



# VASCULAR RINGS, MAGNETIC RESONANCE FINDINGS

Anillos vasculares, hallazgos por resonancia magnética

*Pedro Abad<sup>1</sup>*

*Sara Mesa<sup>1</sup>*

*Ricardo Uribe<sup>2</sup>*

*Ana María Alvarado<sup>2</sup>*

*Sebastián Isaza<sup>2</sup>*



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## Palabras clave (DeCS)

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Arteria pulmonar  
Imagen por resonancia magnética

## Summary

Vascular rings are a spectrum of pathologies where the aortic arch, pulmonary artery and/or the supra-aortic vessels have a different origin or an abnormal course; this may or may not produce tracheal and/or esophageal compression. These entities have a variable clinical presentation, with respiratory symptoms being the most common. They are associated with cardiac, gastrointestinal and other system malformations. We reviewed the available literature about the embryology, epidemiology, clinical and Magnetic resonance imaging findings of the most common vascular rings.

## Resumen

Los anillos vasculares son un espectro de patologías en las que el arco aórtico, la arteria pulmonar o la ramificación de los vasos supraaórticos tienen un origen o trayecto anormal y pueden comprimir la tráquea o el esófago, lo cual genera diferentes síntomas, como los respiratorios, que son lo más comunes. Estas patologías pueden estar asociadas a malformaciones cardíacas, del tracto gastrointestinal y en otros sistemas. Se realiza una revisión de la literatura sobre la embriología, epidemiología, clínica y hallazgos imagenológicos en resonancia magnética de los principales anillos vasculares.

## Introduction

A vascular ring is formed when the trachea and esophagus are completely surrounded by structures derived from the primitive aortic arches (1). These vessels include the aortic arch (single or double), branches of the aortic arch and pulmonary arteries, the ductus arteriosus and the ligament arteriosus (2). They account for approximately 1% of all congenital cardiac malformations (3) and are associated with genetic alterations, such as microdeletions of locus 22q11.2 (2). This compression may result in symptoms originating in the upper airway or dysphagia; however, there is a broad spectrum of clinical manifestations, ranging from asymptomatic patients to newborns with severe respiratory distress. In general, the most common symptoms include stridor,

dysphagia, wheezing, dyspnea, cough, and recurrent respiratory infections (4).

Vascular rings are classified into complete (trachea and esophagus completely surrounded by vascular structures) and incomplete (vascular structures that cause compression, without forming a complete circle) (5) (Table 1).

The most common malformations are the double aortic arch, right aortic arch with aberrant left subclavian, left aortic arch with aberrant right subclavian and the anomalous origin of the left pulmonary artery also known as pulmonary sling (3,6).

X-ray images of the chest and barium digestive studies make this disease suspect and can be used as non-invasive screening methods; however, it must be confirmed with multiple images, such as computed



<sup>1</sup>Radiologist, Instituto de Alta Tecnología Médica (IATM). Medellín, Colombia.

<sup>2</sup>Radiology resident, Universidad CES. Medellín, Colombia.

tomography (CT) or magnetic resonance imaging (MRI) (4). Computerized angiography (CTA) is a rapid method of high sensitivity and specificity and the ability to evaluate the airway, which makes it the test of choice to confirm the diagnosis. However, its disadvantages are high doses of contrast medium and ionizing radiation. Advantages of MRI include contrast resolution, absence of ionizing radiation, complementary evaluation of intracardiac anatomy, ventricular function, and the relationship between the ring and the trachea and esophagus. The main disadvantages of MRI are the long acquisition times, which generally require sedation and intubation, its lower spatial resolution compared to TCA, and its high cost. Additionally, airway evaluation may be limited by orotracheal intubation, especially in this group of patients who may have airway compromise (5,7).

The imaging center where the study was done is specialized in MRI and has become a benchmark for cardiovascular studies in the pediatric population. The cardiovascular MRI protocol includes anatomical images, cinema, 3D angiography, flow quantification, among others. The vascular study leads to finding, in many cases, vascular anatomical variants and even to diagnosing pathologies not previously suspected, which indicates changes in treatment. Vascular rings, complete or incomplete, are visualized in these patients and although traditionally MRI is not the technique of choice, its contribution to the comprehensive assessment of congenital heart disease is important.

**Table 1. Classification of vascular rings**

Complete vascular rings
Double aortic arch and right aortic arch with aberrant left subclavian artery and duct (90 %)
Left aortic arch with right descending aorta and right canal
Right aortic arch with left descending aorta and left duct
Left aortic arch with aberrant right subclavian artery and right canal
Complete vascular rings
Abnormal origin of left pulmonary artery pulmonary sling pulmonary
Innominate artery compression syndrome

Source: Backer and Mavroudis (8).

## Embryology

The aorta begins its development in the third week of gestation (9). According to Edwards’ theory, there is a double-arc system where initially there are two primitive aortas, each formed by a dorsal segment and a ventral segment. These segments are communicated by 6 pairs of aortic or pharyngeal arcs. The ventral segments form the ascending aorta, the dorsals give rise to the descending aorta and the fourth left aortic arch forms the left aortic arch (10, 11). The third arches give rise to the carotid arteries, the fourth right arch together with the seventh intersegmental artery form the right subclavian artery. The fifth arches involute and the sixth arches form the pulmonary arteries and the ductus arteriosus (6, 12, 13). The arch system surrounds the trachea and oesophagus,

so when segments persist or involute inappropriately, vascular rings are formed (10) (Figure 1).

The aortic arch is right or left depending on which bronchus source crosses the arch (10).

### 2.1 Double aortic arch

The double aortic arch (DAA) results from the persistence of the fourth primitive aortic arches. Each arch gives rise to the subclavian arteries and the common carotid arteries separately (14). The largest arch is called the dominant. The right aortic arch is dominant in 75 % to 81 % of cases (1,15), in 16 % the left arch is dominant and in 3 % it is co-dominant (7). The most common configuration is a right arch, with a descending left aorta and a left arteriosus ligament. The left arch may have a small diameter and even be atresic, with a fibrous tract and a diverticulum at the base of the interrupted segment (Figure 2). This tract is usually ipsilateral to the descending aorta and is located distal to the origin of the subclavian artery (2,14). It is important to determine which is the dominant arc to define the surgical approach, which is performed on the non-dominant side (16).

DAA has been reported to be the prevalent symptomatic vascular ring, representing up to 60% of cases (3,17). It is symptomatic in most cases, since the trachea and esophagus are completely surrounded and compressed. The main symptoms are wheezing, stridor, tachypnea, cyanosis and dysphagia. It can be associated with tetralogy of Fallot and transposition of great vessels (5,18,19).

In MRI, there are two aortic arches that originate from the ascending aorta and extend to each side of the esophagus and trachea (Figure 3). The dominant arch is larger and opposite the descending aorta. The sign of “four arteries” can also be found: two ventral carotid arteries and two dorsal subclavian arteries separated uniformly, forming a trapezoidal figure around the trachea (Figure 4) (2,20).

Multiple signs indicate DAA when there is an atrial arch: a) a diverticulum in the proximal region of the descending aorta opposite the permeable aortic arch (21), b) contralateral descending aorta to the aortic arch, c) distorted subclavian artery with a lower and posterior course due to atresia of the distal segment to its origin (22) (Figure 5).

### 2.2 Right aortic arch with aberrant left subclavian artery

The right aortic arch (RAA) is an anatomical variant present in up to 0.1% of the adult population (23,24). The vascular ring occurs with the combination of an RAA and an aberrant left subclavian artery (ALSA) that originates as the last arch emergence. The subclavian artery crosses the mediastinum from right to left with a posterior course to the esophagus forming an incomplete vascular ring (25). Together with a left arteriosus ligament it forms a complete vascular ring (26). RAA and ALSA are related to tetralogy of Fallot, although the association with congenital cardiopathies is relatively low (27).

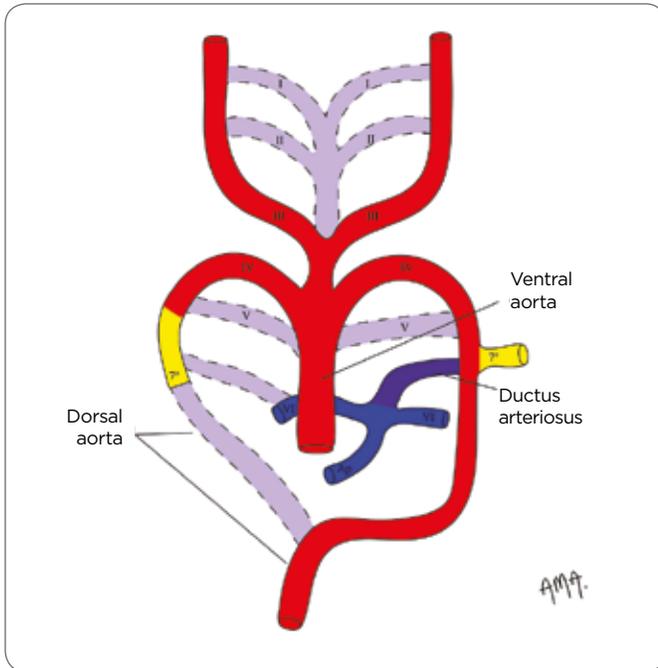


Figure 1. Scheme of Edwards' theory of the double-arc system surrounding the trachea and oesophagus. There are two primitive aortas, each consisting of a dorsal and ventral segment, communicated by 6 pairs of aortic or pharyngeal arcs. The ventral segments form the ascending aorta and the dorsal segments give rise 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 involute the left aortic arch, it gives rise to a right aortic arch. If the two fourth aortic arches persist, a double aortic arch is created. The sixth arches form the pulmonary arteries (PA) and the ductus arteriosus (purple). The seventh cervical intersegmental arteries (7th) form the subclavian arteries. The fourth right arch involute, except for its proximal portion, which together with the seventh intersegmental artery (7th) forms the right subclavian artery. The first, second and fifth arcs involute (dotted lines). Source: made by author Ana María Alvarado.

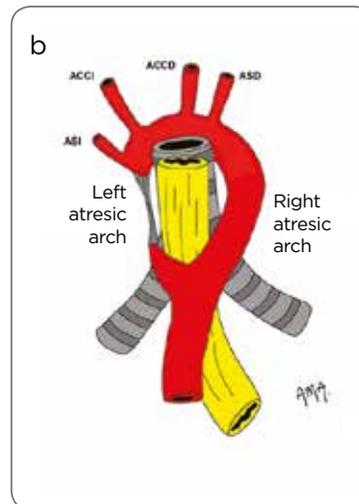
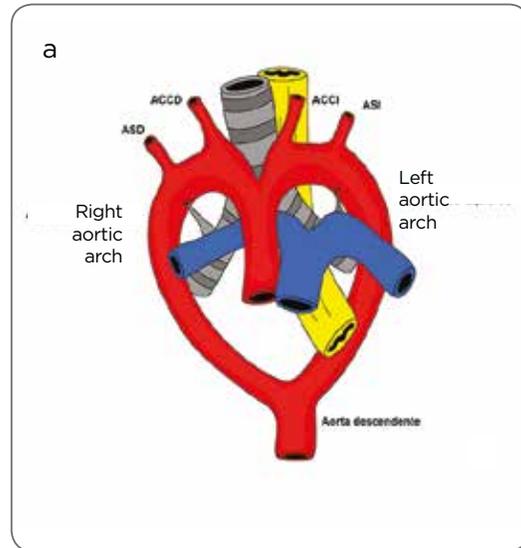


Figure 2. Double aortic arch. a) The illustration shows the appearance of a double aortic arch. The trachea and esophagus are completely surrounded by vascular structures. The left common carotid artery (LCCA) and the left subclavian artery (LSA) originate from the left arch. From the right aortic arch originates the right common carotid artery (RCCA) and the right subclavian artery (RSA). b) Posterior view of a double aortic arch with atresia segment, which connects the left subclavian artery with the descending aorta and produces inferior traction of the aorta. Source: made by author Ana María Alvarado. (RSA).

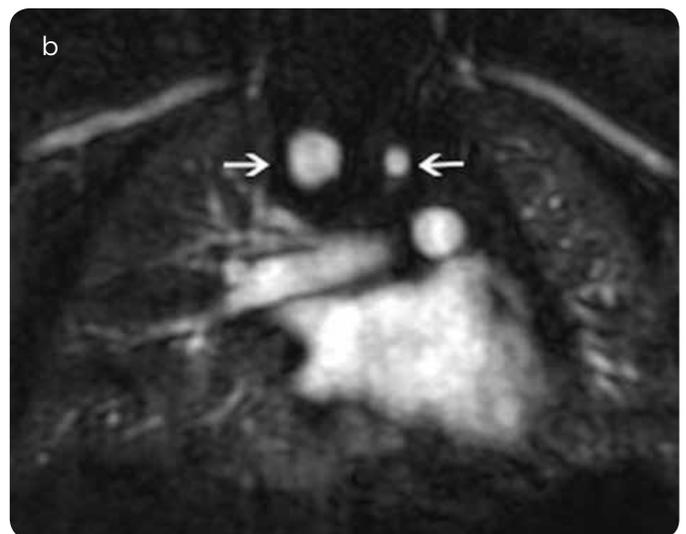
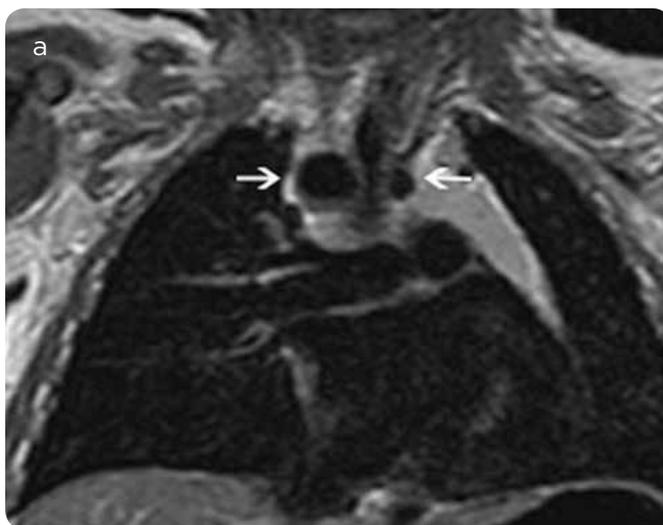


Figure 3. Double aortic arch, with dominant right arch. Patient of 13 months of age with suspicion of anomalous partial pulmonary venous return. a) Coronal with T2 information. b) Coronal reconstruction of 3D angiography. Two vascular structures (arrows) are observed on each side of the trachea, with slight decrease in calibre.

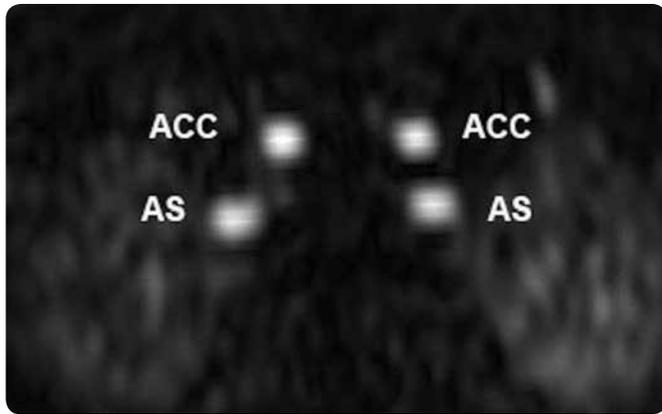


Figure 4. Sign of the four arteries. Axial reconstruction of 3D angiography of the same patient in image 1. 4 supra-aortic arteries are observed, showing a symmetrical trapezoidal shape. The common carotid arteries have a ventral position (CCA) and the subclavian arteries (SA) are located dorsally.

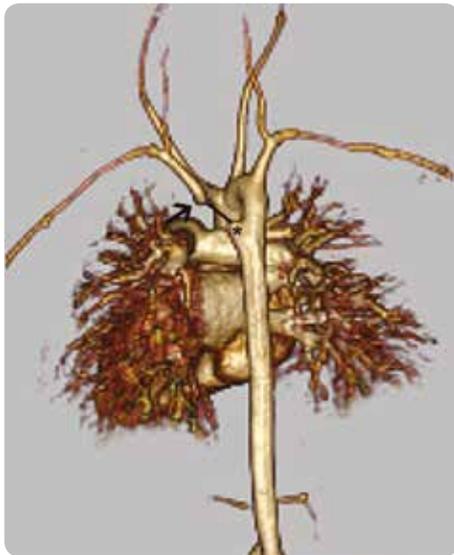


Figure 5. 3D angiography reconstruction, posterior view. Double aortic arch with atrial segment (line) distal to the origin of the left subclavian artery, which is distorted (arrow). Diverticulum in the proximal region of the descending aorta, opposite the permeable aortic arch (asterisk).

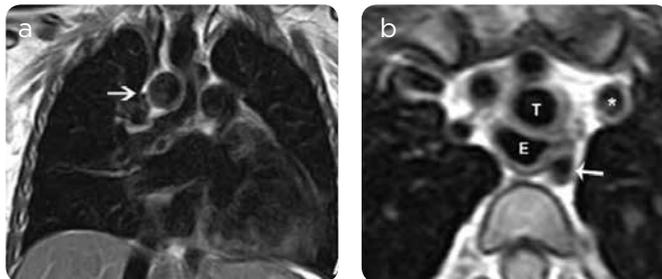


Figure 6. Aberrant left subclavian artery. 8-year-old patient with tetralogy of Fallot. a) Coronal and b) axial, with T2 information: right aortic arch (arrow in a), with vascular structure (arrow in b) that runs posterior to the esophagus (E). This structure corresponds to the left subclavian artery, which has an aberrant origin. Additionally, persistent right vena cava (\*) is identified. Trachea (T).

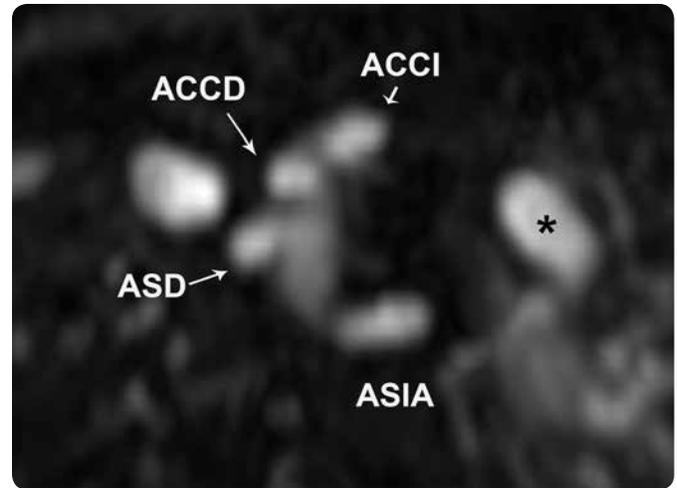


Figure 7. Aberrant left subclavian artery. Multiplane axial oblique reconstruction of 3D angiography, showing the origin of the supraaortic vessels. The left common carotid artery (LCCA) is the first branch of the arch. The left subclavian artery has an aberrant origin and is the last vessel to originate from the arch. Additionally, persistent left vena cava (asterisk) is observed. ALSA: aberrant left subclavian artery; RCCA: right common carotid artery. RSA: right subclavian artery.

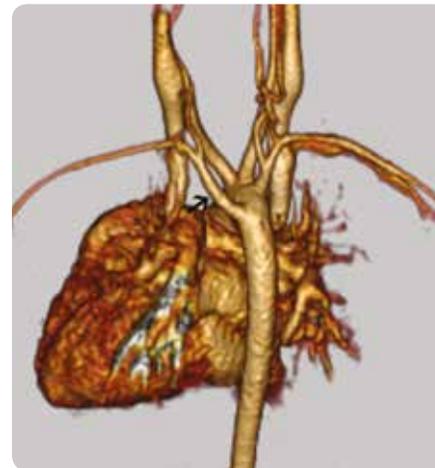


Figure 8. Aberrant left subclavian artery. Reconstruction of 3D angiography, posterior view, shows right aortic arch and aberrant left subclavian artery (arrow).

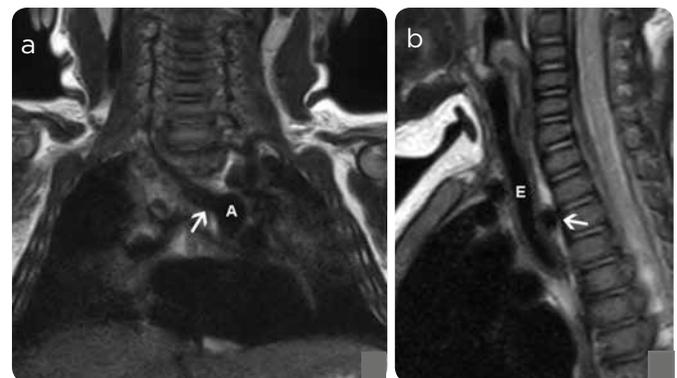


Figure 9. Aberrant right subclavian artery. a) Coronal MRI with T2 information and b) sagittal: left aortic arch (A), with vascular structure (arrow) with retroesophageal course (E). This structure corresponds to the right subclavian artery with aberrant origin.

In some cases, the origin of the aberrant left subclavian artery shows a dilation that has been called “Kommerell’s diverticulum” (28). An enlarged diverticulum can indent the posterior wall of the esophagus and trachea, resulting in symptoms (10).

The embryological origin of DAA and ALSA lies in the interruption of Edward’s hypothetical left arch between the left common carotid and the left ipsilateral subclavian artery (29,30). The branching pattern of the arch is: the left common carotid is the first branch, followed by the right common carotid, the right subclavian arteries and the left aberrant subclavian artery. The descending thoracic aorta is usually located on the right side or near the midline (31). Although this type of configuration is the most symptomatic, most patients are asymptomatic because the ring is lax and has little compressive effect (16).

In patients with a complete vascular ring, axial imaging (CTA and MR) demonstrates 4 separate brachiocephalic branches emerging from the aortic arch. The left subclavian artery is the last branch of the aortic arch with a retroesophageal course; it may originate from Kommerell’s diverticulum (32). The distal trachea may be compressed in varying degrees (16) (Figures 6 to 8).

### 2.3 Left aortic arch with aberrant right subclavian

The left aortic arch (LAA) with aberrant right subclavian artery (ARSA) is the most common congenital anomaly of the aortic arch with a prevalence in the general population of 0.5% to 2% (33). This variant results from the regression of the right arch between the right common carotid artery and the right subclavian artery. The right subclavian artery originates in the proximal descending thoracic aorta, thus configuring the following branching pattern: right common carotid, left common carotid, left subclavian and ARSA (31). The ARSA is then the last branch of the arch and typically has a caudo-cranial oblique retroesophageal course from left to right. Usually the alteration is associated with a left duct, yet a complete vascular ring is not configured because only 3 sides of the trachea and esophagus are compressed by vascular elements (20).

This anomaly is typically found in isolation, but may be less frequently associated with other congenital anomalies including: aortic coarctation, patent ductus arteriosus, ventricular septal defects, and abnormalities of the carotid or vertebral arteries. Patients with trisomy 21 have it, with a high prevalence, up to 35% (10).

Compression of the esophagus may occur in up to 10% of adults with this abnormality, causing difficulty swallowing (34). The dysphagia associated with it is known as lusoria. Dysphagia lusoria is most common in adults in the fourth or fifth decade of life (20).

MRI identifies the aberrant origin of the right subclavian as the last branch of the right aortic arch and its retroesophageal course can be confirmed to the right (Figures 9 and 10). The origin of the aberrant vessel can be dilated and form a Kommerell diverticulum. Vascular alterations, such as calcifications or pathological dilations (20) can be seen.

### 2.4 Right aortic arch with mirror branching with permeable retroesophageal artery ductus arteriosus

It 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 (3,35). The persistence of the arterial ligament or left ductus arteriosus forms the complete vascular ring (12), appears in 75% of patients with right aortic arch and mirrored vessels (20). The arteriosus ligament is born in the proximal descending aorta, passes posterior to the esophagus and trachea and is inserted into the left pulmonary artery (12,20).

The ramification pattern in this configuration of the aortic arch is as follows: left brachiocephalic artery (divided 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 (20,36) (Figures 11 and 12).

66 to 98 % of patients with this kind of vascular ring suffer from congenital heart disease (1,6,12,25), and the most common is tetralogy of Fallot (20). Since this ring is formed by a ligament, no significant airway stenosis is formed, it produces few symptoms and in general, it is an incidental finding (6).

In MRI or TCA it is suspected when a “notch” is observed in the aorta, (2, 12, 20) although in some cases, if the ligament is thick, it can be identified in CT (2). If the ring is secondary to the ductus arteriosus, it can be more easily observed (16). The diagnosis is also made if esophageal or tracheal compression is identified or if there are symptoms of vascular ring (12). The treatment for this vascular ring is ligament dissection surgery (12,35).

### 2.5 Anomalous origin of the left pulmonary artery or pulmonary sling

Tracheal obstruction secondary to the abnormal origin of the left pulmonary artery was first described in 1897 by Glaevecke and Doehle (36). In this pathology, the left pulmonary artery is born from the right pulmonary artery, crosses over the right main bronchus and goes posterior to the trachea and anterior to the esophagus to form an incomplete ring (37-40); this can also be formed if there is a patent ductus arteriosus or ductal ligament (41,42) (Figure 13).

The physiopathology of this entity is not currently known, but the most accepted theory is that the left post-bronchial pulmonary artery does not connect with the sixth left aortic arch, but with the right (43), crosses posterior to the trachea to generate the pulmonary sling. If it is connected in the anterior portion, the pulmonary pseudosling is created (35,38,41,42).

Pulmonary sling is a rare congenital abnormality (39,43), with genetic influence (41,44). Patients in early childhood have respiratory symptoms and difficulty feeding (40). The severity of symptoms varies depending on the compressive effect on the airway and esophagus (16,38,41,45). In some cases, patients may reach adulthood without symptoms, and then present with symptoms such as dysphagia (38).

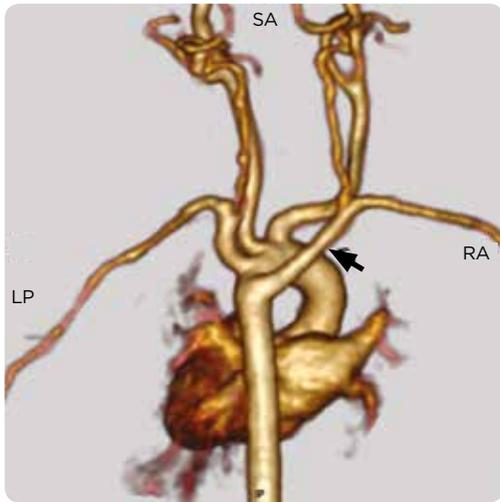


Figure 10. Aberrant right subclavian artery. Reconstruction of 3D angiography, posterior view: left aortic arch and aberrant right subclavian artery (arrow).

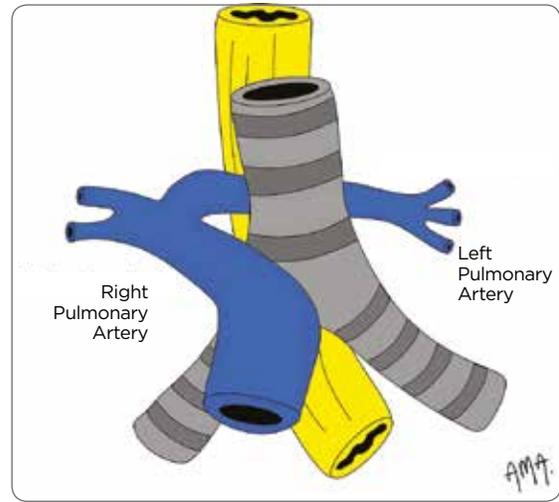


Figure 13. Sling of the pulmonary artery. The illustration shows the anomalous origin of the left pulmonary artery, which originates from the right pulmonary artery, crosses between the trachea and esophagus to reach the right pulmonary hilum. Source: made by author Ana María Alvarado.



Figure 11. Right aortic arch with mirror branching. 5-year-old patient with a history of congenital heart disease treatment. Coronal with T2 information: right aortic arch (arrow) passing to the right of the trachea.

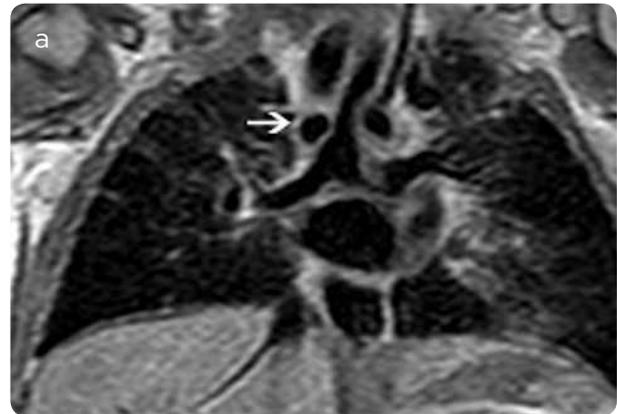


Figure 14. Sling of the pulmonary artery. 3-month-old patient with suspected left pulmonary artery atresia. a) Coronal with T2 information: two vascular structures on each side of the trachea that cause compression. The structure on the right is the left pulmonary artery (arrow). b) Axial with T2 information, white blood: left pulmonary artery originating from the right pulmonary artery and then crossing between the trachea and esophagus (arrow).

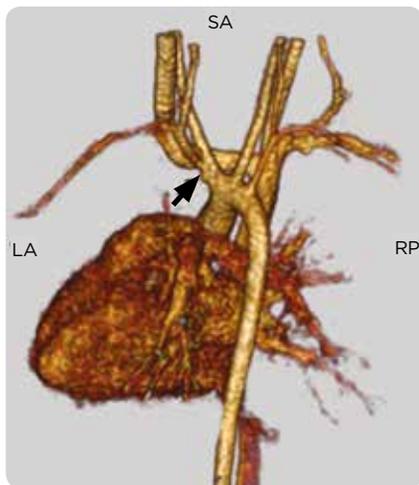


Figure 12. Right aortic arch with mirror branching. Reconstruction of 3D angiography, posterior: right aortic arch. The first branch is the left brachiocephalic trunk (arrow), followed by the right common carotid artery and the right subclavian artery.

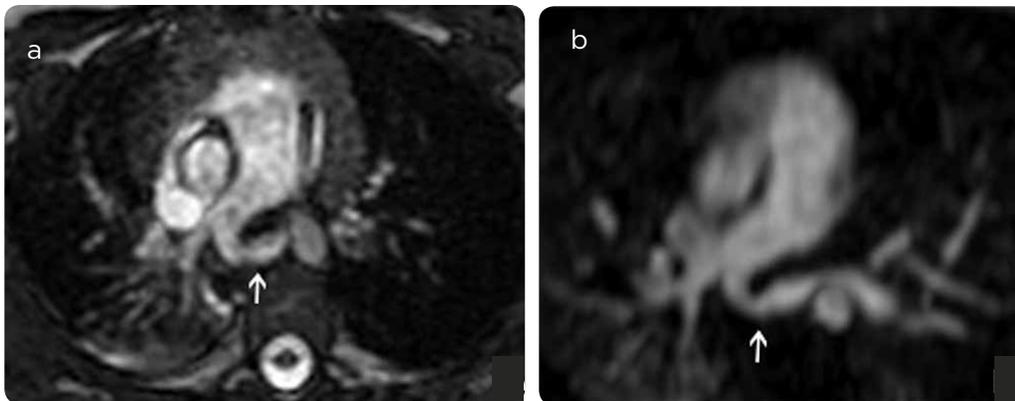


Figure 15. Sling of the pulmonary artery. A 12-month-old patient with suspected vascular ring due to the discovery of hyperlucid left hemithorax. a) Axial MRI with white blood T2 information, b) multiplane reconstruction of 3D angiography: the left pulmonary artery (arrows) originates in the right pulmonary artery, crossing between the trachea and esophagus until reaching the left pulmonary hilum. Also, right atelectasis and left pulmonary hyperinflation causing dextroposition.



Figure 16. Sling of the pulmonary artery. 3D angiography reconstruction: anomalous origin of the left pulmonary artery in the right pulmonary artery.

This abnormality is associated with other malformations, of which the main one is the VACTER spectrum (vertebral abnormalities, imperforate anus, cardiac abnormalities, tracheoesophageal fistula, renal abnormality, limb abnormality) (41). Associated cardiac abnormalities are responsible for patient mortality. These include persistent left superior vena cava (more common), followed by aberrant right subclavian artery, coarctation of the aorta, ventricular and atrial communication, among others (41,46,47). The gastrointestinal abnormalities to which it may be associated are bile duct atresia, Meckel's diverticulum and Hirschprung's disease (41).

### 2.5.1 Classification

It is divided into two types which, in turn, are divided into subtypes (38,41,42).

**Type I:** The *sling* is located above the carina (T4-T5) (12) where it is in contact with the distal trachea and the right main bronchus. It is accompanied by right bronchiomalacia with associated ipsilateral pulmonary hyperinflation (41). On some occasions it may have tracheal bronchus (41) and depending on this finding it is classified as Type IA (absent) and Type IB (present) (41,48).

**Type II:** Es Type II: This is the most common type (39, 47). It may be accompanied by hypoplasia or agenesis of the right lung or in some cases with pulmonary hyperinflation (41).

The *sling* is located at the height of T5-T6 with the carina deviated to the left and T-shaped (12,41) with abnormal bronchial branching (41). It is divided into 2 types:

- Type IIA has a supranumerary bronchus, usually on the right side, with a tracheal bronchus or diverticulum where the carina should be (41).
- In type IIB the trachea is long, with low carina and increased bronchial angle (41). It can be associated with intrinsic tracheal stenosis due to complete cartilaginous rings (6,12,41,48).

The main advantage of MRI is that it does not use ionising radiation, although it requires sedation or anaesthesia and does not have the same resolution in the parenchyma and airway as tomography (39,41). In complex congenital cardiopathies, MRI is useful as part of the protocol for the diagnosis of these pathologies, and in many cases reduces the need for further examination (49) (Figures 14 to 16).

The treatment depends on the type of sling and the symptoms. If it is type I and the patient is asymptomatic, a clinical follow-up is performed, but if the patient has respiratory symptoms, the vessel can be re-implanted. For type II patients it is believed that the reimplantation of the pulmonary artery will not improve respiratory symptoms and therefore surgery for airway stenosis should always be performed (42,50).

## Conclusion

Vascular rings are a group of pathologies that manifest with respiratory symptoms or dysphagia, in which the physical examination provides little information for diagnosis, so diagnostic images are required, ideally tomography. Many of these patients have associated cardiopathies, for which an MRI is performed as part of the study, and there are incidental vascular pathologies that may be important for the comprehensive assessment of the underlying disease. It is essential to know the embryology, the normal anatomy and the anatomical variants of the aorta and the vessels of the neck, in order to identify these pathologies and make an adequate diagnosis, as well as to evaluate all possible associated cardiac pathologies.

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## Correspondence

Ricardo Uribe González  
Resident of Radiology Universidad CES  
Diagonal 29D # 9A south-150  
Medellín, Colombia  
ricardouribegonzalez@gmail.com

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