Imaging Evaluation of the Vascular Rings

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Learning objectives

- To identify the normal embryology and anatomy of the aorta, supra-aortic branches and pulmonary artery

- To review the epidemiology, clinical and imaging findings of the vascular rings
Background

- Vascular rings can be divided in complete and incomplete. Each group with several types of rings with different clinical manifestations.

- A vascular ring is formed when the trachea and esophagus are surrounded by vascular structures from the primitive aortic arches.

- These vascular structures are: the aortic arch (single or double), pulmonary arteries, supraaortic branches, ductus arteriosus or the ligamentum arteriosum.

- The vascular rings account for approximately 1% of all cardiac malformations and are associated with genetic alterations like microdeletions of 22q11.2 locus.

- The vascular rings are diagnosed with great sensitivity and specificity with computed tomography (CT) and magnetic resonance (MR). Thanks to the greater spatial resolution and multiplanar imaging the radiologist may see how many aortic arches are, how the supra aortic vessels originate, and can fully characterize the course and characteristics of the pulmonary arteries. Thoracic x-rays, barium studies and echocardiography can only show some of the signs that may suggest the diagnosis.

- Vascular rings may be classified as complete: when the trachea and esophagus are completely surrounded, and incomplete when at least a side or portion is not completely surrounded.
Findings and procedure details

In this poster we will review the embryology and imaging findings of the most common vascular rings.

EMBRYOLOGY

- The aorta starts to develop at the third week of gestation. Edwards’s theory says that there are two aortic arches with two primitive aorta, each with a ventral and a dorsal segment. These segments are communicated by six paired pharyngeal arches (Fig. 1 on page 11).
- The ventral segments form the ascending aorta and dorsal segments form the descending aorta.
- The third arches form the carotid arteries.
- The fourth left arch forms the left aortic arch and the fourth right arch with the seventh intersegmental artery form the right subclavian artery.
- The sixth arches form the pulmonary artery and the ductus arteriosus.
- The seventh arch forms the subclavian artery.
- The arches surround the trachea and esophagus, that's why if there is an abnormal involution, a vascular ring is formed.
- The aortic arch is considered right or left depending which main bronchus it crosses.
- There are multiple anatomical variants of the supra-aortic vessels (Fig. 2 on page 12).

DOUBLE AORTIC ARCH

- A double aortic arch (DAA) results from the persistence of both fourth primitive aortic arches. Each arch gives rise to the subclavian arteries and the common carotids separately (Fig. 3 on page 13).
- The largest arch is called dominant. In 75% of cases, the right aortic arch is the dominant one. The most common configuration in vascular rings is
a right arch, with a left descending aorta and a left ligamentum arteriosum. The left arch may have a small diameter, and may even be atretic, with a fibrous tract and a diverticulum at the base of the interrupted segment. This tract is generally ipsilateral to the descending aorta and is located distal to the origin of the subclavian artery. Determining the dominant arch is important because it defines the surgical approach.

- It is the most prevalent symptomatic vascular ring, since the trachea and esophagus are completely surrounded and compressed. Symptoms include: wheezing, stridor, tachypnea, cyanosis and dysphagia. It may be associated with Tetralogy of Fallot and transposition of the great vessels.

- **Imaging:**
  - On chest X-rays, two aortic knobs and compression of the trachea may be seen.
  - Barium swallow may demonstrate right or bilateral indentations at the level of the aortic arch and in the lateral projection indentation of the posterior aspect of the esophagus can also be demonstrated.
  - Although conventional radiology images suggest the diagnosis of a vascular ring, a multiplanar image must always be acquired to confirm the diagnosis and for surgical planning.
  - In tomography (CT) and magnetic resonance imaging (MRI) two aortic arches originating from the ascending aorta and extending to each side of the esophagus and trachea may easily be observed. The dominant arch is usually larger and is located in the opposite side of the descending aorta (Fig. 4 on page 13).
  - The four vessel sign can be detected: 2 ventral carotid arteries and two dorsal subclavian arteries uniformly separated and forming a trapezoidal figure around the trachea (Fig. 5 on page 14).
  - Multiple signs indicate the presence of DAA when an atretic arch is present (Fig. 6 on page 15):
    1. Presence of a diverticulum in the proximal region of the descending aorta, opposite to the permeable aortic arch.
    2. Descending aorta on the opposite side to the aortic arch.
    3. Subclavian artery distorted with a lower and posterior course due to the atresia of the segment distal to its origin.

RIGHT AORTIC ARCH WITH MIRROR IMAGE BRANCHING AND LIGAMENTUM ARTERIOSUM
• Occurs when there is persistence of the fourth right arch and partial regression of the fourth left arch between the subclavian artery and the descending aorta.

• Persistence of the ligamentum arteriosum or a persistent left ductus arteriosus form a complete vascular ring, encountered in about 75% of patients.

• The ligamentum arteriosum originates in the proximal descending aorta, passes behind the esophagus and the trachea up to the left pulmonary artery.

• The branching pattern in this configuration of the aortic arch is as follows: Left brachiocephalic artery (divides into left common carotid artery and left subclavian artery), right common carotid artery and right subclavian artery. The descending aorta is located on the right side. (Fig. 7 on page 20 and Fig. 8 on page 21)

• 66 to 98% of patients with this type of vascular ring will present with congenital heart disease, Tetralogy of Fallot being the most common.

• Since this ring is formed by a ligament, a significant stenosis of the airway is not really established producing milder symptoms and it is usually an incidental finding.

• Imaging:
  • In the esophagogram a right aortic arch and posterior compression of the esophagus is seen.
  • CT is better demonstrating the tracheal compression and the course of the vessels.
  • In MRA or CTA the presence of ligament is suspected when a "dimple" is seen in the aorta, although if the ligament is thick enough it may be seen. If the ring is secondary to a patent ductus arteriosus it can be more easily observed.

**RIGHT AORTIC ARCH WITH ABERRANT LEFT SUBCLAVIAN ARTERY**

• It is considered an anatomical variant present in up to 0.1% of the adult population.

• The vascular ring occurs with the combination of an RAA and an aberrant left subclavian artery (ALSA) that originates as the last branch of the arch.
• The subclavian artery crosses the mediastinum from right to left with a course posterior to the esophagus, forming an incomplete vascular ring.

• In the presence of a left ligamentum arteriosum a complete vascular ring is then conformed. RAA and ALSA may be associated with Tetralogy of Fallot, although the association with congenital heart diseases is low.

• In some cases the origin of the ALSA presents a small bulging, that has been called "Kommerell's diverticulum".

• An enlarged diverticulum can indentate the posterior wall of the esophagus and trachea resulting in the onset of symptoms.

• The embryological origin of RAA and ALSA lies in the interruption of Edward's hypothetical left arch between the left common carotid and the ipsilateral left subclavian artery.

• The branching pattern of the arch is: the left common carotid, followed by the right common carotid artery, the right subclavian artery and the left aberrant subclavian artery. The descending thoracic aorta is usually located on the right side or near the midline.

• Although this type of configuration is the most symptomatic, the majority of patients are asymptomatic because this type of ring is relatively lax with little compressive effect.

• **Imaging:**

• A right aortic arch can be identified in the PA projection of the chest X-rays.

• The barium swallow shows posterior and lateral-right indentation of the esophagus.

• In patients with complete vascular ring, axial images (CT and MRI) show 4 separate branches emerging from the aortic arch. The left subclavian artery is the last branch of the aortic arch with retroesophageal course; it may originate from a diverticulum of Kommerell. The distal trachea may be compressed to varying degrees (Fig. 9 on page 15, Fig. 10 on page 16 and Fig. 11 on page 17).

**LEFT AORTIC ARCH WITH ABERRANT RIGHT SUBCLAVIAN ARTERY**

• Most common anomaly of the aortic arch.

• With an estimated prevalence of 0.5 - 2%.
• Occurs when there is regression of the right aortic arch between the right common carotid artery and the right subclavian artery. The right subclavian artery originates from the proximal descending thoracic aorta.

• The ramification pattern is: right common carotid artery, left common carotid artery, left subclavian artery and right subclavian artery (it crosses behind the esophagus from left to right) (Fig. 12 on page 17)

• Most of the times there is a left ductus arteriosus.

• It's an incomplete ring because only 3 sides of the trachea and esophagus are surrounded.

• In the majority of the cases it is not associated to any other congenital anomalies, but it may be accompanied by aortic coarctation and ventricular septal defects.

• It is relatively common in patients with Down syndrome, occurring in up to 35%.

• 10% of the patients will have esophagus compression generating dysphagia. This is more common between the 4th - 5th decades, and it is secondary to calcification of arterial walls and a decrease in esophageal compliance.

• Imaging:

• Up to 40% of Barium esophagram may be normal.

• In CT and MRI there are 4 vessels originating from the aortic arch, being the 4th vessel the right subclavian artery and it may be seen passing behind the esophagus (Fig. 13 on page 18 and Fig. 14 on page 19).

• The origin of the vessel may be dilated configuring a Kommerell’s diverticulum.

PULMONARY SLING

• Glaececke and Doehle described the trachea obstruction secondary to the abnormal origin of the pulmonary artery in 1897.

• In this case the left pulmonary artery originates from the right pulmonary artery and it crosses above the right main bronchus with posterior direction between the trachea and the esophagus forming an incomplete ring (Fig. 15 on page 22). With the presence of a ductus arteriosus or arteriosus ligamentum a complete ring is configured.
• The exact pathophysiology remains unclear, but the most accepted theory is that the left pulmonary artery doesn’t connect with the 6th left aortic arch, instead it connects with the 6th right aortic arch.

• It is a rare congenital malformation with strong genetic link. It is seen in identical twins and Trisomy of the 18th and 21st chromosome.

• Most infants are affected within the first few weeks of life and present with stridor, respiratory distress, cyanosis, wheezing, and/or pneumonia but the severity of symptoms depends on the compression degree over the trachea and esophagus. Some patients may reach adulthood without any symptoms.

• The most common association is VACTERL complex (vertebral anomalies, Anorectal anomalies, Cardiac anomalies, Tracheo-esophageal fistula, Renal anomalies, Limb anomalies).

• The most common associated cardiac anomalies are: persistence of left inferior vena cava; followed by aberrant right subclavian artery, coarctation of the aorta. Gastrointestinal tract malformations include: biliary atresia, Meckel diverticulum and Hirschprung’s disease.

**Classification:**

• Pulmonary Sling may be sub classified in 2 types, as follows:

  **Type I:**
  • The pulmonary sling is located above the carina (T4) where it contacts the distal trachea and right main bronchus. This type is associated with bronchomalacia and ipsilateral hyperinflation.
  • Type IA: with tracheal bronchus.
  • Type IB: without tracheal bronchus.

  **Type II:**
  • Most common type. Associated with right lung hypoplasia or agenesis. The sling is located at the level of T5-T6 displacing the carina to the left giving it a T form. The bronchial ramification is abnormal.
  • Type IIA: There is a supernumerary bronchus in the right and a tracheal bronchus or diverticulum in the place of the normal carina.
  • Type IIB: the trachea is long with an abnormally low carina and an increased bronchial angle. It may be associated with intrinsic tracheal stenosis.

**Imaging:**

• Lateral Chest X Ray for both types: Soft tissue image between the trachea and the esophagus.
• PA Chest X Ray in type I: right lung hyperinflation with partial bronchial obstruction or right bronchomalacia. There may be a right tracheal bronchus.
• PA chest X ray in type II: bilateral lung hyperinflation or volume loss in the right lung.

• Barium esophagogram: findings are nonspecific; there is a soft tissue mass between the trachea and the esophagus, and an indentation in the anterior wall of the esophagus.

• CT: superior evaluation of the airway, lung parenchyma, and vascular structures. Multiplanar examination with 3D reconstruction for surgical planning. Rapid acquisition avoiding the use of general anesthesia.

• MRI: lack of ionizing radiation, but it requires sedation or general anesthesia. CT is better visualizing lung parenchyma and airways. But, MR is better in the diagnosis of cardiac anomalies ( Fig. 16 on page 23, Fig. 17 on page 24 and Fig. 18 on page 24 ).
Fig. 1: Edwards theory of a double arch system that surrounds the trachea and the esophagus. There are two primitive aortas, each formed by a dorsal and ventral segment, communicated by 6 pairs of aortic or pharyngeal arches. The ventral segments form the ascending aorta and the dorsal segments give origin to the descending aorta. The third arches (III) give rise to the carotid arteries. The fourth left aortic arch forms the left aortic arch. If the right aortic arch persists and the left aortic arch involutes, it forms a right aortic arch. If the two fourth aortic arches persist, a double aortic arch arises. The sixth arcs form the pulmonary arteries (Ap) and the ductus arteriosus (purple). The seventh intersegmental cervical arteries (7th) form the subclavian arteries. The fourth right arc involutes, except for its proximal portion, which together with the seventh intersegmental
artery (7th) form the right subclavian artery. The first, second and fifth arcs involute (dotted lines).

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Fig. 2: The illustration shows the appearance of the supra-aortic vessels and their anatomical variants in axial images. A: normal (BA, LCCA and LSA) ; B: bovine aortic arch (the brachiocephalic artery shares a common origin with the left common carotid artery); C: aberrant right subclavian artery (RCCA, LCCA, LSA and ARSA); D: right aortic arch with mirror branching ( left BA, RCCA and RSA) E: aberrant left subclavian artery with right aortic arch (LCCA, RCCA, RSA and ALSA); F: four vessel sing in double aortic arch (common carotid arteries have a ventral position and the subclavian arteries are located dorsally). BA: Brachiocephalic Artery; RCCA: Right Common Carotid Artery; LCCA: Left
Fig. 3: Doble aortic arch. A: Illustration shows the appearance of a double aortic arch. Both the trachea and oesophagus are completely encircled by the vascular structures. Arising from the left arch are the left common carotid (LCCA) and the left subclavian artery (LSA). Arising from the right arch are the right common carotid (RCCA) and then the right subclavian artery (RSA). B: Posterior view of a double aortic arch with atretic left arch. The atretic left arch connects the left subclavian artery to the descending aorta and tetheres it inferiorly.

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Fig. 4: 13-months-old patient with suspicion of partial abnormal pulmonary venous return. Coronal T2 (A) and Coronal reconstruction of 3D angiography (B) show two vascular structures (arrows) on each side of the trachea, with a slight decrease in its caliber. A double aortic arch was diagnosed, with a right dominant arch.

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Fig. 5: Four-vessel sign. Axial reconstruction from a 3D angiography of the same patient of figure 4, shows 4 supra-aortic vessels, forming a symmetrical trapezoidal shape. The common carotid arteries have a ventral position (CCA) and the subclavian arteries (AS) are located dorsally.
**Fig. 6:** 3D angiography reconstruction of the same patient of figure 4 shows a double arch with atretic left segment (line) distal to the origin of the left subclavian artery. A distorted subclavian artery is seen (arrow). There is a diverticular projection in the proximal descending aorta opposite the patent right arch.

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Fig. 9: 8-years-old patient with tetralogy of Fallot. Coronal (A) and axial (B) T2 show right aortic arch (arrow in A), with vascular structure (arrow in B) that runs posterior to the esophagus (E). This structure corresponds to an aberrant left subclavian artery. A persistent left vena cava (asterisk) is also seen. Trachea (T)

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**Fig. 10:** 3D angiography multiplanar reconstruction shows the origin of the supra-aortic vessels. The left common carotid artery (LCCA) is the first branch of the arch. The left subclavian artery has an aberrant origin and is the last vessel of the arch. Persistent left vena cava (asterisk). ALSA: aberrant left subclavian artery; RCCA: right common carotid artery. RSA: right subclavian artery.

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**Fig. 11:** 3D angiography reconstruction shows right aortic arch and an aberrant left subclavian artery (arrow)

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Fig. 12: Coronal (A) and sagittal (B) T2 show a left aortic arch (A), with vascular structure (arrow) that runs posterior to the esophagus (E). This structure corresponds to an aberrant right subclavian artery.

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Fig. 13: Coronal reconstruction of 3D angiography maximum intensity projection shows an aberrant right subclavian artery (arrow)

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Fig. 14: 3D angiography reconstruction shows a left aortic arch and an aberrant right subclavian artery (arrow).

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Fig. 7: 5-years-old patient with history of complex congenital heart disease repair. Coronal T2 shows right aortic arch (arrow) located on the right side of the trachea.

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**Fig. 8:** 3D angiography reconstruction of the same patient as figure 7 shows right aortic arch with mirror branching. The first branch of the aortic arch is the left brachiocephalic artery (arrow), followed by the right common carotid artery and the right subclavian artery.

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Fig. 15: Pulmonary sling. Illustration shows an anomalous left pulmonary artery, which originates from the right pulmonary artery and then crosses between the trachea and oesophagus.

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Fig. 16: 3-months-old patient with suspected left pulmonary artery atresia. Coronal T2 (A) shows two vascular structures on each side of the trachea causing its compression. The structure on the right (arrow in A) is the left pulmonary artery, which has its origin in the right pulmonary artery and later crosses between the trachea and the esophagus (arrow in B).

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Fig. 17: 12-months-old patient with suspicion of a vascular ring due to hyperlucidity in the left hemithorax. Axial T2 white blood (A) and multiplanar reconstruction of 3D angiography (B) show the left pulmonary artery (arrows) originating from the right pulmonary artery, crossing between the trachea and the esophagus until reaching the left pulmonary hilum. Right atelectasis and left hyperinsuflation that caused dextroposition are also seen.

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Fig. 18: 3D angiography reconstruction shows an anomalous left pulmonary artery, which originates from the right pulmonary artery.

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Conclusion

• Vascular rings are a group of pathologies that can present with respiratory symptoms and/or dysphagia. History and Physical exam may be non-specific.

• Therefore, imaging plays a fundamental role in the accurate diagnosis of these pathologies and additionally can give information of other malformations that may be associated with each type of ring.

• The radiologist must know the anatomy and the normal embryology of the aorta and supraortic vessels, to suspect and diagnose in a precise way a possible vascular ring.
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