Occlusion of the Left Main Coronary Artery Os By a Tethered Aortic Valve Cusp

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We report a 4-month-old girl who presented with poor ventricular function and was found to have occlusion of the left main coronary artery os by a tethered aortic cusp. The patient underwent surgical delamination of the aortic valve leaflet, revealing a normal left coronary os. After operative repair, her left ventricular function improved significantly. This diagnosis should be included in the differential in all children presenting with cardiac dysfunction.


The presentation of poor left ventricular (LV) function with mitral regurgitation (MR) in a child aged younger than 1 year is most commonly associated with an anomalous left coronary artery from the pulmonary artery. Other less common diagnoses in this age group include stenosis or atresia of the left main coronary artery (LMCA). Regardless of the cause of coronary insufficiency, MR and LV function can improve significantly if coronary artery repair is feasible; however, knowledge of the cause of coronary artery insufficiency is integral to a successful repair. We report a 4-month-old girl who presented with poor LV function and was found to have occlusion of the LMCA by a tethered aortic cusp. To our knowledge, this is the youngest symptomatic patient reported in the literature.

A 4-month-old girl was referred for a murmur evaluation. The initial examination demonstrated a II/VI blowing systolic murmur at the left lower sternal border. An electrocardiogram showed LV hypertrophy, ST depression in the midprecordial leads, and lateral T-wave changes consistent with ischemia. An echocardiogram demonstrated left atrial and LV dilatation, a decreased LV ejection fraction (LVEF) of 0.30, increased echogenicity of the mitral valve apparatus and portions of the LV endocardium, and mild MR (Fig 1A; Videos 1, 2). The left coronary artery appeared small and without demonstrable antegrade flow (Fig 1B, Video 3). There was abnormal motion of the left coronary cusp of the aortic valve and an eccentric jet of aortic insufficiency (Fig 1C, Video 3). This constellation of findings suggested the diagnosis of occlusion of the LMCA by a tethered left coronary cusp; other diagnoses considered included LMCA stenosis, LMCA atresia, and anomalous left coronary artery from the pulmonary artery.

Cardiac catheterization demonstrated a mixed venous saturation of 60% and an elevated LV end diastolic pressure of 11 mm Hg but otherwise normal hemodynamic data. Selective coronary angiography demonstrated retrograde fill of the left coronary artery from the right coronary artery, with no visualized aortic origin of the LMCA. Angiographic findings were nondiagnostic.

Treatment options considered included cardiac transplantation and surgical exploration of the aortic root. Additional transesophageal images (Fig 2, Video 4) confirmed occlusion of the LMCA by a tethered aortic cusp.

Surgical exploration demonstrated normal right and noncoronary cusps, but the left coronary cusp was multilobe and bulbous. It was subdivided such that half of the leaflet had failed to delamate from the aortic wall, and a dimple was present where the os of the LMCA should have been (Fig 3A). The aortic leaflet was delaminated with a #15 blade (Fig 3B), exposing a normal 2-mm LMCA os (Fig 3C). Partial commissuroplasties of the left/right and left/noncommissures were undertaken to encourage the newly delaminated leaflets to remain separated from the aortic wall (Fig 3D). Pericardium-reinforced sutures of 8–0 Prolene (Ethicon, Somerville, NJ) were used to fortify the leading edge of the leaflet.

The patient’s postoperative course was uncomplicated. She was extubated on postoperative day 1 and discharged home on postoperative day 6. The discharge echocardiogram demonstrated antegrade LMCA flow, trivial aortic insufficiency, and an improved LVEF of 0.46 (Fig 4; Videos 5, 6).

Comment

Occlusion of a coronary os by a tethered aortic cusp is very rare, has usually been described in older children and adults, and typically affects the LMCA, though right coronary artery occlusion has also been reported [1–4]. The lesion is often associated with supravalvar aortic stenosis or other aortic valve abnormalities but can occur as an isolated defect [4–6]. Before this report, the youngest symptomatic patient was aged 4 years, although the lesion was reported as an incidental finding in a neonate with complex congenital heart disease [7]. Presenting signs and symptoms can include MR, chest pain, dyspnea, myocardial infarction, syncope, and sudden death. We report the youngest symptomatic patient and the youngest patient with isolated disease.

The videos can be viewed in the online version of this article [http://dx.doi.org/10.1016/j.athoracsur.2013.10.108] on http://www.annalsthoracsurgery.org.
Occlusion of a coronary os by a tethered aortic cusp most likely results from a failure of cavitation of the left coronary cusp leading to obstruction of the LMCA os [7, 8]. Timing of presentation depends on the extent of coronary artery collateralization. Our patient had functional atresia of the LMCA and presumably had less collateralization, both of which likely contributed to her early presentation with audible MR.

Diagnosis can be made with echocardiography and cardiac catheterization. Echocardiography demonstrates an abnormal, often rudimentary, aortic valve cusp and abnormal flow patterns in the left coronary system. As in this patient, transesophageal echocardiography can better visualize the aortic valve leaflets, potentially facilitating diagnosis. At catheterization, the LMCA cannot be cannulated, and a right coronary artery injection usually demonstrates collateral flow to the left coronary artery. Classically, a “pouch” that separates the left coronary os from the rest of the aortic root can be filled with contrast retrograde after an injection into the right coronary artery. As in our patient, this “pouch” is not always present.

Surgical correction is the preferred treatment, and the procedure depends largely on the aortic valve anatomy. If technically feasible, aortic valve repair with delamination of the tethered cusp should be undertaken. If aortic valve

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**Fig 1.** (A) Apical 4-chamber view demonstrates increased echogenicity of the left ventricular endocardium and mitral valve chordal apparatus. Color flow mapping demonstrates mild mitral regurgitation. (B) A short-axis image demonstrates a small left main coronary artery and left anterior descending artery (arrowheads). (C) A short-axis color compare image demonstrates a hypoplastic, tethered left coronary cusp and the resultant posterior regurgitant orifice.

**Fig 2.** (A) Transesophageal echocardiography at level of the aortic valve leaflets more clearly demonstrates a hypoplastic left aortic cusp with a posterior regurgitant orifice. Color flow mapping demonstrates retrograde flow in the left main coronary artery (LMCA). (B) Illustration demonstrates the regurgitant orifice and complete obstruction of the os of the LMCA. (RCA = right coronary artery.)

**Fig 3.** Illustration demonstrates operative repair. (A) A partially tethered left coronary artery cusp results in obstruction of the left main coronary artery (LMCA) os, with resultant dimple. (B) The aortic leaflet is delaminated with a #15 blade, (C) exposing a normal LMCA os. (D) Partial commissuroplasty of the left/right and left/noncommissures encourages separation of the newly delaminated leaflet from the aortic wall.
repair is unsuccessful or not feasible, replacement with a Ross or mechanical valve is the preferred operation in the pediatric population. Some authors have advocated the Bentall procedure in all patients [4]; however, we recommend avoiding this more invasive approach in the pediatric population when possible.

The patient we present had an LVEF of 0.30 and, once the diagnosis of anomalous left coronary artery from the pulmonary artery was ruled out, was considered for cardiac transplantation. Diagnosing her condition correctly and performing a corrective operation allowed her to recover without transplantation. We recommend that this lesion be included on the differential for all children presenting with poor cardiac function. When the diagnosis is not definitive by transthoracic echocardiography, transesophageal echocardiography should be considered as an alternate imaging modality.

References

Fig 4. A postoperative short-axis image demonstrates antegrade flow in left main and the left anterior descending coronary arteries.