

“ARTERIA LUSORIA”- ABERRANT RIGHT SUBCLAVIAN ARTERY EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

Ganesh Elumala and Muziwandile Bayede Mdletshe

Department of Embryology, College of Medicine, Texila American University, South America.

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ABSTRACT

Patients with (ARSA) are often asymptomatic and discovered accidentally during MRI scans and X-rays ordered by physicians. It is reported that (ARSA) has an incidence of 0.5 % to 2%. The aberrant artery in this anomaly follows a retro esophageal path and it rarely takes a pathway anterior to the trachea or esophagus. Instead of this artery being the first branch with the right common carotid on brachiocephalic artery, it arises on its own as the fourth branch, after the left subclavian artery. It can be associated with chromosomal defects like trisomy 21 and 18.

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Introduction

The most common embryologic anomaly of the aortic arch is aberrant right subclavian artery (ARSA). This anomaly is also known as *Arteria Lusoria* (AL). The first ever descriptive variation of ARSA was provided in 1735 by Hunauld a French anatomist born in Châteaubriant. The medical entity of Lusoria was firstly studied by Bayford. Bayford studied (ARSA) in the year 1787 on a woman patient found to have a long history of dysphagia which caused her difficulty in swallowing. This patient was found to have an aberrant right subclavian artery when an autopsy was done. This anomaly hence, it is also known as Bayford Autenrieth dysphagia.

The human aortic arch normally branching into three vessels patterns called the brachiocephalic trunk (BCT) or innominate artery, the left common carotid artery (LCCA) and the left subclavian artery (LSA) [Ganesh Elumalai et al., 2016]. However, when aberrant right subclavian artery anomaly is present, the brachiocephalic trunk tends to be absent and four large arteries arise from the arch of the aorta. These arteries then go as follows right common carotid artery and the left common carotid artery, the left subclavian artery, and the final one has the best distal left sided origin. The right subclavian artery then arises and is called the *Arteria Lusoria*. This blood vessel travels to the right arm then crossing the sagittal line of the body and usually passing behind the esophagus. If this artery compresses the esophagus it usually may produce a condition called dysphagia Lusoria. Frequently, the *Arteria Lusoria* arises from an aortic arch diverticulum at the proximal descending aorta, which was first described by Kommerell. Most patients with (ARSA) are asymptomatic. Rarely does (ARSA) cause dysphagia (Dysphagia Lusoria) and respiratory symptoms, which usually present in these decades.

Incidence

The most important anomaly of the aortic arch is arguably the presence of an aberrant right subclavian artery (*Arteria Lusoria*). If this vessel compresses the other structures within it reach, several symptoms may be produced by an individual some may be even fatal. Out of 141 cases which were looked at 15 were of cadaveric and 126 were medically documented. The gender dispersion of the subjects was 55.3% female and 44.7% male. The most common symptoms in this group of people were dysphagia (71.2%), dyspnea (18.7%), retrosternal pain (17.0%), cough (7.6%), and weight loss (5.9%). The vascular anomalies parallel with an *Arteria Lusoria* were Truncus Bicaroticus (19.2%), Kommerell's diverticulum (14.9%), cyst of the artery itself (12.8%), and a right side aortic curve (9.2%). ARSA is present to 0.5% to 2.5 % the world's healthy population. However, in this finding it was observed it was common in cases where patient had chromosomal abnormalities and of those cases approximately 30% fetuses with Down Syndrome and only 20 % of fetuses carrying both cardiac defects and chromosomal abnormalities.

Ontogenesis for normal aortic arch and its branching

The aorta develops during the the third week of gestation. The aorta develops in a complex process related to the formation of the endocardial tube at day 21. A primitive aorta is formed consisting of ventral and a dorsal segment that runs continuously through the first Aortic arch. Both the ventral aortae fuse to form aortic sac and the dorsal aortae fuse to form midline descending Aorta. The six paired aortic arches develop between the dorsal and ventral aortae and in addition many intersegmental arteries are given off by dorsal aorta.

The blood vessels derived from each of arch are as follows:

The first pair of blood vessels is assign to formation of both the external carotid and maxillary arteries. The second pair of blood vessels contributes to formation of the stapedial arteries.

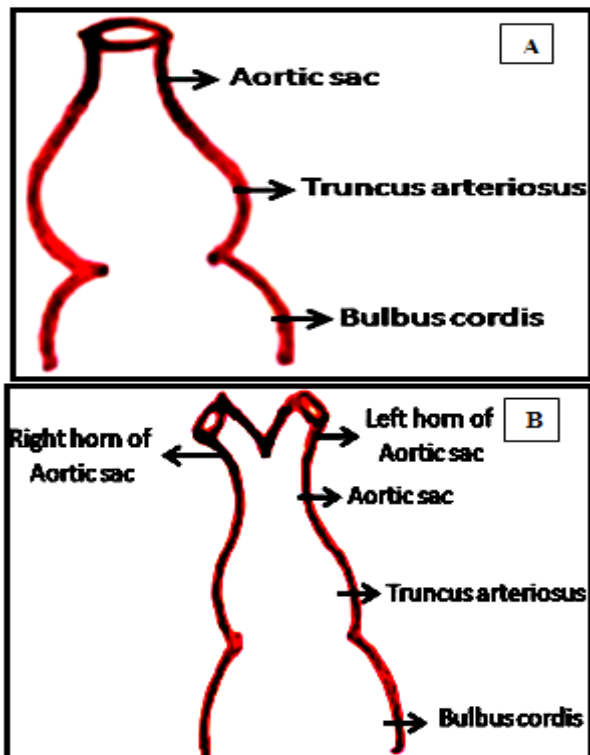


Fig1. The development process of Aortic sacs A. diagrammatic representation of the proximal part of the developing heart tube and B. During the later period, the Aortic sac shows its terminal branches called Right and Left horns.

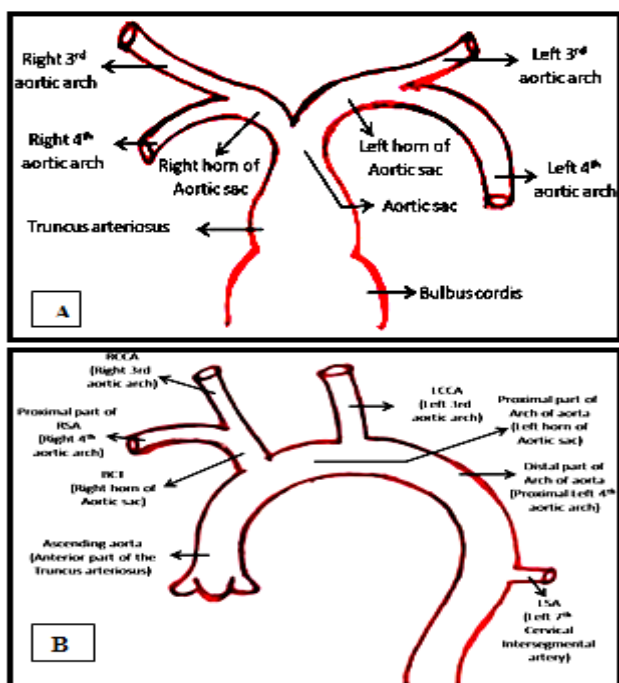


Fig 2.The derivatives of aortic arch arteries. A diagrammatic representation showing the Truncus arteriosus receives the third (III) and fourth (IV) sets (right and left) of Aortic arch arteries, ultimately it opens into the right and left horns of the Aortic sac and B. Derivatives of the Aortic sac horns and third (III) and fourth (IV) sets (right and left) of Aortic arch arteries.(BCT-Brachiocephalic trunk, RSA- Right subclavian artery, RCCA- Right Common carotid artery,

LCCA- Left Common carotid artery and LSA-Right subclavian artery).

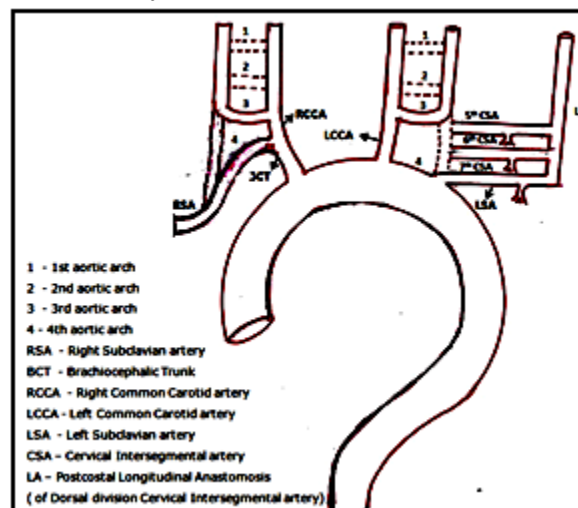


Fig 3.Diagrammatic representation of Aortic Arch and its branches.

The third aortic arch presents the commencement of the internal carotid artery and is known as the carotid arch. Proximal parts of the *third pair* form the common carotid arteries. Together with sections of the dorsal aortae, the distal portions provide to formation of the internal carotid arteries.

The left arch of the *fourth pair* produces the sections of normal left aortic arch between the left common carotid and subclavian arteries. The right fourth arch forms the proximal right subclavian artery whereas, the distal part of the right fourth arterial arch gets regressed. The distal right subclavian artery is derived from a portion of the right dorsal aorta and the right seventh intersegmental artery.

Rudimentary blood vessels that regress early develop out of the *fifth pair*. The left arch of the *sixth pair* provide to the formation of the main and left pulmonary arteries and ductus arteriosus, this duct obliterates a few days after birth has taken place. The right sixth arch provides to formation of the right pulmonary artery. With the caudal migration of the heart in the second fetal month, the seventh intersegmental arteries increase in size and migrate cephalic to form the distal subclavian arteries.

The left subclavian artery is derived mostly from the left seventh intersegmental artery.

Malformations of the aortic arch system can be defined by enduring nature of sections of the aortic arches that usually continue or disappearance of sections that normally remain it can be possible both.

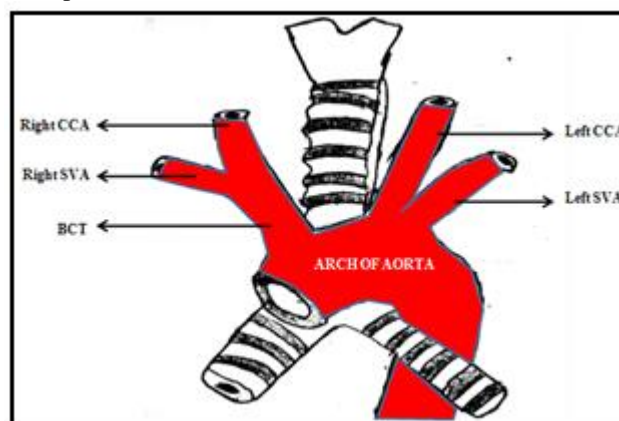


Fig 4.Diagrammatic representation of normal aortic arch.(CCA-Common carotid artery; SVA-Subclavian artery and BCT-Brachiocephalic trunk).

Ontogenesis for the Arteria Lusoria

The congenital anomalies are the most common causes for the death in infants under one year of age [Ganesh Elumalai et al., 2016]. Aberrant right subclavian artery is caused by the involution of the right fourth vascular arch and proximal right dorsal aorta. The obliteration of right fourth aortic arch along with the persistence of the proximal portion of the right dorsal aorta with the right seventh intersegmental artery will results in the “*Arteria Lusoria*”. The abnormal right subclavian artery crosses the midline behind the esophagus and may compress it.

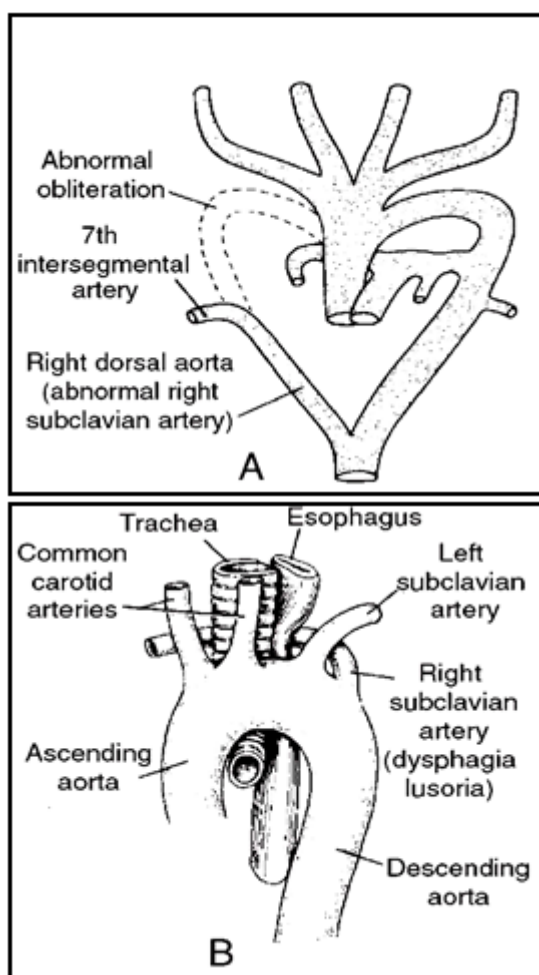


Fig 5. Diagrammatic representation of A. Formation and B. Course of aberrant right subclavian artery.

Discussion

The human aortic arch, normally branches into three vessels patterns called the brachiocephalic trunk (BCT) or innominate artery, the left common carotid artery (LCCA) and the left subclavian artery. (LSA) [Jakanani et al., 2010; Natsiset al., 2009; Boyaciet al., 2015; Budhirajaet al., 2013]. Vascular variations are congenital morphological differences that arise in the human body. Although, for the most part, do not cause injury to the individual, may be important in cases where it is necessary a specific access to the vascular system [Ganesh Elumalai et al., 2016]. The occurrence of arteria

lusoria in analysis is found to be more common in female than male subjects (55.3% versus 44.7%), which is similar to the results recorded and given by Molz and Burri, who note that this anomaly was prevalent more often in females (58%) than males individuals (42%). It is reported that the aberrant right subclavian artery has a very high female predominance.

According to a current bibliographical search, the symptoms of Lusoria compression have been found to be present only in 7 to 10% of adult patients with the anomaly. So it safe to say the anomaly is clinically silent in 90–93% of cases of people with (ARSA). The presence of (ARSA) is also higher in disorders such as Downs, DiGeorge, and Edwards' syndromes. When (ARSA) is present it occurs at the two extremes of life. In infants and children, tracheal obstruction or dysphagia can usually occur. The increased rate of pulmonary infections seen in infants is said to be due to the absence of tracheal rigidity. It is found that 86% of infant patients with ARSA had symptoms of stridor or recurrent respiratory diseases. In infants the trachea is compressible therefore the typical syndrome compression by lusoria are respiratory, such as wheezing, stridor, recurrent pneumonia, and cyanosis while in adults the trachea is more rigid so respiratory symptoms are rare. In an adults individual a congenital vascular anomaly of the aortic arch and its branches is a cause of dysphagia, classically known as “dysphagia lusoria”. A Barium contrast medium examination of the esophagus, showing us a possible characteristic diagonal compression defect at the level of the T3 and T4 vertebrae, is an excellent tool for diagnosing ARSA condition. New imaging techniques are used now such as magnetic resonance imaging and this will contribute to better visualization, especially when a aneurysm is present in the proximal part of the artery [Ogeng'oJAet al., 2010; Patilet al., 2012].

Aberrant right subclavian artery (ARSA) is a very rare form of congenital anomaly that asymptomatic. Symptomatic patients will therefore require surgical intervention. When surgical treatment is needed, a lot of consideration should be given to anatomic re-establishment of orthograde flow into the right subclavian artery. This will avoid sacrificing direct blood flow to the patient's right arm and therefore preventing possible complications. Many surgical approaches can be implemented, such as medial sternotomy, thoracotomies of both left and right side and supraclavicular incision. These surgical practices have been used for the fixation of ARSA. The surgical approach to fix this anomaly has been very controversial.

Conclusion

The clinical relevance from this study to create an algorithm or define a set of factors to alert surgeon and anesthetist's to aware of knowledge [Ganesh Elumalai et al., 2016] about this anomaly. It is very vital to remember that common conditions such as shortness of breath, retrosternal pain, this pain, as is usually described by the patients, stands in closer sense for complaints [Ganesh Elumalai et al., 2016], cough, and weight loss may be major alert symptoms of the compression of adjacent structures by the arteria lusoria. The presence of the ARSA, especially, should be considered to distinguish in between from other causes of dysphagia. the knowledge of such variations may be important for surgeons to reach up to surgical intervention [Ganesh Elumalai et al., 2016].

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