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A left circumflex aorta with a displaced thoracic duct in a 94-year-old male cadaver: a case report with discussion on embryology

P. Ostrowski et al., Left circumflex aorta

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ABSTRACT

A left circumflex aorta (LCA) is an extremely rare variation of the thoracic aorta. It is distinguished by a retroesophageal descending aorta that subsequently travels down the right side of the thoracic vertebrae towards the aortic hiatus. Nonetheless, its embryological origin ought not to be overly generalized, but each case should be considered individually due to its unique vascular patterns.

This study presents a description of a LCA in a 94-year-old male cadaver. The dissection revealed the descending aorta posteriorly from the trachea and esophagus and then laterally on the right from the thoracic vertebral bodies. The branching pattern of the aortic arch was typical, so was the course of the left and right recurrent laryngeal nerves. However, the thoracic duct was placed on the right, and drained into the right internal carotid vein. Due to the normal appearance of the ascending part and the arch of the aorta, it is safe to presume that the variation originated from the persistent right dorsal aorta, with the retroesophageal part from the persistent left dorsal aorta.
Detailed understanding of the variations of the thoracic aorta, and the anomalies associated with the LCA, can help to improve management of these conditions, and with that, improve patients’ overall outcomes. Patients with a LCA, or another vascular ring, can either be asymptomatic or present with esophageal and/or tracheal compression symptoms. Management of this anomaly consists namely of ligation of the patent ductus arteriosus / ligamentum arteriosum and aortic uncrossing.

**Key words:** left circumflex aorta, thoracic duct, anatomy, embryology, anatomical variation

**INTRODUCTION**

A left circumflex aorta (LCA) is an uncommon congenital variation of the cardiovascular anatomy. A circumflex aorta is distinguished by a retroesophageal thoracic aorta that crosses contralaterally to the ascending aorta. The circumflex retroesophageal variation of the aorta was first described by D’Cruz et al. in 1966 [6]. However, a case of a left aortic arch and a right descending aorta was presented by Paul et al. in 1948 [16], matching the description of the left circumflex aorta presented in this case study.

Anomalies of the aortic arch are quite rare in the general population, with their prevalence ranging from 1%-2% [4]. A right circumflex aorta occurs less frequently than a double aortic arch, or right aortic arch with an aberrant left subclavian artery with a Kommerell's diverticulum. A LCA is even less common than the aforementioned anomalies [7].

Anomalies of the aortic arch are often associated with the formation of a vascular ring, with the circumflex aorta being an extreme example of the latter [9]. The circumflex aorta is divided into two groups; a right circumflex aorta, and a left circumflex aorta. The right circumflex aorta can occur with, or without an aberrant left subclavian artery. On the other hand, the left circumflex aorta can appear with, or without an aberrant right subclavian artery. In many cases, a patent ductus arteriosus / ligamentum arteriosum together with a left, or right, circumflex aorta develops a true vascular ring [9].
Unfortunately, the embryological origin of the circumflex aorta is oftentimes confused and has a tangled description in the available literature. The main reason behind this fact is the uniqueness of the found thoracic aorta vascular patterns that ought not to be generalized for the whole population. Instead, it is prudent for the medical professionals to carefully examine each case individually and hence attempt to delineate the possible embryological mechanisms involved.

Many other anomalies of the cardiovascular system can occur in individuals with a LCA. These consist namely of formation of a vascular ring and an aberrant right subclavian artery, amongst others [9, 25]. Furthermore, the prevalence of additional cardiac anomalies in patients with a circumflex aorta is approximately 50% to 60% [21].

Knowledge regarding the different anomalies associated with the circumflex aorta might help increase the overall efficiency of their management, and therefore, aid in attaining better outcomes for the patients.

The following study aimed to present a thorough description of a left circumflex aorta case, its branches and relation to the surrounding anatomical entities. Moreover, the embryology of the circumflex aorta, its symptoms, management, and associated anomalies were comprehensively discussed.

CASE REPORT

During a routine dissection of a 94-year-old male formalin fixed cadaver, anomalies of the thoracic aorta were observed. No pathology that could distort the thoracic cavity was neither noticed upon the dissection, nor was it found in the available medical record of the donor. Two prosectors SP, PO performed the dissection under close supervision by MPZ and JAW.

The cadaver’s thorax was opened with the help of an oscillating saw. The cuts were made at the level of the sternal angle, underneath the clavicles and anteriorly to the long thoracic nerve. Having opened the rib cage and released the connective tissue adhesions, the heart was identified as shifted towards the left lung, compressing it. The left lung was located in the vertebral column segments Th1 to Th8. The segments 3, 4, and 5 of the said lung were enlarged and positioned behind the heart, reaching to the right midclavicular
The right lung was significantly larger than the left lung. The bronchopulmonary segments 3 and 5 of the right lung were partly positioned on the surface of the heart, reaching the left parasternal line.

Both lungs were excised and revealed the presence of the LCA in the posterior inferior mediastinum. Its branches and surrounding structures were carefully dissected, and documented photographically. The external diameters of the aorta and its branches were measured by using a digital caliper (Lux tools, China), and presented as an average of three independent measurements. Figs. 1-4 present the various viewpoints of the variant, whereas fig. 5 is a schematic depiction of the anomaly in relation to the nearby neurovascular structures.

The aorta drained the left ventricle in the usual manner. The ascending aorta was oval in shape, instead of round, hence two dimensions were measured, these were 43.2 mm and 19.8 mm respectively. The aorta ascended from the base of the left ventricle at approximately the Th3 vertebral body, passing to the left of the Th2 vertebral body (where its apex was found), and then turning sharply to the right. The descending aorta began at the Th2-Th3 intervertebral disc level, coursed posteriorly to the trachea and esophagus, and descended on the right side of the thoracic vertebral bodies, laterally from the esophagus. The branches of the aortic arch from both its convex and concave side arose in the classical manner. Going namely proximal to distal: the brachiocephalic trunk was the first branch originating from the aortic arch, giving rise to the right common carotid and right subclavian arteries. The external diameter of the brachiocephalic trunk at its origin was measured at 21.6 mm, and at its branching point 21.6 mm. The next branches of the aortic arch were the left common carotid, and left subclavian arteries. They had an external diameter of 11.6 mm and 15.3 mm respectively.

The ligamentum arteriosum was a thick band of connective tissue connecting the aortic arch with the superior part of the left pulmonary artery. The left recurrent laryngeal nerve passed inferiorly to the ligamentum arteriosum and the aortic arch, however, the right recurrent laryngeal nerve passed underneath the brachiocephalic trunk (instead of the right subclavian artery as classically described). The bronchial arteries were also observed, with the left one originating from the arch of aorta, and the right one originating from the descending aorta.
After giving off the left subclavian artery, the aorta passed the trachea and esophagus posteriorly, and descended in the thoracic cavity leaving a significant impression on the right lung. The aorta was dilated at the point where it crossed the esophagus, with an external diameter of 34.3 mm. At the approximate Th3-Th4 intervertebral disc level (where it was placed laterally on the right of the vertebral bodies), the descending aorta had an external diameter of 29.6 mm. The external diameter remained unchanged as it passed through the aortic hiatus in the diaphragm, which was in close proximity to the esophageal hiatus. All the measurements concerning the external diameter of the aorta and its major branches are presented in Table I. The descending aorta had a winding course in the thoracic cavity, lying anteriorly to the right scoliotic vertebral column.

The azygos vein ascended on the right side of the thoracic cavity, coursing between the vertebral column and the descending aorta. It looped around the aorta, and drained directly into the superior vena cava. The remaining venous tributaries of the thorax were anatomically normal.

The thoracic duct was displaced by the descending aorta to the right side of the vertebral column, and drained into the right internal jugular vein. No vessel draining into the left venous angle was visualized by macroscopic dissection. Moreover, the bronchopulmonary lymph nodes were enlarged and calcified.

Macroscopic evaluation of the heart revealed no visible structural abnormalities except for a small patent foramen ovale that was on average 2.1 mm in diameter. The origin of both right and left coronary arteries was typical, as well as their course and main branching pattern. The patient did not present any other visible vascular abnormalities neither within the thorax, nor the abdomen. Unfortunately, due to the unavailability of the donor’s medical records no conclusions could have been made regarding the presence or absence of any clinical symptoms in vivo, associated with the present variant.

**Ethical concern**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research
committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The material used for this research was obtained from a body donation program.

**DISCUSSION**

The above-mentioned case presents a description of a left circumflex aorta with typical aortic arch branching pattern. The thoracic duct found in the described specimen was displaced to the right side, draining into the right internal jugular vein. To the best knowledge of the authors, this is the first case of a right sided thoracic duct accompanying a LCA in the available literature.

There are numerous descriptions regarding the embryology of the systemic arterial system in the available literature. Following Sadler’s description [18], the ascending aorta arises as the distal (cranial) part of the truncus arteriosus. It gives rise to two ventral aorta, from which the pharyngeal arch arteries (PAA) emerge. As the development process continues, they are brought closer together to form the aortic arch and its major branches, as well as the pulmonary trunk. The arteries in question are related to the 3rd, 4th and 6th pharyngeal arches. Going right to left, the brachiocephalic trunk arises from the right horn of the aortic sac, whereas the proximal aspect of the aortic arch from the left horn. Next, the right common carotid artery has its origin in the third right PAA and the right subclavian artery in its proximal portion from the fourth right PAA (its distal segment comes from the right seventh intersegmental artery). The left third PAA forms the left common carotid, whereas the left fourth PAA is responsible for the creation of the aortic arch between the left common carotid and the left subclavian arteries. Next, the left subclavian artery sprouts from the left seventh intersegmental artery. Lastly, the sixth PAAs give rise to the right and left pulmonary arteries respectively, as well as the ductus arteriosus on the left. The remaining part of the thoracic aorta originates from the fusion of both right and left dorsal aorta [18].

If one of the PAAs fails to regress, a vascular ring encircling the trachea and esophagus may develop [2]. A vascular ring is a rare congenital cardiovascular anomaly, which can be formed as a consequence of a variety of embryological defects, such as
a double aortic arch, a right aortic arch with left ductus arteriosus, a retroesophageal aberrant left subclavian artery, and others [26]. Vascular rings represent approximately 1% of cardiovascular congenital anomalies [13].

Notwithstanding, in the case described herein the ascending aorta and the aortic arch with its major branches were formed in the typically described manner. Henceforth, it is the authors’ understanding that no abnormality in the failed regression of the PAAs could have been involved. The only variation found was the abnormal course of the descending aorta, crossing both the trachea and esophagus posteriorly, to then descend laterally on the right from the thoracic vertebrae. Therefore, bearing in mind the aforementioned appropriate anatomy of the remaining parts of the thoracic aorta, it is safe to presume that the variation encountered in this case originated from the persistent right dorsal aorta below the right seventh intersegmental artery. The retroesophageal portion of the descending aorta was most probably a remnant of the left dorsal aorta that has involuted in the lower thoracic segments, leaving room for the dominance of the right dorsal aorta. It has also occurred to the authors that the displacement of the descending aorta could have resulted from the scoliosis that the patient also presented. Severe scoliosis can result in major arterial dislocation due to the disruption in the normal anatomy and proportions of the thorax, however it would not explain the completely retroesophageal course of the aorta. Henceforth, the authors believe that the variation found was inborn.

Circumflex aortas can be associated with various abnormalities, a prime example being the aberrant subclavian artery. Depending on its branching off point from the aorta, its embryological origin might differ between the patients. The aberrant right subclavian artery, when not accompanied by the circumflex aortic variation, is one of the most common congenital aortic arch anomalies, with a prevalence ranging from 0.16% to 4.4% in the general population [15]. Patients with an aberrant right subclavian artery, when it has a retroesophageal course, can give symptoms of esophageal and/or tracheal compression, and thus will need to be surgically managed [1]. Notwithstanding, the patient in the current paper did not present this variation, but instead the right subclavian artery originated from the brachiocephalic trunk in the typical manner.

Another example may be the Kommerell's diverticulum, or Kommerell's aneurysm, that is a persistent remnant of the fourth primitive dorsal arch due to its failed regression. It
can occur in both left and right sided aortic arches. In a case of the left aortic arch with this anomaly, the right dorsal aorta involutes proximal to the right subclavian artery, and leaves the subclavian artery attached to the left descending aorta via the distal portion of the right dorsal aorta. Individuals with aberrant right or left subclavian arteries are in 20%-60% of the cases associated with Kommerell's diverticulum [22]. Management of this condition can be obtained by either open or endovascular procedures. Some of the open procedures are diverticulum resections, interposition, patch repairs, and more. The endovascular procedure for the management of Kommerell's diverticulum is thoracic endovascular aortic repair using stent grafting [24]. However, in the present case, the LCA had a typical branching pattern without a Kommerell's aneurysm.

Various findings in individual cases make it troublesome for the clinical practitioners to assign a single, unified embryological defect responsible for the creation of the LCA. The description of the LCA's origin provided by Hanneman et al. [8] and Haranal et al. [9] with absent variant right subclavian artery resulting from the involution of the right aortic arch in its part between the right subclavian and the ductus arteriosus most certainly does not pertain to the current case. For that variation to occur, the typical left aortic arch would not be present, contrary to the present findings. The authors would like to acknowledge Sánchez Torres and Roldán Conesa [19], as their case seems to resemble a very similar clinical picture to the one presented in this study, hence likewise we would like to propose and reiterate that the embryological origin of the encountered here variation is the regression of the left dorsal aorta. Henceforth, the authors would like to urge caution whilst overly generalizing the embryological origin of the encountered diversity of the thoracic aorta and rather use an individual approach to a single case to discern its true meaning.

A circumflex thoracic aorta can impede the respiratory pathways, resulting in severe respiratory symptoms and recurrent pulmonary infection [11]. It can also give esophageal compression symptoms with dysphagia being the most common one [12]. The development of a complete vascular ring from a LCA undoubtedly depends on the position of the ductus or ligamentum arteriosum. If the ductus or ligamentum arteriosum is present on the right side, connecting the right sided descending aorta with the right pulmonary artery, a complete vascular ring develops [21]. However, if the ductus or ligamentum arteriosum is located on the left side, the vascular ring is incomplete [9], as found in the
present case. Even though a LCA with a left patent ductus arteriosus does not form a complete vascular ring, it can still cause severe respiratory compression, as shown by Sheth et al. [20] in a case of a 2-month-old patient whose left main bronchus was obstructed. In patients aged two years old or less, the symptoms of a vascular ring are typically respiratory in nature, whereas older children and adults suffer mostly from dysphagia, less frequently the former [25].

The external diameters of the aorta and its branches described herein were higher than average. An example of this is the external diameter of the brachiocephalic trunk, which has an average diameter of 8.3-15.5 mm in females and in males 9.1-14.5 mm [14]. However, the brachiocephalic trunk in the specimen presented in the current study, had an external diameter measured at 21.6 mm at its origin, and 21.6 mm at its branching point. The aortic arch had a dilation at the point where it passed the esophagus, with an external diameter of 34.3 mm. Ectasia at this point could have given symptoms of esophageal compression [23].

Indisputably, symptomatic patients require fast and adequate management of the LCA, so as to prevent the most serious complications that could potentially include death. Pampin et al. [3] described a case of a 26-days-old neonate in whom the LCA was present (with obliterated left ductus arteriosus) who suddenly passed away overnight. The said case contrasts greatly with the current 94-year-old patient, pointing out to the vastly divergent clinical picture of the discussed variation.

There is constant need for advancements in the management of the circumflex aorta due to the fact that a simple ligation of the ligamentum / ductus arteriosus may not necessarily relieve the respiratory or swallowing compressive symptoms associated with it [9]. The first available description of the aortic uncrossing procedure comes from 1984 provided by Planche and LaCoeur-Gayet [17]. The said authors operated on patients who presented with postoperative respiratory and / or swallowing obstructive syndromes following the previously undertaken ligamentum / ductus arteriosus ligations [17]. The aortic uncrossing is a major procedure during which the retroesophageal aorta is transected, mobilized, and reconnected to the ascending aorta on the same side of the airway as the descending aorta, relieving the tracheal and / or esophageal external constriction. Nonetheless, the surgery does not resolve the problem of cartilage
malformation and tracheal stenosis, nor the tracheobronchomalacia that could have resulted from the previous compression [11], hence those issues ought to be addressed by different techniques.

Lastly, it is crucial to bear in mind possible variations of the thoracic duct, which as in the case reported herein can drain into the right sided vessels instead of the typically acknowledged left venous angle. In a report by Cerfolio et al. [5], chylothorax was a complication of 0.42% of general thoracic procedures (47 cases out of 11,315). An unanticipated high level of liquid drainage from the thoracic cavity (not necessarily milky in nature as chylomicrons and triglycerides might be of lower concentrations due to patients not receiving normal diets) might be indicative of thoracic duct’s iatrogenic injury [5]. Henceforth, its anatomy and closeness to the major vessels ought to be always taken into account during a thoracic surgery in furtherance of avoiding complications.

CONCLUSIONS

The above-mentioned study presents a case of a LCA, a rare anatomical variant of the cardiovascular system. A circumflex retroesophageal arch is distinguished by an aortic arch which crosses the midline posteriorly to the esophagus, approximately at the level of the carina, and descends on the contralateral side of the spine [9]. Special caution ought to be in place when examining the available literature for the embryological origin of the LCA, as presence of additional variants and thorough description of the course of the aorta can oftentimes shed light onto a completely different mechanism than previously described. The LCA could be associated with many anomalies, especially those of the cardiovascular system. Henceforth, an extensive knowledge regarding the different variations of the LCA, and the anomalies associated with it, is surely of great importance upon encountering this rare variant to provide the patient with adequate care.

Acknowledgements

The authors are beholden to Mr Jacenty Urbaniak for the technical support and graphical depiction of this variation. “The authors wish to sincerely thank those who donated their bodies to science so that anatomical research could be performed. Results
from such research can potentially improve patient care and increase mankind's overall knowledge. Therefore, these donors and their families deserve our highest gratitude” [10].

**Conflict of interest:** None declared

**REFERENCES**


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<tr>
<th>The measurement point</th>
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<td>Ascending aorta – at origin (oval in shape)</td>
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<td>Aorta – at the junction with esophagus</td>
<td>34.3</td>
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<td>Descending aorta – at Th3/Th4 intervertebral disc</td>
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<td>Brachiocephalic trunk – at origin</td>
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<tr>
<td>Brachiocephalic trunk – at the branching point</td>
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<td>Left common carotid artery – at origin</td>
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<td>Left subclavian artery – at origin</td>
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Table I. The external diameter of the aorta and its major branches.
**Figure 1.** A right-sided view on the posterior mediastinum; 1 — descending aorta; 2 — oesophagus; 3 — azygos vein; 4 — thoracic duct; 5 — right internal jugular vein; 6 — right subclavian vein; 7 — superior vena cava; 8 — left brachiocephalic vein; 9 — right brachiocephalic vein; 10 — ascending aorta; 11 — arch of aorta; 12 — brachiocephalic trunk; 13 — right common carotid artery; 14 — right subclavian artery; 15 — right internal thoracic artery; 16 — right phrenic nerve and pericardiacophrenic vessels; 17 — right vagus nerve; 18 — diaphragm.

**Figure 2.** A right-sided view on the posterior mediastinum with the heart removed; 1 — descending aorta; 2 — oesophagus; 3 — right superior intercostal vein; 4 — azygos vein; 5 — thoracic duct; 6 — right internal jugular vein; 7 — right subclavian vein; 8 — brachiocephalic trunk; 9 — ascending aorta; 10 — right main bronchus.

**Figure 3.** An enlarged view on the left recurrent laryngeal nerve; 1 — arch of aorta; 2 — left vagus nerve; 3 — left recurrent laryngeal nerve; 4 — left phrenic nerve; 5 — left pericardiacophrenic vessels.

**Figure 4.** A right enlarged view on the posterior mediastinum; 1 — ascending aorta; 2 — brachiocephalic trunk; 3 — right common carotid artery; 4 — right subclavian artery; 5 — right vagus nerve; 6 — right recurrent laryngeal nerve; 7 — right internal thoracic artery.

**Figure 5.** A sketch of the LCA and its surrounding structures.