



This family education sheet explains what a double aortic arch is and how it's treated.

### Key points

- The condition can be related to certain genetic syndromes.
- Babies with this condition may need surgery before they turn 1.
- Some babies need surgery soon after they are born.

### What is a double aortic arch?

- A double aortic arch is a type of complete vascular ring.
- The aortic arch is 2-sided instead of 1-sided. The aortic arch is made up of a right aortic arch and a left aortic arch (see Figure 1).
- The ligamentum arteriosum connects the aorta to the pulmonary trunk.
- Usually, both arches and the ligamentum form a "napkin ring" around the trachea (breathing tube) and esophagus (swallowing tube).
- The napkin ring can squeeze the trachea (breathing tube) and/or esophagus (swallowing tube), causing swallowing or breathing problems.
- Children with vascular rings can also have a condition called tracheomalacia. This means that the airway collapses when breathing.

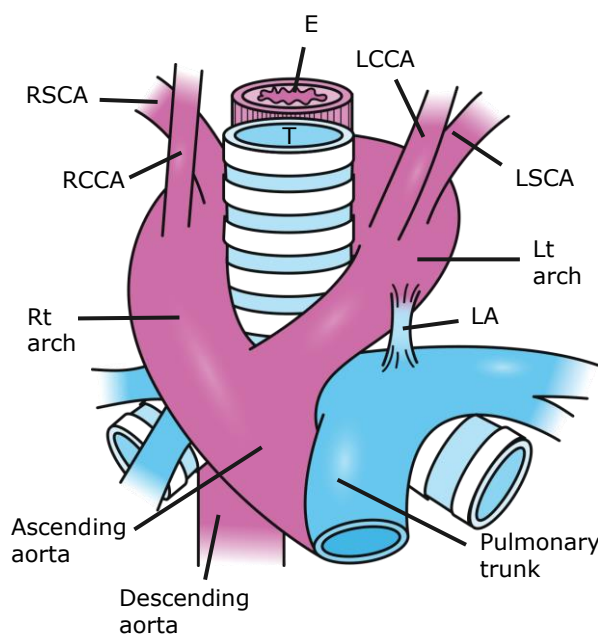


Figure 1.

**Figure 1 Key:** Esophagus (E), trachea (T), left subclavian artery (LSCA), left common carotid artery (LCCA), ligamentum arteriosum (LA), left aortic arch (Lt arch), brachiocephalic artery (RBCA), right common carotid artery (RCCA), right subclavian artery (RSCA), right aortic arch (Rt arch)

### What causes it?

- 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 happens when these vessels form abnormally.

### Is it related to a genetic diagnosis?

- Vascular rings can be related to genetic syndromes such as 22q11.2 (DiGeorge syndrome).
- You can talk about genetic testing options with your child's cardiologist.

### How is it treated?

- Babies born with a double aortic arch may need surgery before they turn 1 year old.
- The timing for surgery usually depends on how serious the symptoms are.
- Some babies with severe symptoms may need surgery soon after birth. This is rare.
- Surgery typically involves a thoracotomy and removing the smaller aortic arch.
- If your child needs other airway and esophageal procedures, such as a tracheopexy and esophageal mobilization, this will happen during the same operation.

### What are the symptoms?

- Most children with a double aortic arch will develop symptoms at some point in their lives.
- Many children develop symptoms in infancy or early childhood.
- Swallowing symptoms can include gagging, choking or coughing when eating or drinking, problems with food getting stuck or pain when swallowing, and/or aspiration (liquids going into the breathing tube when drinking).

- Some children may change their diets to avoid these symptoms by taking a long time to chew food, cutting food in small pieces or taking frequent sips of liquid between bites of food.
- Respiratory symptoms can include a chronic croupy or “barking” cough, noisy/junky breathing, frequent or recurrent respiratory tract infections, “blue spells” or shortness of breath with activities.

## What is the Vascular Ring and Airway Program?

- We are a multidisciplinary program that supports children with vascular rings, related vascular compression anomalies and tracheal anomalies.
- Children with these conditions are complex and need to see multiple specialists to get comprehensive care.
- Medical and surgical specialists on our team include cardiac and general surgeons, gastroenterologists, pulmonologists, otolaryngologists, speech pathologists, cardiologists and nutritionists.

## When should I contact the Vascular Ring and Airway Program about my child?

You can contact our team at any time to get recommendations for monitoring your child’s symptoms, testing, medical treatment or surgery.

### Contact us

#### **The Vascular Ring and Airway Program**

(617) 919-5377, Paulina Bongay, Case Coordinator

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

*This Family Education Sheet is available in Spanish and Arabic.*