Case Reports

Coarctation of Persistent 5th Aortic Arch: First Report of Catheter-Based Intervention

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Persistent 5th aortic arch, originally called double-lumen aortic arch, is a rarely reported cardiac developmental anomaly that results in systemic-to-systemic or systemic-to-pulmonary shunting. When this malformation occurs, other intracardiac defects are almost always present. We report the case of a 7-month-old girl who presented with a heart murmur; she was found to have an interrupted 4th aortic arch and coarctation of a persistent 5th aortic arch, with no other defects. To our knowledge, this is the 3rd report of a pediatric patient with this condition in isolation, and the first report of its diagnosis and treatment by means of cardiac catheterization and balloon angioplasty. ***(Tex Heart Inst J 2014;41(4):411-3)***

Persistent 5th aortic arch, an apparently rare congenital anomaly, is defined as a single aortic arch with separate superior and inferior parts.[1] It was initially called a double-lumen aortic arch.[2] The 5th aortic arch forms between the true aortic arch (a derivative of the 4th aortic arch) and the pulmonary artery (a derivative of the 6th aortic arch).[3] Without additional coarctation of the existing aorta, this anomaly alone has no physiologic significance and does not necessitate surgical intervention.

We report our catheter-based treatment of an infant who had aortic coarctation of a persistent 5th aortic arch with no associated cardiac defects.

Case Report

In July 2012, a 7-month-old infant girl was referred to our department for evaluation of a heart murmur. The healthy-looking patient had a normal prenatal and postnatal history. Her blood pressure was normal, and her systemic oxygen saturation was 98% on room air. Auscultation revealed normal heart sounds and a grade 2/6 midsystolic murmur that was audible at the 3rd left sternal border and at the back. The patient's diminished femoral pulses raised the suspicion of aortic coarctation.

Electrocardiographic results were normal. Echocardiograms showed left ventricular hypertrophy and an unusual double-lumen aortic arch. There was coarctation of the inferior part of the arch (Fig. 1), and mild obstruction was evident upon color-flow Doppler analysis. Continuous-wave Doppler images revealed a 50-mmHg peak instantaneous gradient at the coarctation site, with typical diastolic runoff. Conventional angiograms showed that the common origin of all brachiocephalic vessels from the ascending aorta ended after the left subclavian artery. A distinct vessel in the inferior part of the interrupted arch originated from the ascending aorta and connected with the descending aorta (Fig. 2). We concluded that this lower arch was a persistent 5th aortic arch. The definitive diagnosis was interrupted 4th arch with persistent 5th arch. The coarctation was at the distal end of the 5th arch.

During catheterization, after consulting a cardiac surgeon, we performed balloon angioplasty to correct the coarctation. Afterwards, the peak-to-peak gradient decreased from 50 to 20 mmHg (Fig. 3). At age 1 year, the patient was normotensive. Echocardiograms revealed mild repeat coarctation at the distal end of the persistent 5th arch and a 30-mmHg peak instantaneous gradient across the coarctation site. As of June 2014, we continue to monitor the patient for hypertension and any increase in the gradient. Surgical intervention might be necessary in the future.

Key words: *Aorta, thoracic/abnormalities; aortic arch syndromes/congenital/radiography; aortic coarctation/diagnosis; cardiac catheterization/methods; heart defects, congenital/diagnosis/ultrasonography; treatment outcome*

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Discussion

In our patient, we performed corrective balloon angioplasty during catheterization. To our knowledge, this is the first report of catheter-based intervention for isolated 5th aortic arch coarctation in a pediatric patient.

Persistent 5th aortic arch is a rarely reported developmental anomaly that can manifest itself in various congenital malformations, such as double-lumen aortic arch in which a "subway" vessel occurs beneath the normal aortic (embryonic 4th) arch; interruption of the 4th aortic arch; and a systemic-to-pulmonary arterial connection between the ascending aorta and a derivative of the 6th arch (usually a left pulmonary connection).[3,4] The persistent 5th arch is between the true aortic arch and the pulmonary artery, and no arterial trunks originate from it.[5] Although the aortic arch is subdivided into 2 parts, it does not result in a vascular ring that surrounds the trachea and esophagus. Persistent 5th aortic arch has been classified into 3 variants: 1) double-lumen aortic arch with both lumina patent, 2) interruption of the superior arch with persistence of the 5th arch (inferior part), and 3) systemic-to-pulmonary arterial connection.[6] The first formation appears to be the most common. The clinical presentation is usually silent in patients with the first variant; however, patients with the 2nd variant frequently have coarctation of the distal end of the 5th embryonic arch.[7]

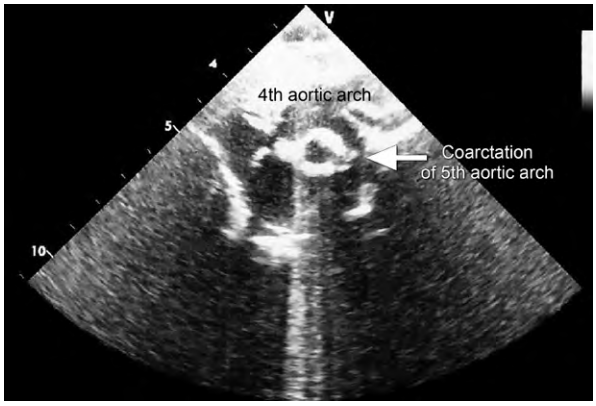

Fig. 1 *Echocardiogram shows left ventricular hypertrophy and a double-lumen aortic arch with coarctation at its inferior part.*

The presentation of the persistent 5th arch depends on the anatomic connections and the associated cardiac defects.[8] The persistent arch is frequently associated with tetralogy of Fallot,[9] complete D-transposition of the great arteries,[6] truncus arteriosus,[10] and ventricular septal defect.[11] Ventricular septal defect is reported most frequently.[12] Double-lumen aortic arch has been diagnosed incidentally.[13] In the presence of pulmonary atresia or true arch interruption, the persistent 5th arch serves as an alternative connection that helps patients to survive.[8] We found only 2 reports of pediatric patients who had an interrupted 4th aortic arch, a persistent 5th arch, and no associated intracardiac defects.[14,15] Our patient's murmur was caused by coarctation of the persistent 5th arch.

Bernheimer and colleagues[11] suggested that echocardiography could enable an accurate diagnosis; however, the condition can be misdiagnosed, because the complex anatomy of the 5th arch can make the study

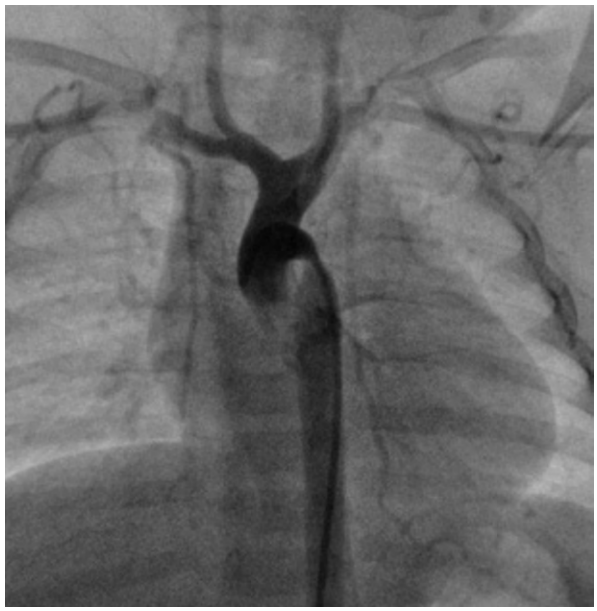

Fig. 2 *Before balloon angioplasty, angiogram shows a distinct vessel in the inferior interrupted aortic arch, originating from the ascending aorta and connecting to the descending aorta.*

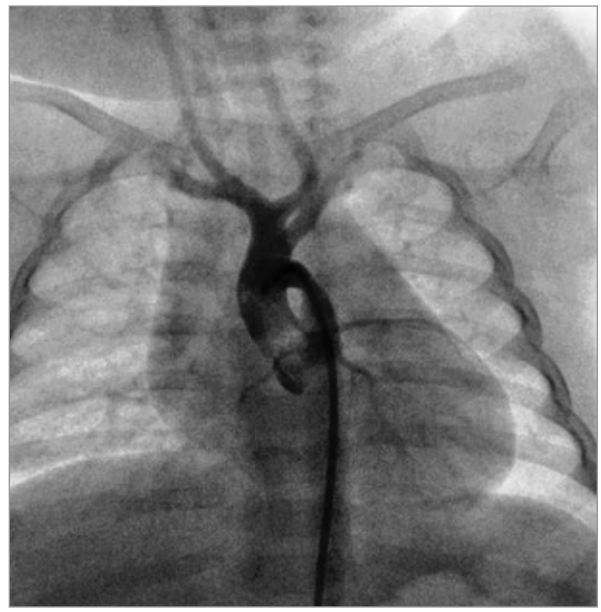

Fig. 3 *After balloon angioplasty, angiogram shows improvement in the obstruction at the coarctation site.*

difficult to interpret. Indeed, persistent 5th aortic arch might be underdiagnosed because of difficulties in echocardiographic evaluation, especially when both lumina are patent.[12,16] Regardless, echocardiography accurately reveals any associated defects. To confirm the diagnosis, conventional angiography[1] or magnetic resonance angiography[17] should also be performed. In our patient, echocardiograms showed a rare double-lumen arch, and we used conventional angiography for definitive diagnosis.

Various methods to repair symptomatic coarctation of persistent 5th arch have been reported.[17] These include patch aortoplasty, end-to-end anastomosis between the descending aorta and the orifice of the 5th arch, and use of a GORE-TEX® tube-graft (W.L. Gore & Associates; Flagstaff, Ariz). A 28-year-old patient with isolated persistent 5th arch and interrupted 4th arch underwent successful graft interposition.[18] The current American Heart Association guidelines state that balloon angioplasty is recommended in children older than 4 to 6 months of age if the gradient exceeds 20 mmHg and the patient's anatomy is suitable.[19,20]

It is crucial to keep persistent 5th aortic arch in mind, especially in patients who have coarctation and pulmonary atresia. To our knowledge, our pediatric patient is only the third to have been diagnosed with interrupted 4th arch, coarctation of a persistent 5th arch, and no other intracardiac defects. We found that balloon angioplasty to correct the coarctation could be performed successfully.

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