

Vascular Ring Due to Right Aortic Arch with Mirror-Image Branching and Left Ligamentum Arteriosus: Complete Preoperative Diagnosis by Magnetic Resonance Imaging

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Abstract. Magnetic resonance imaging (MRI) has been increasingly employed in the evaluation of aortic arch abnormalities. We present a rare form of vascular ring, that of a right aortic arch with mirror-image branching and left ligamentum arteriosus, which was clearly defined preoperatively by MRI. The patient subsequently underwent successful surgical division of the vascular ring followed by improvement in respiratory symptoms.

Key words: Vascular ring — Right aortic arch — Left ligamentum arteriosus — MRI

Case Report

A 22-month-old girl was noted to have frequent respiratory illnesses with associated noisy breathing. Initial treatment modalities had been directed toward a respiratory etiology by her primary physician. Because no significant improvement was realized, attention was directed to the possibility of a vascular ring. A barium swallow revealed a posterior indentation of the esophagus at the level of T5 and a smooth impression on the right side of the esophagus. A referral was made to the pediatric cardiology department. The patient's cardiac exam was normal. A degree of stridor was noted, particularly with agitation. Echocardiographic analysis revealed normal intracardiac anatomy. Arch imaging revealed a right aortic arch with mirror-image branching. Given the high level of suspicion of a vascular ring, magnetic resonance imaging (MRI) was performed. Thin-section (3 mm, 0 gap) axial and coronal T1-weighted sequences were performed with cardiac gating. The trachea was noted to be displaced to the left of the midline and significantly narrowed. A right aortic arch with mirror-image branching and a diverticulum from the descending aorta were clearly defined (Figs. 1 and 2). A ligament coursing from the diverticulum to the main pulmonary artery was identified. The patient subsequently underwent division of the vascular ring via a left thoracotomy. The thick ligamen-

tum was doubly ligated and divided without difficulty. Subsequently, the patient has exhibited improvement of her respiratory symptoms.

Discussion

Investigation for a vascular ring is important in a patient with recurrent respiratory symptoms. Barium swallowing studies can be helpful in determining the presence of a ring but can be nonspecific. Echocardiography can be used to delineate the aortic arch sidedness and great vessel branching pattern. In cases in which echocardiographic visualization is difficult or the ring is of an unusual pattern, angiography can be performed; however, MRI imaging has been increasingly employed.

The two most common forms of a complete ring are a double aortic arch, typically with a larger right arch component, and a right aortic arch with aberrant left subclavian artery, in which a left-sided ligamentum arteriosus completes the ring [2]. Our patient had a significantly rarer form of a ring—that of a right aortic arch with mirror-image branching and left ligamentum arteriosus. Given the infrequent occurrence, exact diagnosis can be more challenging in this situation. One clue was the patient's normal intracardiac anatomy. The vast majority of patients with a right aortic arch with mirror-image branching have associated intracardiac anomalies [5], most commonly that of tetralogy of Fallot and truncus arteriosus. The constellation of a right aortic arch with mirror-image branching, normal intracardiac anatomy, and a vascular ring has previously been defined [4]. As described, the location of the break of the “hypothetical double aortic arch” as proposed by Edwards [3] determines where the ductus arteriosus originates and also appears to impact on the intracardiac anatomy. In-

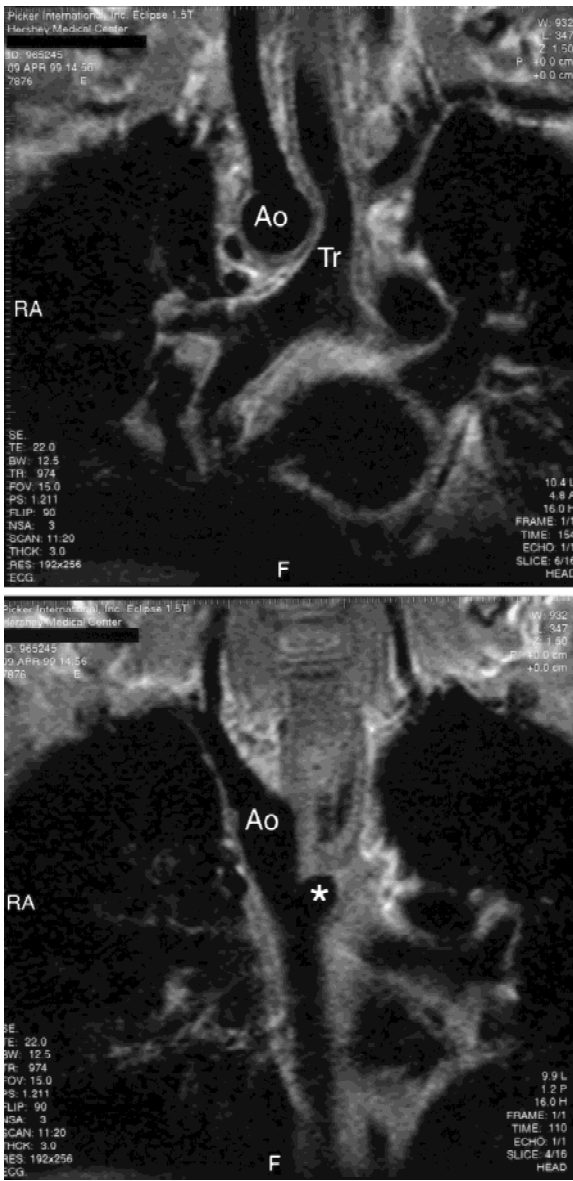


Fig. 1. Coronal sections. (**Top**) The right aortic arch (*Ao*), giving rise to the right common carotid artery, is seen producing an indentation on the right aspect of the trachea (*Tr*). The distal aspect of the left subclavian artery and left carotid artery, which arose from the left innominate, are seen. (**Bottom**) On this more posterior slice, the distal right arch, right subclavian artery, and descending aorta are well visualized. The prominent diverticulum (*asterisk*) is seen arising from the descending aorta.

terruption of the left arch posterior to the left ductus arteriosus results in a right aortic arch with mirror-image branching with the ductus arteriosus originating from the left innominate or left subclavian artery. This was described by Garti et al. [4] as type A (Fig. 3). No ring is formed and intracardiac anomalies are frequently associated. In contrast, interruption of the left arch between the left sub-

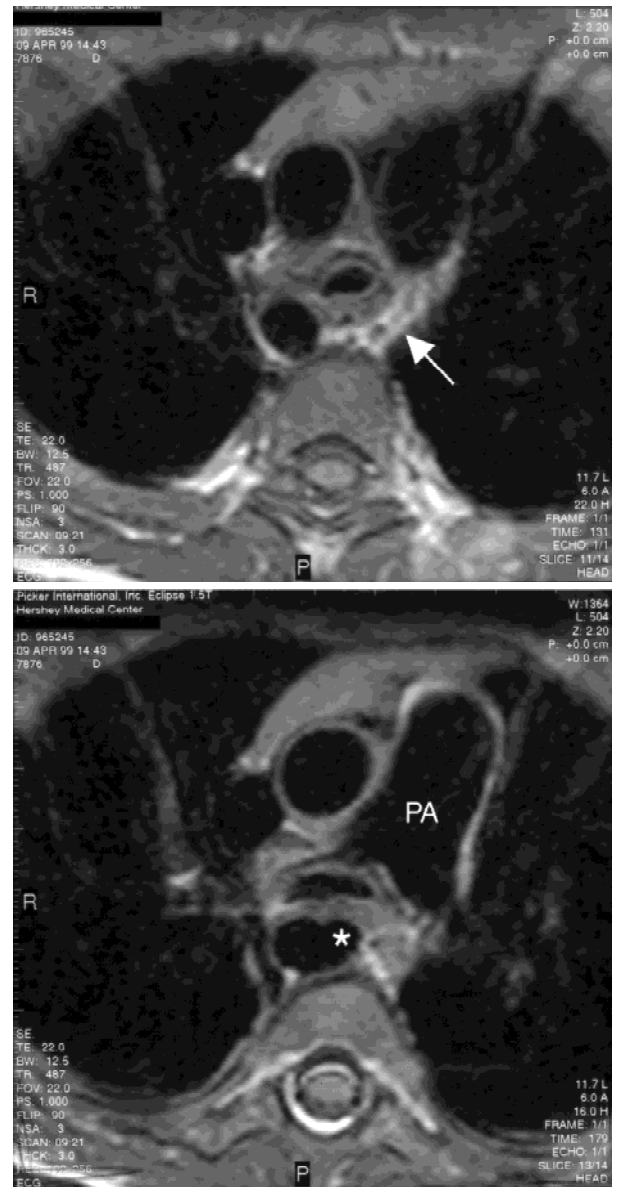


Fig. 2. Transverse sections. (**Top**) The trachea is seen encircled by the vascular ring with a thick posterior ligament (*arrow*) traversing from the descending aorta to the pulmonary artery. (**Bottom**) On this more inferior slice, tracheal compression is noted. The diverticulum (*asterisk*) is seen from the descending aorta. *PA*, pulmonary artery.

clavian artery and left ductus arteriosus results in a right aortic arch with mirror-image branching with the ductus arteriosus arising from the descending aortic diverticulum. This had been referred to as type B. A vascular ring is formed and the intracardiac anatomy is typically normal. A ring of this general nature but with an aneurysm of the left ductus arteriosus has been reported [1]. Definitive diagnosis of these abnormalities prior to surgical intervention had previously relied on angiographic evaluation. MRI evaluation had been employed in a case

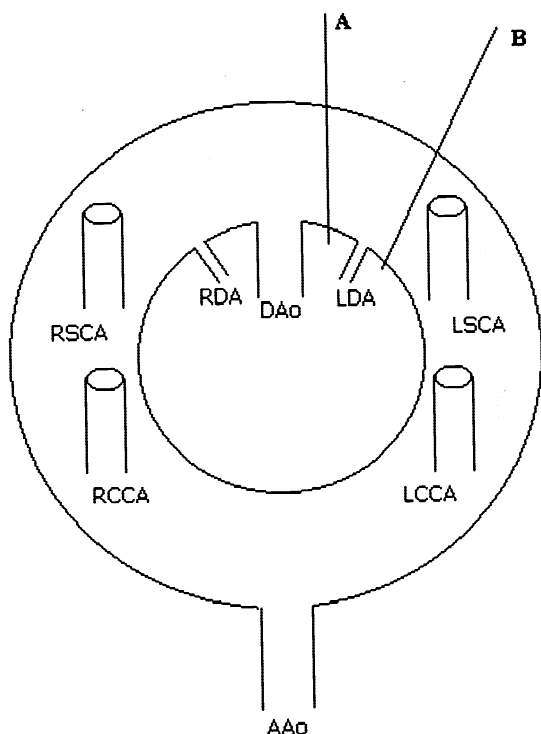


Fig. 3. Limited schematic representation of the “hypothetical double aortic arch” as proposed by Edwards [3]. Locations of breaks in the arch (*A*, *B*) result in different clinical presentations (see text). *AAo*, ascending aorta; *DAo*, descending aorta; *LCCA*, left common carotid artery; *LDA*, left ductus arteriosus; *LSCA*, left subclavian artery; *RCCA*, right common carotid artery; *RDA*, right ductus arteriosus; *RSCA*, right subclavian artery.

similar to ours; however, the presumed preoperative diagnosis was that of a double aortic arch with left arch atresia [6]. The exact nature of the ring in that instance was subsequently identified at the time of surgery. In our case, the presence of a prominent ductal diverticulum and a thick fibrous ligament allowed for complete diagnosis of the anatomy preoperatively.

Unusual forms of vascular rings should be investigated based on clinical suspicions. MRI can be an excellent technique to delineate various forms of vascular rings, including those less commonly encountered.

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