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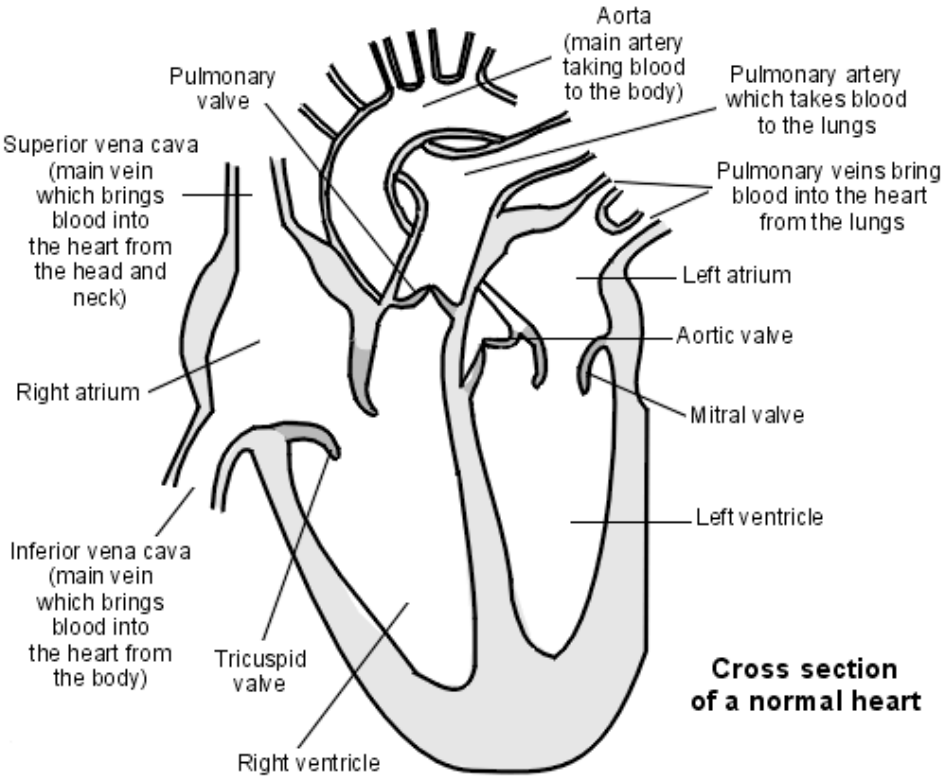
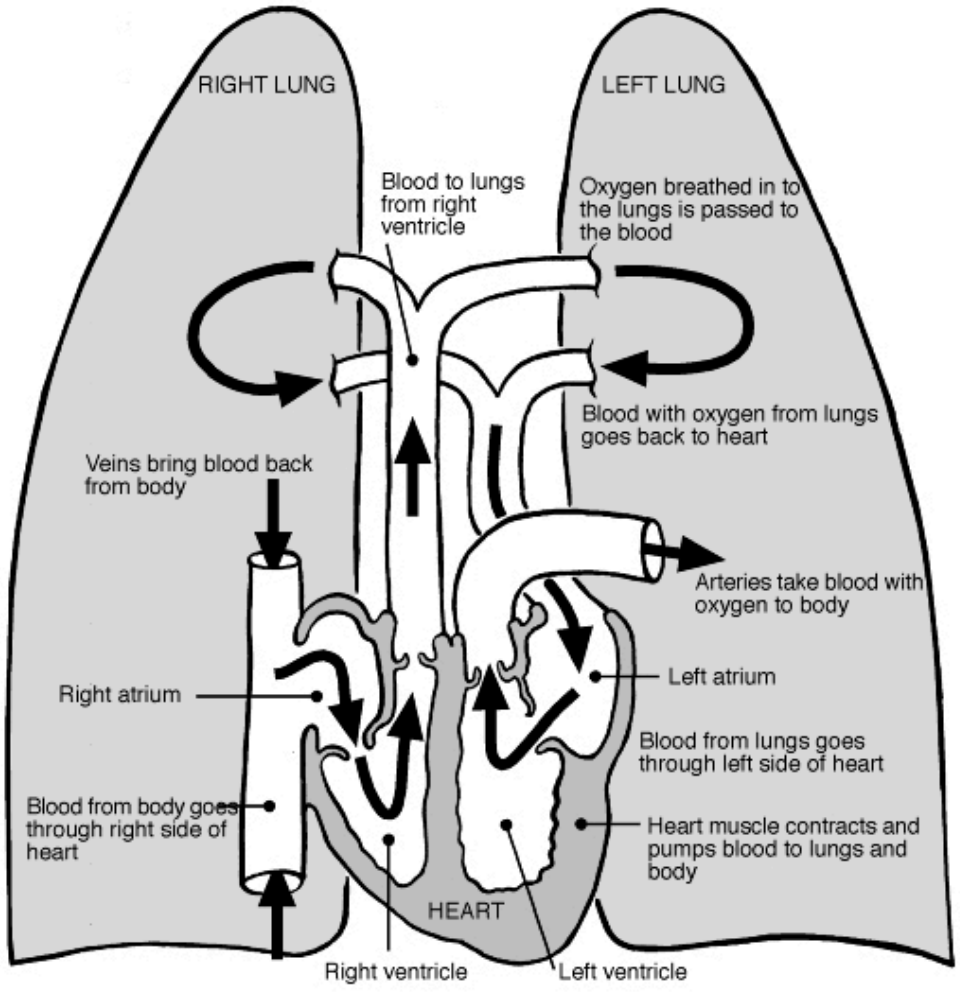
Coarctation of the aorta

Coarctation of the aorta is a type of congenital heart disease where there is a narrowing in part of the aorta, the main artery that leaves the left side of the heart.





Normal blood flow of heart and lungs

In coarctation of the aorta, there is a narrowing in part of the aorta, usually the upper part of the aorta. This is just after where the arteries that supply blood to the head and arms have branched off. Blood cannot pass as easily through the narrowed part of the aorta. This means that the blood supply to the lower half of the body can be affected (less blood is able to get through). Also, because of the narrowing, the heart works harder to pump and try to squeeze blood through the narrowing. Because the heart is working harder to pump, this can cause blood pressure to rise. Often, someone with coarctation of the aorta will have high blood pressure in their upper body and arms (or one arm) and low blood pressure in their lower body and legs. Also, the muscle walls of the left ventricle can increase in thickness (hypertrophy) because of the extra work that they are having to do.

When a baby is growing inside their mother's womb (uterus), the pulmonary artery and the aorta are joined together by a small blood vessel called the ductus arteriosus. It means that most of the blood leaving the right ventricle bypasses the lungs and passes directly into the aorta. Once the baby is born, the ductus arteriosus closes, usually within the first few weeks of life. If a baby has severe coarctation of the aorta, the only way that blood can pass to the lower half of the body is through the ductus arteriosus. So, when the ductus arteriosus closes in these babies, no blood will get through to the lower half of the body, causing severe symptoms. If nothing is done about this, the baby can die.

Congenital heart disease occurs in about 7 in 1,000 pregnancies and coarctation of the aorta is just one type of congenital heart disease. Around 1 in 2,500 babies have coarctation of the aorta. The condition is twice as common in males as it is in females.

Coarctation of the aorta can occur alone, or it can occur with other congenital heart problems. The other congenital heart problems that coarctation of the aorta most commonly occurs with are either:

- A bicuspid aortic valve (the aortic valve is normally made up of three cusps or flaps, a bicuspid aortic valve only has two cusps); **or**
- A ventricular septal defect (a condition where there is a hole in the wall - the septum - that separates the two ventricles).

In some babies, coarctation of the aorta can develop as part of a syndrome (a collection of symptoms) that a baby is born with. For example, coarctation of the aorta can be part of Turner syndrome. (This is a genetic condition that only affects girls. It is a chromosome problem. The most characteristic features of the syndrome are being short, having certain physical features, such as a typical facial appearance, and also having ovaries that do not work properly.)

Symptoms

How much the aorta is narrowed in coarctation of the aorta can be different in different people. Some babies will be born with severe narrowing which can lead to symptoms soon after they are born (see below). However, in others, the narrowing is less severe and symptoms may not be noticed until later. In some people, the narrowing may gradually get worse over time and therefore lead to symptoms. Occasionally, coarctation of the aorta does not cause any symptoms or is not detected until adult life.

Symptoms result from heart failure because the heart is not able to push blood through the narrowed part of the aorta. This causes a back-pressure of blood and congestion of the lungs, leading to symptoms including shortness of breath.

Patients with less severe narrowing will present later as they develop extra blood vessels, called collateral blood vessels, so that some blood is able to bypass the narrowed part of the aorta. These collateral blood vessels may be enough for some time. Symptoms may not become obvious until later in childhood (and sometimes even adulthood). Eventually the heart is no longer able to cope and heart failure develops, with shortness of breath, coughing, feeling tired and swelling of the feet and legs.

More severe narrowing may cause early symptoms, in the first few weeks of life (as the ductus arteriosus closes):

- There may be poor feeding.
- There may be heart failure signs: breathlessness; a rapid breathing rate; swelling of the body.

Are there signs that may be spotted before symptoms develop?

The following may be picked up:

- During a routine check in a baby or child, the healthcare professional may hear a heart murmur when they listen to the child's heart. A heart murmur is an extra sound amongst the heartbeats that the healthcare professional may hear.

- Presence of high blood pressure, with much higher blood pressure in the arms than in the legs.
- Difference in the timing of the pulses between the arms or the arm and legs. The pulses in the legs may be weaker.

What methods are used to diagnose coarctation of the aorta?

Diagnosis of the condition requires various tests and these may include:

- Echocardiogram - this is an ultrasound scan of the heart and will usually show up the narrowing in the aorta. It may also be possible to measure the differences in pressure on either side of the narrowing (called the pressure gradient) and so give an idea of how severe the narrowing is. It will also be able to measure the force with which the heart is contracting and how strong the heart muscle is pumping. If heart failure is present the heart is unable to pump adequately due to heart muscle weakness. This scan may also pick up any other heart defects which may also be congenital.
- Chest X-ray - this may show that the heart is enlarged. It may show fluid on the lungs if there is heart failure. It may also show some 'notching' (or grooves) on the ribs if collateral blood vessels have developed.
- Electrocardiogram (ECG) - this is a test that records the electrical activity of the heart. It can be abnormal in some people with coarctation of the aorta.
- Magnetic resonance imaging (MRI) - scanning of the blood vessel, using magnetic rays. This provides good details of the structure of the heart and the large blood vessels around it.
- Cardiac catheterisation - a catheter is a thin, flexible, hollow tube. Cardiac catheterisation entails a very thin plastic catheter being passed into the chambers of the heart. The catheter can also be passed into the main blood vessels of the heart (the coronary arteries). It is sometimes carried out if someone has coarctation of the aorta. The difference in pressure on either side of the narrowing can be measured to look at how severe the coarctation is.

Treatment options

Treatment to help stabilise symptoms

If a newborn baby has severe coarctation of the aorta, they will first need to have treatment to help control their symptoms. This may include medicines to help treat any heart failure and sometimes they may need to have artificial ventilation to help with their breathing. They may also be given a special medicine that can help to keep the ductus arteriosus open so that blood can pass to the lower part of their body (below the narrowed aorta).

If a child has developed high blood pressure because of coarctation of the aorta, this may need treatment with medicines.

Treatment for the narrowed aorta

The narrowing of the aorta can be repaired using surgery. The surgeon will need to open up the chest so that they can get access to the heart and aorta to operate on them. A clamp is placed across the aorta to stop the flow of blood through it. Then, the narrowed section is cut out and the two normal-sized parts of the aorta are joined back together. Sometimes, if a large part of the aorta has to be removed due to narrowing, a patch (or graft) of special synthetic material is used to fill the gap and repair the aorta.

A newer way of treating coarctation of the aorta is by using balloon angioplasty. This may be used in older children and adults who have been diagnosed with the condition. A balloon catheter (a thin, flexible, hollow tube with a deflated balloon at the tip) is inserted into a large blood vessel, usually an artery in the groin. X-ray guidance is then used and the catheter is passed up through the blood vessels within the body until it reaches the narrowed part of the aorta. The balloon is then inflated within the narrowed section, making the narrowing wider. Sometimes, a small, expandable metal tube (a stent) is then placed in the narrowed segment to keep it open.

Complications

Complications from the surgery

Anyone who has had surgery for coarctation of the aorta will need regular check-ups with a specialist afterwards. Many will have no serious complications due to the surgery and will go on to lead healthy lives afterwards. However, sometimes complications can develop, either at the time of surgery or shortly afterwards. These may include:

- Internal bleeding during the surgery.
- Infection of the wound after the surgery.
- Damage to the kidneys during the surgery.
- Damage to the spinal cord because of the surgery. This is more likely if the operation is more complicated and the aorta needs to be clamped for a longer period of time. (Because the aorta is clamped, it can stop blood getting through to the spinal cord.) If the spinal cord is damaged, this can mean that the child's legs can become permanently paralysed. This complication is rare but, obviously, can be very serious.
- A stroke due to the surgery. However, this is also very rare.

Possible complications following surgery

Some other complications are also possible over a period of time after surgery. They can include:

- Return of the narrowing of the aorta. This is known as recurrent coarctation.
- An aortic aneurysm. This is a widening, or dilatation, of the aorta. An aortic aneurysm can be a complication of coarctation of the aorta that has not been treated with surgery but it is also a rare complication after surgery. The widened part of the aorta is weaker and there is a risk that it can burst (rupture), causing severe internal bleeding.
- Persisting high blood pressure despite the fact that the narrowed part of the aorta has been widened or is no longer present. This may need treatment with medication.

- Infection can develop at the site of the narrowing in coarctation which can lead to blockage of the opening - this is called infective endocarditis. Anyone with coarctation of the aorta (whether they have had surgery or not) has a small risk of getting infective endocarditis. See separate leaflet called Infective Endocarditis for more details.

- About 4 in 100 people who have coarctation of the aorta also have an aneurysm of one of the small blood vessels in their brain, called a berry aneurysm. There is a risk that it may burst. If a berry aneurysm in the brain bursts, this can lead to bleeding in the brain and a subarachnoid hemorrhage.

Pregnancy considerations and precautions

A number of women who have had treatment for coarctation of the aorta go on to have a normal pregnancy with no problems. However, if a woman has coarctation of the aorta, or has had treatment for it, and she is planning to become pregnant, she should discuss this with her heart specialist first. This is because pregnancy can put an extra strain on the heart. It is best to plan a pregnancy when a woman's symptoms and heart condition are stable.

Also, if someone has been born with congenital heart disease, it is more likely that they will have a baby who is also born with congenital heart disease. This risk should be discussed with a heart specialist. It may be possible to arrange for scans of the baby's heart early on in the pregnancy to look for any heart problems.

Further reading

- Balloon angioplasty or stenting for coarctation or recoarctation of aorta; NICE Interventional Procedure Guidance, July 2004
- Vijayalakshmi K, Griffiths A, Hasan A, et al; Late hazards after repair of coarctation of the aorta. BMJ. 2008 Apr 5;336(7647):772-3.
- Ijland MM, Tanke RB; Aortic coarctation. Circulation. 2009 Sep 29;120(13):1294-5.

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