Resection-Plication-Release for Hypertrophic Cardiomyopathy: Clinical and Echocardiographic Follow-Up

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Background. Abnormal positioning and size of the mitral valve contribute to the systolic anterior motion and mitral-septal contact that are important components of obstructive hypertrophic cardiomyopathy (HCM). The RPR repair (resection of the septum, plication of the anterior leaflet, and release of papillary muscle attachments) addresses all aspects of this complex pathology. This study reports outcomes regarding effectiveness of the RPR repair.

Methods. Fifty consecutive unselected patients (average age, 55.8 years) undergoing RPR repair for obstructive HCM from 1997 to 2007 were studied. Each patient underwent preoperative and postoperative transthoracic echocardiograms to document gradient, ejection fraction, degree of mitral regurgitation, and systolic anterior motion. Intraoperative transesophageal echocardiogram was used to guide all surgical repairs. Clinical follow-up included patient interviews to determine New York Heart Association (NYHA) status.

Results. Concomitant operations were performed in 25 patients (50%). Postoperative mortality was 0%. Average mean left ventricular outflow tract gradients decreased from 134 ± 40 to 2.8 ± 8.0. Mitral regurgitation improved from a mean of 2.5 to 0.1 (p < 0.001). Average length of stay was 6.9 ± 2.7 days. NYHA class improved from 3.0 ± 0.6 to 1.2 ± 0.5. Follow-up was 100%, with a mean of 2.5 ± 1.8 years. Average mitral regurgitation at follow-up was 0.9, with no residual systolic anterior motion.

Conclusions. The RPR repair is safe and effective for symptomatic obstructive HCM. Our data support repair of the mitral valve that results in good intermediate outcomes with respect to gradient, mitral regurgitation, and clinical status.

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Hypertrophic cardiomyopathy (HCM) is an uncommon, autosomal-dominant disease characterized by asymmetric septal hypertrophy that was first described by Sir Russel Brock in 1957 [1]. Morrow’s transaortic approach to resection of the septum relieved the bulk of obstruction and was popularized after presentation of his results in 83 patients and their follow-up in 1975 [2, 3]. More recently, multiple large studies have demonstrated excellent outcomes with surgical myectomy for repair of HCM for symptomatic, drug-refractory septal hypertrophy with outlet obstruction [4–9].

The anterior leaflet of the mitral valve plays an important role in obstruction in HCM. Echocardiographic data supports that it is the anterior motion of the mitral valve that causes both subaortic obstruction and loss of coaptation with the posterior leaflet, resulting in mitral valve insufficiency [10, 11]. Previous treatments of mitral regurgitation (MR) with a redundant anterior mitral leaflet have included mitral valve replacement and repair [12, 13]. It is the anterior displacement and the recognition of drag (pushing) forces on the mitral valve that prompted Messmer and colleagues to perform a more extensive myectomy, followed by thinning of the papillary muscle; this allows the anterior leaflet to fall more posteriorly within the ventricular chamber [4].

Our group has proposed horizontal plication of the anterior mitral leaflet, reasoning that this leaflet is often too long in an anteroposterior dimension. Shortening the leaflet horizontally limits its excursion into the outflow tract by decreasing the length in its longest dimension and stiffening the redundant tissue [14–16]. This, in addition to standard extended septal myectomy and release of abnormal papillary attachments, has been termed the resection-plication-release (RPR) repair. This study reviews the clinical and echocardiographic follow-up of our first 50 patients with this repair.
Patients and Methods

Study Group and Patient Selection

From 1997 until 2007, 50 patients underwent RPR repair for HCM at St. Luke’s-Roosevelt Hospital Center. These patients were selected from a group of approximately 450 patients. Most were treated medically to reduce their left ventricular outflow tract (LVOT) gradient and alleviate their symptoms. Patients with symptomatic LVOT obstruction (LVOTO) were given an aggressive trial of pharmacologic therapy including β-blockers, disopyramide, or calcium channel blockers. Those in whom medical therapy failed and who remained symptomatic with LVOTO and gradients exceeding 50 mm Hg at rest or after physiologic provocation were referred for surgical repair.

Selection criteria for patients undergoing the RPR repair included those patients who met the aforementioned criteria and in addition had significant systolic anterior motion (SAM) with an elongated and floppy mitral valve, as judged by echocardiography. The ultimate decision to proceed with an RPR repair was made preoperatively based on transthoracic echocardiography (TTE) characteristics but was subject to change based on intraoperative transeosophageal echocardiography (TEE) and operative findings.

The decision involved an integrated assessment of the extent of SAM, the size of the mitral valve leaflets, the presence of abnormal papillary muscle attachments, and the slack and redundancy of the mitral valve as assessed by direct visualization. Specifically, qualitative preoperative assessment included assessment of the mitral valve as (1) being large relative to the LV chamber and (2) showing large excursion and a large angle of motion as it is pushed into the outflow tract. The intraoperative assessment by the surgeon required that the valve was redundant, enlarged, and slack, as assessed by nerve hook traction.

This study involving human subjects was approved by the St. Luke’s-Roosevelt Institutional Review Board. Individual consent was waived for the study after clearance and use of an approved deidentified database.

Echocardiography

Before admission, TTE was performed for assessment of the LVOT gradient, septal and anterior wall thickness, papillary muscle anatomy, SAM, and MR. Echocardiographic gradients were measured at rest, after the Val-salva maneuver, after standing, and after treadmill exercise. For each patient, the distance between the aortic annulus and the area of mitral–septal contact was carefully measured during diastole. The furthest extent of the myectomy into the LV was determined by measuring the distance from the aortic annulus to the far side of the septal bulge. Mitral valve structure and function were also assessed during outpatient preoperative echocardiograms. Criteria for mitral valve replacement included severe MR in the setting of heavy leaflet calcification and immobility (excluded from study). Any patient with posterior mitral valve prolapse who required a standard mitral valve repair was also excluded from the study.

All patients also received TEEs to remap the location and extent of septal hypertrophy, assess mitral valve function, measure LVOT gradients, and evaluate MR. The degree of MR was scored on a standard scale from 0 to 4.

Operative Technique

A standard median sternotomy was performed, and patients were placed on cardiopulmonary bypass (CPB) using moderate hypothermia. The aorta was cross-clamped, and both antegrade and retrograde cold crystalloid cardioplegia were delivered. Visualization of the septum was achieved with a transverse aortotomy and retraction of the aortic valve leaflets; this allowed for direct evaluation of the degree of hypertrophy.

Extended septal myectomy was performed as previously described by Messmer and colleagues [4, 14]. Stabilization of the muscle was accomplished by use of a trefoil hook. This approach ensures an adequate length of resection into the LV cavity. Two parallel incisions were made into the septal bulge and connected to remove the muscle mass. Further resection was performed after careful palpation of the septum and estimation of residual LV mass. Myectomy was extended to the base of the papillary muscles, when midseptal thickening was present.

Mitral valve pathology was addressed after the myectomy was completed. The horizontal plication of the anterior leaflet was performed to reduce length and decrease leaflet and chordal slack. The leaflet and its degree of redundancy were evaluated in each case and three to four 5-0 polypropylene sutures were placed through the fibrotic area of the leaflet in a horizontal mattress fashion. The extent of plication was determined by integrating the preoperative echo, the degree of SAM, the size of the mitral valve, and the slack and redundancy of the anterior leaflet as assessed with the nerve hook. This usually resulted in a plication of 2 to 5 mm.

The papillary muscles were grasped and pushed medially to visualize the abnormal connections between the papillary muscles and the anterior wall of the ventricle. A blade was used to divide the thickened abnormal attachments. A pituitary rongeur may be used to resect a portion of the junction of the papillary and lateral wall. This reduces the diameter of the papillary muscle and allows for posterior displacement of the anterior mitral leaflet. Division of abnormal attachments and thinning of the papillary muscles is critical for the treatment of SAM. This is followed by extensive irrigation of the LV cavity.

Postoperative TEE was performed after withdrawal of CPB, but before removal of the cannulae, and assessed by the surgeon, anesthesiologist, and referring cardiologist. The repair was closely examined for residual SAM, degree of gradient, presence of ventricular septal defect, and MR. Intravenous dobutamine was used to provoke the patients and measurements and assessments were
repeated. CPB was reinstituted for a persistent gradient greater than 30 mm Hg, mitral septal contact, or MR of moderate or greater degree.

Follow-Up
Follow-up was obtained by direct patient interview, review of patient records, and information supplied by referring physicians. TTE was used for examination of LVOT gradient, MR, and LV function at the outpatient follow-up. Patients were seen 2 weeks postoperatively, with TTE performed at 3 months. These visits were followed by yearly TTE evaluation. Patients being treated medically were evaluated with yearly echocardiography and more frequently as necessary based on changes in treatment or symptoms.

Statistical Analysis
All data were collected retrospectively. Data are presented as mean ± standard deviation. MR is presented as the mean with the ranges included. Continuous variables were compared using paired t test, with significance accepted at values of p < 0.05.

Results
Baseline Characteristics
From August 1997 through May 2007, 50 patients (26 men, 24 women) underwent RPR repair at St. Luke’s-Roosevelt Hospital Center. Patients were a mean age of 55.8 ± 14.6 years (range, 23 to 84 years). The patients were symptomatic, with an average New York Heart Association (NYHA) functional class of 3.0 ± 0.6 (range, 2 to 4). The average resting LVOTO gradient, as measured by echocardiography, was 88 ± 26 mm Hg. The gradients after provocation averaged 134 ± 40 mm Hg (range 66 to 230 mm Hg). Mean ventricular septal thickness was 23 ± 5 mm. The preoperative ejection fraction was 0.60 ± 0.10 (range, 0.35 to 0.85). SAM was present in 96% of patients. The mean degree of MR in this group was 2.5 (range, 1 to 4).

Preoperative characteristics included coronary artery disease in 13 patients (27%), hypertension in 17 (35%), diabetes mellitus in 5 (10%), and prior ventricular arrhythmias in 11 (22%). In addition, 4 patients (8%) had chronic obstructive pulmonary disease, 3 (6%) had a previous pacemaker in place, and 10 (20%) had a history of smoking. Two patients had previous septal myectomy at other institutions.

A positive family history for HCM was present in 15 of 50 patients (30%), of whom 6 (12%) had more than one relative with HCM. Preoperatively, 42 patients (84%) were treated with β-blockers, 18 (36%) were also taking disopyramide, and calcium channel blockers had been used in 6 (12%; Table 1).

Intraoperative Findings
All 50 patients underwent RPR repair as described. Half of the patients had additional procedures, including coronary artery bypass grafting in 12, atrial septal defect repair in 5, repair of atrial septal defect in 2, and radiofrequency atrial ablation in 6. Not all HCM patients received the full RPR repair. Average aortic cross-clamp years (range, 23 to 84 years). The patients were symptomatic, with an average New York Heart Association (NYHA) functional class of 3.0 ± 0.6 (range, 2 to 4). The average resting LVOTO gradient, as measured by echocardiography, was 88 ± 26 mm Hg. The gradients after provocation averaged 134 ± 40 mm Hg (range 66 to 230 mm Hg). Mean ventricular septal thickness was 23 ± 5 mm. The preoperative ejection fraction was 0.60 ± 0.10 (range, 0.35 to 0.85). SAM was present in 96% of patients. The mean degree of MR in this group was 2.5 (range, 1 to 4).

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<td>Aortic valve replacement</td>
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<th>Operative times, min</th>
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<tr>
<td>Cross-clamp time</td>
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<td>CPB time, min</td>
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CABG = coronary artery bypass grafting; CPB = cardiopulmonary bypass; SD = standard deviation.

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<th>Table 3. Complications</th>
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<td>Complication</td>
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| Postoperative bleeding | 1 (2) |
| Atrial fibrillation | 6 (12) |
| Permanent pacemaker | 2 (4) |
| Cerebrovascular accident | 1 (2) |
| Sternal wound infection | 1 (2) |
| Late pericardial tamponade | 1 (2) |
time of all patients was 96.5 ± 25 minutes, with a CPB time of 129 ± 34 minutes (Table 2).

Patients who did not require mitral valve plication due to a shortened or fibrotic anterior leaflet and those who required a mitral valve replacement due to profound MR with severely calcified leaflets (advanced rheumatic disease) were not included in this study because they did not fulfill the criteria for the RPR repair.

Initial postoperative TEEs in the operating room demonstrated a marked reduction in LVOTO gradient to 2.8 ± 8.0 mm Hg (range 0 to 23 mm Hg; p < 0.0001). Significant improvement in MR to 0.1 (range, 0 to 2; p < 0.0001) was also demonstrated. None of the patients had persistent SAM.

After initial assessment by TEE, the decision was made for re-resection in 3 of the 50 patients (6%). These patients required a second pump-run for inadequate initial resection. One patient had persistent and significant MR despite plication and was placed back on CPB to undergo an Alfieri (edge-to-edge) repair of the mitral leaflets, with resulting trace residual MR.

Early Outcomes

No deaths occurred in the postoperative period. Surgical morbidity was 10% overall and included one reoperation for bleeding, one cerebrovascular accident, and one late pericardial tamponade in a patient requiring anticoagulation. Two patients required placement of a permanent pacemaker for complete heart block (4%). The incidence of left bundle branch block in follow-up was 42% (21 of 50 patients). No patients required re-resection of the septum. Overall, the patients had marked improvement in clinical symptoms, with an improvement in NYHA class to 1.2 ± 0.5 (p < 0.0001).

Follow-Up

Echocardiographic or clinical follow-up, or both (closing date, May 2007), was available for all 50 patients (100%). A mean follow-up of 2.5 ± 1.8 years (range, 0.5 to 9 years) was obtained. No deaths occurred during the study period. An examination of the most recent echocardiographic data as of May 2007 for all 50 patients demonstrated that LVOT gradients remained low at 3.4 ± 12.7 (p < 0.0001). Ejection fraction was 0.70 ± 0.10. Maximal septal thickness was 15.7 ± 5.1 mm. The degree of MR remained stable as well, measuring 0.9 (range, 0 to 2; p < 0.0001; Table 4).

Comment

HCM has marked phenotypic variation, leading to significant differences in clinical presentation. The pathophysiology involves variation in ventricular septal thickness, SAM of the mitral valve, and papillary musculature. Detailed echocardiography has led to better understanding of this complex pathophysiology [17–22].

The goal of surgical intervention for HCM is to improve symptoms through relief of LVOTO. Recent large studies show excellent long-term results with extended septal myectomy alone [7–9]. However, HCM operations have been viewed as difficult, with unpredictable results, except when performed at centers with extensive experience. Postoperative SAM and MR have been reported as well as persistent LVOTO [23, 24].

On the basis of echocardiographic data implicating the mitral valve as a key component in obstructive HCM, our group has previously published a novel method of anterior mitral leaflet plication that is performed in addition to extended septal myectomy [15, 16]. This RPR repair technique addresses multiple aspects of HCM pathology. Our surgical approach specifically addresses the problem of the large protruding anterior mitral valve leaflet and its contribution to obstruction. Patients who are candidates for mitral valve plication include those with increased mobility, size, or length of the anterior mitral leaflet. These are patients who are judged morphologically to be at increased risk of residual SAM and obstruction. This study demonstrates that this repair is durable and effective over time, both from a functional and clinical standpoint.

Prior large surgical series of myectomy have not specifically addressed the problem of the mitral valve [7–9]. Repair of the mitral valve itself to relieve SAM is somewhat controversial, although the importance of the anterior leaflet of the mitral valve in the underlying pathology has long been recognized. Previous groups have used mitral valve replacement to eliminate obstruction [12, 25]. Macintosh and colleagues [26] addressed the pathology of the mitral valve with selective mitral valve replacement for those patients with a septal thickness of less than 18 mm. Others have proposed a sliding leaflet or other mitral valve repair for obstructive HCM [27–30]. Redundancy in the anterior mitral leaflet has also been treated with a standard Alfieri edge-to-edge repair [31].

Some groups maintain that extended septal myectomy alone reduces LVOTO and improves any associated MR: Yu and colleagues [32] demonstrated that preoperative MR is proportional to the degree of LVOTO and relief of the LVOTO improved MR with no additional need for an operation. This is a persistent criticism of the RPR tech-

Table 4. Postoperative Outcomes

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<tr>
<th>Outcome</th>
<th>Measurements</th>
<th>Pre-op</th>
<th>Post-op</th>
<th>p Value</th>
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<td>LVOTO, mean ± SD, mm Hg</td>
<td>134 ± 40</td>
<td>2.8 ± 8.0</td>
<td>&lt;0.0001</td>
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<tr>
<td>Mitral regurgitation</td>
<td>2.5</td>
<td>0.1</td>
<td>&lt;0.0001</td>
<td></td>
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<tr>
<td>NYHA, mean ± SD, class</td>
<td>3.0 ± 0.6</td>
<td>1.2 ± 0.5</td>
<td>&lt;0.0001</td>
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LVOTO = left ventricular outflow tract obstruction; NYHA = New York Heart Association.
depends not only on its length and redundancy but also its cavity [34–37]. The cause of SAM appears multifactorially large in HCM and positioned anteriorly in the LV and related to the geometry of the anterior leaflet [38]; it persists after ablation [33]. Furthermore, echocardiography on selected patients who were judged to be at risk for residual SAM and obstruction. Our group has reasoned that the problem of the mitral valve is that it is too long in the anteroposterior direction. This repair shorts and stiffens the leaflet to limit its entry into the LVOTO.

Aside from persistent MR and SAM reported in the surgical literature, additional evidence supports the importance of the mitral valve in this disease. Failures occur after alcohol septal ablation in which persistent SAM is present due to anterior papillary muscle displacement that persists after ablation [33]. Furthermore, echocardiography has revealed a wide spectrum of mitral leaflet abnormalities in HCM: mitral leaflets are morphologically large in HCM and positioned anteriorly in the LV cavity [34–37]. The cause of SAM appears multifactorial and related to the geometry of the anterior leaflet [38]; it depends not only on its length and redundancy but also on the LVOT area and contraction of the LV [39]. The pathophysiologic of the mitral valve component has to do with the pushing force of flow as the dominant hydrodynamic force along with the mitral leaflets being large and anteriorly positioned. SAM, in the setting of mitral septal contact combined with an element of chordal slack, provides an amplifying feedback loop that leads to further obstruction [15]. Limitations of this study include its small size. Although the follow-up was consistent, the average time of follow-up echocardiography is medium-term, because most patients were operated on within the previous 5 years. We were unable to compare these patients with a control group because they were selected for the RPR repair from their specific morphology and pathology, leading to an element of selection bias. Exact measurements of the length of mitral leaflet plication are currently being studied in a retrospective fashion using preoperative and postoperative TEE.

The RPR repair is a safe, reproducible, and effective method of treatment for symptomatic obstructive HCM. The operative risk is low, with marked improved in symptom relief and clinical outcomes.

References


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**DISCUSSION**

**DR JOHN S. IKONOMIDIS** (Charleston, SC): Do you have any data from your institution comparing this technique, which is clearly a little bit more involved, to the standard myotomy/myectomy?

**DR BALARAM:** We have an additional 20 patients who have undergone a standard myotomy/myectomy as opposed to this group of patients. I think it is very important for people to realize that not every patient who comes in for an HCM [hypertrophic cardiomyopathy] repair should have this anterior leaflet plication. These are very specific patients. One hundred percent of them had systolic anterior motion and had measured elongated anterior leaflets of their mitral valve. Some patients come in with small, somewhat fibrotic, or calcified mitral valves that need replacement, and these patients were not included in this study. Because of the low numbers, we have not directly compared the two groups.

**DR EDWARD L. WOODS** (Danville, PA): That was an excellent presentation of a very difficult topic. Could you elaborate a little bit more on how you moved or manipulated the papillary muscles, which at times is one of the biggest aspects of this SAM [systolic anterior motion] and HCM?
DR BALARAM: The important part of the papillary muscles is to release any of the abnormal attachments between the papillary muscles and the wall. Sometimes the papillary muscles actually appear to be completely extruding from the wall itself, and in that situation we would try and thin the papillary muscles somewhat using a rongeur and let the papillary muscles fall more posteriorly so that the whole mitral valve apparatus would fall more posteriorly as well. Of course, it is also important to make sure and irrigate thoroughly to prevent any segments of muscle from causing a stroke in the future.

DR HAROLD G. ROBERTS (Lauderdale Lakes, FL): I enjoyed your presentation very much. The plication of the anterior leaflet, I would like you to elaborate, if you would, more on what guidelines you use in determining how much to reduce the height of the anterior leaflet.

DR BALARAM: One of the problems with this anterior leaflet is the large amount of chordal slack. We base reduction of the anterior leaflet on the preoperative echo. We have not gone back to measure the exact measurements pre- and postoperatively as far as the length of the mitral valve leaflet. We are actually working on that data right now. We use preoperative echo and intraoperative assessment with nerve hooks to determine chordal slack and elongation to typically plicate between 2 to 5 mm of the anterior leaflet.

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