



# Understanding your child's heart

## Transposition of the great arteries



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## About this booklet

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This booklet is written for the parents of babies and children with transposition of the great arteries, and for their relatives and friends. It explains:

- what transposition of the great arteries is and how it is diagnosed
- how transposition of the great arteries is treated
- the benefits and risks of treatments
- how to cope as a parent of a child with transposition of the great arteries
- where to go for more support.

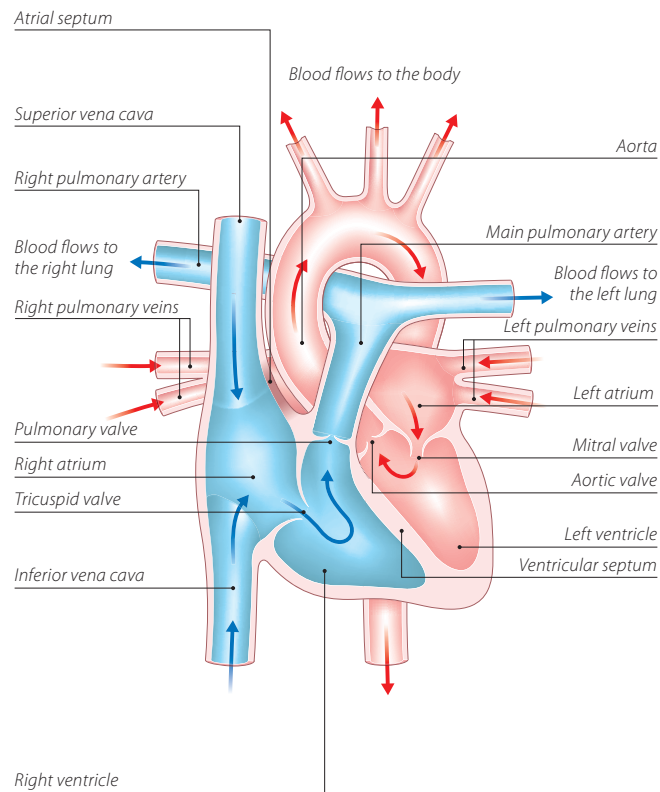
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

## 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?

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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. *Transposition of the great arteries* is one type.

## What causes congenital heart disease?

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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.)

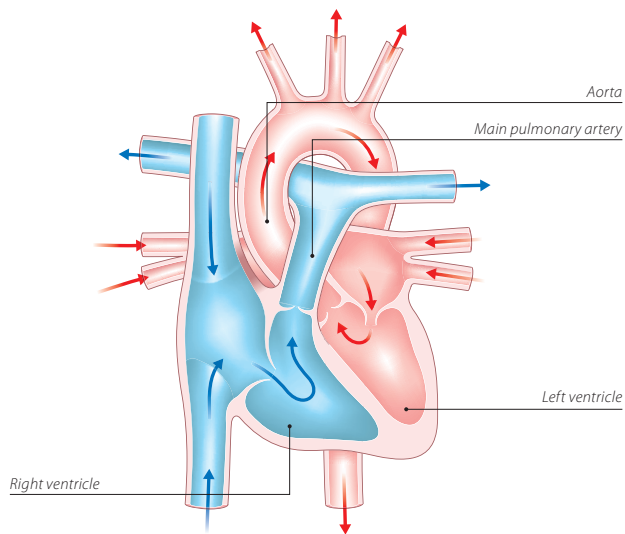
## Why me? Why my child?

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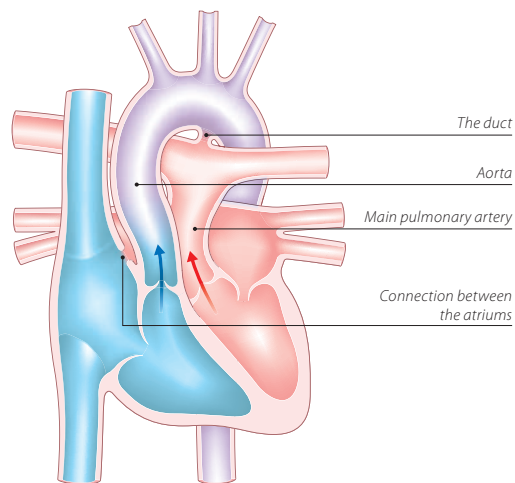
It is not unusual for parents of children with congenital heart disease to blame themselves or to be angry. Anger, disappointment, fear and guilt are all normal feelings to have when you are told that there is something wrong with your child. At first it may be difficult to cope with and it can take 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.<sup>1</sup> 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 transposition of the great arteries?

### The normal heart



### Transposition of the great arteries



Transposition of the great arteries – or TGA for short – means that the pulmonary artery arises from the left ventricle instead of the right, and the aorta arises from the right ventricle instead of the left. See the illustrations above.

In a child with transposition, the blood which doesn't have oxygen in it (blue) gets pumped around the body instead of the blood which does contain oxygen (red). There is a natural connection called the ductus arteriosus – usually called 'the duct' – between the aorta and the pulmonary artery to allow some mixing of blood. This is open while the baby is in the womb but closes shortly after birth. A baby with TGA may die quickly unless he or she has emergency treatment to keep the duct open.

### **What are the symptoms of transposition of the great arteries?**

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The low level of oxygen in your child's blood can make him or her appear blue, particularly on the lips and tongue, inside the mouth, and on the hands. Some children appear pink and just become slightly blue when they cry, while others may appear blue all the time.

Sometimes, transposition of the great arteries is diagnosed before birth, allowing plans for treatment to be made in advance. Your paediatric cardiologist will talk to you about where your baby should be born so that treatment can be given immediately if necessary.

### **What other conditions are associated with transposition of the great arteries?**

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Usually transposition of the great arteries occurs on its own, but it can sometimes occur with other types of congenital heart disease such as a hole in the heart. If your baby has a hole in the heart or another abnormality, your paediatric cardiologist will talk to you about this.

### **How is transposition of the great arteries diagnosed?**

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In most cases, transposition of the great arteries is not diagnosed until 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 child at all.

### **How is transposition of the great arteries treated?**

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The first stage of treatment is to try to keep your child's condition stable by increasing the amount of oxygen-containing blood which gets to different parts of the body.

**Medicines** can help to some extent and most babies will be given a drug called **prostin**, as an injection into a vein. The prostin will help to keep the duct open to keep your baby alive. A side effect of this drug can be that it occasionally interferes with the baby's natural breathing, and so your baby may need the support of a **ventilator**.

### Balloon septostomy

Most babies with transposition of the great arteries will need a procedure called a **balloon septostomy** in the first few days to make sure they get enough oxygen circulating around their body to survive while they are waiting for surgical treatment.

A balloon septostomy involves putting a catheter (a fine, hollow tube), with a small collapsed balloon at its tip, into a vein at the top of the baby's leg or at the umbilicus (belly button). The catheter is then guided up into the heart and across the septum – the wall that divides the two atriums. An ultrasound scan will make sure that the tube and the balloon are in the correct position. The balloon is then gently inflated and the catheter is pulled back across the septum, making a hole in it and allowing oxygenated blood to come from the left side of the heart to the right side. This means that the baby has some oxygen-containing blood circulating around the body. The balloon is then deflated and removed.

Balloon septostomy is usually safe, but there is a very small risk of death, or of your baby having a serious complication such as developing a heart rhythm abnormality.

### Surgery

Once your baby has been stabilised, the surgeon can plan when he or she should have surgery to switch the major arteries back to their normal position. This **switch operation** is usually done within the first three weeks of birth. Although this operation is often called 'corrective surgery', it never makes the heart completely normal.

In some rare cases, a switch operation is not suitable for a baby with transposition of the great arteries. Your paediatric cardiologist will talk to you about alternative treatments if this is the case for your child.

### What happens?

Your baby will be given a general anaesthetic for the switch operation. During the operation, 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. Once the heart has been stopped, the surgeon will switch the arteries around to their normal position. After the operation, your child will have a scar in the middle of the chest along the breast bone.



### **What are the risks of surgery?**

Although the switch operation is a very major one, 97 to 98 in every 100 babies survive it.<sup>2</sup> About one in every 100 babies has complications such as brain damage (sometimes permanent), kidney damage or serious heart rhythm abnormalities.<sup>2</sup> There is also the risk of more minor complications after surgery, such as a lung infection, or fluid collecting around the heart or lungs. These minor complications can be treated, but your baby will need to spend longer in hospital.

### **What happens after surgery?**

Your baby will spend a day or two in intensive care, but most babies are well enough to go home a week to 10 days after the operation.

You will have to bring your baby back to the outpatients department again to see the paediatric cardiologist.

### **What happens as my child grows up?**

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Most babies who have had a successful switch operation will lead normal lives in their childhood, but complications of surgery can occur years later. These complications include narrowing of the artery leading to the lungs (the pulmonary artery), leaking heart valves (most commonly the aortic valve), and narrowing of the small arteries (the coronary arteries) which feed the heart muscle with blood. These complications are rare but, if they are serious, your child may need further surgery. You will need to bring your child for regular visits to the outpatients department throughout his or her life, to check for complications such as these.

The switch operation has been used for about 20 years and we know that most patients who have it survive into adult life. Although doctors are optimistic that life expectancy might be close to normal in many patients who have had a switch operation, they will not know for certain for another 50 years or so.

## The specialist centre for congenital heart disease

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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

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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](http://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](http://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](http://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

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### Endocarditis

Everyone who has transposition of the great arteries 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?

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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.<sup>3</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 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.<sup>3</sup>

## Support groups

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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](http://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](http://www.childrens-heart-fed.org.uk)

## About the British Heart Foundation

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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

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### 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

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### 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

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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](http://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

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## Paediatric nurse

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## Paediatrician

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## Social worker

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## Surgeon

## Others

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## 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

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<b>aorta</b>	The main artery of the heart. It supplies oxygen-rich blood to the body.
<b>atrial</b>	To do with the atrium. (See below.)
<b>atrium</b>	One of the two upper chambers of the heart.
<b>cardiac</b>	To do with the heart.
<b>cardiologist</b>	A consultant specialising in heart disease.
<b>catheter</b>	A fine, hollow tube.
<b>chromosomes</b>	Found in the nucleus of every cell in the body, chromosomes contain the genes, or hereditary elements, which establish the characteristics of an individual.
<b>congenital</b>	From birth.
<b>duct</b>	See <i>ductus arteriosus</i> below.
<b>ductus arteriosus</b>	A natural connection between the aorta and the pulmonary artery. Also called the 'duct'.
<b>ECG</b>	See <i>electrocardiogram</i> .
<b>echocardiogram</b>	An ultrasound scan used to produce pictures of the heart and blood vessels.
<b>electrocardiogram</b>	A recording of the electrical activity of the heart. Also called an ECG.

<b>endocarditis</b>	Infection of the lining of the heart or its valves.
<b>genetic</b>	To do with the information that is passed from parents to children through genes in sperm and eggs.
<b>murmur</b>	An extra sound that is sometimes heard when listening to the heart through a stethoscope.
<b>paediatric</b>	To do with paediatrics – the study of children's diseases.
<b>pulmonary</b>	To do with the lungs.
<b>septostomy</b>	A surgical procedure to make a hole in the septum, to allow blood to pass from the left to the right side of the heart.
<b>septum</b>	The wall that keeps the right and left sides of the heart separate.
<b>ventilator</b>	A machine that helps your child breathe.
<b>ventricle</b>	One of the two lower chambers of the heart.
<b>ventricular</b>	To do with the ventricle or ventricles. (See above.)



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## 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

Greater London House  
180 Hampstead Road  
London NW1 7AW  
Phone: 020 7554 0000  
Fax: 020 7554 0100  
Website: [bhf.org.uk](http://bhf.org.uk)