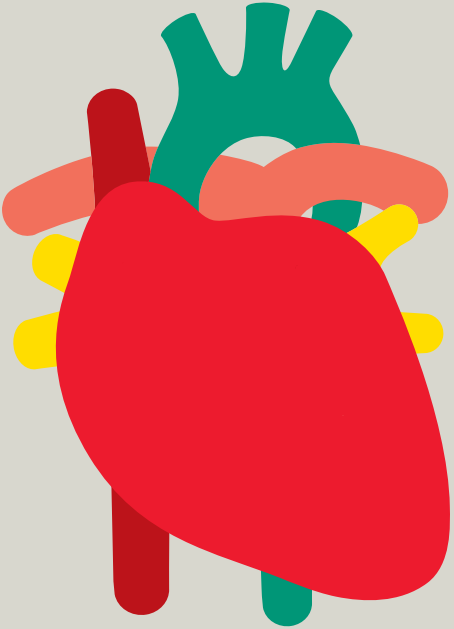
British Heart Foundation

Understanding your child's heart

Double inlet ventricle

10



FIGHT FOR EVERY HEARTBEAT

bhf.org.uk

About this book

If you're reading this book, you've probably just had some very upsetting news and have lots of questions running through your mind.

We've written this book to help answer some of those questions. We'll go through:

- what double inlet ventricle is and how it is diagnosed
- how double inlet ventricle is treated
- the benefits and risks of treatments
- what happens as your child grows up
- where to go for more support.

This booklet shouldn't replace the advice that doctors and nurses may give you, but it should help you to understand what they tell you.

Contents

What is a congenital heart condition?

It's an abnormality of the heart that developed in the womb. Sometimes, a **congenital** heart condition is diagnosed when the baby is still developing in the womb, but most times it's not discovered until after the baby is born. There are lots of different types of **congenital** heart conditions.

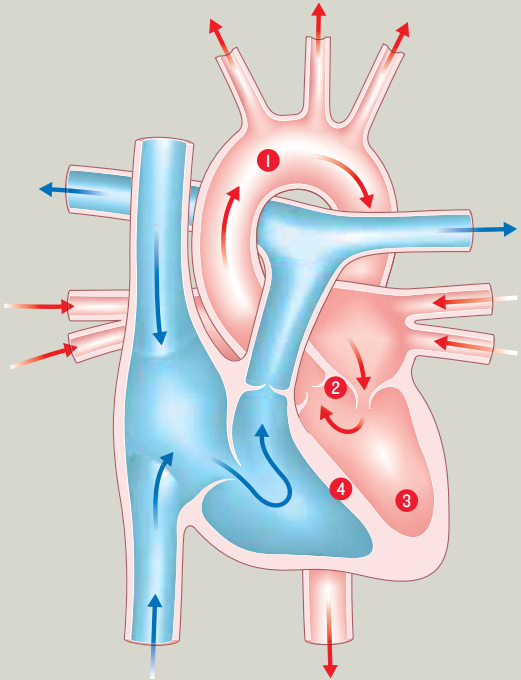
Each day, 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research, visit **bhf.org.uk/research**

What causes a congenital heart condition?

In most cases, it's caused by something going wrong during the very early stages of the pregnancy.

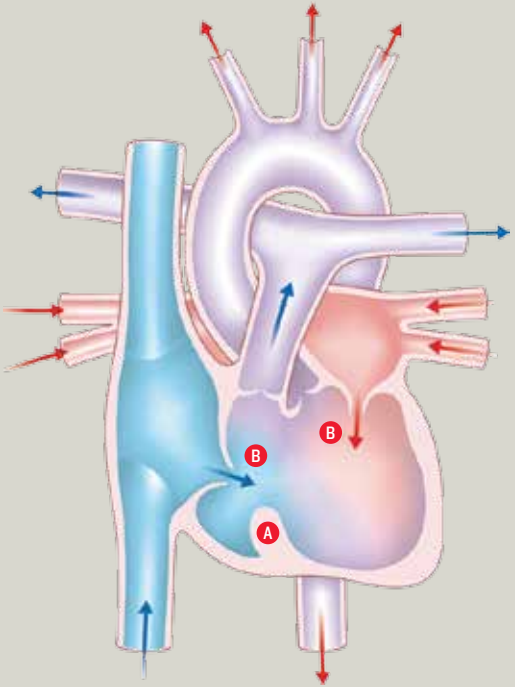
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome means a group of symptoms that commonly appear together as part of a condition).*

Normal heart



1. Aorta
2. Aortic valve
3. Left ventricle
4. Ventricular septum

Double inlet ventricle



A. Ventricular septum (double inlet left ventricle)
B. Blood from both atria flows into one big ventricle

What is double inlet ventricle?

Double inlet ventricle is a serious congenital heart condition. There main abnormalities are:

- Both the left atrium and the right atrium drain into one large ventricle.
- The other ventricle is usually very small.
- There is a hole in the ventricular septum (the wall between the ventricles).

In a normal heart, blood from the right **atrium** drains into the right ventricle, and blood from the left **atrium** drains into the left ventricle. In children with double inlet ventricle, blood from both **atria** drains into one ventricle only. This ventricle then pumps blood through both the pulmonary artery and the **aorta**. *(See the illustration on page 5)*.

Children with a double inlet ventricle may also have other abnormalities of the heart. Some may have a narrowing of the **aorta**, known as *coarctation of the aorta*. In others, there may be narrowing of the pulmonary valve, called *pulmonary stenosis*. The pulmonary valve is the valve that lets the blood out of the heart to the pulmonary arteries and on to the lungs.

What are the symptoms of double inlet ventricle?

The symptoms of double inlet ventricle vary from one baby to another, depending on which additional heart abnormalities they may have. Some babies become breathless early in life if there is too much blood flowing to the lungs. Some will also appear blue because blood mixes in the large ventricle rather than being separated into oxygen-rich and oxygen-poor blood. Babies who are breathless may not gain weight normally.

For more information and support about growing up with a heart condition, visit **bhf.org.uk/heart-health/children-and-young-people**

What other conditions are associated with double inlet ventricle?

Children with double inlet ventricle may also have other abnormalities of the heart, including coarctation of the aorta, pulmonary atresia and pulmonary valve stenosis. Your child's **cardiologist** will discuss this further with you if necessary.

To order booklets on the heart abnormalities mentioned above, please visit *bhf.org.uk/congenital*

How is double inlet ventricle diagnosed?

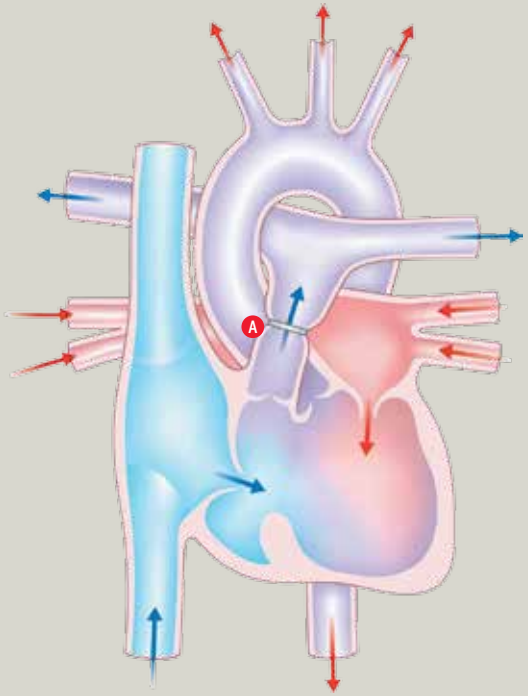
In some cases, the diagnosis is made before birth, but sometimes it is not diagnosed until after the baby is born. Usually, the only test that is needed to make the diagnosis is an **echocardiogram**. This is an ultrasound scan of the heart. It is very similar to the scans that are carried out during pregnancy, and it won't hurt you or your child at all.

How is double inlet ventricle treated?

It's not possible to correct this abnormality with surgery, but there are operations that can help children to have a better quality of life. The type of and timing of surgery recommended for a baby with double inlet ventricle will depend on which additional abnormalities they may have, and how severe they are.

Babies who have too much blood flowing to the lungs need surgery to reduce the blood flow. This surgery is called *pulmonary artery banding*. Babies with too little blood flowing to the lungs need surgery to correct it – called a *shunt operation*. Babies who also have coarctation of the aorta usually need surgery to repair the narrowing in the **aorta** within the first few weeks of life. We describe these surgeries on the following pages. If your child has any of the types of surgery described in this booklet, he or she will need to have a general anaesthetic.

Pulmonary artery banding



A Band

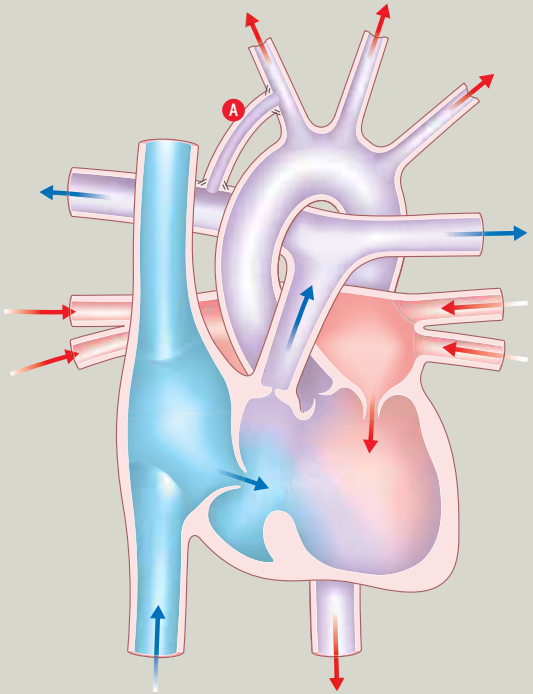
This is an operation to reduce the blood supply to the lungs. The timing of the operation depends on how breathless your baby becomes. Sometimes, it is carried out within the first few months of life.

The surgery involves placing a band around the **pulmonary** artery to narrow it. The band reduces the high blood flow to the lungs, reducing breathlessness and lowering the blood pressure in the **pulmonary** artery, to try to prevent lung damage. *(See the illustration on page 12)*. The surgery usually leaves a scar at the side of the chest rather than in the middle.

Shunt operation

A shunt operation allows more blood to flow to the lungs and makes the baby pinker. The operation involves placing a small tube made of synthetic material between a branch of the **aorta** and the **pulmonary** artery. *(See the illustration on page 14)*. The surgery usually leaves a scar at the side of the chest rather than in the middle.

Arterial shunt operation



A Shunt

Repair of the coarctation of the aorta

If your child needs surgery for **coarctation of the aorta**, the surgeon will place a clamp on the **aorta** to stop the blood flow and make it easier to operate. He or she will then cut out the narrowed part of the **aorta** and sew the ends back together. Or, the surgeon may use a patch made of a special material to enlarge the narrowing. After the operation, your child will have a scar either on the left side of the chest or under their arm, or on the middle of the chest.

What happens after surgery?

How long your child will need to stay in hospital after any of the surgeries mentioned above varies. Many children recover quickly and may only need to stay for about a week. You will have to take your child to the outpatients department to see the **paediatric cardiologist** for a check-up a few weeks after the operation.

Will my child need further surgery?

The exact nature and timing of any further surgery will depend on your child's progress. The type of surgery most commonly carried out is a *cavopulmonary shunt (Glenn Shunt)*, which involves connecting the superior vena cava directly to the **pulmonary** arteries. Further surgery after this usually involves redirecting the blood flow from the inferior vena cava to the **pulmonary** artery. This is called a *total cavopulmonary connection*, or *TCPC* for short.

Cavopulmonary shunt, also called a Glenn shunt

This procedure is used to increase the blood flow to the lungs, and also to reduce the workload of the heart. A cavopulmonary shunt does not correct the underlying heart abnormality.

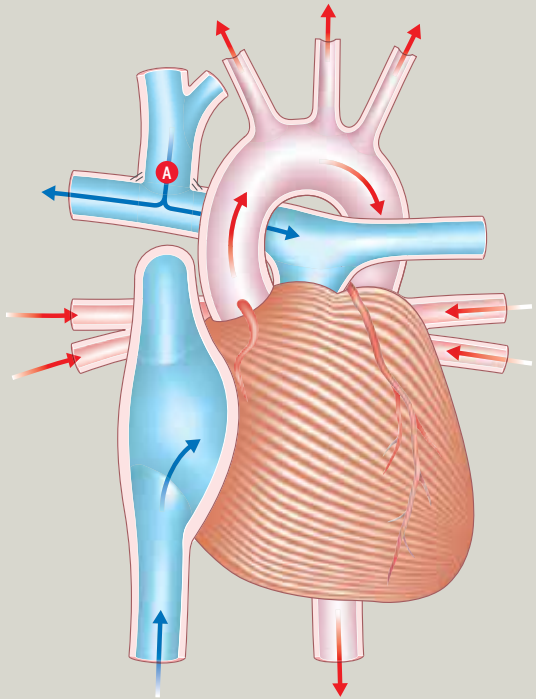
Your child will be given a general anaesthetic. The heart will be stopped and the function of the heart will be taken over by a **heart-lung machine**. The surgeon will connect the superior vena cava directly to one of the arteries that takes blood to the lungs *(the right **pulmonary** artery)*. *(See the illustration on page 18)*. After the surgery, your child will have a scar in the middle of the chest, along the breastbone.

Your child will need to stay in hospital for a few days after the surgery, and will need to visit the outpatients department within a few weeks for a check-up with the **cardiologist**. After that, your child will need regular check-ups.

What are the risks of a cavopulmonary shunt operation?

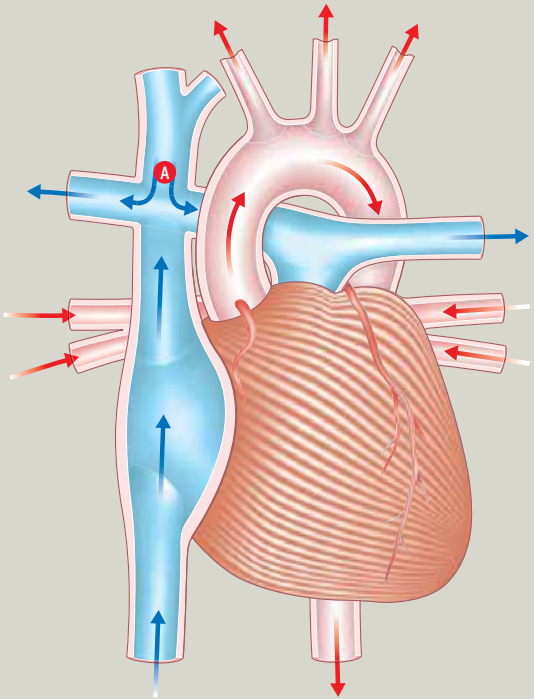
Most children survive this surgery, but they may become more blue and short of breath on exertion as they grow. The risk of death and other complications – such as narrowing where the superior vena cava has been joined to the **pulmonary** artery, brain damage, stroke or internal bleeding – varies based on the exact type of heart condition your child has. Other possible complications include pleural effusion *(fluid around the lungs)* and kidney damage. Your **paediatric cardiologist** or cardiac surgeon will discuss your child's individual risk with you before surgery.

Cavopulmonary shunt



A Superior vena cava connected to pulmonary artery

Total cavopulmonary connection



A Blood flow from both inferior and superior vena cava has been redirected to the right pulmonary artery

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.

Many modifications to the original Fontan operation technique have been developed, including: a *modified Fontan*, a *fenestrated Fontan*, and *total cavopulmonary connection (or TCPC for short)*. Any type of Fontan operation is a major operation. Your child's cardiac surgeon will explain exactly which operation your child needs.

Your child will be given a general anaesthetic. The heart will be stopped and the heart's function will be 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 diagram on page 19)*. The illustration on page 19 shows the TCPC type of Fontan operation. After 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.

What are the risks of the Fontan-type operation?

Most children survive this surgery. The risk of death and major complications – such as brain damage – 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 up?

Children with a double inlet ventricle are almost always limited to some extent in their physical activities. Physical endurance may get worse in teenage years or young adult life, but specific restrictions on exercise are usually not necessary. Your child's **cardiologist** will tell you if there are any specific forms of exercise or activities they should avoid.

Although surgery can give a better quality of life, it is not possible to correct the heart abnormality and it's uncertain how long children with this condition will live for. The longest survivors at present are in their 30s. Heart transplantation may be an option for some patients, although this is rarely considered before adulthood.

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

If you have one child with a **congenital** heart condition, there is around a 1 in 40 chance that if you have another child, they will have a heart condition too[1]. However, this risk may be higher *(or lower)* depending on the type of **congenital** heart condition your child has. Because your risk of having another child with a **congenital** heart condition is higher than it is for other people, your doctor may offer you 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. Do be aware that if you have more than one child with a **congenital** heart condition, the specific condition may not always be the same.

Coping with everyday life

For information on the topics below, please visit **bhf.org.uk/heart-health/conditions/congenital-heart-disease**

- Financial issues
- Low-income benefit
- Disability benefits
- Carer's Allowance
- Fares for visiting your child in hospital

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.

Cardiologist A consultant specialising in heart disease.

Coarctation of the aorta Narrowing of the aorta.

Congenital From birth.

Echocardiogram An ultrasound scan used to produce pictures of the heart and blood vessels.

Heart-lung machine A machine that pumps blood around the body while the heart is stopped during an operation.

Pacemaker A small electrical device fitted in the chest or abdomen. It's used to treat some abnormal heart rhythms (arrhythmias) that can cause your heart to either beat too slowly or miss beats.

Paediatric To do with paediatrics – the study of children's diseases.

References

1. Gill HR, Splitt M, Sharland K, 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.

Acknowledgements

The British Heart Foundation would like to thank all the healthcare professionals involved in the updating of these booklets. Particular thanks are due to:

- Dr James Gnanapragasam, Consultant Paediatric Cardiologist, Southampton General Hospital
- Dr Aaron Bell, Consultant Paediatric Cardiologist, Evelina Children's Hospital

Notes

Notes

For more information and support about children and young people with a heart condition, visit **bhf.org.uk/heart-health/children-and-young-people**

British Heart Foundation

For over 50 years we've pioneered research that's transformed the lives of millions of people living with cardiovascular disease. Our work has been central to the discoveries of vital treatments that are changing the fight against heart disease.

But cardiovascular disease still kills around one in four people in the UK, stealing them away from their families and loved ones.

From babies born with life threatening heart problems, to the many mums, dads and grandparents who survive a heart attack and endure the daily battles of heart failure.

Join our fight for every heartbeat in the UK. Every pound raised, minute of your time and donation to our shops will help make a difference to people's lives.

Text FIGHT to 70080 to donate £3

FIGHT FOR EVERY HEARTBEAT

bhf.org.uk

C10/1117