Pulmonary atresia with intact ventricular septum
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 intact ventricular septum is and how it is diagnosed
- how pulmonary atresia with intact ventricular septum 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.
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. Right ventricle
3. Atrial septum
4. Pulmonary valve
5. Left atrium

Pulmonary atresia with intact ventricular septum

A. The duct
B. Atrial septal defect
C. Blocked pulmonary valve
D. Oxygen-rich and oxygen-poor blood mix in left atrium
E. Small right ventricle

Understanding your child’s heart
Pulmonary atresia with intact ventricular septum is a very serious type of congenital heart disease. There are two main abnormalities:

- The valve which allows blood to flow from the heart to the lungs – the pulmonary valve – is completely blocked. (See the illustration on page 5).
- The main pumping chamber on the right side – the right ventricle – has often not developed normally.

In some babies, the tricuspid valve – the valve which allows blood to flow into the right ventricle – may also be under-developed. In others there can be abnormalities of the coronary arteries (the arteries which supply the heart muscle with blood). These additional abnormalities can have an impact on long-term heart health.

In the normal heart, blood passes through the pulmonary artery to the lungs to collect oxygen. In babies who have pulmonary atresia with intact ventricular septum, blood cannot pass to the lungs in this way. Instead, the blood passes from the aorta to the pulmonary artery through the connection called the ductus arteriosus – often called ‘the duct’. (See the illustration). The duct is open while the baby is developing in the womb, but it usually closes shortly after birth. In a baby with pulmonary atresia with intact ventricular septum, it is vital that the duct stays open after birth if the baby is to survive. We explain more about the treatment to keep it open on the next page.
What are the symptoms of pulmonary atresia with intact ventricular septum?

The low level of oxygen in your baby’s blood can make them appear blue, especially around the lips, tongue, mouth and hands.

What other conditions are associated with pulmonary atresia with intact ventricular septum?

A small proportion of babies with pulmonary atresia with intact ventricular septum may have other abnormalities, including chromosomal abnormalities. Your child’s cardiologist will discuss this further with you if necessary.

For more information and support about growing up with a heart condition, visit bhf.org.uk/heart-health/children-and-young-people
How is pulmonary atresia with intact ventricular septum diagnosed?

In some cases, the diagnosis is made before birth, but sometimes it’s 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 baby at all.

How is pulmonary atresia with intact ventricular septum treated?

The first stage of treatment is to keep the duct open by giving your baby an injection of drugs called prostaglandins to keep the duct open. These drugs can occasionally interfere with your baby’s natural breathing so they may need the support of a ventilator. Without treatment, the duct would close and your baby would die.

Drug treatment is not usually recommended for very long periods, so your baby may need surgery to treat his or her condition within the first few weeks of life. The type of surgery needed will depend on how well the right ventricle has developed and whether your child has any other abnormalities of the heart. Your paediatric cardiologist will discuss your child’s condition with you.
If the right ventricle has developed enough, the next procedure is likely to be a valvuloplasty. This is a procedure to open the blocked valve. We explain more about it on page 13. Babies with too little blood flowing to the lungs need surgery to correct it – called a shunt operation. A small number of children are not suitable for any type of treatment and their life expectancy is very short. If this is the case for your child, your cardiologist will discuss this with you.

Valvuloplasty

In some babies, it may be possible to open the blocked valve using a procedure called a valvuloplasty. Sometimes this can be done using the catheter valvuloplasty procedure, or it can be done with surgery.

Balloon valvuloplasty

Under general anaesthetic, a catheter (a fine, hollow tube) is inserted into a vein at the top of your baby’s leg. It’s guided up the vein into the right side of the heart and put in position under the blocked valve. Once it’s in position, the tip of the catheter is heated to burn a small hole in the valve. Using X-ray pictures for guidance, a small balloon is put through the hole which has been made in the valve. The balloon is inflated, stretching the valve open. The balloon is then deflated and removed.
After the balloon **valvuloplasty**, your baby will need to stay in hospital for a few days to a few weeks. You will have to take your child to the outpatients department to see the **paediatric cardiologist** for a check-up within a few weeks.

### Surgical valvotomy

In more complicated cases, the blocked **pulmonary** valve cannot be opened using the **catheter** technique described above, and open-heart surgery is needed. This operation, called **surgical valvuloplasty**, involves a general anaesthetic and a **heart-lung machine** to pump blood around the body while the heart is stopped briefly and the valve is cut open. The heart is then re-started. After the operation, your child will have a scar down the middle of the chest, along the breast bone.

### What are the risks of valvuloplasty?

Most children survive the catheter **valvuloplasty**, but the risk varies from one child to another. There is also a small risk of complications, such as bleeding around the heart, or occasionally brain damage.

Surgical **valvuloplasty** is usually carried out in children with more complicated types of heart abnormalities, so the risks vary a great deal from one child to another. Your **cardiologist** will discuss this with you.
In some cases, a valvuloplasty may not be possible or suitable, so a shunt operation may be carried out. This will not correct the heart problem, but it helps as a short-term measure to keep your baby alive until he or she can have further treatment. 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 17)*. The surgery usually leaves a scar at the side of the chest rather than in the middle.

**What are the risks of a shunt operation?**

The risks associated with a shunt operation vary a great deal from one child to another. Most children with pulmonary atresia with intact ventricular septum who have a shunt operation survive the surgery. Your cardiologist will discuss this further with you.
What happens after surgery?

The length of time children need to stay in hospital after a surgical valvuloplasty or a shunt operation 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 21). 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.
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, and 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 24). The illustration on page 24 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 who have pulmonary atresia with intact ventricular septum are usually 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’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.

Total cavopulmonary connection

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

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
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<tbody>
<tr>
<td><strong>Aorta</strong></td>
<td>The main artery of the heart that supplies oxygen-rich blood to the body.</td>
</tr>
<tr>
<td><strong>Atrium</strong></td>
<td>One of the two upper chambers of the heart.</td>
</tr>
<tr>
<td><strong>Cardiologist</strong></td>
<td>A consultant specialising in heart disease.</td>
</tr>
<tr>
<td><strong>Catheter</strong></td>
<td>A fine, hollow tube.</td>
</tr>
<tr>
<td><strong>Congenital</strong></td>
<td>From birth.</td>
</tr>
<tr>
<td><strong>Duct</strong></td>
<td>See ductus arteriosus below.</td>
</tr>
<tr>
<td><strong>Ductus Arteriosus</strong></td>
<td>A natural connection between the aorta and the pulmonary artery. Also called the 'duct'.</td>
</tr>
<tr>
<td><strong>Echocardiogram</strong></td>
<td>An ultrasound scan used to produce pictures of the heart and blood vessels.</td>
</tr>
<tr>
<td><strong>Heart-lung machine</strong></td>
<td>A machine that pumps blood around the body while the heart is stopped during an operation.</td>
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<tr>
<td><strong>Paediatric</strong></td>
<td>To do with paediatrics – the study of children’s diseases.</td>
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<tr>
<td><strong>Pulmonary</strong></td>
<td>To do with the lungs.</td>
</tr>
<tr>
<td><strong>Septum</strong></td>
<td>The wall that keeps the right and left sides of the heart separate.</td>
</tr>
<tr>
<td><strong>Valvuloplasty</strong></td>
<td>A surgical procedure to open a narrowed valve.</td>
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<tr>
<td><strong>Ventilator</strong></td>
<td>A machine that helps your child breathe.</td>
</tr>
<tr>
<td><strong>Ventricular</strong></td>
<td>To do with the ventricle or ventricles. (See above).</td>
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References


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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.

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.

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This is a charity donation service for the BHF. Texts cost £3 + 1 standard rate msg. The BHF will receive 100% of your donation to fund our life saving research.

To opt out of calls and SMS text NOCOMMS BHF to 70060, or if you have any questions about your gift call 02032827862.

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