

Thoracic Arterial Arch Anomalies

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THORACIC ARTERIAL ANOMALIES were formerly regarded as having little significance except when they produced tracheal or esophageal obstruction. As thoracic surgical procedures become more prevalent in the smaller or community hospitals, the case concentration will become less, and many of these anomalies may go unrecognized. Recognition of the many types of vascular disarrangement thus assumes a greater clinical significance. Although detailed information is available concerning specific anomalies, general surveys of the over-all problem are lacking. By reviewing 41 selected instances of arterial arch anomalies that we have encountered either on the Thoracic Surgery Service, Walter Reed General Hospital, or in the Cardiovascular Collection of the Armed Forces Institute of Pathology and referring to those described by others, it is hoped to stimulate interest and discussion of these intricate problems.

Many parallels for the study of these anomalies may be found in vertebrate anatomy. Although Hommel¹ had published a beautiful illustration of a constricting double aortic arch in the human in 1737, it remained for Jex-Blake² to suggest in 1926 the feasibility of surgical correction based on the autopsy findings of this anomaly in a puppy which he had observed with stridor and dysphagia. Since Gross³ first successful surgical correction of a vascular ring in June 1945, much progress has been made in the diagnosis and treatment of these anomalies.

The double aortic arch is found normally in some classes of vertebrates. Figure 1 shows the long pairing of the dorsal aortae as seen

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in the reptile. Anyone who has seen a small snake swallow an egg or a rodent is aware that a vascular ring is, not of itself, necessarily productive of difficulty. In other vertebrates, the double arch is found abnormally and may be present with or without symptoms. Figure 2 shows a double aortic arch in a calf. Figure 3 shows another type of vascular ring in the dog, that is the right aortic arch with an encircling left ligament. The German shepherd is said to be particularly disposed toward this anomaly. Because of the frequency of these vascular anomalies in the dog, pig, and calf, the well-read veterinarian is as aware as the physician of this problem.⁴⁻⁶

In the human, the aorta normally arches to the left of the trachea and the esophagus. This is the result of the persistence of the left fourth arch rather than the right fourth arch. In some mammals, however, the reverse is true. In the bird, for example, the arch typically passes to the right. When this avian configuration (fig. 4) is found in humans it is of great clinical interest.

In the Rathke diagram (fig. 5) showing the development of the aortic arch system, the pairing of all structures is emphasized with the exception of the ventral aorta. It is this bilaterality of all structures except the ventral aorta that provides the basis for many anomalies. Shown as a matter of interest in this regard is the unusual vascular anomaly in figure 6. Since two aortae are depicted arising from the heart, this might at first appear to represent an exception to the above statement. It does not, however, as this is a composite of two hearts, an instance of thoracopagus Siamese twinning. In vertebrates a unique exception is found in the crocodile. This mammal has both right and left ventral aortae with an intercommunicating foramen (foramen Panizza). This allows blood to bypass the lungs during submersion.

If one adopts the concept of the "basic

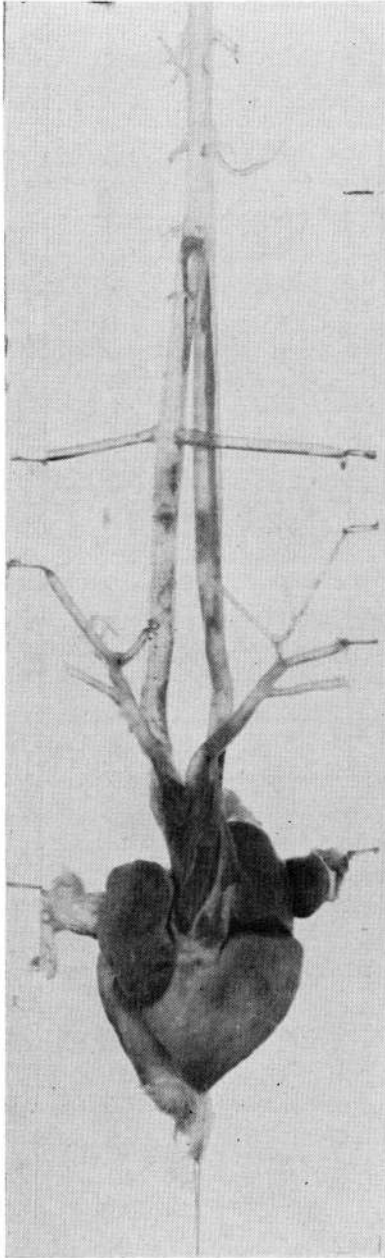


Figure 1

Reptile's double aortic arch. Note looseness of ring.

primitive pattern” of Edwards⁷ as the human embryologic starting point, the complexity of these arch anomalies largely disappears. Recognizing that the aortic arches appear in temporal succession, and by the time the last pair appears, the first two have undergone regression, Edwards has retained only those

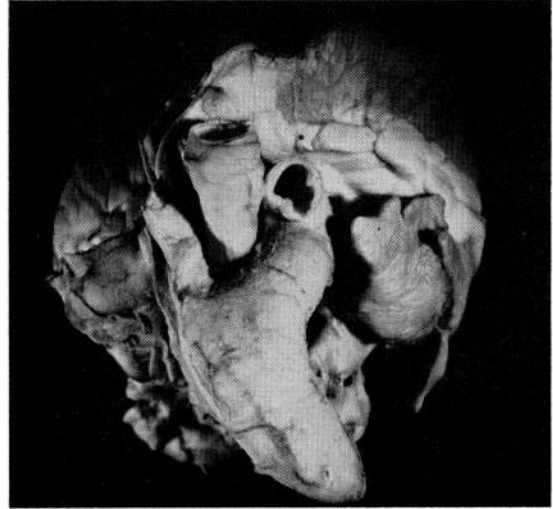


Figure 2

Calf with double aortic arch. In the background is seen the heart with an especially prominent right atrial appendage. A large right arch and a smaller left arch tightly encircle the trachea and esophagus before rejoining.

parts of the Rathke diagram that have remained after many earlier changes (fig. 7). In the operating room, the use of this pattern as a starting point is particularly valuable in that it often permits a quick deciphering of the anomaly with greater certainty and with lessened operative exposure and trauma. This pattern assumes additional importance in that it may be used to visualize the evolution of some 36 theoretically possible vascular arch anomalies if one presumes aortic arch involution at but one point. On the presumption of involution at two or more points, the number of malformations increases as a geometric progression. If this “basic primitive pattern” is presupposed to be the forefather of all subsequent vascular anomalies, how do the latter develop?

To visualize the development of these anomalies, consider the large drawing in figure 8 as representative of the “basic primitive pattern.” A series of changes in this pattern is now assumed. The first of these changes is lateralization of the aorta either to the right or to the left (1-a, 1-b). This shift is secondary to the midline appearance of the vertebral bodies with the subsequent displacement of

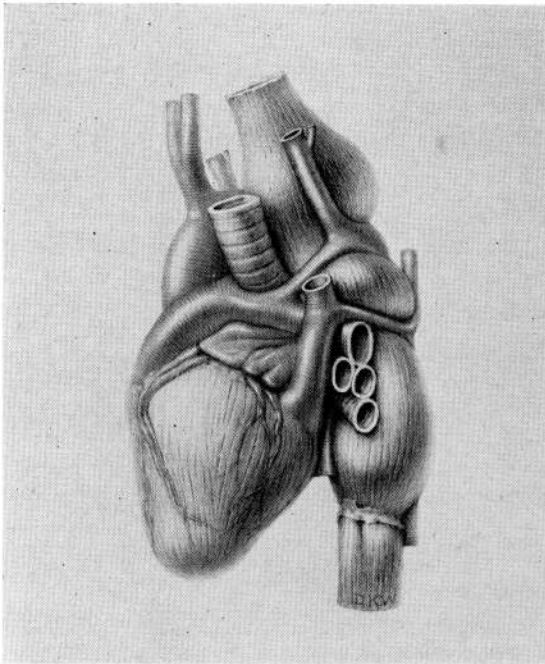


Figure 3

Dog with right arch and a left encircling ligamentum arteriosus causing tracheal and esophageal constriction. Note also the persistent left superior vena cava joined by the hemiazygos vein.

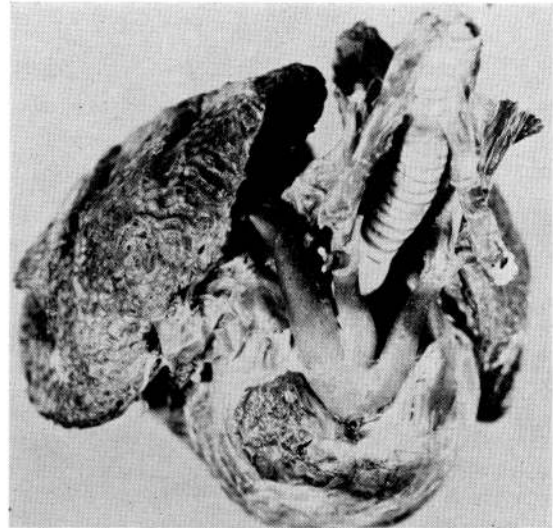


Figure 4

The bird's right arching aorta is seen passing over the bronchus to the right lung. The foreshortened arch has given origin to two brachiocephalic trunks.

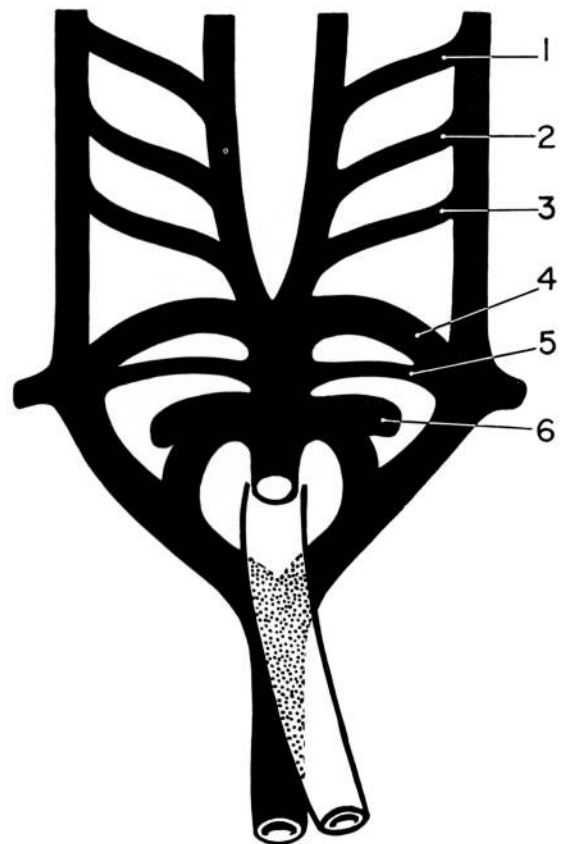


Figure 5

Rathke diagram. The third arches form portions of the carotid arteries; the fourth, the arch of the aorta; the sixth, the distal pulmonary arteries.

the midline dorsal aorta. In our collection, there are three hearts representing this early stage of development (figs. 20, 21, and 22). The second change involves a similar lateralization, but this time of the ductus (a-1, a-2, b-1, b-2). The frequency with which the ductus undergoes lateralization to the left is explained as a function of the left ductus being in a more favorable hemodynamic situation for flow and may be explained as follows. In the normally rotated heart, the ascending aorta lies to the right of the mid-sagittal plane. The pulmonary artery consequently lies to its left. With the pulmonary artery thus lying to the left, the left ductus provides a shorter and more direct route to the left arch than does the right ductus to its arch. Persistence of the left ductus is therefore favored. In situs inversus with dextrocardia, where the relationships of the aorta and pulmonary artery are reversed, the ductus most often persists on the right (fig. 9). The six

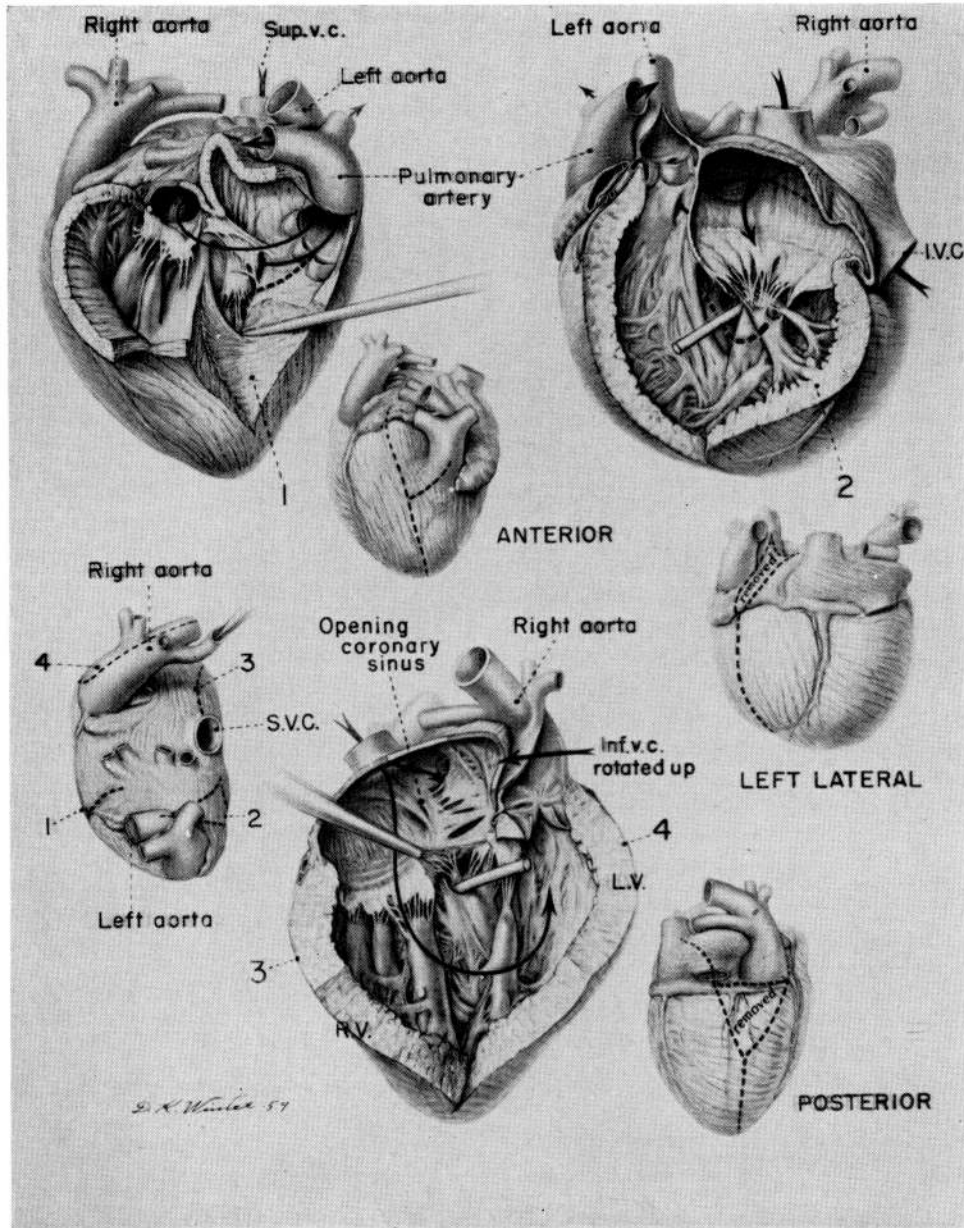


Figure 6

Two aortae arising from the heart. This drawing of the heart in a case of thoracopagus Siamese twinning is a composite of two hearts and thus does not represent an exception to a single ventral aorta of the Rathke diagram.

smaller figures shown in figure 8 constitute six limbs of the genealogy tree of the aortic arch. Each, in turn, can be analyzed further by supposing involution at one of six possible separate points on the arch system. A development of this concept will be carried out

using b-2 or its enlarged counterpart, figure 10.

If involution should occur at point 1, figure 10, the "normal" left aortic arch would result. This arch becoming separated from its point of posterior attachment, retracts and

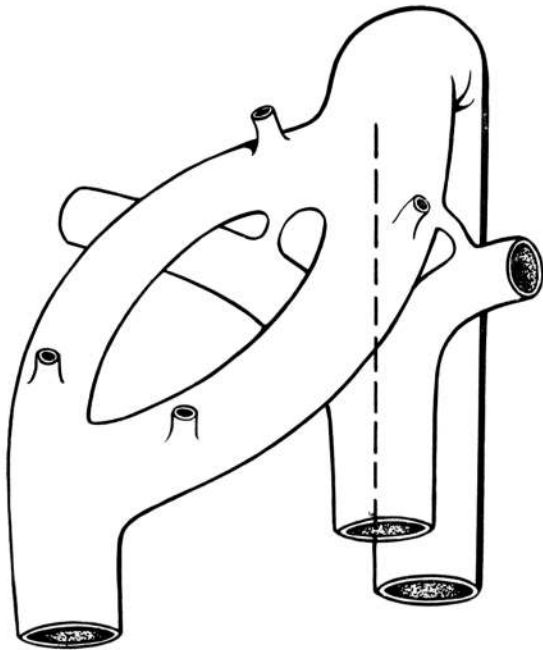


Figure 7

Edward's "basic primitive pattern" consisting of right and left aortic arches, right and left ductus arteriosus, and a midline descending aorta.

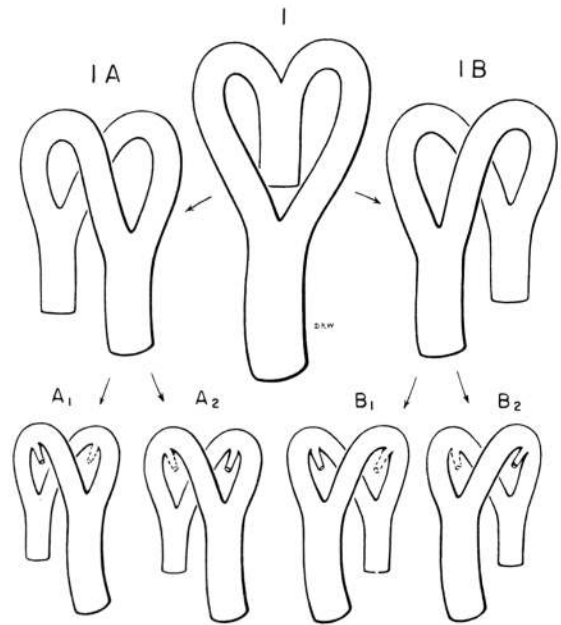


Figure 8

Diagram showing the evolutionary development of six more advanced anomalies from the "basic primitive pattern."

realigns itself. Serving as the common origin of the right carotid and subclavian arteries, the proximal portion now becomes the right innominate artery. It seems teleologically significant that this is the only point at which involution can occur that is not potentially capable of producing some form of tracheal or esophageal obstruction.

Involution at point 2, figure 10, would result in the now caudally arising right subclavian artery, having to cross the midline to reach the right apex. As Holzapfel⁸ indicated, the course taken may be quite variable. Of 133 instances this author studied, six passed anterior to the trachea, 20 passed between the trachea and the esophagus, and 107 passed retroesophageally. Approximately 1 per cent of routine barium studies of the esophagus are said to show the latter anomaly.

There are many aspects of this anomaly, the aberrant right subclavian artery, which still seem poorly understood. If separation of the right arch should occur in such a manner that a portion of this right or posterior arch

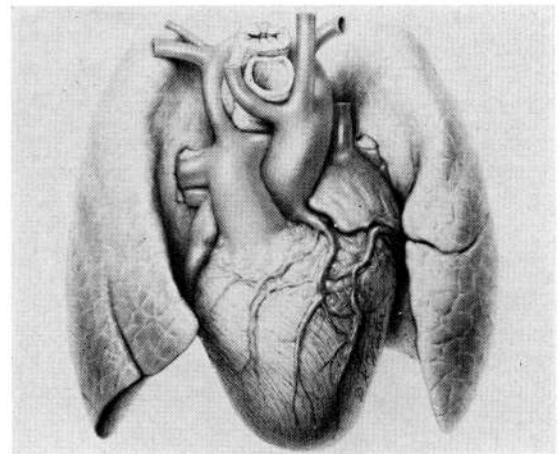


Figure 9

A right ductus and a left arch producing a vascular ring in situs inversus (a situation comparable to an encircling left ductus with a right arch in normal situs).

persists and, in turn, gives rise to the aberrant right subclavian artery, a large aortic diverticulum results (fig. 11), a finding first described by Kommerell.⁹ Generally there is

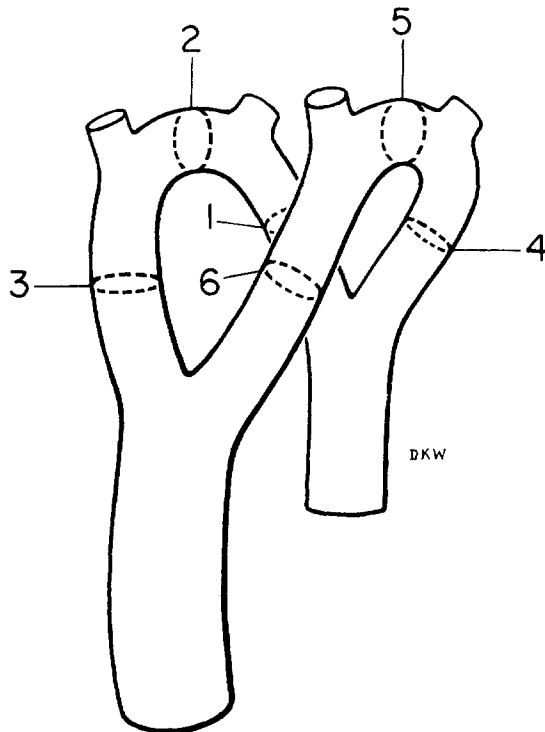


Figure 10

Enlargement of figure 8-b-2. The dotted rings indicate possible points of arch interruption. The left ductus has been omitted.

failure to recognize this diverticulum. Roentgenologically, this large retroesophageal diverticulum may be misinterpreted on the basis of "size" as the posterior limb of a double aortic arch. Not infrequently, the "aneurysmal dilatation" of this artery encountered by the surgeon and possibly thought "too dangerous" to divide, may represent only such a diverticulum. Aneurysmal dilatation and rupture of an aberrant right subclavian artery does occur, although infrequently.¹⁰ Rupture of a diverticulum of Kommerell after resection has also been reported.¹¹ However, the wall of the diverticulum does not appear morphologically to be deficient in structure.

Again it seems worthwhile to re-emphasize the frequency with which an aberrant right subclavian artery occurs in the tetralogy of Fallot, with a left aortic arch. Pattinson and Emanuel¹² reported an incidence of 16 per cent. Blalock^{13a, 13b} has also commented on this anomaly.

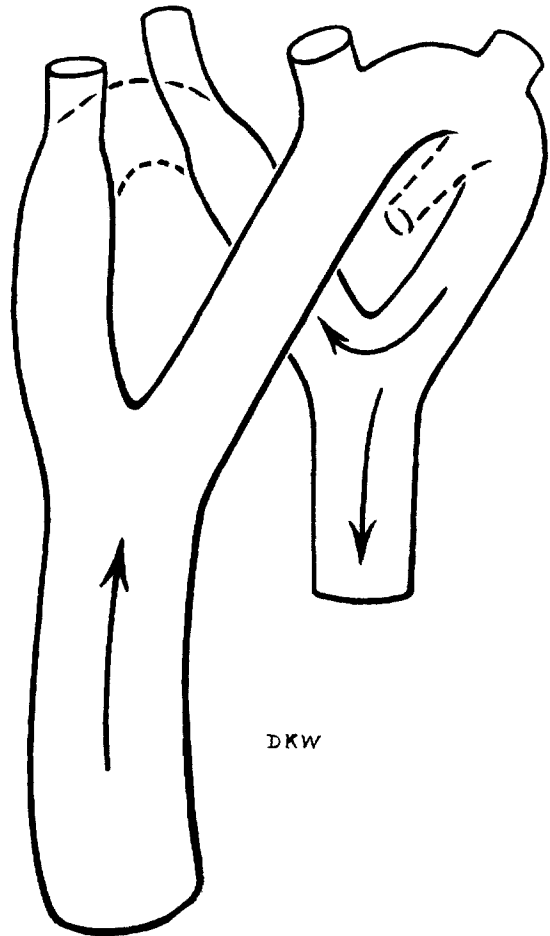


Figure 11

Though the right arch has been interrupted in the dotted area, the distal portion of this arch may persist as an enlargement—the so-called diverticulum of Kommerell.

If involution should occur at points 4, 5, or 6 (fig. 10), a right aortic arch results. As the significance of the right aortic arch has been greatly underestimated, we would like to re-emphasize its importance as a clue to the presence of intrathoracic vascular anomalies.¹⁴ Right aortic arches are associated with a high incidence of intrathoracic anomalies, i.e., several types of vascular rings or slings, the right-sided or bilateral ductus arteriosus, contralateral absence of the pulmonary artery, and large bronchial arteries.

Sprong and Cutler¹⁵ stated that in cases with a right aortic arch and a left innominate

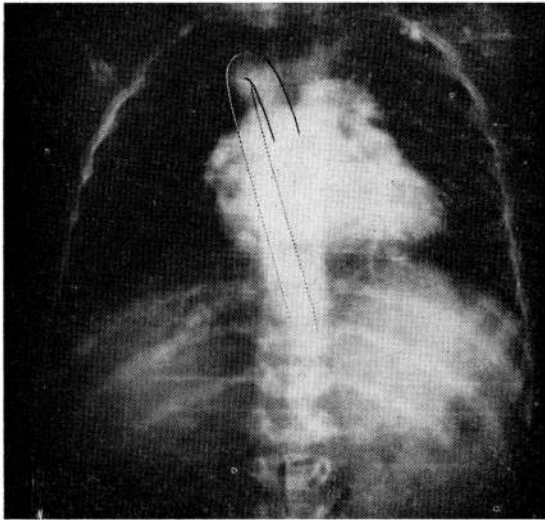


Figure 12

Angiocardiogram of patient with severe congenital cardiac defects. There is a right arch and right descending aorta.

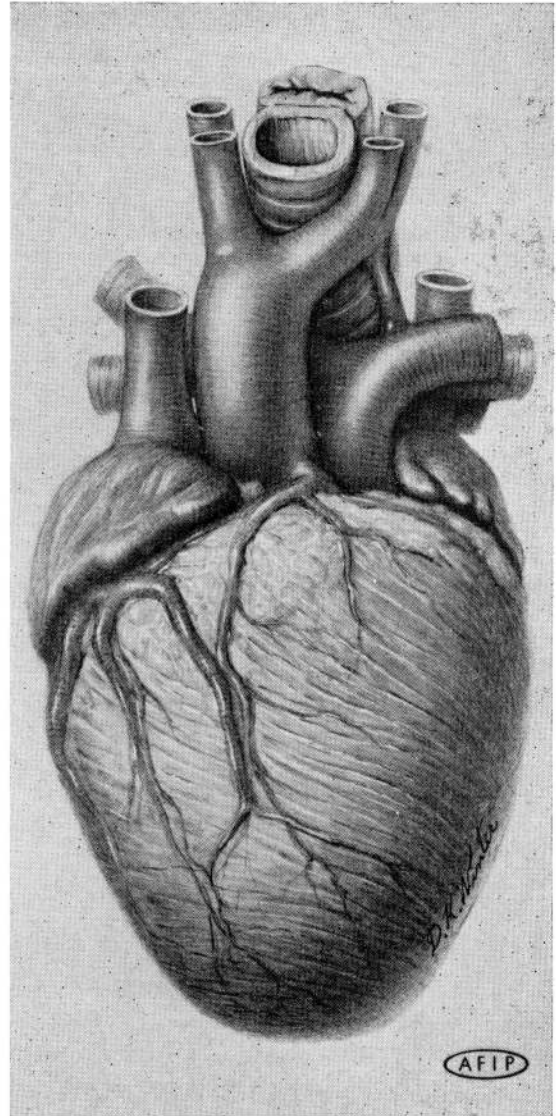


Figure 14

Diagram of heart showing a left anterior ductus.

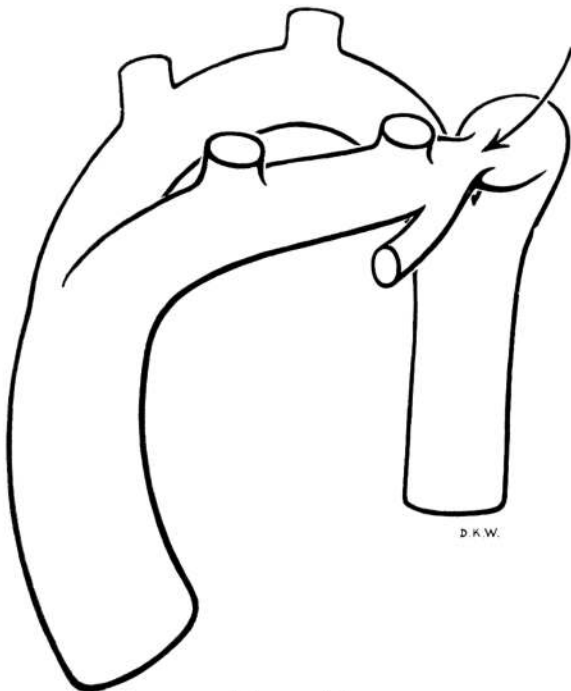


Figure 13

The involutionary process involving the left arch distal to the ductus insertion produces a non-constricting left anterior ductus. Involution proximal to the ductus insertion will produce a posterior encircling ductus and a vascular ring.

artery, intracardiac defects will be found in four of 10 cases. When the right aortic arch is associated with a right descending aorta (the avian configuration—fig. 4), the incidence is much higher. Assmann¹⁶ went as far as to suggest that all such cases are associated with congenital defects of the heart (fig. 12).

With the right arch one might expect the incidence of an obstructive vascular ring composed of the right arch and the posterior en-



Figure 15

Angiocardiogram showing the typical elevation of the left pulmonary artery, the result of an anterior ductus.

circling left ligamentum to be high. That such does not occur frequently is explained diagrammatically by figure 13. The involutionary process, fortunately, most frequently involves the arch system caudal to the entrance of the ductus and results in the formation of a non-constricting left anterior ductus. Should the process involve the left arch proximal to the ductus insertion, a vascular ring will be formed. Blalock¹³ reported that approximately 75 per cent of 144 patients with a right aortic arch and cyanotic congenital heart disease had an anterior ductus (figs. 14 and 15). Thus, only 25 per cent of such individuals are potential candidates for an obstructive vascular ring. Again, due to looseness of the ring, obstructive symptoms are present in extremely few cases. Much clinical skill is necessary to differentiate the symptoms caused by the obstruction and the symptoms of associated congenital heart disease. This

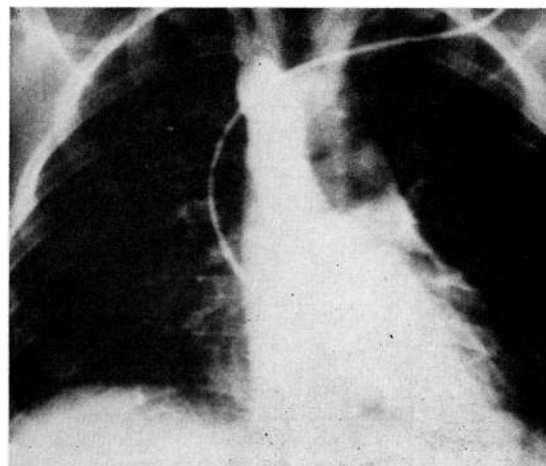


Figure 16A

Angiocardiogram showing right arch giving rise to a left innominate artery.

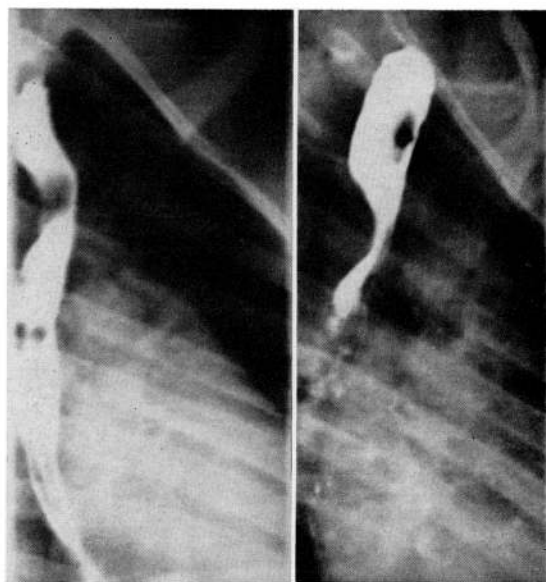


Figure 16B

The esophagogram establishes the posterior passage of the left innominate artery.

is especially so in the patients with a right aortic arch.

Interruption of the arch system at either point 4 or point 5 (fig. 10) may produce a double obstruction. This second point of obstruction is the result of a dextraposed aberrant vessel, which must now cross the midline to reach its definitive position on the left.

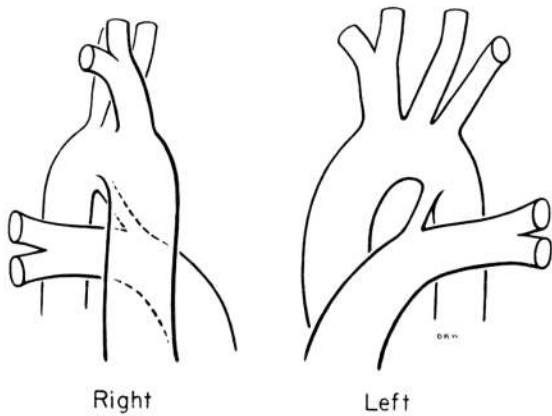


Figure 17

Unilateral absence of either the right or left pulmonary artery often results in the persistence of the opposite arch, ductus arteriosus, and pulmonary artery branch.

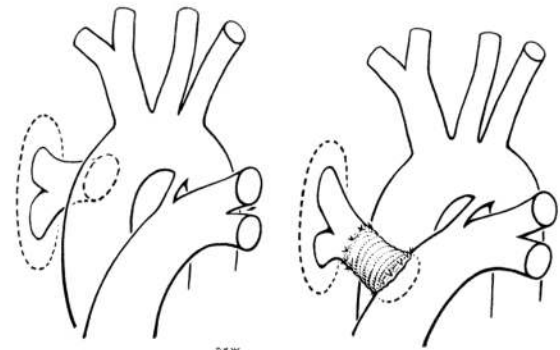


Figure 19

Diagram of absence of proximal portion of right pulmonary artery. The pulmonary artery arises directly from the ascending aorta. This is another example of bilateral persistence of the ductus. A suggested operative correction is shown (after Caro et al.²⁸).

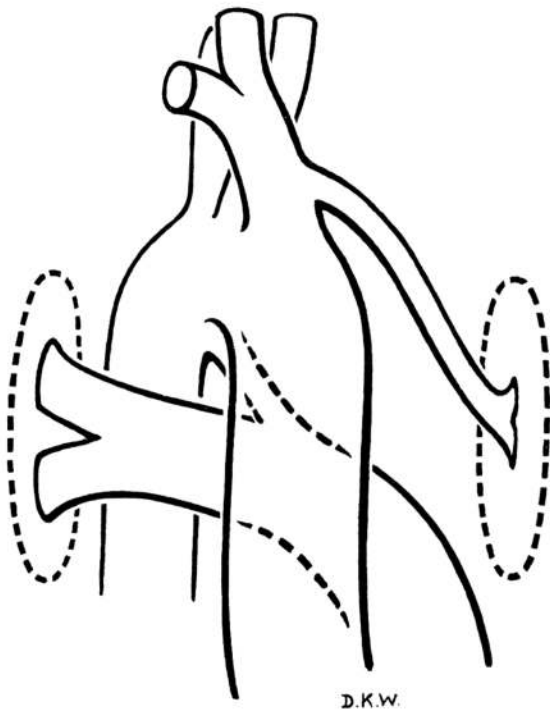


Figure 18

Origin of the blood supply to the left lung from the innominate artery in a case of absence of the left pulmonary artery. This represents a persistence of the ductus arteriosus.

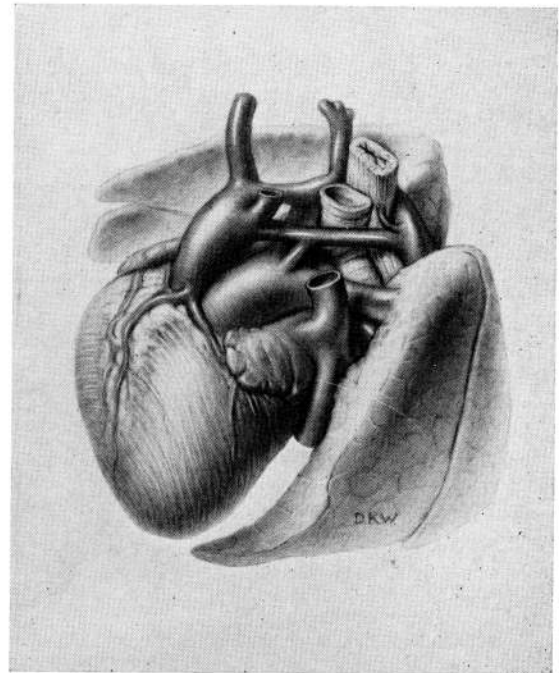


Figure 20

Extremely primitive vascular pattern showing bilateral aortic arches and bilateral patent ductus arteriosus. Note, however, the looseness of the vascular ring, a feature also described by others.²⁹

Gross and Neuhauser¹⁷ have emphasized the anterior "bowstringing" of such a vessel across the anterior trachea. Less well known

is the posterior passage of those vessels. Our material contains examples of posterior passage of both an aberrant left innominate (fig. 16A and B), and an aberrant left subclavian,

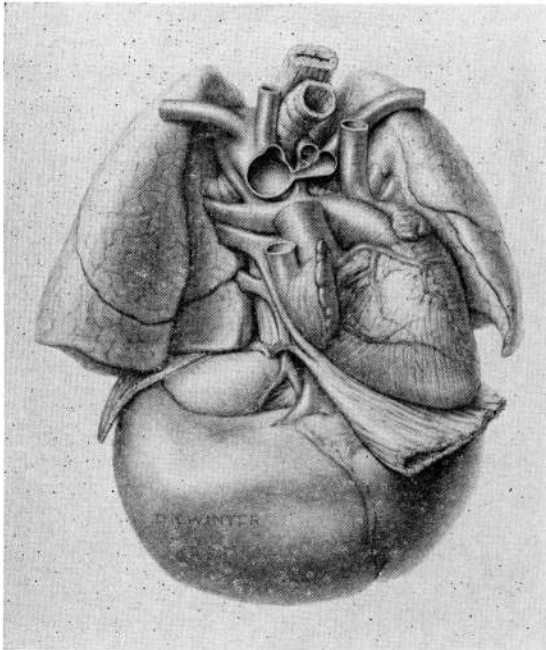


Figure 21

Bilateral aortic arches with patent right and left ductus arteriosus. Additionally, there is anomalous pulmonary venous return to the portal system and a persistent left anterior cardinal vein.

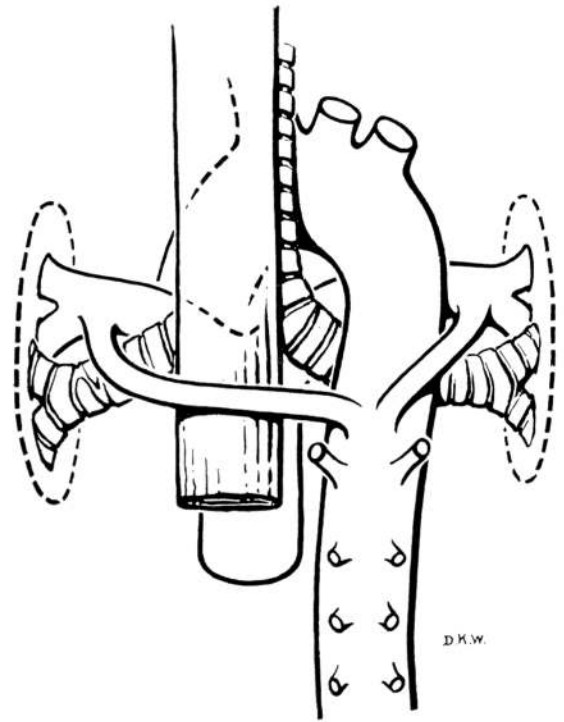


Figure 23

Posterior view. Note vessels arising from the descending aorta to pass to the pulmonary arteries. These vessels probably represent bronchial arteries. (After Findlay and Maier (case 3).²⁵)

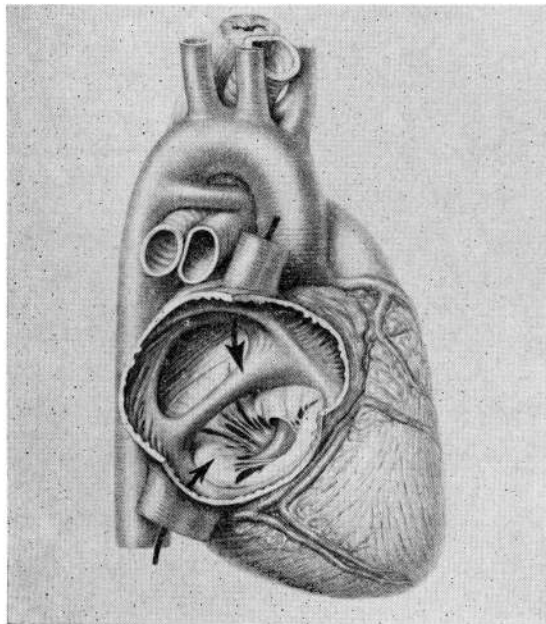


Figure 22

This drawing of a specimen shows double aortic arches, right and left ductus, and an atrioventricular canal; an extremely primitive arrangement.

each in association with a right aortic arch, these vessels being the cause of second points of obstruction. Unawareness of the posterior passage of these vessels may account for incomplete surgical relief in some cases.

A right ductus arteriosus may be seen in conjunction with a right aortic arch. This anomaly was encountered five times in this series and has been encountered and discussed by others.^{18, 19} Its unexpected presence at open-heart surgery may present considerable technical difficulties. For convenience, discussion of the related problem of the bilateral ductus arteriosus is deferred until later.

Absence of the contralateral pulmonary artery is occasionally seen in the patient with a right aortic arch. Since portions of both the pulmonary artery and the ductus have their common origin in the sixth branchial arch, absence of the pulmonary artery may be explained by an exaggerated involution of

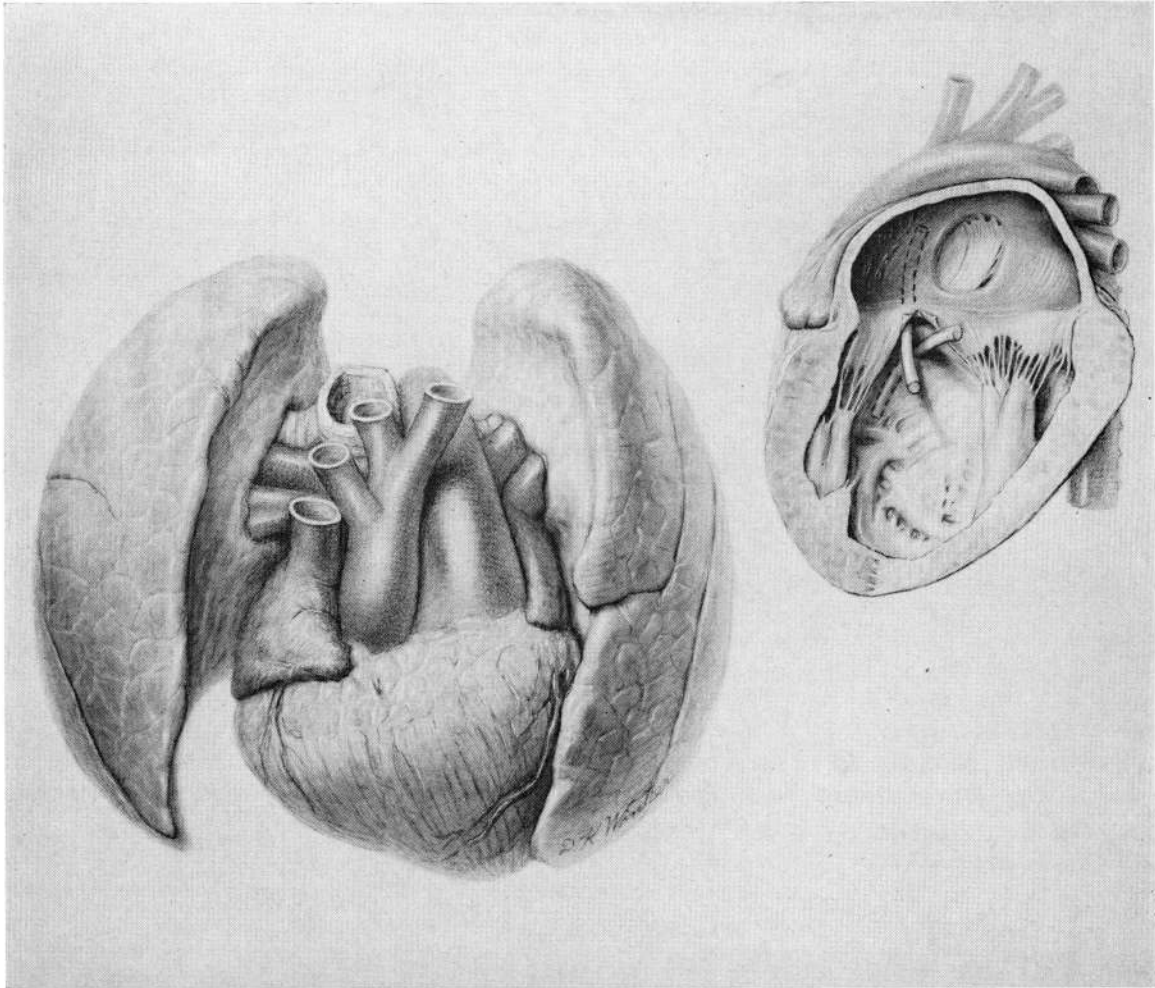


Figure 24

Drawing of the heart and vessels in an atresia or absence of the isthmus. In this case interruption of the arches has occurred at points 1 and 4 in figure 10.

the ductus with involvement of the pulmonary artery. There being no ductus or pulmonary artery, the arch on the affected side is in an unfavorable hemodynamic situation compared to the opposite, and will probably disappear. Thus, this would explain the frequently associated findings of the right aortic arch, right ductus, and an absent left pulmonary artery or its converse; the left aortic arch, left ductus, and an absent right pulmonary artery (fig. 17). The dangers presented by these anomalies during the construction of a pulmonary systemic artery shunt is again emphasized.²⁰⁻²³ Complete correction by means of open-heart surgery should be safer in that the presence

of two pulmonary arteries is not required as for successful accomplishment of a shunt procedure.²⁴

When the pulmonary artery is absent, how is the lung supplied with blood? Findlay and Maier^{25, 26} have described the many possible sources of supply in such situations. Two sources, however, should be emphasized, i.e., the bronchial arteries and vessels arising near the base of the innominate artery. This latter point of origin may often fallaciously be regarded as representing a bronchial artery (fig. 18). We think, however, as do others, that the vessels arising near the base of the innominate artery may represent persistence

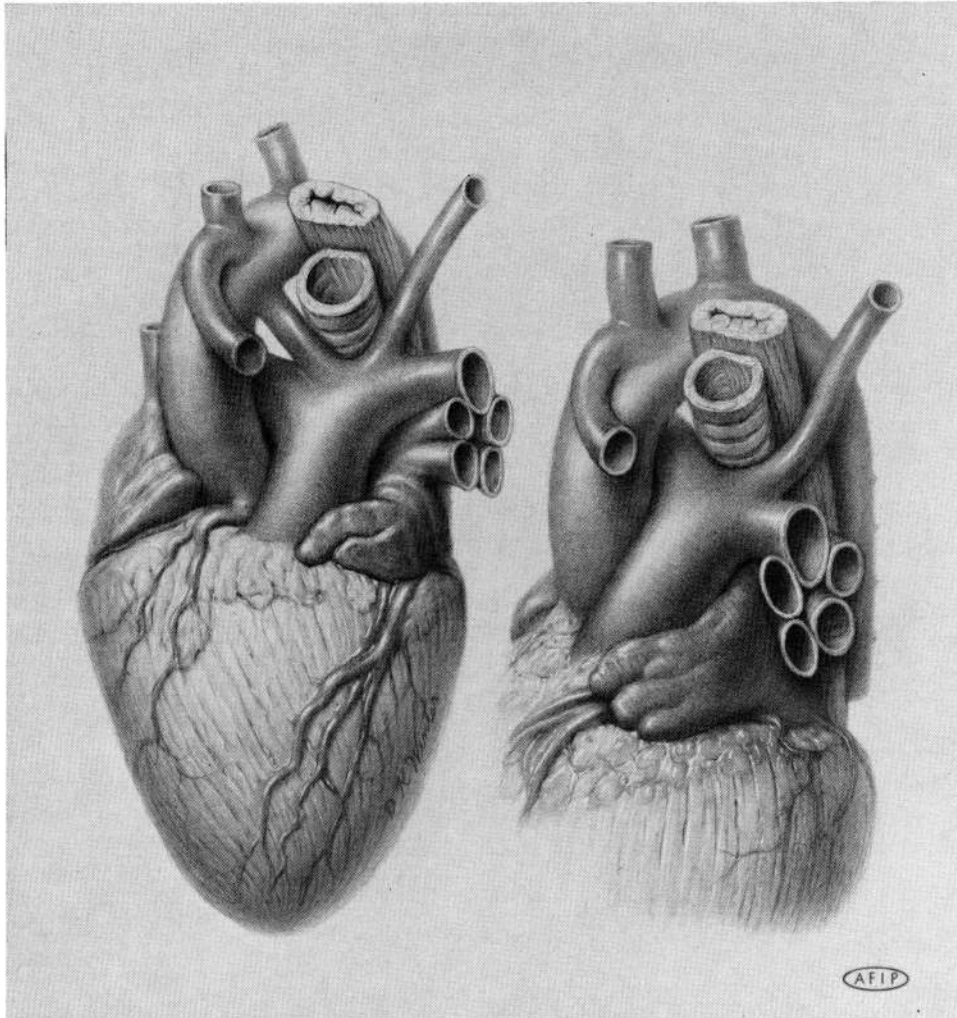


Figure 25

In these cases double interruption of a single arch (left) has occurred at points 4 and 5 in figure 10. Preoperative diagnosis is possible.

of the left ductus.²³ The situation shown in figure 18 closely resembles that shown in figure 14 with the addition that regression has involved the proximal pulmonary arterial portion of the left sixth branchial arch. An analogous situation with the ductus arising near the base of the right innominate artery has been described by Caro et al.²⁸ Shown in figure 19 is a left arch and a left ductus. The proximal portion of the right pulmonary artery is absent, and the blood supply to the right lung is directly from the aorta. This vessel also, we believe, represents a persistent

ductus rather than bronchial artery. The site of origin of this artery from the aorta, that is, at, or near, the origin of the innominate artery is consistent with a right-sided ductus arteriosus. This anomaly is of clinical interest, since the patient presents with unilateral pulmonary hypertension. Severe and repeated unilateral hemoptyses have been described.^{23, 27} The attempted operative correction of such a case by Caro et al.,²⁸ is depicted in figure 19.

Bilateral ductus may occur in several variations. Double aortic arches with double ductus

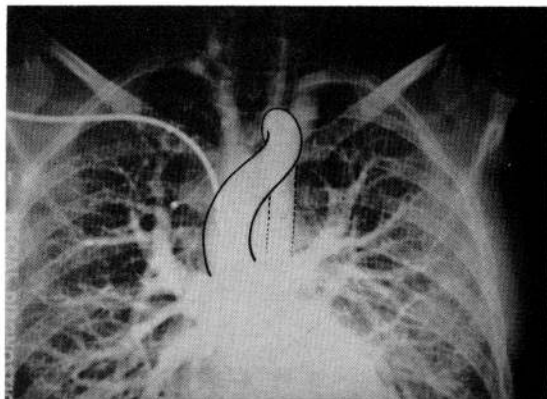


Figure 26

Angiocardiogram in young adolescent girl with congenital elongation of the aorta. A complete inverted U loop is described.

are seen in figures 20, 21, and 22. Previously mentioned were cases where the second ductus arose from the ascending aorta or base of the innominate artery. These are shown in figures 18 and 19. An additional variation is seen in figure 25. Other examples have been reported by several authors.³⁰⁻³⁴

We believe that the case described by Findlay and Maier²⁵ (case 3) probably represents not an example of a bilateral persistence of the ductus, but rather only exaggerated bronchial arteries (fig. 23). Embryologically true pulmonary arteries should arise proximal to the aortic arch branches, otherwise one must assume a transposing of the pulmonary arteries (a derivative of the sixth branchial arch) cephalad to the origin of other vessels, innominate and carotid arteries, derivatives of the third and fourth branchial arches.³⁵ In our experience a right aortic arch is often accompanied by overdevelopment and enlargement of the bronchial arteries. It should be mentioned that esophageal obstruction from an enlarged bronchial artery has been described and corrected.³⁶

Involution of the arch at point 5 (fig. 10) may produce two points of tracheal and esophageal obstruction. While involution at point 6 would result in a right aortic arch, the presence or absence of a vascular ring would depend on the insertion of the left ductus. There

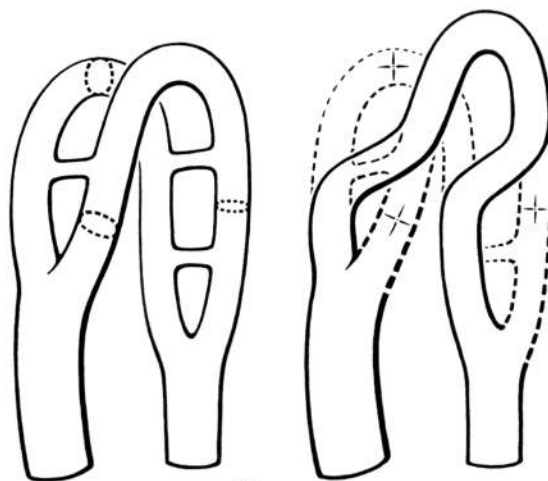


Figure 27

The paired embryonic dorsal aortae have numerous cross connections. Obliteration of alternating points on opposite aortae may cause development of these channels and a resultant elongation or tortuosity of the aorta. Another explanation offered for this elongated loop is a failure of descent from the earlier cervical position.

would, however, be no anterior bowstringing.

So far we have restricted the analysis to but a single limb of the genealogy tree (fig. 8 b-2). Similar analyses might be made for the other five limbs with a result of 36 possible vascular anomalies—all based on the assumption of involution occurring at a single point only. Unfortunately, the problem is not so simple. Involutional changes might occur at two or more points, either on a single arch or at separate points on each arch with the resultant production of aortic atresia, an aberrant origin of a systemic artery from the pulmonary artery, or a congenital elongation or looping of the aorta. Multiple points of regression involving both arches result in aortic arch atresia (fig. 24). We, with others, have successfully treated this lesion surgically, so that this anomaly is worthy of more than academic interest.^{37, 38} Some configurations may lend themselves to a staged surgical correction.

If multiple points of regression affect a single arch, a systemic vessel may arise from a pulmonary artery. In the right-sided por-

tion of figure 25 the anatomic configuration shown was predicted on the basis of the clinical findings of a ductus murmur, a right aortic arch and a weakly palpable left radial pulse. This configuration has been described by others and is quite common in veterinary reports.^{5, 39-41}

Multiple alternating points of involution affecting both arches may cause congenital elongation or "looping" of the aorta.^{42, 43} When this occurs in an infant without evidence of hypertension or arteriosclerosis, the elongation is logically, we believe, attributive to an embryologic maldevelopment (fig. 26). For a possible explanation of the formation of this anomaly, see figure 27. A similar mechanism might explain those cases of right aortic arch in which the aorta quickly crosses the midline to descend on the left side,⁴⁴ or its mirror image, that is the left aortic arch in which the aorta quickly crosses the midline to descend on the right side;⁴⁵ also, the explanation may be similar for the particularly tortuous aorta, in which there is a left arch with the aorta initially passing to the right but quickly returning to the left to exit through the diaphragm. Only with the reporting of additional cases will a better understanding of these anomalies be possible.

Summary

Forty-one well-studied thoracic arterial arch anomalies are related to cases previously reported in the literature and presented with simplified drawings in an attempt to provide a workable understanding of these anomalies.

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