Surgical Myectomy for Hypertrophic Cardiomyopathy in the 21st Century, the Evolution of the “RPR” Repair: Resection, Plication, and Release

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Abstract

Since its first description in the 1950s, the pathophysiology of hypertrophic cardiomyopathy has been clarified by advanced echocardiographic technologies. Improved pharmacotherapy now successfully treats most afflicted individuals. Along with these advances, surgical management has also evolved, as the role of the mitral valve and the subvalvular structures in causing obstruction has been identified. Over the last 2 decades, a variety of options to surgically manage the complex patient with obstruction have been described. Successful surgical management is dependent on the complete evaluation of the causes of obstruction in the specific individual, as the heterogeneity of the anatomy may confound the direction of therapy. Mitral valve replacement may no longer be necessary in individuals who have a relatively thin septum and instead obstruct from an elongated mitral anterior leaflet or the presence of accessory papillary muscles and chords. Techniques for mitral valve plication have been successfully used with mid- to long-term success. A systematic strategy for the evaluation of obstruction in hypertrophic cardiomyopathy and the various surgical options are summarized in a procedure termed RPR for resection (extended myectomy), plication (mitral valve shortening), and release (papillary muscle manipulation). (Prog Cardiovasc Dis 2012;54:498-502)

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Outflow tract obstruction to the left ventricle (LV) thought due to infundibular hypertrophy was first described in London in the mid 1950s by Brock, from a London operating room, while, at approximately the same time, Teare reported massive hypertrophy causing sudden death from a pathology department in the same city. Obstruction was first successfully treated surgically with muscle bar excision by Cleland in 1958. Since then, and now called hypertrophic cardiomyopathy (HCM), a great deal has been clarified about the morphology, pathophysiology, variants, and genetics mostly due to advances in echocardiography and genotype analysis over the last 2 decades. Advances in pharmacologic management now limit the number of obstructed patients who require surgery to approximately 25% of patients screened and managed at HCM centers of excellence. Similarly, since the first large series of patients managed with surgery were described by Morrow et al in the 1960s, many advances in the surgical management have occurred.

Adverse effects of obstruction

Outflow tract obstruction results in a pressure gradient between the LV chamber and the ascending aorta. The pressure gradient causes an inappropriate increase in LV
Guidelines for therapies

Pharmacologic therapy, including β-blockade, calcium-channel antagonists, and disopyramide, is often successful in managing both symptoms and gradients in most patients. Surgical septal myectomy has been successfully performed for 40 years and is the criterion standard of therapy for patients who are refractory to pharmacologic therapy. An alternative therapy, alcohol septal ablation, has recently been introduced. Here, 1 to 2 mL of 100% alcohol is instilled into a proximal septal perforating branch of the left anterior descending. Alcohol septal ablation infarcts an area of the proximal septum causing it to involute and scar and thereby limits its capacity to participate in the obstruction process.

Recent guidelines from the American College of Cardiology and the American Heart Association discussed indications for these 2 interventional therapies for obstructive HCM. Indications for both surgery and ablation include patients (1) who are symptomatic in New York Heart Association classes III and IV, unrelieved by an adequate trial of β-blockers, calcium antagonists, or disopyramide; (2) who have persistent obstruction of LV outflow under basal conditions (subaortic gradient >50 mm Hg) or after physiologic provocation, attributable to the opposition of the mitral valve with the hypertrophied septum; and (3) who have morphology judged reparable by the experienced operator. Surgery was deemed the preferred procedure in most patients. In addition, to differentiate surgery from alcohol septal ablation, specific conditions that favor surgery were delineated: (1) patients who are younger than 21 years; (2) highly recommended for individuals younger than 40 years; (3) obstructive morphology involving complex mitral subvalvular structures, that is, accessory papillary muscle and/or chords; (4) situations where concomitant cardiac surgical therapy is required; and (5) when performed in an experienced center.

Morphological variations driving the evolution of surgical resection

The surgical septal myectomy popularized by Morrow in the 1960s consisted of a resection of subaortic muscle, approximately 1 cm wide, 1 cm deep, and 2 cm in distance into the ventricular chamber. Although this will relieve symptoms in a significant proportion of patients, it has a failure rate of between 10% and 20%. This is thought to be commonly due to an inability to visualize and resect deep enough in the LV cavity and because of variants to morphology predominantly of the mitral valve and papillary muscles that have only been made clearer by advances in echocardiography. The morphological spectrum of HCM includes the heterogeneity of the septal hypertrophy, basal, midventricular, or apical; elongated mitral leaflets, usually the anterior leaflet but, on occasion, the posterior as well; anterior positioning of the papillary muscles; and anatomical variants of the mitral valve attachments and subvalvular structures. In fact, variations in morphology are so common that “it is relatively uncommon to encounter a patient with obstructive HCM at operation in whom septal hypertrophy is both particularly marked and homogeneously distributed so that the standard myotomy-myectomy can be undertaken with no preoperative deliberation regarding the pattern and magnitude of septal thickness.” In the 1990s, Messmer described the extended myectomy that would include enough of the midventricular muscle to allow flow to track more anteriorly and medially along the surgically reduced septum and away from the mitral valve. Flow is thereby made more parallel to the mitral valve and reduces the chances of pushing the anterior leaflet into the outflow tract to cause obstruction. In addition, Schoendube et al, working with Messmer, also included thinning of the hypertrophied papillary muscles to allow them to fall more posteriorly into the ventricular chamber and, in doing so, to bring the anterior leaflet posterior with it, again, out of the way of the flow dynamic within the outflow tract. Echocardiography and gross observation had shown that the papillary muscles are frequently malpositioned within the ventricular chamber in an inappropriately anterior location. As experience in extended myectomy increased, additional variants of papillary muscle attachments were identified. With some frequency, accessory...
papillary muscles are noted, some with attachments directly to the anterior leaflet of the mitral valve without any intervening chordae tendineae. In addition, there are often chordal attachments attaching the anterior leaflet directly to the lateral wall of the LV, prepositioning it anteriorly. These and other similar variations all tend to draw the mitral valve into the outflow tract; these are commonly seen in all obstructed patients that now come to surgery and are the dominant cause of obstruction in patients where the septum may be relatively thin, 18 mm or less.

In summary, then, a variety of morphological variants need to be considered when planning the appropriate surgical management of obstruction: the heterogeneity of the septal hypertrophy, particularly its thickness in the proximal and also the mid-LV; the location of the point of mitral-septal contact; the length and other morphological characteristics of the anterior leaflet of the mitral valve; the location of the papillary muscles and their accessory lateral attachments; and the presence of any abnormal subvalvular segments including accessory papillary muscles attached to the mitral leaflets and thickened chords contributing to obstruction.

Symptomatic relief and significant drops in outflow tract gradients are obtained in most patients undergoing extended myectomy, defined to include manipulation of the subvalvular apparatus, papillary muscle thinning, and the repositioning and possible removal of accessory unneeded papillary muscles and chords. However, these procedures will still not address that morphological variant that consists of a relatively thin septum, less than 18 mm, and an anterior mitral leaflet that, at times, may exceed a length of 4.0 cm. Additional morphological variations include patients where the septum is relatively thin, the anterior leaflet is only mildly elongated, but accessory papillary muscles exist as the primary cause of obstruction.

Role of the mitral valve

Understanding the role of the mitral valve in obstruction, surgeons proposed mitral valve replacement in the 1980s as a solution to the management of the patient with obstruction in the face of a relatively thin septum. However, because many of these patients were relatively young, a solution preserving the native valve was thought to be preferable. In 1992, McIntosh et al proposed mitral valve plication. Because it was known that the mitral valve billows out into the outflow tract, it seemed logical to tighten the leaflet in this area with a plication technique vertically oriented to the valve when oriented in the anteroposterior aspect. This technique was copied with some success but was never applied widely. Nonetheless, the problem of the overly enlarged anterior leaflet continued to represent a surgical dilemma in those cases where extended myectomy is not suitable. Other proposals included patch extension of the anterior leaflet to increase its stiffness and cause lateral displacement of the secondary chords, perhaps moving the leaflet posteriorly out of the left ventricular outflow tract.

We set out to clarify a systematic approach to analyze the complete pathophysiology of the individual patient and tailor a unique collection of surgical procedures to ensure complete resolution of both the outflow tract gradient and mitral insufficiency, in addition to improving stroke volume and cardiac output, both at rest and upon provocation. The hope was that using this methodology, every patient with outflow tract obstruction could be successfully treated with long-term success. We termed this process the RPR procedure, Resection, Plication, and Release (Figure). The resection refers to the extended myectomy, plication is a treatment of the anterior leaflet itself in cases where it is extremely redundant, and release refers to the dividing of any abnormal attachments that the papillary muscles may have to the lateral wall of the LV. In considering the action of the anterior leaflet, it seemed logical that because the leaflet is too long in an anteroposterior dimension, the plication would be better suited horizontally instead of vertically. This would limit the leaflet’s ability to billow out not only by shortening its excursion into the outflow tract but also by stiffening its midportion. Although the vertical plication showed some success in published reports, we found that more often than not, the plication line impacted adversely on the coaptation zone of the leaflet and caused central regurgitation. The horizontal plication leaves the coaptation zone untouched and stiffens the midportion of the leaflet. Moreover, the procedure is technically simple and easy to duplicate; this portion of the leaflet is easy to access from the aortotomy used for the myectomy, and because the working surface of the valve is untouched, the hope is that in the long term, the coaptation area will not reactively thicken and fibrose causing central insufficiency later in life. As a significant proportion of patients with HCM are young, it is possible that leaflets heavily manipulated will fibrose and calcify over a lifetime, not unlike the process that occurs after rheumatic fever where the leaflets become dysfunctional only after a period of decades, although this is purely hypothetical.

On rare occasion, even horizontal plication has not completely relieved outflow tract obstruction. In certain circumstances, the leading edge of the central portion of the anterior leaflet is the culprit anatomical feature causing mitral septal contact, instead of the billowing central portion. In these cases, no amount of central stiffening will limit SAM. Once this is recognized, alternative solutions are necessary and have included simple resection of the terminal portion of the leaflet because again, this area is not involved with coaptation and/or suturing of the...
midportion of the anterior leaflet to the posterior leaflet (Alfieri type stitch). We have used both of these methods with success under the rare circumstances.

In conclusion, the RPR repair addresses the morphological variations present in a significant proportion, if not all, of the HCM population that present for surgical therapy. Appreciation for the morphological variants in HCM is necessary when planning the extent of the myectomy and the need for possible resection and manipulation of abnormal subvalvular attachments including malposition of the papillary muscles and dealing with the overly redundant mitral leaflets. The mitral valve plication easily and reliably deals with the large protruding anterior leaflet causing obstruction in many classically typical HCM cases but particularly those with only modest hypertrophy of the septum. This minimizes the need for mitral valve replacement. Our published reports confirm the effectiveness and durability of this methodology over an extended follow-up.\textsuperscript{21,22}

**Statement of Conflict of Interest**

Drs Daniel G. Swistel and Sandhya K. Balaram have no conflict of interest and no disclosures to make.

**References**


