Vascular Rings and Pulmonary Artery Slings

Aseem Srivastava, Matt Harrison, and Stephanie Fuller

The term vascular ring describes a group of congenital intrathoracic vascular anomalies caused by abnormal regression and differentiation of the aortic arches during fetal development that result in compression of the trachea, esophagus, or both. Albeit rare, these vascular rings can lead to serious and at times life threatening respiratory compromise. Surgical treatment is therefore indicated in all symptomatic patients.

Classification

Vascular rings have previously been classified as complete or incomplete based upon whether the vascular encirclement around the trachea and esophagus is complete or partial. More recently, the International Congenital Heart Surgery Nomenclature and Database Committee has proposed a classification system based on the anatomic configuration of the ring [1] (Table 1).

Double aortic arches are further subgrouped based upon arch dominance (right dominant, left dominant, or balanced). Those vascular rings associated with a right arch are classified as right arch with mirror image branching, right arch with retro esophageal left subclavian artery, or circumflex aorta which denotes a right arch but a left-sided descending thoracic aorta. This can be associated with aortic arch hypoplasia.

A. Srivastava, M.D. • S. Fuller, M.D. (✉)
The Division of Cardiothoracic Surgery, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

University of Pennsylvania, Perelman School of Medicine, Philadelphia, PA, USA
e-mail: fullers@email.chop.edu

M. Harrison, M.D.
Division of Cardiology, The Children's Hospital of Philadelphia, Philadelphia, PA, USA

Embryology

In a human embryo, there are six paired pharyngeal (aortic) arches. They arise from the aortic sac and course posteriorly to connect to the right and left dorsal aorta, respectively. After a series of regression and differentiation events, this embryonic arch system ultimately gives rise to the adult aortic arch, its branches, and the pulmonary arteries. A brief understanding of this embryonic process is helpful to appreciate the anatomy of these vascular rings and for the interpretation of radiological images.

Arches 1 and 2 regress and give rise to maxillary and stapedial arteries. In humans, the fifth arch does not form to any significant degree. The third arch forms the common carotid arteries bilaterally. The sixth arch forms the left and right pulmonary arteries respectively along with the patent ductus arteriosus. Normally, the left fourth pharyngeal arch will form a portion of the adult aortic arch (between the left carotid and left subclavian arteries) whereas the right fourth arch will regress and form the right subclavian artery. A right aortic arch results when this pattern of normal regression is reversed. Similarly, persistence of both right and left fourth arches will form a double aortic arch.

Clinical Presentation and Diagnosis

Depending on the severity of compression of both the esophagus and trachea due to a vascular ring, patients may have symptoms within the first few months of life. However, it is not unusual to see patients that present with significant symptoms at a much later date, even in adulthood. Presentation is generally dependent upon the severity of obstruction based upon the "tightness" of the ring itself. Tighter lesions, like the double aortic arch and pulmonary artery slings, usually present earlier (within 3 months of age) relative to those that are loose such as aberrant right subclavian artery that typically present at greater than 6 months of age [2].

J. Lioy and S.E. Sobol (eds.), *Disorders of the Neonatal Airway: Fundamentals for Practice*,
DOI 10.1007/978-1-4939-1610-8_13,

Table 1 Classification of vascular rings

Double aortic arch
Right arch dominant
Left arch dominant
Balanced double arch
Right aortic arch
Mirror image branching
Retroesophageal left subclavian artery
Circumflex aorta
Pulmonary artery sling
Innominate artery compression

The most common symptoms from these vascular anomalies arise from the external compression of trachea and esophagus. These symptoms include inspiratory stridor, dyspnea, cough, wheezing, and dysphagia. In addition to obstruction of airflow, tracheal compression also impedes mucociliary clearance and, as a result, many patients will also suffer with recurrent respiratory tract infections [3]. Since these symptoms are shared with many other and more prevalent childhood illnesses, these patients can have a significant delay in diagnosis. Often they will receive long-term bronchodilators in an attempt to treat asthma and multiple courses of antibiotics to treat upper respiratory infections before the actual disease is accurately diagnosed. Persistence of these symptoms, especially when they do not respond to routine treatment, should alert the pediatrician of the possibility of a vascular ring. Only a high index of suspicion will ensure these patients receive early diagnosis and appropriate therapy.

Diagnosis

With the availability of more sophisticated imaging techniques, many recent publications give only historical value to the chest radiograph findings in these patients [4]. The chest X-ray is usually the first imaging study performed in a child with airway obstruction and can frequently point toward the presence of a vascular ring. Films should be assessed for the laterality of the aortic arch and for evidence of tracheal compression at the level of the arch, often better visualized on lateral films.

Absence of an aortic knuckle on the left side suggests a right arch, and when present in a child with symptoms of airway obstruction, a vascular ring should be strongly suspected. A right aortic arch may be suspected if the distal trachea is seen slightly deviated to the left instead of the right, because of the aortic arch bowing over the right main stem bronchus instead of the left. Similarly, a double aortic arch is likely if the sidedness of the arch cannot be determined on chest X-ray. Hyperinflation of the left lung with the left hilum lower than the right may suggest a pulmonary artery sling. The location of the descending aorta can usually be determined by inspecting the paraspinal line and the azygoesophageal recess.

A barium esophagogram used to be the most extensively used imaging study for the diagnosis of a vascular ring. Contrast swallow will show a posterior indentation of the esophagus in all vascular anomalies except in the presence of a pulmonary artery sling, which will produce an anterior indentation on the esophagus and an increase in the space between the esophagus and trachea at the same level.

Upon bronchoscopy, vascular rings appear as an external compression of the trachea just above the carina. Routine bronchoscopy is not necessary in a symptomatic patient with a radiologically proven vascular ring. However, it plays an essential role in the diagnostic workup of an infant with airway obstruction and an unclear cause. Bronchoscopy may also reveal additional associated airway lesions, such as tracheomalacia, complete tracheal rings, and tracheal stenosis, which may be commonly associated with vascular anomalies. In fact, vascular rings and innominate artery compression are the most common congenital cause for secondary tracheomalacia [5]. The diagnosis of innominate artery compression is almost always based on its classical findings on bronchoscopy of anterior pulsatile compression of lower trachea.

Both computed tomography (CT) and magnetic resonance imaging (MRI) are extremely useful techniques in the diagnosis of a vascular compression of the airway. The greatest advantage of these imaging modalities is their capability to completely delineate the anomaly along with their relationship to the adjacent structures. Both provide post-processed 3D images that can be very helpful in planning a surgical intervention. However, a limitation common to both these techniques is their inability to directly visualize obliterated structures. Even so, based on branching patterns, the laterality of the arch, and compression of the airway, a diagnosis of vascular ring can usually be reached. There are advantages as well as disadvantages to both modalities. We feel that CT provides excellent tracheal images at a minimal risk of ionizing radiation. MRI, on the other hand, provides a superior assessment of cardiac anatomy and function in the case that additional information is helpful. Yet, scan times are much longer thus requiring sedation and anesthesia in most infants and toddlers.

Echocardiogram can easily identify branching patterns and sidedness of the arch. It can also identify a double aortic arch when both the arches are patent. However, it is not useful to identify an obliterated lumen. It is helpful in the identification of congenital heart disease commonly associated with vascular rings such as the association between right aortic arch and tetralogy of Fallot, the incidence of which can be as high as 30 %.

Before CT and MRI were widely available, angiography was very frequently used to diagnose vascular rings. Since MRI and CT can provide essentially the same information noninvasively, a cardiac catheterization is now very infrequently utilized, unless needed for an associated cardiac disease.

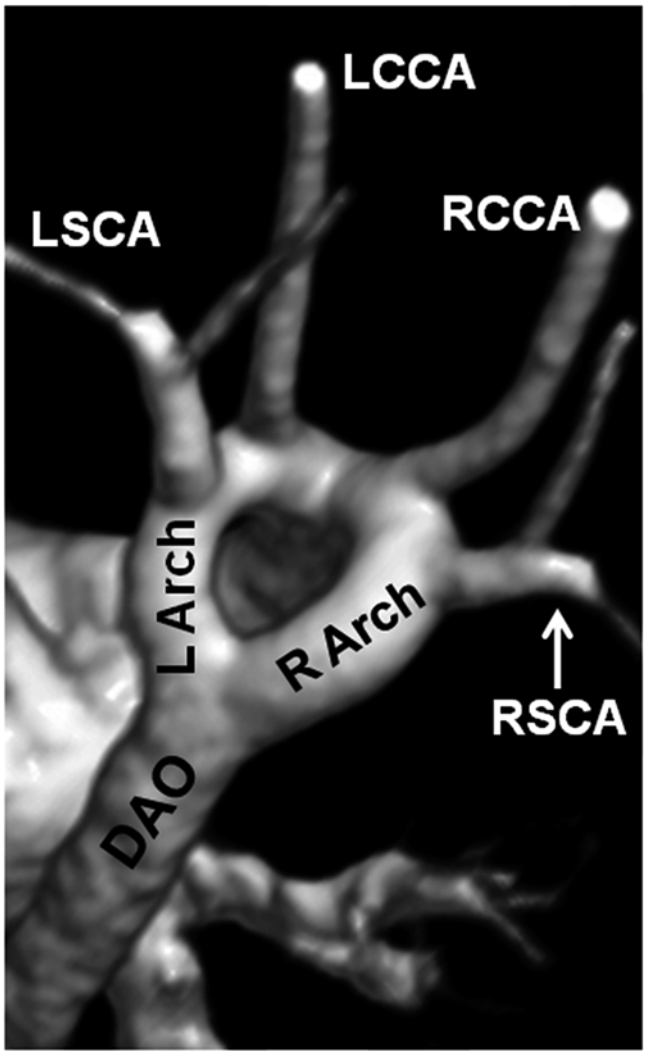

Fig. 1 Magnetic resonance angiogram of an unobstructed double aortic arch viewed from right posterior oblique with cranial angulation. Note that the right aortic arch is slightly larger. Descending Aorta (DAO), Left-Sided Arch (L Arch), Left Common Carotid Artery (LCCA), Left Subclavian Artery (LSCA), Right-Sided Arch (R Arch), Right Common Carotid Artery (RCCA), Right Subclavian Artery (RSCA)

Double Aortic Arch

This is the most common symptomatic vascular ring present in 0.05–0.3 % of the population and results from the persistence of both right and left fourth primordial arches. Two aortic arches arise from the ascending aorta and pass on either sides of trachea and esophagus to meet the descending aorta posteriorly, forming a complete ring. The right and left carotid and subclavian arteries arise from their respective arch. In almost 70 % patients, the right arch is larger (dominant), posterior, and more cephalad than the left, in about 25 % the left arch is dominant and in about 5 % patients both arches are balanced (Fig. 1).

A double aortic arch will usually cause significant narrowing and anterior bowing of thoracic trachea, and this can occasionally be seen on a lateral chest radiograph.

The surgical treatment of a double aortic arch consists of division of the lesser of the two arches.

For most patients, the approach is through a left posterolateral thoracotomy. In rare circumstances, a right thoracotomy is utilized in patients with a dominant left arch. After adequate exposure via thoracotomy, the ligamentum arteriosum, both arches, neck vessels, and descending aorta are clearly identified and both the ligamentum as well as the lesser of the two aortic arches are divided between vascular clamps, usually where the arch joins the descending aorta posteriorly. The divided ends are then oversewn using polypropylene sutures. Frequently, there will be an atretic segment in the lesser aortic arch in which case this site is then ideally suited for division.

Intraoperatively, it is useful to have pulse oximetry and/or pressure monitoring available for both upper and lower limbs. With balanced arches, blood pressure is checked in the lower limbs, while the clamps are alternately applied to each arch. The arch associated with a lesser fall in blood pressure in the lower limbs is then divided. If no appreciable difference is noted, it is preferable to divide the right arch, as its anatomical location poses a higher risk of continued compression of the trachea.

Right Arch

As previously mentioned, a right aortic arch will form if the left fourth arch regresses. Different configurations of vascular rings result depending upon the site of regression.

Right arch and retroesophageal left subclavian artery. This is the most common (65 %) type of right arch seen [10] and results from the involution of the left fourth arch between the left common carotid and left subclavian arteries. As a result, the left subclavian now arises from the descending aorta and passes behind the esophagus to reach the left arm. The ligamentum connects the descending aorta to the left pulmonary artery and, in doing so, completes the ring.

Occasionally, the origin of the aberrant left subclavian will have a bulb like dilation, known as Kommerell's diverticulum (Fig. 2).

This represents the remnant of fourth embryonic arch. When present, it implies a left ligamentum arteriosum and denotes the presence of a ring. Additionally, Kommerell's diverticulum may dilate over time and can compress the trachea and/or esophagus, leading to a recurrence of symptoms.

Surgical approach to these patients is again through a left posterolateral thoracotomy. With careful dissection, the ligamentum, aortic arch, brachiocephalic vessels, and descend-

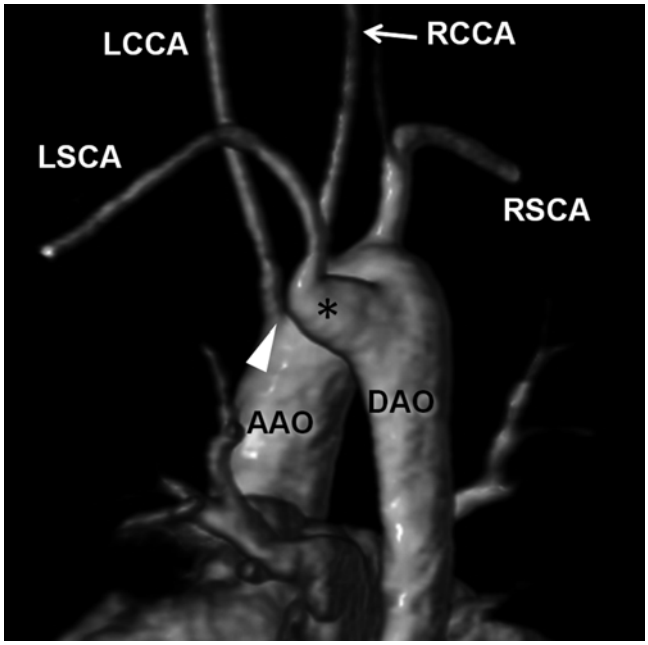

Fig. 2 Magnetic resonance angiogram viewed from left posterior oblique demonstrates a right aortic arch with an aberrant left subclavian artery originating from a retroesophageal diverticulum (diverticulum of Kommerell). The location of the left-sided ligamentum arteriosum (*arrowhead*), though not directly observed, is implied by the regionally dilated proximal left subclavian artery (*) completing the vascular ring. Aberrant Left Subclavian Artery (ALSCA), Aberrant Right Subclavian Artery (ARSCA), Descending Aorta (DAO), Left Common Carotid Artery (LCCA), Right Common Carotid Artery (RCCA)

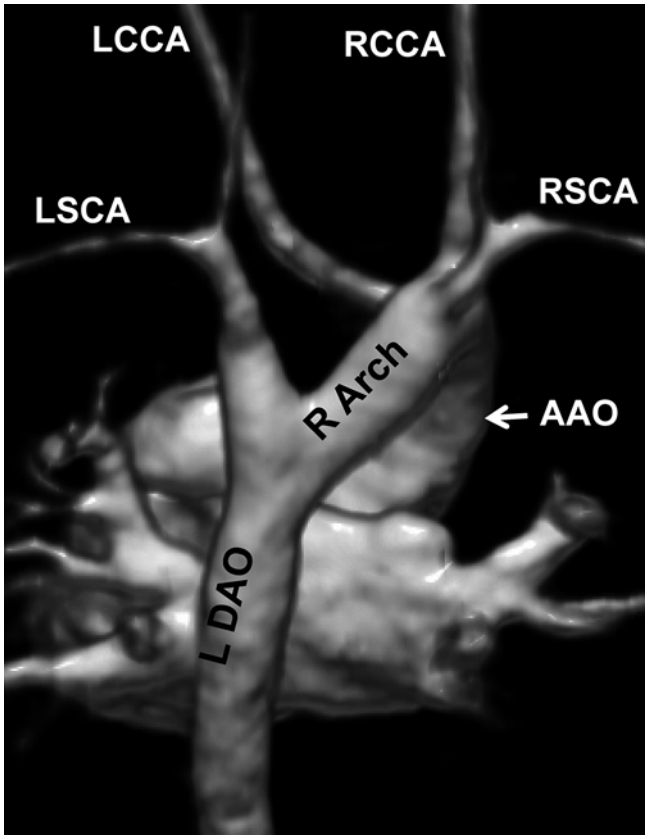

Fig. 3 Magnetic resonance angiogram from a direct posterior view demonstrates a right aortic arch with left-sided descending aorta (circumflex aortic arch). The combination of right arch and left-sided thoracic descending aorta implies a left ligamentum which completes the ring. Left-Sided Descending Aorta (L DAO), Left Common Carotid Artery (LCCA), Left Subclavian Artery (LSCA), Right-Sided Arch (R Arch), Right Common Carotid Artery (RCCA), Right Subclavian Artery (RSCA)

ing aorta are clearly identified. The ligamentum is then divided and its stumps are oversewn.

When a Kommerell's diverticulum is present, controversy exists whether or not it should be resected, or whether the left subclavian artery should be transferred to the left carotid artery. Division of the ligamentum and pexy of the diverticulum may be associated with late dilation of the diverticulum and recurrence of symptoms [6–8].

Right arch mirror image branching. A right aortic arch with mirror image branching occurs when the left fourth arch regresses between the subclavian artery and the descending aorta. As a result, the branching pattern resembles a "mirror image" of the normal left arch. A vascular ring may form if the ligamentum arises from the descending thoracic aorta. However, it most commonly originates from the innominate artery and therefore does not form a compressive vascular ring.

Right aortic arch with left descending aorta (circumflex aorta). This is a rare variant of right aortic arch, where the transverse arch courses posterior to the esophagus and continues as a left descending aorta. A vascular ring is formed in the setting of a left ligamentum (Figs. 3 and 4).

Optimal surgical treatment for this anomaly is known as an "Aortic Uncrossing" and is a more complicated operation, requiring a median sternotomy, the use of cardiopulmonary bypass and deep hypothermic circulatory arrest. The arch along with its branches is extensively mobilized and then divided at a point where it hooks around the esophagus. The proximal end of the arch may be oversewn while the distal retro esophageal segment is brought anteriorly and anastomosed to a counter-incision on the left lateral side of the ascending aorta. Alternatively, both sides of the arch are mobilized anterior to the trachea with great care not to injure both recurrent laryngeal nerves. This lesion may occur in conjunction with severe long segment coarctation of the retro esophageal and descending aorta. In these rare cases, the coarctation requires augmentation using a patch material.

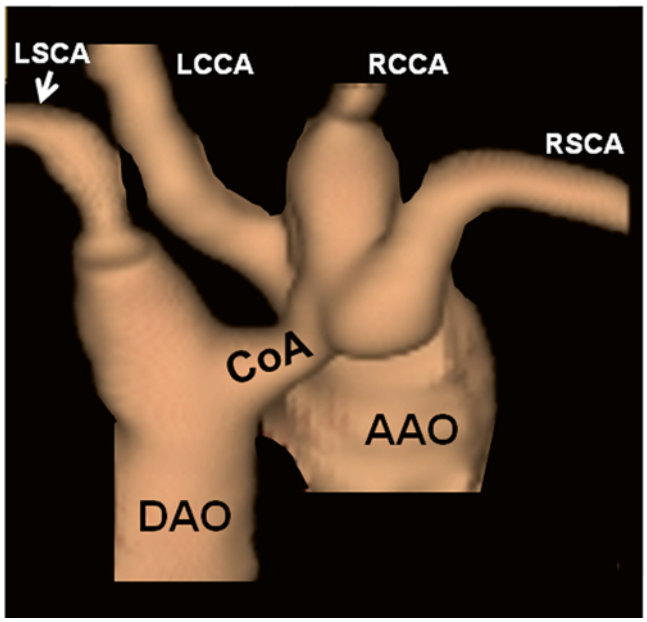

Fig. 4 Magnetic resonance angiogram from a direct posterior view demonstrates a right aortic arch with left-sided descending aorta. The combination of right arch and left-sided thoracic descending aorta implies a left ligamentum which completes the ring. In this particular patient there is also the rare presence of coarctation of the right-sided aortic arch

Pulmonary Artery Sling

This is a relatively uncommon vascular anomaly in which the left pulmonary artery arises extra-pericardially, from the posterior aspect of the right pulmonary artery. It then hooks over the right bronchus and passes between the trachea and esophagus to reach the left pulmonary hilum (Fig. 5).

With this configuration, the left pulmonary artery compresses the lower trachea leading to severe stenosis [9]. Additionally, this anomaly is also strongly associated with tracheal stenosis, due to the presence of complete tracheal rings, in almost 50–65 % patients (known as the ring-sling complex). The tracheal cartilage forms a complete circle with an absence of membranous trachea.

Repair of a pulmonary artery sling requires a median sternotomy and cardiopulmonary bypass [10]. The ductus or the ligamentum is divided. The left pulmonary artery is dissected circumferentially from its origin on the right pulmonary artery to its left hilar branches. It is then divided between clamps at its origin from the right pulmonary artery. It is then brought anterior to the bronchus, and anastomosed to a counter-incision on the main pulmonary artery with great care not to kink the vessel at its neo-origin.

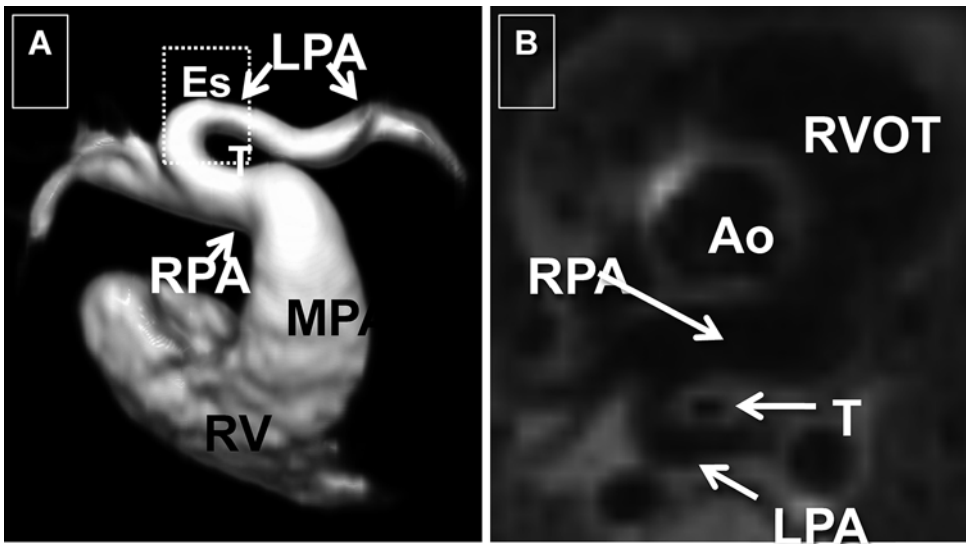

Fig. 5 (**a**) Magnetic resonance angiogram demonstrating a Pulmonary Sling. In this disease, the left pulmonary artery originates from the proximal right pulmonary artery and then passes between the relatively anterior trachea and the relatively posterior esophagus as it heads back toward the left lung. This creates a sling around the trachea with a leftward pull typically producing severe tracheal symptoms. In contrast, vascular rings encompass both the trachea and the esophagus and therefore may cause both stridor and dysphagia. (**b**) Spin Echo Dark Blood Image demonstrating tracheal narrowing at the level where the left pulmonary artery creates a sling around the trachea. Aorta (Ao), Esophagus (Es), Left Pulmonary Artery (LPA), Right Pulmonary Artery (RPA), Right Ventricle Outflow Tract (RV), Trachea (T)

In the presence of complete tracheal rings, a concomitant repair of the trachea (slide tracheoplasty) must be performed.

Innominate Artery Compression

This results from an innominate artery that arises a more distal on the aortic arch than normal and compresses the lower end of trachea as it crosses it anteriorly from left to right (Fig. 6).

The incidence of this anomaly seems to be relatively high. However, only a fraction of these patients are symptomatic, leading to a lot of debate on the optimal management of these patients.

For those symptomatic patients with respiratory distress, persistent stridor and reflex apnea should have an aggressive diagnostic workup followed by aortopexy [11].

The diagnosis is usually made on bronchoscopy by the presence of a pulsatile anterior narrowing of the lower trachea on a child with symptoms of airway obstruction. Additional radiological imaging (CT, MRI) may further support the diagnosis.

Aortopexy, in which the innominate artery and proximal arch are suspended with sutures, to the posterior surface of the sternum, is a simple and effective procedure for the treatment of this malformation. Surgical approach is typically through a small right anterior thoracotomy in the second intercostal space. The right lobe of thymus is resected, taking care to avoid injury to the phrenic nerve. The proximal innominate artery and arch adjacent to its origin are suspended to the posterior sternal periosteum using 3–4 pledgeted mattress sutures that are tied down, at times under direct bronchoscopic guidance. If needed, these sutures can also be passed trans-sternally, and tied on the anterior surface of the sternum. It is important, not to dissect the space between innominate artery and the trachea, as this fibrous tissue will necessary to effectively pexy the anterior wall of trachea.

Left Aortic Arch and Aberrant Right Subclavian

When the right fourth pharyngeal arch regresses between the subclavian and the carotid arteries, the right subclavian artery will arise "aberrantly" from the descending aorta. The incidence of this anomaly ranges between 0.5 and 1.8 %. The majority of these individuals will remain asymptomatic, and only rarely when the ligamentum is right sided will a vascular ring form.

Postoperative Care

Most patients have a relatively straightforward postoperative course after surgical repair. Early extubation is recommended except in those with a concomitant procedure of the trachea or tracheomalacia. Therapies that may prove useful in the early postoperative period include oxygen, humidity, inhaled steroids, bronchodilators, and aggressive early chest physiotherapy. Vocal cord paresis and/or paralysis are a known complication of surgical repair and should be watched for in the post op. Persistent airway obstruction after surgical repair can be due to residual compression, tracheomalacia, or intrinsic lesions of the airway.

Results

Long-term results of surgical repair of vascular rings are generally good, with no reported operative mortality and a very low risk of recurrence as highlighted in many recent publications [11]. Of note, complete resolution of symptoms may not always be evident immediately after surgery, and this is commonly due to an associated tracheal lesion (malacia), and

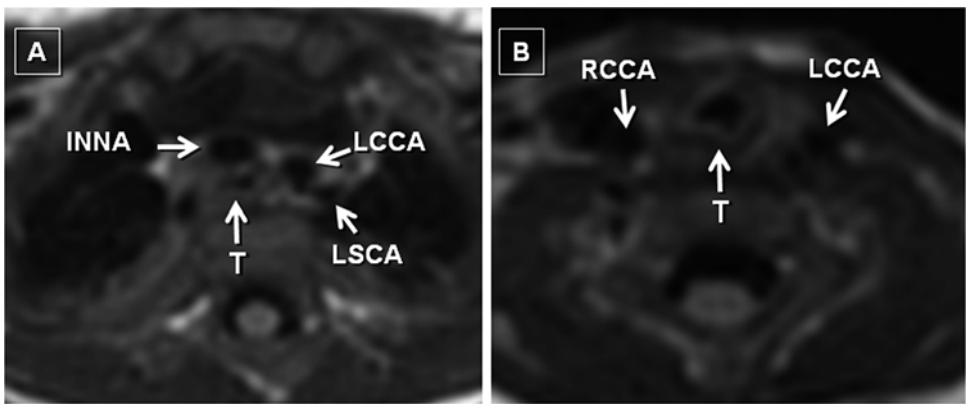

Fig. 6 (**a**) Turbo spin echo dark blood image in transverse view demonstrates severe tracheal flattening at the point where the innominate artery crosses anterior to the trachea. Bronchoscopy demonstrated tracheomalacia of the proximal trachea associated with an anterior pulsating mass. These findings are consistent with innominate artery compression of the trachea. (**b**) More superiorly above the thoracic inlet, the trachea appears round and patent

improves over time. By 1 year, almost 70 % of all patients are expected to be symptom free [7, 12], and most of the remaining patients will have experienced a substantial improvement in symptoms.

Multidisciplinary Considerations

While surgical repair is performed by the cardiothoracic surgeon, management of vascular anomalies often involves multidisciplinary input from neonatology, otolaryngology, pulmonology, and cardiology.

Future Considerations

Over the past few years, there has been a push to expand the role of minimally invasive cardiac surgery in pediatric patients. In 1995, based on an experience with video-assisted interruption of the patent ductus arteriosus, Burke et al. [13] reported their initial results with video-assisted vascular ring division, and recently, robotically assisted vascular ring division represents a further step in this direction [14]. By avoiding muscle division and rib spreading, these minimally invasive operations decrease post-thoracotomy pain, tearing of intercostal ligaments, and may also improve early postoperative respiratory function, in addition to better cosmetic results. However, operative times are longer, conversion rates are still significant, along with a much higher procedural cost.

Importantly, the use of these minimally invasive procedures is limited to relatively bigger kids, and for non-patent (atretic) vascular rings. Recent advances in thoracoscopic and robotic instruments have undoubtedly contributed to an increasing use of these techniques. In the present, however, the use of this technology for surgical correction of vascular rings is very limited and its future application will depend largely on follow-up studies, further advances, and improved cost-effectiveness.

References

1. Backer CL, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: vascular rings, tracheal stenosis, pectus excavatum. Ann Thorac Surg. 2000;69:S308–18.
2. Smith RJ, Smith MC, Glossop LP, Bailey CM, Evans JN. Congenital vascular anomalies causing tracheoesophageal compression. Arch Otolaryngol. 1984;110:82–7.
3. McLaughlin Jr RB, Wetmore RF, Tavill MA, Gaynor JW, Spray TL. Vascular anomalies causing symptomatic tracheobronchial compression. Laryngoscope. 1999;109:312–9.
4. McLaren CA, Elliott MJ, Roebuck DJ. Vascular compression of the airway in children. Paediatr Respir Rev. 2008;9:85–94.
5. Thompson DM, Willging JP, Cotton RT. Lesions of the larynx, trachea and upper airway. In: Coran AG, editor. Pediatric surgery, vol. 2. New York: Elsevier; 2012. p. 837–54.
6. Ruzmetov M, Vijay P, Rodefeld MD, Turrentine MW, Brown JW. Follow-up of surgical correction of aortic arch anomalies causing tracheoesophageal compression: a 38-year single institution experience. J Pediatr Surg. 2009;44:1328–32.
7. Backer CL, Russell HM, Wurlitzer KC, Rastatter JC, Rigsby CK. Primary resection of Kommerell diverticulum adn left subclavian artery transfer. Ann Thorac Surg. 2012;94:1612–7.
8. Ota T, Okada K, Takanashi S, Yamamoto S, Okita Y. Surgical strategy for Kommerell's diverticulum. J Thorac Cardiovasc Surg. 2013;131:574–8.
9. Sailer R, et al. Pulmonary artery sling associated with tracheobronchial malformations. Arch Otolaryngol Head Neck Surg. 1992;118:864–7.
10. Fiore AC, Brown JW, Weber TR, Turrentine MW. Surgical treatment of pulmonary artery sling and tracheal stenosis. Ann Thorac Surg. 2005;79:38–46.
11. Gardella C, et al. Tracheal compression by aberrant innominate artery: clinical presentations in infants and children, indications for surgical correction by aortopexy, and short- and long-term outcome. J Pediatr Surg. 2010;45:564–73.
12. Woods RK, Sharp RJ, Holcomb III GW, Snyder CL, Lofland GK, Ashcraft KW, Holder TM. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg. 2001;72(2):434–8.
13. Burke RP, Rosenfeld HM, Wernovsky G, Jonas RA. Video-assisted thoracoscopic vascular ring division in infants and children. J Am Coll Cardiol. 1995;25(4):943–7.
14. Suematsu Y, Mora BN, Mihaljevic T, del Nido PJ. Totally endoscopic robotic-assisted repair of patent ductus arteriosus and vascular ring in children. Ann Thorac Surg. 2005;80(6):2309–13.