



“RIGHT SIDED AORTIC ARCH” EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

The growth of the aorta and its major branches are from six pairs of branchial arches, which, throughout the fourth week of pregnancy, combine the primitive ventral and dorsal aortas in the embryo. The right-sided aortic is identified when the aortic arch courses to the right of the trachea. When this happens, there is interchanging of the descending aorta and ascending aorta to the right and left respectively. In abnormal condition it can be interruption dorsal segment which may result in any of the three types of right-sided aortic arch. In anomalies which are diagnosed earlier can result in the management of the condition.

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Introduction

The growth of the aorta and its major branches are from six pairs of branchial arches, which, throughout the fourth week of pregnancy, combine the primitive ventral and dorsal aortas in the embryo. Between day 26 and 29, vasculogenesis and angiogenesis result in the development of six pairs of aortic arches from aortic sac. These arteries serve to link the emergent heart with the dorsal aorta. During the fourth and fifth weeks of embryological development, when the pharyngeal arches form, the aortic sac brings forth arteries – the aortic arches. [Ganesh Elumalai et. al., 2016] The aortic sac is the endothelial-lined dilation just distal to the truncus arteriosus; it is the primordial vascular canal from which the aortic arches arise. In humans, these six branchial arches become transformed into the permanent major vessels (the aorta and its main branches) throughout the 5th to the 7th week of embryologic development. The first pair form between day 22 and 24; both arches revert as the second arch forms on day 26. The first three arches target to supply the cranial and cervical sections and form the internal carotid artery among others. The external carotid artery arises later as a separate vessel. The fourth aortic arch develops unsymmetrical. The left fourth arch forms the proximal part of the adult aortic arch, whereas the right one develops into the right subclavian artery. The last two arches, 5 and 6, do not appear in large arch shape. The ductus arteriosus is produced by a left dorsal branch from the dorsal aorta and links the aortic sac with the pulmonary arteries as a shunt. The left subclavian artery is derived from the left seventh intersegmental artery. The aortic valve is derived from conotruncal ridge tissue and forms three triangular valve leaflets.

The carotid arteries are derived from the third pair of arches. Each pharyngeal arch has its own cranial nerve and its own artery; therefore we can conclude that the development of

the aortic and pharyngeal arches are very directly connected. The aortic arches terminate in the right and left dorsal aorta. The dorsal aorta remains paired in the region of the arches, however below this region they fuse to form a single vessel (the descending/thoracic/abdominal aorta).

The pharyngeal arches and their vessels appear in a cephalo-caudal order, so are not all present at the same time. As a new arch form, the aortic sac contributes a branch to it. Right-sided aortic arch is a type of aortic arch identified by the aortic arch coursing to the right of the trachea. There are three classifications of right-sided aortic arch;

Type-I: a Right-sided aortic arch with mirror image branching.

Type-II: a Right-sided aortic arch with the aberrant left subclavian artery.

Type-III: a Right-sided aortic arch with separation of the left subclavian artery.

It is a very uncommon congenital malformation which develops during the 4th and 5th weeks of embryogenesis.

Incidence

This anomaly has been reported to occur in 0.05–0.1% of the general population and about half of these cases are related with an aberrant left subclavian artery. [Du ZD, et. al., 2002]

Ontogenesis of normal development of Arch of aorta

In the development of the aortic arches, the aortic sac receives six right and left sets of Aortic or brachial arterial arch. These arterial arches go through selective apoptosis and the residual branch vessels comprise the development of Aortic arch and its great vessels. Any variations in this normal process will effect in the anatomical variance. The first and second sets of the right and left arterial arch I and II typically get revert. The third pair of right and left arterial arches forms the proximal part of the common carotid arteries bilaterally. The proximal part of the right fourth arterial arch persists as the right subclavian artery up

zto the origin of the internal thoracic artery, while the distal part of the right fourth arterial arch reverts. The distal part of the left fourth arterial arch regresses and its proximal part form a small section of the adult Aortic arch between the origin of the left common carotid artery and the left subclavian arteries. The right and left, fifth arterial arch either regresses or partly formed. The proximal part of the right and left sixth arterial arch forms the pulmonary arteries. The distal part of the right side sixth arch, becomes ductus arteriosus, while in the left side distal part will regress completely (Komiya et al., 2001).

The right horn of the Aortic sac forms the brachiocephalic trunk and the left horn of the Aortic sac usually forms the part of the Aortic arch prevailing between the origins of the brachiocephalic trunk and the left common carotid arteries. Generally, the anterior part of the Truncus arteriosus receives the third and fourth sets of right and left arterial arches. Eventually, it opens into the right and left horns of the Aortic sac. The posterior part of the Truncus arteriosus receives the sixth set of right and left arterial arches, and forms the right and left pulmonary arteries. The formation of the spiral or Conotruncal septum divides the Truncus arteriosus into the anterior ascending aorta and the posterior pulmonary trunk. The anterior part of the Truncus arteriosus continuous above as the Aortic sac, where it links with the third and fourth sets of right and left Aortic or branchial arch arteries. Ultimately, the aortic sac and its horns receive all the derivatives of third and fourth sets of right and left Aortic or branchial arches.

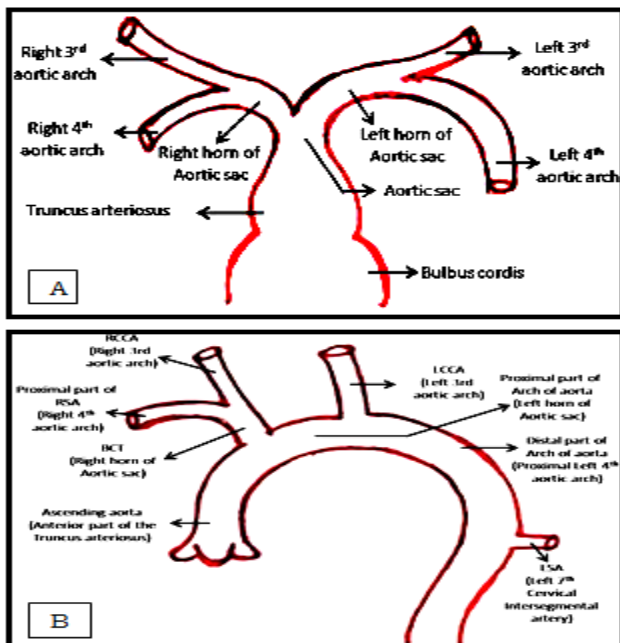


Fig 1. The derivatives of aortic arch arteries A. schematics showing the Truncus arteriosus receives the third (III) and fourth (IV) right and left Aortic arch arteries, which opens into the right and left horns of the Aortic sac and B. Derivatives of the Aortic sac horns and third (III) and fourth (IV) right and left 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) [3].

Ontogenesis of the abnormal development of Right-sided aortic arch.

The embryological origin consists of an abnormal development of the fourth branchial arch. Normally, the aortic

arch is formed by the left fourth aortic arch and the left dorsal aorta. If the right dorsal aorta persists and the distal left aorta disappears, then we have a right aortic arch.

The three-vessel and trachea view is part of the routine exploration of the fetal heart. In the case of the right sided aortic arch, we don't see the normal V shape configuration, because there is a gap between the ascending aorta and the main pulmonary artery. The aortic arch is located to the right of the trachea instead of its normal left position. There are three classifications of right sided aortic arches

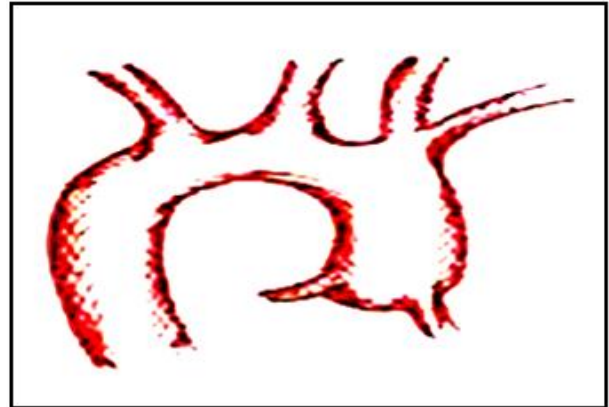


Fig 2.Type I: Right-sided with aortic arch with mirror image branching; it grows from the interruption of dorsal segment of the left arch linking the left subclavian artery and descending aorta, with reversion of the right ductus arteriosus in hypothetical double aortic arch.



Fig 3.Type II: Right-sided aortic arch with aberrant left subclavian artery; it grows from the interruption of dorsal segment of the left subclavian artery with reversion of the right ductus arteriosus in hypothetical double aortic arch.



Fig 4. Type III: Right-sided aortic arch with isolation of the left subclavian artery; develops from the interruption of the left arch of second branches, with first branch between the left common carotid artery and left subclavian artery and the other branch distal to the attachment of left ductus.

Discussion

Development of the aorta and the aortic branches commences between the fourth and seventh week of pregnancy. Six pairs of aortic segments grow and the third pair forms the common and internal carotid arteries. The fourth segment on the left continues to develop the adult aortic arch. The other segments disappear. Persistence of the fourth arch on the right results in the collection of arcus aortae dexter. Conversely, the corresponding segment on the left disappears. A maintenance of both aortic segments results in an aortic ring. Those aortic rings are typically diagnosed in infancy because of airway obstruction or complications in swallowing. In the common type II right-sided aortic arch the left subclavian artery has to cross the mediastinal structures. Although the underlying cause of persisting right-sided arch is not completely known so far, in 24% a deletion of chromosome 22q 11 is associated. A right sided arch does not automatically cause symptoms, but if the physiological abnormalities explained are present, the diagnosis of vascular ring is almost certain. Three types of right sided aortic arch can be distinguished: those with "mirror image branching," in which a left innominate artery arises as the first branch off the aortic arch, dividing into a left common carotid and left subclavian artery, those with an aberrant left subclavian artery, which arises as a fourth branch off the aortic arch and those with isolation of the left subclavian artery. The first type is often associated with cyanotic heart disease, whereas the second one is usually associated with a normal heart. An aberrant left subclavian artery can be confirmed by barium swallow but is not of itself pathological and may be a normal variant in 1% of the population. A left sided ductus arteriosus (or ligamentum arteriosum) completes the ring, and it is this combination that can cause symptoms. The third type rarely exists; it is 0.005% of the population. If the significance of these vascular abnormalities remains uncertain, a fiberoptic bronchoscopy allowing direct examination of the airway should be carried out. Surgery involves dividing the ligamentum arteriosum, thereby splitting the constricting circle. Freeing up the neighboring connective tissues from the trachea, esophagus, and aorta further improves this procedure. After surgery, secondary tracheomalacia is almost always there after the prolonged period of airway compression. Symptoms may not be completely resolved, and abnormalities of the flow volume loop may continue.

Delayed diagnosis of a vascular ring can end in needless investigations and prolonged periods of unsuccessful treatment. Such treatment may be unsafe. Inhaled corticosteroids are an efficient treatment for asthma but may have adverse consequences on growth, bone density, and adrenal function if used incorrectly. Delayed airway compression may also be harmful. Identification of the right diagnosis, with following surgery, leads to a progress in symptoms and withdrawal of needless treatment. Consideration of alternative diagnoses, with careful examination of the chest x-ray film, is essential in the supervision of children with persistent respiratory symptoms. [Ho VB et. al., 1998]

Conclusion

The growth of the aorta and its main branches are very important in the development of the heart. In the three types of right-sided aortic arch, the most common is the aberrant left subclavian artery is not pathological but the presence of ductus arteriosus completes the ring and makes it show symptoms. There are many variations in the development of the heart and

it is still unclear the causes of right-sided aortic arch even though the deletion of chromosome 22q11 is associated with it. With early diagnose it can be manageable.

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