

# Pulmonary atresia with intact ventricular septum

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## WHAT IS PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM?

**Pulmonary atresia with intact ventricular septum is a congenital heart condition. This means that before you were born there was a problem with the development in the structure of your heart.**

Your pulmonary valve sits between the right bottom chamber (ventricle) of your heart, and the arteries that lead to your lungs. Your valve didn't develop properly, and blocked the flow of blood to your lungs. Sometimes the right ventricle doesn't develop properly too, and can be smaller than it should be. You may also have other heart defects.

### THINGS TO TALK ABOUT

- Physical activity
- Healthy lifestyle
- Pregnancy
- Contraception
- Medicines (including warfarin)

Before you were born your mum gave you all the oxygen you needed. Blood went round the blockages in your heart by way of a hole (foramen ovale), and a tube (ductus arteriosus – the duct), that are always in the heart of a newborn baby. They close a few days after birth, so you had a medicine (prostaglandin) to keep them open until you were well enough to have your first operation.

If your right ventricle was developed enough you may have had valve surgery to make a working pulmonary valve. This may have been repeated as you grew, and will need to be followed up regularly by your doctor. You may need more procedures as you grow older.

If your right ventricle was very small you will have had a shunt operation, so a small tube connects your head and neck arteries to your pulmonary arteries, increasing blood flow to your lungs.

For over 50 years we've pioneered research that's transformed the lives of people living with heart and circulatory conditions. Our work has been central to the discoveries of vital treatments that are changing the fight against heart disease. But so many people still need our help.

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.

Once you'd grown you would have needed more blood flowing to your lungs to pick up more oxygen so you needed a second operation. The cavo-pulmonary connection, sometimes called a Glenn shunt or a hemi-Fontan, connects your main vein (superior vena cava), that carries blood from your head and neck, and attaches it on to lung blood vessels (pulmonary arteries). This means that more blood can pick up more oxygen to be pumped around your body. The small tube (shunt) that was put in at the first operation will be removed.

When you were a bit older, you will have had a Fontan procedure or a TCPC (total cavo-pulmonary connection) operation. This connects the vein (inferior vena cava) carrying oxygen-poor blood, from your body to your pulmonary arteries. The hole in your heart (ASD) will be closed. This is the first time that oxygen-poor and oxygen-rich blood will have been separated and so it will have been the first time that you were pinker.

A few of you may have other medical problems or learning difficulties. There is support to help you cope as you go through life, ask your medical team for advice about the help available to you. You will need regular check-ups in a cardiac clinic throughout your life.

### ENDOCARDITIS

**To reduce your risk of getting endocarditis:**

- Keep your teeth and mouth clean and have regular check-ups with a dentist
- Avoid body piercing and tattooing
- Never inject recreational drugs

Revealing the facts  
about your condition

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FOR EVERY  
HEARTBEAT**

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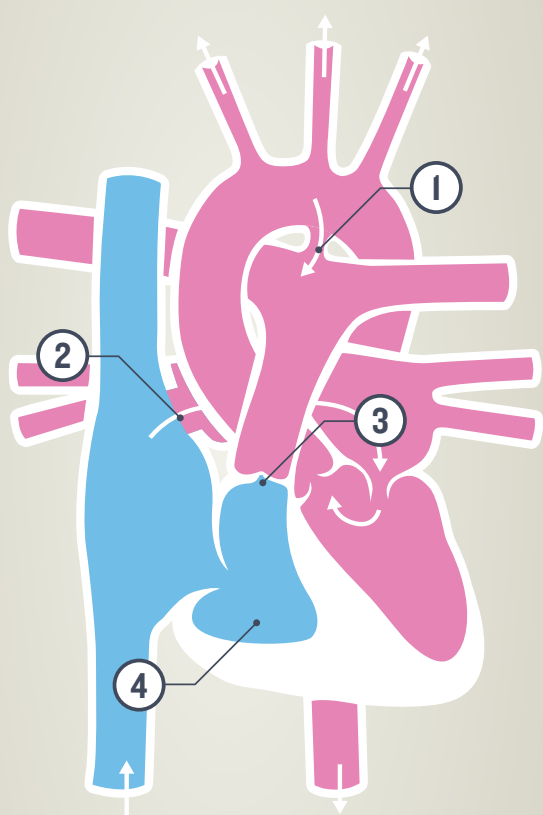
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# YOUR HEART

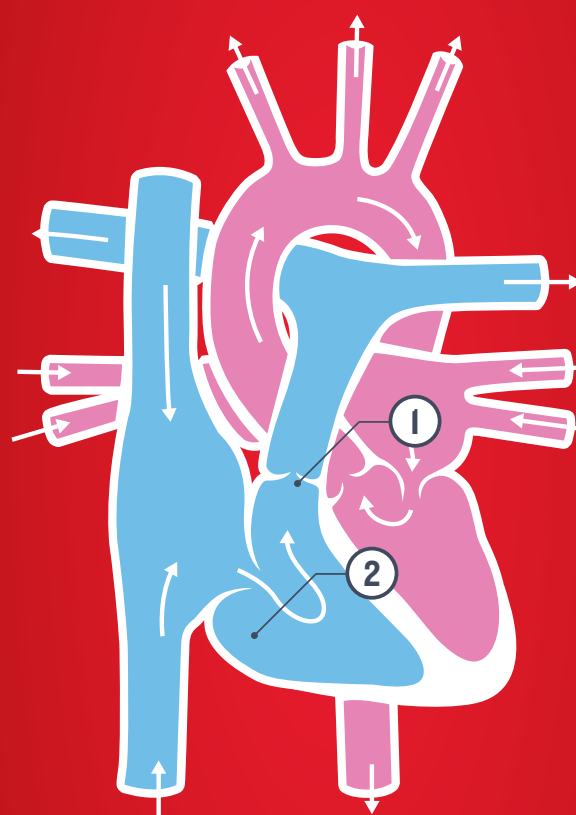
Find out more about your heart:

[yheart.net](http://yheart.net) / [chfed.org.uk](http://chfed.org.uk) / [lhm.org.uk](http://lhm.org.uk)



- ① patent ductus arteriosus /the duct
- ② atrial septal defect (ASD)
- ③ blocked/narrow or unformed pulmonary valve
- ④ small right ventricle.

# THE HEART



- ① pulmonary valve
- ② right ventricle.