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Magnetic Resonance Imaging of a Distorted Left Subclavian Artery Course: An Important Clue to an Unusual Type of Double Aortic Arch

K.W. Holmes,[1] D.A. Bluemke,[2] L.A. Vricella,[3] W.J. Ravekes,[1] K.M. Kling,[4] P.J. Spevak[1]

[1]Division of Pediatric Cardiology, Johns Hopkins Hospital, 600 N. Wolfe Street, Brady 501, Baltimore, MD 21287, USA

[2]Department of Radiology, Johns Hopkins Hospital, 600 N. Wolfe Street, Baltimore, MD 21287, USA

[3]Division of Cardiothoracic Surgery, Johns Hopkins Hospital, 600 N. Wolfe Street, Baltimore, MD 21287, USA

[4]Division of Padiatric Surgery, Johns Hopkins Hospital, 600 N. Wolfe Street, Baltimore, MD 21287, USA

Abstract. The objective of this study was to determine if distortion of the left subclavian artery course distinguishes double aortic arch with atretic left dorsal aorta from right aortic arch with mirror image branching. We performed a retrospective case series at a tertiary care center. Twenty-six patients undergoing magnetic resonance imaging for suspicion of a vascular ring were identified, 6 of whom had an atretic left dorsal aorta confirmed by surgical inspection. Six patients with the diagnosis of right aortic arch with mirror image branching were identified for comparison. The course of the left subclavian artery was assessed using surface-rendered magnetic resonance angiography (MRA) and axial fast spin echo images. All patients with double aortic arch had clinical symptoms suggestive of esophageal or tracheal compression. Six patients had double aortic arch, 3 of whom had an atretic left dorsal aorta. In these 3 patients, the branching patterns on MRA mimicked right aortic arch mirror image branching except for the distortion of the initial course of the left subclavian artery. Surgical observation confirmed the presence of an atretic left dorsal aorta that resulted in tension on the left subclavian artery pulling it posteriorly and inferiorly and completing the vascular ring. Patients with right aortic arch mirror image branching demonstrated no such subclavian artery distortion, and these patients did not have clinical symptoms suggestive of a vascular ring. Our results demonstrate that left subclavian artery distortion due to traction by an atretic left arch is an important diagnostic finding in the evaluation 6 patients with suspected vascular rings.

Key words: MRI — Vascular ring — Congenital heart disease — Aortic arch

Correspondence to: K.W. Holmes, *email:* kwholmes@jhmi.edu

A vascular ring is considered when a young child has symptoms of stridor or dysphagia. The diagnosis of a vascular ring can be made on the basis of characteristic findings on barium swallow or identification of vascular anatomy via echocardiography [7]. Although angiography has been used to further delineate the arch anatomy, recently gadolinium-enhanced magnetic resonance angiography (MRA), or computerized axial tomography (CT), has avoided the need for invasive testing [8].

The most common vascular rings include double aortic arch and right aortic arch with an aberrant left subclavian artery [2, 5, 6]. When the dorsal aortic portion of a double arch is atretic, visualization is problematic for imaging modalities depending on the presence of flow (e.g., color Doppler or MRA). In such patients, the atretic left dorsal aortic segment is usually not visualized and the anatomy mimics right aortic arch with mirror image branching (Fig. 1). A right aortic arch with mirror image branching is usually not a true vascular ring and is a finding often associated with additional intracardiac lesions; therefore, the distinction of a right aortic arch from an double aortic arch with atretic left dorsal aorta is clinically important in the symptomatic patient. We hypothesize that an accurate diagnosis is made by noting left subclavian artery distortion, which is the result of traction by the atretic left dorsal aorta.

Materials and Methods

Patients

Patients identified from the Johns Hopkins Hospital Pediatric Echocardiography Laboratory database were reviewed in this retrospective case series. We selected patients with the diagnosis of vascular ring from September 1995 to January 2004. Thirty patients

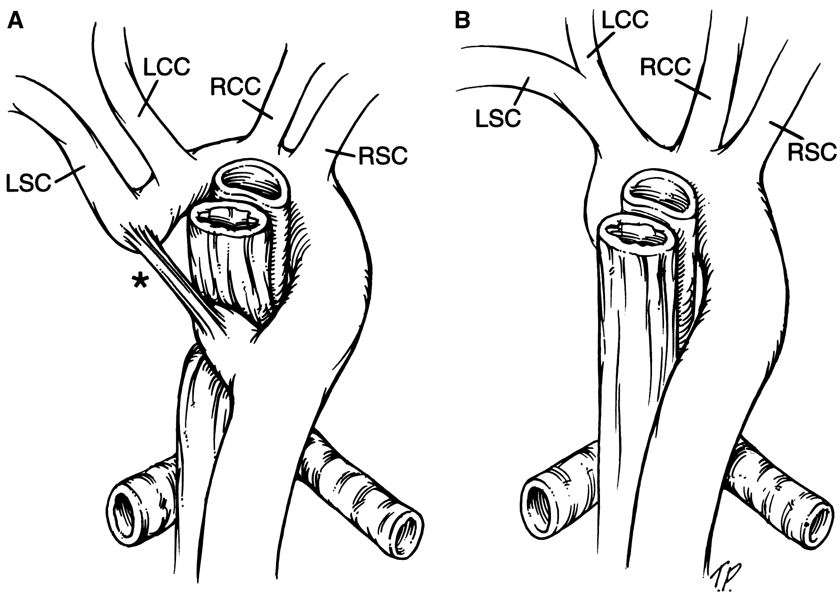

Fig. 1. Drawing contrasting left subclavian artery course in double aortic arch (with atretic left arch) versus right aortic arch (with mirror image branching). (**A**) In double aortic arch with atretic left arch, the vessel branching mimics right aortic arch with mirror image branching. However, the left subclavian artery is tethered inferiorly (*asterisk*) by the atretic left arch, which connects the left subclavian artery to the descending aorta. (**B**) In contrast, the left subclavian artery is not distorted in right aortic arch with mirror image branching. *LSC*, left subclavian artery; *LCC*, left common carotid artery; *RCC*, right common cardotid artery; *RSC*, right subclavian artery.

were identified, 26 of whom underwent magnetic resonance imaging (MRI) for additional definition of the vascular anatomy (Table 1). Seven of the 26 patients were found to have either a double aortic arch or double aortic arch with atretic left dorsal aortat surgery. MRI and MRA were available for review in 6 of these 7 cases. Ages ranged from 3 days to 9 months, and 2 patients were female and 4 male. Ten patients were identified with right aortic arch and mirror image branching; images of 6 patients were available for comparison. None of these patients had symptoms of stridor or dysphagia. Of these 6 patients, 5 were male and 1 was female. Ages ranged from 1 1/2 to 17 years. Five patients had tetralogy of Fallot, and 1 patient had right pulmonary artery stenosis.

The Johns Hopkins Medicine Institutional Review Board approved this protocol.

Magnetic Resonance Imaging/Angiography Protocol and Imaging Analysis

MRI was performed on a 1.5-T scanner (CV/i, General Electric Health Systems, Waukesha, WI, USA). After localizing images, axial double-inversion recovery fast spin echo images were obtained from the thoracic inlet to the diaphragm (TR, 1–2 RR intervals; TE, 5; 20- to 36-cm field of view adjusted for patient size; matrix, 256 × 192–256; slice thickness, 6–8 mm). Sagittal oblique double-inversion recovery fast spin echo images were obtained through the aortic arch using the same parameters. Three-dimensional (3D) MRA was performed after intravenous bolus administration of 0.2 mmol/kg gadodiamide (TR, 4.5–5.5; TE, 1.7; matrix, 256 × 192 interpolated to 512 × 512; 2- or 3-mm slice thickness; coronal acquisition plane). 3D MRA was reconstructed on a separate workstation (Advantage Windows 4.1, General Electric Health Systems) using maximum-intensity projection and surface rendering algorithms.

Table 1. MRI diagnosis of patients referred for evaluation of a vascular ring

Anatomy	No. of patients	%
RAAALS	15	58
LAAARS	4	15
DAA	4	15
DAAAL	3	12
Total	26	100

RAAALS, right aortic arch with aberrent left subclavian; LAAARS, left aortic arch with aberrent right subclavian; DAA, double aortic arch; DAAAL, double aortic arch with atretic left dorsal aorta.

Two observers reviewed all images independently. The presence of posterior and inferior distortion of the left subclavian artery course was used to indicate left subclavian artery distortion.

Results

The course of the left subclavian artery was examined in patients with double aortic arch and compared to that present in the six patients with right aortic arch mirror image branching. In the three patients with double arch and atretic left dorsal aorta, the left subclavian artery course was distorted, resulting in an inferior convexity in the initial takeoff from the innominant artery (Figs. 2a and 2b). The left subclavian artery course was not distorted in any of the three patients in whom both arches were patent

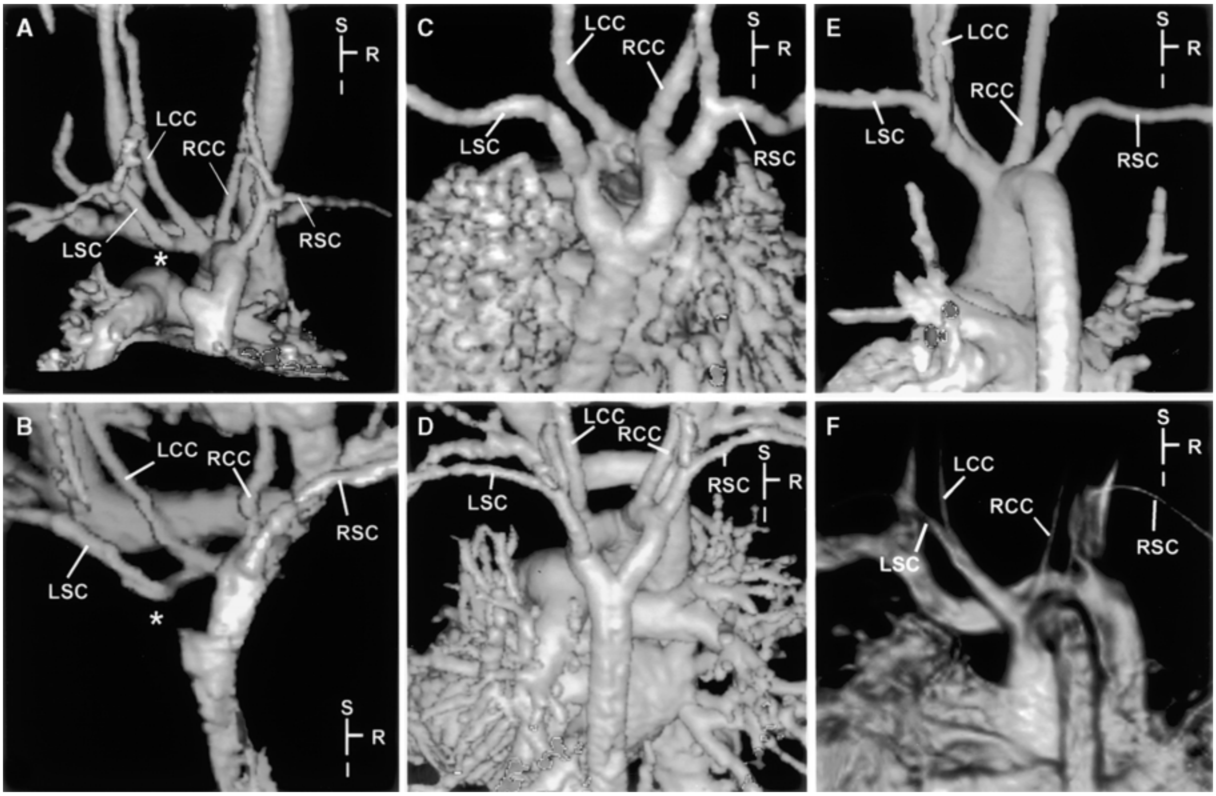

Fig. 2. Three-dimensional surface rendered images. (**A, B**) Double aortic arch with atretic left arch. The left subclavian artery course is distorted (*asterisk*) due to torsion by the ligamentum (i.e., atertic left dorsal arch), which connects the left subclavian artery with the descending aorta. This results in the left subclavian artery having an inferior convexity to its initial course. In contrast, the left subclavian artery course is not distorted in double aortic arch, where both arches are patent. (**C, D**) Not appreciating the left subclavian artery distortion in double aortic arch with an atretic left arch can result in confusion because the vessel branching mimics that seen in right aortic arch with mirror image branching (**E, F**). *LSC*, left subclavian artery; *LCC*, left common carotid artery; *RCC*, right common cardotid artery; *RSC*, right subclavian artery.

(Figs. 2c and 2d) or in any of the patients with right aortic arch mirror image branching (Figs. 2e and 2f).

MRI did not directly visualize the atretic left arch in any patients. Completion of the vascular ring by the atretic arch was inferred to be present based on the distorted left subclavian artery course on MRA and the presence of clinical symptoms including stridor or dysphagia. In addition, in two of three patients, a diverticulum arising from the descending aorta represented the patent portion of the atretic left dorsal aorta (Figs. 2a and 2b). Axial double-inversion recovery images also demonstrated the more posterior subclavian artery origin in patients with double aortic arch and atretic left dorsal aorta (Fig. 3). The atretic left dorsal aorta connecting with the descending aorta tethered the left subclavian artery position posteriorly. Of the three patients with double aortic arch with atretic left dorsal aorta, only one had documented evidence of tracheal compression by MRI axial imaging.

Surgical observation confirmed the MRI findings in all patients referred for intervention (Fig. 4). All patients with double aortic arch on MRI had surgery.

Discussion

Barium swallow, echocardiography, angiography, CT, and MRI all diagnose vascular rings. Barium swallow is an excellent means for screening, although other imaging modalities better delineate the vascular anatomy [1, 3]. Arteriography is now rarely performed due to the availability of noninvasive techniques. Echocardiography achieves a reliable diagnosis in most patient, but adequacy of images is operator dependent [7]. Although both MRI and CT consistently provide reliable images, both have minor associated risks. The later exposes the patient to ionizing radiation, and the former often necessitates general anesthesia and airway intubation in patients with probable airway compression.

The many variations of vascular ring anatomy can confuse the less experienced diagnostician. The most common true rings are double aortic arch and

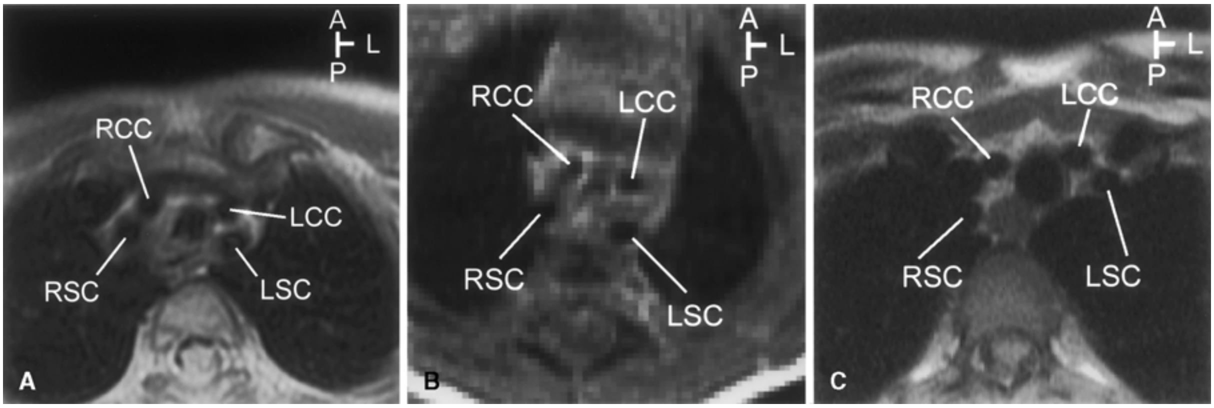

Fig. 3. Axial fast spin echo images demonstrating the position of the left subclavian artery. In both double aortic arch with patent (**A**) and atretic (**B**) left arch, the left subclavian artery position is posterior relative to the position of the trachea. With right aortic arch and mirror image branching (**C**), the left subclavian artery is more anterior. *LSC*, left subclavian artery; *LCC*, left common carotid artery; *RCC*, right common cardotid artery; *RSC*, right subclavian artery.

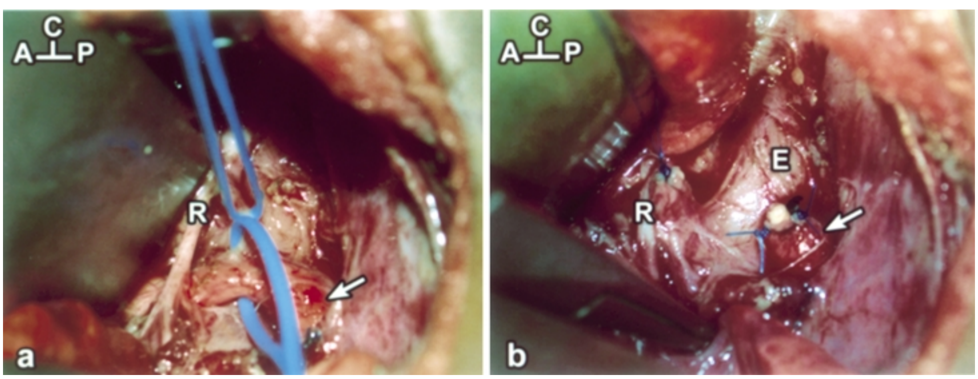

Fig. 4. Intraoperative appearance of double aortic arch with atretic left arch. (**A**) Preoperative appearance via left thoracotomy. The tie encircles the dorsal atretic component of the left arch, and its connection inferiorly with the descending aorta (*arrow*) is noted. The recurrent larnygeal nerve (*R*) crosses the atretic left arch. (**B**) Postoperative appearance after the atretic arch is divided. Note the ties on the two divided segments. By dividing the distal atretic arch, the left subclavian artery vascular ring is released, thereby freeing the esophagus (*E*). A = Anterior, P = Posterior, C = Cranial

right aortic arch with aberrant left subclavian artery. We describe a group of symptomatic patients with a less common and less well-recognized variation—double aortic arch with atretic left dorsal aorta. The vascular ring in these patients was completed by the atretic left dorsal aorta and the imaging appearance mimicked the much more common (and generally asymptomatic) diagnosis of right aortic arch with mirror image branching. Because all patients in our vascular ring group were less than 1 year of age, particular attention is necessary in the interpretation of imaging studies of these symptomatic children.

The incidence of right aortic arch in autopsy and radiographic studies is estimated to be 1 in 1000–1700 individuals [2, 6]. Nearly all patients with right aortic arch and mirror image branching have associated intracardiac anomalies, and rarely do they present with vascular rings [5, 6]. Patients with right aortic arches most often come to clinical attention secondary to cyanosis or a murmur rather than the typical symptoms of stridor and dysphagia seen in patients with vascular rings. Vascular rings in patients with right aortic arch and mirror image branching have rarely been reported; the ring is completed by a left ductus from the descending aorta to the pulmonary artery [4, 9].

Our study identified the course of the left subclavian artery by MRI as a helpful aid in imaging patients with suspected vascular rings. The diverticulum demonstrated in two of our patients is an additional clue to an imperforate vessel or ligamentum connecting the decending aorta to the subclavian artery. Although this study does not have the power to evaluate the sensitivity or specificity of these findings, the diagnostician should consider the diagnosis of double aortic arch with an atretic left dorsal aorta in the symptomatic patient.

In patients with suspected vascular rings, inferior and posterior distortion of the left subclavian artery course, with or without a diverticulum, suggests the presence of a complete ring. Particularly in the symptomatic patient, this important observation aids the clinician in avoiding the misdiagnosis of right aortic arch with mirror image branching, which may result in delayed diagnosis of a vascular ring or unnecessary additional imaging.

References

1. Azarow KS, Pearl RH, Hoffman MA, et al. (1992) Vascular ring: does magnetic resonance imaging replace angiography? *Ann Thorac Surg 53*:882–885
2. Bialowas J, Hreczecha J, Grzybiak M (2000) Right-sided aortic arch. *Folia Morphol (Warsz) 59*:211–216
3. Chun K, Colombani PM, Dudgeon DL, Haller JA Jr (1992) Diagnosis and management of congenital vascular rings: a 22-year experience. *Ann Thorac Surg 53*:597–502
4. Garti IJ, Aygen MM, Levy MJ (1979) Double aortic arch anomalies: diagnosis by countercurrent right brachial arteriography. *Am J Roentgenol 133*:251–256
5. Hastreiter AR, D'Craz IA, Cantez T, Namin EP, Licata R (1966) Right-sided aorta. I. Occurrence of right aortic arch in various types of congenital heart disease. II. Right aortic arch, right descending aorta, and associated anomalies. *Br Heart J 28*:722–739
6. Knight L, Edwards JE (1974) Right aortic arch. Types and associated cardiac anomalies. *Circulation 50*:1047–1051
7. Lillehei CW, Colan S (1992) Echocardiography in the preoperative evaluation of vascular rings. *J Pediatr Surg 27*:1118–1120
8. van Son JA, Julsrud PR, Hagler DJ, et al. (1994) Imaging strategies for vascular rings. *Ann Thorac Surg 57*:604–610
9. Zachary CH, Myers JL, Eggli KD (2001) Vascular ring due to right aortic arch with mirror-image branching and left ligamentum arteriosus: complete preoperative diagnosis by magnetic resonance imaging. *Pediatr 22*:71–73