Pitfalls in Clinical Recognition and a Novel Operative Approach for Hypertrophic Cardiomyopathy With Severe Outflow Obstruction Due to Anomalous Papillary Muscle

Barry J. Maron, MD; Rick A. Nishimura, MD; Gordon K. Danielson, MD

Background—Ventricular septal myotomy/myectomy (Morrow procedure) is the standard surgical option for severely symptomatic patients with hypertrophic cardiomyopathy (HCM) and marked basal obstruction to left ventricular outflow due to mitral valve systolic anterior motion. In some patients, however, congenital malformations of the mitral apparatus may be responsible for outflow obstruction; the failure to recognize this morphology before operation could have adverse consequences.

Methods and Results—We recently evaluated 2 patients with obstructive HCM operated on at Mayo Medical Center in 1997 who demonstrated direct anomalous papillary muscle insertion into the anterior mitral leaflet, producing muscular midcavity obstruction. This anomaly is potentially identifiable with echocardiography by exaggerated anterior displacement of hypertrophied papillary muscles within the left ventricular cavity and the direct continuity between papillary muscle and anterior leaflet associated with a rigid motion pattern of the mitral apparatus. Echocardiographic diagnosis, however, was confused in both patients by the association of systolic anterior motion of the mitral valve, probably produced by freely mobile margins of the mitral leaflet unencumbered by papillary muscle insertion, and in 1 patient probably representing a second and more basal level of obstruction. Because outflow tract morphology was judged unsuitable for conventional myotomy/myectomy, a novel surgical strategy was designed to remove the outflow gradient in which an extensive myectomy trough (wider at its apical than basal extent) was created within the ventricular septum to papillary muscle level; also, in 1 patient, attachment of anterolateral papillary muscle with the lateral free wall was partially severed to increase mobility of the mitral apparatus. After surgery, both patients reported substantial relief of symptoms and improved exercise tolerance and also showed reduced or abolished basal outflow obstruction.

Conclusions—In HCM, outflow obstruction due to anomalous papillary muscle insertion directly into anterior mitral leaflet is challenging to identify but should always be contemplated before operative intervention. This important (but often unsuspected) congenital malformation may require alternative surgical strategies to standard myotomy/myectomy, similar to those described here. (Circulation. 1998;98:2505-2508.)

Key Words: surgery ■ cardiomyopathy ■ echocardiography ■ mitral valve

Surgery has been performed in severely symptomatic patients with hypertrophic cardiomyopathy (HCM) and marked left ventricular outflow obstruction for almost 40 years.1-7 Ventricular septal myotomy/myectomy operation (Morrow procedure) is directed toward relieving the outflow gradient by removal of a small amount of muscle from the basal anterior septum in the anatomic area in which subaortic obstruction develops by virtue of systolic anterior motion of the mitral valve (SAM) and septal apposition.8-12 This approach is standard practice at selected referral centers and is associated with relatively low operative mortality (≤1% to 2%).2-5,7,13 However, in a small but important subset of patients with obstructive HCM, left ventricular outflow obstruction is present in the midcavity region as a result of muscular apposition created by anomalous insertion of a papillary muscle directly into the anterior mitral leaflet (without interposition of chordae tendineae).14,15 Failure to recognize this anomaly may have catastrophic consequences during surgery,14 because standard myotomy/myectomy will not relieve obstruction located in the midventricular region. This malformation, often unanticipated by the operating surgeon, may require innovative surgical strategies to accommodate the unique outflow tract morphology and achieve the desired result.

Patient Selection
Two severely symptomatic men with obstructive HCM (17 and 35 years old) operated on at Mayo Medical Center in 1997 constitute the present report (Table). Each had marked...
Clinical and Morphological Features in 2 Patients With HCM and Outflow Obstruction Due to Anomalous Papillary Muscle Insertion Directly Into Anterior Mitral Leaflet

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Patient 1</th>
<th>Patient 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age/sex</td>
<td>17/M</td>
<td>38/M</td>
</tr>
<tr>
<td>NYHA functional class</td>
<td>Preoperative III</td>
<td>III</td>
</tr>
<tr>
<td>Postoperative</td>
<td>I</td>
<td>II</td>
</tr>
<tr>
<td>Basal LVOTG</td>
<td>Preoperative 100</td>
<td>65</td>
</tr>
<tr>
<td>Postoperative*</td>
<td>0</td>
<td>25</td>
</tr>
<tr>
<td>SAM†</td>
<td>Preoperative 3+</td>
<td>2+</td>
</tr>
<tr>
<td>Postoperative</td>
<td>0</td>
<td>1+</td>
</tr>
<tr>
<td>Mitral regurgitation§</td>
<td>Preoperative Mild</td>
<td>Mild</td>
</tr>
<tr>
<td>Postoperative</td>
<td>Mild</td>
<td>Mild</td>
</tr>
<tr>
<td>LV wall thicknesses‡</td>
<td>Anterior septum 32</td>
<td>19</td>
</tr>
<tr>
<td>Posterior septum</td>
<td>28</td>
<td>19</td>
</tr>
<tr>
<td>Anterolateral free wall</td>
<td>28</td>
<td>19</td>
</tr>
<tr>
<td>Posterior free wall</td>
<td>25</td>
<td>19</td>
</tr>
</tbody>
</table>

LV indicates left ventricular; LVOTG, peak left ventricular outflow tract gradient.

*Postoperative measurements by continuous-wave Doppler; provocative maneuvers were not performed.

†Magnitude of SAM‡: 1+ indicates mitral valve approaches ventricular septum, but without mitral valve-septal contact; 2+, brief mitral-septal contact; and 3+, prolonged mitral-septal contact >30% of echocardiographic systole.

‡Measurements obtained before surgery.

§Assessed with color-flow imaging.

symptoms and functional limitation (New York Heart Association functional class III) as well as substantial peak left ventricular outflow tract gradients of 100 and 65 mm Hg measured under basal conditions at cardiac catheterization or by Doppler echocardiography, respectively. The patients experienced severe symptoms despite trials of drug therapy (with β-blockers and/or verapamil) and dual-chamber pacing; as a consequence, both were referred for operation by one of us (B.J.M.).

Results

Preoperative Echocardiography

Transthoracic echocardiography performed before operation showed diffuse and marked left ventricular hypertrophy in both patients; the pattern of hypertrophy was asymmetrical in patient 1 and symmetrical in patient 2 (Table). Maximum wall thicknesses (in the anterior septum) were 32 and 19 mm, respectively.

Other relevant morphological (echocardiographic) features were present in both patients (Figure 1): (1) particularly exaggerated anterior displacement of the hypertrophied papillary muscles within the left ventricular cavity (as observed in the parasternal short-axis plane); (2) apparent direct continuity between anterolateral papillary muscle and anterior mitral leaflet, associated with decreased mobility of the mitral apparatus and muscular systolic apposition between papillary muscle and ventricular septum, most easily identifiable with the ultrasound beam rotated slightly (off the center of the left ventricular cavity) in the parasternal long-axis plane. SAM was observed in both patients; the anterior mitral leaflet made a characteristic sharp-angled bend and midsystolic septal contact (in patient 1) or a mild bend without septal contact (in patient 2) in the outflow tract ~15 to 20 mm below the aortic valve.

Operative Findings

The operating surgeon (G.K.D.) was initially prepared to perform a standard septal myotomy/myectomy1 in both patients. However, he immediately noted direct insertion of papillary muscle into anterior mitral leaflet (without interposition of chordae tendineae) as well as anterior malposition of the mitral apparatus within the outflow tract. This anomalous insertion involved the anterolateral papillary muscle in patient 2 and both papillary muscles (as well as a smaller third muscle between the 2) in patient 1.

Consequently, a novel operative strategy was devised in the operating suite to substantially reduce the outflow gradient (Figure 2). In both patients, a rectangular muscular resection was extended from just below the aortic valve into the midventricular region to the level of the attachments of the papillary muscles, a distance of up to 7 cm (compared with ~3 cm in the standard Morrow procedure)1; this trough
fascicles of left bundle. Also noted in D is partial liberation of anterolateral papillary muscle (which inserts aberrantly into anterior mitral leaflet) from its attachments to lateral free wall to enhance mobility of papillary muscle and mitral apparatus (performed in patient 2).

Postoperative Assessment

Twelve months after surgery, both patients were in sinus rhythm (with intraventricular conduction delay or left bundle-branch block). Each one subjectively reported striking reduction in limiting symptoms and marked increase in exercise capacity (ie, class III to I) (Table). Continuous-wave Doppler documented sharp reductions in peak instantaneous outflow gradient, to 0 mm Hg in patient 1 and 25 mm Hg in patient 2. Echocardiography identified the myotomy/myectomy trough to be located centrally in the anterior ventricular septum (ie, at the 12 o’clock position in the short-axis plane), with distal extension to the papillary muscle level. After surgery, the left ventricular outflow tract appeared to be widely patent throughout the cardiac cycle. SAM was abolished in patient 1 and was very mild (without mitral-septal contact) in patient 2 (Table).

Discussion

The patients presented here illustrate several important principles relevant to diagnosis and treatment of an important subset within the broad spectrum of HCM. Recognition of midcavity muscular obstruction due to anomalous papillary muscle insertion directly into anterior mitral leaflet is critical before HCM surgery is undertaken and should be contemplated in all operative candidates. Failure to identify this anomaly and relieve outflow obstruction has been reported to result in operative catastrophe.14

The midventricular location of muscular obstruction in our patients differs considerably from the usual morphological form of HCM, in which outflow obstruction is produced solely by subaortic systolic apposition of the mitral valve with the basal ventricular septum.8-11,14,15,17 We previously suggested that clinical recognition of this important congenital malformation in HCM is possible by echocardiographic identification of the direct rigid continuity between anterior mitral leaflet and papillary muscle (and the muscular midcavity obstruction between papillary muscle and septum).14 This image was observed in our patients only when the ultrasound beam was positioned away from the anatomic center of the left ventricular cavity in the parasternal long axis. Lack of mitral leaflet flexibility presumably results from the absence of the chordae tendineae that would normally be interpositioned between mitral valve and papillary muscle. For these reasons, we believe that treatments proposed in HCM as alternatives to surgery, such as DDD pacing18 or alcohol septal ablation,19 are unlikely to relieve outflow obstruction in this variant of the disease.

In contrast to other reported cases of HCM with anomalous papillary muscle,14 our 2 patients also showed SAM with acute systolic bending of the anterior leaflet in the basal outflow tract just below the aortic valve, probably constituting a second level of outflow obstruction and contributing to the measured gradient in patient 1. We propose that SAM resulted when a freely mobile margin of the anterior leaflet (unencumbered by the papillary muscle insertion) moved forward into the outflow tract toward the septum. Therefore, the presence of typical SAM10-12,15,20 does not exclude muscular midcavity obstruction due to anomalous papillary muscle insertion in patients with obstructive HCM.

Mitral valve replacement has previously been advocated in HCM for the surgical correction of direct papillary muscle insertion into the mitral valve, although this strategy is undesirable for young patients.14 Indeed, the novel operative approach described in this report was directed toward avoiding valve replacement in our 2 patients (17 and 35 years old). By creating an extended muscular resection well beyond the site of midven-
tricular obstruction (and liberating the anterolateral papillary muscle from its attachments in one), it was possible to substantially widen the mid and apical left ventricular cavity as well as the basal outflow tract. As a consequence of this surgery, greatly improved and virtually normal hemodynamics and functional capacity were restored to these patients.

References
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