



Understanding your child's heart

Pulmonary atresia with a ventricular septal defect



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You can go directly to the coloured section to read about your child's heart condition



About this booklet

This booklet is for parents of children who have pulmonary atresia with a ventricular septal defect. There is a separate condition called 'pulmonary atresia with intact ventricular septum'. If your child has that condition, it would be more helpful for you to read our booklet *Understanding your child's heart: Pulmonary atresia with intact ventricular septum*. See page 31 for information on how to order the booklet.

This booklet explains:

- what pulmonary atresia with a ventricular septal defect is and how it is diagnosed
- how pulmonary atresia with a ventricular septal defect is treated
- the benefits and risks of treatments
- how to cope as a parent of a child who has pulmonary atresia with a ventricular septal defect, and
- where to go for more support.

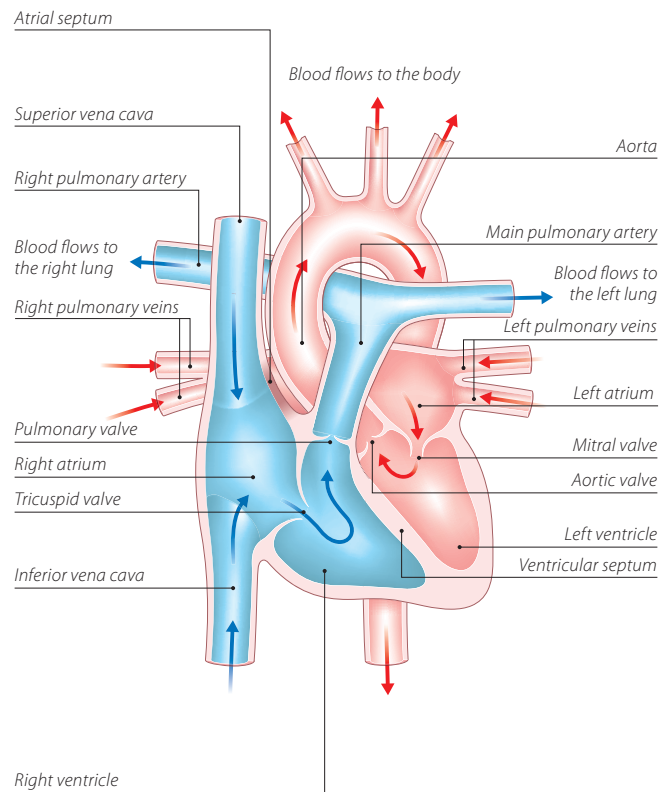
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

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. *Pulmonary atresia with a ventricular septal defect* 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.)

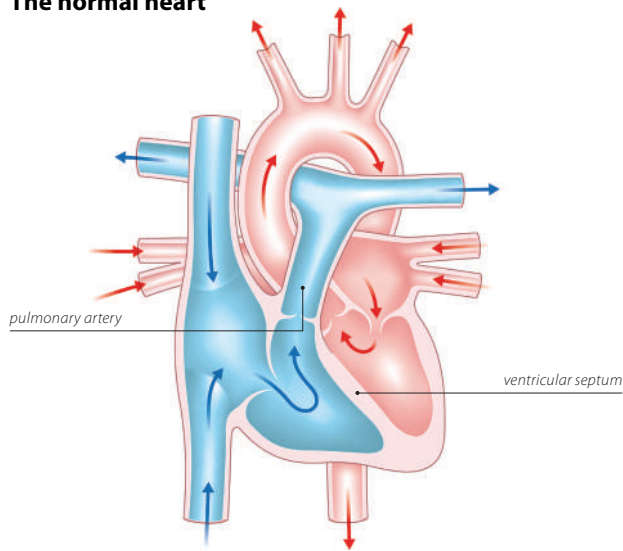
Why me? Why my child?

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. For others it is more serious and, sadly, some children don't survive. However, thanks to advances in diagnosis and treatment, most children can be helped to have a good quality of life.

What is pulmonary atresia with a ventricular septal defect?

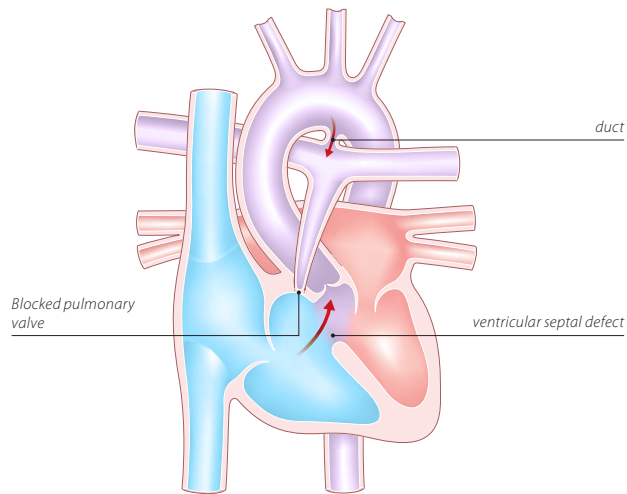
The normal heart



Pulmonary atresia with a ventricular septal defect is a serious type of congenital heart condition. There are two main abnormalities:

- The valve which allows blood to flow from the heart to the lungs – the pulmonary valve – is completely blocked (See the illustration on the opposite page.)

Pulmonary atresia with a ventricular septal defect

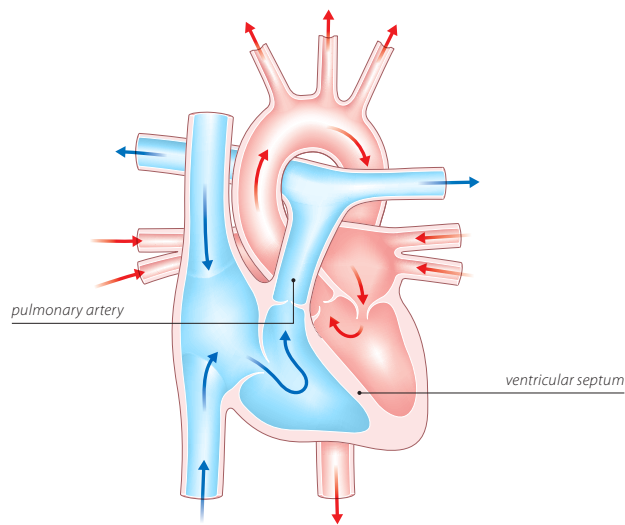


- There is a large hole (ventricular septal defect, or VSD for short) between the two main pumping chambers (ventricles) of the heart.

In the normal circulation, blood passes through the pulmonary artery to the lungs, to collect oxygen. In babies who have pulmonary atresia with a VSD, blood cannot pass

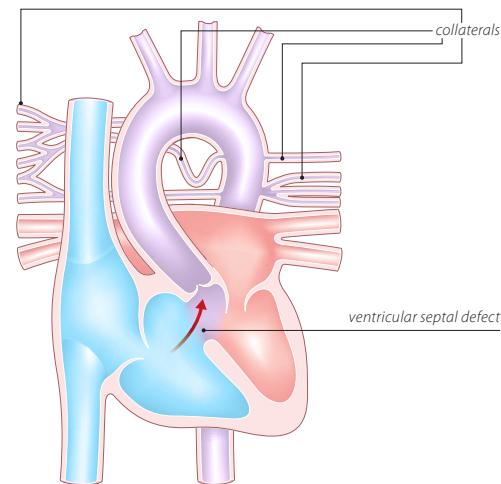
to the lungs in this way. Instead, the blood passes from the aorta to the pulmonary artery through a connection called the **ductus arteriosus** – often called ‘the duct’. (See the illustration on page 11.) The duct is open while the baby is developing in the womb, but it usually closes shortly after birth. In a baby with pulmonary atresia with a VSD, it is vital that the duct stays open after birth if the baby is to survive. We explain more about the treatment to keep it open on page 15.

The normal heart



Some babies with very severe forms of pulmonary atresia with a VSD will have a more complex abnormality. Their pulmonary arteries are extremely underdeveloped and multiple abnormal blood vessels – known as **collaterals** – supply the lungs with blood. (See the illustration below.) Such additional abnormalities can make surgery much more complicated or even impossible. If your baby has any of these complications, your cardiologist will discuss this with you.

Pulmonary atresia with a ventricular septal defect, absent pulmonary artery and collaterals



What are the symptoms of pulmonary atresia with a ventricular septal defect?

Most babies appear normal at birth, but will become blue very quickly and will need to be treated immediately in order to survive.

What other conditions are associated with pulmonary atresia with a ventricular septal defect?

Some babies who have pulmonary atresia with a VSD also have other abnormalities. The most common is a syndrome called '22q11 deletion'. Babies born with 22q11 deletion may have learning difficulties and may also have difficulty fighting some infections. Your paediatric cardiologist will discuss this with you. For more on 22q11 deletion, see our website bhf.org.uk

If your baby has been diagnosed with pulmonary atresia with a VSD before birth, your doctor will discuss with you the option of having a test to check for chromosomal abnormality.

How is pulmonary atresia with a ventricular septal defect diagnosed?

In some cases, pulmonary atresia with a VSD may be detected before birth, but in other cases 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. It doesn't hurt your baby at all.

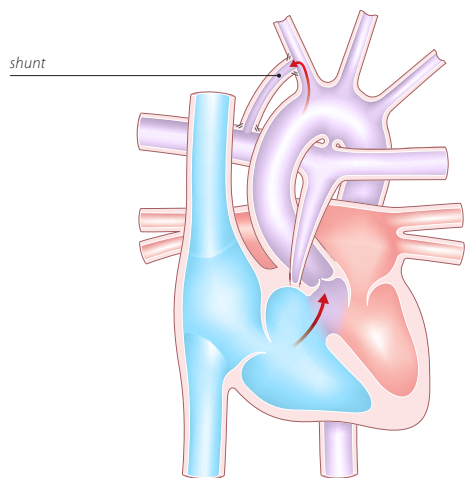
How is pulmonary atresia with a ventricular septal defect treated?

The very first stage of treatment is to keep the duct open. Without treatment, the duct would close and your baby would die. Your baby will be given an injection of a drug called **prostaglandin** to keep the duct open. A common side effect of this drug is that it can interfere with the baby's natural breathing, and so your baby may need the support of a **ventilator**.

Most babies with pulmonary atresia with a VSD will need an operation within the first few weeks of life to treat their condition. This operation is called an **arterial shunt**, often just referred to as a shunt. It allows more blood to flow to the lungs and makes the baby pinker. The operation involves placing a small tube between a branch of the aorta and the pulmonary artery. (See the illustration on page 16.)

This doesn't make the baby's heart normal, but it usually allows the baby to survive until he or she is old enough to consider more major surgery. Even major surgery later in life cannot make the heart completely normal.

After the shunt operation your child will usually have a scar at the side of the chest.



What are the risks of a shunt operation?

About 99 out of every 100 children survive a shunt operation.² But the risk for an individual child will depend very much on the exact abnormalities present. All operations are 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 of the operation for your child. The cardiac surgeon will explain these risks to you before you give your consent for the surgery.

Further surgery

All children with pulmonary atresia and a VSD, and who have had a shunt operation as a baby, will need further treatment when they are older. The exact type of treatment depends very much on how their individual heart abnormality changes with growth.

One of the most important factors is how well the pulmonary arteries grow. If they have grown well, your child may be suitable to have a major repair operation. They will be given a general anaesthetic. The heart is stopped and the function of the heart and lungs are taken over by a 'heart-lung machine' which makes sure that blood is still pumped around your child's body.

During the operation, the surgeon will open up the blockage between the heart and the pulmonary artery. In some cases a human, cow or pig valve may be inserted. The VSD is closed by sewing a patch over it.

After the operation your child will have a scar in the middle of the chest, along the breast bone. Although this operation is often called a 'repair' or 'corrective surgery', it does not make the heart completely normal. Over time the valve between the right ventricle and the pulmonary artery will need to be replaced. The timing of this will vary in every case and will depend upon how well the valve functions as time goes by.

In some children the pulmonary arteries may fail to grow. If this is the case the 'repair' operation is not possible. Further shunt operations may be possible, but in some children no further surgery can be done. If this is the case for your child then unfortunately he or she may die very young.

What happens as my child grows up?

Children who have pulmonary atresia with a VSD are always limited to some extent in their physical activities, but specific restrictions on exercise are usually not necessary. It is usually best to allow children with this condition to join in with their friends' activities, including some sport at school, and for them to judge for themselves what they are able to do.

Pulmonary atresia with a VSD is a complex condition. Although surgery can give a reasonable quality of life, it is not possible to correct the heart abnormality and we do not know how long children with this condition will live for.

Even many years after apparently successful surgery new problems may arise, which require treatment. One of the most common late problems is the development of abnormal heart rhythms. These can usually be treated, but can be serious and in rare cases can even be fatal. So, it's important that your child has regular outpatient reviews with a cardiologist, 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 specialist centre for children with congenital heart disease (paediatric cardiac centre).

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 (some general hospitals have paediatricians who have had special training in cardiology). This means that your child will be looked after locally, but will go to the specialist centre for specialised treatment. (See page 24 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 24.)

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 20). 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 pulmonary atresia with a VSD 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** (similar cost to 01 or 02 numbers) 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

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

Action for Sick Children

32b Buxton Road, High Lane, Stockport SK6 8BH
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

Grown Up Congenital Heart Patients Association

Saracen's House, 25 St Margaret's Green, Ipswich IP4 2BN
Phone: 0800 854759. Website: www.guch.org.uk

Max Appeal

Supporting families affected by 22q11 deletion and other syndromes
Phone: 0800 389 1049.
Website: www.maxappeal.org

www.yheart.net

A website for young people with heart conditions

Child Death Helpline

Offers support and befriending for those affected by child death
Phone: 0800 282 986. Website: www.childdeathhelpline.org.uk

Compassionate Friends

For parents who have lost a child and of any age, including adult children.
Phone: 0845 123 2304. Website: www.tcf.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 Care 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 (similar cost to 01 or 02 numbers)

For information and support on anything heart-related.

Other resources

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/congenital or call the Heart Helpline on **0300 330 3311** (similar cost to 01 or 02 numbers).

Children with congenital heart disease (DVD)

Three families share their experiences from diagnosis to treatment, and staff at the Evelina Children's Hospital offer guidance on parents' common concerns.

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.

To order any of our resources

- 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 DVDs ask for a copy of the *Our heart health catalogue*.

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.
atrium	One of the two upper chambers of the heart.
cardiac	To do with the heart.
cardiologist	A consultant specialising in heart disease.
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.
echocardiogram	An ultrasound scan used to produce pictures of the heart and blood vessels.
endocarditis	Infection of the lining of the heart or its valves.
heart-lung machine	A machine that pumps blood around the body while the heart is stopped during an operation.
pulmonary	To do with the lungs.
ventilator	A machine that helps your child breathe.
ventricle	One of the two lower chambers of the heart.
ventricular septal defect	A hole in the wall between the two ventricles of the heart. Also called VSD.

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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.

2011 is the British Heart Foundation's 50th birthday. Since 1961, we have been the nation's heart charity, dedicated to saving lives through pioneering research, patient care, campaigning for change and by providing vital information. But we urgently need your help. We rely on your donations of time and money to continue our life-saving work. Because together we can beat heart disease.

bhf.org.uk

 **Heart Helpline**
0300 330 3311
bhf.org.uk

Information & support on anything heart-related. Phone lines open 9am to 5pm Monday to Friday. Similar cost to 01 or 02 numbers.

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