A 9-month-old infant girl was referred for management of transverse aortic arch hypoplasia in the setting of a right arch with mirror-image branching of the brachiocephalic vessels and an aberrant left subclavian artery originating from the descending aorta. Because she was asymptomatic and without left ventricular hypertrophy on echocardiogram, no interventions were undertaken. At multiple subsequent clinic visits through 4 years of age, extremity pulses and blood pressures continued to be normal and equal despite persistent arch hypoplasia and Doppler evidence of arch obstruction on serial echocardiograms.

At 4.5 years of age, her left radial pulse was noted to be relatively weaker for the first time, but 4-extremity blood pressures remained equal. An echocardiogram with Doppler revealed a continuous systolic-diastolic forward flow waveform in the abdominal aorta. One year later, her left radial pulse remained relatively weak, and her right leg systolic blood pressure was 30 mm Hg lower than that in her upper extremities. These findings prompted magnetic resonance angiography, which revealed a complex form of right aortic arch with moderate transverse aortic arch hypoplasia measuring 6 mm in diameter, narrowing to 4 mm at the isthmus. Distal to the coarctation, the proximal descending aorta was 11 mm in diameter. Both common carotid arteries arose proximal to the hypoplastic transverse aortic arch. There were no significant aortic-intercostal collateral arteries (Figure 1 and online-only Data Supplement Movie I).

Aortic angiography at the time of interventional catheterization confirmed the magnetic resonance angiography findings and also revealed delayed filling of the left vertebral and aberrant left subclavian arteries from collateral circulation, presumably through the circle of Willis (Figure 2 and online-only Data Supplement Movie II). This retrograde flow through the left vertebral artery contributed to blood flow entering the descending aorta (Figure 3 and online-only Data Supplement Movie III).

After reviewing these findings with the cardiovascular surgery team, we elected to place a transcatheter stent across the narrowest part of the coarctation (marked with a * in Figure 2), reducing the pressure gradient from 30 to 10 mm Hg (Figure 4). After the stent was placed, improved antegrade flow in the aberrant left subclavian artery was also established (online-only Data Supplement Movie IV).

Coarctation of a right aortic arch is an extremely rare anomaly. In a review of 240 patients with a right aortic arch, 4.1% had coarctation, of whom 60.0% had an aberrant left subclavian artery or retroesophageal diverticulum of Kommerell similar to this patient.1 Similarly, in a review of 11 276 children with congenital heart disease, whereas 5.7% had coarctation with a left aortic arch, only 0.1% had coarctation with a right aortic arch, of whom 64.0% had an aberrant left subclavian artery.2 In a literature search for specific descriptive reports of a right aortic arch with coarctation, we found only 2 articles totaling 26 patients.3,4 This paucity of cases reflects observations that right-sided arches are much more commonly associated with conotruncal defects with pulmonary outflow obstructive lesions. These defects are typically associated with right-to-left intracardiac shunting and left-to-right shunting across the patent ductus arteriosus during embryonic development, resulting in more forward flow across the juxta ductal aortic arch segment, a watershed region.5 As a result of the increased blood flow across this segment of aortic arch, coarctation is a very uncommon association in conotruncal defects, such as tetralogy of Fallot. The intracardiac anatomy in this patient, however, was normal.

Although coarctation of the aorta can be diagnosed by imaging, the decision to intervene is typically based on clinical data. In cases of severe coarctation, infants often present in left heart failure or even cardiogenic shock, requiring immediate intervention. In older children or less severe cases, coarctation tends to be asymptomatic or to have only vague symptoms of claudication even when hypertension or left ventricular hypertrophy is present. In these cases, the hemodynamic consequences develop over time, and the cardiovascular system has time to compensate. In such individuals, the diagnosis is suspected when there is a discrepancy in pulses or blood pressures between the upper versus lower extremities. Therefore, the routine physical examination of children to rule out coarctation of the aorta traditionally includes a comparison of these findings in the
right arm and either of the lower extremities. However, certain assumptions are made in this approach. First, one or both of the subclavian arteries (at least the right) arise proximal to the site of coarctation, which is true for most left-sided aortic arch forms. Second, collateralization has not formed to the degree that lower extremity pulsation is sufficient enough to mask differences in pulse amplitude and timing (causing delay).

We present an unusual case of severe coarctation of the aorta associated with rare anatomy that masked the severity of the obstruction for several years. Because neither subclavian artery arose proximal to the coarctation, neither arm’s pulse represented ascending aortic pressure. Perfusion of the lower extremities was supported by retrograde flow in the left subclavian artery via a vertebral artery steal phenomenon. This case illustrates a potential pitfall of an abbreviated examination to rule out coarctation of the aorta when it includes only the right arm and lower extremities. Examining carotid pulses should be part of the routine cardiovascular examination in children, and discrepant pulses from any extremity should lead to further evaluation for coarctation or other arch anomalies.

Figure 1. Three-dimensional (3D) reconstruction of magnetic resonance angiography (MRA) showing the aortic coarctation (marked with a *) and the anatomy of the brachiocephalic arteries from the left posterolateral view. The severity of the coarctation is exaggerated because of the 3D reconstruction algorithm, as well as the resolution limits of the original MRA sequence. Figure shows the ascending aorta (AAo), left common carotid artery (LCCA), right common carotid artery (RCCA), right vertebral artery (RVA), right subclavian artery (RSA), aberrant left subclavian artery (LSA), and descending aorta (DAo).

Figure 2. Aortic angiogram from the right anterior oblique view, showing the aortic coarctation (marked with a *) and the anatomy of the brachiocephalic arteries. The aberrant left subclavian artery, which branches distal to the coarctation, is not visualized because of washout from retrograde filling via the left vertebral artery. Figure shows the left common carotid artery (LCCA), right common carotid artery (RCCA), right vertebral artery (RVA), right subclavian artery (RSA), and descending aorta (DAo).

Figure 3. Angiograms from the same right anterior oblique view, showing the aberrant left subclavian artery (LSA) revealed by direct injection into the diverticulum of Kommerell (DoK). Image A was obtained just before image B, in which layering of contrast can be seen in the LSA because of competitive flow via the left vertebral artery (LVA).

Figure 4. Angiogram from the right anterior oblique view, showing the position of the transcatheter stent (S) placed across the coarctation. The stent is located between the origin of the right subclavian artery (RSA) and the diverticulum of Kommerell (DoK). Figure shows the ascending aorta (AAo), right vertebral artery (RVA), and descending aorta (DAo).
Disclosures

None.

References


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