

## **Surgical Myectomy for Obstructive Hypertrophic Cardiomyopathy for Symptoms Resistant to Medications. The “RPR” Repair: Resection, Plication and Release.**

Outflow tract obstruction to the left ventricle thought due to infundibular hypertrophy was first described in London in the mid 1950's 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 somewhere around 25 percent 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 in the 1960's, many advances in the surgical management have occurred.

**Adverse effects of obstruction:** Outflow tract obstruction results in a pressure gradient between the left ventricular chamber and the ascending aorta. The pressure gradient causes an inappropriate increase in left ventricular wall tension and decreases coronary blood supply to the myocardium at the same time that myocardial oxygen consumption is elevated. The most common cause of obstruction is contact between anterior leaflet of the mitral valve and the hypertrophied septum commonly referred to as Systolic Anterior Motion (SAM). SAM also displaces the mitral valve and compounds an already precarious hemodynamic situation by causing mitral insufficiency. Obstruction also causes a sudden decrease in mid-LV ejection velocities and flow termed the “lobster-claw” abnormality. This instantaneous drop in LV systolic performance likely contributes to the inability to increase stroke volume with exertion. In obstruction, increase in wall tension, increased myocardial oxygen consumption, impaired systolic performance, and mitral insufficiency can culminate in debilitating symptoms of shortness of breath exercise intolerance, angina and syncope. Obstruction has been shown to decrease survival.

**Guidelines for Therapies:** Pharmacologic therapy, including beta blockade, calcium channel antagonists and disopyramide, are often successful in managing both symptoms and gradients in the majority of patients. Surgical septal myectomy has been successfully performed for 40 years and is the gold standard of therapy for patients who are refractory to pharmacologic therapy. An alternative therapy, alcohol septal ablation, has recently been introduced. Here, one to 2 ccs 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 who are 1) symptomatic in NYHA association class III and IV, unrelieved by an adequate trial of beta-blockers, calcium antagonists or disopyramide; 2) persistent obstruction of left ventricular 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) morphology judged reparable by the experienced operator. Surgery was deemed the preferred procedure in most patients. Additionally to differentiate surgery from alcohol septal ablation, specific conditions that favor surgery were delineated: 1) patients who are under the age of 21; 2) highly recommended for individuals under the age of 40; 3) obstructive morphology involving complex mitral subvalvular structures, ie. accessory papillary muscle and/or chords; 4) situations where concomitant cardiac surgical therapy is required and 5) when performed in an experienced center.

**Morphologic Variations driving the Evolution of Surgical Resection:** The surgical septal myectomy popularized by Morrow in the 1960's, consisted of a resection of subaortic muscle, approximately one centimeter wide, one centimeter deep and two centimeters in distance into the ventricular chamber. Though 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 left ventricular 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, either 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 anatomic 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 1990's, Messmer and his associates 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. Additionally, Schoendube, 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, with attachments directly to the anterior leaflet of the mitral valve without any intervening chordae tendinae. And, there are often chordal attachments attaching the anterior leaflet directly to the lateral wall of the left ventricle, repositioning 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.

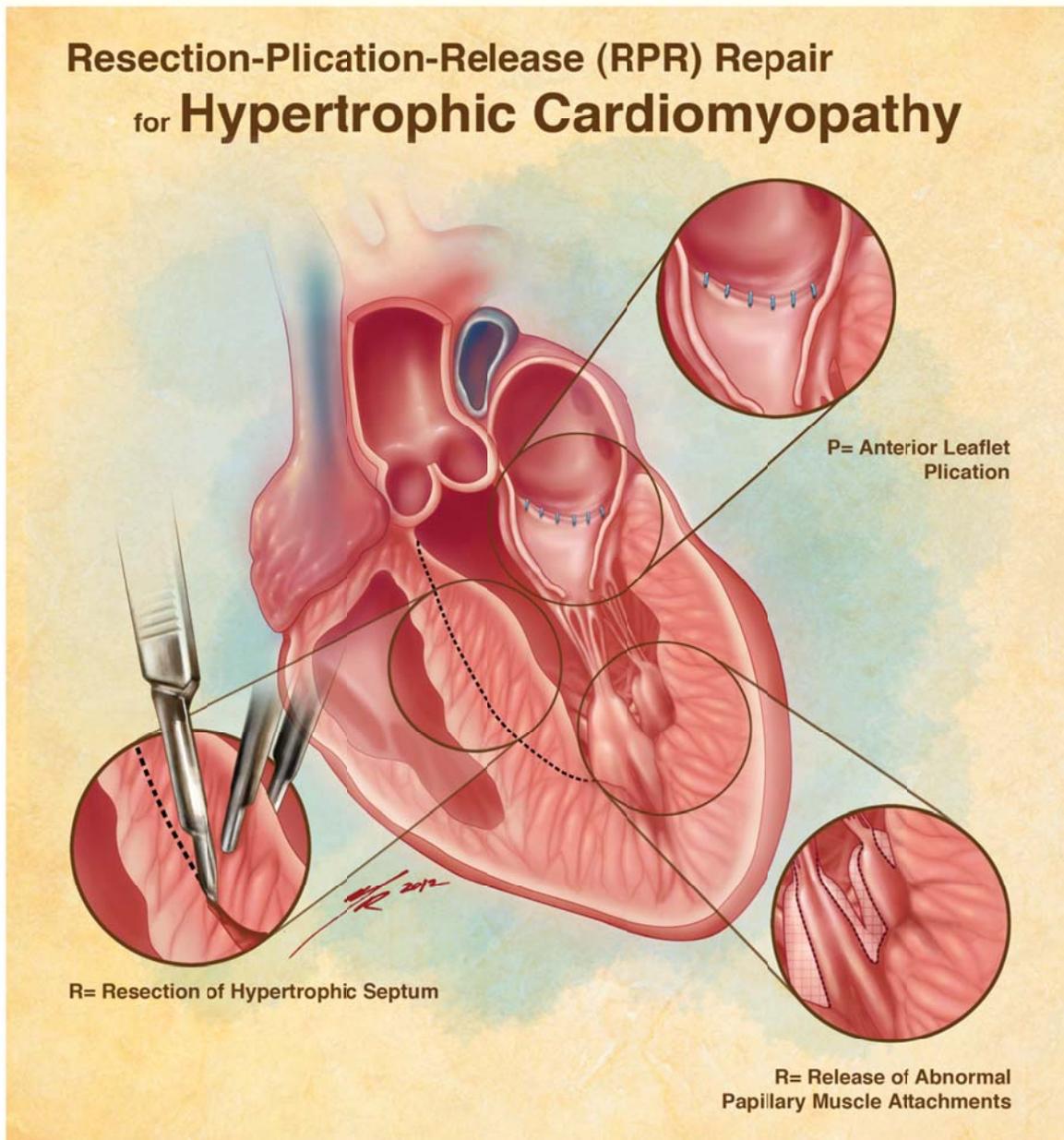
In summary, then, a variety of morphologic variants need to be considered when planning the appropriate surgical management of obstruction: the heterogeneity of the septal hypertrophy, particularly its thickness in the proximal and also the mid LV, the location of the point of mitral-septal contact, 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 the majority of 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. But these procedures will still not address that morphologic variant that consists of a relatively thin septum, less than 18mm, and an anterior mitral leaflet that at times may exceed a length of 4.0 cm. Additional morphologic 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 1980's as a solution to the management of the patient with obstruction in the face of a relatively thin septum. However, since many of these patients were relatively young, a solution preserving the native valve was thought to be preferable. In 1992, McIntosh proposed mitral valve plication. Since 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 antero-postero 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 LVOT.

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*. 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 the papillary muscles may have to the lateral wall of the left ventricle. In considering the action of the anterior leaflet, it seemed logical that since the leaflet is too long in an antero-postero 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 stiffen 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 mid-portion 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 since the working surface of the valve is untouched, the hope is that 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, though this is purely hypothetical.



Schematic representation of the hypertrophied heart in hypertrophic cardiomyopathy depicting the morphologic variations leading to obstruction and the potential surgical options for management including *Resection* (extended myectomy), *Plication* (horizontal mitral plication) and *Release* (manipulation of the subvalvular structures).

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 anatomic 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 since again, this area is not involved with coaptation, and/or suturing of the mid-portion of the anterior leaflet to the posterior leaflet (Alfieri type stitch). We have utilized both of these methods with success under the rare circumstances.

In conclusion, the *RPR* repair addresses the morphologic variations present in a significant proportion, if not all, of the HCM population that present for surgical therapy. Appreciation for the morphologic variants in HCM is necessary when planning the extent of the myectomy, 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 period of time.

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