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Review Article

Vascular rings and pulmonary arterial sling: from respiratory collapse to surgical cure, with emphasis on judicious imaging in the hi-tech era

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VASCULAR RINGS, AND PULMONARY ARTERIAL slings, are relatively rare vascular anomalies, but are important lesions to recognize, since if left untreated, patients may experience life-threatening respiratory compromise and "near death" episodes. Surgical therapy, however, is almost always curative. As most infants and children initially present with respiratory symptoms, they are frequently referred to and managed by pulmonologists and otorhinolaryngologists before being presented to a surgical team. A high index of suspicion is necessary to direct the diagnostic work-up, which may rapidly reveal the correct diagnosis, and allow for timely surgical repair.

In the current era, an increasing number of diagnostic procedures are performed preoperatively, often yielding repetitive or overlapping information. Nonetheless, the hi-tech escalade of new imagery has not provided better results, and is substantially more expensive.

This article reviews the history of surgical correction of rings and slings, the standard surgical approaches, the results, and the long-term outcomes. Emphasis is given on a suggested algorithm for diagnostic work-up. If followed, this may avoid the frequent unnecessary delay, along with superfluous investigations in achieving a correct diagnosis, and lead more rapidly to successful surgical treatment.

Historical background

Although the earliest description of tracheal or esophageal compression by a vascular structure was first given in 1737 by Hommel,[1] over two centuries elapsed before the first surgical correction of a complete vascular ring was performed, by Robert Gross in 1945.[2] In his landmark article, he described the technique for repair of double aortic arch through a left anterior thoracotomy. He described two other cases, another double aortic arch, and a complete ring formed by a right aortic arch with retroesophageal left subclavian artery and left-sided arterial ligament.[2] Gross subsequently performed the first successful operations for partial vascular rings, namely an aberrant right subclavian artery causing dysphagia lusoria in 1946, followed by suspension of the brachiocephalic artery to the sternum in 1948.[3]

Willis Potts was the first to describe, in 1954, a successful operation for pulmonary arterial sling, performed at Children's Memorial Hospital in Chicago.[4] This was achieved through a right thoracotomy, with division and reimplantation of the left pulmonary artery anterior to the trachea. The first successful combined repair of pulmonary arterial sling and complete tracheal rings with a pericardial patch using cardiopulmonary bypass was also performed at Children's Memorial Hospital, by Farouk Idriss in 1982.[5] Since then, surgical approaches and techniques have varied little, and the minor controversies in treating the syndromes of vascular compression lie more among diagnostic preferences and surgical timing.

Symptoms and diagnostic work-up

A combination of respiratory symptoms are often the presenting mode for vascular rings. They include stridor, a "seal-bark" cough, asthma, recurrent pneumonia, respiratory distress, cyanosis, and apnea. Complete vascular rings present at a younger age

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and more severely. They are more commonly associated with the "seal-bark" cough, which is a typical early finding in patients with double aortic arch.[6] Apnea is prominent in patients with compression from the brachiocephalic artery, and in those with complete tracheal rings. Patients with compression produced by the brachiocephalic artery often have a characteristic posture, with opisthotonos and hyperextension of the head.[6] Dysphagia and choking episodes occur in older children subsequent to intake of solid food, or inadvertent swallowing of miscellaneous objects.[6] Feeding difficulties in general may lead to failure to thrive with concomitant stunting of weight.[6]

Unfortunately, the diagnostic work-up is frequently uncoordinated, and many examinations are performed unnecessarily, even when a correct diagnosis has already been established. Ideally, minimal imagery that suggests a diagnosis leading to the correct surgical approach should suffice. It is important to stress that more than nine-tenths of all lesions producing vascular compression may be corrected through a left thoracotomy.[6–9] As intraoperative dissection and identification of all structures represents the ultimate diagnostic confirmation,[10] only certainty in the incisional approach is the one absolute preoperative must. Precise anatomic delineation with invasive or expensive procedures may be desirable for some surgeons, but are truly necessary only in the very rare cases requiring a right thoracotomy, or when the diagnosis is altogether still in doubt. Along the same line of thought, more surgeons are advocating a median sternotomy as the standard approach, as it potentially allows repair of virtually any anomaly.[10]

The chest X-ray may give the first clue to the diagnosis of extrinsic airway compression, and eventually to the type of vascular anomaly.[9,11] Indentation upon the trachea is visible on a plain or high voltage chest X-ray (Figs 1 and 2). Atelectasis or hyperinflation of various dependent lung lobes should hint at an abnormal vascular pattern.[6,7,12] Hyperinflation of the entire right lung is typically seen in patients with pulmonary arterial sling.[6] The side of the arch, in particular a right aortic arch, can be readily diagnosed on a plain film. A double aortic arch should be suspected if the side of the arch is indeterminate, being hidden in the middle of the X-ray, overlapped by the various mediastinal structures and the spine.[6]

A barium swallow is the single most important diagnostic procedure in the work-up,[6,7,9,11,12] and the second to perform after an X-ray. It is specific, relatively non-invasive, cost-effective, and should in itself be sufficient to proceed with surgery without further undue escalade in imagery.[6,7,9,11–13] Even in

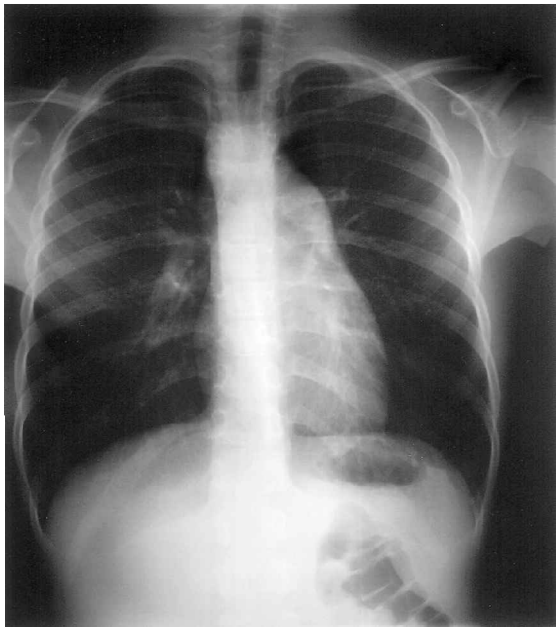

Figure 1.
Antero-posterior chest X-ray revealing indentations upon the trachea in a patient with double aortic arch.

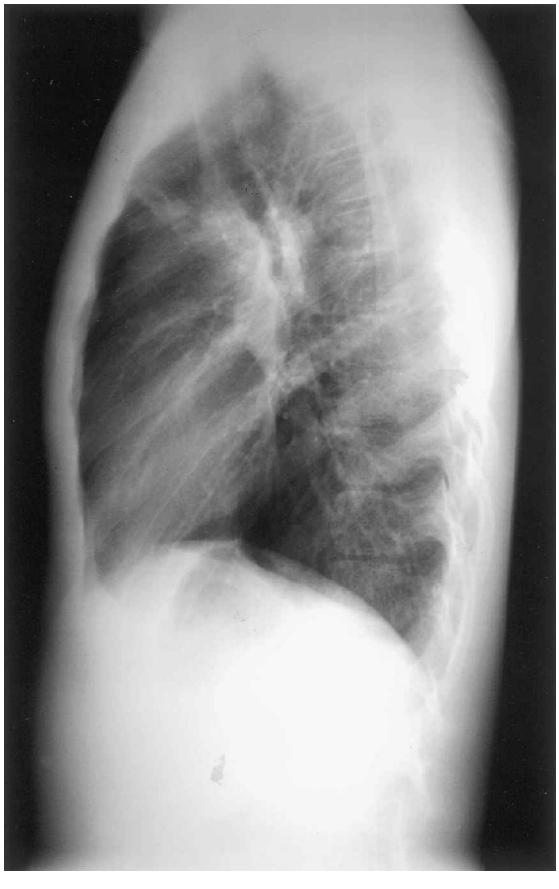

Figure 2.
Lateral chest X-ray from the same patient with an anterior indentation upon the trachea.

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Figure 3.
Antero-posterior barium oesophagram of a patient with double aortic arch and dominant right arch. Note the higher and more pronounced indentation on the oesophagus on the right side as compared to the left. This image is indistinguishable from that of a right aortic arch with left ligament.

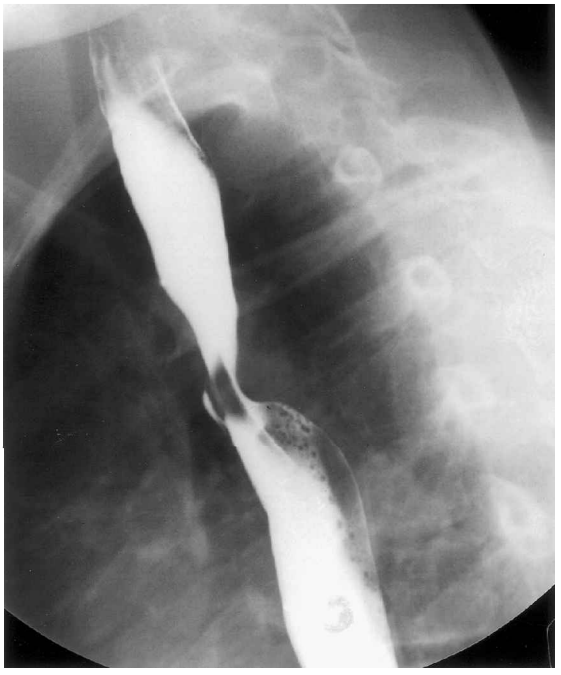

Figure 4.
Lateral oesophagram of the same patient with double aortic arch showing a deep posterior indentation on the oesophagus.

reports advocating the virtues and specificity of more novel imagery as compared to traditional ones, the correct diagnosis was achieved by an oesophagram in two-thirds to nine-tenths of cases.[11–15] A double aortic arch will cause compression on both sides of the esophagus on an anteroposterior view (Fig. 3), with a posterior indentation on a lateral view (Fig. 4). Most commonly, the right arch is dominant, causing a higher and more pronounced indentation on the right side of the oesophagus than the left.[6,9,11,12] This image is identical in patients with a right aortic arch and left-sided arterial ligament,[6] but as the surgical approach is the same in both situations, namely a left thoracotomy, further investigation is unnecessary. In the more rare cases of double aortic arch with a dominant left arch, the left indentation is higher up than the right one.[6,7,9] In a patient with symptoms compatible with a vascular ring and a normal barium oesophagram, compression by the brachiocephalic artery should strongly be suspected. Finally, the pulmonary arterial sling causes an anterior indentation on the lateral view of an esophagram, without a posterior component.

Rigid bronchoscopy is a diagnostic must in patients with compression from the brachiocephalic artery, which will reveal the level and degree of an anterior pulsating mass causing stenosis of the tracheal lumen.[6,11,16] The compression progresses from most severe on the right side to less severe on the left, following the course of the brachiocephalic artery as it originates from the aortic arch.[6] Pressing the bronchoscope anteriorly against the tracheal wall will typically suppress the right radial or right carotid arterial pulses.[6,12] Bronchoscopy is also indispensable in the assessment of congenital tracheal stenosis from complete tracheal rings, which is frequently associated with pulmonary arterial sling.[17] It is estimated that up to three-quarters of patients with the sling have tracheal stenosis from complete tracheal rings.[17] hence the term "ring-sling" complex,[18] and that one-third of patients with complete tracheal rings have an associated pulmonary arterial sling.[17,19] Bronchoscopy may define the level, length, and degree of luminal narrowing, and allows precise surgical planning.

In patients with complete tracheal rings, distal tracheo-bronchomalacia is often associated and underscored.[19] The extent of distal airway involvement may help to predict the weaning potential from mechanical ventilation postoperatively.[17] Defining the severity of distal disease preoperatively is essential, therefore, as extensive bronchomalacia of the smaller

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airways may discourage or contraindicate surgical repair of the major airway. To assess this, concomitant bronchography is useful, both to confirm and delineate the precise location of compression upon the trachea from the sling, but also to disclose disease of the smaller airways.[17]

Computer tomography scanning and magnetic resonance imaging are both useful when the diagnosis, but not the precise anatomy of a vascular ring, is still uncertain after the aforementioned procedures. They may reveal the classic "four-vessel sign", namely the presence of 4 brachiocephalic vessels rising separately from the aortic arch. The same image is common in patients with a right aortic arch and a left-sided arterial ligament. Magnetic resonance imaging is advocated by some as the diagnostic procedure of choice[8,13,15,20,21] instead of bronchoscopy and the oesophagram, although this view is by far not universally shared, and not advocated by the authors.

The usefulness of angiography is currently debatable,[7,14,15,22] although it is still routinely used in many centers to confirm compression by the brachiocephalic artery. Angiography may be the only imaging modality which provides anatomic definition in rare vascular anomalies.[10,15,22–26] It may be indicated when an associated cardiac anomaly is suspected, and when concomitant surgical correction of a heart defect is planned. Alternatively, echocardiography is often sufficient, both to rule out a pulmonary arterial sling, and to assess intracardiac anatomy.[6,27] The potential iatrogenic morbidity of angiography should be remembered when investigating children, namely allergic reactions, embolic phenomenons, vascular injury at the site of arterial puncture, radiation, the need for general anesthesia, and hence prolonged periods of immobilization and stay in hospital,[6,15] not to mention considerations of cost.

Classification and surgical technique

Vascular rings are traditionally divided into three groups, namely complete and partial rings, and the pulmonary arterial sling (Fig. 5). The more common complete rings include double aortic arch (Fig. 6), right aortic arch associated with a left-sided arterial ligament with or without an aberrant left subclavian artery, and left aortic arch associated with a right descending aorta and a right-sided arterial ligament. Partial rings include aberrant right subclavian artery and compression by the brachiocephalic artery. When a right aortic arch is present with mirror imaged branching, the left-sided ligament usually arises from the left brachiocephalic artery, and a complete ring is *not* formed[6,9,12] (Fig. 7). This is seen

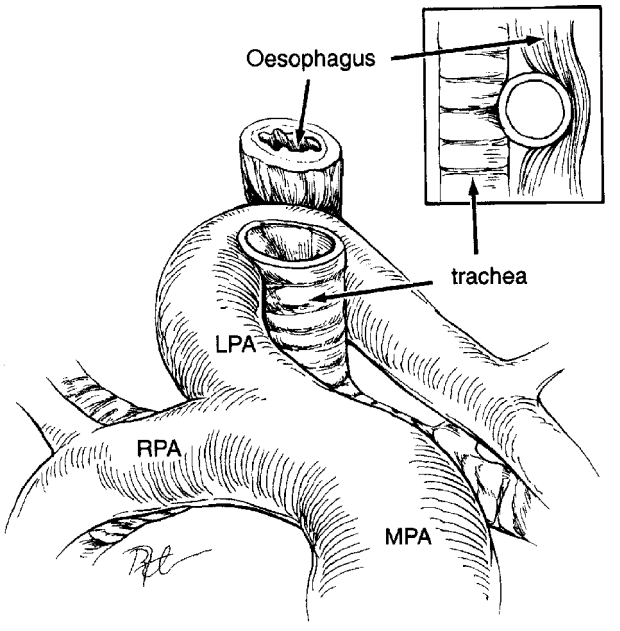

Figure 5.
Pulmonary arterial sling. The left pulmonary artery (LPA) originates from the right pulmonary artery (RPA) and courses between the oesophagus and trachea to reach the left lung. Inset shows a lateral relationship of the left pulmonary artery to the oesophagus. It is this view that can be diagnostic on a barium swallow. MPA: main pulmonary artery.

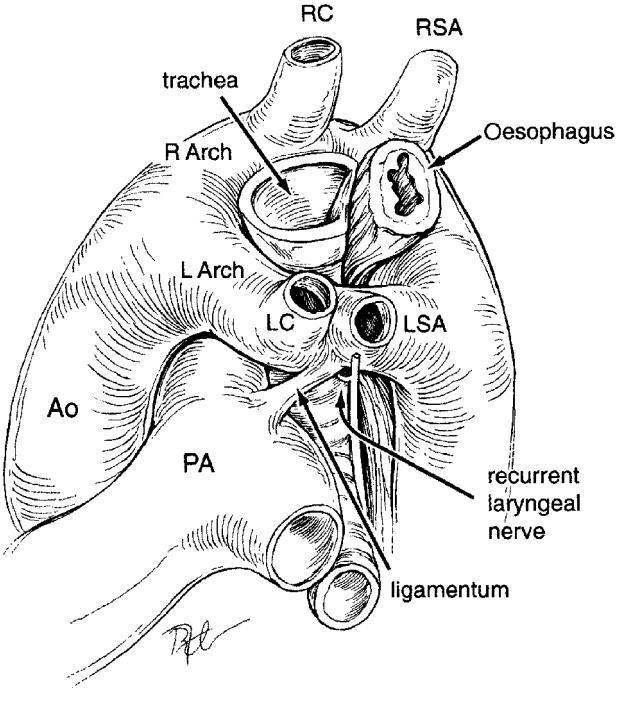

Figure 6.
Double aortic arch, right arch dominant. Left carotid artery (LC) and left subclavian artery (LSA) originate separately from the left aortic arch (L Arch). Ao: aorta; PA: pulmonary artery; R Arch: right aortic arch; RC: right carotid artery; RSA: right subclavian artery.

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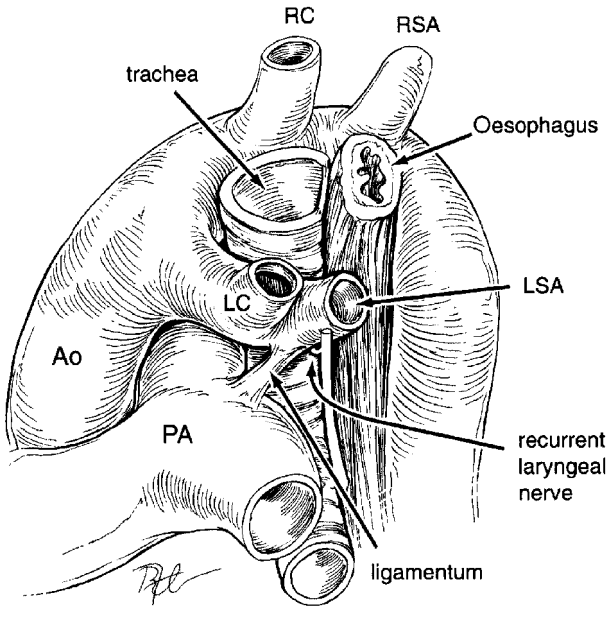

Figure 7.
Right aortic arch with mirror imaged branching. The ligament is often from the left pulmonary artery (PA) to the left brachiocephalic artery and a complete vascular ring is not *formed. Ao: aorta; LC: left carotid artery; LSA: left subclavian artery; RC: right carotid artery; RSA: right subclavian artery.*

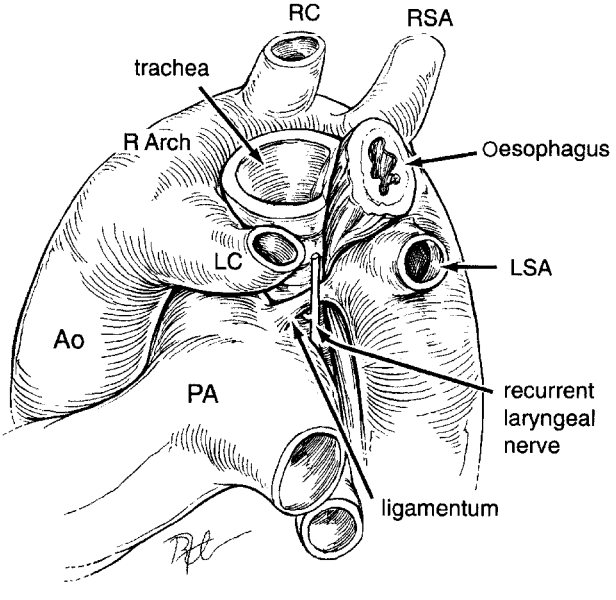

Figure 8.
Right aortic arch (R Arch) with retroesophageal left subclavian artery (LSA) and left ligament. The ring is formed by the right arch, pulmonary artery (PA), and ligament. Ao: aorta; LC: left carotid artery; RC: right carotid artery; RSA: right subclavian artery.

in up to one-fifth of patients with tetralogy of Fallot.[8,9] If the ligament arises from the aorta behind the oesophagus, however, and attaches to the left pulmonary artery, there will be a compressive

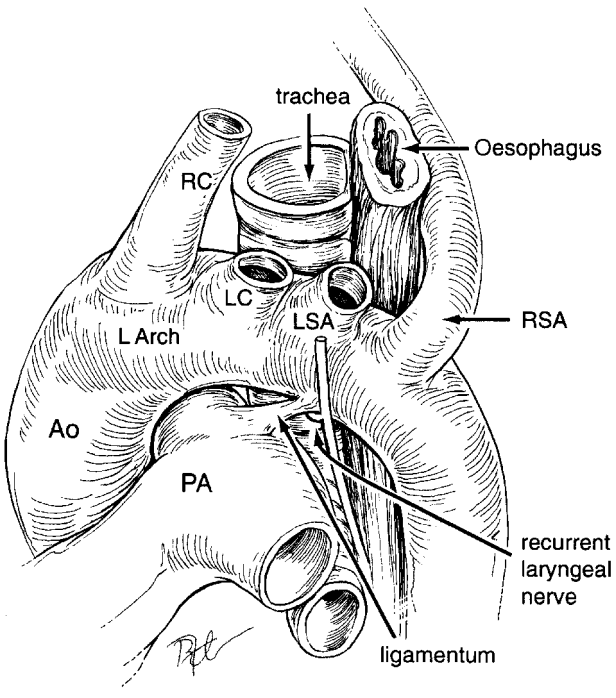

Figure 9.
Left aortic arch (L Arch) with aberrant right subclavian artery (RSA). The right subclavian artery is the last brachiocephalic vessel from the aortic arch. Ao: aorta; LC: left carotid artery; LSA: left subclavian artery; PA: pulmonary artery; RC: right carotid artery.

ring[6,8,9] (Fig. 8). Right aortic arch with left-sided arterial ligament, therefore, may be classified either as a complete or partial ring.

Patients with a right aortic arch and left ligament frequently have persistence of Kommerel's diverticulum.[6] This structure is a remnant of the left fourth arch, and typically arises at the origin of the left subclavian artery. As it may form an aneurysm, and compress adjacent structures, most agree to incorporate excision of the diverticulum or aortopexy at the time of repair of the vascular ring.[6,9,14]

Aberrant origin of the right subclavian artery is the most common vascular anomaly of the aortic arch, found in up to 2% of the population[8] (Fig. 9). As it is almost always asymptomatic,[6,27] some question the validity of its incorporation amongst partial vascular rings.[6] The relative prevalence of the most common vascular lesions, and their operative approach, is given in Table 1.

Surgery is indicated in all symptomatic patients,[6,9,12] as attempts at medical management result in a mortality of 90%.[12] Timing of the intervention need not be delayed upon diagnosis,[6,9] as any type of vascular ring places the patient at a potential risk for severe respiratory compromise, hypoxia, or apneic episodes. Other potential complications of unrepaired lesions include aortic dissection and

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Table 1. Classification and surgical approaches.

Anomaly	Incidence (%)	Approach
Complete rings (75%)		
Double aortic arch	55	
Right arch dominant	73[6]–80[8]	Left thoracotomy
Left arch dominant	15[8]–20[6]	Right thoracotomy
Arches equal	5[8]–7[6]	Left thoracotomy
Right aortic arch with left ligament and mirror branching along with aberrant right subclavian artery	45	Left thoracotomy
Left aortic arch with right descending aorta and right ligament	<1	Right thoracotomy
Partial rings (20%)		
Compression by brachiocephalic artery		Right thoracotomy or sternotomy
Right aortic arch with left ligament and mirror branching and aberrant right subclavian artery		Left thoracotomy
Pulmonary arterial sling (5%)		Median sternotomy or left thoracotomy

aneurysm.[28] In intubated patients, earlier repair is performed to lessen the iatrogenic consequences of mechanical ventilation, namely erosions into the trachea, aorta, or esophagus.[6,9]

Some controversy exists as to the indication and timing of surgery in asymptomatic patients with compression by the brachiocephalic artery. Backer et al.[6] use a criterion of narrowing of the tracheal lumen of three-quarters, as assessed by bronchoscopy, before considering patients for surgery. Filston et al.[29] reported on 20 patients, of whom only four were symptomatic and underwent surgery. They state that none of the asymptomatic patients, representing four-fifths of the total group, needed surgical intervention, claiming that all remained asymptomatic,[29] although the follow-up interval from the time of diagnosis was not specified. Mustard et al.[30] also reported almost nine-tenths of patients in their series as being asymptomatic, all of whom were successfully managed non-operatively, postulating that the artery moves up and off of the trachea as the child grows.

Almost all vascular rings are approached through a left thoracotomy. These include double aortic arch and the combination of right aortic arch and left ligament, which together represent more than three-quarters of all vascular rings and slings. After full dissection of all pertinent vascular and ligamentous structures, along with maximal mobilization from adjacent mediastinal and thoracic adhesions, relief of tracheo and/or oesophageal compression is achieved by dividing the compressing vessel or ligament. The mediastinal parietal pleura is typically not sutured closed, as formation of scar tissue may contribute to repeat compression, as originally caused by the vascular ring.[6]

Exceptions to an approach through a left thoracotomy are the more rare compression by the brachiocephalic artery, the combinations of left aortic arch and right descending aorta with a right ligament to the right pulmonary artery, left aortic arch with aberrant right subclavian artery and right ligament, and double aortic arch with atresia of the right (posterior) arch. All of these are approached through a right thoracotomy,[22] required in less than one-twentieth of all patients with vascular rings and pulmonary arterial sling.

Of note, the compressing brachiocephalic artery may also be repaired through a median sternotomy, and is the preferred approach by some. The repair of such compression consists of placing multiple pledgetted sutures through the base of the artery and distal ascending aorta, and suspending them from the posterior wall of the sternum (Fig. 10). Intraoperative bronchoscopy will simultaneously visualize widening of the tracheal lumen, and decrease or disappearance of the pulsating vascular compression, as the sutures are pulled upwards towards the sternum.[6,29] It is important to avoid any dissection between the brachiocephalic artery and the tracheal wall which lies posterior to the vessel.[6,29] The adhesions binding the two will ensure traction by the vessel on the trachea, as both are attached to the sternum.

Pulmonary arterial slings are most commonly approached through a median sternotomy, which allows access for cardiopulmonary bypass and concomitant repair of intracardiac defects and/or complete tracheal rings.[6,27] Cardiopulmonary bypass may be conducted at normothermia, or at moderate hypothermia maintaining a beating heart. Techniques include either transection of the trachea and its reanastomosis posterior to the left pulmonary artery[31] or, more commonly, transection of the left pulmonary artery and reimplantation anterior to the trachea.[27] The advantage of the latter technique is the avoidance of manipulation or sutures into the tracheal wall,

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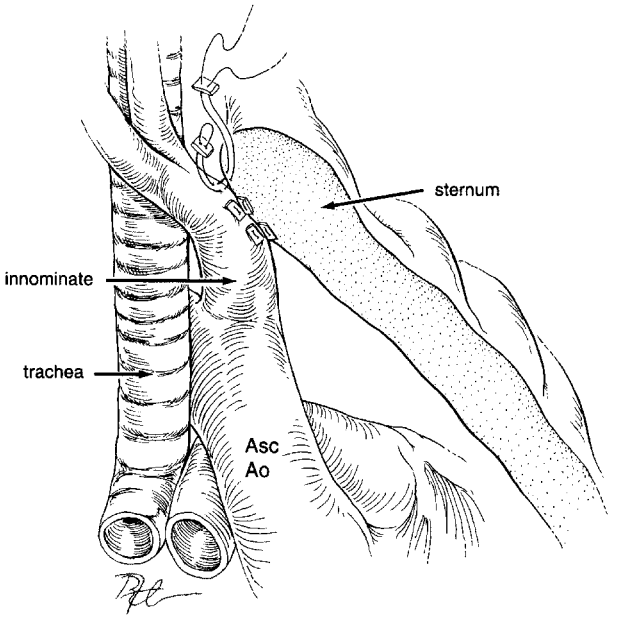

Figure 10.
Suspension of the brachiocephalic artery. Fixing the adventitia of the artery to the posterior table of the sternum with pledgetted sutures pulls the artery anteriorly and actively pulls forward and opens the tracheal wall. Asc Ao: ascending aorta.

which commonly produce granulation tissue with resultant restenosis at the tracheal anastomosis.[17] The technique also avoids kinking of the left pulmonary artery.[27] Ligation and reimplantataion of the left pulmonary artery may potentially create iatrogenic stenosis at the vascular anastomosis, although excellent results with complete patency of the left pulmonary artery have been achieved through meticulous surgical technique.[27]

Postoperative care, operative and long-term results

Immediate or early extubation postoperatively is an important factor in reducing stay in intensive care and respiratory morbidity.[27] Even up to years after successful surgery, patients may typically retain noisy breathing, and the urge to reintubate them should be resisted. Rather, high humidity oxygen hoods, chest physiotherapy, and nasopharyngeal suctioning are performed to prevent atelectasis and pulmonary surperinfection.[6,8,9] Morbidity is encountered in up to one-third, in the form of chronic wheezing, pneumonia, injury to the recurrent laryngeal nerve, pneumothorax, chylothorax, and respiratory insufficiency requiring tracheostomy.[6–8] It is important preoperatively to specify to the parents of these patients that respiratory symptoms may not abate immediately despite a satisfactory surgical result.[6,7,17] Indeed, the malacia resulting from extrinsic tracheal compression may take months to solidify after surgical relief, resulting in improvement that is clinically noticeable.

Table 2. Surgical results (excluding associated complete tracheal rings).

	Number of patients	Mortality (%)
Van Son[13] (1994)	39	2.6
Backer[16] (1993)	249	3.6
Chun[14] (1992)	39	5.1
Azarow[15] (1992)	21	4.8
Marmon[12] (1984)	54	1.9
Roesler[7] (1983)	51	3.9
Arciniegas[9] (1979)	53	3.8

Mortality for surgical repair of vascular rings ranges between 0–6%.[6,8] The cause of death is most frequently from complicating upper respiratory tract infections, pneumonia, and respiratory failure. The reported mortality after repair of pulmonary arterial sling ranges between 0–5%.[6,12,27] Using the technique of relocation of the left pulmonary artery with cardiopulmonary bypass, Backer et al.[27] report complete patency of the left pulmonary artery as assessed by angiography, and flow of almost half the output from the right ventricle, as documented by nuclear scanning. When complete tracheal rings are associated with the pulmonary arterial sling, survival rates are in the range of 80%,[6,17,19] and most deaths are related to residual airway stenosis. Surgical results of modern series are given in Table 2.

The long-term outcome of surgery is sparsely documented, as follow-up is undertaken by a heterogeneous group of physicians, resulting in scattering of information. Surgical series claim a complete relief of respiratory symptoms in the majority of cases.[6,7,10,12,14] In a more objective study, Marmon et al.[12] reported pulmonary function studies in 17 asymptomatic patients at a mean of 6.1 years after surgical repair. More than half had abnormal findings indicating fixed or variable obstruction of the central airway. No relationship was found between age of diagnosis and surgery, type of vascular lesion, or severity of presenting symptoms, and the abnormal pulmonary function tests. They concluded that the absence of symptoms despite persistent spirometric aberrations could result from a relative reduction in the percentage of luminal obstruction, owing to growth of the tracheal diameter.[12] More long-term data is desirable, using objective quantification of respiratory function, before confirming the eventual curative nature of surgical therapy.

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Comment

Anomalies of the aortic arch causing tracheo-oesophageal compression account for 1–3%[8,12] of all congenital cardiac malformations. Despite their rare nature, an awareness and a high index of suspicion is required quickly to orientate the diagnostic work-up, and suggest an underlying vascular abnormality as the culprit of the respiratory symptoms. Once diagnosis is established in a symptomatic patient, surgery is indicated without delay, both to avoid problems in feeding with a resulting decrease in somatic growth, ongoing respiratory compromise, cyanosis, and foremost to anticipate impending "near death" episodes. Although it seems intuitive that earlier repair would lead to a shortened interval of extrinsic compression, and hence diminished insult to the tracheal wall,[6,7] no correlation has been demonstrated between the severity of preoperative symptoms, age at repair, type of vascular anomaly, surgical technique, interval to recovery from respiratory symptoms, and long-term results of pulmonary function.[7,12]

Imaging modalities are multiple, and are often more complementary and overlapping than necessary and insightful. Given the overwhelming predominance of the left thoracotomy approach to achieve successful repair of most forms of vascular rings, diagnostic work-up should limit itself to the minimal number of examinations which will allow, with near certainty, an appropriate surgical incision. In the majority of cases, this may be accomplished only on the basis of a chest X-ray and a barium swallow.[6,7,9,11–15] Intraoperatively, surgical dissection and identification of all non and vascular structures will allow for the ultimate and precise diagnosis. Moderating this statement, in the unusual patients that do not fit into the neat classic categories of rings and slings, where the diagnosis is unclear and the approach uncertain, imaging should be continued as far as possible, until the surgeon can confidently perform an incision that will permit successful relief of tracheo-oesophageal compression.[10] Another alternative is routinely to perform a median sternotomy, which provides adequate exposure to repair most lesions, although its advantages and disadvantages compared to a thoracotomy need to be weighed out for each patient.

Surgical technique has not changed considerably for decades, and in isolated vascular rings, where mortality rates are already very low, it is historically difficult to demonstrate a trend of improvement in surgical results. In patients with pulmonary arterial sling and associated complete tracheal rings, improving results are probably due to a combination of better preoperative respiratory management, a widening scope of repair possibilities, facilitation of surgical exposure achieved through cardiopulmonary bypass,[27] and anticipation of postoperative problems relating to healing airways, and the therapeutic armamentum dealing therewith.[17]

Postoperatively, after a variable interval with ongoing respiratory symptoms, the majority of patients become asymptomatic, defining surgery as the mainstay therapeutic option. This is achieved with acceptable morbidity which is limited in time, and carries very low mortality. Nonetheless, structural deformities of the airway, and complete tracheal rings in particular, may not be perfectly corrected by surgical intervention. This is illustrated by the persistence of symptoms and respiratory morbidity in some, and by subnormal pulmonary function tests long after repair in otherwise asymptomatic patients.

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