Rings and Slings Revisited

Brandon M. Smith, MD[a,*], Jimmy C. Lu, MD[a,b], Adam L. Dorfman, MD[a,b], Maryam Ghadimi Mahani, MD[b,c], Prachi P. Agarwal, MBBS, MS[c]

KEYWORDS

• MR imaging • Vascular ring • Pulmonary sling • Pediatrics

KEY POINTS

- Vascular rings and pulmonary artery slings present with symptoms of tracheal and esophageal compression during infancy and childhood.
- Double aortic arch and right aortic arch with aberrant left subclavian artery and left patent ductus arteriosus (PDA) are the 2 most common types of vascular ring.
- Atretic segments cannot be directly visualized on MR imaging, but their presence can be inferred from other indirect imaging signs.
- MR imaging/magnetic resonance angiography (MRA) can diagnose these vascular anomalies and evaluate the airway and esophagus without the use of ionizing radiation.

INTRODUCTION

Vascular rings and pulmonary artery slings are rare congenital anomalies of the aortic arch branches or pulmonary arteries that often present with symptoms of tracheal or esophageal compression during childhood. Diagnosis of these conditions can be made using various imaging modalities, including radiography, esophagography, echocardiography, CT, and MR imaging.[1] The purpose of this review is to highlight the advantages of MR imaging for the diagnosis of vascular rings and pulmonary artery slings and to provide an insight into the diagnosis of specific vascular ring types using case examples.

ADVANTAGES OF MR IMAGING

MR imaging possesses unique advantages over other imaging modalities in the diagnosis of vascular rings and pulmonary artery slings. Chest radiography can detect a right aortic arch or tracheal compression, and barium esophagography may demonstrate esophageal compression,[2] but these modalities do not directly image vascular structures and cannot provide conclusive anatomic diagnosis, which can be essential for surgical planning.[3] Echocardiography can diagnose aortic arch sidedness and branching pattern, double aortic arches, and pulmonary artery anatomy. Tracheal and esophageal compression cannot be evaluated, however, and due to suboptimal acoustic windows may limit diagnostic accuracy for vascular rings.[4] Angiography can image patent vascular structures but cannot demonstrate tracheal and esophageal compression, requires invasive vascular access with a potential for complications, and requires ionizing radiation and nephrotoxic contrast agent.

Disclosures: No financial relationships or conflicts of interest to disclose.

[a] Division of Pediatric Cardiology, Department of Pediatrics and Communicable Diseases, University of Michigan, 1540 East Hospital Drive, Ann Arbor, MI 48109, USA; [b] Division of Pediatric Radiology, Department of Radiology, University of Michigan, 1500 East Medical Center Drive, Ann Arbor, MI 48109, USA; [c] Division of Cardiothoracic Radiology, Department of Radiology, University of Michigan, 1500 East Medical Center Drive, Ann Arbor, MI 48109, USA

* Corresponding author. University of Michigan Congenital Heart Center, C.S. Mott Children's Hospital, 1540 East Hospital Drive, Ann Arbor, MI 48109-4204.

E-mail address: bsmi@med.umich.edu

Magn Reson Imaging Clin N Am 23 (2015) 127–135

http://dx.doi.org/10.1016/j.mric.2014.09.011

1064-9689/15/$ – see front matter

Due to the ability to delineate vascular anatomy with multiplanar and 3-D reconstruction, and simultaneously demonstrate tracheal and esophageal compression, CT and MR imaging have become the preferred imaging modalities for the diagnosis of vascular rings and pulmonary artery slings. MRA can demonstrate vascular anatomy similar to CT without using ionizing radiation, which is particularly desirable in this young population.[5]

MR IMAGING PROTOCOL

MR imaging protocol for vascular ring and pulmonary artery sling evaluation typically consists of gadolinium-enhanced angiography and ECG-gated black blood and bright blood sequences.

- Black blood imaging in axial and oblique coronal (oriented along the trachea) planes is useful for assessment of vascular anatomy, tracheobronchial tree, and esophagus.
- Unless contraindicated, 3-D gadolinium-enhanced MRA provides comprehensive assessment of vascular anatomy.
- ECG and respiratory navigator-gated 3-D steady-state free precession (SSFP) imaging can simultaneously assess vascular, airway, and esophageal anatomy in a 3-D data set without the need for gadolinium.

VASCULAR RINGS

A vascular ring is defined as an abnormality of the aortic arch, its branches, or remnants that results in encircling of the trachea and esophagus with variable degrees of compression. Vascular rings represent 1% to 3% of congenital cardiac anomalies[6] and may remain asymptomatic or present at various times throughout childhood with stridor, dyspnea, chronic cough, wheezing, recurrent respiratory infection, or dysphagia, which may be relieved by surgery.[7]

Normal Aortic Arch Development

Normal aortic arch development involves variable regression of 6 paired aortic arches from the aortic sac to the paired dorsal aortae. The first, second, and third arches form parts of the maxillary, stapedial, and common carotid arteries, respectively.[8] Remnants of the fourth arch form the distal left aortic arch and proximal right subclavian artery. The fifth arches usually regress, and the sixth arches form parts of the pulmonary arteries and distal ductus arteriosus. The spectrum of aortic arch anomalies and vascular rings can be easily understood using the Edwards hypothetical double arch system (Fig. 1). Normal left arch anatomy occurs after involution of the distal right fourth arch and regression of the right ductus (Fig. 2). Abnormal patterns of regression of these paired aortic arches can result in a vascular ring.

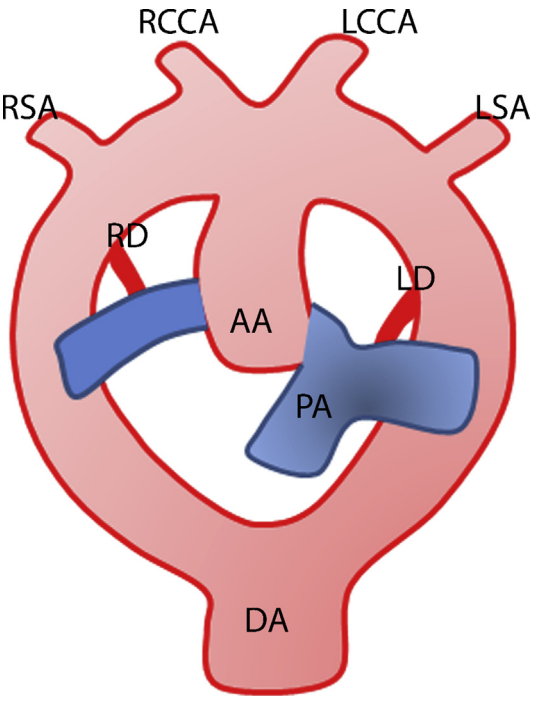

Fig. 1. Schematic diagram of Edward's hypothetical double aortic arch. The 2 arches give rise to the respective carotid and subclavian arteries and ductus arteriosus. AA, ascending aorta; DA, descending aorta; LCCA, left common carotid artery; LD, left ductus arteriosus; LSA, left subclavian artery; PA, pulmonary artery; RCCA, right common carotid artery; RD, right ductus arteriosus; RSA, right subclavian artery.

Aortic arch sidedness is defined by the location of the aortic arch in relation to the trachea (left, right, or double) as it passes over the mainstem bronchus (Fig. 3).[9] On echocardiography and angiography, this determination is indirect and may be inaccurate. MR imaging directly visualizes both the aorta and tracheobronchial tree, thus allowing conclusive diagnosis.

The spectrum of vascular rings (Table 1), with their distinctive MR imaging appearance, is described later.

Double Aortic Arch

Double aortic arch, a persistence of the right and left 4th embryonic arches, is the most common form of vascular ring, comprising approximately 50% of cases, and often presents from birth through childhood with respiratory symptoms,