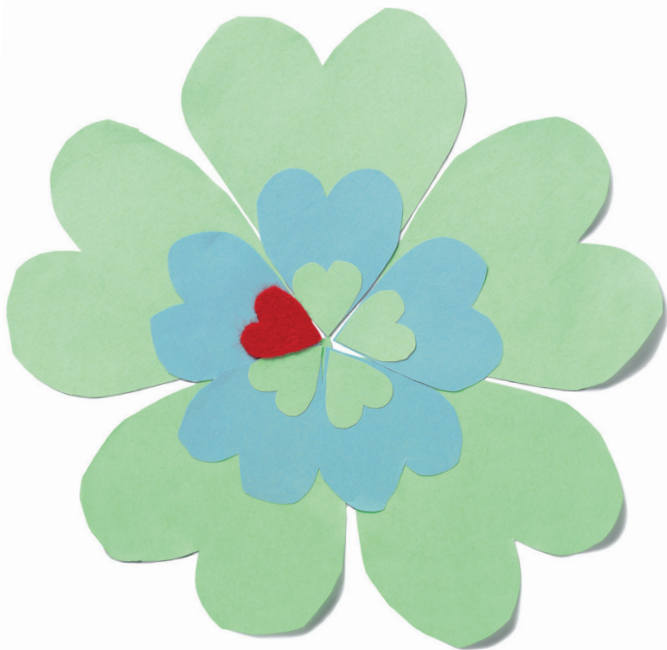




Understanding your child's heart

Complete & partial atrioventricular septal defect



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 an atrioventricular septal defect?	10
What are the symptoms of an atrioventricular septal defect?	13
What other conditions are associated with an atrioventricular septal defect?	13
How is an atrioventricular septal defect diagnosed?	13
How is an atrioventricular septal defect 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?	25
Support	26
About the British Heart Foundation	27
References	29
Contacts	30
Hospital visits	31
The medical terms and what they mean	32
Index	33
Have your say	39



About this booklet

This booklet is for parents of children with a complete or partial atrioventricular septal defect.

This booklet explains:

- what a complete or partial atrioventricular septal defect is and how it is diagnosed
- how a complete or partial atrioventricular septal defect is treated
- the benefits and risks of treatments
- how to cope as a parent of a child with a complete or partial atrioventricular septal defect, and
- where to go for more support.

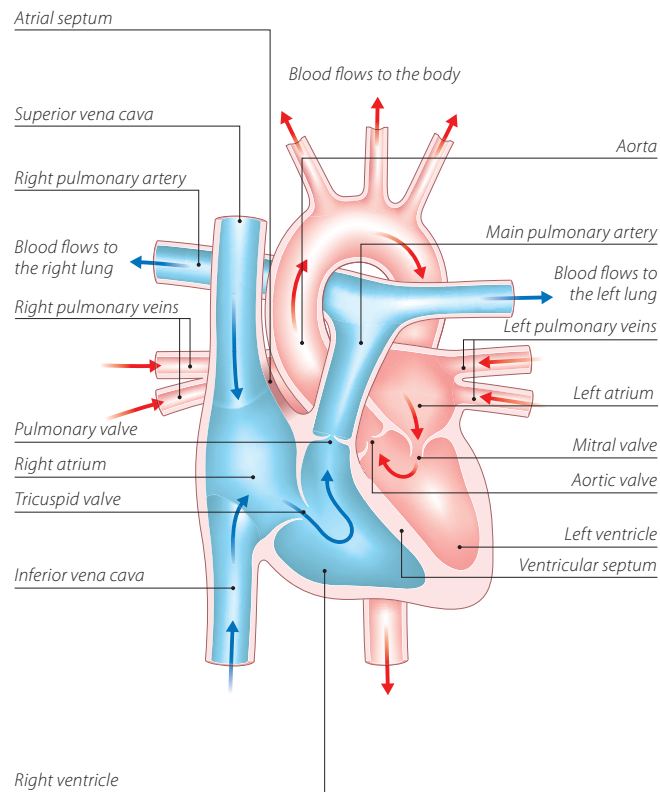
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

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. ***Atrioventricular septal defect (AVSD)*** 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.)

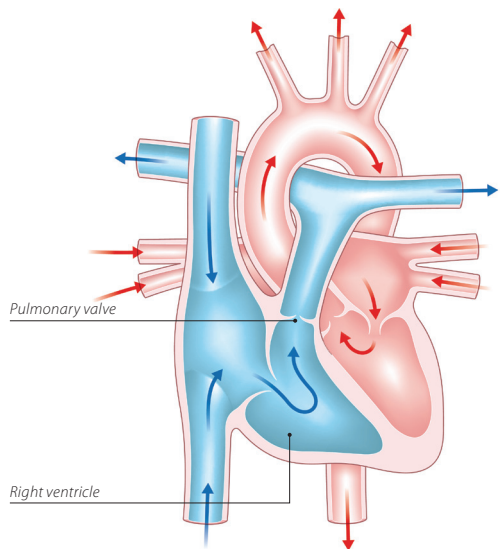
Why me? Why my child?

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 an atrioventricular septal defect?

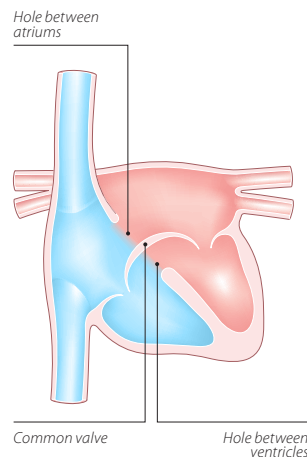
The normal heart



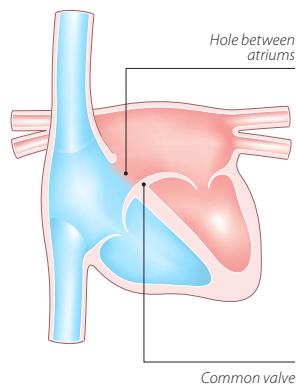
There are two types of atrioventricular septal defect (AVSD) – complete and partial. Both are described in this booklet.

- A **complete AVSD** consists of a complicated hole between the right and the left side of the heart. The hole

Complete atrioventricular septal defect



Partial atrioventricular septal defect



is in the centre of the heart, effectively producing two holes – one between the atriums, and the other between the ventricles. (See the illustration on the left, above.)

- A **partial AVSD** is very similar but there is no hole between the ventricles. (See the illustration on the right, above.)

In both types the inlet valves of the heart are also abnormal. Normally, there are separate right and left-sided inlet valves – the tricuspid valve on the right side, and the mitral valve on the left. In children with either a complete or partial AVSD, the middle part of the two valves is shared between the left and right sides of the heart. This is often called a ***bridging leaflet*** or a ***common valve***.

Normally the valves open to allow the ventricles to fill with blood and then they close to allow all the blood to be pumped out through the two main arteries leaving the heart. The abnormal valves may not be ‘watertight’, so when they close, some blood may leak backwards from the ventricles into the atriums.

The degree of the valve abnormality will affect the success of the operation and its long-term result. We talk more about the operation on page 14.

In some children with either types of AVSD, either the right or the left side of the heart may be underdeveloped. If this is severe, the child will require more complicated surgery and their longer-term outlook would be more uncertain.

AVSD may be associated with many other heart conditions which can also affect the treatment available and the outcome for your child. Your cardiologist will discuss your child’s condition with you.

What are the symptoms of an atrioventricular septal defect?

Children with an AVSD usually look perfectly well when they are born. Children with a complete AVSD often gradually become breathless over the first month or so after birth. Children with a partial AVSD usually remain well.

What other conditions are associated with an atrioventricular septal defect?

Some babies with an AVSD may not have any other abnormality, but many do. The most common are chromosomal abnormalities which are often associated with learning difficulties. Down’s Syndrome is the most common example. See page 26 for where to get more information on Down’s Syndrome. If your baby has been diagnosed with an AVSD before birth, your doctor will discuss with you the option of having a test to check for chromosomal abnormality.

How is an atrioventricular septal defect diagnosed?

Many cases can be diagnosed while the baby is still in the womb, but some are not detected until after birth. Usually, the only test that is necessary to make the diagnosis is an ***echocardiogram***. This is a scan of the heart. It is very similar

to the ultrasound scans that are carried out during pregnancy. It does not hurt your baby at all.

How is an atrioventricular septal defect treated?

Most babies with an AVSD don't need immediate treatment, but babies who become breathless may be given medicines to help improve their symptoms until surgery is carried out.

Surgery

Babies with a complete AVSD will usually need major surgery to repair the defect when they are about three to six months old. In babies with partial AVSD the operation is usually not necessary until they are a few years old.

The operations are carried out under general anaesthetic. The heart is stopped and the function of the heart and lungs 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(s) in the heart. The common valve will also be divided into separate right and left parts. Once the heart is repaired, it is restarted.

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

If your baby has additional heart abnormalities, surgery may be much more complicated. Your cardiologist will discuss this with you.

What are the risks of surgery?

About 98 out of every 100 babies who have a complete AVSD with no other heart abnormalities survive the surgery. About 99 out of 100 babies who have a partial AVSD survive the surgery.² For both groups there are serious risks including 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.

The valve on the left side (the mitral valve) always leaks to some extent after the holes have been closed. This is called **mitral regurgitation**. If the mitral valve leaks just a little, this may not matter and no further surgery may be needed. However, if it leaks a lot, it can be a serious problem and further major surgery will be needed. Where possible the valve is repaired, but in some cases it may need to be replaced. Repairing or replacing the mitral valve is very difficult in young children and it carries a further risk to the baby's life.

There is also a small risk that the heart's electrical system may be damaged during the operation. If this happens, your child's heart rate will be slower than normal and a further operation may be needed to implant a pacemaker to make his or her heart beat faster.

Further surgery

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

If your child has a replacement valve, he or she will outgrow the replacement valve, and will therefore need to have it replaced again with a larger one. The age at which this surgery needs to be done varies from one child to another.

What happens after surgery?

Most children need to stay in hospital for about a week after the major repair operation, although if there are complications they may need to stay for longer than this. Your doctor may prescribe some medicines for you to give to your child after leaving hospital, but he or she will probably need to take these only for a month or so.

What happens as my child grows up?

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

After the operation you will need to take your child for regular visits to the outpatients clinic, to make sure that the mitral regurgitation does not become worse as time goes by.

After repair of the AVSD a small proportion of children can develop narrowing below the aortic valve. This is known as **subaortic stenosis** and it may need further surgery as time goes on.

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 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 atrioventricular septal defect 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

Down's Syndrome Association

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

www.yheart.net

A website for young people with heart conditions

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*.

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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.
pacemaker	A device that is implanted in the chest to stimulate the heart to beat.
pulmonary	To do with the lungs.
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.

Index

aorta	6, 7
aortic valve	6, 7, 17
atrium	6, 7
benefits	21
breathlessness	13, 14
bridging leaflet	12
Carer's Allowance	22
causes of congenital heart disease	8
centre: specialist centre	18, 19
chromosome abnormality	13
common valve	11, 12, 14
complications of surgery	15
congenital heart disease	8
diagnosis	13
Disability Living Allowance	21
Down's Syndrome	13
echocardiogram	13
endocarditis	23
fares to hospital	22
finance	20
future	17, 23
heart: how the normal heart works	6
infection	23
long term outlook	17
medicines	14, 16

mitral regurgitation	15, 17
mitral valve	6, 7, 12, 15
money	20
normal heart	6
operation	14–16
pregnancy	24
professionals	18
pulmonary artery	6, 7
pulmonary valve	6, 7
pulmonary veins	6, 7
risks: of surgery	15
septum	6, 7
specialist centre	18, 19
specialist team	18, 19
subaortic stenosis	17
support groups	26
surgery	14–16
symptoms	13
travel expenses	22
treatment	14
tricuspid valve	6, 7, 12
valves	6, 7, 10, 11, 12, 14, 15
ventricle	6, 7

Notes



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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