Congenital anomalies and variants of the aortic arch. A pictorial essay based on an embryological model.

Poster No.: C-2135  
Congress: ECR 2017  
Type: Educational Exhibit  
Authors: N. S. Trujillo Calderon, J. F. Forero, J. I. Castaño Quintero, R. J. Valdés, J. M. PEREZ, M. C. perez; Bogotá/CO  
Keywords: Congenital, Screening, Barium meal, MR, Digital radiography, CT, Paediatric, Cardiovascular system, Cardiac  
DOI: 10.1594/ecr2017/C-2135

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Learning objectives

LEARNING OBJECTIVES

- To present an embryological model (The Edwards model) as a useful tool to understand the morphology as well as manifestations of most congenital anomalies and variants of the aortic arch.
- To describe the radiologic features and signs of each anomaly and variant of the aortic arch.
- To describe the role of X ray studies, multi-detector CT and MRI in diagnosing and characterization of the anomalies and variants of the aortic arch.

Abbreviations: Ascending aorta (AA), descending aorta (DA), right common carotid artery (RCCA), left common carotid artery (LCCA), right subclavian artery (RSA), left subclavian artery (LSA).
Background

The Edwards model describes the embriological origin of the aortic from a double arch system that depending on the site in which it is interrupted, explains the resultant configuration of the aortic arch and its branches.

These anomalies and variants of the aortic arch and its branches could lead to compression of the airway and esophagus and have been associated with other cardiac and airway malformations. Based on this understanding, certain patterns of compression of displacement of the airway and the esophagus can be detected on chest radiography and barium swallow which are a useful first step approach.

The literature suggests that vascular rings correspond to 1% of congenital cardiovascular abnormalities, however, it is believed that it could be greater due to underdiagnosis in cases of asymptomatic patients (1). Most of these alterations are sporadic but have been associated with genetic factors such as the microdeletion of the long arm of chromosome 22 (2).

CT and MRI can confirm the underlying abnormality as well as differentiate some of them which have similar presentations.
Findings and procedure details

From an embriological model, the variants and anomalies of the aortic arch are presented as well as the basis for their clinical and radiological presentation.

PATHOGENESIS

In 1948, Edwards (3) created a model in order to understand the radiological manifestations of the defects of the aortic arch. The model shows the normal embryological development of the double aortic arch, the ventral ascending and dorsal descending aorta are localized in the midline and the branches are the common carotid artery, the subclavian artery and a ductus arteriosus on each side (4).

Normally, there is an interruption of the dorsal segment of the right arch between the RSA and the DA by the involution of the ductus arteriosus, allowing the formation of the left aortic arch(4-5). The aorta descends in the left side and its branches are the innominate artery (RSA and RCA), the LCCA and the LSA. The arterial ligament is located on the left side and runs between the left pulmonary artery and the DA(6).

Based on Edwards' model the defects are classified according to the location of the ductus arteriosus and the dissolution site of the double arches. The vascular rings are usually classified according to the laterality of the arch, the course of the DA, the pattern of aortic branching and the laterality and course of the ductus arteriosus(4).

Double aortic arch

It is a true vascular ring and the most frequent alteration, usually symptomatic. The ascending aorta originates anterior to the trachea and is divided into a left anterior arch and a right posterior arch, enclose the trachea and the esophagus and come together to form a common DA (4). There are two common carotid and two subclavian arteries (4). The right arch is usually larger, longer, posterior, and cephalic than the left arch (2-4). Chest x-ray may show a lateral lateral indentation of the trachea and opacities in both sides of the airway column of the trachea(2).

Right-sided aortic arch

Type I: Right-sided aortic arch with mirror image branching.
There is an interruption of the left arch between the DA and the LSA. The arch and the DA are on the right, a left ductus arteriosus connects the left pulmonary artery to the ipsilateral subclavian, so it is not a vascular ring (4-5). There is a right arch with 3 branches: the innominate, the CCA and the right subclavian artery. The proximal portion of the left arch forms an innominate artery with two branches: the left CCA and the LSA (31). There isn't retroesophageal component.

Chest x-ray shows the presence of a right aortic arch with a descending aorta to the right of the midline. It is usually associated with congenital heart disease in 98% and Tetralogy of Fallot in 90% (4).

Type 2: Aberrant left subclavian.

It is the most common of the right vascular rings. There is an interruption of the left arch between the CCA and the LSA. The persistence of the ductus arteriosus or a ligament completes the vascular ring and leads to compression of the airway (5). It is usually associated with cardiac malformations as tetralogy of Fallot in 71%, CIA 21%, and aortic coarctation in 7% (4-5).

Four vessels arise from the right aortic arch in the following order: left common carotid artery, right common carotid artery, right subclavian artery and aberrant left subclavian artery (5). The proximal portion of the left arch forms the CCA that emerges as the first branch of the aorta, the distal portion of the left arch may persist and be enlarged (Kommerell's Diverticulum) and the LSA originates from this aortic diverticulum at the junction of the right arch and the DA(5).

The findings are similar to the double arch Aortic by conventional radiography, and depend to a large extent on the location of the ductus arteriosus, usually the ductus is on the left side and runs from the aortic diverticulum to the left pulmonary artery, forming an indentation in the posterior wall of the esophagus (5). Other findings include right paratracheal opacity with tracheal indentation, and loss of normal left aortic arch shadow (4). Often, aortic diverticulum can be visualized on the chest X-ray as a soft convex tissue density to the left of the trachea at the position of a normal aortic button(5).

Type 3: Right-sided aortic arch with isolated left subclavian artery

It is an infrequent alteration. There is an interruption of the left aortic arch at two levels, the first between left common carotid and left subclavian arteries and another one distal to the left ductus junction (5). The proximal portion of the left arch becomes the left common artery and arises as the first branch of the right arch followed by the right CCA and RSA.
The left subclavian artery joins the left pulmonary artery through the ligament or left ductus arteriosus (4-5).

4. Right-sided aortic arch with left descending aorta.

The descending aorta crosses from right to left posterior to the esophagus (4-5). A mass can be visualized as a mediastinal opacity, especially in elderly patients (7). The brachiocephalic artery could be sited in the left side.

**Left-sided aortic arch**

1. Aberrant right subclavian artery.

This is the most frequent anomaly of the aortic arch and is usually an incidental finding (8). There is an obliteration of the right ductus arteriosus so the RSA born distal to the left subclavian artery. The RSA surround the posterior wall of the esophagus and goes to the right arm (8-9). The frontal X-ray projection can be normal in up to 40% of the cases (8).

2. Compression syndrome by the innominate artery or the brachiocephalic trunk.

The origin of the brachiocephalic trunk is at the left side in 95% of the cases and cross obliquely the trachea and ascend through the mediastinum to the neck (10). A persistent anterior constriction of the trachea is usually seen in the lateral chest radiograph at the level of the thoracic operculum (8).

**Cervical aortic arch**

It is a rare malformation caused by the persistence of the third aortic arch which rises from the mediastinum to the neck, usually behind the esophagus and becomes the descending aorta (1). It is associated with anomalies of the vessels of the right aortic arch such as the left aberrant subclavian artery or aortic coarctation (1). The chest X-ray shows a high aortic opacity, and in the lateral projection there is a anterior displacement of the trachea (1).

**Pulmonary artery sling**

The left pulmonary artery arises from the right pulmonary artery, then cross and compress the posterior wall of the trachea and the anterior wall of the esophagus (8). Often the
distal trachea and carina are displaced to the left (8). It is the only vascular ring that leads to the anterior indentation in the esophagus (11). The most frequent radiological signs include the presence of a right tracheal indentation and there is usually hyperinflation of the right middle and lower lobe (2) or asymmetric insufflation of the lungs with a low position of the left hilum and an anterior arch of the lower part of the trachea (8).

Two types of pulmonary sling have been described; in type I the carina has a normal location between T4-T5 with the normal epiarterial bronchi (type IA) or a tracheal bronchus for the right upper lobe with hyperinflation of the right lung (type IB).

In type II the carina is caudal to T6 and is associated with stenosis of the intermediate bronchus (12). Type II is less favorable than other vascular rings due to association with anomalies such as complete cartilage rings in 60-80% (2) or other associated tracheobronchial malformations (types IIA and IIB) (10). If is associated with cartilaginous rings, there is a horizontal course and thickening of the main bronchus this finding is known as "T-shaped carina" (2). In the lateral projection a high density structure is identified between the trachea and the esophagus (2, 12).
**Fig. 1:** Aortic arch development based on the Edwards model. A. Anatomic representation showing the normal interruption zone (opaque) from a system with two aortic arches, regression of the right dorsal aortic root (between the RSA and the DA) and the right ductus arteriosus results in the normal left aortic arch. B. Schematic representation of the aortic archs with an arterial circle extending from the ascending to the descending aorta via two arches. Both common carotid and subclavian arteries emerge from each aortic arch. Gray lines represent the potencial locations of the ductus arteriosus.

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Fig. 3: Double aortic arch. Barium swallow A. Frontal images shows the reverse S indentation of the contrast column B. In the lateral view there is a posterior indentation in the esophagus wall (*).

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**Fig. 2:** Double aortic arch. A. Frontal chest X-Ray shows a "reverse S" shape of the trachea due to the location of the right (R) and left (L) aortic arches. B. Axial contrast CT, the Ascending aorta is anterior to the trachea and divides in an anterior left arch (LA, smaller) and a posterior right arch (RA), both arches enclose the trachea and the esophagus, and join to form the descending aorta.

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**Fig. 4:** Right-sided aortic arch with mirror image branching. A. There is an interruption of the left arch between the DA and the LSA (*). B-C. M, 16 years old with Tetralogy of Fallot. CT and MRI. The arch and the DA are on the right, the right arch has 3 branches: the innominate, the RCCA and the RSA. The proximal portion of the left arch forms an innominate artery (arrowhead) with two branches: the LCCA and the LSA. The innominate artery compress the trachea anteriorly. This patient also has a persistent left superior vena cava (red arrow).

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**Fig. 5:** Right aortic arch with left aberrant subclavian artery. A. Barium swallow that shows posterior compression and displacement of the esophagus (*). B. Axial image from contrast enhanced MRI shows the right arch and the retroesophageal course of the left subclavian artery. The dotted line shows the level of the interruption of the left arch between the left common carotid artery and left subclavian artery.

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**Fig. 6:** Right-sided aortic arch with isolated left subclavian artery. There is an interruption of the left aortic arch between left common carotid and left subclavian arteries and another one distal to the left ductus junction. The proximal portion of the left arch becomes the left common artery and arises as the first branch of the right arch followed by the right
CCA and RSA. The left subclavian artery joins the left pulmonary artery through the left ductus arteriosus (yellow line).

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**Fig. 7:** Right-sided aortic arch with left descending aorta. Enhanced thorax CT shows the descending aorta crossing from right to left posterior to the trachea (T) and the esophagus (E).

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Fig. 8: Left arch with aberrant right subclavian artery. A. Barium swallow shows compression of the posterior and left lateral wall of the esophagus (*). B. MIP projection shows the left arch with a right subclavian artery passing posterior to the esophagus. The dotted line shows the level of the interruption of the right arch between the right common carotid artery and right subclavian artery.

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**Fig. 9:** Cervical aortic arch A. Angiogram shows an elongated aortic arch extending above the level of the clavicles

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Fig. 10: Pulmonary artery sling with associated tracheal anomaly. MIP projection in the axial plane that shows the left pulmonary artery (yellow arrow head) arising from the posterior wall of the right pulmonary artery (black arrow head) coursing to the left lung posterior to the trachea (T) and anterior to the esophagus (E).

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Fig. 11: Pulmonary artery sling Type IB. A. Frontal chest X-Ray shows a hyperinsuflation of the right lung secondary a tracheal bronchus for the right upper lobe (arrowhead), B Angiogram in the main pulmonary artery shows the left pulmonary artery (LPA) arising from the posterior wall of the right pulmonary artery (RPA).

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Conclusion

The spectrum of anomalies of the aortic arch have been associated with vascular rings and other congenital cardiovascular diseases. Opportune detection and accurate characterization play an important role in their treatment. The Edwards model helps to understand the origin and potential manifestations of each of this conditions as well as its radiological presentation.
References


