CONDITION

Double aortic arch

Double aortic arch is a birth difference in which the aorta forms a ring around the windpipe and food pipe.

WHAT IT IS
A complete blood-vessel ring around the windpipe and food pipe

BODY SYSTEM
Heart, circulation, breathing, and swallowing

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

WHAT CAUSES IT
Altered aortic development before birth

HOW IT IS DIAGNOSED
CT angiography or magnetic resonance angiography

MAIN TREATMENT
Surgery to release the vascular ring

CAN IT BE CURED?
Sometimes - surgery removes compression, but airway symptoms may persist

IS IT AN EMERGENCY?
Sometimes, with severe breathing difficulty or blue lips

LONG-TERM OUTLOOK
Usually good after repair, though breathing recovery may be gradual

Overview

Double aortic arch is a congenital condition, meaning it develops before birth. The aorta is the large artery carrying blood from the heart to the body. Normally, part of it forms one arch over the heart.

In double aortic arch, the aorta divides into two arches. One passes on each side of the trachea, or windpipe, and the oesophagus, or food pipe. They join again behind these structures, creating a complete vascular ring .

The ring can press on the windpipe, food pipe, or both, causing noisy or difficult breathing, cough, feeding problems, or difficulty swallowing. Symptoms often begin in infancy, but mild compression may not be recognised until later in childhood or adulthood.

One arch is usually larger and carries most of the blood. The other may be smaller or partly closed but still complete the ring. Double aortic arch does not usually affect how the heart pumps, although another heart or major blood-vessel difference may also be present.

Symptoms

Symptoms result mainly from pressure on the windpipe or food pipe. Their severity and timing vary.

Breathing symptoms often appear during the first months of life and may include:

Pressure on the food pipe can cause coughing, choking, arching, or tiredness during feeds. Feeding may take a long time, and some children gain weight slowly because eating requires extra effort.

Older children and adults may feel that solid food, particularly bread or meat, sticks. This is called dysphagia , meaning difficulty swallowing. Some people compensate by eating slowly, chewing carefully, or avoiding certain foods.

Symptoms can be mistaken for asthma, reflux, croup, or repeated infections. A vascular ring may be considered when symptoms are persistent, unusual, or do not improve as expected. Some people have few or no symptoms and are diagnosed after imaging for another reason.

- Noisy breathing, including a harsh sound called stridor
- Wheezing that does not improve as expected with usual treatment
- A persistent or seal-like, barking cough
- Fast or difficult breathing
- Repeated bronchitis, pneumonia, or other chest infections
- Noisier breathing during feeding, crying, or respiratory infections

Causes and risk factors

Double aortic arch develops early in pregnancy while the aorta and its branches are forming. Several temporary arches normally appear during development, and some disappear as the usual single arch forms. In double aortic arch, sections that would normally disappear remain and create a ring.

Nothing a pregnant person did or did not do is known to cause this change. It is not caused by how a baby is fed, held, or delivered.

Most cases have no clear inherited cause. The condition can occasionally occur with another congenital heart, blood-vessel, chromosome, or genetic difference. Genetics assessment may be offered when there are other congenital differences, developmental concerns, or a relevant family history.

Diagnosis

Diagnosis begins with the pattern of breathing, feeding, or swallowing symptoms. A clinician may hear noisy airflow, but an examination cannot confirm the condition.

An echocardiogram uses sound waves to examine the heart and nearby blood vessels. It can identify other heart differences and may suggest an abnormal aortic arch, but it may not show the complete ring or the degree of airway compression.

CT angiography is commonly used to define the anatomy. It combines computed tomography with contrast material given into a vein. Images show both arches, their branches, and their relationship to the windpipe and food pipe. CT is quick but uses radiation.

Magnetic resonance angiography, or MRA, produces detailed images without radiation. It takes longer, so younger children may need sedation or a general anaesthetic to remain still.

Other tests may include:

Aspiration means food, drink, or saliva entering the airway. These tests assess the ring's effects, but CT angiography or MRA is usually needed to confirm the blood-vessel anatomy.

- A barium swallow to show an indentation on the food pipe
- Bronchoscopy to examine the windpipe with a small camera
- Lung function tests for older children or adults
- Swallowing studies to assess coughing, choking, or aspiration during meals

Treatment and management

Treatment depends on symptoms, the degree of compression, and the anatomy shown on imaging. Care may involve congenital heart specialists, surgeons, respiratory or ear, nose and throat specialists, and feeding professionals.

Surgery is the main treatment when the ring causes significant breathing or swallowing problems. The surgeon usually divides the smaller or less important arch and any fibrous tissue completing the ring. The larger arch continues carrying blood from the heart. The exact

operation depends on the person's anatomy.

Symptoms do not always disappear immediately after surgery. Long-term pressure can make the windpipe softer than usual, a condition called tracheomalacia . The airway may partly narrow during breathing, coughing, or illness. It often becomes firmer with growth, although some people need further assessment or treatment.

Supportive care may include feeding therapy, altered food textures, treatment of respiratory infections, or help clearing mucus. These measures manage symptoms but do not remove the ring.

People without symptoms may sometimes be monitored rather than treated immediately. Follow-up depends on the anatomy and risk of future compression.

Complications

Ongoing windpipe compression can cause retained mucus, repeated chest infections, or persistent noisy breathing. Severe compression may interfere with breathing, particularly during respiratory illnesses.

Food-pipe pressure may lead to choking, food avoidance, slow feeding, or poor growth. Aspiration can irritate the lungs and increase infection risk.

Possible surgical complications include bleeding, infection, fluid in the chest, or injury to nearby structures. Uncommon injury to a nerve controlling a vocal cord may cause a weak cry, hoarse voice, or swallowing difficulty.

Some people continue to have cough, wheeze, noisy breathing, or swallowing symptoms after repair. Possible causes include tracheomalacia, airway irritation, scarring, reflux, or another breathing condition. Persistent or returning symptoms may require repeat imaging, bronchoscopy, or another procedure.

Prevention

Double aortic arch cannot currently be prevented. It forms during early development before birth, and no action is known to reliably cause or prevent it.

If it occurs with a genetic condition or several congenital differences, genetic counselling may help a family understand test results and the chance of a similar condition in another pregnancy. Early recognition cannot prevent the anatomy, but it can reduce the time spent with untreated airway or swallowing compression.

Living with it and what to expect

Many children recover well after surgical release of the ring. Feeding may become easier, and repeated respiratory problems may decrease. Breathing often improves gradually, especially if the windpipe was compressed for a long time.

During the first months after surgery, cough or noisy breathing may become more noticeable during colds. Follow-up helps determine whether this reflects expected recovery or needs further investigation.

Children with tracheomalacia may need respiratory follow-up, airway-clearance support, or feeding help. The airway often becomes more stable with growth, but recovery differs between children.

Once recovered, many people can take part in school, play, exercise, and other usual activities. Double aortic arch alone does not usually require lifelong activity limits. Restrictions are more likely to relate to recent surgery, another heart condition, or continuing airway symptoms.

Routine follow-up may assess breathing, swallowing, growth, and surgical recovery. New or returning symptoms should be discussed with the care team.

Adults diagnosed later may have had unexplained swallowing or breathing symptoms for years. Treatment can still relieve compression, although recovery may be slower after long-term pressure.

When to get medical help

Arrange a routine medical assessment for persistent noisy breathing, repeated chest infections, coughing during meals, slow feeding, poor weight gain, or a feeling that solid food sticks. These symptoms have several possible causes and do not by themselves confirm a vascular ring.

Contact the usual care team promptly if known symptoms become more frequent, feeding becomes harder, or breathing does not return to its usual pattern after an infection.

Seek urgent medical help for:

Call emergency services for severe breathing difficulty, blue or grey lips or skin, pauses in breathing, collapse, or inability to speak, cry, or feed because of breathlessness.

- Increasing effort to breathe, including pulling in around the ribs or neck
- Trouble swallowing liquids or saliva

- Repeated choking or coughing without recovering normally
- Unusual sleepiness or difficulty staying awake
- Signs of dehydration, such as very little urine

Common questions

Is double aortic arch a heart defect?

It is a congenital difference involving the aorta, the main artery leaving the heart. It usually affects breathing and swallowing more than the heart's pumping function. Some people also have another congenital heart difference.

Does double aortic arch always need surgery?

No. Surgery is the main treatment when the ring causes important airway or swallowing compression. Someone with little or no compression may sometimes be monitored by a specialist team.

Will surgery stop the noisy breathing?

Surgery removes the ring's pressure, but noisy breathing may continue while the windpipe recovers. Tracheomalacia can make improvement take months or longer. Persistent or worsening noise needs follow-up.

Can double aortic arch be mistaken for asthma?

Yes. Wheeze, cough, and repeated breathing problems may resemble asthma. Symptoms caused by fixed windpipe pressure may not improve as expected with asthma treatment. Imaging is needed to identify a vascular ring.

Can an adult have double aortic arch?

Yes, although it is usually recognised in childhood. Adults may have swallowing difficulty, repeated respiratory symptoms, or an unexpected finding on a scan. Congenital heart and chest specialists can assess whether the ring is causing symptoms.

Can double aortic arch come back after surgery?

The divided arch does not usually reform into the original ring. Symptoms can persist or return because of a soft airway, scar tissue, remaining compression, reflux, or another breathing or swallowing problem. New symptoms should be assessed.

Sources

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