Evolving Treatment for Patients With Hypertrophic Obstructive Cardiomyopathy*

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Dynamic left ventricular outflow obstruction is a major cause of exertional symptoms in patients with hypertrophic cardiomyopathy. The obstruction is secondary to systolic anterior motion of the mitral valve abutting against a hypertrophied septum, resulting in diastolic dysfunction, myocardial oxygen supply-demand mismatch, reduced forward output, and secondary mitral regurgitation. Medical treatment directed at decreasing contractility, improving diastolic filling, and slowing heart rate can be successful in reducing symptoms in many patients. However, there is a subset of patients who remain severely symptomatic, despite optimal medical therapy.

Although surgical resection of the septal hypertrophy has been an accepted treatment for these severely symptomatic patients for >5 decades, techniques have evolved and become highly refined. Most experienced surgeons favor wider (“extended”) resection of septal muscle (1,2) rather than resection of a narrower portion of muscle in the early reports of Morrow (3). Other interventions have been utilized to treat left ventricular outflow obstruction, including dual chamber pacing (4), alcohol septal ablation (5,6), radiofrequency septal ablation, and, most recently, implantation of a MitraClip (7). Less invasive and less demanding procedures appeal to patients and to many clinicians, especially those who treat obstructive HCM infrequently, but transaortic septal myectomy has remained the optimal therapy for carefully selected patients with severely symptomatic hypertrophic cardiomyopathy. When performed by experienced surgeons, myectomy can now be performed with a low operative risk (<1% for isolated myectomy) and minimal complications, with documented sustained improvement in symptoms. These patients, who were severely limited before the operation, are generally able to return to a full active lifestyle.

In current practice at centers treating large numbers of patients, complications of septal myectomy, including septal defects and heart block, are now seen in <1% of patients undergoing this procedure. The extension of the myectomy distally has been found to be important for prevention of residual post-operative obstruction. Special surgical approaches have also been successful in the treatment of patients with the mid-ventricular and apical variants of hypertrophic cardiomyopathy (8,9).

The underlying pathophysiologic mechanisms producing the systolic gradient are complex: they are related to septal hypertrophy, but are also due to abnormalities of the mitral valve apparatus. Together, they produce drag forces resulting in systolic anterior motion of the mitral valve. There are abnormalities of the mitral valve present in most patients with hypertrophic cardiomyopathy, including anterior displacement of the papillary muscles, anomalous attachments of the papillary muscles and chordae, and marked elongation of the leaflets themselves. Because mitral regurgitation is a major underlying pathophysiological mechanism in severely symptomatic patients, mitral valve replacement with a low-profile prosthesis was initially suggested as a surgical option (10,11). Even though this procedure relieves both the outflow gradient and mitral regurgitation, it has been avoided due to the additional adverse consequences of a prosthetic valve. To optimize the hemodynamic benefit of surgery, others have proposed additional mitral valve repair techniques besides surgical myectomy, including mobilization of papillary muscles and extension or plication of the mitral leaflets (12-16).

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However, experienced surgeons have found that a complete, well-performed myectomy alone is sufficient to obliterate the outflow obstruction and relieve the mitral regurgitation, unless there is an intrinsic primary abnormality of the mitral valve, such as ruptured chordae or fibrosed immobile mitral leaflets (17).

In this issue of the Journal, Ferrazzi et al. (18) report on the clinical and hemodynamic results of a surgical approach in 39 patients with only “mild” septal hypertrophy, in which cutting thickened secondary chordae is added to a shallow septal muscle resection. The results are comparable to the excellent results from other series of patients with increased septal thickness undergoing conventional myectomy with residual outflow gradients <10 mm Hg and a marked improvement in functional class at follow-up of nearly 2 years. They demonstrated that the anterior leaflet-annulus ratio was greater and the tenting area under the mitral valve was smaller than in patients who had myectomy alone, indicating that the procedure moved the mitral apparatus more posteriorly, thereby “enlarging the outflow tract.” They conclude that this unique procedure is an important step forward in the treatment of this subgroup of patients, whom they consider would otherwise have received a mitral valve replacement.

Ferrazzi et al. (18) should be congratulated on the outcome of their surgical experience in this subgroup of patients. Relief of symptoms is the primary goal of surgery, which was achieved in all their patients. Avoidance of a mitral valve replacement is of great long-term benefit for any patient. However, the patients were included in this study if the septal thickness was considered “mild” and <20 mm diameter. In our own practice, isolated septal myectomy has been effective from both the hemodynamic and clinical outcome standpoints for patients with septal thickness as small as 15 mm. The mechanism of a myectomy is not only to mechanically enlarge the outflow tract, but also to interrupt the abnormal flow patterns that cause a drag force on the elongated and abnormally displaced mitral leaflets.

Thus, a myectomy in the presence of a thinner septum will be effective if it is extended distally, where the abnormal flow patterns during systole begin. It is interesting that Ferrazzi et al. (18) compared the detailed echocardiographic measurements of mitral valve position from 25 patients with myectomy alone versus the 39 patients who had secondary chordal resection. However, there is limited information on the hemodynamic and clinical outcome of these 25 patients who underwent myectomy alone. It is unclear to us how often it is necessary to add an additional mitral valve procedure to an adequately performed myectomy to relieve obstruction and mitral valve regurgitation. In a recent review of our experience, additional mitral valve procedures were performed in 2.2% of 1,905 operations, and indications included redundant leaflet tissue thought to contribute to LV outflow tract obstruction, minor clefts in leaflets, and iatrogenic injury to chorda during myectomy or inadvertent injury of the mitral valve. No patients had a mitral valve procedure or valve replacement because of “inadequate” septal thickness. Dividing secondary chordae at the time of myectomy may decrease incidence of SAM and postoperative gradients in selected patients, but it is important to recognize that the most common cause of residual outflow tract obstruction is failure to carry the myectomy far enough towards the cardiac apex (19).

For the cardiologist who is caring for these patients, it is of great importance that there is an understanding of the outcomes of treatment, particularly for those patients who remain severely symptomatic despite optimal medical therapy. All would agree that operations on patients with symptomatic hypertrophic cardiomyopathy by experienced surgeons are truly life-changing interventions that will restore normal activity to those who were severely limited before the operation. The finding of abnormalities such as the “relatively thin septum” or “elongated redundant mitral leaflets” should not deter one from referring the severely symptomatic patient to centers of excellence with experienced surgeons for optimal therapy.

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