REVIEW TOPIC OF THE WEEK

The Mitral Valve in Obstructive Hypertrophic Cardiomyopathy
A Test in Context

Mark V. Sherrid, MD,a Sandhya Balaram, MD,b Bette Kim, MD,c Leon Axel, MD, PhD,d Daniel G. Swistel, MD

ABSTRACT

Mitral valve abnormalities were not part of modern pathological and clinical descriptions of hypertrophic cardiomyopathy in the 1950s, which focused on left ventricular (LV) hypertrophy and myocyte fiber disarray. Although systolic anterior motion (SAM) of the mitral valve was discovered as the cause of LV outflow tract obstruction in the M-mode echocardiography era, in the 1990s structural abnormalities of the mitral valve became appreciated as contributing to SAM pathophysiology. Hypertrophic cardiomyopathy mitral malformations have been identified at all levels. They occur in the leaflets, usually elongating them, and also in the subvalvular apparatus, with a wide array of malformations of the papillary muscles and chordae, that can be detected by transthoracic and transesophageal echocardiography and by cardiac magnetic resonance. Because they participate fundamentally in the predisposition to SAM, they have increasingly been repaired surgically. This review critically assesses imaging and measurement of mitral abnormalities and discusses their surgical relief. (J Am Coll Cardiol 2016;67:1846–58) © 2016 by the American College of Cardiology Foundation.

Left ventricular outflow tract (LVOT) obstruction due to systolic anterior motion (SAM) of the mitral valve is a frequent cause of disabling symptoms in hypertrophic cardiomyopathy (HCM). First-line therapy is pharmacological, with beta-blockade, disopyramide, verapamil, or their combinations (1). However, in patients with systolic gradients ≥50 mm Hg who fail to reach relief of symptoms with pharmacotherapy or who have side effects, surgical myectomy is recommended by international guidelines as the preferred modality for relief of obstruction. An appreciation of mitral abnormalities in HCM has accumulated over the past 20 years (2-13). There has been a natural response by surgeons to this greater understanding of the contribution of mitral pathology to SAM. At myectomy, they have tried to avoid leaving unrepaird pathology by repairing the mitral valve (7-10,12-19). Diagnostic echocardiographic and cardiac magnetic resonance (CMR) discovery of mitral abnormalities in an elderly patient may directly lead to a judgment for surgical septal myectomy rather than alcohol septal ablation (ASA), because ablation only addresses the septal thickening. Patients with mitral valve abnormalities may be left with persistent SAM, gradients, and mitral regurgitation (MR) after ASA (20).

Guidelines support decisions to select surgery for patients with mitral structural abnormalities. The 2011 American guidelines state: “Additionally, specific abnormalities of the mitral valve and its support apparatus can contribute significantly to the generation of outflow tract obstruction, suggesting the potential value of additional surgical approaches (e.g., plication, valvuloplasty, and papillary muscle relocation) and making myectomy more appropriate than alcohol septal ablation in some patients” (21). The

From the aDivision of Cardiology, New York University Langone Medical Center, New York, New York; bDivision of Cardiac Surgery, Mount Sinai St. Luke’s and Mount Sinai Roosevelt Hospitals, New York, New York; cDivision of Cardiology, Mount Sinai St. Luke’s and Mount Sinai Roosevelt Hospitals, New York, New York; dDepartments of Radiology, Medicine, and Neuroscience and Physiology, New York University Langone Medical Center, New York, New York; and the eDivision of Cardiac Surgery, New York University Langone Medical Center, New York, New York. The authors have reported that they have no relationships relevant to the contents of this paper to disclose. Barry J. Maron, MD, served as Guest Editor for this paper.

Manuscript received November 18, 2015; revised manuscript received January 3, 2016, accepted January 14, 2016.
2015 European guidelines are more explicit: “Septal myectomy, rather than SAA, is recommended in patients with an indication for septal reduction therapy and other lesions requiring surgical intervention (e.g., mitral valve repair/replacement, papillary muscle intervention) Class I, Level of Evidence: C” (22).

HISTORICAL APPRECIATION OF MITRAL VALVE ABNORMALITIES IN HCM

In 1969, Shah et al. (23) reported with echocardiography that LVOT obstruction in HCM was caused by SAM and mitral-septal contract. Anterior displacement of papillary muscles in the left ventricle (LV) was noted in 1974 (24) and was followed by related publications (4,7,25,26). These investigators described how this displacement positions mitral leaflets anteriorly into the flow stream and also reduces their restraint, causing mitral slack, a necessary component of dynamic SAM (25,27). Investigators from the National Institutes of Health (later from Minneapolis) described direct insertion of an anomalous anteriorly displaced papillary muscle into the middle of the anterior mitral leaflet without intervening chordae (9). These displaced anterior papillary muscles can contact the septum with each beat, causing submitral LV obstruction. Papillary muscle or anomalous chordae insertion into the midleaflet can also tent the mitral leaflet anteriorly into the flow stream. Mitral leaflet elongation and its importance was prominently brought to light by these same investigators (2,3), who also described isolated posterior leaflet SAM occurring when this leaflet is particularly elongated (28). At about the same time, transesophageal echocardiography (TEE) showed elongated leaflets in patients coming to surgery (29).

The most common cause of MR in obstruction occurs when the posterior leaflet is not long or mobile enough to move anteriorly with the anterior leaflet, resulting in poor coaptation (29–31); MR is reduced or eliminated by abolition of SAM, either surgically or with optimal pharmacological therapy (12,29,31). The important point is that resolution of the SAM is the therapeutic goal; this improves coaptation and the MR. In this review, mitral abnormalities are discussed according to their prominence and relative frequency in patients coming to surgery. TEE is necessary to resolve any ambiguities about the cause of obstruction when pre-operative imaging is suboptimal and always in the operating room immediately before and after cardiopulmonary bypass. The final operative plan should be confirmed after review of TEE immediately before cardiopulmonary bypass. Because CMR has been increasingly used to precisely demonstrate abnormalities of the mitral apparatus in HCM, we will discuss its utility in detail.

ELONGATED ANTERIOR AND POSTERIOR MITRAL LEAFLETS

The large majority of patients with obstructive HCM have elongated anterior and posterior mitral leaflets, as compared with normal subjects. Elongation has been noted pathologically, on echocardiography, and on CMR (2,3,5,6,12,29,32). In obstructive HCM, the anterior leaflet averages 34 mm versus 24 mm in normal hearts (5,12,29). We have termed this abnormality the “nightcap” mitral valve because of its typical appearance on apical views at the moment of systolic coaptation (Figure 1). Elongated leaflets extend (protrude) into the LV cavity well above the plane of the mitral annulus, a mean of 26 mm above the annulus compared with 13 mm in normal hearts (5). Often, a residual portion of the anterior mitral leaflet extends past the point of coaptation (12,33), which is important in SAM pathophysiology because it is not

FIGURE 1 "Nightcap" Mitral Valve

Apical 3-chamber view in obstructive hypertrophic cardiomyopathy patient just after leaflet coaptation. Tips of the elongated leaflets protrude 24 mm above the mitral annulus, higher into the left ventricle than normal (double red arrow). As flow courses around the septal bulge, it catches mitral leaflets and sweeps them into the septum.
constrained by the LV–left atrium pressure difference. Rather, it is only bounded by the LV and thus freely moves with LV flow, even at low velocity (33). The residual leaflet usually contacts the septum first (Figure 2).

The protruding mitral leaflet plays an important role in the pathophysiology of SAM. Late diastolic and early systolic flow strike the posterior surfaces of the protruding leaflets with a high angle of attack and push them into apposition with the septum (5,34,35). After mitral-septal contact, the pressure difference itself pushes the obstructing mitral leaflet further into the septum (34). Mitral leaflets are longer in patients with obstructive HCM than with nonobstructive HCM (5). An increased ratio of anterior leaflet length to LVOT diameter is associated with resting and provokable obstruction (6). LVOT obstruction may occur after exercise in patients with completely normal LV wall thickness; here the mitral leaflets are elongated and papillary muscles are anteriorly positioned in the LV cavity (36). Echocardiographic and some CMR reports have used different methods for measuring the anterior mitral leaflet: echocardiographic papers have included the aortic-mitral continuity (5,12,29), whereas some CMR papers have excluded this region (6).

A variety of surgical approaches have been devised to correct anterior leaflet elongation. These have included plication parallel to the long axis of the valve, termed “vertical plication” (14); and plication perpendicular to the long axis, termed “horizontal plication” (15,16,19,37,38). In short-axis views, the body and tip of the anterior leaflet (usually A2) billow out into the outflow tract, catching the forward flow of blood. Decreasing leaflet width in a medial/lateral dimension through vertical plication limits billowing (14). However, we have found that vertical plication often perturbs the coaptation line of the anterior leaflet and causes central MR. With horizontal plication, as devised by Dr. Swistel, a line of sutures is placed horizontally across the pliable main body of the leaflet, within the aortomitral curtain. This stiffens the anterior leaflet, limits billowing, decreases its area exposed to drag, shortens it, and prevents its leading edge from reaching the septum (12,16,19). Shortening of the leaflet is achieved without disturbing the coaptation line. Moreover, the procedure is quite simple. Done through the aortotomy, 4 or 5 sutures of 5-0 prolene are placed, using vertical mattress technique, horizontally across the leaflet, just superior to the bulk of the chordal attachments (Figure 3). The amount of plication is easily chosen from 2 to 6 mm, depending on need, as determined by pre-operative echocardiography (12) (Figure 4, Online Videos 1 and 2).

When excessive residual leaflet length contributes to obstruction, excision has been utilized as an ancillary or alternative procedure to leaflet plication. A2 is often an isolated segment of excessive length, with 2 or 3 extremely slack and thinned-out chordae. Pre-operative analysis of the echocardiogram often reveals this segment to be inconsequential to coaptation and competence, and direct examination in the operative field suggests that these chordae play no role in preventing flail if removed. A segment of 2 to 5 mm can at times be simply and safely excised.

FIGURE 2 3-Dimensional TEE of the Residual Leaflet at the Moment of Mitral-Septal Contact

(Left) The 2-dimensional frame for orientation, showing systolic anterior motion and mitral-septal contact (white arrow). (Right) Corresponding 3-dimensional frame, orientation is viewing mitral valve en face from left ventricular (LV) cavity, looking up at LV outflow tract. Obstructing residual leaflet is shown with a white arrow. LV outflow tract orifices, displayed in black, are on either side of the residual leaflet, shown with red arrows. High drag profile of the residual leaflet can be appreciated. TEE = transesophageal echocardiography.
(Figure 5). As in plication, judgment on the basis of experience must be exercised when determining the safety of residual leaflet excision.

Another method to stiffen the mitral leaflet has been termed anterior mitral leaflet extension. Here, the anterior leaflet is vertically incised and a stiff pericardial patch is inserted to prevent billowing of the leaflet (17). This underscores the principle that alternate surgical approaches can be used in an individual patient to achieve optimal relief of LV outflow tract obstruction (18). However, inserting a pericardial patch requires incising the valve, and can be time-consuming. The simplest, least time-consuming technique should be used to repair mitral abnormalities, to avoid increasing the complexity of an already complex operation. Note that virtually all leaflet repairs described in this entire review are performed without insertion of an annuloplasty ring.

**ANTERIOR AND BASILAR DISPLACEMENT OF THE ANTEROLATERAL PAPILLARY MUSCLE**

Anterior displacement of the papillary muscles results in an anterior position of the coaptation plane of the mitral valve in the LV cavity (4,7,26,39). This causes a crucial overlap of the inflow and outflow portions of the LV that predisposes to SAM. We have made direct surgical inspection of the papillary muscles in HCM patients after extended myectomy and contrast them to those of hypertrophied patients with aortic stenosis undergoing valve replacement. The 2 most common pathogenic abnormalities of the papillary muscles in HCM are: 1) an anterior and basilar displacement of the base of the anterolateral papillary muscle (Figure 6); and 2) abnormal muscular connections between its head and the anterolateral wall, inserting into or near the A1 scallop (12). Related CMR research reported by Kwon et al. (40) has shown anterior displacement of the anterolateral papillary muscle and a higher frequency of bifid papillary muscles; there was closer proximity between the superior papillary muscle and the septum in patients with obstruction due to SAM (Figure 6). Obstruction
magnitude correlated with anomalies, independent of septal thickness. Bifid papillary muscle, seen on CMR in Figure 6B, can appear on the echocardiographic short-axis view as an anterior extension and displacement (Figure 6A).

The extended myectomy, as described by Messmer (7) and by Schoendube et al. (8), includes release of the anterolateral papillary muscle by extending the resection laterally into the free wall above its base and thinning the hypertrophied heads (12). Muscular connections between the papillary muscle head and LV free wall are easily visualized when examining the ventricular chamber during myectomy. We divide and excise these attachments, usually with a pituitary rongeur (15,16,19). These techniques allow the anterior papillary muscle to fall posteriorly, drawing the anterior mitral leaflet with it, explicitly out of the ejection stream. Papillary release brings the plane of the mitral annulus and aortic valve into a more normal parallel orientation (41). Other surgeons have devised novel surgical approaches for the contribution of anomalous papillary muscles to SAM (40,42).

**INSERTION OF ANTEROLATERAL PAPILLARY MUSCLE DIRECTLY INTO THE MIDANTERIOR MITRAL VALVE LEAFLET**

Insertion of a head of the anterolateral papillary muscle directly into the middle of the anterior mitral valve leaflet without intervening chordae is demonstrated in Figures 7 to 9, by CMR in Online Videos 3 and 4, and by 3-dimensional echocardiography in Online Video 5. It may cause obstruction by apposition of the papillary muscles with the septum on every beat (9,18,43); when this occurs, flow accelerates in the submitral region. Off-axis echocardiographic views may be necessary for demonstration of this abnormality (18). CMR (through its superior lateral resolution) and TEE can reliably demonstrate this morphological abnormality in the LVOT, which can alter management decisions. They should be performed whenever there is ambiguity about the level of obstruction (43). With both techniques, tomographic planes can be chosen that show the relevant abnormalities and their anatomic relationships. Surgeons from the Mayo Clinic showed that accessory papillary muscle heads and anomalous chordae could be removed if they did not support the leading edge of the valve leaflet (Figure 7) (18). Alternatively, the most anterior component can be excised, leaving a posterior component attached to minimize the risk of flail leaflet. When the anomalous papillary muscle is very large and directly obstructs, but excision is not possible, we have successfully used a longitudinal resection to thin it, even to its base. This is combined with extended myectomy, to the degree allowed by the septal thickness.

**ANTERIOR MITRAL TENTING BY PAPILLARY MUSCLES AND FIBROTIC, RETRACTED SECONDARY CHORDAE**

Anomalous anterior papillary muscles or fibrotic, retracted secondary mitral chordae may also contribute to obstructive pathophysiology and SAM, even if they do not directly obstruct in the submitral area. They lift and tent the mitral valve anteriorly, so it is pre-positioned in the flow stream, predisposing to SAM (18,44) (Figure 9B).

**ELONGATED POSTERIOR LEAFLET WITH SAM**

When an isolated elongation of the posterior leaflet causes SAM, the residual leaflet protrudes through the spaces between the chordae to contact the septum (28) (Figure 10, Online Video 6). Although an uncommon cause of mitral-septal contact, it nonetheless can cause severe obstruction, and its identification before surgery is essential. Surgical alleviation has always included some degree of septal resection; thus, the initial exposure has been via the standard transaortic approach. With this exposure, one can often access the posterior leaflet to excise a portion of the residual leaflet. Alternatively, if the posterior leaflet is not
accessible, we have approached the valve via left atrial incision and performed a narrow triangular resection, without an annuloplasty ring.

**CALCIFICATION OF THE MITRAL LEAFLETS OR ANNULUS**

Occasionally, calcification of the mitral leaflets or annulus occurs in a patient who also has SAM with mitral-septal contact. It is generally the goal of 21st-century myectomy surgery to have patients leave the operating room with their native mitral valves, to avoid the sequelae of mitral valve replacement (MVR) (i.e., chronic anticoagulation or late prosthesis degeneration) (45). However, it is occasionally impossible to salvage the native mitral valve. A clue that calcification or fibrosis is causing MR is anterior or mid-direction of the regurgitant jet. With regurgitation caused only by SAM, the jet is usually posteriorly and laterally directed.

With even moderate annular calcification or anterior leaflet fibrosis, manipulation of the tissue is

**FIGURE 6** Anterior Displacement of Hypertrophied Anterolateral Papillary Muscle

(A) Echocardiographic short-axis view of extra anteriorly displaced papillary muscles (red arrows). This bifid anteriorly displaced papillary muscle positions the mitral valve leaflets anteriorly in the left ventricular chamber, where they overlap with the ejection flow stream. (B) Cardiac magnetic resonance 4-chamber view showing hypertrophied bifid anterolateral papillary heads (red arrows), with the superior head in close proximity to the septum (yellow arrows). Anterior displacement also decreases posterior restraint on mitral leaflets.

**FIGURE 7** TEE of Hypertrophied Anomalous Anterolateral Papillary Muscle Head Inserting Directly Into Midanterior Mitral Leaflet Without Intervening Chordae

(A) Diastolic 0° view. Red arrows indicate anomalous papillary muscle in frames A to C. (B) Systolic frame, 0° view, showing apposition of the anomalous papillary muscle with the septum, causing severe obstruction. (C) Systolic frame, 120°, showing apposition of the anomalous papillary muscle with the septum, causing severe obstruction. (D) Post-operative transthoracic parasternal view. Because the papillary muscle did not support the leading edge of the anterior leaflet, it could be safely removed. Yellow arrowheads indicate septal myectomy. TEE = transesophageal echocardiography.
not possible. On the basis of generalized results with mitral valve repair for other pathologies (i.e., rheumatic disease), MVR might be necessary. However, if the primary pathology is an excessive residual leaflet that does not participate in coaptation, then residual leaflet excision with myectomy may be performed.

**RELATIVELY THIN SEPTUM**

Another situation in which MVR was previously the primary option is the presence of a relatively thin septum ≤18 mm. A limited myectomy would be performed because of concern that a ventricular septal defect might result; alone, this would not be enough of an intervention to alter flow adequately to relieve SAM. However, these patients invariably have one or multiple of the mitral valve abnormalities that lend themselves to the operative repairs described previously. Combined with a necessarily limited myectomy, this has sufficed to relieve SAM (12,42,44). We now routinely perform a limited myectomy on these patients, along with mitral repair without MVR. A recent report of transaortic chordal cutting in patients with relatively mild septal thickening has underscored the utility of mitral repair in this group (44). In 39 patients, intraoperative TEE and direct surgical inspection detected that the anterior mitral leaflet was tented up into the outflow tract by retracted and fibrotic secondary chordae. A median of 3 secondary chordae were resected, in addition to necessarily

**FIGURE 8** CMR of Bifid Anomalous Anterolateral Papillary Muscle With Insertion Into the Midanterior Mitral Leaflet

(A) Early systolic 4-chamber view. Red arrows indicate anomalous muscle head in frames A and B. Note its proximity to the septum. (B) Early systolic 3-chamber view. Note that papillary muscle tents up the middle of the anterior mitral leaflet (yellow arrow). (C) The 3-chamber view later in systole from a sequence demonstrating flow velocities in outflow tract. Narrowing of the outflow tract between the papillary muscle and the septum causes flow acceleration due to obstruction. Note that septal thickening is modest in this patient. See Online Videos 3, 4, and 5. CMR — cardiac magnetic resonance.

**FIGURE 9** Obstructing Anomalous Papillary Muscle

(A) Cardiac magnetic resonance (CMR) showing midsystolic apposition (large arrowheads) of ventricular septum and anomalous papillary muscle (arrows) inserting directly into the anterior mitral leaflet (small arrowheads). Reprinted with permission from Rowin et al. (43). (B) Anomalous papillary muscle (red arrow) tapers and then inserts into the midportion of anterior leaflet and tents it anteriorly (yellow arrow). Orange arrow indicates leaflet coaptation. There was no papillary muscle–septal contact. Rather, there was mild resting systolic anterior motion, and severe systolic anterior motion with high gradient after exercise. Ao — aorta; LA — left atrium; VS — ventricular septum.
shallow myectomy; there was virtual abolition of high resting gradients and decrease in tenting. The Central Illustration shows common anomalies of the mitral leaflets and papillary muscles that contribute to obstruction in HCM.

CLINICAL AND ECHOCARDIOGRAPHIC RESULTS OF THE RESECT-Plicate-RELEASE OPERATION

From 1997 to 2015, we operated on 252 patients with obstructive HCM for symptoms and gradients resistant to pharmacological therapy (1,15,16). The resect-plicate-release (RPR) techniques used were adopted and modified from the reported surgical experience of others (7,8,9,14,18). The mean age was 56 ± 14 years; 55% were men. The average New York Heart Association functional class was 2.8 ± 0.6. The majority (96%) of these patients had surgery for SAM and mitral-septal contact. The remaining patients had mid-LV obstruction.

In the patients who had SAM with mitral-septal contact, LVOT gradients were 61 ± 8 mm Hg and provoked gradients were 111 ± 41 mm Hg. Septal thickness was 24 ± 5 mm (range 13 to 40 mm). Preoperative echocardiography (both transthoracic and TEE) and now CMR provide the basis for the extent of septal resection; echocardiography and direct
Anomalies of Mitral Valve and Papillary Muscles

surgical inspection provide the basis for individual case-by-case judgments regarding the need for mitral plication and papillary muscle release. Plication was performed in 66% of patients; papillary release was performed in 86%. MVR was necessary in 6%. Concomitant procedures were performed in 35%, with coronary artery bypass graft as the most common (16%).

**Text in red and in quotations** denote our shorthand method for categorizing our pre-operative assessment of which procedures should be done.

*Additionally, direct surgical inspection of anterior leaflet for slack determines indication for and extent of plication. Extensive mitral calcification of anterior leaflet or annulus is a contraindication to plication. Patients with extensive calcification require mitral valve replacement. †Papillary muscle release is predicted on pre-operative echocardiography by anterior extension of the lateral papillary muscle. §Dual-chamber pacing with short AV delay is also a therapeutic option for frail elderly patients. AML ¼ anterior mitral leaflet; ASA ¼ alcohol septal ablation; AV ¼ atrioventricular; HCM ¼ hypertrophic cardiomyopathy; MVR ¼ mitral valve replacement. Modified with permission from Halpern et al. (12).
Operative mortality was 0.4%. The post-procedural complication rate was 15%; the majority of complications were respiratory compromise in older patients with multiple comorbidities. Complete heart block requiring pacemaker insertion occurred in 6%. Early reoperation was required in 2 patients (0.4%): 1 for ventricular septal defect repair and 1 for MVR. Length of stay was 7 ± 5 days. There was marked improvement in New York Heart Association functional class for 96%. In our 2012 report, survival at 1, 5, and 10 years was 98%, 98%, and 92%, respectively (16).

We recently reported on detailed echocardiography before and after the resect-plicate-release repair in 77 patients. Pre-operative echocardiographic anterior mitral length provides a basis to recommend performance of plication and defines its extent. We recommend plication when the anterior leaflet is >30 mm long or >17 mm/m2. Plication decreases anterior leaflet length by 16%, residual leaflet length by 33%, and protrusion into the LV above the mitral annular plane by 24%. After papillary muscle release, there was a decrease in the distance from mitral coaptation to the posterior LV wall. Surgery abolished severe SAM and resting gradients and reduced MR (12). An algorithm describing our use of echocardiography, both transthoracic and TEE, to select which of the “resect,” “plicate,” or “release” procedures to apply in an individual patient is presented in Figure 11.

**SHOULD MITRAL SURGERY ACCOMPANY MYECTOMY?**

There is controversy about whether mitral valve abnormalities should be surgically repaired at the time of myectomy (46). Some centers generally perform mitral repair in addition to extended myectomy (8,10,14–17), whereas it is the judgment of others to usually just perform myectomy (1,47). It may be a natural surgical approach to comprehensively address all discovered contributors to a patient’s SAM, and to not leave unrepaired pathology. However, because a clinical trial has not been performed comparing results of myectomy plus mitral repair versus myectomy alone, these decisions currently rest in the realm of clinical judgments.

Myectomy is considered to be among the most challenging of all surgical procedures for acquired heart disease. The paucity of centers applying isolated myectomy provides silent evidence of a precipitous learning curve; there is difficulty gauging “too much versus too little myectomy.” Residual obstruction requiring reoperation was previously reported after isolated myectomy (48,49); which we have not seen in our experience (12,15,16). For inexperienced surgeons, isolated myectomy is an imprecise tool; there is a reported myectomy hospital mortality of 6% at non-referral centers in the U.S. Nationwide Inpatient Sample (50). In contrast, there is very low 30-day post-operative mortality, 0.5%, in patients operated on at dedicated HCM specialty centers (51).

Concomitant mitral surgery, particularly, RPR myectomy operation, provides a buffer against failure that may occur because of imprecision in depth and extent of myectomy. Mitral repair remediates the mitral half of obstructive HCM physiology; myectomy addresses only the flow abnormality related to the septal bulge. The decision of whether to repair detected abnormalities of the mitral valve at myectomy should be undertaken jointly by the referring cardiologist and surgeon. Technical aspects of mitral plication are undemanding and can be readily learned. Papillary muscle release is more technically challenging and requires experience. However, echocardiographic and clinical results of its usefulness for relieving SAM suggest that an effort to master the technique should be pursued. Given the experience required to master these surgical techniques, patients who are resistant to maximal pharmacological therapy should be referred for evaluation and treatment to dedicated HCM centers (51).

**WHY IS THE MITRAL VALVE ABNORMAL IN HCM, WHICH IS CAUSED BY MUTATIONS IN GENES CODING FOR SARCOMERIC PROTEINS?**

Leaflet elongation has been observed in subjects who have an HCM-associated mutation but who have not yet developed thickening, suggesting that mitral elongation is a primary phenotypic expression of HCM and is not acquired due to stretch from SAM (6). Elongated leaflets are not myxomatous (2). Abnormal cell lines that elongate to form valves appear to originate from epithelial-derived cells from coelomic mesothelium that migrate to endocardial cushions (52). These cells may respond to paracrine or to local mechanotransduction signals with the observed developmental abnormalities (11,53).

**CONCLUSIONS**

In many patients with HCM, mitral valve abnormalities play a major contributing role in LV outflow...
obstruction. In patients who come to surgery at many centers, the judgment is made to repair contributing mitral abnormalities whenever they are detected, regardless of the magnitude of septal hypertrophy. Mitral abnormalities play a particularly important role in patients with mild or moderate hypertrophy. These patients should not be treated by septal reduction alone (ASA in particular); surgical mitral valve repair in association with septal myectomy is the preferred approach. A number of surgical techniques are available to repair the mitral valve in HCM; given the great diversity of mitral abnormalities, the proper technique should be selected for the individual patient.

**ACKNOWLEDGMENTS** The authors thank Drs. Waseem Shami and Robert Nampiaparampil for their technical expertise.

**REPRINT REQUESTS AND CORRESPONDENCE:** Dr. Mark V. Sherrid, Division of Cardiology, New York University Langone Medical Center, 530 First Avenue, New York, New York 10016. E-mail: mark.sherrid@nyumc.org.

**REFERENCES**

Mitral Valve in Obstructive Hypertrophic Cardiomyopathy

et al
Sherrid
JACC VOL. 67, NO. 15, 2016


Kwon DH, Seter RM, Thamilarasan M, et al. Abnormal papillary muscle morphology is independently associated with increased left ventricular outflow tract obstruction in hypertrophic cardiomyopathy. Heart 2008;94:1295-301.


