

Vascular Rings

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Vascular rings are a group of aortic arch anomalies (occasionally involving the PA) that can result in tracheoesophageal compression. By definition, true (“complete”) vascular rings consist of vascular structures that entirely surround the trachea and the esophagus. The 2 most common variants are the double aortic arch and the right aortic arch with aberrant left subclavian artery (arising from a diverticulum of Kommerell) and a left ligamentum arteriosum (or ductus arteriosus, if patent) (Figure 29-1). Other potential causes of vascular tracheobronchial compression such as innominate artery syndrome (anterior compression of the trachea by the innominate artery) and PA sling (origin of the left PA from the right PA and subsequent course between the trachea and esophagus, Figure 29-1, C) are not truly vascular rings but are sometimes called “incomplete” vascular rings. PA slings may be associated with complete tracheal rings causing further tracheal obstruction.

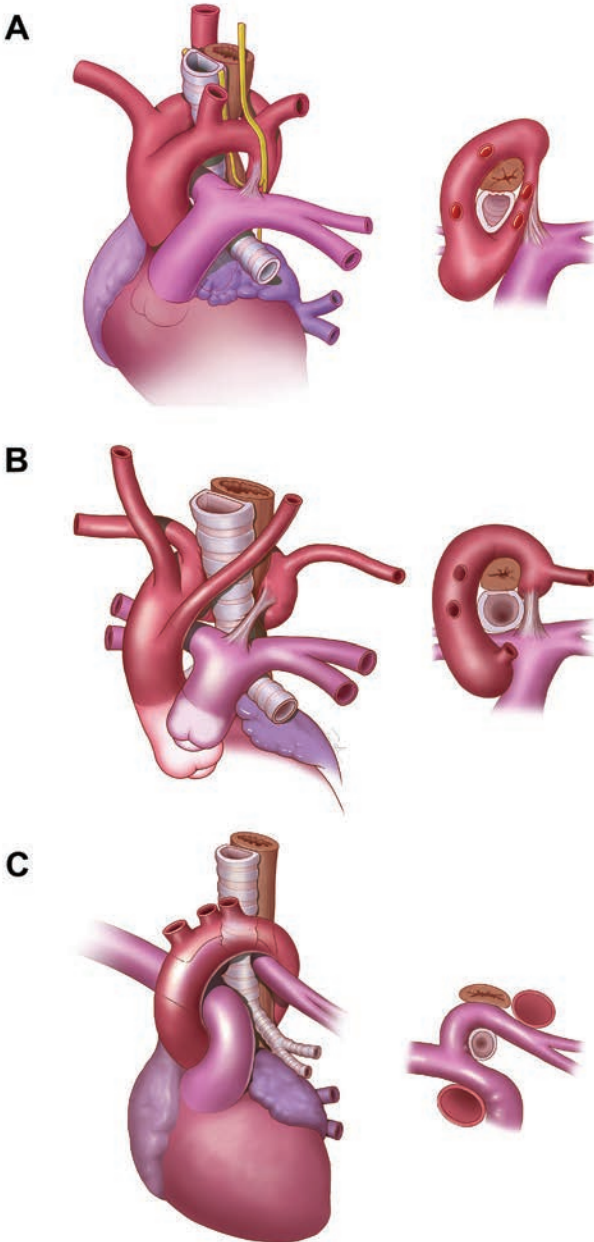
Pathophysiology, Clinical Presentation, and Associated Abnormalities

Vascular rings occur when there is abnormal persistence or regression of the various components of the embryonic totipotential arch. The location and severity of tracheoesophageal compression determines the timing of presentation. With the exception of critical airway obstruction, symptoms are rare in neonates. Those with a double aortic arch may present earlier, during the first year of life. Many patients are asymptomatic, but over time may develop stridor, cough, increasing distress with intercurrent respiratory illnesses, and reflux or dysphagia with the introduction of solids in the diet. Some children may be noted to favor nonsolid foodstuff. Dysphagia is more common in older children and adolescents.

Vascular rings may be associated with cardiac anomalies such as tetralogy of Fallot, VSD, and coarctation of the aorta. Noncardiac anomalies include tracheoesophageal fistulae, and cleft lip and palate. Children with DiGeorge and Down syndromes have a higher incidence of vascular rings.

Diagnosis

- **Fetal echocardiogram.** Color Doppler imaging may reveal vascular structures around the trachea during cephalad transducer sweeps from a three-vessel view.
- **CXR (Figure 29-2).** May be shown as right aortic arch indentation on the distal trachea and anterior bowing of trachea on lateral view due to the retroesophageal vessel (aberrant subclavian artery).
- **Barium esophagram (Figure 29-3).** Not necessary but may show a posterior indentation on the esophagus from an aberrant subclavian artery or anterior indentation from a pulmonary sling.
- **CTA/MRI (Figure 29-4).** Definitive diagnosis of vascular ring requires a CTA or MRI. CTA is the preferred modality due to fast acquisition times without the need



for sedation, superior resolution, and superior airway information compared to MRI. It allows for detailed 3D reconstructions and characterization of the type of vascular ring that assist with surgical planning. The differentiation of a right aortic arch with aberrant left subclavian artery from a double aortic arch with an atretic left arch can be challenging (Adachi et al. 2011).

Indications/Timing for Intervention

The presence of a vascular ring alone is not necessarily an indication for operation. In general, intervention is warranted if the patient has symptoms attributable to the vascular ring (dysphagia or airway obstruction) and characteristic imaging studies (e.g., esophagram or CTA). Intervention may be considered for asymptomatic patients that show significant compression on imaging studies since the risk of intervention is relatively low. It is important to rule out the presence of significant cardiac anomalies necessitating intracardiac repair.

Figure 29-1. Vascular rings and other vascular causes of tracheobronchial compression. A) Double aortic arch. B) Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum. C) Left PA sling.

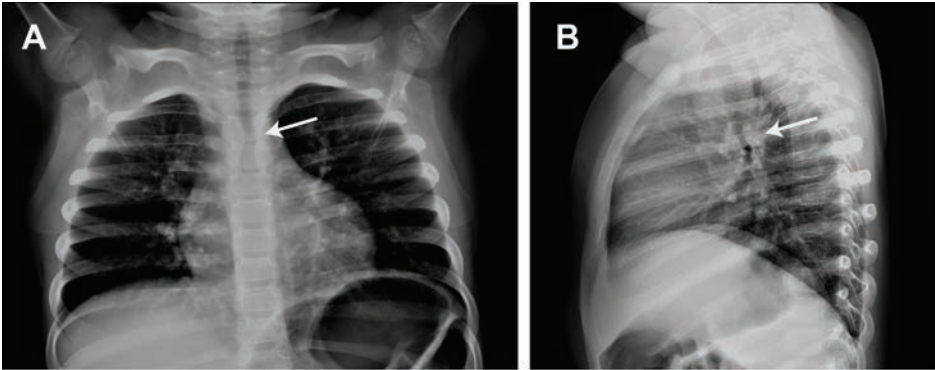


Figure 29-2. Anteroposterior (A) and lateral (B) views of a CXR of a patient with a vascular ring (right aortic arch with aberrant left subclavian artery), showing tracheal compression (arrow).

subclavian artery (i.e., dysphagia lusoria) is not a vascular ring and very rarely (if ever) requires intervention.

Surgical Repair

The type of surgical intervention depends on the type of vascular ring. Most procedures are performed through a left posterolateral thoracotomy through the 4th intercostal space. The goal of the operation is to completely relieve the tracheoesophageal compression. Patients may benefit from a direct laryngoscopy and bronchoscopy (DL&B) by ENT at the beginning of the procedure and sometimes a flexible bronchoscopy at the end to confirm relief of tracheobronchial compression. The surgical procedures for the two most common types of complete rings are:

- **Right aortic arch with aberrant left subclavian artery.** The left ligamentum arteriosum (between the origin of the left subclavian artery and the left PA) is divided, therefore releasing the ring. The descending aorta may be fixed posteriorly to the spine (i.e., aortopexy) to increase the space clearance. If there is a large diverticulum of Kommerell at the base of the left subclavian artery or if simple division of the ligamentum fails to create a wide-enough space, the diverticulum is excised and the left subclavian artery is translocated to the left carotid artery (Figure 29-5).
- **Double aortic arch.** The atretic or smallest portion of the double arch (most commonly the posterior left arch, just distal to the origin of the left subclavian artery) is divided and oversewn. It is important to also divide the ligamentum arteriosum that travels from this portion of the arch to the left PA in order not to leave a residual ring.

A circumflex aortic arch (an aortic arch that travels on one side of the trachea, crosses behind the trachea and esophagus, and descends on the other side) poses a difficult challenge. Patients with a right aortic arch, aberrant left subclavian artery, and a circumflex arch (left descending aorta) may benefit from a simple repair of the vascular ring (division of the left ligamentum arteriosum and translocation of the left



Figure 29-3. Esophagram of a patient with a vascular ring (right aortic arch with aberrant left subclavian artery) showing posterior indentation of the esophagus.

subclavian artery to the left carotid artery). In some instances, patients may require a more complicated procedure (e.g., aortic uncrossing procedure) through a median sternotomy (Backer et al. 2016).

Anesthetic Considerations

Preoperatively, anesthetic management should begin with a thorough chart review of radiologic images defining the anatomy of the vascular ring. Clear communication is needed between the surgical and anesthetic teams with regards to lung isolation, regional anesthesia, and invasive monitoring lines, as occlusion of blood vessels are likely included in the surgical plan. Most children will not be large enough to accommodate double-lumen tubes for lung isolation. However, there are other alternative

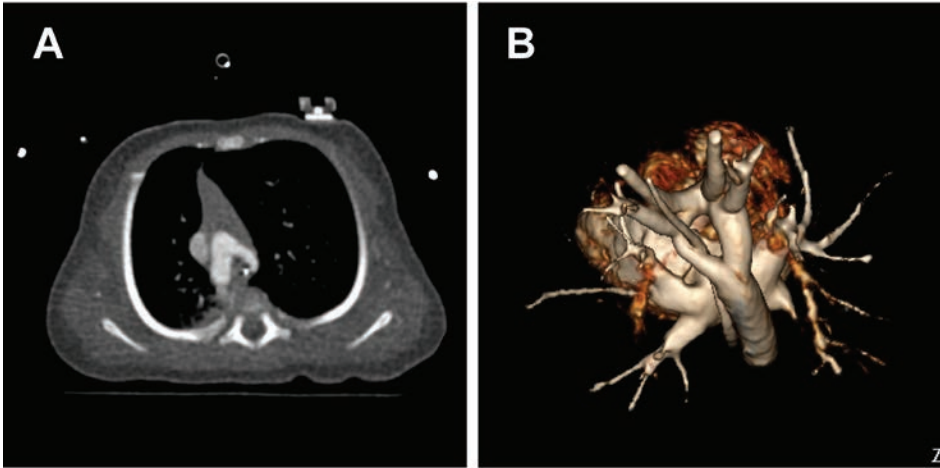


Figure 29-4. CTA of a patient with a double aortic arch with axial cuts (A) and 3D reconstruction (B). The right arch is dominant, compared to the left arch.

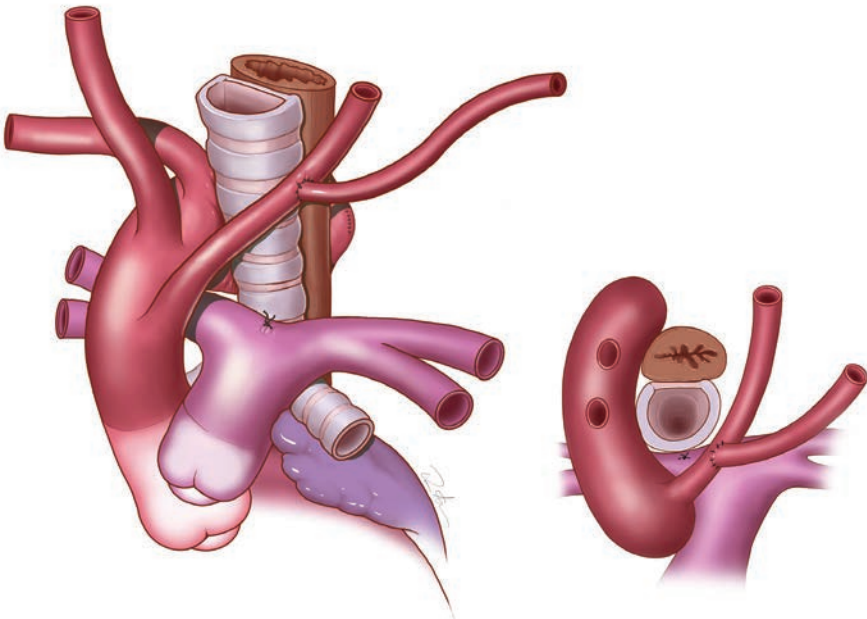


Figure 29-5. Surgical repair of a right aortic arch with an aberrant left subclavian artery, left ligamentum arteriosum, and a prominent diverticulum of Kommerell consisting of division of the ligamentum, excision of the diverticulum, and translocation of the left subclavian artery to the left carotid artery.

TCH experience with vascular rings (1996-2018)

(Binsalamah et al. 2019)

Number of patients: 148

Median age: 1 year (interquartile range 0.4-5.2 years)

Median weight: 12.8 kg (interquartile range: 7.5-26.5 kg)

Types of ring:

- Double aortic arch: 72 (49%)
- Right aortic arch with aberrant left subclavian artery: 69 (47%)
- Others: 7 (5%)

Perioperative mortality: 0

Complications:

- Chylothorax: 12%
- Vocal cord paresis: 1 (<1%)

techniques for pediatric lung isolation (Hammer et al. 1999), including the use of bronchial blockers and right-mainstem intubation.

Tracheal compression by the vascular ring seldom precludes endotracheal tube placement or easy bag-mask ventilation during inhalational induction. However, the use of orogastric or nasogastric tubes is discouraged, especially in patients with tight vascular rings, to avoid esophageal perforation. If adequate analgesia is obtained, early extubation may be considered, as cardiac dysfunction and bleeding are rarely encountered in this surgery.

Postoperative Management

- **Fluids.** 100% maintenance with D5%/0.45% NS is standard. Large volume fluid resuscitation is seldom necessary.
- **Analgesia and sedation.** The use of regional anesthesia, including intercostal and paravertebral peripheral nerve blocks and thoracic epidurals, may be utilized for supplemental postoperative pain control. The Acute Pain Service will assist with co-managing analgesia if a catheter is placed. A PCA with continuous and intermittent opioid dosing is commonly utilized with scheduled adjunctives such as acetaminophen and ketorolac. Adequate analgesia is key to enable adequate airway clearance.
- **Mechanical ventilation.** Most patients are extubated in the OR, or considered for expedited extubation, if otherwise hemodynamically stable. Postoperative stridor, typically treated with steroids, is relatively common. Retractions or “seal-bark” cough may be present due to residual tracheobronchomalacia. Depending on the level of residual bronchomalacia, patients may need non-invasive positive mechanical ventilation (NIMV) after extubation. In some instances, even mild residual tracheobronchomalacia in association with postsurgical chest wall pain may need NIMV temporarily. Steroids may play a role when there is secondary inflammation.
- **Nutrition.** Clear fluids can be started 4 hours postextubation if the patient is otherwise hemodynamically stable. This may be advanced to a regular diet in the absence of postoperative dysphagia. Chest tubes should be monitored for chylous output, particularly when fat-containing food is introduced.

Complications

- **Neuropathy.** The vagus, recurrent laryngeal, and phrenic nerves are close to the area of surgical dissection. The recurrent laryngeal nerve may be particularly prone to injury, often times transient.
- **Chylothorax.** Complication due to the presence of large lymphatic nodes and channels in the area.

Suggested Readings

Adachi I, Krishnamurthy R, Morales DL. A double aortic arch mimicking a right aortic arch with an aberrant subclavian artery. *J Vasc Surg* 2011;54:1151-1153.

Backer CL, Mongé MC, Popescu AR, et al. Vascular rings. *Semin Pediatr Surg* 2016;25:165-175.

Binsalamah ZM, Ibarra C, John R, et al. Contemporary mid-term outcomes in pediatric patients undergoing vascular ring repair. Presented at the 55th Annual Meeting of the Society of Thoracic Surgeons, San Diego, CA, 2019.

Hammer GB, Fitzmaurice BG, Brodsky JB. Methods for single-lung ventilation in pediatric patients. *Anesth Analg* 1999;89:1426-1429.

Ramos-Duran L, Nance JW, Schoepf UJ, et al. Developmental aortic arch anomalies in infants and children assessed with CT Angiography *AJR Am J Roentgenol* 2012;198:W466-W474.