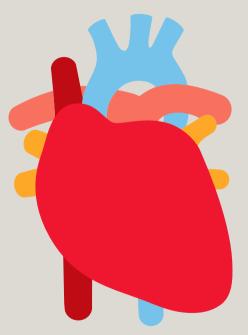




Understanding your child's heart

Transposition of the great arteries





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

Please be aware that this booklet shouldn't replace the advice your doctors or nurses may give you. But it should help make what they tell you that little bit clearer.

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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.

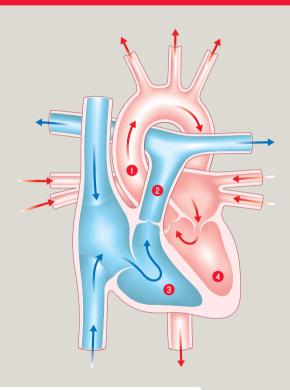
Each day 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research visit **bhf.org.uk/research**

What causes a congenital heart condition?

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

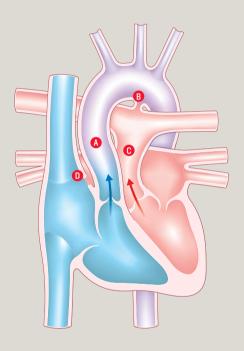
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. (A syndrome is a group of symptoms that commonly appear together as part of a condition).

Normal heart



- Aorta
- 2 Pulmonary artery
- 3 Right ventricle4 Left ventricle

Transposition of the great arteries



- A Aorta
 B The duct
- Pulmonary artery
- Connection between the atria (atrial septal defect)

What is transposition of the great arteries?

Transposition of the great arteries (TGA) means that the **pulmonary** artery arises from the left ventricle instead of the right, and the **aorta** arises from the right **ventricle** instead of the left. (See the illustration on page 5).

In a child with TGA, oxygen-poor blood gets pumped around the body instead of oxygen-rich blood. There is a natural connection called the ductus arteriosus – ('the duct') between the aorta and the pulmonary artery to allow some mixing of blood. This is open while the baby is in the womb but closes shortly after birth. It's important the duct stays open in order for a baby with TGA to survive, unless they have another type of defect such as a hole in the heart that already allows the mixing of blood.

What are the symptoms of transposition of the great arteries?

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

Often, TGA can be diagnosed before birth, allowing plans for treatment to be made in advance. Your child's **paediatric cardiologist** will be able to advise you on the best possible place for your baby to be born, so that treatment can be given immediately if necessary.

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

Most of the time, TGA occurs on its own. But it can sometimes occur with other types of **congenital** heart disease such as a hole in the heart.

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

How is transposition of the great arteries diagnosed?

In some cases, TGA is not diagnosed until after the baby is born, but in others it can be detected before birth using an **echocardiogram**. This is an ultrasound scan of the heart and it won't hurt your baby at all.

How is transposition of the great arteries treated?

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

Medicines can help to some extent and most babies will be given prostaglandin drugs as an injection into a vein. This will help to keep the **duct** open and allow blood to circulate around your baby's body. A side effect of this drug can be that it occasionally interferes with the baby's natural breathing, so your baby may need the support of a **ventilator**.

Balloon septostomy

Most babies will need a procedure called a balloon septostomy in the first few days to make sure they get enough oxygen circulating around their body while they are waiting for surgical treatment. A balloon **septostomy** involves putting a **catheter** (a thin, hollow tube), with a small collapsed balloon at its tip, into a vein at the top of the baby's leg or at the belly button. The catheter is then guided up into the heart and across the **septum** – the wall that divides the two atria. The balloon is then gently inflated and the catheter is pulled back across the septum, making a hole in it and allowing oxygenrich blood to come from the left side of the heart to the right side. This will allow some oxygen-rich blood to circulate around your baby's body. The balloon is then deflated and removed.

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

Surgery

This switch operation to swap the major arteries back to their normal position is usually done within the first three weeks of birth. Although this operation is often called 'corrective surgery', it doesn't make the heart completely normal. In some rare cases, a switch operation is not suitable for a baby with TGA. If this is the case, your child's paediatric cardiologist will talk to you about alternative treatments.

Your baby will be given a general anaesthetic for the switch operation. During the operation, the heart is stopped and the function of the heart is taken over by a 'heart-lung machine' which makes sure that blood is still pumped around your baby's body. Once the heart has been stopped, the surgeon will switch the arteries around to their normal position. After the operation, your child will have a scar in the centre of the chest along the breast bone.

The surgeon will also need to move the small arteries, swapping the coronary arteries from the old aorta to the new one. When the coronary arteries are abnormal, this can make the operation more difficult, and so slightly higher risk.

What are the risks of surgery?

The switch operation is a very major one, but the good news is success rates are high.

Very rare complications can include brain damage, kidney damage or serious heart rhythm abnormalities. More minor complications after surgery, such as a lung infection, or fluid collecting around the heart or lungs are also uncommon but may mean your baby will need to spend longer in hospital.

What happens after surgery?

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

What happens as my child grows up?

Most babies who have had a successful switch operation will lead normal lives in their childhood, do be aware that unfortunately complications of surgery can occur years later. These complications include narrowing of the artery leading to the lungs (the pulmonary artery) and leaking heart valves (most commonly the aortic valve). These complications are rare but, if they are serious, your child may need further surgery.

You will need to bring your child for regular visits to the outpatients department throughout their life, to check for complications such as these. The switch operation has been used for about 30 years and we know that most patients who have it survive into adult life.

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 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.

Do 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 **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/congenital**

- Financial issues
- Low-income benefits
- 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.

Atrial To do with the atrium. (See below).

Atrium One of the two upper chambers of the heart.

Cardiac To do with the heart.

Cardiologist A consultant specialising in heart disease.

Catheter A fine, hollow tube.

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.

Duct See ductus arteriosus below.

Ductus Arteriosus A natural connection between the aorta and the pulmonary artery. Also called the 'duct'.

ECG See electrocardiogram.

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

Electrocardiogram A recording of the electrical activity of the heart. Also called an ECG.

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

Pulmonary To do with the lungs.

Septostomy A surgical procedure to make a hole in the septum, to allow blood to pass from the left to the right side of the heart.

Septum The wall that keeps the right and left sides of the heart separate.

Ventilator A machine that helps your child breath.

Ventricle One of the two lower chambers of the heart.

References

1. 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.

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



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.

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