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Cervical Origin of the Right Subclavian Artery in Aortic Arch Interruption: Pathogenesis and Significance

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In 14 of 21 infant hearts (66%) with aortic arch interruption between the left common carotid and left subclavian arteries (type B of Celoria and Patton), the right subclavian artery (SA) arose anomalously. In 9 (43%) it arose from the descending aorta and in 1 heart from the right pulmonary artery (5%). In 4 hearts (19%), it arose high in the neck from the right common carotid artery as a trifurcation with the internal and external carotid arteries, an anomaly not previously reported. In these latter cases, the SA descended down the neck to enter the right arm, with the right recurrent laryngeal nerve coursing around its take-off. Anomalous origin of the right SA is common in type B aortic arch interruption, and we believe this to be the result of a strong tendency for both fourth aortic arches to disappear early in development. If involution occurs in embryos of <14 mm crown-rump length, before the right ductus caroticus and dorsal segment of the right sixth arch have become attenuated, 3 alternate routes are available to the embryo to form a right SA.

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Aortic arch interruption (AAI) is often associated with anomalous origin of the right subclavian artery (SA),[1-4] particularly if the interruption is between the origins of the left common carotid and subclavian arteries, the type B of Celoria and Patton.[5] Molz and Burri[3] found 38 subjects with the combination among 153 reported cases of AAI (24%), 37 of which were type B. Of 17 cases described by Oppenheimer-Dekker et al,[4] the right SA arose anomalously in 7 (41%); 6 of these had type B AAI. In most reported cases the anomalous right SA arises from the descending aorta just distal to the origin of the left SA. Occasionally, it arises from the right pulmonary artery. A third type of anomalous origin from the right common carotid artery at the level of its bifurcation was encountered at autopsy in 4 of 21 infants with AAI. Because the anomaly may be of practical importance and because of its theoretical significance relative to the understanding of the nature and pathogenesis of the various forms of AAI, we describe the anomaly and discuss what we believe to be its pathogenesis.

Methods

Since July 1966, we have studied at necropsy 21 infants with AAI. In all autopsies on children with heart disease, we dissect the aortic arch arteries and follow them distally until identity and topography have been determined, after which they are doubly ligated and cut. Pulmonary arteries and veins and the great systemic veins are similarly treated. The superior vena cava is cannulated and the heart is fixed in the distended state by infusing 10% formalin in 95% alcohol at a pressure of about 20 cm of water while the heart is submersed in the same fixative.

Results

In all 21 infants the AAI was between the left common carotid and left subclavian arteries. In 19 of the 21 cases there was a ventricular septal defect (VSD), in 17 of these it had the characteristic appearance as described by Van Praagh et al[2] and there was encroachment of the left ventricular outflow tract. In 17 of the 19 cases with VSD (15 of those with characteristic VSD), the aortic valve was bicuspid. Two infants had double inlet left ventricle; in 1 both great arteries arose from the rudimentary outflow chamber but were normally interrelated; in the other, there was ventricular inversion as well as transposition of the great arteries. The right SA arose anomalously in 14 (66%) of the 21 cases: in 9 from the descending aorta (43%, Fig. 1A), in 1 from the right pulmonary artery (5%), and in 4 cases its origin was high in the neck at the level of the thyroid gland from the right common carotid artery as a trifurcation with the internal and external carotid arteries (19%, Fig. 1B). After take-off, the SA descended down the neck to enter the right arm (Fig. 2). The right recurrent laryngeal nerve coursed around the take-off of the right SA (Fig. 2B), much higher than normal.

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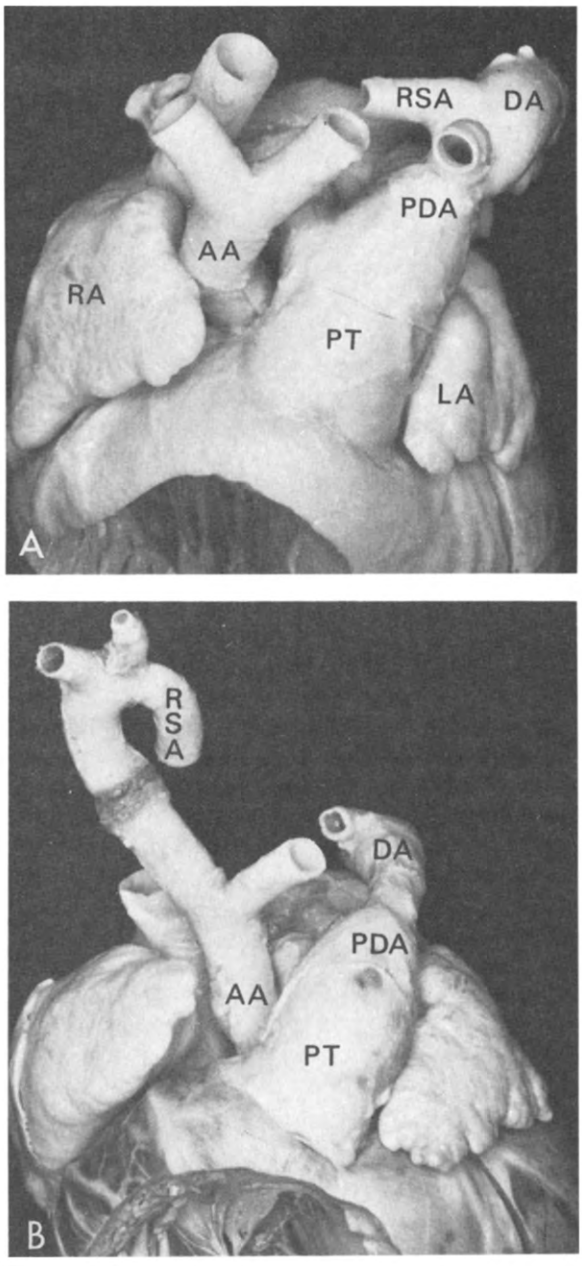

FIGURE 1. Aortic arch interruption with anomalous origin of the right subclavian artery from the descending aorta (**A**) and the right common carotid artery (**B**). The dark band on the right common carotid artery is an artifact. AA = ascending aorta; DA = descending aorta; LA = left atrium; PDA = patent ductus arteriosus; PT = pulmonary trunk; RA = right atrium; RSA = right subclavian artery.

Discussion

AAI between the origins of the left common carotid and left SA appears to be due to early involution of the left fourth pharyngeal arch artery (fourth aortic arch).

In normal development (Fig. 3A) the third aortic arches become the common carotid arteries, the left fourth arch forms part of the definitive aortic arch and the left sixth arch contributes the proximal part of the left pulmonary artery and the ductus arteriosus.[6,7] (As in all mammals the fifth arches in human embryos are small, imperfectly developed and have a fleeting existence, normally disappearing without a trace.)

The right fourth arch together with a part of the right dorsal aorta and the right seventh intersegmental artery forms the right subclavian artery. The dorsal part of the right sixth arch involutes very early in development and disappears; the proximal part becomes the right pulmonary artery. Segments of the dorsal aortae between the third and fourth arches (ductus caroticus) and the distal part of the right dorsal aorta disappear in normal development.

Since the origin of the right SA is normally formed by the right fourth aortic arch (Fig. 3A), its anomalous origin, common in cases of AAI, is readily explained by early involution and absence of both the right and the left fourth pharyngeal arch arteries. The pathogenesis of the 3 types of anomalous origin of right SA can be derived from the normal developmental pattern of the aortic arch system. If the right fourth arch involutes early, i.e., in embryos of <14 mm crown-rump length, before the right ductus caroticus and dorsal sixth arch have become attenuated, 3 alternate routes are available

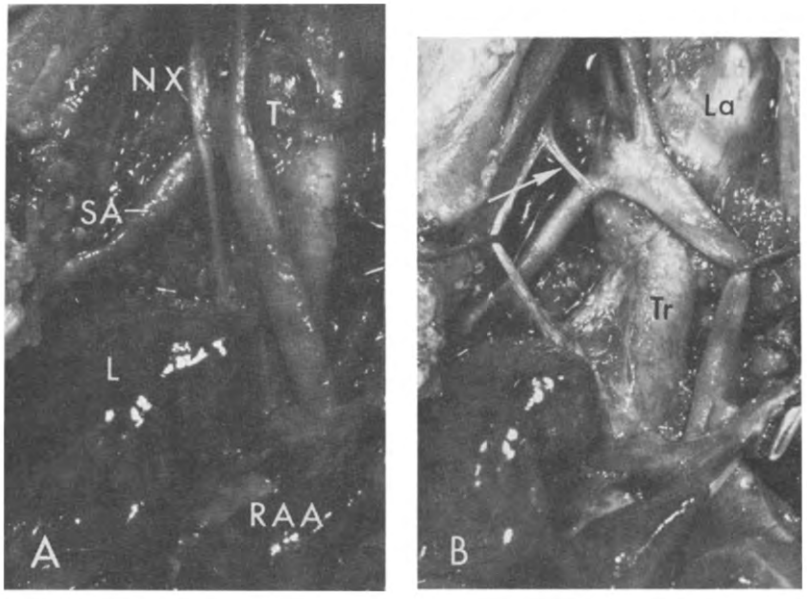

FIGURE 2. Topography of right subclavian artery in cases with cervical origin from the right common carotid artery. In **panel B,** the right common carotid artery has been pulled to the left and the vagus nerve to the right, exposing the recurrent laryngeal nerve (**arrow**). L = lungs; La = larynx; NX = vagus nerve; RAA = right atrial appendage; SA = subclavian artery; T = thyroid gland; Tr = trachea.

to the embryo to form an anomalous right SA. Most often, the distal part of the right dorsal aorta is retained, which, with the right seventh intersegmental artery, forms the anomalous vessel (Fig. 3B). Origin from the right pulmonary artery may be explained as being the result of retention of the dorsal segment of the right sixth aortic arch, the equivalent of the ductus arteriosus on the left side, which, with a segment of right dorsal aorta and the right seventh segmental artery, forms the SA (Fig. 3C). This genesis is similar to that of the normal right SA: The right sixth arch is simply substituted for the absent fourth arch.

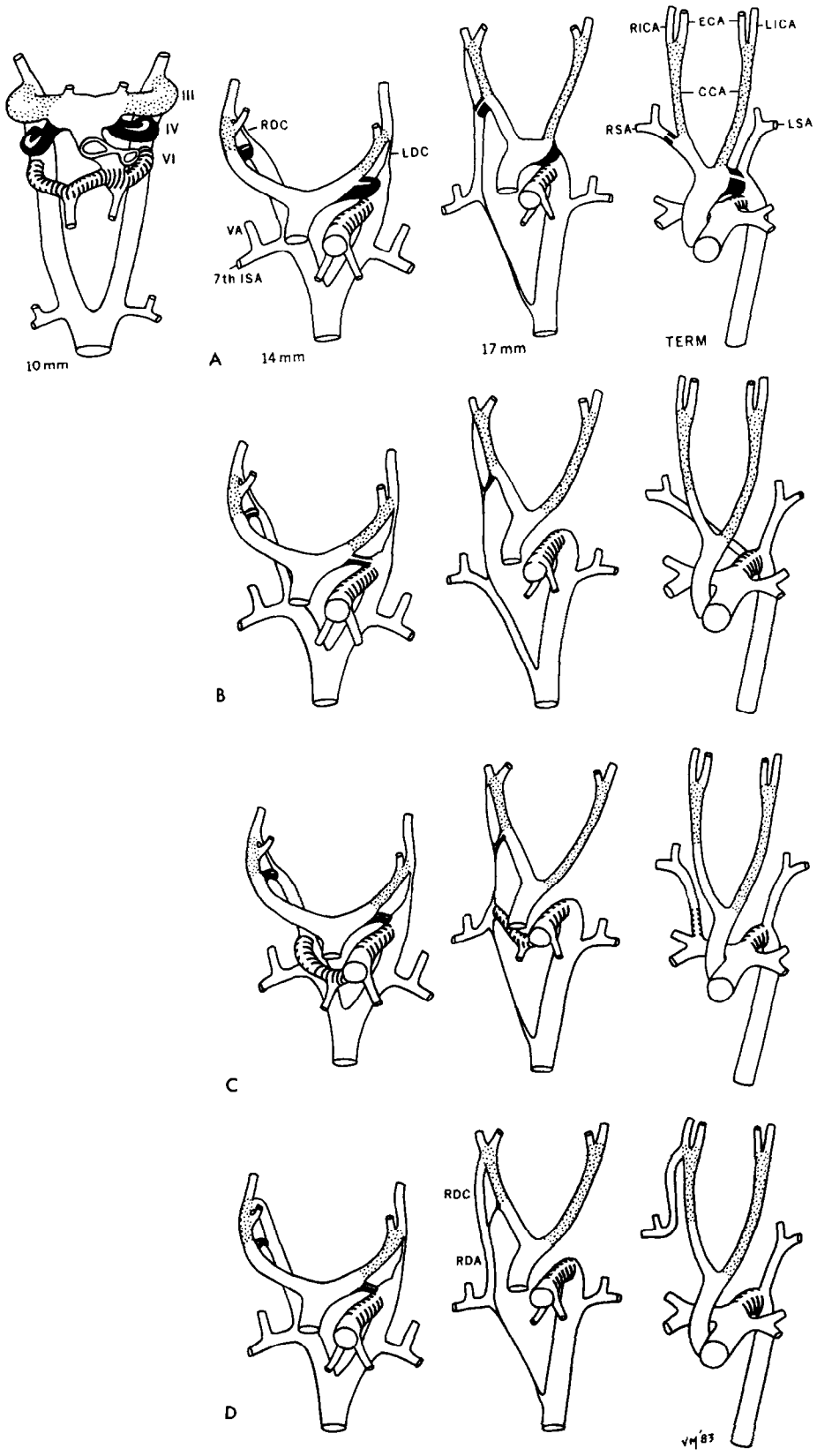

FIGURE 3. A, development of the normal aortic arch system. Stages shown are from embryos of 10, 14 and 17 mm crown-rump length, and at term. Redrawn after Congdon.[7] **B, C** and **D,** hypothetical stages in the pathogenesis of aortic arch interruption associated with anomalous origin of the right subclavian artery from the descending aorta (**B**), the right pulmonary artery (**C**) and the right common carotid artery (**D**). III, IV, VI = third, fourth and sixth aortic arches; 7th ISA = seventh intersegmental artery; CCA = common carotid arteries; ECA = external carotid artery; L(R)DC = left (right) ductus caroticus; L(R)ICA = left (right) internal carotid artery; L(R)SA = left (right) subclavian artery; RDA = right dorsal aorta; VA = vertebral artery.

We believe that in cervical origin of the right SA the right ductus caroticus, the segment of the right dorsal aorta between the third and fourth arches (Fig. 3D) is retained. This segment normally begins to involute in human embryos of about 14 mm crown-rump length, or about the seventh week of gestation. This pattern of development explains why the anomalous SA originates at the level of the bifurcation of the right common carotid artery, as well as the location of the right recurrent laryngeal nerve. Initially, this nerve courses around the sixth aortic arches on both sides. On the left side this position is retained and the recurrent laryngeal nerve courses around the ductus arteriosus (ligamentum arteriosum), which is derived from the left sixth aortic arch. Since on the right side the corresponding sixth arch segment involutes and disappears early in development, the right recurrent laryngeal nerve normally runs around the right fourth arch, i.e., the origin of the right SA. When the fourth arch also disappears the nerve moves 1 arch higher, hence its course around the origin of the cervical SA.

Clearly anomalous origin of the right SA is the rule in type B AAI. Cervical origin of the SA is one of the 3 options available to the embryo for alternate routing of the right upper limb arterial supply if the ipsilateral fourth arch involutes in early development. This anomaly has not been described previously, probably because it has been overlooked, not because it is rare. The caliber of the right common carotid artery in these cases is the same as the usual innominate (brachiocephalic) artery, since it has to carry the same amount of blood. If it is cut in the usual way at autopsy there is nothing to indicate that the stump is anything but the innominate artery.

Persuasive arguments have been put forward[8-10] in support of the concept that hypoplasia or atresia of an aortic arch segment, juxtaductal coarctation and AAI are secondary anomalies induced by altered prenatal hemodynamics resulting from intracardiac defects, such as certain malalignment type VSDs, which are associated with anomalous muscle bundles encroaching upon the subaortic area or bicuspid or stenotic aortic valve and which favor pulmonary blood flow at the expense of aortic flow. The intracardiac anomalies present in the 21 cases of AAI presented here support the above concept.

However, one might question why there should be such a high incidence of anomalous origin of the right SA; that is, why is the right fourth aortic arch so often absent as well? It might be argued that the same hemodynamic alteration—reduced flow through the right fourth arch—is responsible, since this arch is also supplied by the ascending aorta. Substitution of the dorsal segment of the right sixth arch (right ductus arteriosus), or the distal part of the right dorsal aorta, for the right fourth arch represents what appears to be a logical and plausible alternate pathway in the formation of the right SA, since both of these arterial segments are supplied by the pulmonary trunk. However, this cannot explain the substitution of the right third arch and ductus caroticus in cases of cervical origin of the right SA, since that pathway is supplied by the ascending aorta. Another difficulty is that anomalous origin of the right SA, i.e., disappearance of the right fourth arch, is only common in type B AAI, but is rare in type A and in coarctation of the aorta.

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