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"ABNORMAL ORIGIN OF THE RIGHT SUBCLAVIAN ARTERY FROM THE RIGHT PULMONARY ARTERY" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Isolation of the right subclavian artery occurs when the right subclavian artery takes origin from the pulmonary artery through a ductus arteriosus. The right subclavian artery normally takes origin from the brachiocephalic trunk, specifically the brachiocephalic artery, however due to this anomaly it takes origin from the pulmonary trunk, specifically the right pulmonary artery. As a result of this aortic arch malformation, there is a continuous filling of blood of blood into the right pulmonary artery; this phenomenon is known as subclavian steal phenomenon. As blood continues to flow in the right pulmonary artery, it gives rise to other complication s such as pulmonary over circulation.

Introduction

Subclavian artery is the artery, which is located within the thoracic region of the human body [1]. There are two types of subclavian arteries, both of which are located behind the clavicle, specifically behind the left Sternoclavicular joint. The left and right subclavian arteries are both responsible for supplying the brain, upper limbs and chest region with oxygenated blood [2-3].

On the left side it originates from the arch of aorta and then moves upward in the neck just below the clavicle [3]. The right subclavian artery takes origin from the brachiocephalic artery, which also ascends upto the sternoclavicular joint which is located just below the clavicle [4-5]. The right subclavian artery is divided into three parts, all of which have relations to the scelanus anterior, which a scalene muscle is found in the cervical region in the neck [6]. The first part is located medially to the scelanus anterior, the second part is located posteriorly or behind of the scelanus anterior and the third part is located laterally to the scelanus anterior connecting to the lateral border of the first rib [7].

The isolation of the subclavian artery is a very rare anomaly, whereby the subclavian artery arises from a pulmonary artery instead of aortic arch [8]. The isolated subclavian artery is connected to the pulmonary artery either by a closed or patent ipsilateral ductus arteriosus.

This form of anomaly usually involves the left subclavian artery and is usually involved with Intracardiac or aortic arch anomaly. The abnormal origin of the right subclavian artery from the pulmonary artery is an extremely rare congenital anomaly that occurs in the aortic arch [8-10].

Incidence

The epidemiological data pertaining to the isolated right subclavian artery is reported to range from 0.4% to 1.7% of the population every two years [7-10].

Ontogenesis for normal aortic arch and its branching

The aorta develops during the third week of gestation. The aorta develops in a complex process related to the formation of the endocardial tube at day 21 [11]. 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 [12].

Embryological development of aortic arch begins at the third week of gestation, the proximal part of the right subclavian artery originates from the fourth aortic arch of the right dorsal aorta, which is located between the fourth and seventh intersegmental artery [13-15]. The remaining part of the right subclavian artery, which consists of the distal portion, is originated from the seventh intersegmental artery [14]. The left subclavian artery completely originates from the seventh intersegmental artery, which arises from the dorsal aorta opposite the attachment of the fourth aortic arch. As embryological development continues, the point at which subclavian artery is originated from the aorta will slowly move upward to lie adjacent to the common carotid artery [16]. In normal embryonic development, the subclavian arteries, which supply the head, neck and upper limbs, are formed mainly by the relocation and dissolution of the pharyngeal arches. These pharyngeal arches proceeds symmetrically to reach the dorsal aorta. In the developmental process the components are

formed within the distal portion of the outflow tract of the pulmonary artery [17]. The remaining pericardial components of both the left and right pulmonary artery are formed with the pharyngeal mesenchyme of the sixth aortic arch. The intersegmental artery migrates during the normal development and eventually becomes the right subclavian artery, which takes origin from the brachiocephalic trunk, which arises from the fourth aortic arch [18]. The distal components of the fourth and fifth aortic arches regress, which then leads to the right fourth arch becoming the definitive aortic ach and the fifth aortic arch is then proceeds to form the right ductus arteriosus [18-19].

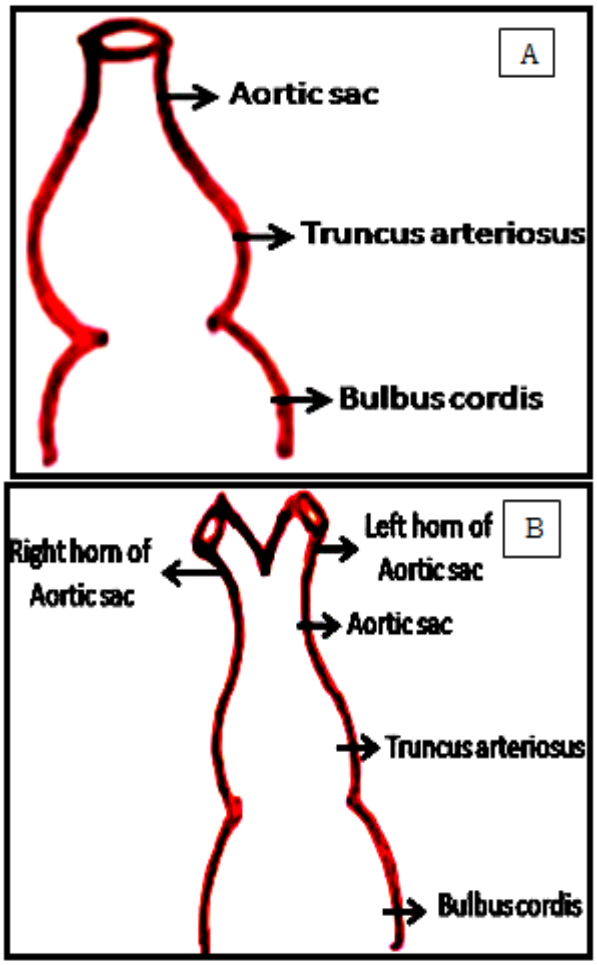


Fig 1. The development process of Aortic sacs A. diagrammatic representation of the proximal part of the developing heart tube and B. During the later period, the Aortic sac shows its terminal branches called Right and Left horns.

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 [20]. The second pair of blood vessels contributes to formation of the stapedial arteries. The third aortic arch presents the commencement of the internal carotid artery and is known as the carotid arch [20].

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 [21-23].

The left arch of the fourth pair produces the sections of normal left aortic arch between the left common carotid and subclavian arteries [23]. 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 [24].

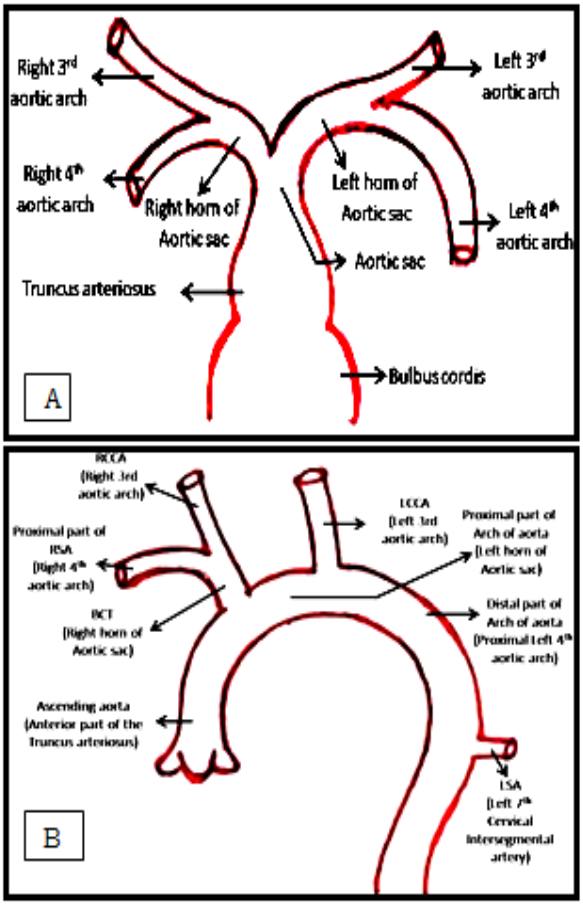


Fig 2. The derivatives of aortic arch arteries. A diagrammatic representation showing the Truncus arteriosus receives the third (III) and fourth (IV) sets (right and left) of Aortic arch arteries, ultimately it opens into the right and left horns of the Aortic sac and B. Derivatives of the Aortic sac 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).

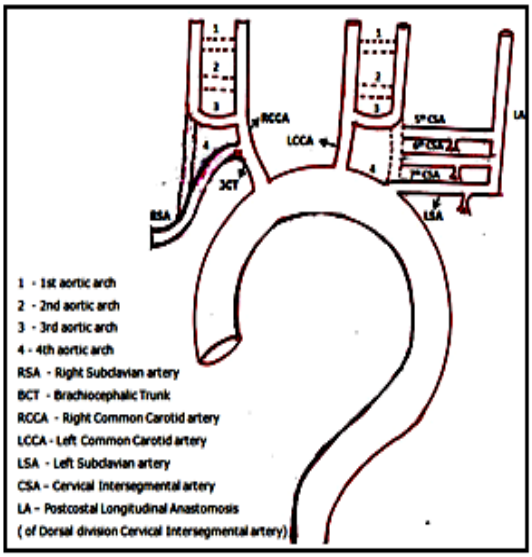


Fig 3.Diagrammatic representation of Aortic Arch and its branches.

Rudimentary blood vessels that regress early develop out of the fifth pair [25]. 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 [26]. 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 [26].

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 [27-28].

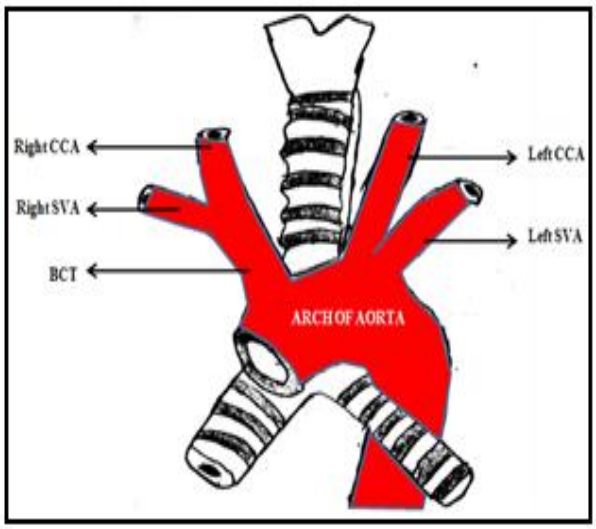


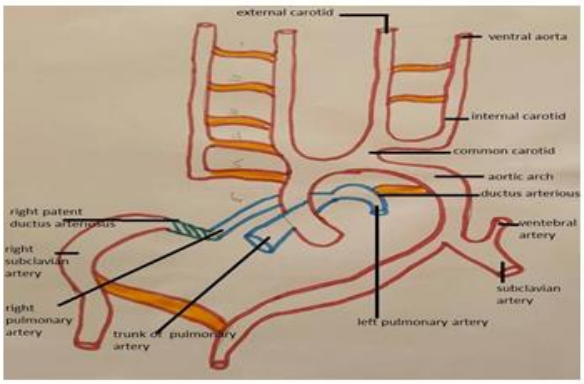
Fig 4. Diagrammatic representation of normal aortic arch. (CCA-Common carotid artery; SVA-Subclavian artery and BCT-Brachiocephalic trunk).

Ontogenesis of the abnormal development of the right subclavian artery

The development of the aortic arch occurs during the third week of gestation. The anomalous origin of the right subclavian artery usually occurs when the right fourth aortic arch and the right dorsal aorta, which locates cranial to the seventh intersegmental artery regresses abnormally [28].

Normally the right subclavian artery originates from the right fourth aortic arch and the right dorsal aorta, which proceeds distally to the seventh intersegmental artery. In the case of the abnormal development of the right subclavian artery, it is dissolute from the right fourth aortic arch and forms a connection at the distal end of right sixth aortic arch and forms continuity with the distal end of the right pulmonary artery via a ductus arteriosus [29]. The right subclavian artery then begins to move distally. The right fourth aortic arch has no connections pertaining to the right subclavian artery in this anomaly. The right fourth aortic arch begins to dissolute and the subclavian artery loses it connection, the subclavian artery then persists to the sixth aortic arch forming the ductus arteriosus and connection with the right pulmonary artery [29]. The right subclavian artery then proceeds from its proximally distally to the portion where it is said to supply the head, neck and upper limbs of the body [30].

Due to the right pulmonary artery which transports deoxygenated blood to the lungs making a connection with the right subclavian artery which supplies oxygenated blood upper limbs, a patient with this anomaly can run into sever complications, which causes swelling and over saturation in face, upper limbs and chest regions of the body [31].



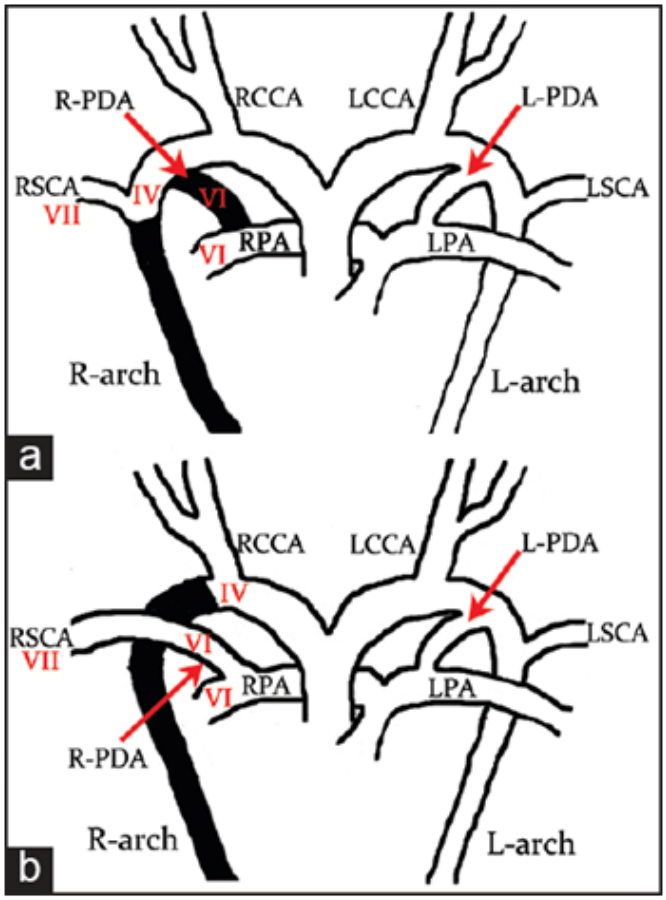


Figure 5. (a) Diagram of the normal embryological origin of the RSCA from the seventh (VII) intersegmental artery with subsequent cranial migration, and dissolution (in black) of the R-PDA and right fourth (IV) aortic arch (R-arch). (b) Isolation of the RSCA occurs when there is dissolution (in black) of the right IV aortic arch but persistence of the right sixth (VI) arch from which the R-PDA and RPA take their origin. L-arch = Left aortic arch, LCCA = left common carotid artery, L-PDA = left ductus arteriosus, LSVC = left subclavian artery, LPA = left pulmonary artery, RCCA = right common carotid.

Discussion

Isolation of the right subclavian artery is an anomaly whereby the subclavian artery develops at the level of the right sixth aortic arch and forms a connection to the distal portion of the right pulmonary artery via a ductus arteriosus [32-33].

In an effort to elaborate on the congenital anomaly of the aortic arch, which is the isolation of the right subclavian artery from the right pulmonary artery, a lot of information was

gathered, with the main focus being on the development of the normal and abnormal origin of the subclavian artery [34].

The right subclavian artery usually have variations in its origin, it can take origin from the third, fourth, fifth or even sixth aortic arch [36]. In majority of cases the right subclavian artery normally derives from the fourth aortic arch and dorsal aorta, which travels distally to the seventh intersegmental artery. In the case of abnormal development of the right subclavian artery, there is dissolution of the right fourth aortic arch with its connection with the right subclavian artery and instead the right subclavian artery joins to the distal end of the right pulmonary artery via a ductus arteriosus [36].

The diagnosis of this anomaly can be seen through Magnetic Resonance Imaging and transthoracic Echocardiography.

This anomaly can be corrected by an arterial switch operation. The long ductal tissue which originates from the pulmonary artery is trimmed and the right ductus arteriosus is tie up in this procedure. The artery is then fixed in its correct anatomical position next to the right common carotid artery [37-38].

The right subclavian artery gives raise several different branches before it supplies the upper limbs of the body. These branches include the right vertebral artery, the right thyrocervical trunk, the right costocervical trunk and right internal mammary artery [38]. These branches are connected via collateral vessels to enlarged upper right intercostal arteries, which take origin from the descending aorta [39]. There is usually reverse flow within the right subclavian artery due to its higher density than the right pulmonary artery.

The manifestations of the isolation of the right subclavian artery depend merely on the ductus arteriosus [40]. In cases whereby the ductus arteriosus is exterminated, the isolated subclavian artery is being supplied by collaterals, which comes from contralateral subclavian artery and the ipsilateral vertebral artery; this is called the subclavian steal's phenomenon. In cases where the patent ductus arteriosus connects the subclavian artery to the pulmonary artery, there is a demonstration of a left to right shunt due to the lower resistances of the pulmonary artery compared to that of the systemic vascular resistance [39-40]. This shunt would increase the amount of steal from vertebral artery. However, when the pulmonary resistance is high, there may be flow from pulmonary artery to subclavian artery, which is the right to left shunt [40].

There can be a case of pulmonary steal phenomenon, where according to Doppler sonography report there is a complete retrograde flow of blood in the right vertebral artery and the upper part of the subclavian artery towards the right pulmonary artery [41]. This continuous filling of blood into the right pulmonary artery by the right subclavian artery is a demonstration of the left to right shunt. Due to this phenomenon a patient may suffer from pulmonary over circulation, vertebrobasilar insufficiency and weakness of right upper limb [42].

According to the incidence of the occurrence of this anomaly, it can be implied that this anomaly is extremely rare. The subclavian artery is a vessel, which lies in the thoracic region and supplies the upper limbs and chest region with oxygenated blood directly from the arch of aorta [42]. The subclavian artery is of two types, the left and right subclavian artery.

The right subclavian artery gives rise to several different branches, these branches forms a connection with the enlarged right intercostal arteries, which takes their origin from the descending aorta [43]. In the development of the abnormal origin of the right subclavian artery, it stated the position in which the subclavian artery is derived from depends on the ductus arteriosus, which makes the connection form the right subclavian artery and the right pulmonary artery. Due to this malformation, it may give rise to the pulmonary steal syndrome. According to Doocer's sonography this syndrome causes continuous flow of blood into the pulmonary artery from the subclavian artery. This phenomenon can give rise to May other complications in the body [43].

It is also believed that the isolated right subclavian artery does not only have to take origin from the pulmonary artery to be referred to as isolation of the subclavian artery [43]. Isolation of the subclavian artery occurs when the subclavian artery originates from any point other than the normal, which is the brachiocephalic artery for the right and the aortic arch for the left. It is also believed that since there can be continuous blood flow to the pulmonary artery, this process can also be applied to the opposite direction. Due to this hypothesis, the subclavian steal phenomenon can also be involved [43-44].

Conclusion

The congenital anomaly of the aortic arch, whereby there is an abnormal origin of the right subclavian artery occurs very rarely [28-31]. This anomaly can lead to heart failure and subclavian steal syndrome. The main emphasis were placed normal abnormal development of this particular aortic arch anomaly and how it affects the functionality of the aortic arch in the mediastinum [30]. The diagnosis of this anomaly can be made through MRI scanning and Transthoracic Echocardiography and correction can be made through Arterial switch operation, whereby the artery is relocated to its normal anatomical position [41].

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