

CASE REPORT

## Coarctation of a Right-Sided Aortic Arch and an Unusual Vascular Ring

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### ABSTRACT

The clinical presentation, diagnostic workup, and surgical therapy of a coarcted right-sided aortic arch associated with a vascular ring are described.

*Key Words:* Coarctation; Right aortic arch; Vascular ring.

### INTRODUCTION

Coarctation of the aorta with right aortic arch is a rare occurrence, with few cases described in the medical literature (Ad and Vidne, 1999; McElhinney et al., 1999). Right aortic arch is found in 0.1% of the population and usually is accompanied by other forms of congenital cardiovascular disease. When intracardiac defects such as tetralogy of Fallot are associated, mirror image branching is usually found. When intracardiac anatomy is normal, usually a diverticulum of Kommerel is found with the left subclavian artery arising from it, creating a vascular ring. Coarctation of

the right aortic arch is extremely rare. A recent new classification of right aortic arch was proposed by Ad and Vidne with a review of cases reported and the type of surgical repair performed (Ad and Vidne, 1999). A type D right aortic arch was described with involution of the proximal left aortic arch and origin of the left common carotid and subclavian artery off the descending aorta. This type of arch has been previously documented (Grollman et al., 1968). We describe the first known case of this arch anomaly and coarctation of the right aortic arch. This anomaly produces a vascular ring completed by a left sided ductus ligamentum.

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## CLINICAL PRESENTATION

The patient was a 7-month-old female noted to have a murmur on routine physical examination at 6 months by the primary care physician. Cardiology consultation was requested and revealed a healthy-appearing 7-month-old with a weight of 10.7 kg (greater than the 95th percentile), height of 73 cm (greater than 95th percentile). Blood pressure was difficult to obtain in any extremity. The chest was symmetric with bilaterally clear breath sounds: no rales, wheezes, or rhonchi. Cardiovascular exam demonstrated a quiet precordium with normal first and second heart sounds and a grade 3/6 medium pitched, harsh systolic murmur maximally heard in the mid back. There were no associated clicks, gallops, or diastolic murmurs. Dermatological exam revealed a pink, lacy, flat vascular birthmark on the right hand and right shoulder but no other rashes were noted. The remainder of the exam was normal.

## Electrocardiogram

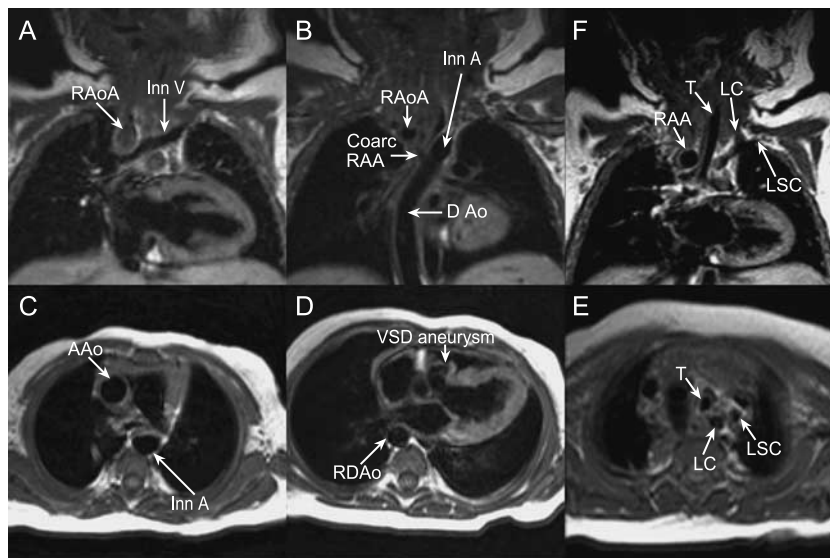
Borderline biventricular hypertrophy.

## Echocardiogram

Demonstrated a moderately hypertrophied left ventricle with a small tissue deficiency in the perimembranous ventricular septum that was covered over by a thin membranous “windsock” preventing any ventricular level shunting. A patent foramen ovale was present with a trivial volume left to right shunting at the atrial level. A mild amount of mitral regurgitation was present. Doppler interrogation of the descending aorta revealed a 4 m/sec velocity with diastolic run-off pattern consistent with coarctation of the aorta. Precise aortic arch anatomy was suboptimally defined.

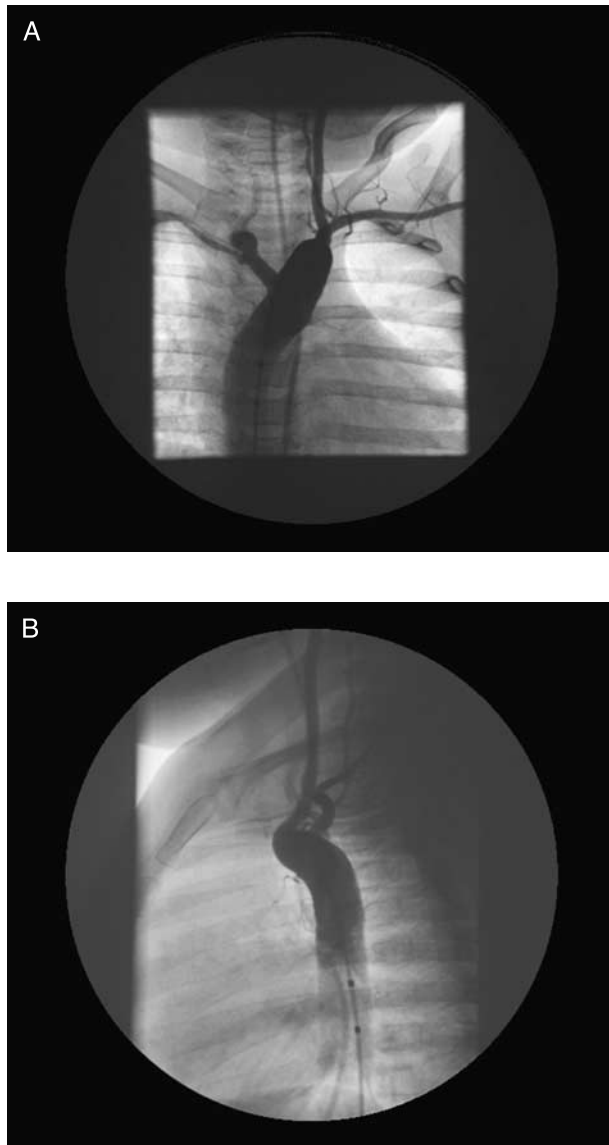
## Magnetic Resonance Imaging

A sedated cardiovascular magnetic resonance imaging/angiography MRI/MRA study was performed on a Phillips 1.5 Tesla magnet using a cardiac Synergy<sup>®</sup> coil. Scout localizing images were performed followed by high-resolution T1 spin echo echo-planar images in the axial and coronal planes. Gradient echo (white blood) studies were performed in similar planes followed by a gadolinium enhanced magnetic



**Figure 1.** Spin echo echo planar T1 weighted images. A: Coronal plane demonstrating the innominate vein (Inn V) coursing underneath the right-sided and ascending aorta (RA<sub>O</sub>A). B: A more posterior coronal view demonstrating coarctation in the right side of the aortic arch (coarc RAA), as well as the proximal innominate artery (Inn A). The oblique course from the patient’s left to right of the thoracic aorta is visualized to the descending aorta (DA<sub>O</sub>). C: Axial image demonstrating the ascending aorta (AA<sub>O</sub>) and the proximal innominate artery (Inn A), which is very posterior. D: Axial image inferior to IC demonstrating not only the ventricular septal defect (VSD) aneurysm, but also the right-sided descending aorta (DA<sub>O</sub>). E: Coronal image slightly posterior to IA demonstrating the branching of the innominate artery into the left carotid (LC) and left subclavian (LSC). The trachea (T) is labeled. F: Axial view slightly superior to IC demonstrating branch of the innominate artery into the left carotid (LC) and left subclavian (LSC). The trachea (T) and right aortic arch (RAA) are seen.

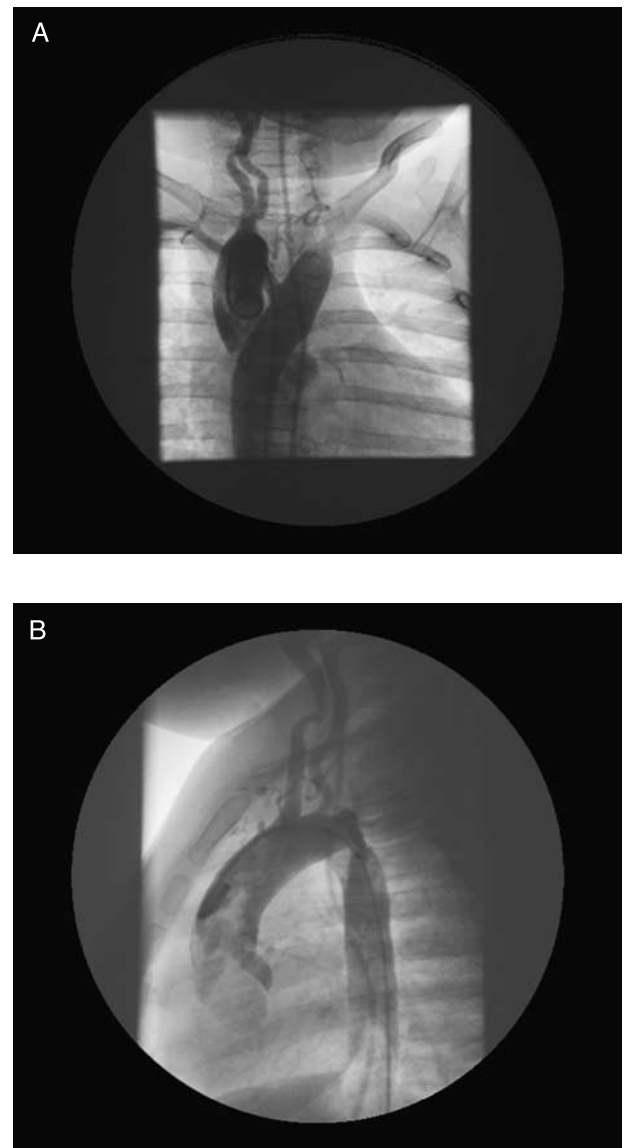




**Figure 2.** A: Anterior/poster A/P and B: lateral aortogram with the catheter positioned in the descending aorta. Best seen in the A/P view is the tubular coarctation of the right arch and proximal stenosis of the right subclavian artery.

resonance angiography with 3D volume reconstruction. Figures 1A, 1B, 1C, and 1D represent selected axial and coronal slices of the T1 spin echo echo-planar images. A right aortic arch with right descending aorta was found with a retroesophageal aberrant innominate artery arising from a diverticulum of the descending aorta, and coarctation of the distal right arch was seen. The left carotid and left subclavian artery are branches of the innominate artery depicted in Figs. 1E, and 1F. A stenosis of the proximal right subclavian artery was suspected from the 3D reconstructed gadolinium

magnetic resonance angiogram. Although not directly seen, a vascular ring was proposed based on a ductus ligament attachment of the diverticulum to the pulmonary artery. In addition to the arterial abnormalities, the innominate vein coursed underneath the right aortic arch prior to entry into the right superior vena cava. Given the rarity of the proposed anatomy and at the request of the surgical team, angiography was performed to confirm the anatomy described at MRI/MRA.



**Figure 3.** Anterior/posterior aortogram demonstrating the distal aortic arch with an NG tube present as a reference point within the esophagus. Structures are identified, including the coarctation of the right-sided aortic arch and a slight narrowing in the right subclavian artery.

**Angiography**

At cardiac catheterization, pressure in the ascending aorta was 125/55 mmHg with mean of 85 compared to a pressure in the descending thoracic aorta below the area of coarctation of 80/50 mmHg with mean of 66. There was a small gradient into the right subclavian artery where the pressure was 75/55 mmHg with mean of 65. This was representative of proximal stenosis of the right subclavian artery with origin in the coarctation segment of the distal right aortic arch. Figures 2 and 3 represent anterior/posterior and lateral aortograms.

**Artist Rendition**

Figure 4 is a diagrammatic depiction of the patient's anatomy.

**Surgical and Clinical Course**

The procedure was approached through a median sternotomy with full exposure of the right aortic arch, left ductus ligament, left-sided diverticulum, and aberrant innominate artery. The right-sided subclavian artery and coarctation were located very posterior.

ductus ligament was ligated and divided and the patient was placed on cardiopulmonary bypass with arterial cannulation in the ascending aorta and in the upper descending aorta. In an effort to avoid prosthetic material, the right aortic arch was transected immediately proximal to the coarcted segment and origin of the right subclavian artery. The right arch was then translocated anteriorly and leftward of the trachea with reconstitution of the aorta by direct anastomosis to the top of the descending aorta with absorbable 5-0 suture material. The right subclavian artery was too short to reimplant in the ascending aorta without an interposition graft and it was therefore left attached to the narrow distal right arch. The patient was readily separated from bypass in normal sinus rhythm.

The patient's postoperative course was very satisfactory. Upper airway problems were anticipated but she was extubated in less than 24 hours without difficulty, despite significant mobilization of both recurrent laryngeal nerves. Oral feeding was advanced slowly without aspiration and the patient was discharged from hospital on the 7th postoperative day. There was no evidence of flow acceleration by transthoracic Doppler flow studies prior to discharge. Full pulses were present in the lower extremities and left arm with a diminished but easily palpable pulse in the right arm. Carotid pulses were full bilaterally.

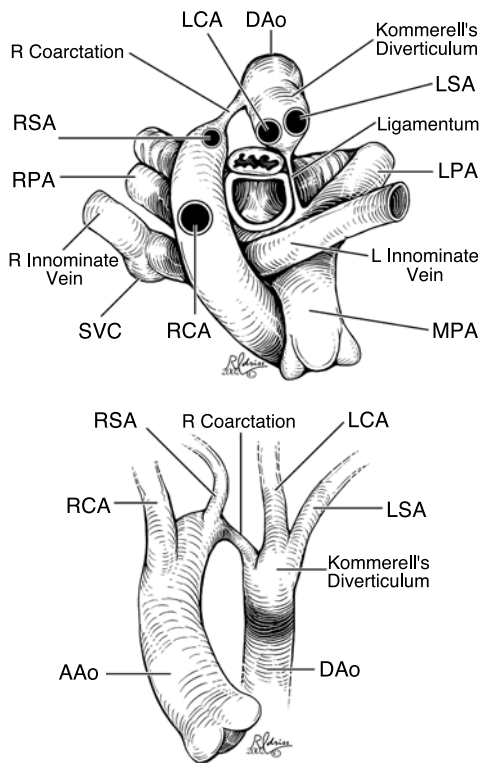


Figure 4. Artist renditions of the patient's anatomy.

**DISCUSSION**

Ad and Vidne described a simplified classification of right aortic arch dependent on the region of involution of the left arch. Table 1 summarizes their classification. Given the rarity of right aortic arch with coarctation, multiple imaging modalities including echocardiography, MRI/MRA, and conventional angiography were performed in the preoperative evaluation of this child. The anatomy implied involution of the left aortic arch proximal to take-off of the left carotid and left subclavian artery. This was described as a type D involution by Ad and Vidne. A left-sided ductal ligament completing a vascular ring predicted from the

Table 1. Classification of right aortic arch.

Type	Site of involution of left arch
A	Distal to left ductus
B	Between left subclavian and left ductus
C	Between left carotid and left subclavian
D	Proximal to left carotid

From Ad and Vidne, 1999.



MRI and conventional angiogram was confirmed at surgery and divided. No evidence of a ligamentous band from the ascending aorta to the innominate artery was found at surgery.

The MRI/MRA provided detailed 3-D anatomy of the unusual aortic anatomy and the relationship to the airway and esophagus. In addition to the above complex anatomy, the patient had an innominate (left brachiocephalic) vein, which coursed under the right aortic arch seen on MR images and confirmed surgically. Although cardiac catheterization with angiography confirmed the anatomy and provided additional hemodynamic data, it would not be used today given our improvements and increased surgical confidence in MRI/MRA.

The surgical correction described above leaves both subclavian arteries arising distal to the arch repair site, and therefore, arm/leg blood pressure will be inadequate for accurate prediction of a future gradient across the arch repair. The right subclavian artery was left in place within the coarcted segment and may possibly need transcatheter balloon dilation in the future. Repeat imaging is planned. The reconstruction was performed without prosthetic material and allowed for division of the left ligamentum, which had completed the vascular ring. The MR and angiographic images depicted this very unusual anatomy and allowed the surgeon to plan a unique translocation of a right aortic arch to left of the trachea to bypass a coarctation in the distal right arch, avoid use of prosthetic material, and relieve an unusual vascular ring.

## CONCLUSION

Right aortic arch associated with coarctation and vascular ring is an extremely rare anomaly. Preoperative identification of the anatomy is mandatory for optimal surgical repair. Magnetic resonance imaging can guide surgical management by providing excellent delineation of the presurgical anatomy in this condition.

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