Encouraged by the favorable results of this limited series, we evaluate all patients with thoracic aortic aneurysms extending proximal and distal of the left subclavian artery for this new treatment modality, the frozen elephant trunk technique.

References

Mitral valve-sparing operation in subaortic stenosis caused by anomalous papillary muscle and discrete subaortic stenosis

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Left ventricular outflow tract (LVOT) obstruction caused by subaortic stenosis covers a wide range of anatomic lesions. Most cases in children were caused by the discrete subaortic stenosis (DSAS). In adult patients LVOT obstruction caused by abnormal insertion of papillary muscle into the base of the anterior mitral leaflet in hypertrophic cardiomyopathy has been reported. Isolated LVOT obstruction caused by an anomalous papillary muscle (APM) has been recognized also. The procedures, including entire resection of the APM and mitral valve replacement, have been thought necessary for the successful correction. Report of LVOT obstruction caused by the combination of DSAS and APM is rare. We report on such a patient who was treated with extensive resection of the APM, resection of the DSAS, and careful mobilization of the anterior mitral leaflet, resulting in successful relief of the obstruction without mitral valve replacement.

Clinical Summary
A 48-year-old woman presented with progressive shortness of breath on exertion and chest tightness over a period of 3 years. Holter monitoring showed sinus rhythm. A 12-lead electrocardiogram revealed voltage criteria for left ventricular hypertrophy with strain pattern. On auscultation, a grade 4/6 harsh systolic murmur maximal at the right upper sternal border was audible. Transthoracic echocardiography showed that the LVOT was obstructed by the membranous DSAS and an anomalous insertion of a papillary muscle to the anterior mitral leaflet with a peak instantaneous gradient of 90 mm Hg (Figure 1). The aortic valve had 3 cusps and was normal in size and appearance. The ventricular septum and the left ventricular posterior wall were mildly thickened. Neither systolic anterior motion nor localized hypertrophy of the ventricular septum was noted. Coronary angiographic results were normal.

The operation was performed through a lower half ministernotomy with standard cardiopulmonary bypass and cold oxygenated crystalloid cardioplegic arrest. On inspection through the oblique aortotomy, the aortic valve was found to be mildly sclerotic and competent. The LVOT was explored, and a papillary muscle 12 mm in diameter was found extending from the anterior mitral leaflet to the apicoseptal portion of the left ventricle without any chordal interposition. A membranous DSAS 6 mm in width was found just below the left coronary cusp as well (Figure 2). The APM and the DSAS were resected. Then an extensive myectomy trough was carried within the left ventricular midcavity. Finally, the anterior mitral leaflet was fully mobilized, with careful shaving of the residual papillary muscle and the secondary chords, which were attached over the ventricular surface of the leaflet (Figure 3). Cardiopulmonary bypass was discontinued unevent-
fully, and the gradient across the LVOT measured directly was 22 mm Hg. Predischarge echocardiography on the seventh postoperative day confirmed a peak instantaneous gradient across the LVOT of 36 mm Hg, with trivial mitral and aortic regurgitation. Follow-up transthoracic echocardiography 3 weeks after the operation revealed the gradient regressed to 25.4 mm Hg, and the LVOT was not obstructive and 15.5 mm in diameter (Figure 4). Neither systolic anterior motion nor mitral regurgitation was noted. The patient has been drug free since then.

Discussion

LVOT obstruction caused by subaortic stenosis is a complex subject because of the peculiarities and a wide range of anatomic features of the LVOT. In children varying degrees of severity have been described, ranging from a simple membranous DSAS to a tunnel subaortic stenosis. Abnormalities of the subvalvular structures have been implicated in causing significant subaortic stenosis. Most commonly, in hypertrophic cardiomyopathy the thickened septum affects the systolic flow in the LVOT, resulting in systolic anterior motion of the anterior mitral leaftet. Some muscular structures, such as APM in isolated form, in association with hypertrophic cardiomyopathy or an abnormal mitral muscle band, have been well documented to cause LVOT obstruction, although they are rare. Our patient has a very unusual cause of LVOT obstruction. Del Guzzo and Sherrid have documented the only report regarding LVOT obstruction caused by the combination of DSAS and APM in an adult female patient; unfortunately, she died 1 day after admission without operation.

In the surgical treatment of hypertrophic cardiomyopathy and severe LVOT obstruction caused by APM, Maron and associates

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Figure 1. Parasternal long-axis echocardiogram showing the APM (arrowhead) directly inserted into the anterior mitral leaftet. The membranous DSAS (arrow) was also noted.

Figure 2. Operative picture of the aortic root showing the APM, extending from the anterior mitral leaftet to the apicoseptal portion of the left ventricle. The membranous DSAS just below the left coronary cusp was demonstrated as well. AML, Anterior mitral leaftet; NCC, noncoronary cusp.

Figure 3. Operative picture of the aortic root after resection of the DSAS, resection of the APM, and creation of the extensive myectomy trough (arrows) with anterior mitral leaftet mobilization revealing a widely opened LVOT. AML, Anterior mitral leaftet; NCC, noncoronary cusp.

Figure 4. Postoperative parasternal long-axis echocardiogram showing the completely removed APM and the DSAS. The measured LVOT diameter is 15.5 mm. Neither systolic anterior motion nor mitral regurgitation was noted.
have designed a technique in which an extensive myectomy trough was created to remove the outflow gradient in 2 patients. They also partially severed the papillary muscle to increase mobility of the mitral apparatus in 1 patient (mobilization of the anterior mitral leaflet). On the other hand, complete resection of the APM and mitral valve replacement have been thought necessary for the successful correction of the isolated APM.4,5 Valve replacement with a mechanical prosthesis or bioprosthesis is not without drawbacks. This strategy is undesirable for young patients.

The case presented here illustrates several issues regarding diagnosis and treatment of this rare congenital anomaly. Recognition of the left ventricular midcavity obstruction caused by an APM insertion directly into the anterior mitral leaflet before the usual DSAS resection is critical.2,3 Clinical recognition of these unusual findings is possible by means of 2-dimensional echocardiography.2-6 Identification of the direct continuity between the base of the anterior mitral leaflet and apicoseptal portion of the left ventricle is diagnostic in our patient. This clear recognition allows us to adopt the mitral valve–sparing strategy described by Maron and associates.3 Indeed, an extensive myectomy trough within the left ventricular midcavity and the fully mobilized anterior mitral leaflet by careful shaving off of the residual papillary muscle over the ventricular surface of the leaflet substantially widen the mid to basal outflow tract. Mitral valve is spared with good mobility and competency as well.

In conclusion, LVOT obstruction caused by a combination of the DSAS and APM is rare and challenging to identify. The failure to recognize this anomaly before and during operation could be disastrous. Sophisticated surgical planning and adopting of the meticulous procedures, including extensive myectomy trough creation, careful mitral leaflet mobilization, and adequate membranectomy, will restore the patient’s normal hemodynamic and functional capacity.

References