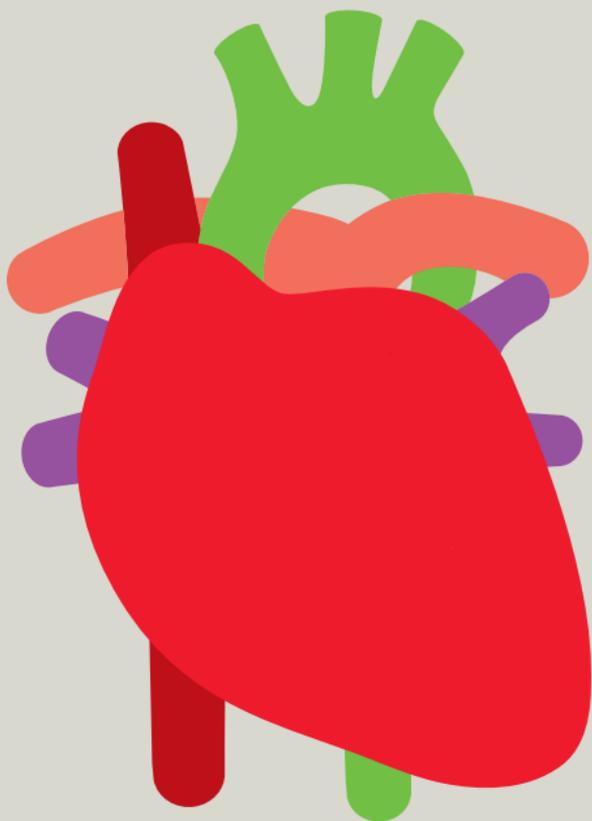




Understanding your child's heart

Tetralogy of Fallot

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**FIGHT
FOR EVERY
HEARTBEAT**

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About this book

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 tetralogy of fallot is and how it is diagnosed
- how tetralogy of fallot is treated
- the benefits and risks of treatments
- what happens as your child grows up
- where to go for more support.

Please be aware that this booklet shouldn't replace the advice your doctors or nurses may give you. But it should help make what they tell you that little bit clearer.

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What is a congenital heart condition?

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.

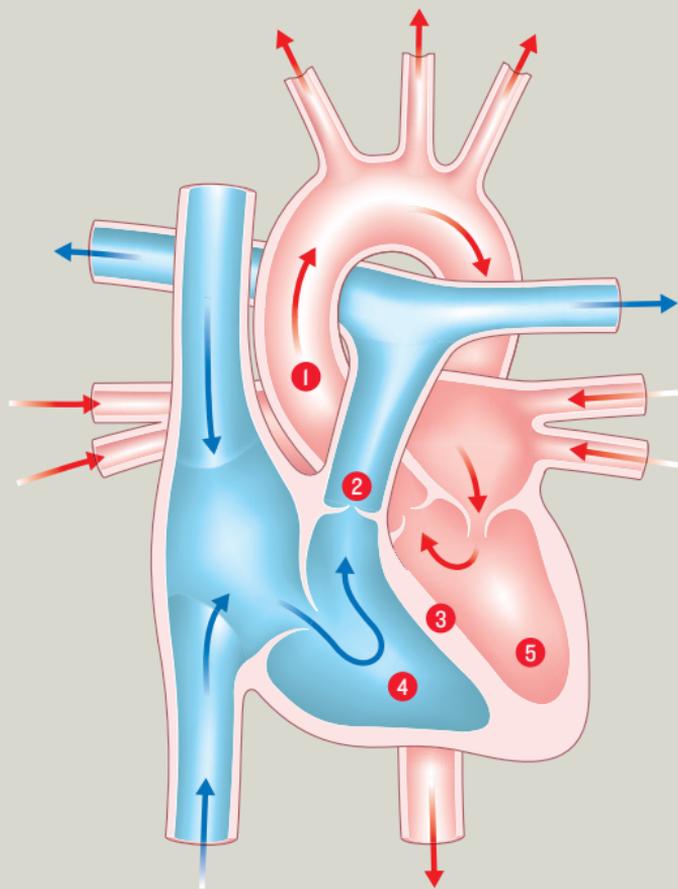
Each day 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research visit [bhf.org.uk/research](https://www.bhf.org.uk/research)

What causes a congenital heart condition?

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

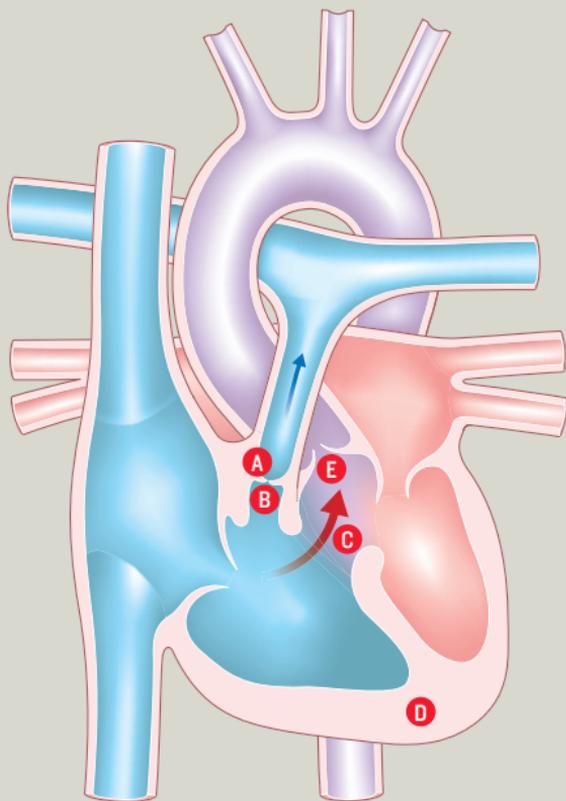
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome is a group of symptoms that commonly appear together as part of a condition).*

Normal heart



- | | |
|----------------------|-------------------|
| 1 Aorta | 4 Right ventricle |
| 2 Pulmonary valve | 5 Left ventricle |
| 3 Ventricular septum | |

Tetralogy of Fallot



- A** Narrowed pulmonary valve
- B** Narrowing beneath pulmonary valve
- C** Ventricular septal defect
- D** Thickened muscle
- E** Overriding aorta

What is tetralogy of fallot?

There are four main problem abnormalities with tetralogy of fallot. (See the illustration on page 5).

- The **pulmonary** valve is narrow (*pulmonary stenosis*) and the muscle below it is thickened.
- There is a large hole – called a **ventricular septal defector VSD** – between the two main pumping chambers of the heart (*the right and left ventricles*).
- The **aorta** sits over the left and right **ventricles** instead of just the left **ventricle** (*overriding aorta*).
- The right **ventricle** is enlarged and more muscular (*right ventricular hypertrophy*).

In tetralogy of fallot, oxygen-rich and oxygen-poor blood mix because of the **VSD**. This means that the blood being pumped around your child's body doesn't contain as much oxygen as it should, and they can appear blue.

Your baby will need to have an operation to correct the problem. This usually takes place when they're 4 to 6 months old, but the timing varies from one baby to another. We explain more about the surgery later on.

What are the symptoms of tetralogy of fallot?

The low level of oxygen in your baby's blood can make them appear blue, particularly on the lips and tongue, inside the mouth, and on the hands. How blue your baby looks depends on how severe the **pulmonary stenosis** is. Some babies appear pink and just become slightly blue when they cry, while others may appear blue all the time.

Some babies may have **hypercyanotic attacks** where they suddenly become very blue, or very pale or floppy, or faint. You should tell the **paediatric cardiologist** immediately, as these attacks can be very serious and can be life-threatening. It is usually possible to control these attacks with medication, but often they are a sign that it is time to plan surgery.

What other conditions are associated with tetralogy of fallot?

Some babies might have a syndrome called '**22q11 deletion**'. Others have syndromes such as Down's Syndrome. If your baby is diagnosed with tetralogy of fallot, your doctor will talk you through the option of having a test to find out if he or she also have a **chromosomal** abnormality. If the condition is diagnosed before birth, this test can be carried out before your baby is born.

How is tetralogy of fallot diagnosed?

In most cases, tetralogy of fallot is diagnosed after the baby is born using an **echocardiogram**, but in some cases it can be detected before birth. An echocardiogram is an ultrasound scan of the heart, and it won't hurt your baby at all.



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

How is tetralogy of fallot treated?

Most babies can go home as normal soon after birth, as they don't need any immediate treatment. However, your baby will need major surgery later on, usually when they are between 4 and 6 months old.

It's important to be aware that without this surgery, most children with tetralogy of fallot would sadly will not live to adulthood.

Surgery

If the **pulmonary** artery is small, your baby may first need to have a shunt operation to improve the blood supply to the lungs, and later on have a major operation to repair the heart.

In most cases the pulmonary artery has formed well, so your baby will probably have a single major repair operation under general anaesthetic. The heart is stopped and the function of the heart is taken over by a 'heart-lung machine', which makes sure that blood is still pumped around your baby's body. The surgeon will close the hole between the two pumping chambers of the heart (the **VSD**) by sewing a patch over it.

The narrow **pulmonary** valve will also be widened and any narrowing in the **pulmonary** artery can be treated too. After the operation, your baby will have a scar in the centre of their chest along the breast

bone. Although this operation is often called a 'repair operation' or 'corrective surgery', it will never make the heart completely normal.

A shunt operation usually involves placing a small tube between the pulmonary artery and the artery that feeds blood to the right arm. Very occasionally a **valvuloplasty** may be done instead of a shunt operation. This is when a **catheter** (*a thin, hollow tube*) with a small balloon at its tip, is used to stretch the narrowed **pulmonary** valve open.

What are the risks of surgery?

The good news is, most children will survive surgery and have a very good quality of life. However, all major heart operations are very serious and carry a small risk of fatality, or of major complications such as brain damage, kidney

damage or lung complications such as pneumonia. The cardiac surgeon will explain the risks before you give your consent for the surgery.

The risk associated with the shunt operation is usually low, but varies from one child to another. There is a small risk that the heart's electrical system may be damaged during the operation. If this happens, your baby's heart rate will be slower than normal and they might need to have another procedure to implant a pacemaker to make the heart beat faster.

Other more common but less serious complications include a fast heart rate in the first few days after surgery, fluid collecting around the heart, or a wound infection.

What happens after surgery?

Most babies need to stay in hospital for about a week after the major repair operation. Some babies may need to stay in for longer if there are complications. Your baby may be prescribed some medicines to take after getting home from hospital, but they will probably only need to take these for a short while.

What happens as my child grows up?

Most children with repaired tetralogy of fallot will lead normal, active lives after their operation. Your **cardiologist** will tell you if your child should avoid any specific forms of exercise.

Anyone who has had a tetralogy repair operation will have an abnormal **pulmonary** valve which does not close effectively. This means that, after the right ventricle has pumped blood into the **pulmonary** artery, some of this blood leaks back into the right **ventricle**. This is called **pulmonary** regurgitation, and the right **ventricle** has to work harder than usual, sometimes causing it to get enlarged. This rarely needs treatment early on, but further surgery will most probably be needed in teenage or adult life to replace the valve.

A donated tissue valve (from a cow, pig or human) is used to replace the **pulmonary** valve. A tissue valve will not last forever, and will need to be

replaced approximately every 10-15 years. This is most commonly done by open heart surgery, but some people may be suitable for keyhole surgery.

Even many years after apparently successful surgery, your child may develop a new problem – such as an abnormal heart rhythm. An abnormal heart rhythm is often very treatable, but it can be serious, and in very rare cases can even be fatal. It's important that your child has regular outpatient reviews with a specialist, even if they appear well.

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

If you have one child with a **congenital** heart condition, there is around a 1 in 40 chance that if you have another child, they will have a heart condition too.¹ However, this risk may be higher (*or lower*) depending on the type of **congenital** heart condition your child has. Because your risk of having another child with congenital heart condition is higher than it is for other people, your doctor may offer you a special scan at an early stage in future pregnancies, to look at the baby's heart.

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

Coping with everyday life

For information on the topics below, please visit [bhf.org.uk/congenital](https://www.bhf.org.uk/congenital)

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

The medical terms and what they mean

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

Cardiac To do with the heart.

Cardiologist A consultant specialising in heart disease.

Catheter A fine, hollow tube.

Chromosomes Found in the nucleus of every cell in the body, chromosomes contain the genes, or hereditary elements, which establish the characteristics of an individual.

Congenital From birth.

ECG See electrocardiogram.

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

Electrocardiogram A recording of the electrical activity of the heart. Also called an ECG.

Hypercyanotic attack When someone suddenly becomes very blue, or very pale or floppy, or faint.

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

Pulmonary To do with the lungs.

Pulmonary stenosis When the pulmonary valve is very narrow.

Stenosis See pulmonary stenosis.

Valvuloplasty A procedure to stretch open a narrowed valve.

Valvotomy A surgical procedure to open a narrowed valve.

Ventricle One of the two lower chambers of the heart.

Ventricular Septal Defect A hole between the two ventricles of the heart. Also called VSD.

VSD See ventricular septal defect.

References

1. Gill HR, Splitt M, Sharland GK, 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.

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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://www.bhf.org.uk/heart-health/children-and-young-people)



**British Heart
Foundation**

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.

But cardiovascular disease still kills around one in four people in the UK, stealing them away from their families and loved ones.

From babies born with life threatening heart problems, to the many mums, dads and grandparents who survive a heart attack and endure the daily battles of heart failure.

Join our fight for every heartbeat in the UK. Every pound raised, minute of your time and donation to our shops will help make a difference to people's lives.

Text FIGHT to 70080 to donate £3

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