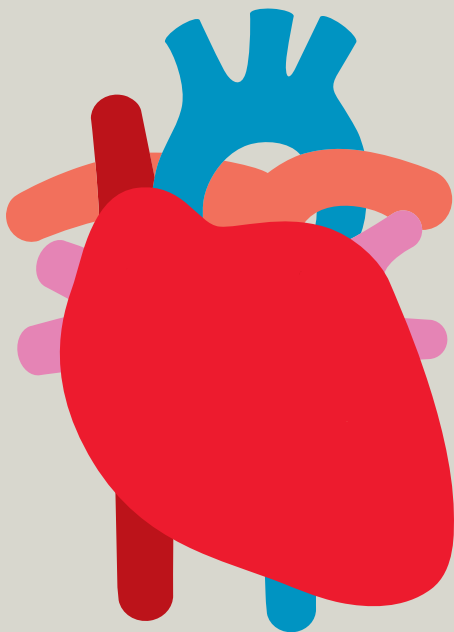


Pulmonary atresia with a ventricular septal defect



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 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
- 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.

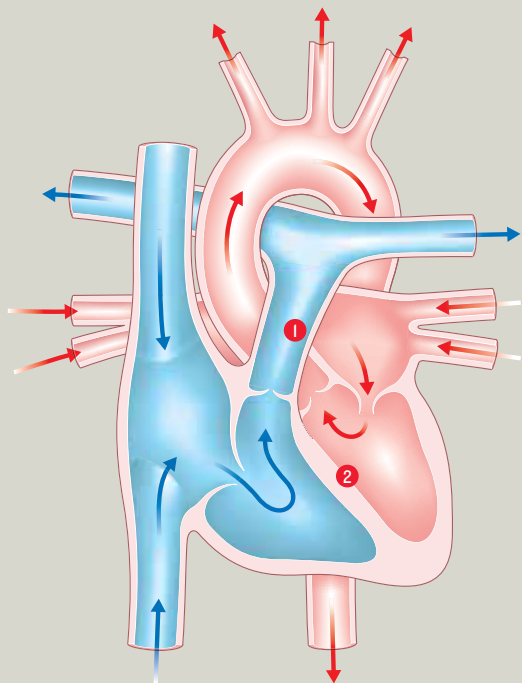
Each day 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research visit bhf.org.uk/research

What causes a congenital heart condition?

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

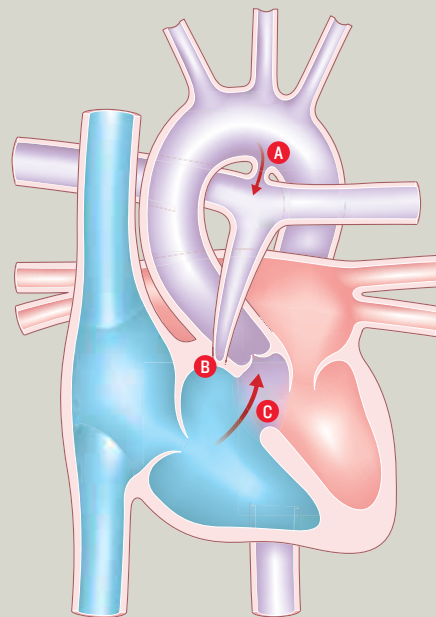
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome is a group of symptoms that commonly appear together as part of a condition).*

Normal heart



- ① Pulmonary artery
- ② Ventricular septum

Pulmonary atresia with a ventricular septal defect



- A The duct
- B Blocked pulmonary valve
- C Ventricular septal defect

What is pulmonary atresia with a ventricular septal defect?

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

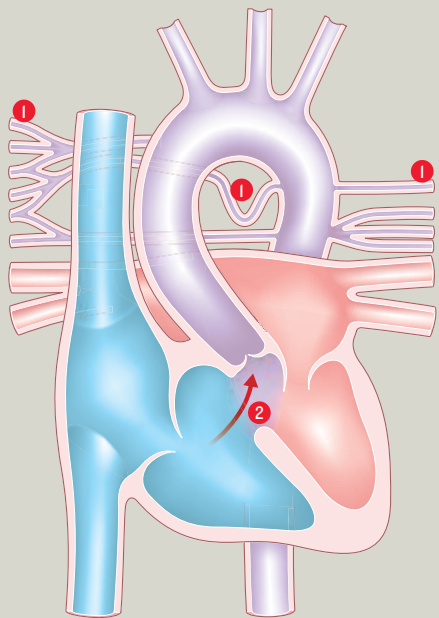
- The valve which allows blood to flow from the heart to the lungs – the **pulmonary** valve – is completely blocked
- There is a large hole (*ventricular septal defect* - *VSD*) between the two main pumping chambers (*ventricles*) of the heart.

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 (*‘the duct’*).

The duct is open while a baby is developing in the womb, but it usually closes shortly after birth. In order for a baby with **pulmonary** atresia with a *VSD* to survive, the duct must stay open after birth. We explain more about the treatment to keep it open later on.

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 on page 8*). This can make surgery much more complicated or regrettably, sometimes impossible.

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



- 1 Collaterals
- 2 Ventricular septal defect

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 need to be treated immediately.



For more information and support about growing up with a heart condition, visit [bhf.org.uk/heart-health/children-and-young-people](https://www.bhf.org.uk/heart-health/children-and-young-people)

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

Some babies will 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.

If your baby has been diagnosed before birth, your doctor will talk you through 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 using an **echocardiogram**. This is an ultrasound scan of the heart and it doesn't hurt your baby at all.

In babies where the pulmonary arteries are small or difficult to see it may be necessary to do additional tests. This can include a CT scan, MRI scan or cardiac catheter. Your doctor will talk these through with you if this is the case for your baby.

How is pulmonary atresia with a ventricular septal defect treated?

The very first stage of treatment is to keep the duct open. It's very important your baby has this treatment to keep the duct open, in order to survive. 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 a baby's natural breathing, so your baby may need the support of a **ventilator**.

Most babies with pulmonary atresia with a VSD will need an operation a few weeks after they're born to treat their condition. This operation is called an arterial shunt, which allows more blood to flow to the lungs. The operation involves placing a small tube between a branch of the **aorta** and the **pulmonary** artery. While this won't make your baby's heart normal, it usually allows your baby time until they're old enough to consider more major surgery. After the shunt operation your child will usually have a scar at the side of their chest.

What are the risks of a shunt operation?

Shunt operations are usually very successful and the fatality risks are low. But all operations are serious and carry a small risk of major complications such as brain damage, kidney damage and pneumonia. It's 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. If the **pulmonary** arteries have grown well, your child may be suitable to have a major repair operation.

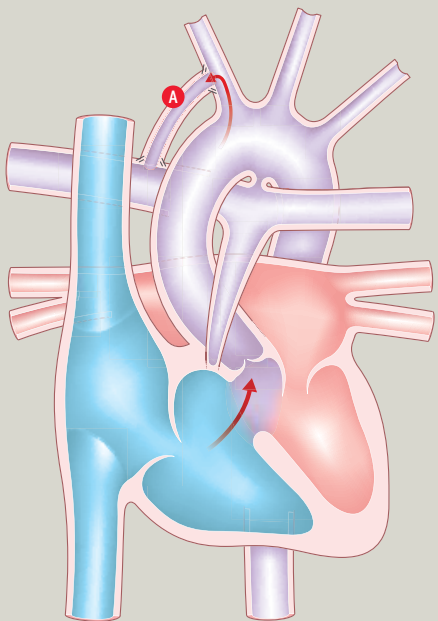
For the further surgery, your child will be given a general anaesthetic. The heart is stopped and the function of the heart and lungs are taken over by a '**heart-lung machine**' which makes sure that blood is still pumped around your child's body.

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 centre of their chest, along the breast bone.

After the operation your child will have a scar in the centre of their 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 will need to be replaced. The timing of this will vary in every case, but will be approximately every 10-15 years.

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 they may die very young.



A Shunt

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.

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.

Even many years after successful surgery new problems may arise, which require treatment: most commonly the development of abnormal heart rhythms. The good news is, while these can be serious, they can usually be treated. So, it's important that your child has regular outpatient reviews with a **cardiologist**, even if they appear well.

For babies with an absent pulmonary arteries with collateral arteries, these will often narrow in later life and need stretching with a balloon catheter.



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

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.

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.

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.

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.

Acknowledgements

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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



**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.

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