Figure 2. Echocardiogram at 51 months follow-up. **RVOT**, Right ventricular outflow tract; **RA**, right atrium; **AO**, aorta; **PA**, pulmonary artery; **RV**, right ventricle.

The ventriculoarterial septal defect that we describe in our patient is a combination of a subcoronary window between the aortic and pulmonary valvular sinuses and a DcVSD, with the leaflets of the arterial valves in fibrous continuity and bridging the two defects. The window present between the sinuses of the arterial roots is the consequence of incomplete separation during development of the intermediate part of the developing outflow tract.^{1,2} It would be a mistake to consider the lesion as a deficiency of aortopulmonary septation, not only because, in the normal heart, there is no septum between the aorta and the pulmonary trunk, each possessing its own discrete walls,^{1,2} but also because the intrapericardial trunks in our patient were indeed separate one from the other. The abnormal communication represents inappropriate fusion of the cushions that normally separate the developing aortic and pulmonary valves within their persisting cuff of outflow myocardium.¹

The DcVSD is the consequence of failure of muscularization of the proximal parts of the outlet cushions, which during normal development become converted to the free-standing subpulmonary infundibulum. The upper margin of such a defect is formed by the conjoined leaflets of the pulmonary and aortic valves, which are at the same level, and are separated only by a thin rim of fibrous tissue. In our patient, the confluence of the aortopulmonary sinus window and the interventricular communication was spanned by the conjoined leaflets of the arterial valves, just as rarely the bridging leaflets of the valve found in hearts with common atrioventricular junction can be fused to each other, producing separate right and left atrioventricular orifices, yet still float within the atrioventricular septal defect.⁴

The use of a DKS anastomosis to “avoid” creating aortic regurgitation by closure of the DcVSD and subcoronary window is a difficult choice. It would seem that the patient is still at risk for regurgitation with a DKS anastomosis because there is no support for a portion of the aortic valve. We did consider closing the DcVSD and window on the pulmonary side. However, partitioning the outlet valves with only a thin rim of fibrous tissue in continuity is unpredictable in terms of valvular competency. The fallback option of aortic valve substitution in a small infant is rather unfavorable.

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Papillary muscle realignment for symptomatic left ventricular outflow tract obstruction

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Video clips are available online.

Left ventricular outflow tract obstruction (LVOTO) in hypertrophic cardiomyopathy typically results from proximal septal hypertrophy and systolic anterior motion (SAM) of the anterior mitral leaflet caused by the Venturi effect.¹ Mitral valve and subvalvular anomalies can also produce LVOTO.² Reported are 2 patients with abnormal orientation of the papillary muscles resulting in LVOTO treated with the technique of transaortic papillary muscle realignment.

Figure 1. Cardiac mitral regurgitation before (A) and after (B) the papillary muscle realignment procedure (see supplemental video clips online).

Clinical Summary

A 35-year-old man presented with exertional dyspnea and a provokable gradient of 100 mm Hg on transthoracic echocardiography. SAM of the mitral leaflet was evident on transesophageal echocardiography and magnetic resonance imaging. The intraventricular septum was 1.8 cm thick. Intraoperatively, the anterolateral and posteromedial papillary muscles had multiple heads, with the anterior or septal-most head pointing directly along the axis of the aortic valve rather than that of the mitral valve (Figure 1, A). There also was a great deal of laxity of the papillary muscles, with only a small apical attachment.

A 53-year-old man presented with shortness of breath and a provokable gradient of 184 mm Hg on transthoracic echocardiography. SAM was evident, and there was moderate (2+) mitral regurgitation. The intraventricular septum was 2 cm thick. In addition to a myectomy, the anterior leaflet was shortened at the free edge of the A2 segment by 1 cm. It was also noted that the medial head of the anterolateral papillary muscle pointed along the aortic valve axis and was also very lax. After the procedure, intraoperative isoproterenol (Isuprel) provocation produced a peak gradient of 64 mm Hg, with severe SAM and mitral regurgitation. The septal thickness was 1.5 cm. An additional 3- to 4-mm myectomy was performed, and the anterolateral papillary muscle was realigned.

Figure 2. Papillary muscle realignment procedure. A, Anomalous papillary muscle head directed toward the aortic valve, causing left ventricular outflow tract obstruction by systolic anterior motion of the anterior mitral leaflet. B, Pledgetted mattress stitches are placed to change the orientation of the anomalous papillary muscle. C, Completed procedure.

The realignment procedure was performed through the aortic valve. A pledgetted mattress suture is placed posterior to the most posterior papillary muscle head, avoiding entanglement with the chordae. It is then passed through the anterior head, avoiding twisting of the papillary muscle. One or more mattress sutures might be necessary to realign the papillary muscles, and both the anterolateral and posteromedial papillary muscles have been realigned (Figure 2). LVOTO was eliminated without the development of mitral regurgitation.

Discussion

Orientation of the papillary muscles and their attachment relative to the left ventricular outflow tract is another possible cause of severe SAM of the mitral valve and LVOTO.³ Abnormalities of the mitral valve have been reported in as many as 20% of patients undergoing surgical intervention for hypertrophic obstructive cardiomyopathy.⁴ In 1991, Klues and colleagues² reported anomalous insertion of the left ventricular papillary muscles in 13% of patients. Direct insertion of both or either papillary muscle into the anterior mitral leaflet resulted in left ventricular outflow tract gradients of 70 to 150 mm Hg. The papillary muscle abnormality we have described must be added to the now numerous mitral valve abnormalities associated with hypertrophic cardiomyopathy and should be considered a potential cause of LVOTO.

In the cases presented, the orientation of 1 or more papillary muscle heads along the aortic valve axis appeared to contribute to LVOTO. By realigning the papillary muscle toward the mitral valve axis, we easily eliminated the obstruction (Figure 1, B). Thus the papillary muscle realignment procedure we have described is easily accomplished through the aortic valve, eliminates SAM, and preserves mitral valve function.

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