Editorial

Vascular Rings, Slings, and Tracheal Rings

The phrase “vascular ring” was coined by Robert E. Gross in 1945. This landmark article described Gross’ experience with the diagnosis and successful surgical management of an infant with a double aortic arch. Gross had performed an autopsy 14 years earlier on a 5-month-old baby who had had wheezing respiration since birth and difficulty with swallowing. At the time of autopsy, a “ring” of blood vessels was found that encircled the intrathoracic portion of the esophagus and trachea in such a way that, the esophagus was indented from behind, whereas the trachea was compressed on its anterior surface. The pathological findings at once suggested that a division of some part of the so-called “vascular ring” during life would probably have relieved the pressure on the constricted esophagus and trachea.

The operation Gross performed was on a 1-year-old child who had had multiple hospitalizations for complaints of wheezing respirations, cough, respiratory distress, fever, and stridor. Vascular ring was diagnosed on the basis of a barium esophagogram and a tracheogram. Through a left anterolateral thoracotomy, a diagnosis of a double aortic arch was confirmed, and the smaller anterior (left) arch was doubly ligated and divided. The child recovered satisfactorily and had no respiratory difficulties at the time of dismissal from the hospital.

In the same article, Gross described the two types of true (complete) vascular rings. In the first type, the aortic arch is missing from its normal position in front of the trachea, and instead courses behind the esophagus. The pulmonary artery lies in a normal position in front of the trachea, but it is anchored to the distal part of the aortic arch by a patent ductus arteriosus—or a ligamentum arteriosum—at the left side of the esophagus and trachea. Hence, the esophagus is compressed from behind by the aortic arch and the trachea is encroached on anteriorly by the pulmonary artery. It is conceivable that division of the patent ductus arteriosus or the ligamentum arteriosum would allow the pulmonary artery to fall forward and give sufficient room for the esophagus and trachea.

This anomaly is now commonly referred to as a right aortic arch with retroesophageal left subclavian artery and left ligamentum. The second type of vascular ring described by Gross was a “divided” or “split” aortic arch:

At a short distance above its valve, the aorta divides into two limbs. One of these passes posteriorly and then to the left behind the esophagus, and the other courses to the left in front of the trachea, the two merging in the left side of the mediastinum and forming the descending aorta.

This developmental abnormality is now commonly called a double aortic arch.

Gross subsequently described division of an aberrant right subclavian artery for dysphagia lusoria in 1946 and suspension of the innominate artery to the sternum for compression of the trachea in 1948. These conditions are not anatomically complete (true) vascular rings but rather “partial” rings; they are vascular anomalies that also cause symptoms from tracheal or esophageal compression.

The tradition of caring for children with vascular rings at Children’s Memorial Hospital in Chicago began in 1947, when Willis Potts successfully divided a double aortic arch in an 8-year-old boy. In 1954, Potts became the first surgeon to describe a successful operation for an infant with pulmonary artery sling. Potts operated on a 5-month-old child with recurrent attacks of dyspnea and cyanosis and diagnosed a pulmonary artery sling intraoperatively. He ligated, divided, and reimplemented the left pulmonary artery anterior to the trachea through a right anterolateral thoracotomy. Many infants with pulmonary artery sling have, in addition, complete tracheal rings—the so-called ring-sling complex. For these critically ill infants, Idriss and associates described a technique of pericardial patch tracheoplasty, which has considerably improved their outcome. Improvements in the diagnosis, pathoanatomic comprehension, and surgical management of children with vascular rings have led to superior results as well as low morbidity and mortality.

Evidence of an excellent outcome is clearly shown in this issue of the Mayo Clinic Proceedings (pages 1056 to 1063), in which van Son and associates review a 45-year experience with 37 patients who underwent an operation for relief of tracheobronchial obstruction from a vascular ring. In this series, 18 patients had a double aortic arch, 12 had a right aortic arch with a left ligamentum, 4 had a left aortic arch with aberrant origin of the right subclavian artery, 2 had a pulmonary artery sling, and 1 had a left aortic arch with a right ductus. The report includes superb illustrations and describes an extremely long follow-up (mean, almost 20 years). The only death in this series occurred in a 6-week-old patient in 1947. Only 3 of 37 patients had difficulty with...
residual tracheomalacia. The authors conclude that magnetic resonance imaging is the imaging technique of choice for vascular rings.

Our experience with vascular rings at Children's Memorial Hospital between 1947 and 1993 is summarized in Table 1. In this series, 19 deaths occurred, 10 of which were in infants with major associated anomalies. No operative or late deaths from an isolated vascular ring have occurred since 1967. On long-term follow-up, 92% of these patients are free of respiratory symptoms. A comparison of the experience at Children's Memorial Hospital and at the Mayo Clinic is of interest.

**Complete Vascular Rings.**—In patients with complete vascular rings, van Son and colleagues conclude that magnetic resonance imaging is the imaging technique of choice for adequate delineation of the vascular and tracheal anatomy. Although magnetic resonance imaging clearly reveals the vascular ring anatomy, other issues must be addressed when an imaging technique is selected. From a practical standpoint, my colleagues and I believe that more expensive diagnostic maneuvers have not added substantially to the information that can be acquired from simpler tests. In our experience, most patients underwent successful division of a vascular ring through a left thoracotomy without necessarily having a magnetic resonance imaging or computed tomographic scan and with the diagnosis based solely on chest roentgenography and barium swallow study.

Selective diagnostic processing beginning with chest roentgenography and followed by barium swallow study discloses patients who have (1) no evidence of vascular ring and who require no further study, (2) classic vascular ring and who require no further diagnostic intervention preoperatively, or (3) an unusual anatomic configuration or nondiagnostic test result that, in some cases, may necessitate either a computed tomographic or a magnetic resonance imaging scan. Of note, we have not used angiography to diagnose vascular ring for more than 15 years. Once the diagnosis has been determined, further tests to refine the diagnosis or to provide various nuances of anatomic information are unnecessary. Because of the increasing cost constraints on health care, finding the most economically effective and efficient way of making the diagnosis will be imperative so that these children may have prompt operative intervention.

**Innominate Artery Compression Syndrome.**—That no children in the Mayo Clinic experience were treated for innominate artery compression syndrome is surprising. This syndrome results from anterior compression of the trachea by the innominate artery, which can cause stridor, respiratory distress, cyanosis, reflex apnea, and even death. Diagnosis is made by rigid bronchoscopy, which shows a characteristic pulsatile anterior compression of the trachea that extends from left to right. Innominate artery compression syndrome is treated with a right anterolateral thoracotomy and suspension of the innominate artery to the sternum. In our series, the median age at the time of innominate suspension was 9 months. We have performed this procedure in 79 patients; the symptoms resolved in more than 90% of the infants, but 2 patients required reoperation because of technical failure.

**Pulmonary Artery Sling.**—Pulmonary artery sling causes compression of the trachea and right main-stem bronchus, and most infants with this anomaly have severe respiratory distress within the first year of life. We have operated on 15 children with pulmonary artery sling at Children's Memorial Hospital (Table 1), a third of whom have had associated complete tracheal rings (the “ring-sling complex”) and life-threatening tracheal obstruction. The membranous portion of the trachea is absent, and the cartilage rings are completely circumferential. Many infants transferred to our institution cannot be intubated with even the smallest endotracheal tube. Computed tomography or magnetic resonance imaging will demonstrate both the pulmonary artery sling and the complete tracheal rings. The pulmonary artery sling is repaired through a median sternotomy with use of extracorporeal circulation. The left pulmonary artery is reimplemented into the main pulmonary artery anterior to the trachea. A pericardial patch tracheoplasty is performed under bronchoscopic guidance for the associated complete tracheal rings. Of the six infants with the ring-sling complex in our operative series, five are long-term survivors.

**Aberrant Right Subclavian Artery.**—In the series of patients with vascular rings at Children's Memorial Hospital, no patient with a left aortic arch and aberrant right subclavian artery has undergone operation since 1973. In most patients who have dysphagia and aberrant right subclavian artery, our impression has been that the dysphagia is caused by some factor other than the aberrant right subclavian artery. Beabout and coworkers reported 106 cases of aberrant right subclavian artery from the Mayo Clinic, and some degree of dysphagia was present in only 5 cases. In none of

| Table 1.—Vascular Rings: Children's Memorial Hospital, 1947 Through 1993 |
|--------------------------|----------------|
| Type of vascular ring     | No. of patients |
| Complete                 | 249             |
| Double aortic arch        | 80              |
| Right aortic arch with left ligamentum | 68          |
| "Partial"                |                 |
| Innominate artery compression syndrome | 79            |
| Pulmonary artery sling    | 15              |
| Left aortic arch and aberrant right subclavian artery | 7          |
| Total                    |                 |

those five patients was the dysphagia thought to be attributable to the presence of the aberrant right subclavian artery. The exception is the adult patient in whom a large aneurysm develops at the base of the aberrant right subclavian artery, which can compress the esophagus and impose an additional risk associated with rupture. In their review, van Son and colleagues had four such patients, three of whom were adults (one with a large aneurysm). Their approach to a patient with a left aortic arch and a retroesophageal right subclavian artery through a right thoracotomy is noteworthy.

Left Aortic Arch and Right Ductus Arteriosus.—One patient in the Mayo Clinic experience who deserves special attention is the 2-year-old child with a left aortic arch and a right ductus arteriosus. This type of vascular ring necessitates a right thoracotomy for effective division. In our experience at Children’s Memorial Hospital with 148 patients with complete vascular rings, a child with this lesion has not been encountered, similar to the rare frequency in other reports. Fortunately, a left aortic arch with a right descending aorta and right ligamentum arteriosum (or ductus arteriosus) has a distinctive appearance on an esophagogram. An extrinsic indentation is evident in the upper left posterior aspect of the esophagus at the level of the second thoracic vertebra, much higher and leftward than the typical vascular ring. This finding should prompt further evaluation with either computed tomography or magnetic resonance imaging before operative intervention.

Conclusion.—The history of the surgical management of infants and children with vascular rings is fascinating. The evolution of surgical techniques has led to an excellent outcome in almost all cases. The major remaining issue is actually making the initial diagnosis of vascular rings, which necessitates a high index of suspicion on the part of primary-care physicians who first encounter these children. The difficulty for these physicians is that vascular rings are rare, as evidenced by a total of only 37 patients in the currently reported 45-year review from the Mayo Clinic. The most important single message of the article by van Son and colleagues may simply be to increase the index of suspicion and heighten the awareness of pediatricians, family practitioners, and otolaryngologists for the possible diagnosis of a vascular ring in any child with respiratory distress, stridor, “barky” cough, apnea, or dysphagia. The Mayo Clinic experience suggests that once a vascular ring has been diagnosed, the treatment is straightforward and extremely effective.

Carl L. Backer, M.D.
Division of Cardiovascular-Thoracic Surgery
The Children’s Memorial Hospital
Chicago, Illinois

REFERENCES