Congenital mitral stenosis: A rare presentation and novel approach to management

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Congenital mitral stenosis is an uncommon congenital heart defect, typically presenting with symptoms in early childhood.1,2 This condition remains a surgical challenge, with a trend toward early single-stage complete repair.3 We present the case of a 15-year-old patient given a diagnosis of severe mitral stenosis and pulmonary hypertension. The patient underwent successful complex mitral valve repair with mitral valve ring annuloplasty and neochordae placement and achieved dramatic improvement in symptoms and quality of life.

Clinical Summary
A 15-year-old boy with a history of asthma was transferred to our hospital with apparent status asthmaticus. He was initially treated for continued shortness of breath and had difficulty attending school.

Follow-up echocardiography indicated a significant decrease in estimated right ventricular and pulmonary artery systolic pressure (60-65 mm Hg). Severe mitral stenosis was again evident, along with significant tricuspid valve regurgitation. His pressure half-time was 238 ms, with a peak gradient of 49 mm Hg and an estimated mitral valve area of 0.9 cm². The posterior leaflet was tethered to the free wall of the left ventricle, and the anterior leaflet was tethered to the tips of the papillary muscles, with essentially no chordae tendineae. The leaflets were normal in appearance. The valve orifice was fixed and severely restrictive.

At the time of the operation, the echocardiographic diagnoses were confirmed. The patient underwent a complex mitral valve repair within weeks of his initial presentation by using the following techniques: (1) transection of trapped chordae tendineae to both the anterior and posterior mitral valve leaflets from both papillary muscles, leaving normal-sized and normal-appearing pliable leaflets without any chordae tendineae; (2) mitral valve ring annuloplasty with a no. 26 Cosgrove annuloplasty ring; and (3) placement of multiple neochordae (5-0 Gore-Tex polytetrafluoroethylene sutures; W.L. Gore and Associates, Flagstaff, Ariz), 2 to the anterior leaflet and 3 to the posterior leaflet. Neochordae were used because of the congenital absence of functional chordae.

After the procedure, the patient had a dramatic reduction in tricuspid regurgitation. He was able to resume activities normal for an adolescent boy without shortness of breath. A postoperative echocardiogram was obtained. The echocardiogram demonstrated no residual mitral stenosis and a trivial jet of mitral valve regurgitation. The Doppler estimated right ventricular systolic pressure was 49 mm Hg. Moderate tricuspid valve regurgitation was noted. Mitral valve flow on Doppler scanning was 178 cm/s. Our patient continued to be asymptomatic at 1-year follow-up.
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cimitar syndrome is a congenital cardiac malformation in which pulmonary flow from the right lung is partially or totally drained via a venous channel that joins the inferior vena cava (IVC), usually just below the level of the diaphragm. Options for repair include the creation of a long baffle from the orifice of the scimitar vein within the IVC to the atrial septal defect (ASD) that directs the anomalous pulmonary venous flow to the left atrium, division with reimplantation of the scimitar vein into the right atrium with an intra-atrial baffle that directs blood flow through an ASD to the left atrium, partitioning of the IVC into anterior and posterior compartments with a pericardial baffle that channels pulmonary venous flow directly into the left atrium, or direct anastomosis of the divided scimitar vein to the left atrium. These techniques may be complicated by obstruction of the scimitar vein owing to either baffle thrombosis or kinking of the venous channel. We describe an alternative method for repair of scimitar syndrome using a short polytetrafluoroethylene (Gore-Tex; W. L. Gore & Associates Inc, Flagstaff, Ariz) interposition tube graft to ensure freedom from tension or kinking when directly connecting the scimitar vein to the left atrium.

Clinical Summary

Two patients aged 4 and 14 years have undergone repair for scimitar syndrome with a pulmonary venous extension technique. Both patients had the entire venous return from the right lung via the scimitar vein with a relatively short length of vein from the hilum of the lung to the junction with the IVC. Both patients had a secundum ASD. Preoperative echocardiogram showed right atrial and ventricular dilation. Cardiac catheterization demonstrated the anomalous pulmonary venous return from the right lung associated with an ASD. The pulmonary/systemic flow ratio was 2.6:1 (4-year-old) and 1.4:1 (14-year-old).

Cardiopulmonary bypass with bivacal cannulation was established through a median sternotomy. The patients were cooled before aortic crossclamping and administration of cold blood cardioplegic solution. The scimitar vein was isolated from its point of emergence from the pulmonary parenchyma to its junction with the IVC near the reflection of the diaphragm. Despite dividing the scimitar vein flush with the IVC, there was concern that a direct anastomosis to the left atrium would create acute angulation and kinking. Therefore, a short polytetrafluoroethylene interposition graft (12-mm in the 4-year-old and 16-mm in the 14-year-old) was sewn end to end to the divided scimitar vein; then, through a right atriotomy and via the ASD, the rightward superior lateral aspect of the left atrium was probed and an atriotomy was made. The polytetrafluoroethylene interposition graft was cut to appropriate length and anastomosed end to side to the left atriotomy (Figure 1). The ASD was closed with an autologous pericardial patch. After repair, an

References


