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GUEST EDITOR'S PAGE

Identification and Management of Vascular Rings and Slings

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Vascular rings and slings are an uncommon phenomenon accounting for only 1% to 3% of congenital heart malformations.[1] They are often associated with other congenital heart defects, most commonly conotruncal abnormalities. Tetralogy of Fallot is the most commonly associated defect. Vascular rings may also be associated with chromosomal abnormalities such as 22q11 deletion, also known as DiGeorge syndrome. A vascular ring is an anomaly that involves encirclement of the trachea or esophagus. This encirclement can create a stricture that impedes normal airway mechanics, esophageal motility, or both. The most common type of symptomatic vascular ring involves the presence of a right sided aortic arch with an aberrant left subclavian artery. Rings can be classified as either complete or incomplete. A complete ring occurs when both the trachea and the esophagus are encircled by a vascular anomaly, and an incomplete ring involves less than full encirclement of both structures.[2] Pulmonary artery slings are a distinct anomaly with the potential to exert pressure on neighboring structures. In this type of sling, the left pulmonary artery arises embryologically from the right pulmonary artery. This results in a course that loops in between the esophagus and trachea.

In this issue of *JACC: Case Reports,* Sangupta et al[3] and Naaraayan et al[4] both have offered different perspectives on vascular ring and pulmonary artery sling phenomena. These 2 cases highlight the need for a high index of suspicion by pediatricians and adult cardiologists alike to consider advanced imaging. Sangupta et al[3] report the case of a 9-year-old child with persistent symptoms and multiple prior aortopexies, whereas Naaraayan et al[4] describe a newly identified defect in a 51-year-old adult. In each case, the patient presented with primarily respiratory symptoms, not the more common symptoms of chest pain or pressure.

Sengupta et al[3] discuss the case of a child who had 2 prior aortopexies for vascular ring symptoms yet had persistent symptoms. A circumflex aorta was identified and is comprised of the right aortic arch with a left-sided ligamentum arteriosum and a descending aorta that crosses posteriorly from right to left above the level of the tracheal carina. Despite 2 prior successful aortopexies, this third uncrossing procedure finally offered symptom relief.

As Naaraayan et al[4] report, it is more common for pulmonary artery sling patients to present in early childhood with overwhelming respiratory symptoms. In the very small number of previously reported adult cases, the most common presentation is one of obstructive pulmonary physiology presenting as a pneumonia as theirs does as well. As advanced cardiac imaging becomes increasingly used, we may find more cases of previously undiagnosed left pulmonary artery sling similar to the Naaraayan case. It is even more uncommon to find 2 anomalies in the same case as they have identified with a persistent left superior vena cava to coronary sinus.

The development of the vascular system begins in the third week of gestation. The thoracic aorta is divided into 3 segments: ascending, transverse arch, and descending thoracic aorta. Each of these has separate development stages. The ascending aorta arises from the primitive heart tube with the truncus arteriosus forming as the base of the pulmonary and aortic roots. The proximal, medial, and distal aortic arches develop from distinct pharyngeal arches. These arches come in pairs but eventually there is regression of some and dominance of others. The right and left third arches persist as the right and left

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carotid arteries. The left fourth arch persists as the transverse arch. The left sixth arch persists as the ductus arteriosus. The right and left subclavian arteries arise from the seventh intersegmental arteries along the posterior body wall and are remodeled into the final aortic arch. The descending thoracic aorta develops from the dorsal aorta which initially begins as a paired structure eventually fusing into a single structure. An aberrant left pulmonary artery arises from a lack of development of the sixth aortic arch. Pulmonary artery slings are considered a subtype of vascular ring anomalies.[1] Infants with vascular rings are often noted to be snuffling breathers. Young children and adolescents may present with exertional shortness of breath. As the aorta dilates with exertion to accommodate increased cardiac output, there is compression of the trachea leading to dyspnea, and often an incorrect hypothesis of exercise induced asthma. Failure to thrive in infancy is associated with esophageal compression. Difficulty in swallowing is seen in older children and adults. There can be a dynamic component to symptomatology in complete encirclement wherein the dilated aorta may further compress both the esophagus and the trachea. The wide range of presentations from critical airway obstruction in a neonate to an adult who self-limits activities secondary to exertional symptoms necessitate a high level of clinical suspicion in evaluating these patients. A transthoracic echocardiogram to visualize the aorta, arch vessels, and pulmonary arteries is needed. If the diagnosis remains unclear, advanced vascular imaging with computed tomography angiography or with cardiac magnetic imaging may further define aortic anatomy. Depending on presenting symptoms, a bronchoscopy and an esophagogram may both be needed.

Surgical ligation or resection is the only treatment for symptomatic patients and can be performed minimally invasively in most cases for the typical vascular ring. In most cases, surgical ligation or resection can resolve symptoms; however, sometimes a tracheopexy may be indicated. In general, asymptomatic patients diagnosed incidentally with a vascular ring require no intervention or long-term monitoring.[5] There is no role for medical management. Surgical repair of a pulmonary artery sling involves re-implantation of the anomalous pulmonary artery and potential need for repair of tracheal stenosis. Long-term survival is excellent, although early mortality can be related to the need for concomitant tracheal surgery.[6] Vascular rings and pulmonary artery slings as highlighted by Sangupta et al[3] and Naaraayan et al[4] create a window into an uncommon anomaly with 2 equally uncommon presentations. It is, as always, critical to use every tool at our disposal to better understand symptoms in this heterogenous population.

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REFERENCES

1. Davies M, Guest PJ. Developmental abnormalities of the great vessels of the thorax and their embryological basis. *Br J Radiol*. 2003;76:491.

2. Haranal M, Srimurugan B, Sivalingam S. Circumflex aorta: an uncharted territory. *Asian Cardiovasc Thorac Ann*. 2022;30:217.

3. Sengupta A, Brigger M, Lamberti J, Myrthy R. Recurrent pediatric respiratory distress from a challenging vascular anomaly: the uncrossing operation for circumflex aorta. *J Am Coll Cardiol Case Rep*. 2024;29:102305.

4. Naaraayan A, Buza V, Gerula C, Waller AH. Co-existing pulmonary artery sling and persistent left superior vena cava. *J Am Coll Cardiol Case Rep*. 2024;29:102315.

5. Woods RK, Sharp RJ, Holcomb GW 3rd, et al. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. *Ann Thorac Surg*. 2001;72:434.

6. Saran N, Dearani J, Said S, et al. Vascular rings in adults: outcome of surgical management. *Ann Thorac Surg*. 2019;108(88):947.