CONDITION

Vascular ring

A vascular ring is an arrangement of major blood vessels that presses around the windpipe or food pipe.

WHAT IT IS
Congenital blood vessels encircling the windpipe and food pipe

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Noisy breathing, cough, swallowing difficulty, repeated chest infections

WHAT CAUSES IT
Aortic arch development before birth

HOW IT IS DIAGNOSED
CT angiography or MRI showing the vessel anatomy

MAIN TREATMENT
Surgical release for significant symptoms or compression

CAN IT BE CURED?
Yes - surgery can release the ring

IS IT AN EMERGENCY?
Sometimes - severe breathing trouble or blue lips

LONG-TERM OUTLOOK
Usually good after repair; airway symptoms may linger

Overview

A vascular ring is a congenital condition, meaning it develops before birth. It affects the aorta and nearby blood vessels. The aorta is the large artery that carries blood from the heart to the body.

In a vascular ring, these vessels form a complete or partial circle around the trachea, which is the windpipe, and the oesophagus, which carries food to the stomach. The ring can press on either structure.

Some vascular rings cause breathing or feeding problems in infancy. Others cause mild symptoms that are not recognised until later childhood or adulthood. A person may occasionally have no symptoms, with the condition found during imaging for another reason.

A vascular ring usually does not stop blood from reaching the body. It matters mainly because pressure on the windpipe can restrict airflow, while pressure on the oesophagus can make swallowing difficult.

Symptoms

Symptoms depend on which structures are compressed and how easily the windpipe collapses under pressure.

Breathing symptoms often begin during infancy or early childhood. They may include:

Symptoms may become more noticeable during feeding, crying, exercise, or a respiratory infection.

Pressure on the oesophagus may cause slow feeding, gagging, vomiting, or difficulty swallowing. Older children and adults may notice that solid food seems to stick. Some people take very small bites, chew for a long time, or drink frequently while eating without realising they are adapting to the problem.

Infants may gain weight slowly if breathing and feeding require extra effort. Symptoms do not show how severe the compression is in every case, so imaging is needed to understand the anatomy.

- Noisy, high-pitched breathing called stridor
- A harsh or barking cough
- Wheezing that does not improve as expected with usual asthma treatment
- Rapid breathing or extra effort when breathing
- Repeated bronchitis, pneumonia, or other chest infections

Causes and risk factors

A vascular ring forms while the major arteries are developing early in pregnancy. Several temporary blood vessel arches normally appear and then reshape. If they develop or disappear in a different pattern, vessels or fibrous tissue can remain around the windpipe and oesophagus.

The two main complete rings are:

There are also partial rings and related blood vessel patterns that can cause similar compression. A pulmonary artery sling is often discussed alongside vascular rings, although it

involves a lung artery passing between the windpipe and oesophagus.

Most vascular rings occur without a known action or exposure causing them. They are not caused by diet, exercise, or something a parent did during pregnancy.

Some people have another congenital heart condition. Certain aortic arch patterns are also associated with chromosome changes, including a deletion known as 22q11.2 deletion. This does not mean everyone with a vascular ring has a genetic condition. Genetic assessment may be discussed when the vessel pattern, other health findings, or family history suggests it could be useful.

- A double aortic arch, in which two arches pass around the windpipe and oesophagus
- A right aortic arch with an unusual left subclavian artery and a band of tissue called the ligamentum arteriosum completing the ring

Diagnosis

A healthcare professional may suspect a vascular ring from breathing sounds, swallowing problems, repeated chest infections, or symptoms that have not responded as expected to other treatment. An examination alone cannot confirm it.

An echocardiogram uses sound waves to examine the heart and the start of the major arteries. It can identify associated heart conditions, but it may not show the full ring or its effect on the windpipe.

Detailed blood vessel imaging confirms the anatomy:

The choice between CT and MRI is based on age, available expertise, the detail needed, and whether remaining still for the scan is possible.

A contrast swallow, sometimes called a barium swallow, uses X-rays while contrast liquid passes through the oesophagus. It may show where the oesophagus is indented, but it does not define every blood vessel.

Bronchoscopy involves passing a small camera into the airway under anaesthesia. It can show where the windpipe is compressed and whether its walls are unusually soft. This softness is called tracheomalacia. Bronchoscopy is especially useful when symptoms are severe, the diagnosis is uncertain, or airway treatment may be needed.

A vascular ring may be suspected during a pregnancy ultrasound. Imaging after birth is generally needed to confirm the exact arrangement and assess compression.

- CT angiography uses X-rays and contrast dye to create detailed images of the blood vessels, windpipe, and oesophagus
- MRI uses a magnetic field to show the vessels without ionising radiation, although the scan takes longer and some children need sedation

Treatment and management

Treatment is based on the vessel anatomy, symptoms, airway or oesophageal compression, and any other heart or airway condition.

A person without symptoms may be monitored rather than treated immediately. Follow-up can check for new breathing, feeding, or swallowing problems. Medicines cannot remove the ring, although they may treat a separate infection, reflux, or another condition contributing to symptoms.

Surgery is the main treatment when a vascular ring is causing important breathing or swallowing problems. The operation releases the ring by dividing the vessel or fibrous band that completes it. The surgeon may also address an enlarged vessel segment or reposition a blood vessel when necessary.

Many repairs are performed through an incision on the side of the chest. The exact operation depends on the anatomy, and some complex patterns require a different approach. Blood usually continues to reach the body normally after the ring is released.

Some people also need treatment for a soft or flattened windpipe. In severe cases, an operation may support the airway by attaching it to nearby tissue so it remains more open. Feeding support may be used when swallowing has been difficult or weight gain has been affected.

After surgery, pain control, breathing exercises, and gradual return to eating are common parts of recovery. Follow-up may involve cardiology, surgery, respiratory medicine, ear, nose and throat care, or feeding specialists.

Surgery corrects the ring, but breathing sounds or cough may not disappear immediately. A windpipe that was compressed for a long time can remain soft while it recovers.

Complications

Ongoing airway compression can contribute to repeated infections, difficulty clearing mucus, or episodes of significant breathing difficulty. Oesophageal compression can lead to choking,

poor food intake, or food entering the airway, which is called aspiration.

Long-term pressure may contribute to tracheomalacia. This can cause a persistent barking cough, noisy breathing, or airway collapse during illness or forceful breathing.

Possible surgical complications include bleeding, infection, fluid leaking from lymph vessels in the chest, or injury to nerves that help control the voice box or diaphragm. Residual compression can remain if the anatomy is complex. The surgical team can explain risks that apply to the planned procedure.

Prevention

A vascular ring cannot usually be prevented. It forms during early development before birth, often for no identifiable reason.

There is no evidence that routine food, activity, or behaviour during pregnancy causes a vascular ring. If a chromosome change or another congenital condition is identified, genetic counselling can explain what it may mean for the person and their family. It cannot change a ring that has already formed.

Living with it and what to expect

Before treatment, symptoms may stay stable or become more noticeable as feeding changes, activity increases, or respiratory infections occur. A young child may have more trouble with solid foods than with liquids. Adults may have years of unexplained swallowing difficulty, cough, or repeated chest symptoms before diagnosis.

After repair, swallowing and breathing often improve, but recovery is not identical for everyone. Pressure on the oesophagus may ease soon after surgery. Airway symptoms can take weeks or months to settle, particularly when tracheomalacia is present.

Some people continue to have a noisy cough or breathing problems during colds even though the ring has been released. Follow-up helps distinguish expected airway recovery from persistent compression, scarring, or another condition.

Once recovery is complete, many people can eat, exercise, attend school, and work without special limits. Restrictions may be needed temporarily after surgery or because of an associated heart or airway condition.

Routine follow-up may include checking breathing, swallowing, growth, and recovery from surgery. Repeat imaging or airway tests are used when symptoms continue or return, rather

than being necessary in the same way for everyone.

When to get medical help

Call emergency services if you or your child has:

These symptoms need emergency assessment, whether or not a vascular ring has already been diagnosed.

Seek prompt medical advice for increasing noisy breathing, new difficulty swallowing, repeated vomiting with feeds, signs of dehydration, or a chest infection with worsening breathing. After surgery, prompt assessment is also appropriate for breathing that is getting worse, fever with an unwell appearance, or redness, swelling, drainage, or opening of the wound.

Arrange a routine appointment for ongoing noisy breathing, a long-lasting barking cough, repeated chest infections, food sticking during swallowing, slow feeding, or poor weight gain. These symptoms have several possible causes and do not confirm a vascular ring, but they deserve assessment.

- Severe difficulty breathing or is struggling to draw breath
- Blue, grey, or unusually pale lips or skin
- Pauses in breathing, collapse, or reduced responsiveness
- Choking that does not clear
- An inability to swallow saliva with breathing distress

Common questions

Is a vascular ring a heart defect?

It is a congenital blood vessel condition involving arteries close to the heart. The heart itself may be structurally normal. Some people have a vascular ring together with another congenital heart condition.

Can a child outgrow a vascular ring?

The blood vessels do not move into a normal arrangement as a child grows. Symptoms can change, but apparent improvement does not prove that compression has resolved. Monitoring or treatment decisions are based on the anatomy and its effects.

Does vascular ring surgery fix the problem?

Surgery can release the ring and remove its direct pressure. Many people improve, but symptoms caused by a soft windpipe may continue during recovery. Complex anatomy or another airway condition can also affect the result.

Can adults have a vascular ring?

Yes. A mild ring may not be recognised in childhood. Adults may be assessed after years of difficulty swallowing solid food, unexplained cough, noisy breathing, or chest infections.

Can a vascular ring come back after surgery?

A divided ring does not usually grow back. Symptoms can persist or return because of airway softness, scar tissue, remaining vessel compression, or another breathing or swallowing condition. New or returning symptoms should be reviewed.

Is a vascular ring inherited?

Most cases are not clearly inherited. Some occur with chromosome changes or other congenital conditions. Genetic counselling may be offered when other findings or family history raise that possibility.

Sources

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