Vascular Ring Due to Left Aortic Arch and Right Descending Aorta

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SUMMARY Two children presented with symptoms suggestive of a vascular ring. Barium esophagrams in each case showed a posterior impression caused by a retroesophageal aortic arch. Angiographic studies defined a rare form of vascular ring resulting from an ascending left-sided aorta, a descending right-sided aorta and a right-sided ligamentum arteriosum. At surgery, the diagnosis was confirmed and the anomaly was corrected in each patient. Left pulmonary anomalies were present in each patient.

Vascular rings surrounding the trachea and esophagus are a cause of dysphagia and respiratory distress during infancy. Most symptomatic vascular rings result from a double aortic arch system or the coexistence of a right-sided aortic arch with a left-sided patent ductus arteriosus or ligamentum arteriosum.1,2

We report the cases of two infants with marked dysphagia associated with an extremely unusual form of vascular ring. Feeding problems were managed successfully by surgery.

Case Reports

Case 1

This 1-month-old male infant was born after an uneventful, full-term gestation. Birth weight was 6 pounds, 3 ounces. He presented with a 2-week history of regurgitation, choking and nasopharyngeal reflux after all bottle feedings. He had no history of respiratory distress or cyanosis.

On physical examination, vital signs were normal and weight was 8 pounds. The patient was acyanotic and the lungs were clear. Cardiac examination revealed normal pulses, a quiet precordium, a normal liver size and a grade II/VI high-pitched systolic murmur along the upper right and left sternal borders. The infant choked and regurgitated formula repeatedly through the nose and mouth during feeding.

Laboratory evaluation showed normal electrolytes, blood count and urinalysis. A chest x-ray showed a normal heart size, normal pulmonary vascular markings, and scant right perihilar and right upper lobe infiltrates. An ECG was normal. A barium esophagram showed marked reflux of contrast material into the nasopharynx and extrinsic, posterior compression of the proximal esophagus (fig. 1).

A vascular anomaly of the great vessels was suspected, so cardiac catheterization was performed. There was mild right ventricular and pulmonary artery hypertension (40-mm Hg systolic pressure). No intracardiac or extracardiac shunts were detected. Angiography revealed no anatomic intracardiac defects. Left ventriculograms showed that the aorta arose normally from the left ventricle and ascended anteriorly and to the left in a normal fashion. In its terminal portion, the aortic arch turned medially, crossed the midline behind the esophagus, which it compressed, and descended to the right of the thoracic spine (fig. 2). There was an aberrant right subclavian artery arising from the descending thoracic aorta (fig. 3). The left pulmonary artery was smaller in caliber than the right.

Surgery was performed 2 days later through a right lateral thoracotomy. The surgical findings confirmed the presence of a vascular ring. A ligamentum arteriosum, which originated from the descending aortic arch together with the anomalous right subclavian artery and inserted into the right pulmonary artery, completed the vascular ring and tethered the distal aortic arch anteriorly against the esophagus. The ligamentum was divided, and the esophagus was dissected and mobilized. Since surgery, the infant has been free of feeding difficulties or other symptoms and has gained weight appropriately.

Case 2

This 3½-year-old female had a history of chronic dysphagia, recurrent lower respiratory infections and easy fatigability. Physical examination showed normal peripheral pulses and blood pressures, a right ventricular lift, an increased intensity of the pulmonic component of the second heart sound and a grade II/VI systolic ejection murmur. A chest x-ray showed right ventricular enlargement and pulmonary venous congestion. The ECG showed right atrial and right ventricular hypertrophy. A barium esophagram showed a posterior impression on the esophagus. Cardiac catheterization revealed no intracardiac shunts; the pulmonary artery pressure was 72/25 mm Hg. The left pulmonary artery was not seen (fig. 4). The aortic arch ascended to the left, but descended on the right; the right subclavian artery arose from a diverticulum on the descending aorta.

Through a right thoracotomy, a right-sided liga-
mentum arteriosum was divided and the esophagus was mobilized. The patient recovered from surgery uneventfully and no longer complains of dysphagia.

**Discussion**

In 1948, Edwards hypothesized that a left-sided aortic arch with a retroesophageal component and a right-sided descending aorta, in conjunction with an
anomalous right subclavian artery and a ligamentum arteriosum joining the origin of the right subclavian artery to the right pulmonary artery, could form a vascular ring about the trachea and esophagus (fig. 5). Later that year, Paul reported two children (ages 7 and 11½ years) in whom a left-sided aortic arch and descending right-sided aorta were documented for the first time; however, normal origin of the right subclavian artery was present in each case and a complete vascular ring was not present. In 1948, Edwards also reported the autopsy findings from a 17-month-old male who died from small bowel obstruction after abdominal surgery. Although that infant had no dysphagia or respiratory distress during life, pathologic examination of the heart and great vessels revealed a left-sided aortic arch with a retroesophageal component, a right-sided descending aorta, anomalous origin of the right subclavian artery and a right-sided ligamentum arteriosum. Since 1948, nine additional cases have been reported in which a left-sided aortic arch has coexisted with a right-sided descending aorta. Four of the patients were adults, in whom the aortic anomaly was suspected from fluoroscopic examination. Five of these patients were children. Although each had a left-sided aortic arch and a right-sided descending aorta, a symptomatic vascular ring of the type described by Edwards was present in only one of the cases.

Our cases had a true vascular ring about the trachea and esophagus. The aortic arch arose from the left ventricular outflow tract and coursed leftward, anterior to the trachea, in the normal fashion. Following a course that was somewhat higher than normal into the upper thoracic cavity, the aorta then coursed posteriorly and to the right behind the esophagus. The esophageal impression from the descending portion of the aortic arch was evident on the barium esophagram. The aorta then coursed inferiorly, descending on the right side of the spine. The great vessel branching pattern was abnormal. The first branch of the ascending aorta was the right common carotid artery, followed by the left common carotid and the left subclavian arteries. The right subclavian artery arose from the descending, right-sided portion of the thoracic aorta. It appeared to arise from a diverticulum-like outpouching of the anterior aortic wall, from which the ligamentum arteriosum also arose. The ligamentum arteriosum attached to the right pulmonary artery and completed the vascular ring about the trachea and esophagus (fig. 5). The left pulmonary artery was hypoplastic in case 1 and absent in case 2. Left pulmonary artery anomalies have not been reported in association with this pattern of aortic arch malformation.

The clinical findings in these two cases were suggestive of a vascular ring. The catheterization and angiographic findings detailed the anomalous course of the aortic arch. Because the ductus arteriosus was not patent, all the structures that contributed to the vascular ring could not be visualized angiographically. Hypothetical and autopsy reports by Edwards, however, alerted us to the possibility that these infants’ symptoms were caused by this extraordinarily rare condition.

These case reports emphasize the complexity and diversity of vascular malformations that may constrict the trachea and esophagus. They also point out the contributions made to clinical medicine by detailed and meticulous studies of the embryology and pathology of congenital malformations.

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