



Understanding your child's heart

Tetralogy of Fallot



Contents

About this booklet	5
The normal heart	6
What is congenital heart disease?	8
What causes congenital heart disease?	8
Why me? Why my child?	9
What is Tetralogy of Fallot?	10
What are the symptoms of Tetralogy of Fallot?	12
What other conditions are associated with Tetralogy of Fallot?	13
How is Tetralogy of Fallot diagnosed?	13
How is Tetralogy of Fallot treated?	14
What happens as my child grows up?	17
The specialist centre for congenital heart disease	18
Coping with everyday life	20
General advice for the future	23
What is the risk of having another child with congenital heart disease?	24
Support groups	25
About the British Heart Foundation	26
References	27
Contacts	28
Hospital visits	29
The medical terms and what they mean	30
Index	32
Have your say	35



About this booklet

This booklet is written for the parents of babies with Tetralogy of Fallot, and for their relatives and friends. It explains:

- what Tetralogy of Fallot is and how it is diagnosed
- how Tetralogy of Fallot is treated
- the benefits and risks of treatments
- how to cope as a parent of a baby with Tetralogy of Fallot
- where to go for more support.

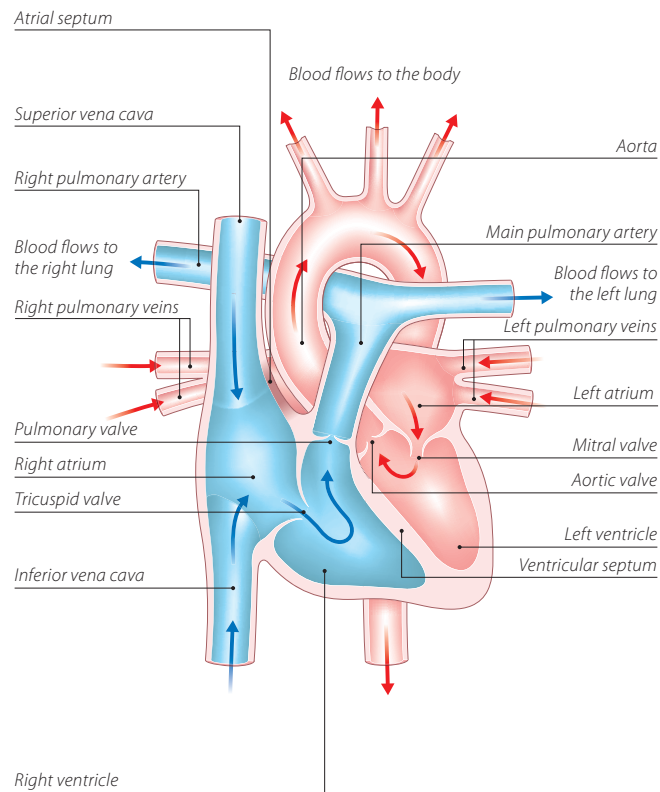
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

The normal heart

The heart is a muscular pump which pumps blood through the body and lungs. There are four chambers in the heart. The two upper ones are called the right atrium and left atrium. These are separated by a wall called the atrial septum. The two lower chambers are called the right and left ventricles, and are separated by a wall called the ventricular septum. See the illustration opposite.

On each side of the heart, blood passes from the atrium, through a heart valve – the tricuspid valve on the right, and the mitral valve on the left – into the ventricle. The ventricles are the main pumping chambers of the heart. Each ventricle pumps blood out into an artery. The right ventricle pumps blood – blue in the illustration – into the pulmonary artery (the blood vessel that takes blood to the lungs). The left ventricle pumps blood – red in the illustration – into the aorta (the blood vessel that takes blood to the rest of the body). Blood flows from the right side of the heart, through the pulmonary valve into the pulmonary artery, and then to the lungs where it picks up oxygen. The oxygen-rich blood flows back into the left side of the heart through the pulmonary veins. The left ventricle then pumps the oxygen-rich blood out of the heart through the aortic valve and into the aorta, and all around the body. The blood then returns to the right side of the heart through two main veins – one from the upper body (superior vena cava), and the other from the lower body (inferior vena cava).

The normal heart



What is congenital heart disease?

Congenital heart disease is an abnormality of the heart that developed in the womb. In some cases, congenital heart disease is diagnosed when the baby is still developing in the womb, but in most cases the problem is not discovered until after the baby is born. There are lots of different types of congenital heart disease. *Tetralogy of Fallot* is one type.

What causes congenital heart disease?

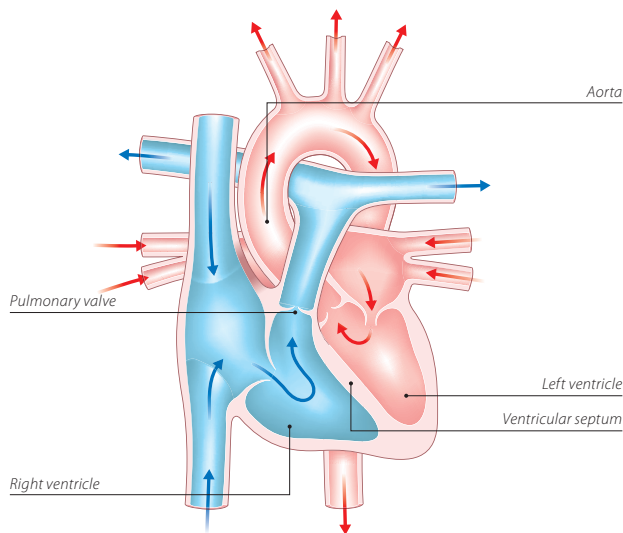
We know that, in most cases of congenital heart disease, something has gone wrong in the early development of the fetus, at the very early stages of the pregnancy. In most cases, we don't understand why the baby's heart did not develop normally. In some cases, congenital heart disease can be part of a syndrome that the baby is born with. (A syndrome means a group of symptoms that appear together.)

Why me? Why my child?

It is not unusual for parents of children with congenital heart disease to blame themselves or to be angry. Anger, disappointment, fear and guilt are all normal feelings to have when you are told that there is something wrong with your child. At first it may be difficult to cope with and it can take a while for the news to sink in. Many pregnant women or mothers, and their partners, ask themselves what they did during their pregnancy that could have caused their baby's heart to develop with heart disease. But the reality is that it can happen to anyone. In fact congenital heart disease happens in about 7 in every 1,000 pregnancies.¹ For more than half of these children, the heart disease is only a minor problem which either doesn't need any treatment, or which can be successfully corrected with surgery.

What is Tetralogy of Fallot?

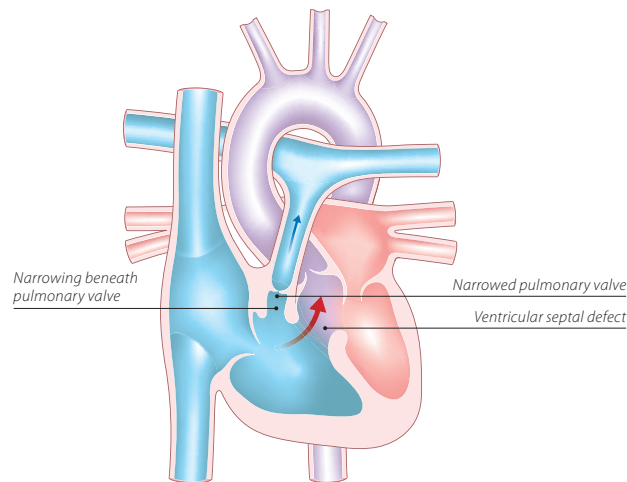
The normal heart



Tetralogy of Fallot is a serious heart abnormality. There are two main problems. (See the illustration on the next page.)

- The pulmonary valve is narrow (**pulmonary stenosis**) and the muscle below it is thickened.
- There is a large hole – called a **ventricular septal defect** or **VSD** – between the two main pumping chambers of the heart (the right and left ventricles).

Tetralogy of Fallot



In the normal circulation, blood passes through the lungs to collect oxygen (as described on page 6). In babies with Tetralogy of Fallot, the narrowing in the pulmonary valve and the thickened muscle below it mean that less blood can flow through to the lungs. This means that the level of oxygen in the blood is low.

Your baby will need to have an operation to correct the problem. This usually takes place when the baby is 6 to 12 months old, but the timing varies from one baby to another. We explain more about the surgery, and the benefits and risks involved, on page 14.

What are the symptoms of Tetralogy of Fallot?

The low level of oxygen in your baby's blood can make him or her 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 with Tetralogy of Fallot may have attacks where they suddenly become very blue, or very pale or floppy, or faint. These are known as *hypercyanotic attacks*, and are sometimes called *spells*. If your baby has attacks like this, you should tell the paediatric cardiologist immediately, because the attacks can be very serious and may even be life-threatening. It is usually possible to control these attacks with medication, but they can often mean that it is time to plan surgery.

What other conditions are associated with Tetralogy of Fallot?

Most babies with Tetralogy of Fallot don't have any other abnormality, but some do. Some have a syndrome called '22q11 deletion'. Others have syndromes such as Down's Syndrome. Your paediatric cardiologist will talk to you about these. For more on 22q11 deletion, see our website bhf.org.uk. And see page 25 for where to get more information on Down's Syndrome.

If your baby is diagnosed with Tetralogy of Fallot, your doctor will discuss with you the option of having a test to find out if he or she has a chromosomal abnormality. If the Tetralogy of Fallot 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, but in some cases it can be detected before birth. 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. It doesn't hurt your baby at all.

How is Tetralogy of Fallot treated?

Most babies can go home as normal soon after birth, as they don't need any special treatment immediately. However, your baby will need major surgery later on, usually when he or she is between 6 and 12 months old. Without this surgery, most children with Tetralogy of Fallot would die before adulthood.

Surgery

The type and timing of treatment depends very much on how blue your baby becomes, and on how well the pulmonary artery grows. If the pulmonary artery doesn't grow well, 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 (see below). 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 **valvoplasty** may be done instead of a shunt operation. This is when a catheter (a fine, hollow tube) with a small balloon at its tip, is used to stretch the narrowed pulmonary valve open. You will need to take your baby for outpatient visits after any of the treatments described above.

If the artery has grown well, your baby will probably have a single **major repair operation**. Your baby will be given a 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.

During the operation, 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. If there are also narrowings in the pulmonary artery, these can be treated at the same time, by sewing a patch into the wall of the artery.

After the operation, your baby will have a scar in the middle of the chest along the breast bone.

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

What are the risks of surgery?

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 death, or of major complications such as brain damage, kidney damage or lung complications such as pneumonia. It is important to understand the risks associated with your baby's operation. The cardiac surgeon will explain to you the risks for your baby 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. Your surgeon will discuss this with you.

Ninety-eight in every 100 children survive the *major repair operation*.² The risk of getting brain damage as a result of this operation is small – probably around 1 in 100.² There is also 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 he or she might need to have another operation 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 doctor may prescribe some medicines for your baby to take after getting home from hospital, but he or she will probably only need to take these for a short while. After the operation, you will need to take your baby to the outpatients clinic.

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*. This means that the right ventricle has to work harder than usual, sometimes causing it to get enlarged as the years go by. This rarely needs treatment early on, but further surgery may be needed in teenage or adult life to replace the valve.

Even many years after apparently successful surgery, your child may develop a new problem – such as an abnormal heart rhythm – which may need treatment. An abnormal heart rhythm can usually be treated, but it can be serious, and in very rare cases can even be fatal. So, it's important that your child has regular outpatient reviews with a specialist, even if he or she appears well.

The specialist centre for congenital heart disease

Your child will continue to have check-ups at a specialist centre for congenital heart disease throughout their life. Up to the age of about 16, he or she will go to a centre which may be called a 'specialist centre for children with congenital heart disease', or a 'specialist paediatric centre'. (Paediatric means to do with children.)

If the specialist centre is quite a distance from your home, it may be possible to make arrangements for your child to be looked after under a 'shared care' system. This means that your child will be looked after locally, but will go to the specialist centre for specialised treatment. (See page 22 for information about claiming travel expenses for visits to the specialist centre.)

The specialist team

At the specialist centre, a large multidisciplinary team of people will be involved in caring for your child and your family's needs. (Multidisciplinary means that it includes several different types of health professionals.) The team usually consists of:

- a consultant paediatric cardiologist
- a specialist registrar (a doctor who is specifically training in children's heart problems)
- a cardiac nurse specialist or cardiac liaison nurse
- a consultant paediatric surgeon or paediatric cardiothoracic surgeon
- cardiac technicians

- a paediatric physiotherapist
- a paediatric dietitian
- ward-based paediatric nurses
- a social worker
- an occupational therapist
- a speech and language therapist, and
- a play specialist.

All of these people are involved in planning the care for each patient. You probably won't need to see all of them, but it is important to know who is there and available to help you with any problems you may come across. For example, the social worker can be a very useful source of information on what benefits you might be entitled to claim, and whether you can claim your travel expenses for visiting your child in hospital, or for visits to the specialist centre. (For more on this, see page 22.)

The specialist centre will also have access to psychology services which you can use to help your child or your family if you need help at difficult times.

Specialist adult congenital heart disease centres

When your child grows into adulthood, it is important that he or she carries on going to a specialist centre for check-ups. There are several specialist centres in the UK for adults with congenital heart disease. Your child's care will be transferred to an adult specialist centre usually at around the age of 16. These centres usually have a multidisciplinary team with the same mix of professionals as in the children's specialist centre (see

page 18). When your child is nearing adulthood, your specialist paediatric centre will start preparing you and your child for the move to the adult specialist centre, to make the transition as smooth as possible.

Coping with everyday life

Having a child with a heart condition in hospital can be very difficult for a number of reasons. You are having to care for your child in hospital, and may be anxious about the treatment your child has to have or how well he or she is recovering. You might also be worrying about being away from home, or about your other children and who's looking after them, or about your work or your finances.

Hospital staff recognise the stress that you and your family might be under. Ask the nurse who is looking after your child about the support services available within the hospital – such as psychology services, welfare rights advisers and social workers. Below we give a brief guide to the benefits and help you may be able to get.

Financial issues

Financial problems may arise because you need to stay in hospital with your child. The hospital may be a long way from home, and you may not know how long you will need to stay there for. This can affect your finances, as you may have to take time off work and will have extra costs such as travel expenses and buying meals while in hospital. This can be an extra anxiety, and difficult to cope with.

If you are worried about your finances, it is important to discuss your situation with a hospital social worker or cardiac liaison nurse, or with the Citizens Advice Bureau. They may be able to advise you on the benefits you can claim. Also, an organisation called Working Families can give you advice on the phone – on 0800 013 0313 – about your rights as an employee if you need to take time off work to be with your child.

Low-income benefits

Benefits for people on a low income include Income Support, housing and council tax benefits, and Tax Credits. If your income goes below a certain amount, you may be able to claim benefits. However, you have to meet certain criteria in order to get these benefits. The criteria vary from one benefit to another, so you should get specialist advice from a hospital social worker, Citizens Advice Bureau or Jobcentre Plus.

Disability benefits

Some children with congenital heart disease will qualify for a **Disability Living Allowance (DLA)**, but most will not. Ask the specialist nurse or social worker for advice. If your child needs extra care because of his or her condition, you may be able to apply for this benefit. There is a mobility and a care component to the benefit. It can be difficult to get Disability Living Allowance for a child. You will need to show that your child needs more attention or supervision than other children of the same age. To get a claim form for Disability Living Allowance, call 0800 88 22 00. Or you can get a form from your GP or your local Jobcentre Plus office, or claim online at www.direct.gov.uk

Carer's Allowance

You may be entitled to *Carer's Allowance* if your child is awarded the middle or higher care component of Disability Living Allowance. However, you will only get Carer's Allowance if your earnings are below a certain limit. To get a claim form for the Carer's Allowance, call 01253 856 123. Or you can get a claim form from your local Jobcentre Plus, or claim online at www.direct.gov.uk

Fares to hospital

If you get Income Support, or have a Tax Credit exemption card, or are assessed as being on a low income, you are entitled to get reimbursed for your fares to hospital appointments and inpatient treatment. If you think you might qualify because of your income, complete a form HC1. To get one, call 0845 850 1166. The forms are usually also available from Jobcentre Plus offices and NHS hospitals.

Fares for visiting your child in hospital

If you are getting Income Support, income-based Jobseeker's Allowance, or Pension Credit, you may be able to get help towards the cost of fares for hospital visiting, from the Social Fund. You should apply for a Community Care Grant on form SF300, which is available from your local Jobcentre Plus office or from the website of the Department for Work and Pensions at www.dwp.gov.uk. However, these payments are not given in all cases.

The benefits system is very complex, so it is important to get specialist advice on what you may be entitled to, from the hospital social worker or Citizens Advice Bureau.

General advice for the future

Endocarditis

Everyone who has Tetralogy of Fallot is at risk of getting **infective endocarditis**, both before and after surgery or treatment. Infective endocarditis is a rare condition where the inner lining of the heart, most commonly one of the heart valves, becomes infected.

Infective endocarditis is a serious condition which can be life-threatening if it's not treated quickly. Nowadays, if it is diagnosed early, most people with it will recover well with antibiotic treatment, although some damage may occur to the heart valves as a result of the infection.

Endocarditis is caused by a bacterium, or (rarely) another type of infective organism that is in the bloodstream, which settles onto the abnormal structure or defect in the heart. Although it is not possible to prevent all bacteria from getting into the bloodstream, there are some things your child can do, as he or she grows up, to reduce the risk of getting endocarditis:

- Maintain good oral hygiene and have regular check-ups with a dentist
- Avoid body piercing and tattooing
- Never inject recreational drugs.

If your child develops flu-like symptoms with a temperature **which persist for over a week**, you should visit your GP as your child may need a blood test. Make sure that the GP knows that your child is at increased risk of getting endocarditis. You can

do this by showing the GP an *Endocarditis warning card*. You can get this card from the British Heart Foundation (BHF) by calling either the Heart HelpLine on **0300 330 3311** (local rate number) or the BHF Orderline on **0870 600 6566**.

Pregnancy

If you have a daughter with congenital heart disease, you need to be aware that pregnancy could carry risks to both the mother and the baby. So, when your daughter gets older, it's particularly important that she avoids having an unplanned pregnancy. You will need to discuss this with your daughter in whichever way you, as a parent, think is appropriate for her. If your daughter wants to have a baby, it's best that she speaks to her cardiologist about it first, so that the pregnancy can be planned for when your daughter's heart condition is most stable.

People who have congenital heart disease themselves have an increased risk of having a child with a heart problem. This applies to both males and females. You can discuss this with your cardiologist. Early scans in pregnancy can be arranged to look for heart disease in the baby.

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

If you have one child with congenital heart disease, there is about a 1 in 50 chance of having another child with congenital heart disease.³ 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 had two children with congenital heart disease, the risk of having another child with heart disease rises to about a 1 in 10 chance. This may sound like a high risk, but you still have a much better chance of the baby's heart being normal than abnormal. If there is a recurrence, the heart disease may not always be of the same type.³

Support groups

The following support groups may be able to offer you further information, advice and support:

Action for Sick Children

36 Jacksons Edge Road, Disley, Stockport SK12 2JL
Phone: 0800 074 4519. Website: www.actionforsickchildren.org

Children's Heart Federation

Level One, 2-4 Great Eastern Street, London EC2A 3NW
Phone: 0808 808 5000. Website: www.childrens-heart-fed.org.uk

Down's Syndrome Association

Langdon Down Centre, 2a Langdon Park, Teddington TW11 9PS
Phone: 0845 230 0372. Website: www.downs-syndrome.org.uk

About the British Heart Foundation

The British Heart Foundation is the nation's heart charity, saving lives through pioneering research, patient care and vital information.

What you can do for us

We rely on donations to continue our vital work. If you would like to make a donation to the BHF, please ring our Supporter Services team on **0844 847 2787** or contact us through our website at **bhf.org.uk/donate** or send it to us at the address on the back cover.

For more information

British Heart Foundation website

bhf.org.uk

For up-to-date information on heart disease, the BHF and its services.

Heart HelpLine

0300 330 3311 (local rate number)

For information and support on anything heart-related.

Booklets

To order any of our booklets

- call the BHF Orderline on **0870 600 6566**
- email **orderline@bhf.org.uk** or
- visit **bhf.org.uk/publications**

You can also download many of our publications from our website.

For information on other BHF booklets, and on DVDs and videos, ask for a copy of the *Heart health catalogue*.

Understanding your child's heart series

This booklet is one of the booklets in the *Understanding your child's heart* series. For a full list of the booklets available in this series, see our website **bhf.org.uk** or call the Heart HelpLine on **0300 330 3311** (local rate number).

Operation Fix-it

A short story book about eight-year-old Tom's experience in hospital for a heart operation. Prepares children for their hospital visit in an interesting and sometimes humorous way.

References

1. Petersen S, Peto V, Rayner M. 2003. *Congenital Heart Disease Statistics 2003*. Oxford: British Heart Foundation Health Promotion Research Group.
2. Specific procedures national data. The congenital heart disease website. London: The Information Centre. Accessed from: www.ccad.org.uk
3. 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.

Contacts

Use this page to keep contact details of the health professionals who are caring for your child.

Paediatric cardiologist

Paediatric nurse

Paediatrician

Social worker

Surgeon

Others

Hospital visits

Use this page to write down the dates of your hospital visits.

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The medical terms and what they mean

aorta	The main artery of the heart. It supplies oxygen-rich blood to the body.
atrial	To do with the atrium. (See below.)
atrium	One of the two upper chambers of the heart.
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 <i>electrocardiogram</i> .
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.
endocarditis	Infection of the lining of the heart or its valves.
genetic	To do with the information that is passed from parents to children through genes in sperm and eggs.

hypercyanotic attack	When someone suddenly becomes very blue, or very pale or floppy, or faint.
murmur	An extra sound that is sometimes heard when listening to the heart through a stethoscope.
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.
septum	The wall that keeps the right and left sides of the heart separate.
stenosis	See <i>pulmonary stenosis</i> .
valvoplasty	A procedure to stretch open a narrowed valve.
ventricle	One of the two lower chambers of the heart.
ventricular	To do with the ventricle or ventricles. (See above.)
ventricular septal defect	A hole between the two ventricles of the heart. Also called VSD.
VSD	See <i>ventricular septal defect</i> .

Index

22q11 deletion	13
aorta	6, 7
aortic valve	6, 7
atrium	6, 7
benefits	21
blueness	12
Carer's Allowance	22
causes of congenital heart disease	8
centre:	
specialist centre for adults	19
specialist centre for children	18
chromosome abnormality	13
complications of surgery	15
diagnosis	13
Disability Living Allowance	21
Down's Syndrome	13, 25
echocardiogram	13
endocarditis	23
fares to hospital	22
finance	20
future	17, 23
heart: how the normal heart works	6
hypercyanotic attacks	12
infection	16, 23
mitral valve	6, 7
money	20
normal heart	6
operation	14
pacemaker	16
pregnancy	24
professionals	18

pulmonary artery	6, 7, 14
pulmonary stenosis	10
pulmonary valve	6, 7, 17
pulmonary veins	6, 7
regurgitation	17
repair operation	14, 16
rhythm: heart rhythm	17
risks of surgery	15
septum	6, 7
shunt operation	14, 16
specialist centre:	
for adults	19
for children	18
specialist team	18
spells	12
stenosis	10
support groups	25
surgery:	
major repair surgery	14, 16
risks of surgery	15
shunt operation	14
symptoms	12
team	18
travel expenses	22
treatment	14
tricuspid valve	6, 7
valves	6, 7
valvoplasty	14
ventricle	6, 7, 17
ventricular septal defect	10, 15
VSD	10, 15



Have your say

We would welcome your comments to help us produce the best information for you. Why not let us know what you think? Contact us through our website at [bhf.org.uk/contact](https://www.bhf.org.uk/contact) Or, write to us at the address on the back cover.

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- Dr John Gibbs, Consultant Paediatric Cardiologist, Leeds General Infirmary.

HEART HELPLINE

For information and support on anything heart-related



0300 330 3311

local rate number



bhf.org.uk

Phone lines open 9am to 6pm Monday to Friday



British Heart Foundation

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