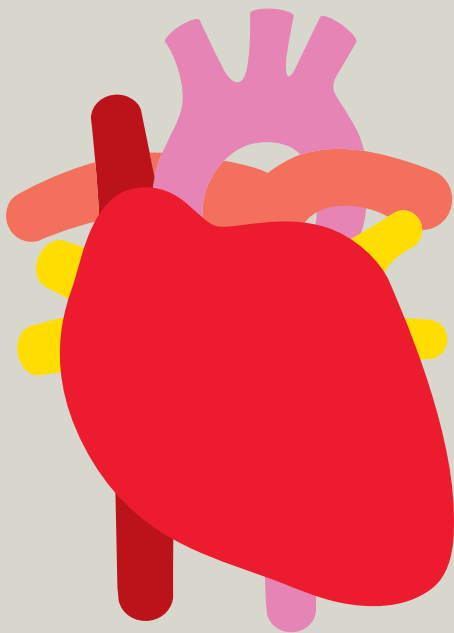


Understanding your child's heart

# Hypoplastic left heart syndrome

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**FIGHT  
FOR EVERY  
HEARTBEAT**

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## About this book

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If you're reading this book, you've probably just had some very upsetting news and have lots of questions running through your mind.

We've written this book to help answer some of those questions. We'll go through:

- what hypoplastic left heart syndrome is and how it is diagnosed
- how hypoplastic left heart syndrome is treated
- the benefits and risks of treatments
- what happens as your child grows up
- where to go for more support

This booklet shouldn't replace the advice your doctors or nurses may give you, but it should help you to understand what they tell you.

## Contents

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What is a congenital heart condition?	2
What causes a congenital heart condition?	3
What is hypoplastic left heart syndrome?	6
What are the symptoms of hypoplastic left heart?	8
What other conditions are associated with hypoplastic left heart?	9
How is hypoplastic left heart diagnosed?	10
How is hypoplastic left heart treated?	11
The Norwood procedure	12
The hybrid procedure	15
Will my child need further surgery?	18
What happens as my child grows up?	24
What is the risk of having another child with a congenital heart condition?	25
Coping with everyday life	26
The medical terms and what they mean	27
References & acknowledgements	29

## What is a congenital heart condition?

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It's an abnormality of the heart that developed in the womb. Sometimes, a **congenital** heart condition is diagnosed when the baby is still developing in the womb, but most times it's not discovered until after the baby is born. There are lots of different types of **congenital** heart conditions.

Each day, 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research, visit [bhf.org.uk/research](https://bhf.org.uk/research)

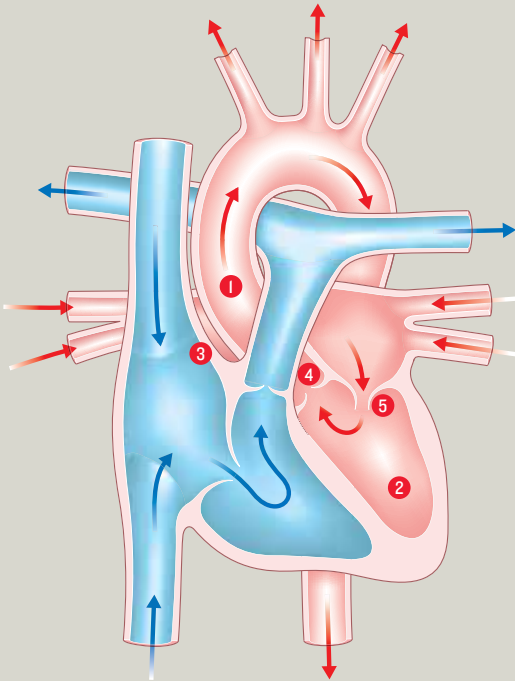
## What causes a congenital heart condition?

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In most cases, it's caused by something going wrong during the very early stages of the pregnancy.

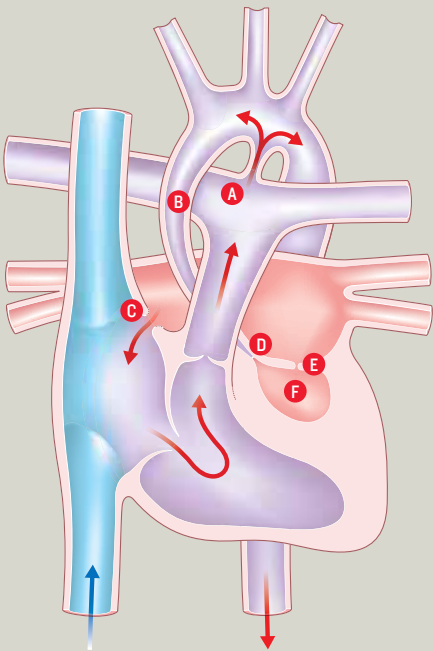
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome means a group of symptoms that commonly appear together as part of a condition).*

# Normal heart



- |                  |                |
|------------------|----------------|
| 1 Aorta          | 4 Aortic valve |
| 2 Left ventricle | 5 Mitral valve |
| 3 Atrial septum  |                |

# Hypoplastic left heart syndrome



- |                        |                                  |
|------------------------|----------------------------------|
| A The 'duct'           | D Aortic valve is narrow/blocked |
| B Small aorta          | E Mitral valve is narrow/blocked |
| C Atrial septal defect | F Small left ventricle           |

# What is hypoplastic left heart syndrome?

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Hypoplastic left heart syndrome (HLHS) is a very serious type of **congenital** heart condition. The main abnormalities are:

- The mitral valve and/or the aortic valve are either very narrow or completely blocked.
- The main pumping chamber on the left side of the heart (*the left ventricle*) is very under-developed.
- The **aorta** is small and under-developed.
- There is a hole between the top chambers of the heart (*This is called atrial septal defect or ASD*).

In the normal heart, oxygen-rich blood flows from the lungs to the left atrium, and then into the left ventricle. From there it is pumped into the aorta and around the body. In babies with HLHS, the blood cannot flow normally through the left side of

the heart. Instead, it passes from the left atrium to the right atrium through the ASD and into the right ventricle. From there it is pumped to the pulmonary artery and back to the lungs. (*See the illustration on page 5*).

Before babies are born, the two main arteries – the **pulmonary** artery and the **aorta** – are connected by a short channel called the **ductus arteriosus** (*'the duct'*). During pregnancy the **duct** allows blood to bypass the lungs because the foetus is not breathing but is instead receiving oxygen from the placenta. In a normal heart the **duct** closes shortly after birth because it is no longer required. However, in babies with HLHS, the only way blood can flow to the **aorta** and the body is through the **duct**. So, if the **duct** closes, blood will not be circulated around the body and the baby will die. Please see page 11 for more information about prostaglandin treatment.

## What are the symptoms of hypoplastic left heart?

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Most babies with HLHS look normal immediately after birth, but they become breathless and very ill if they are not treated quickly.



For more information and support about growing up with a heart condition, visit [bhf.org.uk/heart-health/children-and-young-people](https://bhf.org.uk/heart-health/children-and-young-people)

## What other conditions are associated with hypoplastic left heart?

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Children with HLHS only have the heart abnormalities described on page 6 and 7. However, a small proportion of babies may have other abnormalities, including chromosome abnormalities. Your child's **cardiologist** will discuss this further with you if necessary. Children with HLHS also have a greater risk of having learning disabilities than other children.

## How is hypoplastic left heart diagnosed?

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In most cases, HLHS is detected before birth, but sometimes it is not diagnosed until after the baby is born. Usually, the only test that is needed to make the diagnosis is an **echocardiogram**. This is an ultrasound scan of the heart. It is very similar to the scans that are carried out during pregnancy and it won't hurt your baby at all.

## How is hypoplastic left heart treated?

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It's not possible to correct HLHS with surgery, but there are operations that can help your child to have a better quality of life. Your child will need to have several operations during their lifetime. The first stage of treatment is to keep the **duct** open by giving your baby an injection of drugs called prostaglandins. These drugs can occasionally interfere with your baby's natural breathing, and in some rare cases, they may need the support of a **ventilator**.

Often, the initial surgery is carried out within a few days of birth. The first operation for most babies with HLHS is called the Norwood procedure. However, some babies who are very small or very ill may not be suitable for this and may be offered the hybrid procedure. We describe both of these procedures in the next section.

# The Norwood procedure

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The Norwood procedure is an open-heart operation. Your baby will be given a general anaesthetic, and the function of the heart is taken over by a heart-lung machine. The aim of the operation is to use the right ventricle to pump blood into the **aorta**. To do this, the surgeon separates the main pulmonary artery from the right and left branches and joins it to the upper part of the **aorta**. (See the illustration on page 14). The blood supply to the right and left pulmonary arteries is then provided by a 'shunt' – a tube, made of synthetic material, which is placed between the **aorta** and the pulmonary artery or between the right ventricle and the pulmonary artery.

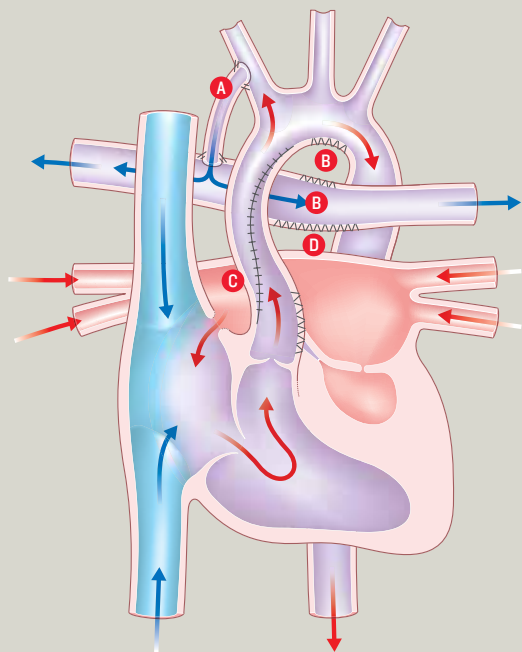
After the surgery, your baby will have a vertical scar in the centre of the chest, along the breast bone. Most babies are allowed home within a few weeks. After the operation, you will need to take your baby to the outpatients department to see the **cardiologist** for regular check-ups.

## What are the risks of the Norwood procedure?

Most babies survive this first stage of surgery, but there is a small risk that your baby may have brain damage or internal bleeding. Kidney damage can also occur, but this usually recovers with treatment. Your cardiologist will discuss these risks with you in more detail.



## The Norwood procedure



- A** Shunt
- B** The 'duct' is closed
- C** New 'aorta'
- D** Pulmonary artery is disconnected from the heart

## The hybrid procedure

This is a technique that involves a combination of surgery and catheter treatment.

The hybrid procedure helps to improve your baby's circulation while allowing them to grow before they have more major surgery. It also helps to protect the lungs by reducing the amount of blood being delivered to them and it keeps the **duct** open.

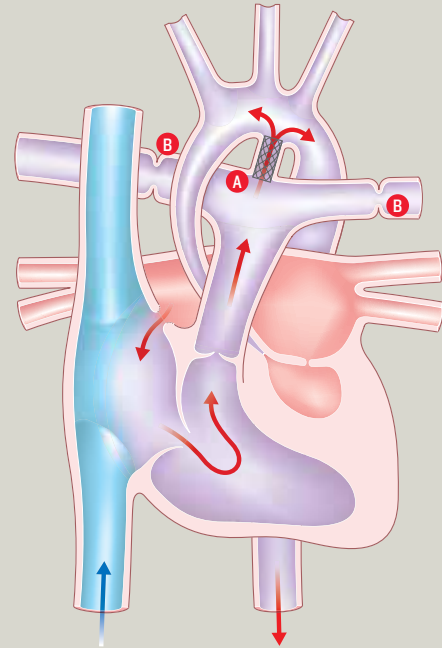
Your baby will be given a general anaesthetic. The surgeon will place an expandable metal mesh tube – called a **stent** – inside the **duct**, to stop it from closing. (See the illustration on page 17). This means that blood can flow into the **aorta** to be delivered to the rest of the body.

Small bands are also placed around the right and left pulmonary arteries, to reduce the amount of blood flowing to the lungs. This helps to protect the lungs. After the surgery, your baby will have a scar down the centre of their chest.

## The hybrid procedure

## What are the risks of the hybrid procedure?

This is a relatively new procedure and the technique is still being evaluated. Your child's **cardiologist** will discuss this with you in more detail.



**A Stent in duct**  
**B Band**

## Will my child need further surgery?

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The exact nature and timing of any further surgery will depend on your child's progress. The type of surgery most commonly carried out is a *cavopulmonary shunt (Glenn Shunt)*, which involves connecting the superior vena cava directly to the **pulmonary** arteries. Further surgery after this usually involves redirecting the blood flow from the inferior vena cava to the **pulmonary** artery. This is called a *total cavopulmonary connection*, or *TCPC* for short.

### Cavopulmonary shunt, also called a Glenn shunt

This procedure is used to increase the blood flow to the lungs, and also to reduce the workload of the heart. A cavopulmonary shunt does not correct the underlying heart abnormality.

Your child will be given a general anaesthetic. The heart will be stopped and the function of the heart will be taken over by a **heart-lung machine**. The surgeon will connect the superior vena cava directly

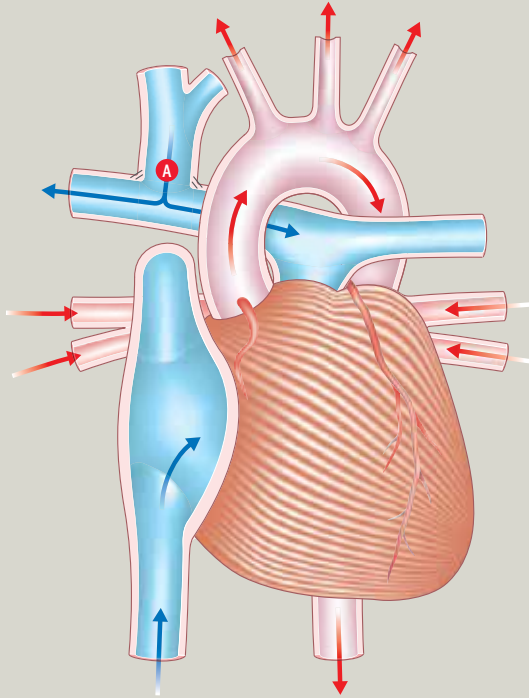
to one of the arteries that takes blood to the lungs (the right **pulmonary** artery). (See the illustration on page 20). After the surgery, your child will have a scar in the middle of the chest, along the breastbone.

Your child will need to stay in hospital for a few days after the surgery, and will need to visit the outpatients department within a few weeks for a check-up with the **cardiologist**. After that, your child will need regular check-ups.

### What are the risks of a cavopulmonary shunt operation?

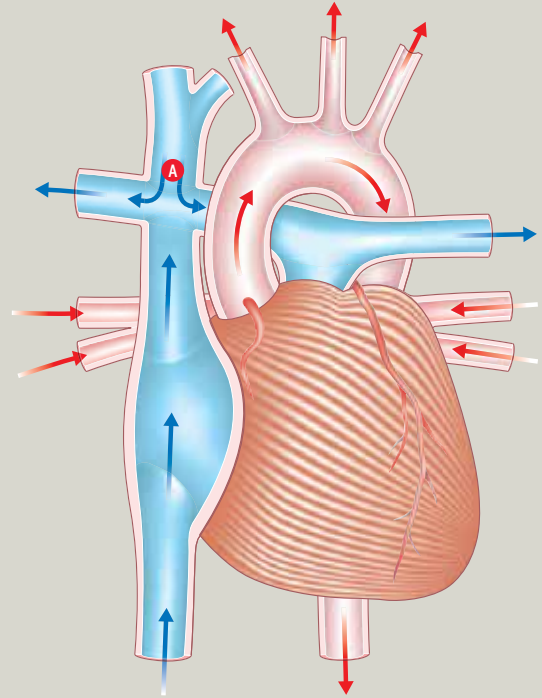
Most children survive this surgery, but they may become more blue and short of breath on exertion as they grow. The risk of death and other complications – such as narrowing where the superior vena cava has been joined to the **pulmonary** artery, brain damage, stroke or internal bleeding – varies based on the exact type of heart condition your child has. Other possible complications include pleural effusion (*fluid around the lungs*) and kidney damage. Your **paediatric cardiologist** or cardiac surgeon will discuss your child's individual risk with you before surgery.

## Cavopulmonary shunt



**A** Superior vena cava connected to pulmonary artery

## Total cavopulmonary connection



**A** Blood flow from both inferior and superior vena cava has been redirected to the right pulmonary artery

## Fontan-type operation

The purpose of this operation is to improve the amount of oxygen in the blood and in most cases to improve exercise capacity. This is achieved by connecting both the inferior and superior vena cava to the **pulmonary** artery.

Many modifications to the original Fontan operation technique have been developed, including: a *modified Fontan*, a *fenestrated Fontan*, and *total cavopulmonary connection (or TCPC for short)*. Any type of Fontan operation is a major operation. Your child's cardiac surgeon will explain exactly which operation your child needs.

Your child will be given a general anaesthetic. The heart will be stopped and the heart's function will be taken over by a **heart-lung machine**. The surgeon will redirect the flow of blood from the inferior vena cava to the **pulmonary** artery. In most cases, the superior vena cava has already been connected. (See *diagram on page 21*). The illustration on page 21 shows the TCPC type of Fontan operation. After surgery, your child will have a scar in the middle of

the chest, along the breastbone. A Fontan-type operation will not make your child's heart normal, but – if the operation is successful – it should allow an adequate blood supply to the lungs to allow your child to grow.

## What are the risks of the Fontan-type operation?

Most children survive this surgery. The risk of death and major complications – such as brain damage – varies depending on the exact type of heart condition your child has. Other possible complications include pleural effusion (*fluid around the lungs*), pericardial effusion (*fluid around the heart*), and kidney damage. Some children can develop heart rhythm disturbances which need to be treated with medicines, or less commonly with a **pacemaker**.

The length of time your child will need to stay in hospital will vary, depending on how well he or she recovers after surgery. There is an increased risk of developing a blood clot after the surgery, so most children will need to take either warfarin or aspirin to help prevent this. See our factsheet on warfarin at **bhf.org.uk**

## What happens as my child grows up?

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HLHS is a complex condition, and some children will not survive all three stages of surgery. Although surgery can give a better quality of life, it is not possible to correct the heart abnormality and it's uncertain how long children with this condition will live for. The longest survivors at present are in their late 20s. Heart transplantation may be an option for some patients, although this is rarely considered before adulthood.

Children with HLHS are always limited to some extent in their physical activities. Physical endurance may get worse in teenage years or young adult life, but specific restrictions on exercise are usually not necessary. Your child's **cardiologist** will tell you if there are any specific forms of exercise or activities they should avoid.

## What is the risk of having another child with a congenital heart condition?

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If you have one child with a **congenital** heart condition, there is about a 1 in 40 chance of having another child with **congenital** heart disease<sup>1</sup>. However, this risk may be higher (*or lower*) depending on the type of **congenital** heart disease your child has. Because your risk of having another child with **congenital** heart disease is higher than it is for other people, you may be offered a special scan at an early stage in future pregnancies, to look at the baby's heart.

Ask your midwife or GP for more information on having a scan earlier than usual. If you have more than one child with **congenital** heart disease, the specific condition may not always be the same.

## Coping with everyday life

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For information on the topics below, please visit [bhf.org.uk/heart-health/conditions/congenital-heart-disease](https://bhf.org.uk/heart-health/conditions/congenital-heart-disease)

- Financial issues
- Low-income benefit
- Disability benefits
- Carer's Allowance
- Fares for visiting your child in hospital

## The medical terms and what they mean

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**Aorta** The main artery of the heart. It supplies oxygen-rich blood to the body.

**Atrial septal defect** A hole between the top two chambers of the heart.

**Atrium** One of the two upper chambers of the heart.

**Cardiologist** A consultant specialising in heart disease.

**Congenital** From birth.

**Duct** See ductus arteriosus below.

**Ductus Arteriosus** A natural connection between the aorta and the pulmonary artery. Also called the 'duct'.

**Echocardiogram** An ultrasound scan used to produce pictures of the heart and blood vessels.

**Heart-lung machine** A machine that pumps blood around the body while the heart is stopped during an operation.

## The medical terms and what they mean

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**Pacemaker** A small electrical device fitted in the chest or abdomen. It's used to treat some abnormal heart rhythms (arrhythmias) that can cause your heart to either beat too slowly or miss beats.

**Paediatric** To do with paediatrics – the study of children's diseases.

**Pulmonary** To do with the lungs.

**Ventilator** A machine that helps your child breathe.

**Ventricle** One of the two lower chambers of the heart.

## References

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1. Gill HR, Splitt M, Sharland K, Simpson JM. 2003. *Patterns of recurrence of congenital heart disease: An analysis of 6,640 consecutive pregnancies evaluated by detailed fetal echocardiography.* Journal of the American College of Cardiology; 42: 923-9.

## Acknowledgements

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## Notes

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For more information and support about children and young people with a heart condition, visit [bhf.org.uk/heart-health/children-and-young-people](https://bhf.org.uk/heart-health/children-and-young-people)



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For over 50 years we've pioneered research that's transformed the lives of millions of people living with cardiovascular disease. Our work has been central to the discoveries of vital treatments that are changing the fight against heart disease.

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