Vascular Rings and Pulmonary Slings

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18.1 Introduction

Development of the mediastinal great vessels is the result of the growth and resorption of vessels derived from the two dorsal aortae and all the derived arches (Fig. 18.1). An alteration in the process that originates the great vessels can result in the development of a complete ring surrounding the trachea and esophagus. Depending on the type of alteration, several anatomical variants are possible, all causing an obstruction at respiratory and esophageal levels. The indication for corrective surgery to relieve the obstructive symptoms is a consequence of their tendency to constrict the trachea and/or esophagus [1]. Vascular rings comprise an estimated 1% of cardiovascular malformations that are managed surgically.

18.2 Vascular Ring

18.2.1 Etiology and Pathogenesis

Vascular development in the embryo starts from the connection of the ventral and dorsal aortae by the six pairs of aortic/branchial arches. Usually the first, second, and fifth arches as well as a segment of the right fourth arch regress, giving the normal left aortic arch. The residual third arch gives origin to the common carotid arteries, whereas from the residual portion of the right fourth arch and the seventh intersegmental artery develop the right subclavian artery. The proximal portion of the pulmonary arteries and the ductus arteriosus are formed by the sixth arch. Failure of this process causes erroneous persistence or reabsorption of segments of the primitive vascular system, thereby accounting for most of the congenital vascular rings and vascular slings [2].

18.2.2 Classification

Several anatomical variants can be present, and we report here the most frequently described. Most of these variants have a coexistent right aortic arch. A left dominant aortic arch is very rare but needs to be considered for a surgical approach through a right thoracotomy.

18.2.2.1 Double Aortic Arch

A double aortic arch is caused by persistence of the right dorsal aorta. In these patients, left and right aortic arches are present. The right arch gives rise to the right common carotid artery and the right subclavian artery, whereas

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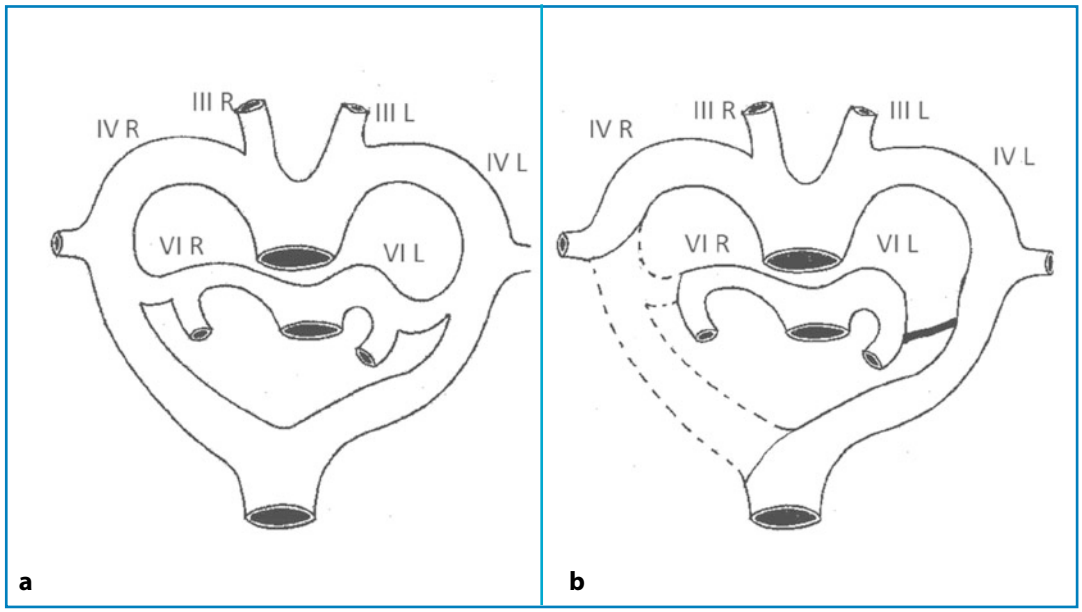

Fig. 18.1 The developing aortic arch (**a**) and the resulting normal left-sided aortic arch with all the vascular segments that disappear (*broken lines*; **b**) (schematic). Roman numbers refer to the embryological aortic arches

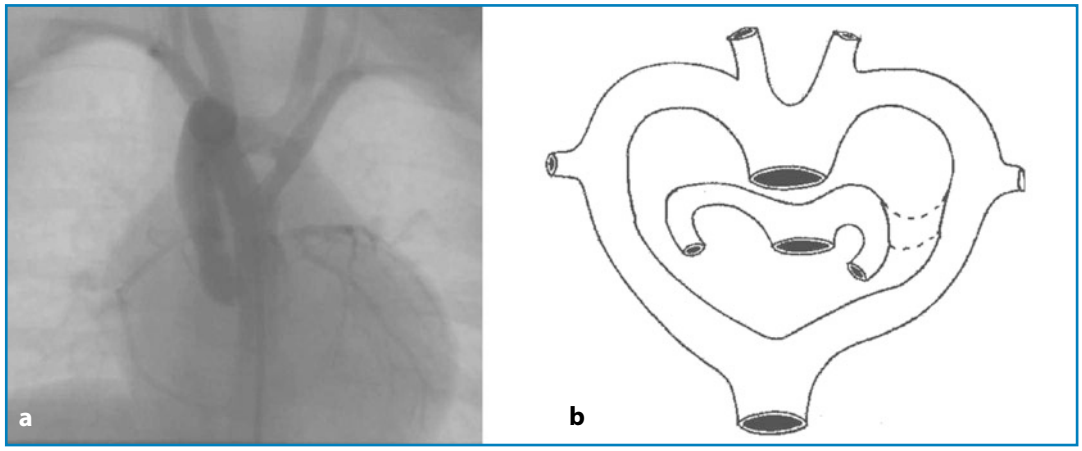

Fig. 18.2 Angiography in a double aortic arch (**a**) and schematic view showing the embryological origin (**b**)

the left arch gives rise to the left common carotid and left subclavian artery. In approximately 70% of cases, there is a dominant right arch and a hypoplastic (or even atretic) left arch that joins the descending aorta at its emergence from behind the esophagus near the insertion of the ligamentum arteriosus. The descending aorta is usually left-sided, but can be right-sided or in the midline (Fig. 18.2).

18.2.2.2 Right Aortic Arch, Aberrant Left Subclavian Artery and Left Ligamentum Arteriosus

This anomaly originates from persistence of the right fourth aortic arch with resorption of the left fourth aortic arch. The right aortic arch gives origin to the left common carotid artery, the right common carotid artery, the right subclavian artery and the left subclavian artery

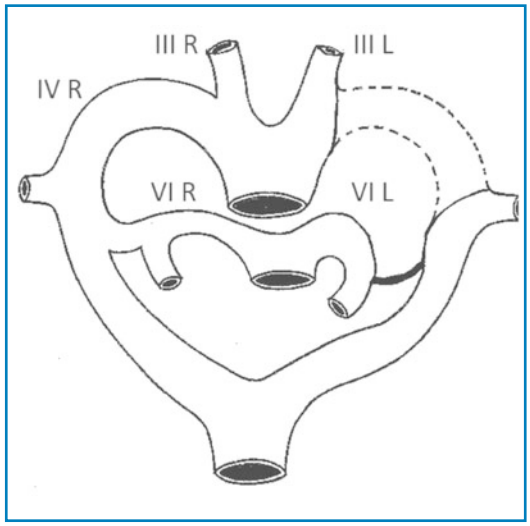

Fig. 18.3 Right aortic arch, aberrant left subclavian artery and left ligamentum arteriosus. The portion of the arch that is reabsorbed is represented by the broken lines. The *black bold line* is the ligamentum arteriosus. *Roman numbers* indicate the embryological aortic arches

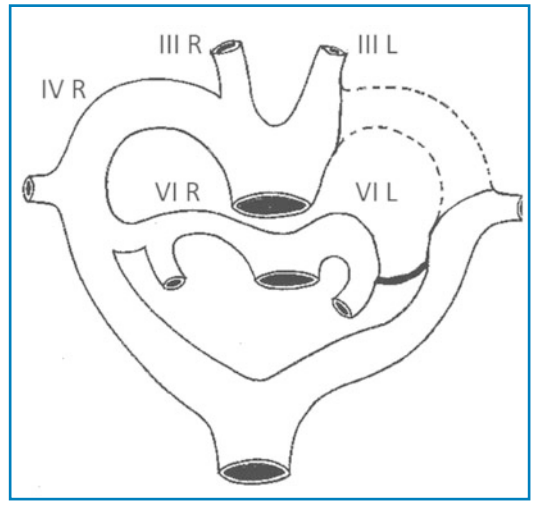

Fig. 18.4 Right aortic arch, mirror-image branching, and retro-esophageal ligamentum arteriosus. The portion of the arch that is reabsorbed is shown by broken lines. The *black bold line* refers to the retro-esophageal ligamentum arteriosus. *Roman numbers* refer to the embryological aortic arches

crossing the midline behind the esophagus. The ligamentum arteriosus, arising from the left subclavian artery, is connected to the left pulmonary artery originating a complete vascular ring that embraces the trachea and esophagus. Usually, the ligamentum arteriosus is on the same side as the aortic arch, but can develop on the contralateral side to the arch and is responsible for completing the vascular ring. A small diverticulum named "diverticulum of Kommerell" can occur at the site of the aberrant subclavian artery, and represents the aortic attachment of the ligamentum arteriosus (Fig. 18.3).

18.2.2.3 Right Aortic Arch, Mirror-image Branching and Retro-esophageal Ligament

This anomaly derives from persistence of the right fourth aortic arch and resorption of the fourth left aortic arch. A short portion of the distal end of the fourth left arch persists as the diverticulum of Kommerel. From the right aortic arch arise in sequence the left brachiocephalic trunk, the right common carotid artery and the left subclavian artery. A fibrous ligament originating from the prominent diverticulum passes leftward behind the esophagus and then anteriorly to connect to the left pulmonary artery, closing the ring around the trachea and esophagus (Fig. 18.4).

18.2.2.4 Left Aortic Arch, Mirror-image Branching, Right Descending Aorta and Atretic Right Aortic Arch

A normal left-sided aortic arch is present in this anomaly. After passing the midline in a retro-esophageal position, it joins the right descending thoracic aorta. An atretic right aortic arch, immediately superior to a right ligamentum arteriosum connecting to the right pulmonary artery, gives origin to the vascular ring.

18.2.2.5 Left Aortic Arch, Right Descending Aorta and Right-sided Ligamentum Arteriosum to the Right Pulmonary Artery

This anomaly is characterized by the mirror image of the right aortic arch that gives rise in sequence to the following arteries: right common carotid, left common, left subclavian and

right subclavian. The arch passes to the left of the trachea and then behind the esophagus to join the right descending thoracic aorta. At this level, an atretic right arch is found forming a diverticulum.

18.2.3 Clinical Features

Common symptoms include stridor, wheezing, respiratory distress, cough or evidence of dysphagia, or difficulty with feeding that can also worsen respiratory symptoms. In less serious cases, patients can remain asymptomatic throughout their life. In more severe cases, stridor is common as early as the neonatal period. Feeding can also be difficult, especially when progressing to solids. Reflux and respiratory infections are quite common [3, 4].

18.2.4 Associated Anomalies

A double aortic arch is usually an isolated lesion without other cardiovascular anomalies. In about 20% of cases, it is associated with a deletion in chromosome band 22q11. This deletion is responsible for DiGeorge, velocardiofacial, and conotruncal anomaly face syndromes. These syndromes are often referred to using the terms "catch-22 syndrome" or "chromosome band 22q11 deletion syndrome". For this reason, the most common associated defects are ventricular septal defect, tetralogy of Fallot and all the congenital cardiopathies referable to conotruncal anomalies.

18.2.5 Diagnosis

Chest radiography alone or barium esophagography can give strong clues to the definitive diagnosis by showing signs of compression in the airway or esophagus. However, more comprehensive imaging details are required, and magnetic resonance imaging (MRI) and computed tomography (CT) have sensitivities approaching 100% [5] (Fig. 18.5). In both cases, all the details necessary to plan corrective surgery can be provided [4, 6]. Bronchoscopy may be necessary to disclose pulsatile external airway compression.

18.2.6 Management

In a symptomatic child, surgery is indicated. Symptoms can be present in the neonatal period and prompt relief of obstruction may be indicated. Less serious symptoms may be diagnosed in the older infant (usually recurrent infection or other respiratory symptoms rather then stridor or dysphagia). In these cases, elective surgery is indicated.

18.2.6.1 Surgical Procedure

The first attempts at surgery for vascular rings were by Gross in 1945 [7]. Preoperative evaluation of imaging and clinical findings is crucial to plan the side of the thoracotomy [8]. In >95% of cases, a left thoracotomy is preferred. A right thoracotomy is necessary only if a left dominant aortic arch is present. In patients with a double aortic arch, the dominant one must be disclosed (which can be difficult). This evaluation is fundamental to choose the side of the thoracotomy, which will be on the right in case of dominant left arch and *vice versa*. Intraoperatively, arterial pressure in one of the arms and legs must be ascertained to check the effect of temporary closure of the smaller arch. This measurement ensures that a pressure gradient is not present at the level of the dominant aortic arch.

The patient is positioned in the lateral decubitus position. A conventional posterolateral thoracotomy (usually in the fourth intercostal space) is carried out. After retracting the lung anteriorly, descending aorta, distal arch, and all the vessels that can be reached are exposed and surrounded with a vessel loop (Fig. 18.6a). The vagus nerve (giving origin to the recurrent laryngeal nerve that passes around the ligamentum arteriosus) can be a useful landmark. In the case of a double aortic arch, the distal portion of the diminutive arch is clamped to check that

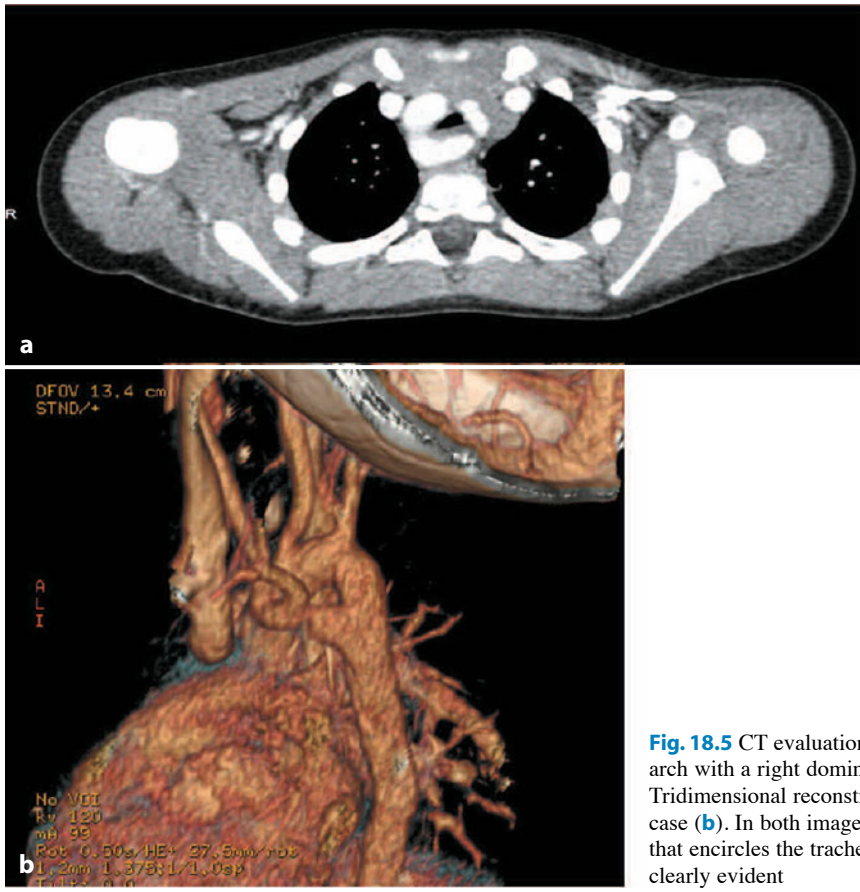

Fig. 18.5 CT evaluation in a double aortic arch with a right dominant arch (**a**). Tridimensional reconstruction of the same case (**b**). In both images, the vascular ring that encircles the trachea and esophagus is clearly evident

a pressure gradient between arms and legs is present. The arch is transected between the clamps and the two sides closed with a double running suture (Fig. 18.6b). This causes spontaneous retraction of the two stumps, indicating the tension with which the ring surrounds the trachea and esophagus. If a ligamentum is not present, this must be identified and transected. In both cases, careful dissection of the vessels from the surrounding tissue is important to provide space to the trachea and esophagus. Care must be taken not to dissect the esophagus, which can necrotize for the metameric vascularization that characterizes this organ. Moreover, checking for serous leakage and eventually solving the problem is very important because it can cause postoperative chylothorax.

18.2.7 Outcome and Follow-up

Mortality and morbidity have been reported to be very low [1, 4]. In infants, severe symptomatic tracheomalacia may be associated with long-standing compression. In these cases, relief of all respiratory symptoms can be progressive, and complete and rapid improvement in feeding is always obtained [3].

18.3 Pulmonary Artery Sling

A pulmonary artery sling is a rare anomaly in which the left pulmonary artery originates from the right pulmonary and, by surrounding the distal part of the trachea and right main-stem bronchus, runs between the trachea and esophagus [9] (Fig. 18.7).

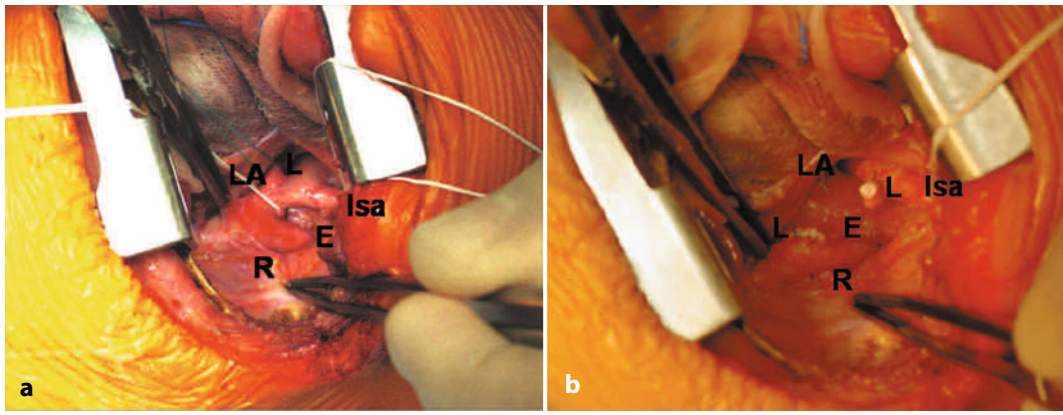

Fig. 18.6 Intraoperative view of a double aortic arch with a dominant right aortic arch. A left thoracotomy has been carried out and the two arches can be visualized (**a**). After division of the left arch, the two stumps move away, leaving more space for the esophagus (**b**). *L*, left arch; *R*, right arch; *LA*, ligamentum arteriosum; *lsa*, left subclavian artery; *E*, esophagus

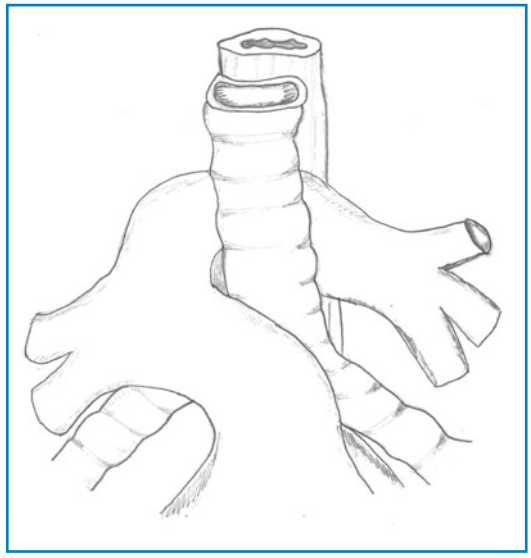

Fig. 18.7 Pulmonary artery sling (schematic). The left pulmonary artery arises from the right pulmonary artery and courses posteriorly to the trachea

18.3.1 Etiology and Pathogenesis

A pulmonary artery sling is a rare anomaly, accounting for <1% of all congenital heart defects. It is caused by abnormal progression of the aortic sac and/or development of the sixth arch. Such abnormal progression results in an extrapericardial origin of the left pulmonary artery from the posterior aspect of the right pulmonary artery. It then encircles the right main-stem bronchus and courses posteriorly between the trachea and esophagus. The ligamentum arteriosum or the ductus itself originates from the main pulmonary artery and passes anteriorly to the left main bronchus and anomalous left pulmonary artery to join the aorta and complete the vascular ring.

18.3.2 Clinical Features

The clinical features are secondary to the associated tracheal lesions and coexisting cardiac anomalies. In isolated cases (and different to vascular rings), symptoms are exclusively of a respiratory nature. In approximately 65% of cases, stenosis of the tracheobronchial tree is present. Tracheomalacia is not usually found in these patients.

18.3.3 Associated Anomalies

In approximately 50% of cases, other congenital cardiac defects are coexistent. The most common are atrial or ventricular septal defects, patent ductus arteriosus, and tetralogy of Fallot.

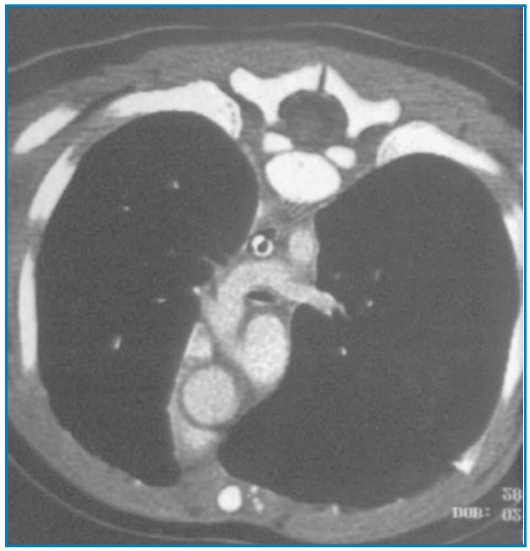

Fig. 18.8 MRI of a pulmonary artery sling

18.3.4 Diagnosis

The diagnosis is usually obtained by echocardiography (which can also disclose coexisting cardiac anomalies). Chest radiography can be useful if airway compression is suspected. Bronchoscopy is essential to exclude tracheal stenosis secondary to complete tracheal rings. CT or MRI can give the complete diagnostic picture and realistic reconstruction of the anatomy [10] (Fig. 18.8).

18.3.5 Management

The diagnosis of a pulmonary artery sling is an indication for surgery. In asymptomatic, isolated cases, a conservative attitude is preferable. Surgery alone can address the pulmonary artery sling or necessitate tracheal reconstruction if a stenosis or complete tracheal ring is present [11].

18.3.5.1 Correction of an Isolated Pulmonary Artery Sling

Surgery is usually *via* median sternotomy (Fig. 18.9). After extensive dissection of the left pulmonary artery, cardiopulmonary bypass (CPB) is established. The decision of undertaking the procedure on a beating heart or under cardioplegic arrest is left to the surgeon. The left pulmonary artery is divided at its origin and dissected free from the posterior aspect of the trachea and left bronchus. The proximal portion is closed with a running suture and, after relocating the artery in its usual position, it is implanted on the main pulmonary artery [12].

18.3.5.2 Correction of a Pulmonary Artery Sling and Tracheal Stenosis

Using a median sternotomy and CPB, extensive dissection of the left pulmonary artery is

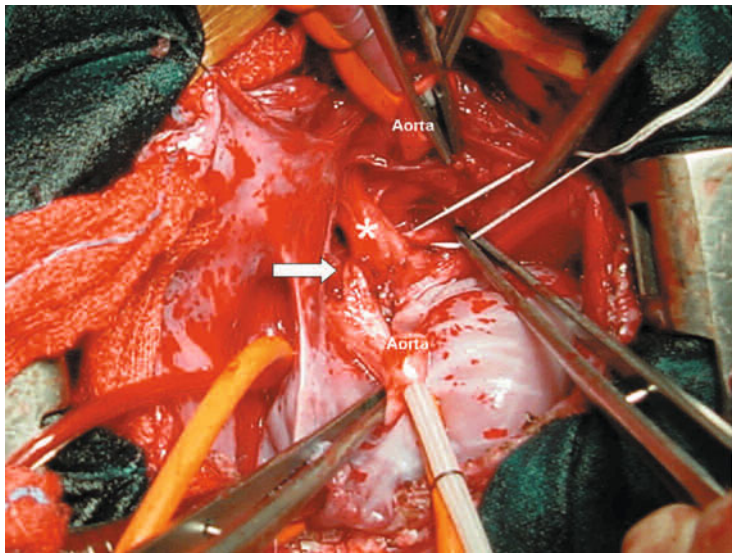

Fig. 19.9 Intraoperative view of a pulmonary artery sling. A median sternotomy has been carried out. The two stumps of the divided ascending aorta (*Aorta*) disclose the trachea (*asterisk*). The *white arrow* refers to the left pulmonary artery, which originates from the right pulmonary artery and courses behind the trachea. Its distal portion is surrounded by a white snare and is indicated by the forceps

achieved. The stenotic segment of the trachea is exposed and repaired, depending on the anatomy of the lesion, by: resection and end-to-end anastomosis; pericardial tracheoplasty; tracheal autograft; tracheal resection; cartilage tracheoplasty; slide tracheoplasty; or aortic homograft patch tracheoplasty [6]. In the case of transection of the trachea, the left pulmonary artery is moved anteriorly and can be left in this position unless a "kinking" at the origin is observed. In this case or if transection of the trachea is not carried out, the left pulmonary artery is re-implanted as described previously [13].

18.3.6 Outcome and Follow-up

Morbidity and mortality are consequences of the involvement of the respiratory tree. In these cases, the necessity to also undertake tracheal correction affects short- and long-term results. In isolated cases, results are very good and mortality has been reported to be <1%.

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