



Understanding your child's heart

Single ventricle circulation



Contents

About this booklet	5
The normal heart	6
What does single ventricle circulation mean?	8
What are the symptoms of single ventricle circulation?	8
What does treatment involve?	8
What happens as my child grows?	17
The specialist centre for congenital heart disease	20
Coping with everyday life	22
General advice for the future	25
Support	26
About the British Heart Foundation	28
Contacts	30
Hospital visits	31
The medical terms and what they mean	32
Index	36
Have your say	39



About this booklet

This booklet is for the parents of children with complex congenital heart disease where there is effectively only one main pumping chamber in the heart. Your child may already have had one or more operations. This booklet describes the treatment your child may need later on, and the long-term outcomes for children with this condition.

The booklet explains:

- what single ventricle circulation is
- how single ventricle circulation is treated
- the benefits and risks of treatments
- how to cope as a parent of a baby who has single ventricle circulation
- where to go for more support.

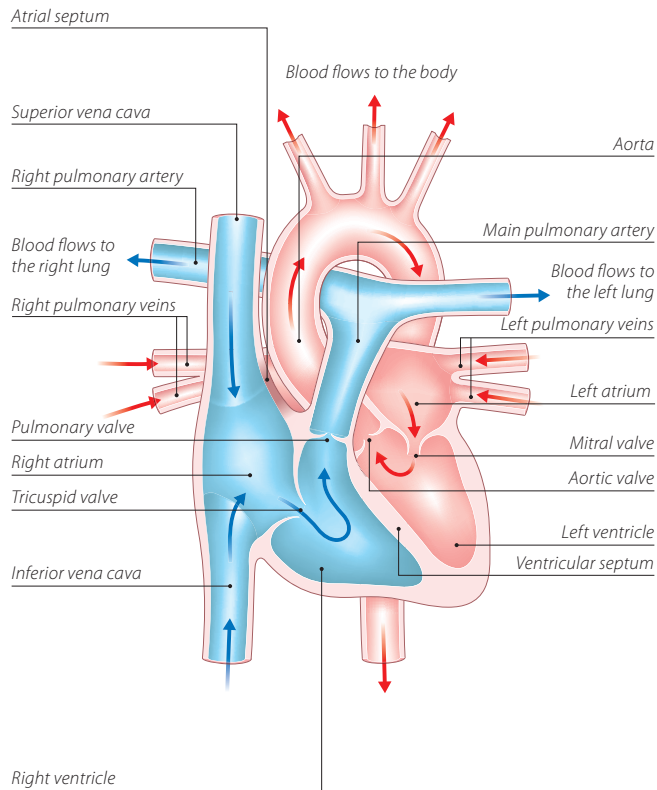
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

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 does single ventricle circulation mean?

Single ventricle circulation is a term used to describe a wide variety of very serious heart abnormalities which are too complicated to be corrected. In most of these conditions, one of the heart's lower chambers (the ventricles), or one of the inlet valves of the heart (the mitral valve or the tricuspid valve), has failed to develop normally. Children with single ventricle circulation effectively have only one ventricle rather than the normal two. This significantly affects the way blood is delivered to the lungs and to the rest of the body.

What are the symptoms of conditions associated with a single ventricle circulation?

Some children appear relatively well, despite their complex heart condition. However, others may appear blue, especially around the lips, tongue, mouth and hands. Most children are in some way limited in the amount of physical exercise they can do.

What does treatment involve?

Most children born with these very complex heart conditions will already have had at least one operation in infancy, and further major operations later on in early childhood or adolescence. Operations such as an *arterial*

shunt operation or *pulmonary artery banding* are usually carried out when the baby is quite small, to help stabilise his or her condition. All of these operations are palliative (which means that they improve the circulation, but don't correct the heart abnormality), and are carried out in stages in preparation for major surgery later. For more information on the initial treatments available for the type of condition your child has, see the *Understanding your child's heart* series of booklets (see page 28).

This booklet describes two types of surgery and explains what they aim to do:

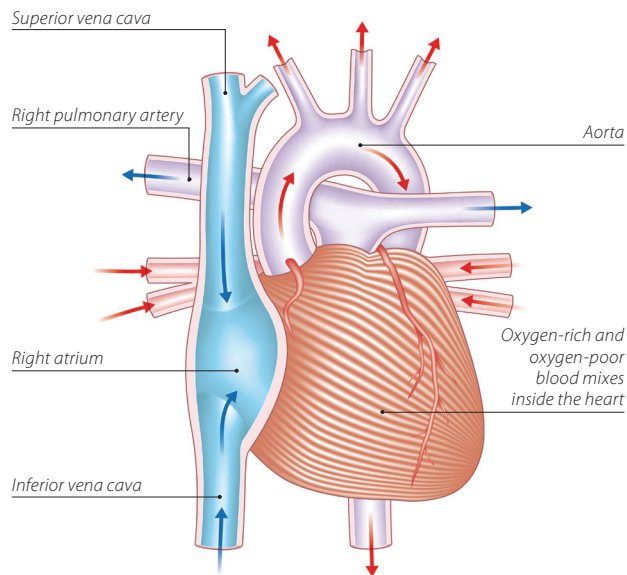
- the cavopulmonary shunt, which is sometimes called a Glenn shunt, and
- a Fontan-type operation.

Cavopulmonary shunt *Also called a Glenn shunt*

The purpose of inserting a cavopulmonary shunt is to increase the blood flow to the lungs, and also to reduce the amount of work the heart's pumping chambers need to do. A cavopulmonary shunt will help your baby but does not correct the underlying complex heart abnormality.

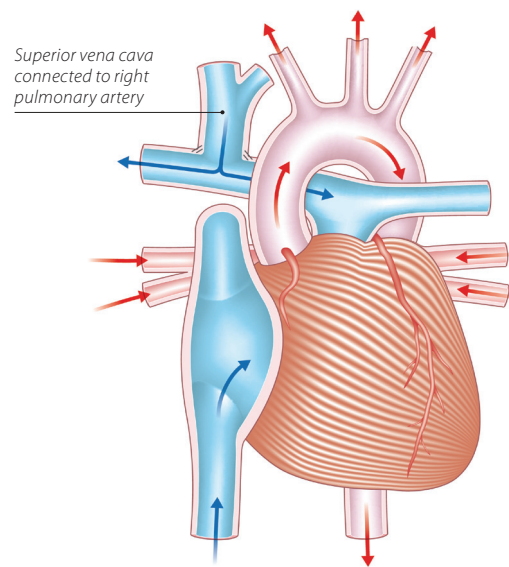
Your child will be given a general anaesthetic. The heart is stopped and the function of the heart is taken over by a heart-lung machine. The surgeon will then connect the superior vena cava directly to one of the arteries that takes

Single ventricle circulation



blood to the lungs (the right pulmonary artery). (See the illustrations above.) After the surgery, your child will have a scar in the middle of the chest, along the breastbone.

Cavopulmonary shunt



Your child will need to stay in hospital for a few days after the surgery. You will need to take your child to the outpatients department within a few weeks, for a check-up

with the cardiologist. After that, your child will need further regular check-ups.

A cavopulmonary shunt is not suitable for every child with single ventricle circulation, especially if the pressure in the pulmonary artery is high. If this is the case with your child, your cardiologist will discuss what other types of treatment might be suitable.

Even after a cavopulmonary shunt operation, most children will become more blue and short of breath on exertion as they grow.

What are the risks of a cavopulmonary shunt operation?

There is a risk of your child dying either during or shortly after the operation, but this risk will vary depending on the exact type of heart condition your child has. For example, children with hypoplastic left heart have a higher risk of death than children with other types of congenital heart disease. Your paediatric cardiologist or cardiac surgeon will discuss your child's individual risk with you before surgery.

There are also risks of other complications, such as a narrowing where the superior vena cava has been joined to the pulmonary artery, brain damage, stroke or internal bleeding. There may also be kidney damage, but this usually

recovers with treatment. Some children are also at risk of fluid collecting around the lungs.

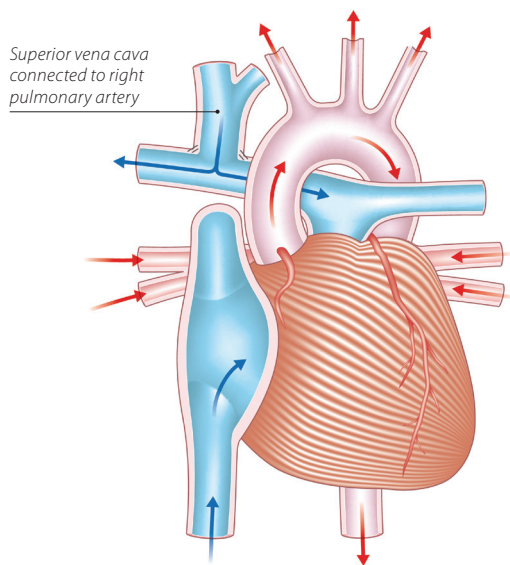
Because these children have very complex congenital heart disease, they are also at risk of complications that are not related to their operations such as heart rhythm disturbances or even sudden death. This is more likely to happen in some children such as those with hypoplastic left heart than in others.

Fontan-type operation

The purpose of this operation is to improve the amount of oxygen in the blood and in most cases to improve exercise capacity. This is achieved by connecting both the inferior and superior vena cava to the pulmonary artery. There are several ways of doing this.

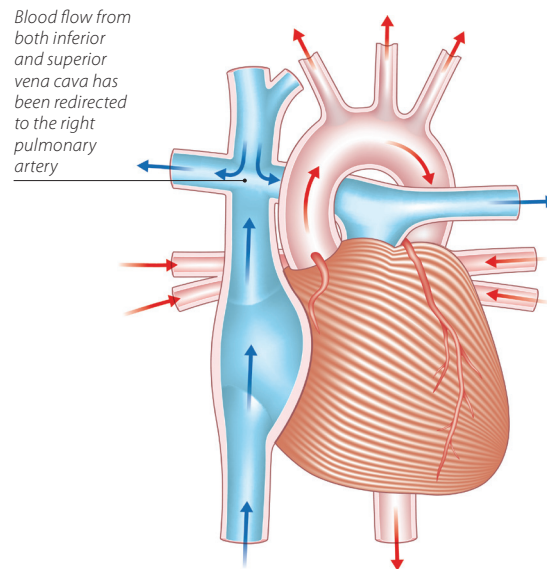
The Fontan operation was first carried out in 1968. Since then, several modifications to the original technique have been developed including: a **modified Fontan**, a **fenestrated Fontan**, and a **total cavopulmonary connection** (or **TCP** for short) which is the most common nowadays. Any type of Fontan operation is a major operation, and the timing of surgery will depend on the exact condition your child has. Your child's cardiac surgeon will explain exactly which type of Fontan operation your child needs. Below, we describe the basic principles of the Fontan-type operation.

Cavopulmonary shunt



Your child will be given a general anaesthetic. The heart is stopped and the heart's function is taken over by a heart-lung machine. The surgeon will redirect the flow of blood from the inferior vena cava to the pulmonary artery. In most cases the superior vena cava has already been connected – see above.

Total cavopulmonary connection



The illustration above shows the TCPC type of Fontan operation.

After the surgery, your child will have a scar in the middle of the chest, along the breastbone.

A Fontan-type operation will not make your child's heart normal but, if the operation is successful, it should allow an adequate blood supply to the lungs to allow your child to grow.

A Fontan-type operation is not suitable for every child with single ventricle circulation, especially if the pressure in the artery that takes blood to the lungs (the pulmonary artery) is high. If this is the case with your child, your cardiologist will discuss other possible treatments with you.

What are the risks of a Fontan-type operation?

While this is a major operation, most children survive the surgery. The risk of death and major complications such as brain damage following a Fontan-type operation varies depending on the exact type of heart condition your child has. Other possible complications include pleural effusion (fluid around the lungs), pericardial effusion (fluid around the heart), and kidney damage. Some children can develop heart rhythm disturbances which need to be treated with medicines, or less commonly with a *pacemaker*.

The length of time your child will need to stay in hospital will vary, depending on how well he or she recovers after surgery. There is an increased risk of developing a blood clot after the surgery, so most children will need to take either warfarin or aspirin to help prevent this. See our factsheet on warfarin at bhf.org.uk

What happens as my child grows?

Children who have a single ventricle circulation are always limited to some extent in their physical activities whatever treatment they have had, but specific restrictions on exercise are usually not necessary. It is usually best to allow children with complex heart disease 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.

Even after many years of being well, young people with single ventricle circulation can develop heart rhythm disturbances. A pacemaker or medication may be needed to treat this.

There may also be abnormal function of the intestine in later life – called 'protein-losing enteropathy'. Damage to the liver can also occur later in life. Some people may also develop heart failure (when the heart muscle becomes less efficient at pumping blood). All of these are serious complications with a significant impact on quality of life.

A small proportion of people who develop heart failure may be suitable for a heart transplant. Your child's cardiologist will discuss this with you if appropriate.

Everyone with single ventricle circulation will need to have regular and life-long follow-up appointments with a cardiologist.

Long term outlook

All types of single ventricle circulation are complex, and even with surgical treatment they cannot be corrected. Although surgery can give a reasonable quality of life, we do not know how long children with these conditions will live. The longest survivors are at present in their 30s, but some children die at a much younger age. The long-term survival varies depending on the type of condition the child has, as well as the complications they may have.

Sadly some children may die very young. For support on coping as a parent with a child who is dying, see page 26. For support with helping a young child with the death of a brother or sister, see the BHF bereavement booklet *The small creature*. For information on how to order this booklet, see page 29.

Pregnancy

If you have a daughter with single ventricle circulation, you need to be aware that pregnancy could carry high risks to both the mother and the baby. So, it's particularly important that your daughter avoids having an unplanned pregnancy. In some cases, young women with single ventricle circulation are advised to avoid pregnancy because of the high risk of death associated with it. If your daughter wants to have a baby, it is essential that she speaks to her cardiologist about it first.

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

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 single ventricle circulation 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**.

Support

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

Little hearts matter

11 Greenfield Crescent, Edgbaston, Birmingham,
West Midlands, B15 3AU
Phone: 0121 455 8982. Website: www.lhm.org.uk

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

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

The Compassionate Friends

For parents who have lost a child 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 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.

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** (local rate number).

One of the following titles might be relevant to your child:

- Double inlet ventricle
- Hypoplastic left heart
- Pulmonary atresia with intact ventricular septum
- Tricuspid atresia.

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.

The small creature

This beautiful, award winning resource is to help children under 11 who are bereaved.

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 *Our heart health catalogue*.

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.
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.
genetic	To do with the information that is passed from parents to children through genes in sperm and eggs.
heart-lung machine	A machine that pumps blood around the body while the heart is stopped during an operation.
pacemaker	An electrical instrument which stimulates contractions (beats) of the heart.
paediatric	To do with paediatrics – the study of children's diseases.
pulmonary	To do with the lungs.
ventricle	One of the two lower chambers of the heart.

Notes

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Index

aorta	6, 7
aortic valve	6, 7
arterial shunt operation	8
atrium	6, 7
benefits	23
blueness	8, 12
Carer's Allowance	24
cavopulmonary shunt	9, 11, 12
centre: specialist centre	20, 21
complications of surgery	12
Disability Living Allowance	23
endocarditis	25
fares to hospital	24
fenestrated Fontan	13
finance	22
Fontan operation	13–16
future	17, 25
Glenn shunt	9
heart: how the normal heart works	6
infection	25
long term outlook	17
medicines	16
mitral valve	6, 7
modified Fontan	13
money	22
normal heart	6
operation	8–16
pacemaker	16
pregnancy	19
professionals	20
pulmonary artery	6, 7

pulmonary artery banding	9
pulmonary valve	6, 7
pulmonary veins	6, 7
rhythm: heart rhythm	16
risks: of cavopulmonary shunt	12
of Fontan-type operation	16
septum	6, 7
specialist centre	20, 21
specialist team	20, 21
support groups	26
surgery	8–16
symptoms	8
TCPC	13, 15
team	20
total cavopulmonary connection	13
travel expenses	24
treatment	8–16
tricuspid valve	6, 7
valves	6, 7
ventricle	6, 7
warfarin	16



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 Or, write to us at the address on the back cover.

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- Dr James Gnanapragasam, Consultant Paediatric Cardiologist, Southampton General Hospital
- 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
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Fax: 020 7554 0100
Website: bhf.org.uk

C7/0210