COARCTATION OF THE AORTA

Essay

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General Surgery

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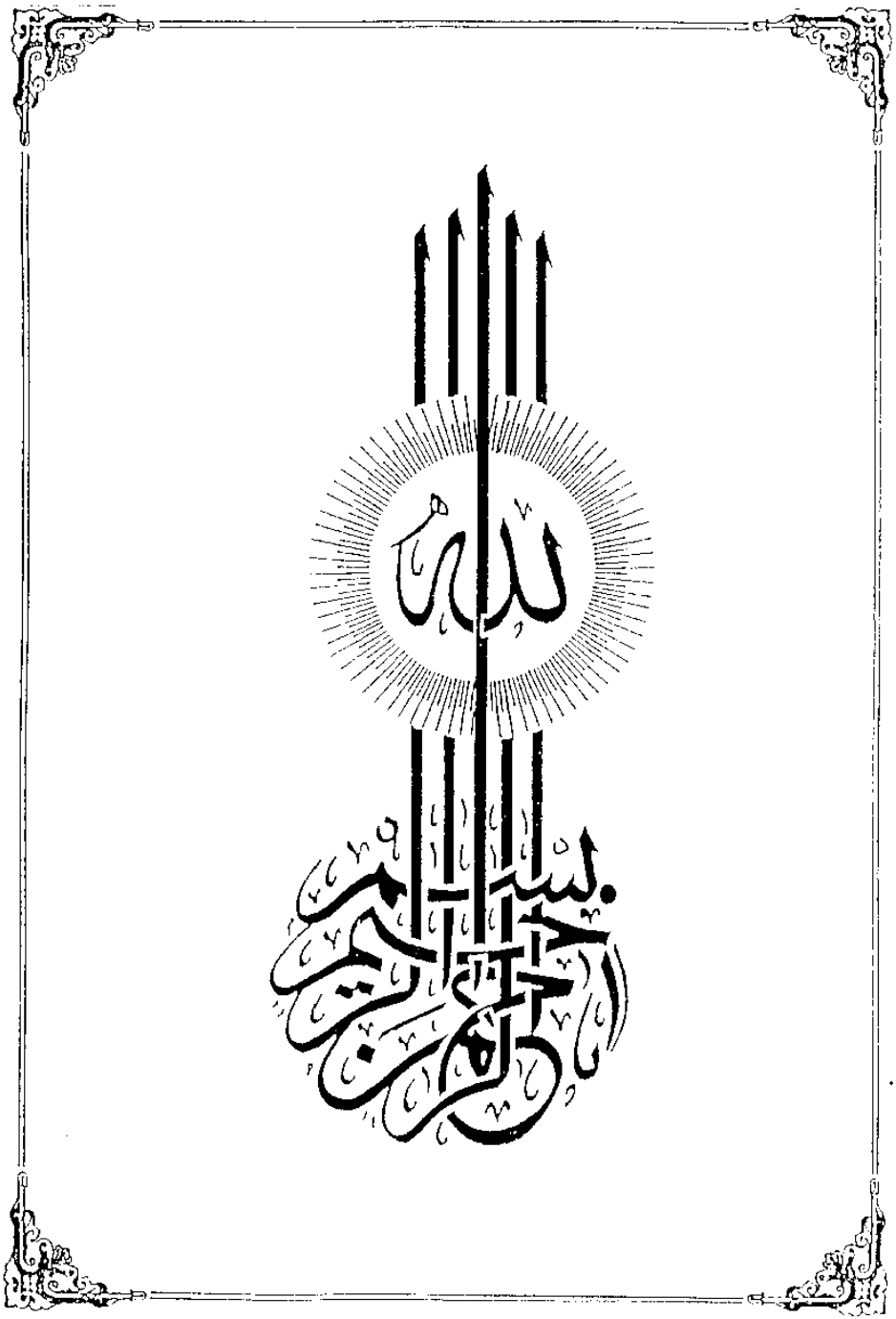

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INTRODUCTION AND HISTORICAL ASPECT

Coarctation of the aorta signifies a constriction of the aorta in the region of its juncture with the ductus arteriosus or its vestige **(Blackford, 1928).** The term congenital stenosis of the isthmus is also employed. The advantage of this term is that it is more early understood by persons who do not read English readily **(King, 1926).** The disadvantages outweigh this, for the term is clumsy and subject to ambiguous abbreviations. Many authors have maintained that the condition develops after birth. The anatomic isthmus is that part of the aorta between the origin of the left subclavian and the mouth of the ductus arteriosus; this definition excludes the large number of cases reported in which the constriction is distal to the insertion of the ligamentum arteriosum. However, **in 1839 Mercier** wrote "Coarctation" in French; ten years later Diesterweg, "Coarctatio" in Latin, and the English word coarctation has an honorable history since 1876 **(Blackford, 1928).**

In the year 1760, Morgagni performed a postmortem on a monk and drew attention to a constriction of the aorta a short distance from the heart.

The first case definitely described with the typical pathological findings and collateral circulation was recorded by **Paris in 1791 (Barié, 1886).**

The occurrence of coarctation of the aorta is not rare. **Blackford in 1928** estimated that coarctation of the aorta was found

once in every 1550 autopsies. Evans in 1932 stated that it occurred once in every 1000 autopsies. Abbott, in her series of 1000 cases of congenital heart disease, records coarctation of the aorta as occurring 178 times, 105 of these or 60% were of the adult type. Perlman found it occurring once in 10000 men **(Calodney, 1950).**

Although an increasing number of anatomical descriptions appeared in the intervening years, it was not until 1835 that Le Grand made the first diagnosis of aortic obstruction based on the presence of collateral circulation and decreased femoral pulsations **(Jarcho, 1962).**

Also **in 1839, Mercier** diagnosed an obstruction to the thoracic aorta, but Oppolzer **(Quoted by Barié, 1886),** was the first to make the diagnosis of coarctation at the site of election; he did this twice and saw the diagnosis verified post mortem both times. Little or nothing had been added to Oppolzer's diagnostic criteria **(Blackford, 1928).**

In 1828, Reynaud considered coarctation a persistence of the fetal state in some way influenced by the involution of the ductus. **Rokitansky, (1852),** was in essential agreement **(Blackford, 1928).**

In 1887, Loriga maintained that the ductus arteriosus did not play a part in the pathogenesis, and that the anomaly could be traced

back to intrauterine life, the differing pathologic types corresponding to the differing periods at which the lesion began.

Dickinson and Fenton (1900), Minkowski (1901), Monckeberg (1925), and Meixner (1925) believed it is of developmental origin.

The prenatal development of coarctation in all cases has never been universally accepted. **Zenoni (1911), Kaufmann (1922),** and others attributed it, in some cases at least to the propagation of an inflammatory process in the ductus Botalli to the wall of the aorta.

In 1844, Hamernjik observed that the constriction might be proximal to, at the site of, or distal to the aortic opening of the ductus arteriosus, and accordingly divided these cases into three types, he accepted Raynaud's theory for the first, but seems to have supposed that prolongation of the thrombus, hypothesized in the closure of the ductus, was responsible for the other two forms. **In 1855, Skoda** argued that fibres from the ductus extended into the aortic wall and that these fibres contracted with the contraction of the ductus in the process of its obliteration **(Blackford, 1928).**

In 1900, Schesinger reviewed theories of both Reynaud and Skoda. Three years later, Bonnet elaborated these two hypotheses, and according to him, constriction of the aorta proximal to the mouth of the ductus arteriosus dates from intrauterine life and, if not a transient persistence of the normal fetal state, is incompatible with

life for more than a brief period. On the other hand, he explained constriction at or distal to the ductus by the Skoda's theory, and found it devoid of influence on the health or longevity of the person except that it may predispose to endocarditis. He called these, respectively, the infantile (type du nouveau né) and the adult types. Abbott offered this explanation in 1927 **(Blackford, 1928).**

In has been intimated several times that syphylis may cause *coarctation , but striking arguments have not* been adduced in support of the theory **(Edelmann and Maron, 1922).**

In 1903, Bonnet classified the condition into the infantile and adult types. In the former, the coarctation lies above the ductus and may be localized or diffuse, in the later, the constriction is just below the entrance of the ductus and is commonly localized. This concept is unvalid because the adult type may give rise to severe congestive heart failure and death in infancy, while the infantile type occasionally is asymptomatic in adulthood, in addition anatomically the coarctation may stride the entrance of the ductus arteriosus **(Moss, 1968).**

In the clinical diagnosis of coarctation, precordial murmurs are of little consequence as they are sometimes absent, and they have been described in such a variety of terms that nothing pathognomonic can be deduced. The murmur in the interscapular space, more often to the left of the vertebral column, is of greater importance. Oppolzer attributed this to the stream of blood rushing through the

greatly enlarged superior intercostal artery (Intercostalis suprema). Walshe attributed it to blood passing through the stenosed area, and thought that its absence would prove atresia **(Dickinson and Fenton, 1900).**

Woltman and Shelden, (1920) , diagnosed the condition on the basis of absence of pulsations in the abdominal aorta and femorals. **Laubry, (1926),** verified the diagnosis made on such grounds by postmortem examination, he believed that marked disproportion in the blood pressure of the brachial and femoral arteries is enough basis for the diagnosis **(Blackford, 1928).**

Van den Berg, (1911), King, (1926), and others have remarked that the electrocardiograph does not show any important or constant changes.

Loriga, 1887, advanced the original idea that exophthalmic goiter must be considered in the differential diagnosis on account of the bruits and thrills in the neck.

Maude Abbott, (1928), collected 200 autopsy cases from the literature of patients over 2 years old. Only 28 were diagnosed in life.

Rhodes and Durbin, (1942), stated that the total number of cases of coarctation diagnosed clinically in children reported in the entire literature had increased to 47 **(Calodney and Carson, 1950).**

In 1945, both Crafoord and Gross independently reported a significant achievement in great blood vessel surgery. They successfully operated on patients with coarctation of the aorta. The operation consisted in excising the constricted portion of the aorta and anastomosing the two ends. In November 1944, Potts, Smith, and Gibson reported the successful anastomosis of the aorta to the pulmonary artery. They used a special aortic clamp which permits part of the blood to pass through the aorta during the operation. **Clagett, (1947),** reported the first successful anastomosis of the left subclavian artery to the aorta bypassing the coarctation in a human subject.

Gross, (1948), reported the successful use of human arterial grafts in the treatment of coarctation of the aorta. These achievements of modern surgery and possibilities, made the diagnosis of coarctation in the newborn and young infants a matter of vital importance **(Calodney and Carson, 1950).**

EMBRYOLOGY

Development of the aortic arch system :

In an embryo of about 3-mm. the first pair of aortic arches is large and the second pair is just forming (Fig. 1-A). The dilated junction of the truncus arteriosus and the first pair of aortic arches is the aortic sac, from which subsequent aortic arches will originate, new arches being added caudally. A true ventral aorta is not present in mammalian embryos. Caudally the dorsal aortas fuse to form a single vessel; this fusion progresses craniad.

In a 4-mm. embryo the first arch has largely disappeared with only a part of it persisting as a portion of the maxillary artery (Fig. 1-B).

The second aortic arch has also retrogressed; eventually all that remains in the tiny stapedial artery. The third aortic arch is well developed and the fourth and sixth arches are being formed as ventral and dorsal sprouts of the aortic sac and dorsal aorta respectively. The ventral portion of the sixth arch already has as its major branch the primitive pulmonary artery even though the arch itself has not yet been completed. The fifth aortic arch in mammals is rudimentary and present only for a very brief time.

In a 10-mm. embryo the first two aortic arches are no longer present as such and the third, fourth and sixth are large (Fig. 1-C). The truncoaortic sac has been divided by the formation of the aorticopulmonary septum so that the sixth arches are now continuous

with the pulmonary trunk. Of the cervical intersegmental arteries the seventh pair will play an important role in the formation of the subclavian arteries. They are located about the level where the dorsal aortas join each other .

In a 14-mm embryo the aortic arch system has lastly lost its original symmetrical pattern (Fig. 1-D). The segments of the dorsal aortas between the third and fourth arches, the carotid ducts, have disappeared, and the third arches begin to elongate as the heart descends farther into the thorax. This descent has also caused a relative shortening of the paired portion of the dorsal aorta. The dorsal portion of the right sixth arch has disappeared; its counterpart on the left persists until birth as the ductus arteriosus. The seventh intersegmental arteries have migrated craniad. The aortic sac has been " Pulled out" on both sides on the right it forms the brachiocephalic (innominate) trunk; on the left it becomes part of the definitive arch of the aorta up to the origin of the left third arch (common carotid artery).

In a 17-mm embryo the right dorsal aorta between its junction with the left dorsal aorta and the origin of the right seventh intersegmental artery has become attenuated and later disappears (Fig. 1-E) . The remainder of the right dosal aorta persists and with the right fourth aortic arch, forms the proximal subclavian artery.

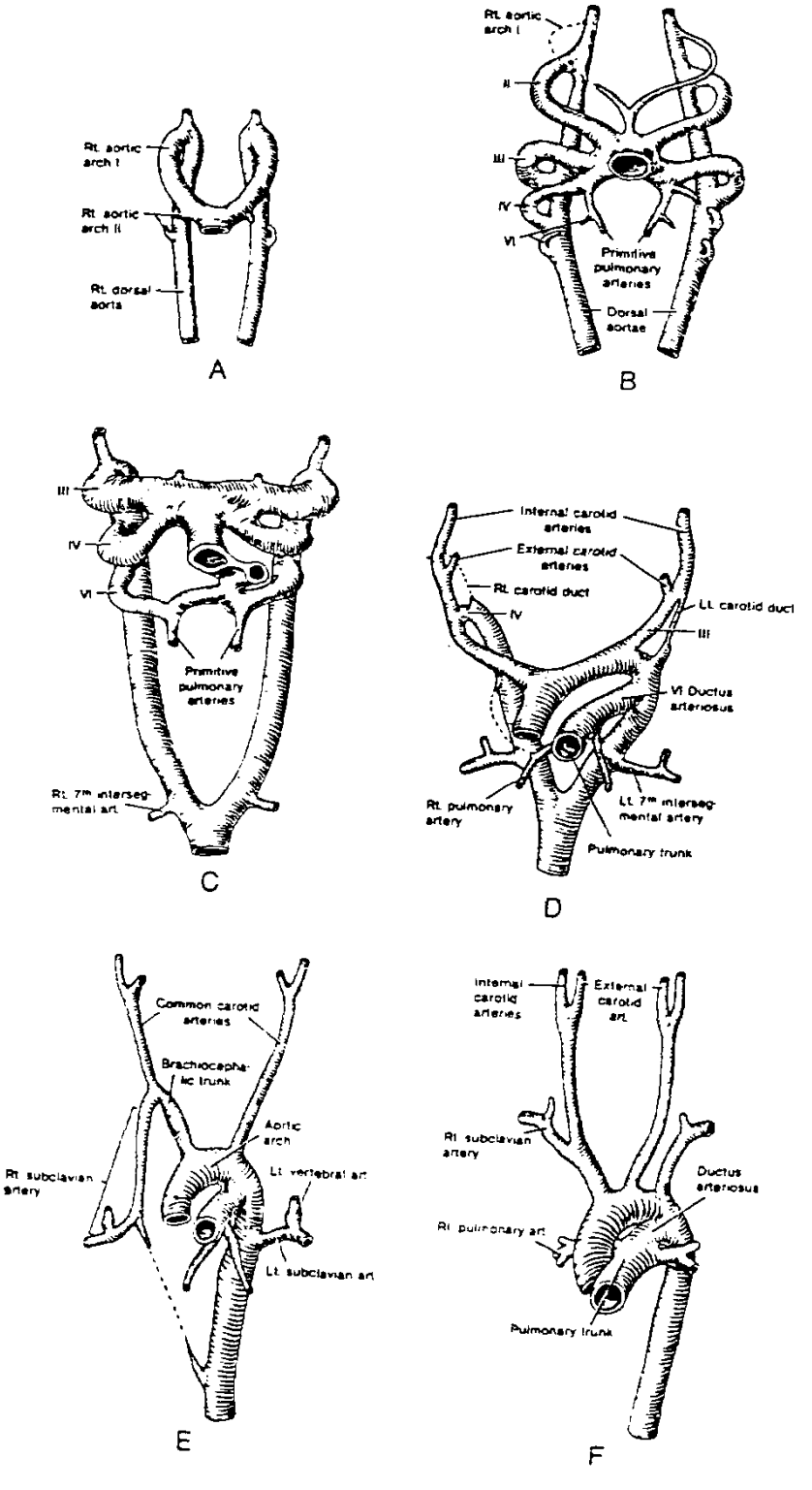

Fig. (1) : Development of the aortic arch system.