Vascular Rings and Aberrant Brachiocephalic Trunk

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Key Points

1) Vascular rings can develop in many different individual patterns because they result from abnormal persistence of embryologic structures from the multiple parts of the right and left aorta dorsal and ventral segments, as well as the interconnecting pharyngeal arches and arteries.
2) Not all vascular rings are symptomatic, but when symptoms are present, they are due to compression of the trachea or the esophagus by the abnormal structures, resulting in dyspnea or dysphagia.
3) In general, the best operative approach for any vascular ring is a lateral thoracotomy performed on the side of the patient that has a descending aorta. Different approach is possible.
4) Advanced imaging techniques result in more vascular rings being diagnosed, often during the prenatal period, and with excellent localization of anatomy, leading to discussion for treatment without symptoms.

1) Introduction

At first sight vascular anomalies causing compression of neighboring structures do not appear to be simply categorized because of their diversity.

The development of the aortic arch and its branches from two dorsal and two ventral parts and their interconnecting pharyngeal arches/arteries is substantiated by embryology leading to numerous possibilities of ring development, but they have

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only isolated clinical relevance (Fig. 1). A definite simplified classification serving >95% of the clinically described variations is as follows:

A) Double aortic arch (Fig. 2)
B) Right-sided aortic arch/left-sided arterial ligament
C) Aberrant origin of the right-sided brachiocephalic trunk causing tracheal stenosis.

The common feature between these anomalies is the compression of the neighboring structures such as the trachea and esophagus causing shortness of breath and sometimes difficulties in swallowing. In infancy with predominantly liquid food as mother's milk/formula and a rather soft trachea, the most common symptoms are respiratory. With increasing age, difficulties in swallowing/dysphagia increase with the introduction of solid food causing more pronounced dysphagia. However,

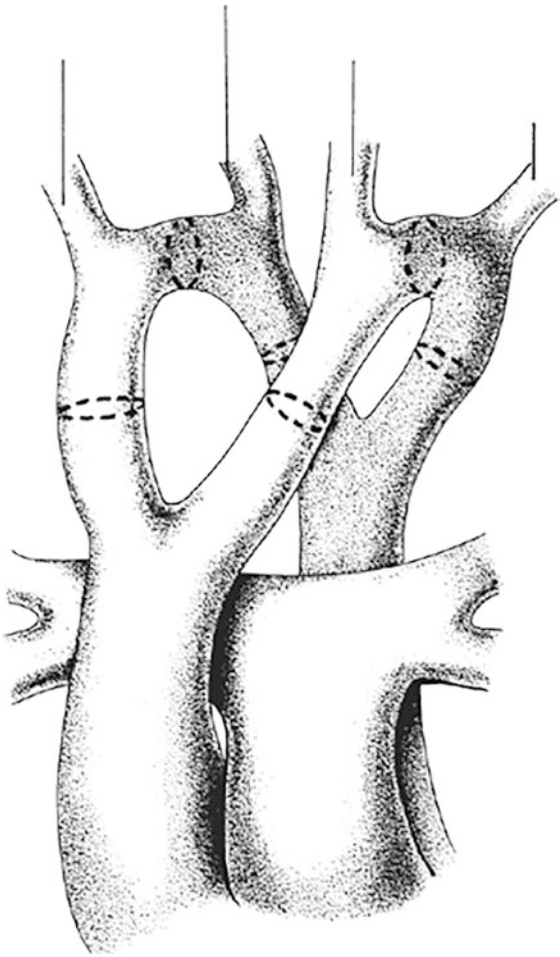

Fig. 1 Primitive aortic arch, the dashed lines indicate all possible interruptions, which explain all possible forms of normal and abnormal aortic arch and vascular rings

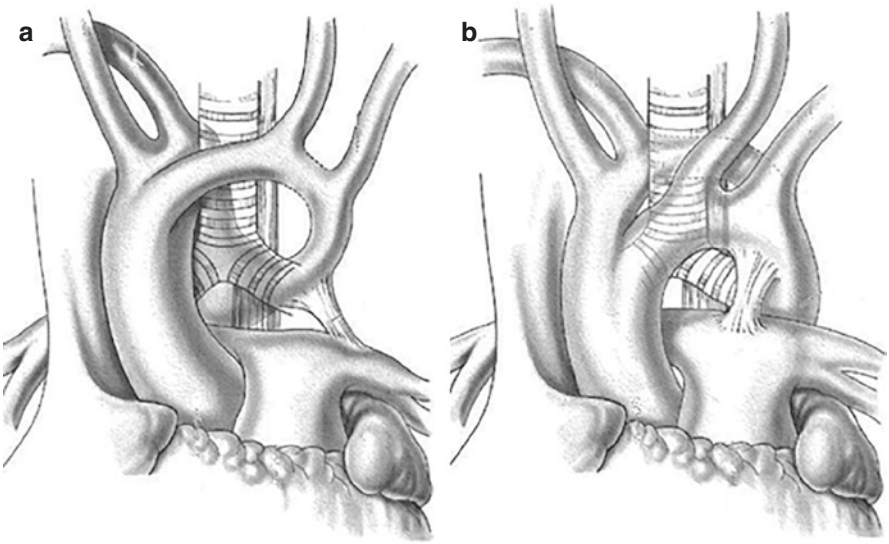

Fig. 2 Double aortic arch. (**a**) The most common form of double aortic arch: right-sided aortic arch with a hypoplastic left arch. The hypoplastic arch can become extremely hypoplastic or even atretic either between the supra-aortic vessels or distal to them (**b**). The rare form of a dominant left-sided aortic arch with a hypoplastic retroesophageal segment of the right-sided arch with the descending aorta on the left

patients who never experienced normal swallowing before, may not be able to report their objective dysphagia.

Modern imaging tools, such as computed tomography (CT)/magnetic resonance imaging (MRI), improve the accuracy of localizing defects. The timely and up-to-date implementation of surgery brings about immediate regression and abolishment of symptoms in most patients with excellent long-term results.

The variations in the course of the thoracic aorta do not necessarily cause symptoms. Contrary to other congenital heart diseases, anomalies of intrathoracic great vessels are only operated upon when clinical symptoms are present. However, there are two exceptions:

A. An asymptomatic vascular anomaly associated with intracardiac defects requiring surgery
B. An asymptomatic complete double aortic arch as it will become symptomatic with age and growth, but surgery is much easier in neonates and infants.

A new clinical challenge is the increasing number of asymptomatic vascular rings being diagnosed during fetal life.

The radical resection of the encircling vascular and fibrotic structures is recommended. The importance of the Kommerell diverticulum as an independent cause for recurrent symptoms has been recognized over the years and addressed now for primary surgery.

Video-assisted thoracoscopic repair as well as totally endoscopic robotic-assisted approach is reported. These approaches, however, do not meet the abovementioned principles of radical resection.

Symptomatic pulmonary artery sling is not part of this chapter, as it most often requires additional tracheal surgery, which nowadays is performed via sternotomy, employing extracorporeal circulation, and therefore done in pediatric cardiac surgery units.

2) Aim of Surgery for Vascular Rings

In our view, with the exception of an aberrant brachiocephalic trunk, for all compressing thoracic vascular anomalies, the approach, a lateral thoracotomy, should always be performed on the side of the descending aorta. The argument in favor of such an approach is to obtain safe intraoperative control of the structures to be cut, especially of the descending aorta. A complete resection starts at the descending aorta and extends beyond the esophagus on the other side if not limited by specific anatomic structures/details we are not aware of before operation.

The resection of a patent vascular segment can be carried out by side bite clamping or under certain circumstances in cross-clamping of the descending aorta above and below the structure to be resected. To perform a continuous running suture to close the resected structure, flush to the aortic wall should take only a few minutes, and for that there is no need for further protective measures like including a left heart assist.

It must be mentioned that most surgeons perform a division of any vascular ring irrespective of the side of the descending aorta via a left-sided posterior thoracotomy, which occasionally leads to safety issues when following the principles of radical resection. An advantage may be the option of re-implantation of an aberrant subclavian artery on the side of thoracotomy.

The immediate intraoperative sign of effectively having taken care of the symptomatic vascular ring is the significant retraction of the divided ring structures.

Vascular rings are associated only rarely with a primary structural tracheal stenosis caused by hypoplasia in contrast to a pulmonary (artery) sling. A certain degree of tracheomalacia can be the reason for a residual stridor in infants in the first postoperative month and in most cases does not require additional surgery.

3) Double Aortic Arch/Complete Vascular Ring

For all types of vascular rings, the following anatomic and hemodynamic factors have to be taken into consideration when planning for surgery:

A) Position of the descending aorta, especially in relation to the esophagus
B) Position of the dominant aortic arch
C) Possibility of transection of currently patent vascular structures.

The sidedness of the descending aorta in relation to the esophagus is important because the access to the aorta can be challenging when the esophagus has to be passed.

The proportions of size between the anterior and posterior parts of the arches vary. Double aortic arch may consist of two equally and well-developed arches. Commonly there is a dominant right aortic arch with a hypoplastic left arch.

In case of a double aortic arch, the nondominant part should be divided. If there is no side preference reported, accessibility dictates the arch to be transected.

Surgery of a balanced (co-dominant) double aortic arch follows the principles of careful retroesophageal dissection and resection as mentioned above. Prior to actual division, the strength of the right and left carotid and radial pulses should be evaluated with the vascular clamps applied. In the absence of any changes in proximal and distal blood pressure and pulse curves, even the bigger of the two aortic arches can be divided. To divide the bigger of two arches in double aortic arch may be thought of, if the bigger arch is the one with a retroesophageal course. Precise documentation is required.

In case of a dominant aortic arch with retroesophageal course, a simple transection of the anterior hypoplastic ring segment may not be sufficient to relieve all, but it may diminish some clinical symptoms.

4) Incomplete Vascular Ring

Right-sided aortic arch with left-sided Ductus Botalli/Ligamentum Arteriosum

The right-sided aortic arch is curving to the right of the trachea. There are three theoretical possible varieties, two of which may create a vascular ring. The fundamental structure is always the retroesophageal vascular structure or its remnant (arterial duct vs. arterial ligament).

The first variety, the most commonly seen patologic form (65%), is the combination of an aberrant left subclavian artery and an arterial duct located between the left pulmonary artery and aortic isthmus. A short aortic segment is addressed here indeed as the isthmus from which the left subclavian artery and the duct arise (Fig. 3).

Both structures can arise separately from the aorta or jointly from a Kommerell diverticulum. The existence of a Kommerell diverticulum is not obligatory for this kind of symptomatic vascular ring.

The second variety (33%) is the mirror image of the normal left aortic arch: Right aortic arch with a left-sided brachiocephalic trunk giving rise to the left subclavian artery which runs regularly to the left and anteriorly of the trachea. The ring is created by a retroesophageal duct respectively ligamentum arteriosum, only, coming off the right sided isthmus.

The third variety, also with a mirror image branching, does not create a vascular ring, as the ductus originates from the left subclavian artery directly. This form occurs sporadically (2%) and is associated with a tetralogy of Fallot without causing any vascular ring type symptoms.

5) Kommerell Diverticulum

The Kommerell diverticulum is named after Burckhard Kommerell, staff radiologist at Charité Hospital, Berlin, Germany, in the 1930s. It is a remnant of the dorsal right or left root of the aortic arch as a pendant to the contralateral existing complete aortic arch (Fig. 3). This structure, like when first described, is not referring to a certain kind of vascular ring but can occur, not obligatory, in a number of anomalies. The diverticulum is an aorta-sized outpouching of the descending aorta

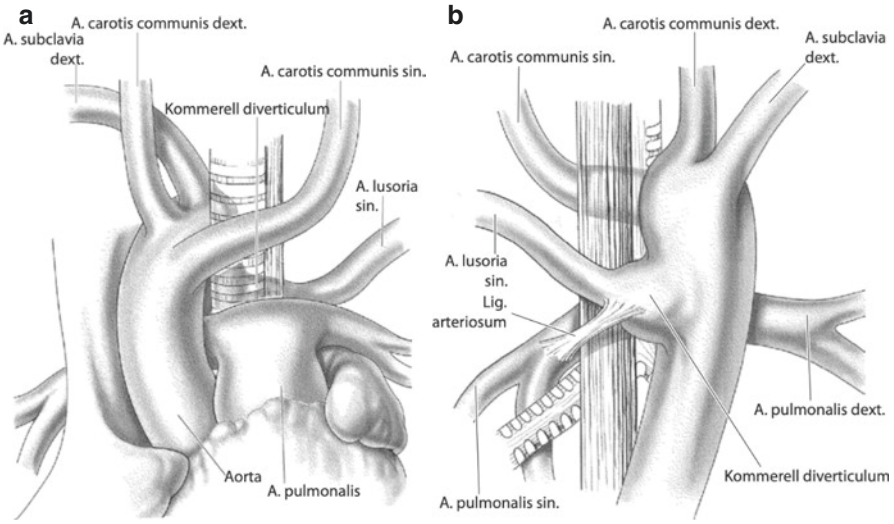

Fig. 3 Incomplete vascular ring: View of a Kommerell diverticulum with a right-sided aortic arch and aberrant left subclavian artery. View from ventral (**a**) and view from dorsal (**b**)

ending anteriorly in a nondominant/atretic aortic arch or in an arterial duct and aberrant subclavian artery (lusorian artery). It was originally described in a patient with a left-sided aortic arch and retroesophageally located right subclavian artery.

The diverticulum can cause obstruction as part of a vascular ring compression, or probably only because of its retroesophageal volume. This is confirmed by the fact that a Kommerell diverticulum is a frequent cause of a symptomatic recurrence or persistence of symptoms following surgery of division of a vascular ring without resection of the diverticulum.

6) Less Common Findings of a Vascular Ring

The diversity of these findings published mostly as a pathological case report or as an isolated clinical case is quite understandable given the complex embryologic development (Fig. 1). As these forms account for only about 1% of the clinical cases with vascular ring malformation, they cannot be focused on individually.

7) Compression Caused by an Aberrant Origin of the Brachiocephalic Trunk/Innominate Artery

This anomaly is caused by an atypical distal take-off of the innominate artery. It does not form a vascular ring. The artery leaves the arch distally posteriorly. Coming from the left, it takes a course ventral to the right around the trachea heading into a posterior direction. This at times causes a compression of the trachea. The ventral indentation of the trachea seen by bronchoscopy is pathognomonic for this anomaly; no other vascular anomaly causes an isolated indent like that (Fig. 4).

For minor stridor symptoms, the treatment is mostly conservative because with the development of the cartilaginous rings during the second and third year of life

symptoms diminish or may even disappear completely. This may be also due to the changing size relation of the structures with growth. A symptomatic, significant tracheal stenosis (70–80%) or apneic spells during infancy and early childhood should be an indication for surgical treatment.

This anomaly is mainly seen in infants and causes a typical stridor. Even episodes of short apnea are described.

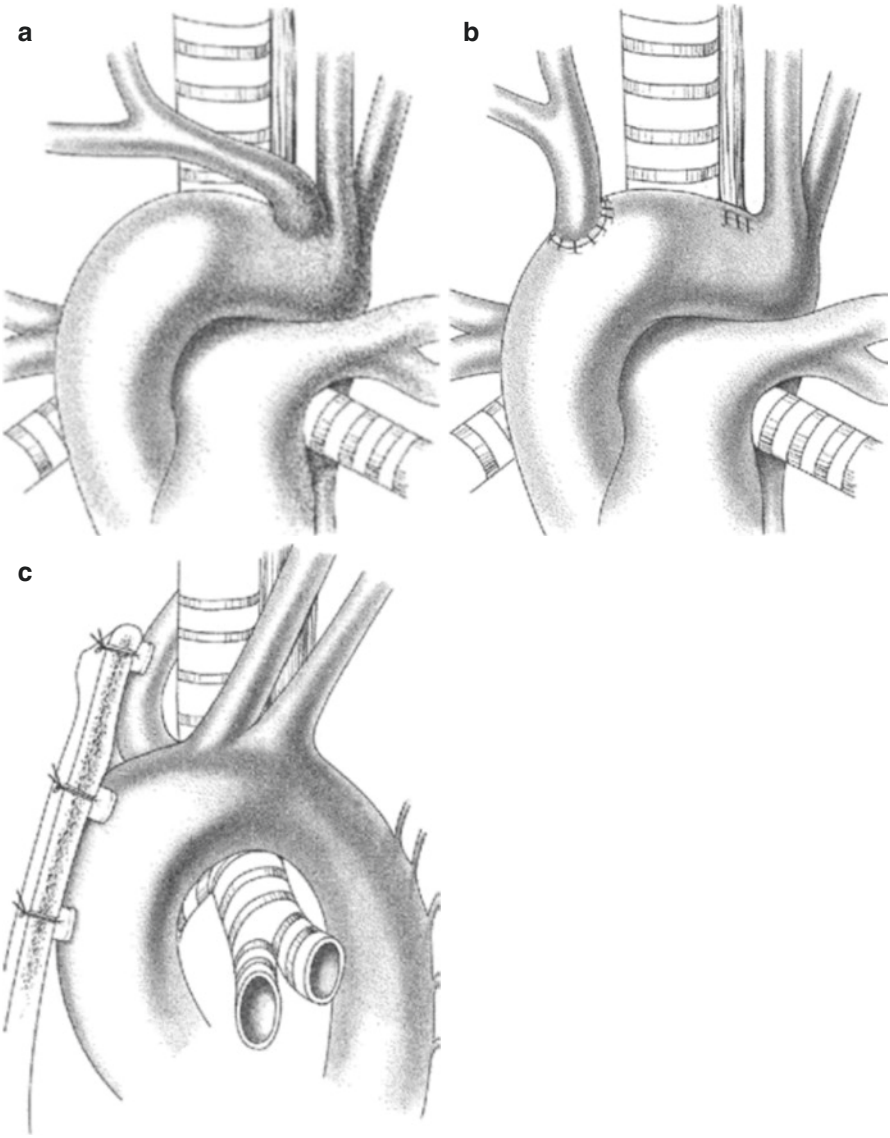

Fig. 4 Aberrant brachiocephalic trunk/innominate artery. (**a**, **b**) Anterior translocation. (**c**) Anterior aortotruncopexy

An established therapy is the classical ventral truncopexy—tacking the innominate artery to the sternum (Fig. 4c)—or, as a more complex alternative, the transsternal resection followed by a proximal re-implantation of the innominate artery into the proximal aortic arch or the distal ascending aorta (Fig. 4b), our preferred choice. We always employ extracorporeal circulation and mild hypothermia for this procedure. During the procedure, Near Infrared Spectroscopy (NIRS) is used to monitor brain perfusion.

For truncopexy, the original surgical approach described by Gross is the left thoracotomy. Other authors prefer a right anterolateral thoracotomy with a inframammary incision line. When we do a pexy rather than a translocation, we use a median partial upper sternotomy. The argument in favor of such access is the safe anchoring of the transsternal sutures.

We use three U-figured pledged sutures (3.0 or 4.0). Instead of a mere truncopexy, we perform an aortotruncopexy (Fig. 4c): we place the first suture subadventially at the distal ascending aorta, the second at the base of the innominate artery, and the third at the distal trunk just above the innominate vein. The sutures are fed transsternally through the right part of the sternum and tightened fast after the removal of the retractor. With that the pulse oximeter measurement applied at the right arm should be unchanged, and the bronchoscopy, which should be performed obligatorily, should show an expansion of the tracheal lumen, ideally with disappearance of the pulsations. Late re-sternotomies for any reason may find the aortotruncopexy loosened without recurrence of tracheal symptoms.

Additional Notes

Further notes on the Kommerell diverticulum: An increasing number of case reports describe an aneurysmatic dilatation of an originally asymptomatic diverticulum occasionally leading to aortic dissection later in life. For this reason, also, it is highly recommended to resect a diverticulum during primary surgery of any vascular ring if present. Endovascular procedures may be able to exclude a retroesophageal aneurysm from the circulation, but they may not relieve the gastrointestinal symptoms if present before.

Study Questions

1) What is a common stage in life for a child with a vascular ring to begin experiencing symptoms of dysphagia?

   a. Shortly after birth
   b. With transition from liquid to solid foods

c. During the growth spurt of adolescence
d. With scarring of structure following surgical correction

Answer to Question 1: (b.) With transition from liquid to solid foods. Clinically significant vascular rings result in compression of neighboring structures such as the trachea and esophagus. This causes the typical symptoms of shortness of breath and dysphagia. During infancy, when babies consume liquid food and the trachea is relatively soft, the most common symptom is shortness of breath. With increasing age, the introduction of solid food can result in more pronounced dysphagia. Growth of the trachea with stronger cartilaginous rings and greater diameter can improve respiratory symptoms.

2) Which lesion can cause obstructive symptoms after surgical correction of a vascular ring?

a. Duplicate aortic arch
b. Aneurysmal dilatation of a Kommerell diverticulum
c. Ligamentum arteriosum
d. Aberrant brachiocephalic trunk/innominate artery

Answer to Question 2: (c.) Aneurysmal dilatation of a Kommerell diverticulum. It is an aorta-sized outpouching of the descending aorta ending anteriorly in a non-dominant/atretic aortic arch or in an arterial duct and aberrant subclavian artery (lusorian artery). The Kommerell diverticulum is a frequent cause of a symptomatic recurrence or persistence of symptoms following surgery of division of a vascular ring without resection of the diverticulum.

Further Reading

Ziemer G, Kaulitz R. Congenital anomalies of the great vessels. 21.2.2 Aortic anomalies causing compression of neighboring structures. In: Cardiac surgery. Operations on the heart and great vessels in adults and children, Ziemer G, Haverich A, editor. 1st engl edition. 1158 pp. Berlin: Springer; 2017, p. 688–99.