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Q and A: Coarctation of the Aorta

What is coarctation of the aorta?
People with coarctation of the aorta are born with an aorta that is too narrow. The aorta is the longest blood vessel in the body. Blood is pumped from the left ventricle into the aorta. It then branches out into the right arm, head, and left arm. In coarctation the aorta can be narrow in different places and in different ways. Some people have a short narrow section. Others have long areas of their aorta that are too narrow. If the aorta is very narrow, blood will not be able to pass. This kind of coarctation is usually found at birth. If the narrowing is less severe, it may not be detected until later in life.

How common is coarctation?
Coarctation is a common heart defect. About 5–8% of people with heart defects have coarctation. This means there are about 16,000 people in Canada living with coarctation.

How does coarctation develop?
Right now there are several theories about what causes the aorta to become too narrow. Doctors have noticed that coarctation often occurs near a heart structure called the ductus arteriosus. This is an artery that is open before birth and normally closes in the newborn. It is possible that when the ductus closes it pinches the aorta. This may result in a coarctation.

It is also possible that coarctation develops because the blood flow in the growing aorta is not normal. This might be caused by problems in the heart itself. Another theory is that people with coarctation have an in-born problem in the cells that make up the walls of their aorta.

What causes coarctation?
We do not know enough about why coarctation happens. In 10–20% of cases there is a known family history and/or genetic factor, such as Turner's Syndrome. Scientists suspect that as we continue to study coarctation new genetic factors may emerge. Coarctation is not known to be caused by environmental factors during pregnancy.

If I have coarctation, do my relatives have a higher risk for heart problems?
If you have coarctation, your parents, sisters, and brothers are more likely to have certain kinds of heart defects. These can include bicuspid aortic valve, abnormal mitral valve, and coarctation. Your family members may want to get tested for heart defects. This usually involves an echocardiogram, which is an ultrasound picture of the heart.

What other defects are common in coarctation?
50-85% of coarctation patients also have a bicuspid aortic valve. This means the aortic valve has two flaps instead of three. Some people with COA are also born with a hole in the lower wall of the heart (ventricular septal defect). A smaller number have problems with other heart valves such as the mitral valve. People with coarctation are also at risk from swelling in their blood vessels (aneurysms). The causes and risks of aneurysms are discussed below.

How is coarctation diagnosed?
When coarctation is severe it is usually diagnosed at birth. These babies may have heart failure or go into shock. They are often very ill. Surgery is needed right away to repair the coarctation.

In undiagnosed adults a heart murmur or high blood pressure might lead to a diagnosis. One common clue is a difference in the blood pressure between the arms and legs. This can indicate that there is a narrowing in the aorta. The diagnosis can be confirmed by using an echocardiogram, CAT scan, MRI, or heart catheterization.

What is the treatment for newly-diagnosed coarctation?
There are now different options available to widen the aorta. In most centers the narrow section of the aorta is removed surgically. The two ends of the aorta are then re-attached. In the past coarctation was also treated by widening the narrow section with a patch. However, this procedure is no longer common because of higher rates of long-term complications.

Another, less-common option is to open up the aorta with a catheter. A catheter is a long, thin tube that the doctor guides into the heart. An attached "balloon" can be used to widen the aorta. Sometime the doctor will also place a mesh tube (stent) in the aorta to help keep it open. Over time, all repaired coarctation patients are at risk of having their aortas re-narrow. This risk seems to be higher in patients who have been treated via catheter.

If you are an adult who needs coarctation repair or re-repair, it is very important that you seek treatment from specialists in adult congenital heart disease (ACHD). You and your ACHD doctors can discuss risks and benefits of surgery and other treatments.

What are common long-term complications in repaired coarctation?
All repaired coarctation patients are at risk of developing the following problems:

Narrowing of the Aorta
In some patients the aorta re-narrows. This can occur in those who had surgery as well as catheter treatment. In others, the original repair may have left the aorta too narrow. Some patients will need additional treatment to re-open their aorta.

High Blood Pressure
Many people with coarctation continue to have high blood pressure. Some people may have normal pressure at rest but get high blood pressure when they exercise. Some patients will need to be treated to keep their blood pressure low.

Swelling in the Aorta (Aneurysms)
Some patients develop swollen areas in their aorta (aneurysms). If these grow too quickly and/or get too big they can burst. This can be a fatal event. When diagnosed early, aneurysms can be treated to prevent this from occurring. It is important that coarctation patients have regular testing to check the size of their aorta.

Brain Aneurysm/Stroke
Some people with coarctation also have abnormalities in the brain's blood vessels which can also swell and burst. Coarctation patients should know how to identify signs of stroke and should report any worrisome symptoms to their doctor promptly. New methods exist to diagnose and treat these problems before they cause damage. Good control of high blood pressure may also help prevent this problem.

Valve Problems
As discussed above, the majority of coarctation patients have a bicuspid valve. This valve can develop leaking over time. It can also become too narrow. It is important that this valve's function continue to be checked regularly.

Coronary Artery Disease
Some patients with repaired coarctation develop coronary artery disease earlier than expected. This may be due to the high blood pressure that many patients experience. It is important that coarctation patients get regular screening for acquired heart disease. As in all CHD patients, heart healthy habits are advised.

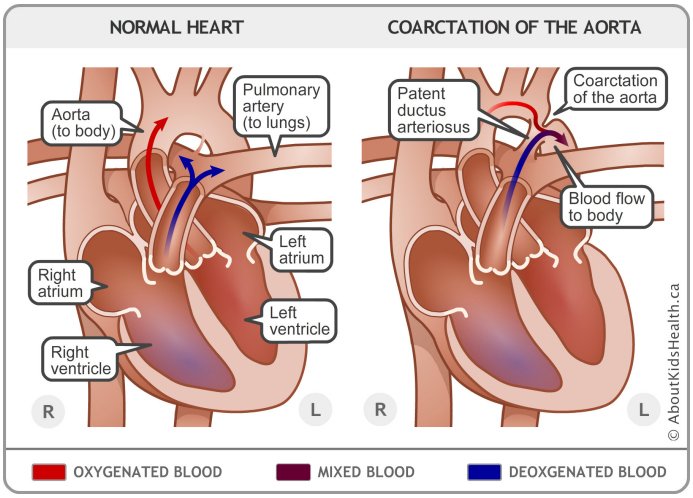
Infection
Some people with coarctation are at risk of heart infection (endocarditis). They may also be prone to infection in the aorta (endarteritis). This includes patients with a history of heart infection and patients who have recently received a stent or graft. It is important that patients with coarctation protect themselves from infection as directed by their cardiac team.

Can repaired coarctation patients exercise normally?
Most patients with repaired coarctation can exercise normally. Some repaired coarctation patients should avoid heavy weight lifting. Your ACHD doctor can tell you more about what exercise is right for you.

Can women with coarctation have children?
Coarctation should be repaired before you become pregnant. Women with unrepaired coarctation are most at risk for problems during pregnancy.

Most women with repaired coarctation can have successful pregnancies. But it is very important that all women with coarctation see an ACHD specialist before getting pregnant. The ACHD specialist should order a detailed image of your entire aorta. It is important to make sure that you do not have any new narrowing or areas of swelling. Pregnancy is a bigger risk if your aorta is still narrow or an aneurysm is forming. You may need additional treatment to make pregnancy safer for you.

About 15–20% of women with coarctation have high blood pressure during pregnancy. All pregnant women with coarctation should have their blood pressure checked regularly. A small number (10%) have miscarriages. If you have coarctation, your risk of having a child with a heart defect is considered to be between 3–10%.



What kind of cardiology care is recommended for adults with repaired coarctation?
We used to think that coarctation was a simple defect. But we now realize that it is much more than a narrowed aorta. People with coarctation may have an in-born abnormality in the cells throughout all their arteries. This would explain why so many patients experience new narrowing and other problems after coarctation is repaired.

It is important that coronary artery disease be found and treated. This means you may need treatment for high cholesterol. It also means that controlling your weight is especially important. If you smoke, try to get the help you need to stop.

Coarctation of the aorta is a lifelong disorder. But the good news is that most people with coarctation continue to do well. There are now many treatments available to prevent and treat the complications described above. By getting the ACHD care you need, and taking good care of your heart health, you can help your "funny plumbing" continue to thrive.

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