

## NOTES ON THE ANOMALIES OF THE AORTIC ARCH AND OF ITS LARGE BRANCHES

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TWO HELIOTYPE PLATES (FOUR FIGURES)

Variations and anomalies of blood vessels are frequently met with and are thought to be due to various developmental factors, among which we may name the following: the potency of the fertilized ovum, the mechanical influence of the neighboring organs, and the direction, rapidity, and pressure of the blood stream. There is, as yet, much confusion concerning the development of even the large blood vessels. In clearing up this confusion the interpretation of every experiment of nature—every anomaly and variation—is important. The anomalies described here do not illustrate extremely rare cases. However, certain authors state that, for a complete explanation of the variations and anomalies of blood vessels, there is still a deficiency of material illustrative of the less rare cases. Therefore, we are describing the following two cases of anomalies which were found in the anatomical laboratory of the University of Latvia during the dissection of two adult, normally developed, male bodies.

### CASE I. ANOMALOUS RIGHT SUBCLAVIAN ARTERY (figs. 1 and 2)

The heart is normal. The aortic arch and the thoracic aorta have a normal shape, size, and an almost normal course; the thoracic aorta shows a bend to the right. The branches of the aortic arch are: the right common carotid, the left common carotid, the left subclavian, and the right subclavian. The right common carotid arises from the upper

surface of the aortic arch, where the latter is placed on the right side of the trachea, and goes directly upward. The left common carotid originates immediately to the left side of the right common carotid (with which it corresponds in size) and from the ventral surface of the aortic arch. It runs over the ventral surface of the trachea to the left side of it. Close to the origin of the left common carotid is the beginning of the left subclavian, which is of normal size and direction and branches normally. The right subclavian artery arises from the upper part of the dorsal surface of the aortic arch and at the left side of the left subclavian artery. The origin of this artery is just to the left of the oesophagus and trachea; then the vessel passes over the dorsal surface of the oesophagus to the right side; where it crosses the oesophagus it is 5 cm. distal from the beginning of that organ. On the right side of the oesophagus it curves downward and branches.

The right vagus nerve, in the beginning, follows a normal course along the right side of the oesophagus and trachea and in front of the right subclavian artery. It has no recurrent nerve. All the branches passing to the larynx, trachea, and oesophagus are independent and arise close to the level of their final distribution. The general direction of these branches is downward and medialward. The left vagus nerve is normal in its direction and branching.

CASE II. RIGHT AORTIC ARCH (figs. 3 and 4)

The heart is enlarged. The beginning of the aorta is covered by the pulmonary artery and the right auricle of the heart. The ascending part of the aorta passes directly upward along the right side of the trachea and oesophagus, then curves suddenly backward and to the left. On the left side of these organs it makes a sharp curve downward; on its farther course it is on the dorsal surface of the oesophagus, in this position exhibiting a curve toward the right. On the right side of the ascending part of the aortic arch is the superior vena cava, which receives the azygos vein lying on the dorsal surface of the aorta. The origin of the pulmonary

artery is normal; this artery passes to the left and, at the level of the bifurcation of the trachea, divides into its two branches. From the upper surface of the bifurcation originates the ligamentum arteriosum, which crosses the ventral surface of the trachea on the left side running upward to join the left subclavian artery. The branches of the aortic arch are: the left common carotid, the right common carotid, the right subclavian, and the left subclavian arteries. The first branch arises on the medial border of the ventral surface of the descending aorta, where the latter reaches the right side of the trachea and then swings around the ventral surface of the trachea passing somewhat forward in its course. The right common carotid arises a little more cranially and to the right of the first branch, while the right subclavian artery takes its origin from the uppermost part of the aortic arch and still farther to the right. The fourth branch—the left subclavian artery—arises from that part of the aortic arch which begins to curve downward, the origin, which is conical in form, being on the ventral surface of the arch. This conical enlargement lies just at the left of the oesophagus and is joined by the ligamentum arteriosum. The walls of the aortic arch are sclerosed, and the vessel itself is enlarged. The relations between the trachea and the oesophagus are normal. In addition to the abnormal course of the aortic arch, the courses of the vagi and the recurrent nerves are not quite normal.

The left vagus nerve has a normal course; its recurrent nerve goes around the ligamentum arteriosum to its place of distribution. The right vagus nerve courses over the dorsal surface of the aortic arch, then goes between the azygos vein and the lower surface of the arch, where it gives off the recurrent nerve, which passes under the aortic arch running in the ventromedial direction to reach its normal position.

## DISCUSSION

The literature concerned with the variations and anomalies of the aortic arch and its branches is large, particularly that dealing with the abnormal course of the right subclavian artery. The first extensive compilations of the different anomalies and variations of this kind were given by Quain ('44) and by Henle ('74). In 1883, Brenner referred to the relations between these anomalies and the course of the vagi and recurrent nerves. Holzapfel listed all the cases (200) of the right subclavian artery as the last branch of the aortic arch which had been described in the literature up to 1899, and gave very complete details of the characteristics of this anomaly; he also described the relations existing between this anomaly and the neighboring organs. This contribution gives the most complete consideration of this question to the present time. All later cases (from 1899 to 1907) of this anomaly have been compiled by Banchi. Milianitch and Kadanoff, in 1924, published two cases of the aortic-arch anomalies; Kadanoff also cited the most recently published cases. The last paper, within our knowledge, concerned with this question is the one published in 1925 by Cairney. This author described two cases of the right subclavian artery as the last branch of the aortic arch, and gave, also, a review of such cases of this anomaly as had been published in England and America. It appears to us that the authors of some of these later papers have arrived at certain erroneous conclusions by reason of their unfamiliarity with the earlier literature; this criticism would apply particularly to the work of Milianitch.

The two anomalies described in the present paper illustrate the most frequent and most characteristic cases, and therefore it is unnecessary to discuss them more completely. From the literature, it may be seen that the most frequent anomaly in the development of the aortic-arch system is the abnormal beginning and course of the right subclavian artery (case I). Up to the present time, there have been published about 260 such cases (231 up to 1907). Of course, there are undoubtedly

many cases which have not been published; for example, in the Anatomical Museum of the University of Michigan there are four such cases. On an average, this anomaly is found in 0.4 to 0.8 per cent of cases, i.e., nearly 1:167. In such specimens the course of the right subclavian artery behind the oesophagus is to be regarded as the usual position. Out of the 165 cases compiled by Banchi ('07), the right subclavian artery coursed between the oesophagus and trachea in twenty-one cases (12.5 per cent) and in front of the trachea in seven cases (4 per cent). The most frequent disposition of the aortic-arch branches is: the right common carotid, the left common carotid, the left subclavian, and the right subclavian arteries. A true right recurrent nerve does not exist when the right subclavian has this anomalous origin. All the nerve branches which usually are included in the right recurrent nerve pass gradually to the region of distribution directly from the right vagus nerve. Krause and Telgman (*Die Nervenvarietäten des Menschen*, Leipzig, 1868) were the first to give an explanation for such a course of the branches of the right vagus nerve. Much more rare are the cases where the right recurrent nerve exists and embraces the vertebral, the inferior thyroid, or the right subclavian arteries. Some cases are known (Pinge, Dubrueil, Banchi) where the right vagus nerve passed behind the right subclavian artery.

The anomalies in which the normal left aortic arch is substituted for by a right aortic arch are more rare than the previously mentioned group of anomalies. Twenty-eight such cases have been described in the literature, our case making the twenty-ninth. In all the published cases the right aortic arch reached the right border of the oesophagus and then passed behind this organ and to the left of it. Only in Dubrueil's case (*Les anomalies artérielles*, Paris, 1847) it passes between the trachea and the oesophagus. The left subclavian artery, in the presence of a right aortic arch, lies behind the oesophagus, with the exception of the case described by Kadanoft, in which it passed between the oesophagus and trachea. When there is a right aortic arch, the *ligamentum arteriosum*

(sometimes a persistent ductus Botalli) is, as a rule, on the right side. Cases with a right ligamentum arteriosum are very seldom found. Such cases have been published by Abernethy (1793), Quain ('44), Brechet (1826), and Brenner ('83). Usually, when the right aortic arch is present, the right recurrent nerve embraces the aortic arch and the left recurrent nerve swings around the ligamentum arteriosum. Gottschau's case ('87) represents the only one where both recurrent nerves embraced the subclavian arteries. The course of the right vagus nerve behind the aortic arch can be very simply explained in the case we are presenting (figs. 3 and 4). By turning the aortic arch from the right to the left, the anterior surface becomes the posterior surface and the nerve is displaced in the same direction.

In connection with the two groups of anomalies just discussed, cases of double aortic arches should be mentioned, i.e., the reptilian form of the aortic arch. These anomalies are comparatively rare, only about fifteen such cases being described in the literature. In these cases the double arches join behind the oesophagus. Only one case (Zagorsky, 1824) is reported where the junction occurred between the trachea and the oesophagus; in this last-mentioned case the right aortic arch passed between the trachea and the oesophagus. We shall not consider further the different characteristics of these three groups of anomalies, since detailed discussions of them are easily available in the works of Brenner ('83), Holzapfel ('99), Cairney ('25), and others. Our aim is to consider here the factors underlying the occasional development of parts of the aortic-arch system which ordinarily disappear and, likewise, those factors which occasionally produce the disappearance of such parts of the aortic system as are normally present.

Rathke and von Baehr were the first to point out that the development of the heart and of the six doubled arches is the same in all vertebrates. Turner ('62), on the fundamental basis of a common embryological origin, systematized all the variations of the aortic arch and of its branches.

Turner's work was extended by Krause ('56 and '76, in Henle's Anatomy) by means of diagrams. From the embryological point of view, it is not difficult to explain many of the different anomalies of the aortic arch. It is not easy, however, to explain those cases in which the blood vessels pass ventral to the oesophagus.

In this connection it is necessary to mention that Cairney, relying on the work of Geddes ('11), is inclined to deny entirely the possibility of the presence of the right subclavian artery in front of the trachea; he says there is "no reliable case in which the artery is shown to pass in front of the trachea." Cairney doubts all those cases referred to by Holzappel in which the right subclavian artery passes between the oesophagus and the trachea, and states that only one case (Bradley, '71) has been published in which such a course was found. Cairney, at the time his paper was published, did not have a first-hand acquaintance with Holzappel's work (he writes, "Holzappel, whose paper I have unfortunately not been able to consult"), and it is also evident that he was unfamiliar with the cases described by Dubrueil, Zagorsky, and Kadanoff. There is no doubt that, when certain aortic-arch variations and anomalies are present, the pre-oesophageal course of the large blood vessels occurs, i.e., the course between the trachea and the oesophagus and in front of the trachea. These last-named anomalies cannot be explained by the aid of Rathke's diagrams; indeed, thus far we have not been able to find an adequate explanation for them, although hypotheses, attempting to explain the conditions, have been given by several authors. Holzappel regarded these cases as dependent upon the existence and considerable development, in early embryologic stages, of pre-oesophageal anastomoses between the roots of the ascending aortic arches and the anlage of the subclavian artery. The Kadanoff figures (nos. 4 and 5) illustrate this theory. Banchi tried to explain the anomalies on the basis of a total or partial persistence of the fifth aortic arch. That this last hypothesis is not acceptable has been clearly shown by

Kadanoff, who pointed out that the relations between the recurrent nerves and the right subclavian artery demanded by the acceptance of this theory are quite different than the relations existing when the actual anomalies are present. Cairney wrote: "One can but suggest that in this case [i.e., the Bradley case], the fusion of the paired aortae to form the unpaired aorta may have occurred, at any rate in its cephalic part, ventral to the alimentary canal instead of dorsal to it; and that subsequently the anomalous artery was developed by the incorporation of the terminal segment of the right dorsal aorta in the usual manner." This audacious suggestion of Cairney, in explanation of these anomalies, is very difficult to accept; in fact, it appears impossible. Regarding these hypotheses, Kadanoff ('24) wrote as follows: "Diese Erklärung [i.e., Holzappel's] scheint mir die Einzige zur Zeit mögliche sein. Sie hat nur den Wert einer Notbrücke, solange nicht weitere Befunde vorliegen."

It is evident, from the preceding brief discussion, that Rathke's diagram needs further consideration and modification. Congdon, working with the human embryo, has already given some important results in this direction. The question about the relations existing between the different anomalies of the aortic arch and the course of the vagus and recurrent nerves has been thoroughly solved, from an embryological point of view, by Brenner ('83). We know definitely now that these relations depend upon the transformation of the branchial and aortic arches and upon the displacement of the heart.

In connection with the above-mentioned discussion, attention should be called to Backman's suggestion (based mainly upon Funcius' investigations) that the heart is a 'punctum fixum' and that the anterior part of the body grows cranialward along the heart; this latter condition giving the appearance of a descent of the heart. It seems to us that both conditions must be recognized here; that is, the heart does reach its final position by a caudal displacement, but that at the same time there is forward growth of the anterior part of the body, particularly of the neck region.



With reference to the underlying causes of the different forms of aortic-arch anomalies and of the anomalies of its branches, most authors agree that they are caused by the different directions of flow of the blood stream itself. However, we have been unable to find any definite statements in the literature regarding the causes underlying the changes in direction of the blood stream. We regard it, at present, as dependent upon three factors: 1) incomplete development or total lack of development of some part or parts of the aortic-arch system; 2) different external, mechanical influences, which do not allow the blood to pass through some portion or portions of the aortic arches—for instance, pressure of the neighboring tissues or organs, and, 3) different position of all or of a portion of the heart, as, for example, of the truncus arteriosus, atria, etc. It is possible that sometimes two of these factors or even all of them may operate together. The first two factors mentioned above are probably the main causes for the appearance of those anomalies where the single parts of the dorsal or descending aortae disappear, i.e., do not develop; for instance, the anomalous cases of the right subclavian as the last branch of the aortic arch. In some of these cases Cairney's explanation may be the correct one. It must be admitted that, up to the present time in the cases described, no anomaly has been explained on the basis of the first two factors enumerated above by us, but this does not preclude the existence of such factors. Of course, the possibility of finding direct evidences of the influence of these factors in the fully developed aortic system is very slight, because these factors act during very early embryological stages and the traces of their action may entirely disappear later on.

The last of the three factors mentioned above has been taken in account by various authors in considering the anomalies of the ventral or ascending parts of the aortic arches; but here also the possible operation of the first two factors must not be neglected. For example, von Baehr (*Entwicklungsgeschichte der Tiere*, Tome II, Königsberg,

1837) wrote: "dass die Stellung der beiden Herzkammern und die davon abhängige Richtung des Blutstromes das eine Mal vorwaltende Benutzung und Ausweitung des linken vierten Gefassbogens bedingt, das andere Mal wieder des rechten." Rathke (Müller's Archiv, 1843) stated that the difference in the development of the aortic-arch system depended upon the direction of the truncus arteriosus and the axis of the heart itself. From this statement one can draw the conclusion that, in cases showing the development of a right aortic arch, the heart, in early development, must have the same position, which is normal for birds where the fourth right arch always gives the normal existing arch. Holzapfel did not give detailed consideration to the question, but said: "weshalb der Blutstrom in einzelnen Fällen sich andere Wege offenhält, kann weder im allgemeinen, noch im Speciellem angegeben werden." Kegel wrote: "die verschiedenen Fehlbildungen am Herzen und den grossen Gefässen sind nach Ziegler wohl besser als Hemmungsbildungen aufzufassen (wie wohl auch mancher Sit. inver. part. eine Solche sein dürfte)."

It seems to us that the development of the right aortic arch depends mainly upon the position of the axis of the heart. However, when we accept the statement that the development of the right or the left aortic arch is dependent upon the direction of the blood stream in connection with the different positions of the heart, then we must go further and ask: Upon what does the position of the heart itself depend? To this question scientists cannot, as yet, give a definite answer, although certain hypotheses have been offered. Kegel ('25), in his work "Über Situs inversus totalis," considered in detail the different theories explaining the position of the heart. Since the presence of a right aortic arch in human is really a case of partial situs inversus, Kegel's statements also demand consideration in the explanation of such an anomaly. Of all the theories discussed by Kegel, the two following appear to us to be the most acceptable for explaining a right aortic arch: 1) The first theory takes into con-

sideration the well-known fact that the embryo lies with the left side normally toward the yolk sac. Any change from this normal position might well induce a corresponding shift in the direction of the axis of the heart (von Baehr, Remak, Schulz, Dareste, and Ahlfeld). 2) A second theory which deserves consideration is that a change in the normal spiral turn of the embryo may be accompanied, presumably, by an alteration in the direction of the developing heart's axis (D'Alton and Günther). When, in the future, further investigations shall have been made along these lines, it may be possible to explain why in mammals the left aortic arch develops, in birds the right aortic arch, and in reptiles the double aortic arch; we shall also understand the causes of some of the aortic anomalies. Furthermore, such investigations will give a better basis for estimating the truth represented by the diagrams of Rathke. Cairney wrote in regard to this matter: "Reference is made to the recent research of Congdon on the developmental changes in the aortic arch system in the human embryo, from the study of which it appears evident that the classical descriptions of this portion of embryology as they appear in most of our standard text-books require revision." The investigations in this direction, it is hoped, will throw further light on the preoesophageal course of the large blood vessels of the aortic arch—a condition existing in certain of the anomalies. We are convinced that we shall have a better conception of the different anomalies and variations of the aortic arch and its branches when, in the future, other cases have been described with care and exactness.

In conclusion, we wish to mention that the occasional development of the embryologic right fourth aortic arch as the adult aortic arch of man or certain other mammals (as rabbit or hedgehog) has significance in the discussion of the isotopy of all homologous body parts of the metazoan organism. The occasional presence of the right aortic arch in mammals indicates that the isotopy of this structure, which has been in abeyance for many, many thousand years, may

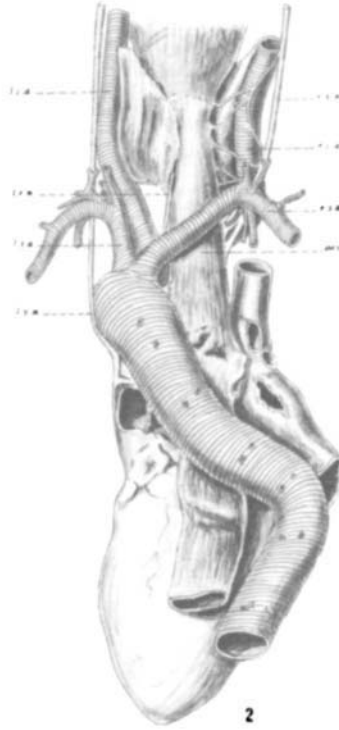
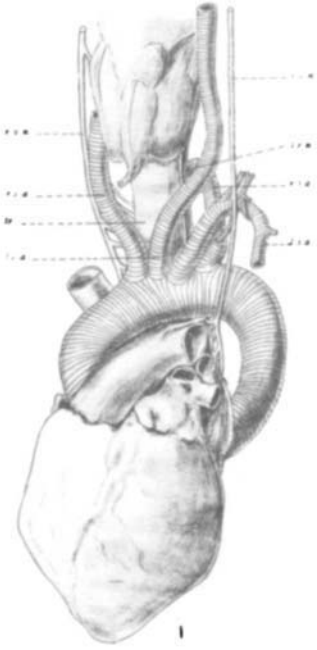
sometimes, under certain circumstances, become active and make possible the development of an organ functionally normal in all respects. We may even regard this isopotency—i.e., this latent morphogenetic potency—as not really quiescent in such a case, but as developing with the left side of the body, i.e., with the fourth left aortic arch. Only on the basis of the existence of this isopotency is the possibility given of the development of the right aortic arch. According to Lebedinsky, we have in the right aortic arch of mammals and of man an example of the isopotency of the homotypic parts of the body.

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## PLATE 1

### EXPLANATION OF FIGURES

1 Anomalous right subclavian artery; heart and major vessels, case 1, ventral view; one-third natural size. *l.c.a.*, left carotid artery; *l.r.n.*, left recurrent laryngeal nerve; *l.s.a.*, left subclavian artery; *l.v.n.*, left vagus nerve; *r.c.a.*, right carotid artery; *r.s.a.*, right subclavian artery; *r.v.n.*, right vagus nerve; *tr.*, trachea.

2 Anomalous right subclavian artery; heart and major vessels, case 1, dorsal view; one-third natural size. *l.c.a.*, left carotid artery; *l.r.n.*, left recurrent laryngeal nerve; *l.s.a.*, left subclavian artery; *l.v.n.*, left vagus nerve; *oes.*, oesophagus; *r.c.a.*, right carotid artery; *r.s.a.*, right subclavian artery; *r.v.n.*, right vagus nerve.

## PLATE 2

### EXPLANATION OF FIGURES

3 Right aortic arch; heart and major vessels, case 2, ventral view; one-third natural size. *l.c.a.*, left carotid artery; *l.r.n.*, left recurrent laryngeal nerve; *l.s.a.*, left subclavian artery; *l.v.n.*, left vagus nerve; *lig.art.*, ligamentum arteriosum; *oes.*, oesophagus; *r.c.a.*, right carotid artery; *r.s.a.*, right subclavian artery; *r.v.n.*, right vagus nerve; *tr.*, trachea.

4 Right aortic arch; heart and major vessels, case 2; one-third natural size. *l.c.a.*, left carotid artery; *l.r.n.*, left recurrent laryngeal nerve; *l.s.a.*, left subclavian artery; *l.v.n.*, left vagus nerve; *oes.*, oesophagus; *r.r.n.*, right recurrent laryngeal nerve; *r.v.n.*, right vagus nerve; *v.a.*, vena azygos.



