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Vascular rings

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ABSTRACT

The term vascular ring refers to congenital vascular anomalies of the aortic arch system that compress the esophagus and trachea, causing symptoms related to those two structures. The most common vascular rings are double aortic arch and right aortic arch with left ligamentum. Pulmonary artery sling is rare and these patients need to be carefully evaluated for frequently associated tracheal stenosis. Another cause of tracheal compression occurring only in infants is the innominate artery compression syndrome. In the current era, the diagnosis of a vascular ring is best established by CT imaging that can accurately delineate the anatomy of the vascular ring and associated tracheal pathology. For patients with a right aortic arch there recently has been an increased recognition of a structure called a Kommerell diverticulum which may require resection and transfer of the left subclavian artery to the left carotid artery. A very rare vascular ring is the circumflex aorta that is now treated with the aortic uncrossing operation. Patients with vascular rings should all have an echocardiogram because of the incidence of associated congenital heart disease. We also recommend bronchoscopy to assess for additional tracheal pathology and provide an assessment of the degree of tracheomalacia and bronchomalacia. The outcomes of surgical intervention are excellent and most patients have complete resolution of symptoms over a period of time.

Introduction

The first surgical repair of a vascular ring was performed by Dr. Robert Gross[1] from Boston Children's Hospital. He first described a vascular ring diagnosed at autopsy: "A ring of blood vessels was found encircling the intrathoracic portion of the esophagus and trachea…. The pathologic findings at once suggested that a division of some part of the vascular ring during life would have relieved the pressure on the constricted trachea and esophagus." Gross made this observation and then applied the interpretation clinically to successfully divide a double aortic arch in a 1-year-old child with persistent wheezing and recurrent hospital admissions for serious upper respiratory tract infections.

The term vascular ring is now loosely used to refer to all congenital vascular anomalies of the aortic arch system that cause compression of the trachea and/or esophagus. The symptoms relative to these two structures are primarily noisy breathing, cough, and dysphagia. An example of a CT image of a child with a double aortic arch is shown in Figure 1.

Gross[1] was also the first to describe the other type of commonly seen true vascular ring, which is a right aortic arch with an aberrant or retroesophageal left subclavian artery and a left ligamentum arteriosum. It is in this group that we have recently recognized that the Kommerell diverticulum that serves as the source of blood flow to the left subclavian artery, may be an independent contributor to the compression of the trachea and esophagus and require removal.

At our institution Dr. Willis J. Potts[2] reported the first successful vascular ring repair (double aortic arch) in 1948. Potts also was the first surgeon to report successful repair of the anomaly now known as pulmonary artery sling.[3] This is a rare vascular anomaly where the left pulmonary artery originates from the right pulmonary artery. The left pulmonary artery encircles the distal trachea as it travels to the left lung. This compresses the distal trachea and right mainstem bronchus. The classification scheme that we have

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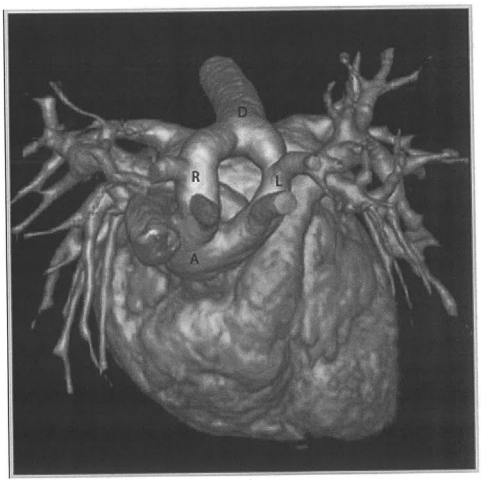

Fig. 1. Computed tomogram (3D reconstruction) of a child with a double aortic arch with balanced right and left arches. A, ascending aorta; D, descending aorta; L/R, left/right arch. (Reproduced with permission from Backer CL, Popescu AR, Rastatter JC, Russell HM. Vascular rings and slings. In: daCruz E, Ivy DD, Jaggers J, eds. *Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care*. London: Springer-Verlag; 2014, vol. 4, 2219–2238.)

used at Ann & Robert H. Lurie Children's Hospital of Chicago for vascular rings is that endorsed by the Society of Thoracic Surgeons' Congenital Nomenclature and Database Project.[4] The surgical experience with vascular rings at Lurie Children's is illustrated in Table 1 (1947–2015; DAA; RAA; PAS). This article will review the clinical presentation and diagnosis, embryology, indications for surgery, surgical techniques for the different vascular rings, and the outcomes of surgical intervention.

Clinical presentation and diagnosis

The classic clinical presentation of a child with a vascular ring is noisy breathing and a barky cough. Many clinicians have compared the bark to that of a seal, hence the phrase "seal-bark cough." Other frequent symptoms are recurrent upper respiratory tract infections, wheezing, dyspnea on exertion, and dysphagia (Table 2). Some infants have apparent life-threatening events (ALTE) or apnea. In rare cases, children with severe compression and tracheomalacia may develop respiratory distress requiring intubation. Dysphagia is not usually a symptom until the child is old enough to take solid foods. Children who are taking breast milk or formula usually have no difficulty with swallowing, despite a tight vascular ring. The careful clinician can elicit these symptoms even when the patient is referred as being "asymptomatic." Many older children have learned to chew their food very carefully and the parents will note that they are always the last to leave the dinner table. A careful history in an older patient may also reveal that as an infant the child had episodes of croup, was diagnosed with "asthma," was treated with inhalers, and had other respiratory issues which may not have risen to the level of being thought to be "symptoms."

Table 1
Lurie Children's Hospital experience 1947–2015.

Vascular ring	No. of patients
Double aortic arch	155
Right aortic arch/left ligamentum	172
Pulmonary artery sling	46
Total	373

Table 2
Symptoms leading to clinical presentation in patients with vascular rings.[a]

	Double aortic arch (n = 80)[b]	Right aortic arch (n = 78)[b]
Stridor	46 (57%)	18 (23%)
Recurrent upper respiratory tract infections	22 (27%)	18 (23%)
Cough	17 (21%)	8 (10%)
Dysphagia	12 (15%)	12 (15%)
Respiratory distress	8 (10%)	13 (17%)
Ventilator preoperatively	7 (9%)	3 (4%)

Reproduced with permission from Backer et al[5].

[a] More than one symptom occurred in many patients.

[b] Our records did not provide symptoms for the earlier patients in the series.

The diagnosis of a child with vascular ring has changed substantially in the last 15 years. However, a key point is that the diagnostic evaluation should proceed in a stepwise fashion without obtaining excessive studies. Once the diagnosis has been made and the surgeon is comfortable with the diagnosis and knows exactly what operation to proceed with, it is unnecessary to obtain further imaging studies.

The first diagnostic study is the chest radiograph. Careful analysis of the radiograph can usually show the location of the aortic arch. In patients without a vascular ring the knob of the left aortic arch can usually be appreciated. In patients with a double aortic arch, it actually may be difficult to tell which side the arch is on (this is a clue!). If a patient has a right aortic arch, the compression on the right side of the trachea usually can be visualized. Although a barium esophagram historically was the study used to diagnose vascular ring, in the current era we and others prefer CT imaging.[5] With the new generation dual source CT scans, the evaluation can be completed in less than 1 s without the need for intubation and with a considerable reduction in radiation dose. The information obtained from a CT scan would allow for precise surgical planning of the surgical strategy.[6] Some centers prefer magnetic resonance imaging (MRI) but this requires a longer period of time to obtain and often requires sedation and intubation. MRI currently does not give as clear a picture of the tracheal anatomy and lumen as does CT imaging. Figure 2 is a CT image of a double aortic arch and Figure 3 is a CT image of a right

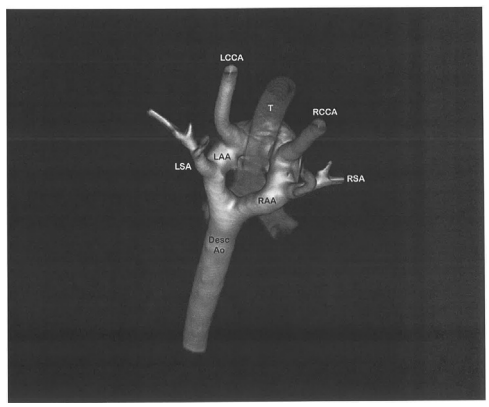

Fig. 2. Contrast-enhanced CT angiogram with 3D multiplanar reformat of a child with a double aortic arch. This shows a posterior view of a child with balanced aortic arches. Desc Ao, descending aorta; LAA, left aortic arch; LCCA, left common carotid artery; LSA, left subclavian artery; RAA, right aortic arch; RCCA, right common carotid artery; RSA, right subclavian artery; T, trachea. (Reproduced with permission from Backer CL, Popescu AR, Rastatter JC, Russell HM. Vascular rings and slings. In: daCruz E, Ivy DD, Jaggers J, eds. *Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care*. London: Springer-Verlag; 2014, vol. 4, 2219–2238.)