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CASE REPORT

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Right-sided aortic arch

Jacek Białowąs[1], Jolanta Hreczecha[2], Marek Grzybiak[2]

[1]*Department of Anatomy and Neurobiology, Medical University of Gdańsk, Poland*
[2]*Department of Clinical Anatomy, Medical University of Gdańsk, Poland*

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Congenital abnormalities of the aortic arch arise due to a defect in the unilateral disappearance of arteries of the IVth and exceptionally of the IIIrd primary branchial arches and also of the appropriate sections of paired dorsal aortas. Apart from the cases of complete "situs inversus" and a double aortic arch, the following anatomical possibilities can be distinguished: A — a left-sided aortic arch with a properly established system of branches, B — a left-sided aortic arch with an aberrant right subclavian artery, C — a left-sided aortic arch with a retro-esophageal course and right-sided descending aorta or retro-esophageal course of the brachiocephalic trunk onto the right side, D — a right-sided aortic arch of the „symmetric" type usually coexisting with cyanotic congenital heart lesions, E — a right-sided aortic arch with a retro-esophageal bulge and an aberrant left subclavian artery, and F — a right-sided aortic arch with an aorta descending left-sidedly or brachiocephalic trunk going left-sidedly behind the esophagus. At the Department of Anatomy from 1945 to 1998, 1700 adult cadavers were examined. Throughout this time, one case of each of the types E and C and two cases of the type B were noted in the material. Regardless of the rare occurrence among adults (about 0.01%), the abnormal course of the aortic arch can be the reason for atypical clinical symptoms such as esophageal compression and dysphagia or insufficient cerebral blood supply.

key words: right-sided aortic arch, embryology, clinical manifestations

INTRODUCTION

Congenital developmental abnormalities of the aortic arch and its branches can occur as an isolated lesion giving no serious clinical symptoms or can correlate with disturbances in the heart development and cyanotic lesions. Heretofore, Hunauld described the aberrant subclavian artery in 1735. Fioratti and Aglietti first reported on the right-sided aortic arch in 1763, and in 1818 Corvisart confirmed its presence in a case of Tetralogy of Fallot [cited after 7,8,12].

At the stage of a 30 mm embryo, a ventral aortic sac and paired dorsal aortas join at different times with 6 pairs of arteries of the branchial arches. Sections of the first three arches and their ventral and dorsal aortal connections create the carotid artery system. The paired dorsal aortas join together forming a descending aorta later usually moving onto the left side. The ventral portion of the right fourth arch develops into the right subclavian artery running off a common trunk with the right common carotid artery from the aortic arch, stemmed from the left fourth arch; however, a part of the right dorsal aorta from the outflow of the subclavian artery to the place of fusion with the contralateral one disappears. In this way, the most common normal type A develops — Figure 1A. The arteries of the fifth arches atrophy altogether, however those of the sixth take part in creating the pulmonary arteries and the ductus arteriosus. Sometimes atrophy of the right fourth arch can occur and then to the right subclavi-

Address for correspondence: Jacek Białowąs, MD, Department of Anatomy and Neurobiology, Medical University of Gdańsk, ul. Dębinki 1, 80–211 Gdańsk, Poland, tel: +48 58 349 14 01, fax: +48 58 349 14 21

an artery blood courses through the remaining lower section of the right dorsal aorta running beyond the esophagus (type B; Fig. 1B — aberrant right subclavian artery). If the artery of the third right arch also disappears, then the right brachiocephalic trunk leaves the aortic arch as the last branch and runs beyond the esophagus. Sometimes a retro esophageal course of the aortic arch from the left side to the right and a right-sided descending aorta occurs (type C; Fig. 1C). Analogically, three possibilities exist in the development of a right-sided aortic arch from the artery of the right fourth branchial arch with disappearance of appropriate sections of vessels on the left side (type D, E, F; Fig. 1D, E, F). It is important to note that type D is "symmetrical" to the normal type A, and is most often associated with cyanotic congenital heart lesions [4,5,7,8,15,21]. However, type E can be diagnosed already in infancy [14,17,18,21] or in adolescence [5] or can not be recognized at all, as in our case, due to light and atypical clinical symptoms.

CASE DESCRIPTIONS

In the Department of Anatomy at the Medical University of Gdańsk from 1945 to 1998, 1700 adult cadavers of both sexes were scrutinized. In this period of time, one case of a right-sided aortic arch (type E) was observed in the material, one case of a retroesophageal course of the aortic arch (type C) and two cases of aberrant subclavian arteries (type B).

CASE 1

During the preparation of the cadaver of a 70-year-old male who died owing to status epilepticus and pneumonia, upon opening the thoracic cage a nor-

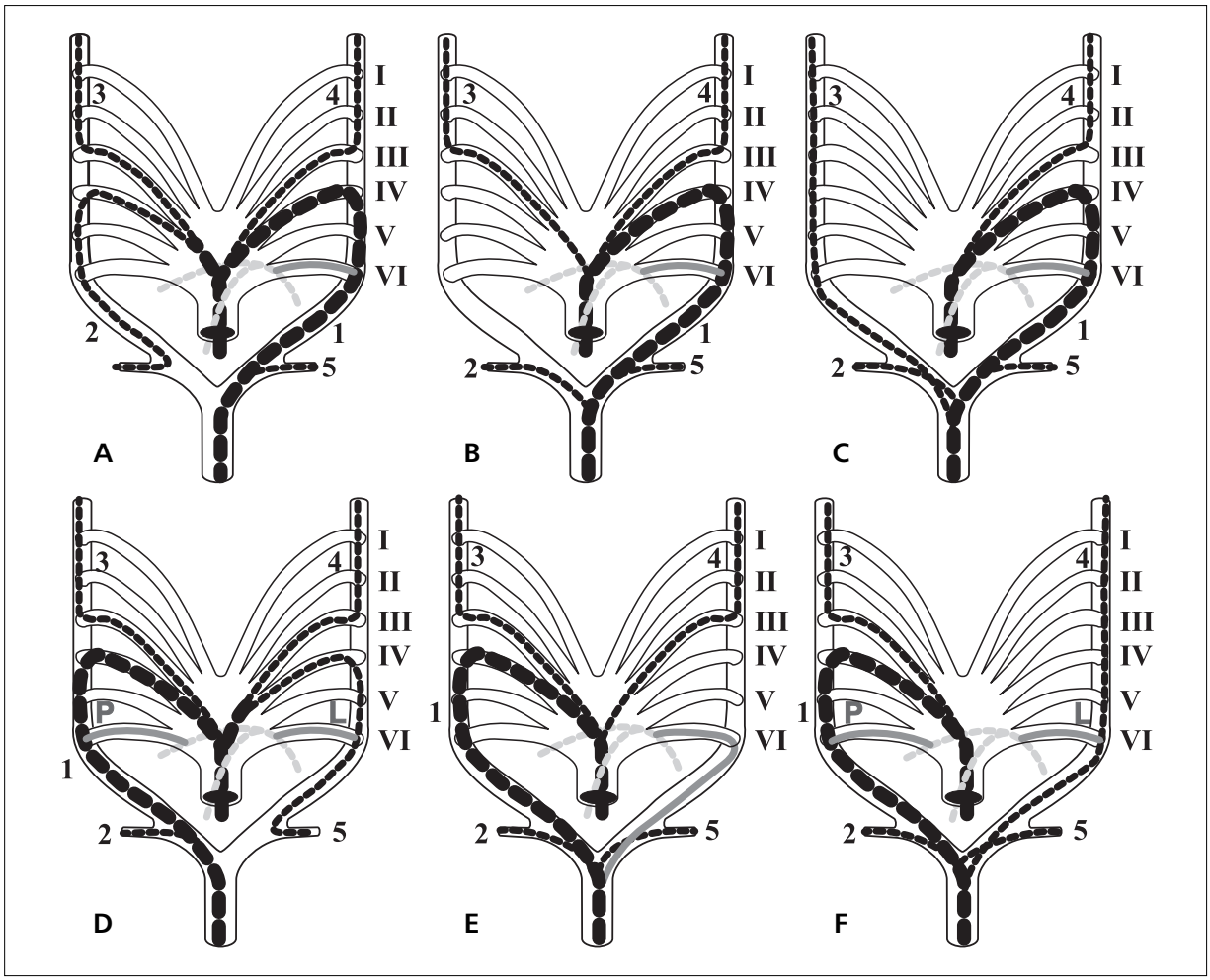

Figure 1. Types of course and branching of the aortic arch superimposed on the schemes of branchial arch arteries (I to VI) and dorsal aortas. Dashed lines — aorta and its branches after completion of the development. Gray lines — pulmonary arteries. Black line — arterious ligament. 1) Aortic arch; 2) Right subclavian artery; 3) Right common carotic artery; 4) Left common carotic artery 5) Left subclawian artery. A), B), C) — left and D), E), F) — right aortic arch. Detailed description of particular types from A to F in text.

mally built and located heart was observed. The aortic arch ran above the right main bronchus, along the right side of the trachea and esophagus (in an X-ray carried on at the hospital no other abnormalities apart from inflammatory changes were noted in the chest). Next, the aorta, somewhat widening, entered beyond the esophagus and the distal end of its arch was visible on the left. Here the left subclavian artery branched off upwards, and the anteroinferior part of the bulge joined the thick ligamentum arteriosum. From this point the descending aorta moves beyond the esophagus running towards the diaphragm along the right side of the vertebral column; the aortic hiatus was located posteriorly and subtly on the right of the esophageal hiatus. From the initial part of the aortic arch the following vessels branched off respectively: the left common carotid artery crossing the trachea in front, the right subclavian artery and right common carotid artery (Fig. 2). Recurrent nerves ran under the subclavian arteries. Therefore, the trachea and the esophagus were somewhat shut in a ring of large vessels — the aorta and pulmonary arteries connected by the ligamentum arteriosum.

The remaining three cases were observed by investigating museum preparations from the early

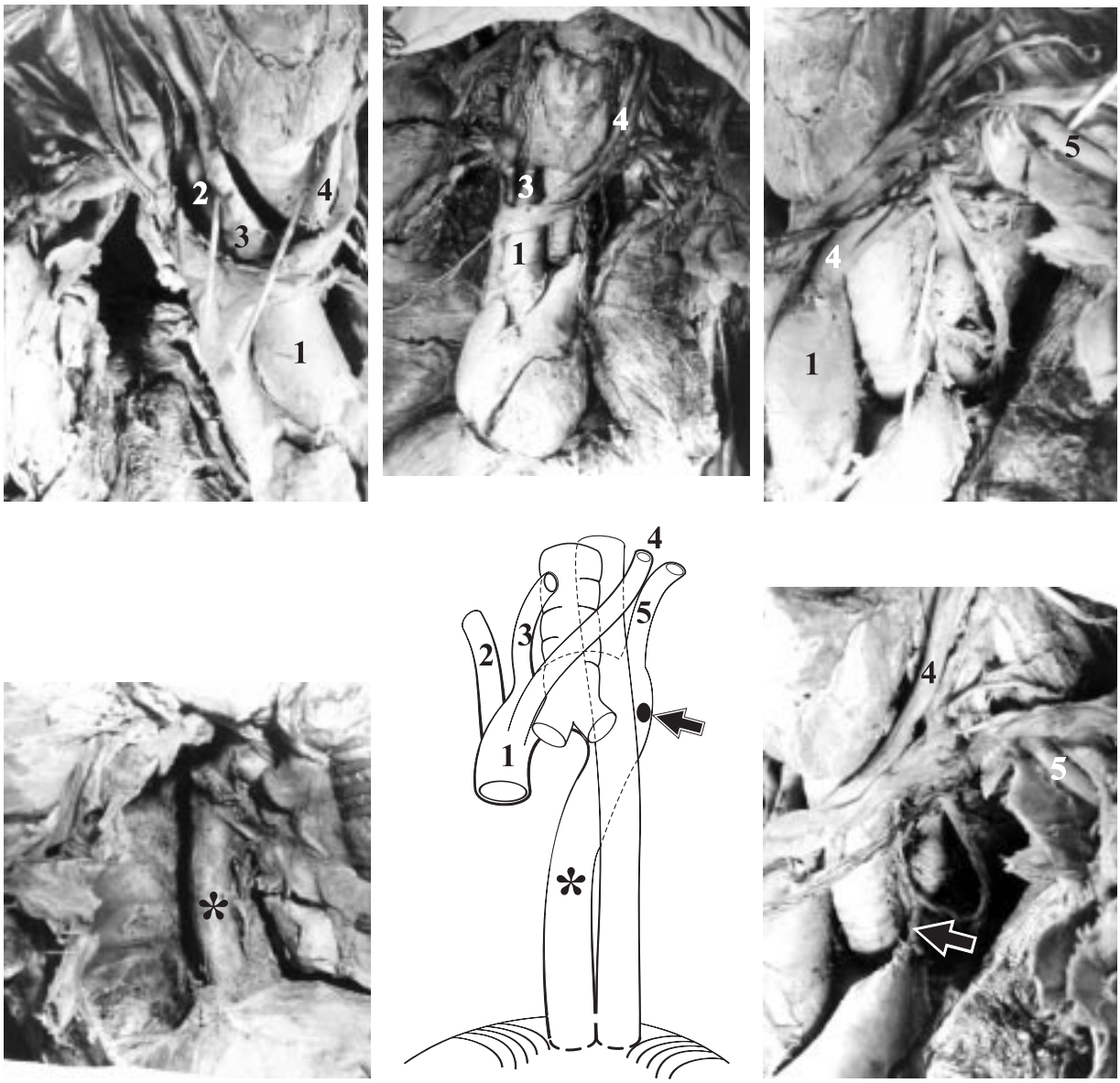

Figure 2. Case 1 — Type E from figure 1. Photos taken during preparation mentioned above, right and left respectively to the central schematic drawing. 1 to 5 as in Fig. 1. Asterisk — right descending aorta. Arrow — arterious ligament.

years of the Department of Anatomy. They were preserved as isolated preparations containing only the aortic arch with its branches as well as the trachea and esophagus.

CASE 2

The aortic arch ran beyond the esophagus, from the left to the right, where the following arteries branched off: the left subclavian artery, the left common carotid artery (both near each other), and next on the right: the right subclavian artery and right common carotid artery, also near each other (Fig. 3A).

CASE 3 AND 4

The arches of the aorta ran properly along the left side; the right aberrant subclavian arteries extended off as the last fourth branch of the arch and ran beyond the esophagus from the left onto the right (Fig. 3B, C).

DISCUSSION

According to the arrangement prepared by Hastreiter et al [8], a right-sided aortic arch in adults occurs in about 0.1%. Baron [4] maintains that distinctively in case of a small retro-esophageal bulge, it can be overlooked completely during the evaluation of a typical chest X-ray, as it happened in our case.

In a Polish anatomical literature various types of aortic arch branches have been presented by Aleksandrowicz [2], Karcz and co-workers [10], Lize [16] and Niżankowski [19] but no case of the right-sided aortic arch was found.

In a Polish clinical literature, only a few cases of a double aortic arch or of right-sided course have been described [20,21]. As the authors underline, the early diagnosis and treatment is very important. Sometimes, after entering a particular age, the symptoms resulting from constriction of the respiratory tract and the esophagus that are caused by abnormally coursing vessels can decrease or vanish completely.

As it has already been mentioned in the introduction, in the time of individual development closure and atrophy occur unilaterally as one of three sections of the previously symmetrical vascular systems take part in forming of the aortic arch and its branches. In the etiopathogenesis of developmental lesions of both the large vessels and the heart, other than infective or teratogenic factors, the role of the cranial sections of the neural crest is underlined. It is a source of so-called ectomesenchymal cells that take part in creating the vascular system [13]. It is important to underline that the shaping of the aortic arch occurs earlier that the final division of the outflow from the heart, therefore, developmental malformations of the aorta and heart can, but do not have to, occur simultaneously.

If, with one-sided atrophy of one out of three mentioned sections, constriction or atrophy of any given section of the contralateral side occurs, then we have constriction or a congenital absence of the aortic arch [22]. In this lesion, three different types can also be differentiated, connected developmen-

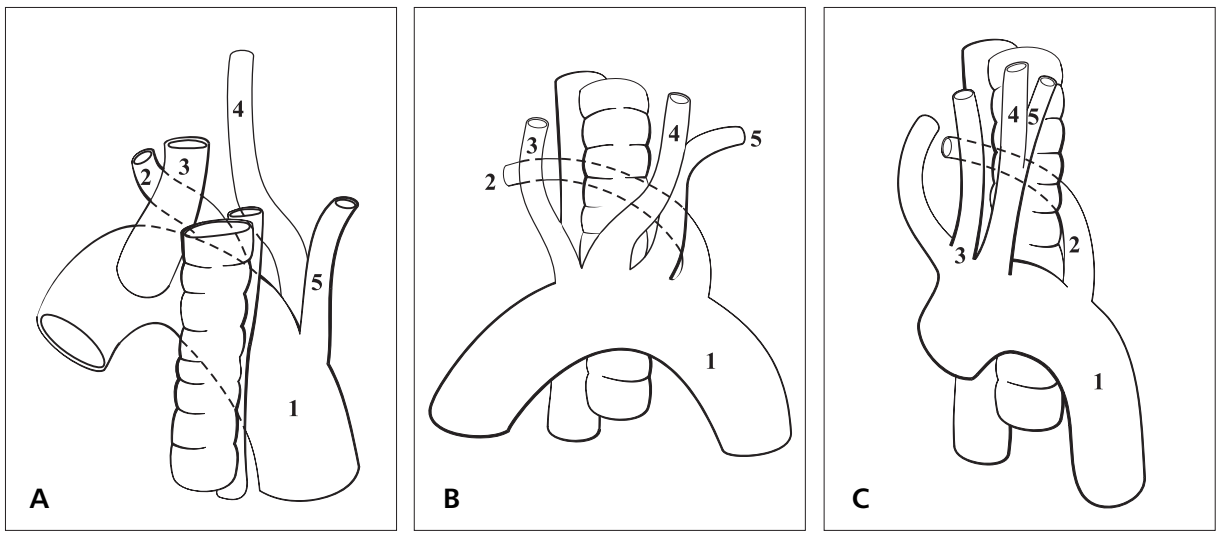

Figure 3. Drawings from preparations of cases 2, 3 and 4. A) Retroesophageal course of aortic arch — Type C from figure 1; B) and C) Right aberrant subclavian artery — Type B from Fig. 1. 1 to 5 as in Fig. 1.

tally with the closure of one out of three previously described parts of arteries of the branchial arches and dorsal aortas.

In literature, two types of right-sided aortic arches are generally distinguished. The first type, the "anterior" one, is identical to our developmental type D, very often associated with cyanotic congenital heart disease; and the second type, the „posterior", with a retro-esophageal bulging part of the arch and a left-sided subclavian artery running off as the last branch of the aortic arch [3,5,7,8,11,14,15]. Type two correlates to our case study number 1 and type E.

Case number 2 requires a separate discussion. Niżankowski [19] describes a case of a right-sided brachiocephalic trunk leaving the aortic arch as the last branch and running behind the esophagus (type C according to our description) at which he notes that he did not find a similar case in literature, including Adachi's monograph [1]. Hastreiter et al [8] describe the possibility of a retro-esophageal course of the aortic arch and the occurrence of the descending aorta on the contralateral side to the arch in both right and left -sided courses. In our case number 2, we deal with a left to right course. It seems that developmentally it belongs to type C, such as Niżankowski's case [19], however, the descending aorta has been pulled by the vessels branching entirely from this initial section onto the opposite side of the arch.

It is important to observe the possibility that the aortic arch can stem from the artery of the third branchial arch with the atrophy of the fourth arch exists. This anomaly, the cervical aortic arch, occurs extremely seldom and most often on the right side [9]. The only symptom that seems to be visible and palpable is the pulsation in the supraclavicular area. One of the authors of this research personally observed such a case in a patient treated for an external ear disease. For an ethical reason, lack of any clinical symptoms, the patient was not proposed any vascular examinations with contrast; the typical chest examination did not reveal any abnormalities.

Of much significance is the situation of the ductus arteriosus or the ligamentum arteriosum [17,20,21]. Symptoms associated with the vascular rings that constrict the trachea and esophagus occurs most often in small children and in general it requires urgent surgical intervention. If symptoms are not considerable, the developmental lesion can go unnoticed to a late age. Nonetheless, an abnormal vascular course can be the reason of insufficient cerebral blood flow and can lead to encephalopathy [6]. It may be that the neurological symptoms diagnosed in our case number 1 were due to the disturbances in blood flow to the brain.

In the description of case number 1 it is mentioned that a ring of great vessels closed by arterious ligament surround the esophagus and trachea. Such a ring occurs also in cases of double aortic arches, in an abnormal course of the pulmonary arteries behind the trachea and esophagus, or even in the presence of a single aberrant subclavian artery.

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