

This family education sheet explains what a vascular ring is and how it may be treated.

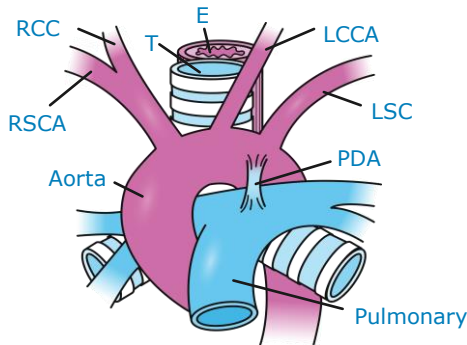
Key points

- A vascular ring is an abnormality in the formation of the body's main artery called the aorta.
- Some children with vascular rings never develop symptoms.
- Not all children with vascular rings need surgery.

What is a vascular ring?

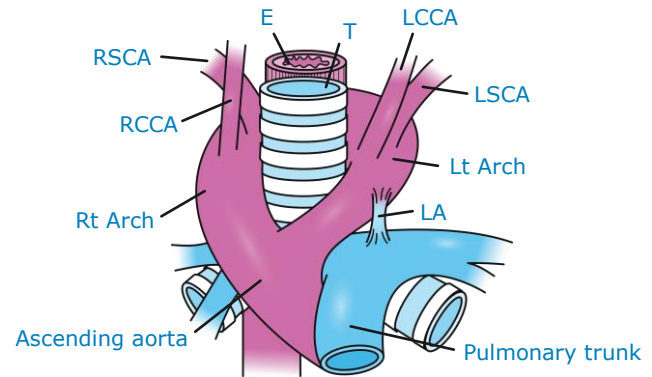
- A vascular ring is a rare abnormality of the aorta – the body's main artery that carries blood out of the heart.
- A vascular ring causes the aorta or its branches to form a "ring" around the trachea (breathing tube) and/or esophagus (swallowing tube).
- Sometimes the vascular ring only partly surrounds the trachea and esophagus (incomplete ring). Other times, it forms a full circle or "napkin ring" around these structures (complete ring).
- Incomplete and complete rings can compress (put pressure on) the trachea (breathing tube) and esophagus (swallowing tube), causing breathing and/or swallowing problems.

Normal aortic arch anatomy – The aortic arch is on the left side, the left subclavian artery and the left common carotid artery are connected to the aorta, and the brachiocephalic trunk is connected to the aorta. The brachiocephalic trunk then branches to the right subclavian artery and the right common carotid artery.

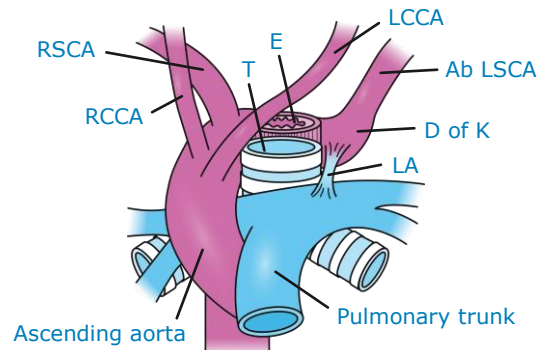


What are the types of vascular rings?

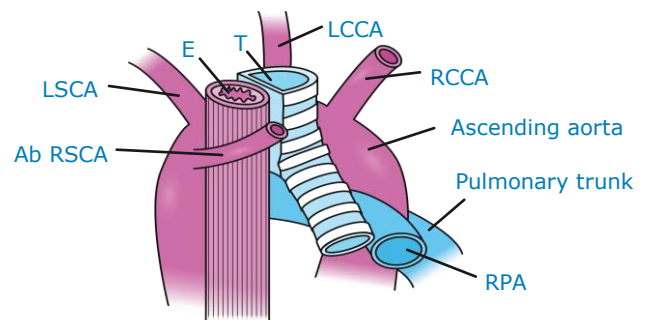
Double aortic arch (complete ring) – The aortic arch is 2-sided instead of 1-sided. The aortic arch is made up of both a left aortic arch and a right aortic arch.



Right aortic arch with an aberrant left subclavian artery with diverticulum of Kommerell (complete ring) – The aortic arch is on the right side, and the left subclavian artery is connected by the diverticulum of Kommerell (outpouching attached to aberrant left subclavian artery) to the aorta. A ligamentum completes the ring.



Left aortic arch with an aberrant right subclavian artery (incomplete ring) – The aortic arch is left-sided, and the aberrant right subclavian artery typically passes behind the airway (breathing tube) and esophagus (swallowing tube). Aberrant means the subclavian artery is located in an atypical position.



Key

Rt arch - Right aortic arch	LCCA - Left common carotid artery
Lt arch - Left aortic arch	RCCA - Right common carotid artery
RSCA - Right subclavian artery	LA - Ligamentum arteriosum
LSCA - Left subclavian artery	D of K - Diverticulum of Kommerell
Ab LSCA - Aberrant left subclavian artery	E - Esophagus
Ab RSCA - Aberrant right subclavian artery	T - Trachea
	RPA - Right pulmonary artery

What causes a vascular ring?

- A vascular ring is formed during a baby's development in the womb.
- The aorta develops from vessels that are formed very early in pregnancy.
- A vascular ring occurs when these vessels form abnormally.

Is a vascular ring associated with a genetic diagnosis?

- The right aortic arch with an aberrant left subclavian artery can be associated with DiGeorge Syndrome, 22q11.2 deletion syndrome.
- The left aortic arch with aberrant right subclavian artery and double aortic arch are less likely to be associated with DiGeorge Syndrome.
- Genetic testing options can be discussed during pregnancy.

Will my baby need treatment before birth?

Treatment isn't needed before your baby's birth.

What should we plan at time of delivery?

- An echocardiogram is sometimes done before a baby leaves the hospital.
- Other testing isn't typically needed right away.

Will my baby need treatment after birth?

- Not all babies with vascular rings need surgery.
- Symptoms typically guide the timing of surgery, if needed.
- Some people may need surgery in childhood. This decision is made based on the type of vascular ring, how serious the symptoms are and the results of certain medical tests.
- We recommend that your child be closely followed by a pediatrician and cardiologist. Your child may also be referred to other specialists based on their symptoms.
- Children with complete rings (double aortic arch, right aortic arch with aberrant left subclavian artery) may need surgery before they're 1 year old.
- Many children with a left aortic arch and aberrant right subclavian artery don't develop symptoms or need surgery. Surgery is typically recommended for children who develop moderate to severe symptoms.

What are the symptoms of a vascular ring?

- Some children with vascular rings never develop symptoms.
- Others develop symptoms as an infant, child or adult.
- Swallowing symptoms can include gagging, choking or coughing when eating or drinking, problems with food getting stuck when swallowing, pain when swallowing or aspiration (liquids going into the breathing tube when drinking).
- Some children eat differently to avoid these symptoms. Some children benefit from taking their time chewing food, eating food cut up into small pieces or taking frequent sips of liquid between bites of food.
- Respiratory symptoms include a chronic croupy or "barking" cough, noisy/junky breathing, frequent or severe respiratory tract infections, "blue spells" or having shortness of breath during activities.

What is the Vascular Ring and Airway Program?

- We are a multidisciplinary program that was developed to better support patients with vascular rings, associated vascular compression anomalies and tracheal anomalies.
- The children we see are complex and need several specialists to give them comprehensive care.
- Medical and surgical specialists on our team include cardiac and general surgeons, gastroenterologists, pulmonologists, otolaryngologists, speech pathologists, cardiologists and nutritionists.
- The program is supported by advanced practice providers from cardiac surgery, cardiology and general surgery.

When should I contact the program about my child?

Our multidisciplinary team can review your child's case at any time to make recommendations for monitoring symptoms, testing, medical treatment or surgery. An evaluation before your child's birth isn't usually needed. However, we are happy to provide anticipatory guidance if requested.

Contact us

The Vascular Ring and Airway Program

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[Second Opinion Portal at Boston Children's](#)