTH

After the baby is born, symptoms of Hypoplastic Left Heart Syndrome may not be present immediately, but some signs can quickly develop after the first few hours or days after birth.

SIGNS TO LOOK OUT FOR AFTER BIRTH



Breathing problems



Pounding heart



Bluish skin colour



Weak pulse

During a physical examination, a doctor might hear a heart murmur, which is an abnormal whooshing sound caused by blood not flowing properly. If a murmur is heard or other signs are present, your child's doctor may request more tests. An echocardiogram is the most common type of test that will be used.

Hypoplastic Left Heart Syndrome can also be detected with newborn pulse oximetry screening, which is a simple test used to determine the amount of oxygen in the baby's blood. Low levels of oxygen can be a sign of HLHS.

HYPOPLASTIC LEFT HEART SYNDROME SYMPTOMS

Difficulty breathing

Difficulty feeding

Lack of energy

Rapid heartbeat but weak pulse

Sweaty, clammy or cool skin

Greyish or bluish skin

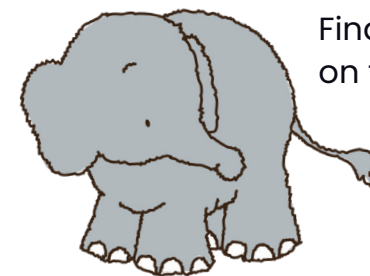
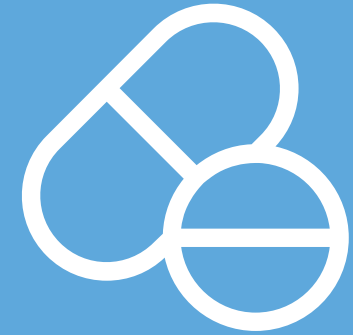
Discoloured nails and lips

Newborns don't always show symptoms of HLHS immediately, but they will usually develop within a few hours or days after birth.

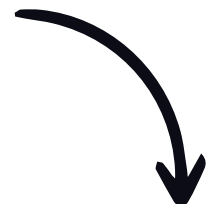
TREATMENTS FOR HYPOPLASTIC LEFT HEART SYNDROME

MEDICINE

Some babies may need medicine to help strengthen the heart muscle, lower the blood pressure and help to get rid of the extra fluid.



Find out about surgery on the next page



HYPOPLASTIC LEFT HEART SYNDROME SURGERY

1

NORWOOD PROCEDURE

This surgery is usually done in the first 2 weeks of a baby's life. Surgeons create a 'new' aorta and connect it to the right ventricle. A tube is also placed from either the aorta or the right ventricle to the vessels to supply the lungs with oxygen. After the surgery, the baby's skin may still look discoloured because oxygen-rich and oxygen-poor blood is still in the heart.

BIDIRECTIONAL GLENN SHUNT OPERATION

At 4 - 6 months of age, the next procedure is complete. This procedure creates a connection between the pulmonary artery and the vessel returning oxygen-poor blood to the heart. This reduces the work the right ventricle has to do by allowing blood returning from the body to flow directly to the lungs.

2

FONTAN PROCEDURE

This procedure is conducted during the period when a baby is aged 18 months to 3 years of age. Doctors will connect the pulmonary artery and the vessel returning oxygen-poor blood to the heart, allowing the rest of the blood coming back from the body to go to the lungs. Once this surgery is complete, the oxygen-rich blood will no longer mix with the oxygen-poor blood in the heart.

3

Sometimes your child's surgeon may recommend a heart transplant instead of the 3-part surgery. However, babies who have a heart transplant will need lifelong medications.



AS YOUR CHILD GROWS UP

As your child grows up, they will need lifelong follow up visits at least once a year with a cardiologist.

These appointments will ensure that their heart, lungs and other vital organs continue to function as they should be.

When your child becomes an adult, they will need to move to seeing specialists in adult congenital heart disease.

Most children who have had surgery for Hypoplastic Left Heart Syndrome will need some form of heart medication for the rest of their life. They will also likely need antibiotics before any other surgeries to help prevent the risk of a heart infection.

CONDITIONS ASSOCIATED WITH HYPOPLASTIC LEFT HEART SYNDROME

A small proportion of babies may have other abnormalities, including those relating to chromosome irregularities such as Turner Syndrome and Jacobsen Syndrome. Your child's cardiologist will discuss this further with you if necessary.

MEDICAL TERMS & WHAT THEY MEAN

AORTA The main artery of the heart that supplies oxygen-rich blood to the body.

ATRIA Either of the two upper chambers in the heart.

ATRIUM one of the two upper chambers of the heart.

CONGENITAL Present from birth.

ECHOCARDIOGRAM A scan to look at the heart and nearby blood vessels.

HEART MURMUR A whoosing or rasping sound heard during a heartbeat.

VENTRICLE One of the two lower chambers of the heart.



Respite and support of any kind is invaluable.

Lagan's Foundation, a Charitable Incorporated Organisation in England and Wales (1154208) aims to physically help support families who have infants and children up to 19 years old, diagnosed with complex health conditions specialising in heart defects and feeding difficulties. Lagan's Foundation's purpose is to provide unique at home and in hospital care to support parents in their caring role.

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