INTRODUCTION

Vascular rings are rare malformations of pharyngeal arches that result in the trachea and esophagus being encompassed by the aortic arch and accompanying structures, potentially causing stridor and dysphagia. The most common variation is a double aortic arch; when both arches are patent, diagnosis is easily achieved by echocardiographic evaluation. When any portion of the double arch is absent, recognition is more difficult owing to absence of flow across that segment. We report a double arch with proximal left arch atresia versus interruption, which to the best of our knowledge is the first case to be diagnosed prospectively by echocardiogram.

CASE PRESENTATION

A 38-week gestation female was born to a G4P3 female with gestational diabetes controlled with insulin. The prenatal obstetric ultrasound was concerning for a right aortic arch with otherwise normal intracardiac anatomy. Apgars were 8 and 9, and birth weight was 3.67 kg. Peripheral oxygen saturations were 99%-100% on room air, with no discrepancy between pre- and postductal measurements. The electrocardiogram was normal for age. A grade I-II systolic ejection murmur was auscultated at the left upper sternal border at 1 day of life. A cardiology consult with echocardiogram was obtained. The echocardiogram revealed mild pulmonary valve stenosis (peak instantaneous gradient 35 mm Hg) and a patent right aortic arch. The right common carotid and right subclavian artery origins were identified from the transverse arch (Figure 1). The left common carotid and left subclavian artery origins were from a separate vascular structure that connected to the midline descending thoracic aorta (Figure 2). Color Doppler evaluation revealed retrograde filling of this vessel from the descending aorta (Figure 3). A small, left-sided arterial duct was patent. The diagnosis of double aortic arch with atretic versus interrupted proximal left arch was made based on the lack of antegrade flow from the ascending aorta to the left common carotid artery. Diagnosis was confirmed by computed tomography scan (Figure 4), and minimal narrowing of the trachea was noted. At three months of age, the infant is growing well (50th percentile on growth chart) with no respiratory symptoms. The pulmonary valve stenosis has not progressed.

DISCUSSION

Abnormalities of the aortic arch are rare and represent approximately 1% of congenital cardiac malformations. A vascular ring occurs when the trachea and esophagus are completely encompassed by the aortic arch and its associated vessels. The most common variation is the double aortic arch, accounting for 50% of cases, followed by a right aortic arch with left-sided arterial duct/ligament in 26%. A variety of very rare additional subtypes of vascular rings have been diagnosed, but this usually requires a high index of clinical suspicion, and prospective recognition by echocardiogram is not common in these situations.

In order to fully comprehend the different combinations of vascular malformations that may result in a vascular ring, an understanding of the formation of the normal human aortic arch is imperative. There are five symmetric pharyngeal arches on either side, named 1 through 4 and 6 (the fifth arch has been identified as an incomplete structure in only a solitary human embryo). Through a series of consequential changes, the third, fourth, and sixth arteries give rise to the aortic arch. The third arteries on both sides form the respective common carotid arteries, while the fourth artery on the right remains only as the proximal right subclavian artery. The left fourth artery forms the definitive transverse arch, while the distal left sixth arch remains as the left-sided arterial duct. The distal right sixth regresses. This results in the formation of a left-sided aortic arch with a right innominate artery and a left arterial duct.
When both fourth pharyngeal arches persist, a double aortic arch is encountered. The right arch gives rise to the right common carotid and the right subclavian arteries, while the left arch gives rise to the left common carotid and left subclavian arteries. In our patient, both fourth arches persisted with an atretic or interrupted proximal left fourth arch (Figure 5) and a patent distal left fourth arch and left dorsal aorta. This arch malformation is best understood by a classification system proposed by Garti et al., in which a double arch with left atresia is classified as type A, B, C, or D. In type A, the left arch is atretic distal to the arterial duct/ligament, while in type B, the affected segment is in between the arterial duct and the left subclavian artery. Atresia proximal to the left subclavian artery is type C, and proximal to the left common carotid, as seen in our patient, is type D. In this classification system, the absent segment remains as an atretic ligament, thus forming a complete vascular ring. When the segment is entirely interrupted with no residual ligament, it is categorized as a right aortic arch rather than a double aortic arch. According to Garti’s classification system, our patient has either double aortic arch type D or right aortic arch type D (also known as right aortic arch with retroesophageal left innominate artery), depending on whether the segment is atretic versus interrupted, respectively. Unfortunately, identification of an atretic ligament is possible only via intraoperative or postmortem inspection. Thus, an alternative classification system was proposed by Bogren and Porter, where the term “double aortic arch types A, B, C, or D” included both atresia and interruption of left arch segments. Using this approach, our patient would be classified as double aortic arch with type D interruption versus atresia.

Review of the reported cases in the literature confirms that the diagnosis of atresia versus interruption is not straightforward. In our patient, the presence of a left ductal ligament forms a "loose
ring.” Lack of symptoms at 3 months of age clinically suggests that no atretic ligament is present. However, the descending aorta is midline (Figure 6), and the left common carotid and left subclavian artery arise from a vessel that runs parallel to the right aortic arch with a relatively upward course of both vessels (Figure 7), which has been suggested as evidence of a left arch with an atretic ligament. Regardless of the anatomy of the proximal left arch, our patient demonstrates a vascular ring composed of a patent right arch with a left ligamentum, which may present with symptoms even at an older age.\(^\text{11}\)

To the best of our knowledge, this particular variation of a double aortic arch type D has only been diagnosed twice before: in a 39-year-old man in whom diagnosis was made by cardiac magnetic resonance imaging\(^\text{10}\) and in a 5-day-old infant with aortic atresia in whom the atretic segment of the proximal left arch was not appreciated by echocardiogram.\(^\text{12}\) It is important to note that in the 39-year-old, the diagnosis has not been confirmed intraoperatively.

**Figure 6** (A) Computed tomography scan in a coronal plane demonstrates the thoracic descending aorta (arrow) is at midline relative to the spine. (B) Three-dimensional reconstruction of the chest cavity demonstrates a longitudinal view of the midline descending aorta.

**Figure 7** Three-dimensional reconstruction demonstrates the left transverse arch traveling parallel to the right transverse arch. The left transverse has a relatively horizontal course as the left common carotid and left subclavian arteries arise at a perpendicular angle.

**Figure 8** The origins of the four brachiocephalic vessels in the suprasternal frontal plane with posterior angulation, with the typical appearance of the corners of a rectangle, consistent with a double aortic arch. The midline is depicted with the white line. 1: right common carotid; 2: right subclavian; 3: left common carotid; 4: left subclavian artery.
Diagnosis of vascular rings by echocardiography was vastly improved with the introduction of new techniques by Murdison et al., in which emphasis was placed on delineating the origin and course of the brachiocephalic vessels via suprasternal frontal sweeps. The lack of a bifurcating brachiocephalic vessel should alert the clinician to the presence of an aberrant/isolated subclavian artery or a double aortic arch. In a double arch, the appearance of the origins of the four brachiocephalic vessels is characteristic and described as forming "the corners of a rectangle," as seen in our patient (Figure 8, Video 1). Care must be taken to assess this just superior to the aortic arch(es). The addition of color Doppler allowed visualization of retrograde flow in the patent portion of the left arch, which confirmed the diagnosis of proximal left arch atresia.

In conclusion, we believe echocardiography is sufficient to diagnose rare arch abnormalities but requires attention to detail and a thorough understanding of embryologic arch formation. Additional imaging such as computed tomography scan and/or magnetic resonance imaging may assist in preoperative planning.

SUPPLEMENTARY DATA
Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.01.003.

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