

“DOUBLE AORTIC ARCH”

EMBRYOLOGICAL BASIS AND ITS SURGICAL IMPLICATIONS

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ABSTRACT

Double aortic arch is class of congenital anomalies caused by a chromosome band 22q11 deletion, which are often referred to CATCH-22 syndrome or chromosome band 22q11 deletion syndrome using the unified terms. it is responsible for formation of Double aortic arch ,both right and left aortic arch arise from the ascending aorta common form of complete vascular ring, encircling both the esophagus and trachea are compressed by the aortic arches and their derivatives, resulting in non-cardiac morbidity. This study aimed to through an insight on the embryological basis, its clinical and surgical implications on the double arch aorta.

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Introduction

Double Aortic Arch (DAA) is a type of congenital defect of an embryonic aorta development .normal development, each primitive arch either progression into a functional vascular structure or either involutes. In Double aortic arch develops due to the persistence of the fourth right position and left arches and dorsal aortas, resulting in the malformation of complete vascular rings that encircling trachea and esophagus [1]. DAA is a splitting of the ascending aorta into 2 segments, which pass on the both side of the esophagus and trachea and join together in a single descending aorta. In the most common variation, both the aortic arch segments are patent ,the right segment is larger (posterior) or dominant than the left segment (anterior) is smaller or hypoplasia ,the right arch is behind and the left arch is in front of the trachea and esophagus [2]. The two arches joins to form the descending aorta which is mostly left sided (but maybe right sided or in the midline. Less frequently, end of smaller left aortic arch closes (atretic left arch), and the vascular tissue is represented by a fibrous cord [2].

Human genetic studies suggest that chromosome 22q11 deletions may be due to the important cause of abnormality development of the aortic arch. Similarly recurrence of the double aortic arch has been noted, a genetic causes for this aortic anomaly [3]. The signs and symptoms are related to the compression of the wind pipe and esophagus or both by the complete vascular ring. Mostly the patients are presently suffering with stridor (harsh vibrating noise while breathing) recurrent respiratory infections or dysphagia during the first 6 month of life. Diagnosis are made by barium esophageal, chest x-ray Echocardiography, Computed Tomography (CT), Magnetic Resonance Imaging (MRI) these tests shows the relationship of Aortic arches to the trachea and esophagus also the degree of tracheal stenosis. Bronchoscopy can be useful in internally assessing the degree of Tracheo malacia, tracheomalacia is often occurs in newborn due to the poor

development of cartilage in trachea [3]. The presentation of windpipe or trachea was floppy, soft and weak.

Treatment is surgical and is indicated in all symptoms that seen in the patients. In the current findings the risk of mortality or significant morbidity of the surgical division of the lesser arch is low. And the preoperative stage of Tracheo malacia has an important impact on postoperative recovery [4]. In certain patients it may take several months upto 1 to 2 years for the obstructive respiratory symptoms like wheezing to disappear.

Incidence

Complete vascular rings comprise an estimated of 0.5 – 1.0 % of all Congenital Cardiovascular malformation. The majority of these are double aortic arches [4]. In most surgical series, 45-65 % of patients undergoing repair of vascular ring have double aortic arch. There is no known gender preference, that is the male and female are equally affected. There is also no known ethnic or geographic predisposition cardiovascular anomalies that are associated with Double aortic arch are found in about 10-15 % of the patients [5].

Ontogenesis for the normal development of Left aortic arch

Martin heinrich rathke was a German embryologist, credited with much of work leading to the understanding of the normal embryological development of the branchial arches. The development of the branchial apparatus begins at the second week of gestation and it is completed by seven weeks [5]. The apparatus consists of six branchial arches in the wall of the anterior part of the gut. The branchial arches are numbered one to six from cephalad to caudad. Then each of the branchial arches connects paired dorsal and ventral aortas. In reality the arches appear and disappear at different times [5-6]. arches connects paired dorsal and ventral aortas. In reality the arches appear and disappear at different times [5-6].

The six branchial aortic arches normally develop into the thoracic aorta its branches. The first two arches intricate before development of the 6th arch, and the 5th arch is atretic or never fully develops. The 3rd arch and portions of the ventral and dorsal aortic arches forms the head and neck arteries [6]. The 4th arch becomes the aortic arch on the left and a part of subclavian artery in the right side, and the right and left pulmonary arteries develop from the 6th branchial arches from both the sides. At right side the dorsal contribution of the 6th arch disappears, and at the left it persists as the ductus arteriosus. The cervical seventh intersegmental arteries migrate and form the subclavian arteries [7].

Normally, the left arch aorta develop from the following sources,

- First part: Left horn of truncus arteriosus – form the portion between the ascending aorta and the brachiocephalic trunk
- Second part: Left 4th arch artery - form the portion between the brachiocephalic trunk and left common carotid artery
- Third part: Part of left dorsal aorta - form the portion between the left common carotid artery and left subclavian artery.

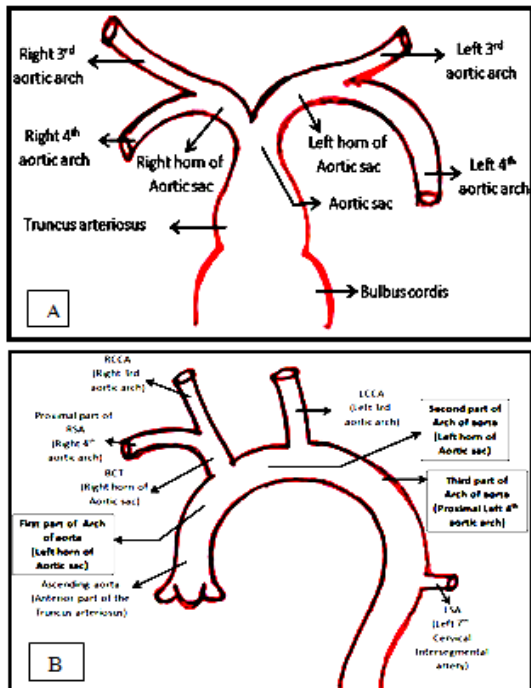


Fig 1. The derivatives of aortic arch arteries A. schematics showing the Truncus arteriosus receives the third (III) and fourth (IV) sets (right and left) of Aortic arch arteries, ultimately it opens into the right and left horns of the Aortic sac and B. Different source and development of left aortic arch.

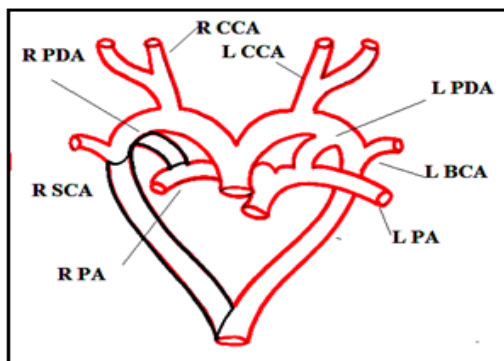


Fig 2. The above Schematics showing the Segments of the pharyngeal arch system that regress (shown in black) in

the normal formation of the thoracic great arteries. (Primitive Pharyngeal arch left (L) and right (R) external carotid (EC), internal carotid (IC) pharyngeal arches – (IV) and (VI) pharyngeal arches Dorsal Aortas (DA), intersegmental arteries (VII)).

Ontogenesis for the anomalous development of Double aortic arch

On observing the bilateral system of pharyngeal arch vessels in the early embryo during embryonic morphogenesis, the six pairs of pharyngeal arch arteries develop in conjunction with the branchial apparatus [8]. First through sixth arches appear in sequential with left-to-right symmetry, and constitute the primitive vascular supply to the brachiocephalic structures, running from the aortic sac to the paired dorsal aortas [9]. As normal cardiovascular morphogenesis proceeds, a patterned regression and persistence of the various arches and right-sided dorsal aorta occur resulting in the mature configuration of the thoracic aorta and its branches. The 3rd and 4th aortic arches along with the 7th intersegmental arteries and the left dorsal aorta, are the primary contributors to the normal aortic [9-10].

The connection between the distal part from the 4th arch on the right and the dorsal aorta is lost and it soon disappears. Bilateral or double aortic arch is formed when both 4th arches and both dorsal aortas remain present [10].

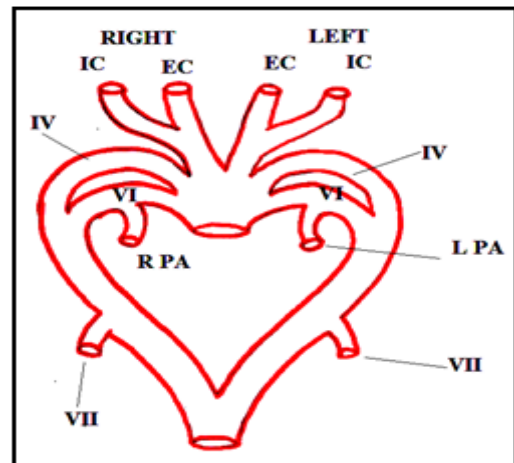


Fig 3. The above schematics diagram showing) sixth arches develop into the proximal pulmonary arteries and the distal 6th arches become the ductus arteriosus; Cervical (VII) intersegmental arteries develop into the subclavian arteries. (Primitive Pharyngeal arch left (L) and right (R) external carotid (EC), internal carotid (IC) pharyngeal arches, (IV) and (VI) pharyngeal arches dorsal aortas (DA), intersegmental arteries (VII)).

Discussion

Aortic Arch anomalies, classification are based on the absence, position, or course of the aortic arch. Due to arch anomalies may be characterized based on the order or pattern of branching of the great vessels, and also arch anomalies may be characterized as interrupted, right sided, left sided, or double in configuration, Anomalies may be left-sided, right-sided, double, or cervical aortic arch [11-16].

Many arch anomalies are asymptomatic, Aortic arch anomalies can also be classified by their morphology or clinical presentation [17]. But, anomalous isolation of the subclavian, brachiocephalic arteries or carotid, and vascular rings can cause clinical symptoms. On the base of the clinical presentation arch anomalies can be classified into, 1) asymptomatic cases, 2) cases with clinical symptoms caused by tracheobronchial and also may or may not be by

esophageal compression by vascular rings, 3) cases in which there is isolation of aortic arch branches and alteration of the normal blood flow with a “steal” phenomenon from the cerebral circulation [18-20].

Double Aortic Arch (DAA) is a common congenital aortic anomaly. The most common of the complete vascular rings, in which the trachea and esophagus are compressed by aortic arches and their derivatives [21-22]. Recognition of anomalies of aortic arch began in 1737 when Hommel first described DAA, is due to the presence of right and left aortic arches encircling and compressing the trachea and esophagus. The first successful surgical correction of DAA was performed by Gross in 1945 on a 1-year-old boy at Boston Children's Hospital through a left anterolateral chest approach and he further elucidated the principle involved in the surgical division of vascular rings [22-26]. The International Congenital Heart Surgery Nomenclature and Database Committee have classified DAA into three categories based on dominance as Right aortic arch, Left aortic arch or Balanced arches. Of which balanced type of DAA is the least common [27].

There is only one left aortic arch normally. In DAA the ascending aorta arises normally from the left ventricle but then as it exists the pericardium it divides into 2 right and left aortic arches which encircle the trachea and esophagus and join posteriorly to become the descending aorta [28]. DAA has various forms when arches may be patent the right or left arch may be larger, or they may be similar in size, or an atretic segment may be present at one of the several locations in either of the arch. The ligamentum arteriosum or ductus arteriosus runs between the left pulmonary artery, which is inferior to the junction between the left aortic arch and the descending aorta. The aortic arches give rise to the ipsilateral subclavian and common Carotid arteries and the innominate artery is absent [29].

DAA patients present with symptoms which range from nonspecific complaints to life-threatening respiratory distress or can be asymptomatic. Symptoms of stridor a harsh vibrating noise when breathing caused by obstruction of the larynx [30]. Dysphagia is the difficulty or discomfort in swallowing, Chronic barking cough, bronchopneumonia susceptibility, head retraction, onset during early infancy, seen increase in respiratory distress during feeding were described by Wolman (1939).

Chest radiography (chest x-ray), barium esophagography, echocardiography, medium chain triglycerides (MCT), magnetic resonance imaging (MRI), angiography and conventional cardiovascular angiography is used for the diagnosis of DAA. Echocardiography is recommended to rule out associated congenital cardiac defects [30]. Barium esophagogram shows the bilateral indentation of the esophagus. The development of cross-sectional imaging has facilitated the accurate and right diagnosis and surgical treatment of compression caused by vascular anomalies. MCT helps in highly accurate preoperative evaluation of the vascular anatomy and its relationship to the trachea and esophagus. Due to the presence of separate common carotid and subclavian arteries on both sides, four vessel sign can be seen in the superior mediastinum [29].

Conclusion

There is no predilection in sex has been documented in patients with double aortic arch. Due to limited data, no racial predilection is apparent. Double aortic arch can be corrected by surgery [31-32]. Long-term prognosis for patients with

repaired double aortic arch is excellent. Most common adverse outcomes are persistent respiratory symptoms.

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