STATE OF THE ART REVIEW

Intraoperative Two- and Three-Dimensional Transesophageal Echocardiography in Combined Myectomy-Mitral Operations for Hypertrophic Cardiomyopathy

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Transesophageal echocardiography is essential in guiding the surgical approach for patients with obstructive hypertrophic cardiomyopathy. Septal hypertrophy, elongated mitral valve leaflets, and abnormalities of the subvalvular apparatus are prominent features, all of which may contribute to left ventricular outflow tract obstruction. Surgery aims to alleviate the obstruction via an extended myectomy, often with an intervention on the mitral valve and subvalvular apparatus. The goal of intraoperative echocardiography is to assess the anatomic pathology and pathophysiology in order to achieve a safe intraoperative course and a successful repair. This guide summarizes the systematic evaluation of these patients to determine the best surgical plan. (J Am Soc Echocardiogr 2018;31:275-88.)

Keywords: Hypertrophic cardiomyopathy, Echocardiography, Cardiac anesthesiology, Left ventricular outflow tract obstruction, Cardiac surgery

Hypertrophic cardiomyopathy (HCM) is a genetic disease of the myocardium characterized by unexplained left ventricular (LV) hypertrophy.[1] Epidemiologic studies estimate a prevalence of approximately 0.2% of the general population, or one in 500 individuals.[2,3]

Patients affected by HCM may present in a variety of ways. Signs of disease progression include heart failure with exertional dyspnea, angina, syncope, and atrial fibrillation; infrequently, the course of HCM is unexpectedly ended by sudden cardiac death.[1] An important therapeutic goal in these patients is to decrease or eliminate obstruction. Initially, first-line pharmacologic treatment includes β-blockers, disopyramide, verapamil, or a combination of these medications.[4] In patients with persistent symptoms refractory to medical management or who experience significant side effects, surgical intervention may be required.[5] Guidelines indicate that alcohol septal ablation should be reserved for patients who are too old, too frail for surgery, or inalterably opposed to sternotomy.[1]

Transthoracic echocardiography (TTE) is critical to the initial preoperative workup.[1] A preliminary plan for surgical intervention can be formed on the basis of this examination, and the patient can be recommended for surgery on the basis of the findings. Exercise stress echocardiography may also be used to assess inducible LV outflow tract (LVOT) gradients, mitral regurgitation (MR), and provokable arrhythmia.[6] Increasingly, magnetic resonance imaging is used in the initial evaluation of patients with HCM, including preoperative evaluation of the septum and mitral valve apparatus,[7-10] though it is our impression that it is not as useful as echocardiography for planning surgery, unless echocardiographic imaging is suboptimal. In such patients, elective outpatient preoperative transesophageal echocardiography (TEE) is indicated to clarify any ambiguity before surgery.

Surgical septal myectomy is a technically demanding procedure. Inadequate resection results in persistent obstruction and symptoms, while excessive or poorly placed myectomy can produce iatrogenic heart block and ventricular septal defects and consequent high morbidity and mortality, reported as high as 5.9%.[11] In sharp contrast, data collected from dedicated HCM centers have shown remarkably decreased operative mortality (0.4%), explained primarily by surgical experience, advances in surgical technique, improved myocardial preservation, and decreased cardiopulmonary bypass times.[12,13]

Patients with HCM not only have myocardial hypertrophy but also malformations of the mitral valve and subvalvular apparatus.[14] Current guidelines recommend surgical intervention over alcohol septal ablation in patients with concomitant abnormalities of the mitral valve and the submitral apparatus, as a class I recommendation.[1,15] The decision of whether to intervene on the mitral valve and the subvalvular apparatus in addition to myectomy is controversial. Dedicated HCM centers have found success with both approaches: some perform extended myectomy with mitral valve or subvalvular interventions in selected patients[16-21]; at other centers, extended myectomy is performed without operating on the mitral valve.[22-24]

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Abbreviations

3D = Three-dimensional

AML = Anterior mitral valve leaflet

HCM = Hypertrophic cardiomyopathy

LV = Left ventricular

LVOT = Left ventricular outflow tract

MR = Mitral regurgitation

SAM = Systolic anterior motion

TEE = Transesophageal echocardiography

TTE = Transthoracic echocardiography

Clinical judgment and experience determines the best operative choices for these patients.

PREOPERATIVE EXAMINATION

The comprehensive assessment of these abnormalities in a systematic fashion directs the surgical plan for the operation.[19,20,25,26]

According to the 2011 American College of Cardiology Foundation/American Heart Association guidelines, TEE is recommended for the intraoperative guidance of surgical myectomy as a class IB recommendation.[1] Because the patient already has confirmation of HCM on preoperative TTE, TEE is generally performed after the induction of general anesthesia in the operating room; a final surgical plan may then be formed among the cardiologist, cardiac anesthesiologist, and surgeon at that time. Because HCM pathophysiology is complex and may involve, to varying degrees, septal thickening of a heterogeneous nature, mitral leaflet elongation, and subvalvular abnormalities, at our hospital we find it useful for the referring HCM cardiologist to participate in operative planning; pursuant to this end, we have established a secure Internet-based system so that live images in the operating room can be viewed remotely from the office and discussed.

Interventricular Septum

LV hypertrophy, particularly of the septum, is a cardinal feature of HCM. The nomenclature for HCM has been refined in recent years. Idiopathic hypertrophic subaortic stenosis has been largely discarded after the realization that patients with nonobstructive HCM can have limiting symptoms and be prone to sudden cardiac death; current nomenclature acknowledges this by the dichotomy between obstructive and nonobstructive HCM.

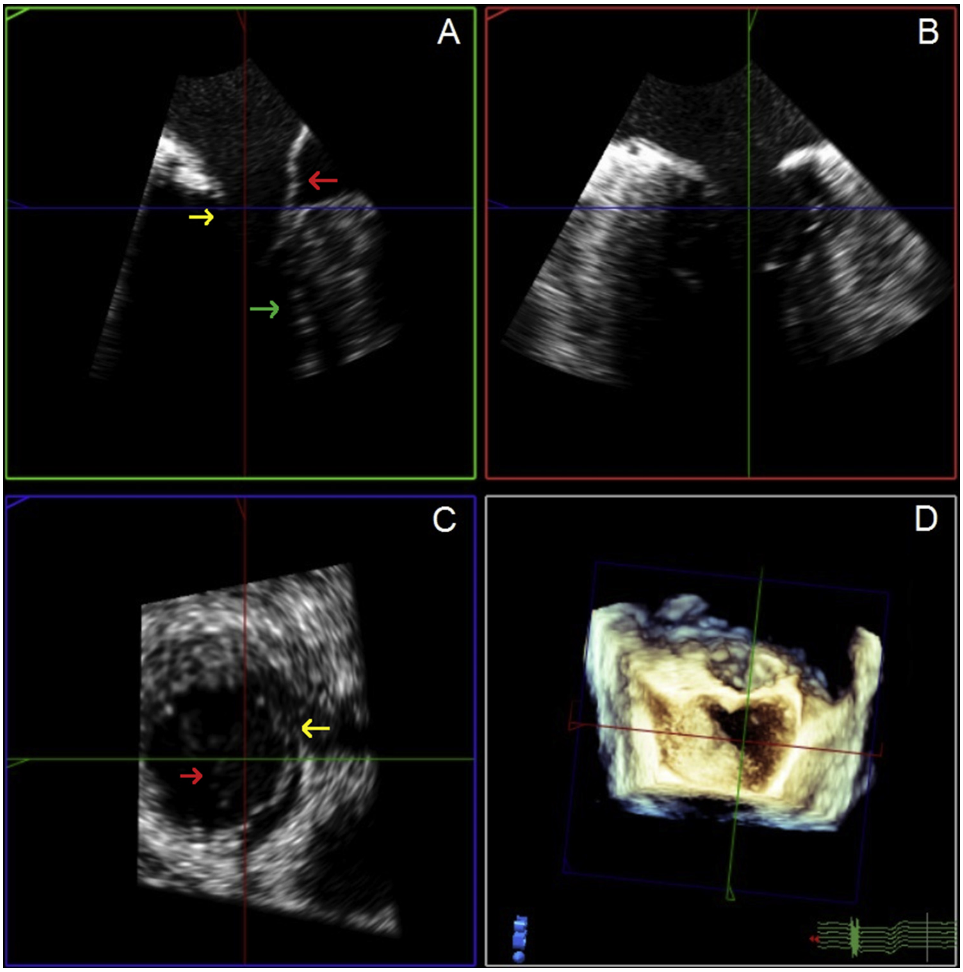

Figure 1 Multiplanar reconstruction is useful to determine accurate septal wall measurements. In this figure, the blue plane is aligned at the mitral valve leaflets **(A, B)**. Using this as a reference point, the green plane is positioned to bisect the mitral valve along the anterior leaflet *(red arrow)* and posterior leaflet *(yellow arrow)* at A2:P2 **(B, C)**. The septum visualized in the green plane will therefore be the anterior septum *(green arrow)* **(A)** and can be measured precisely. Additionally, the green plane can be shifted in either direction to determine wall thickness along the entire septum. **(D)** A 3D reconstruction with the position of the different planes.

HIGHLIGHTS

- Intraoperative two- and three-dimensional TEE is crucial for HCM patients.
- Measure septal thickness, mitral leaflet length, and assess subvalvular apparatus.
- The septum is best measured from the three-chamber long-axis and transgastric views.
- Use 3D TEE from the LV side, “looking up”, to visualize mitral valve.
- Interactive communication with the surgeon before and after bypass is key.

Determining the extent and location of interventricular septal wall thickening is crucial and directs the resection. The reader should be aware that the thickness of the septum may vary from initial TTE to the ensuing preoperative TEE and to the subsequent arrested heart on cardiopulmonary bypass. Most anesthetics are vasodilators, and positive pressure ventilation after induction of general anesthesia decreases venous return. Perhaps because of both of these effects, the heart may be unloaded. We have noticed that wall thickness is greater on postinduction TEE than on preoperative ambulatory TTE. Decreased chamber size could possibly explain the increase in wall thickness; alternatively, this might be explained by differences in planes between imaging modalities.

The best views to evaluate the interventricular septum are the midesophageal long-axis view (135°), transgastric midpapillary short-axis view (0°), and midesophageal four-chamber view (0°).

The midesophageal long-axis view is useful for measuring anterior septal wall thickness, which is the principal target for extended myectomy. This view is correlated with wall thickness on the transgastric short-axis views. The myectomy is directed primarily toward the anterior septum. The goal is to thin the septum adequately to divert outflow anteriorly, away from the mitral valve, and allow it to leave the heart without catching the mitral valve leaflets.[27,28] To accomplish this goal the myectomy often must be extended down to the level of the papillary muscles, resecting the mid-anterior septum.[16,19,23,25,27,28] Measurements of the interventricular septum are made near the end of diastole at or before the R wave, when the ventricle is at its largest diastolic dimension.

The portion of the septum just below and adjacent to the aortic annulus generally is not involved in the pathophysiology of obstruction in HCM. Thinning this portion of the septum should be avoided during myectomy to preserve the atrioventricular node and to avoid the thin membranous portion of the septum.[25]

Three-dimensional (3D) volume acquisition can provide precise confirmatory midesophageal long-axis views by acquiring a 3D data set and then aligning a plane to bisect the mitral valve at A2:P2. The septum in this view correlates with the anterior septum (Figure 1). Although one can align a plane to create a midesophageal five-chamber view (0°) from the 3D data set to assess the anterior septum, we prefer the midesophageal long-axis plane, as this view and measurement correspond anatomically to the surgical approach through the aortotomy. Tomographic measurements of the anterior septal thickness can then be checked at both the subaortic and midventricular levels (Figure 2).

The transgastric midpapillary short-axis view is useful for measuring the anterior septum, the posterior septum, and the anterior LV free wall in the same plane. As we advance the probe, we progressively visualize the basal, mid, and apical portions. Alternatively, anteflexion of the probe shows more of the basal layer, while retroflexion reveals more of the apical segments. Biplane imaging can be used to ensure the image is a transverse cut of the left ventricle. These measurements supplement findings from the midesophageal long-axis view (Figure 3).

The midesophageal four-chamber view will often show portions of the posterior septum. Generally, less myectomy is performed in the subaortic posterior septum to avoid complete heart block, though the excision may be extended into the mid-LV posterior septum, so its thickness at this level should be defined.

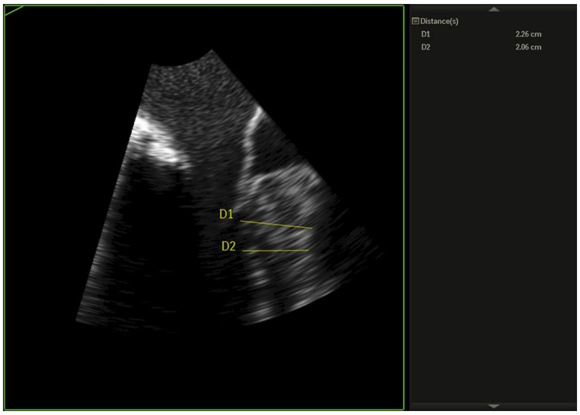

Figure 2 The green plane can be magnified so precise measurements are obtained for the anterior septum at subaortic and midventricular levels.

Mitral Valve

Septal hypertrophy and anterior displacement of the mitral valve by anteriorly positioned papillary muscles decreases the distance from the septum to the plane of mitral valve coaptation. There is a crucial overlap between the inflow and outflow portions of the left ventricle.[29] As flow sweeps around the septal bulge, flow drag, the pushing force of flow, catches the mitral valve from behind and sweeps the anterior mitral valve leaflet (AML) into the septum.[30] Systolic anterior motion (SAM) can disrupt mitral valve coaptation and result in MR. MR secondary to SAM often is directed posteriorly and laterally (Figure 4). A regurgitant jet aimed anteriorly suggests a structural defect in the mitral valve itself, such as calcification, thickening, or a cleft.[1] During the postinduction examination, patients often have decreased preload and decreased afterload, both of which can predispose to SAM.[31]

Compounding the problem of anterior mitral displacement, the majority of patients with HCM have elongated mitral valve leaflets.[7,14] The anterior leaflet may protrude into the LVOT and contribute to obstruction. We have called this the “nightcap mitral valve” because of its characteristic appearance on the apical three-chamber view on TTE, midesophageal long-axis view on TEE, or transgastric long-axis view on TEE (Figure 5).

Leaflet lengths are measured in the midesophageal long-axis view (135°). On two-dimensional imaging, we have measured this length from the tip of the AML in A2 to the aortic annulus, at the insertion of the noncoronary leaflet.[32] This measure includes the intervalvular fibrosa but is more reproducible than the distance from the tip to the hinge point of the leaflet. Although one may measure only the length of the leaflet up to the point of coaptation,[33] we believe it is important to include the entire leaflet, particularly the portion past the

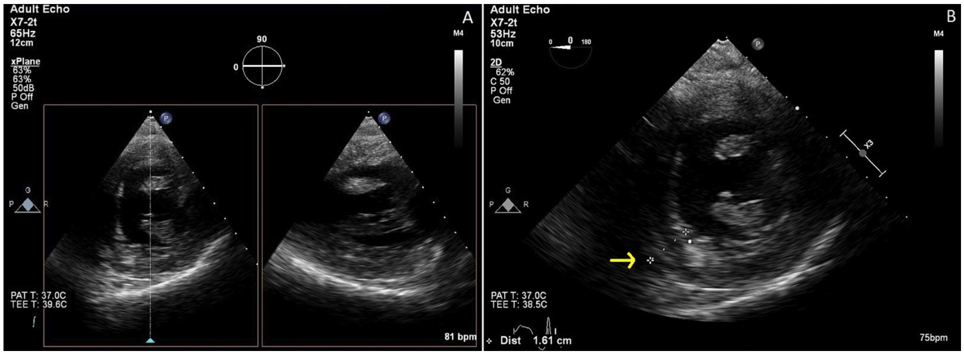

Figure 3 Midtransgastric view is obtained to measure septal wall thickness. Biplane mode can be used to confirm the cut is appropriately aligned **(A)**. Subsequently, a measurement of the anterior septum can be performed *(yellow arrow)* **(B)**.

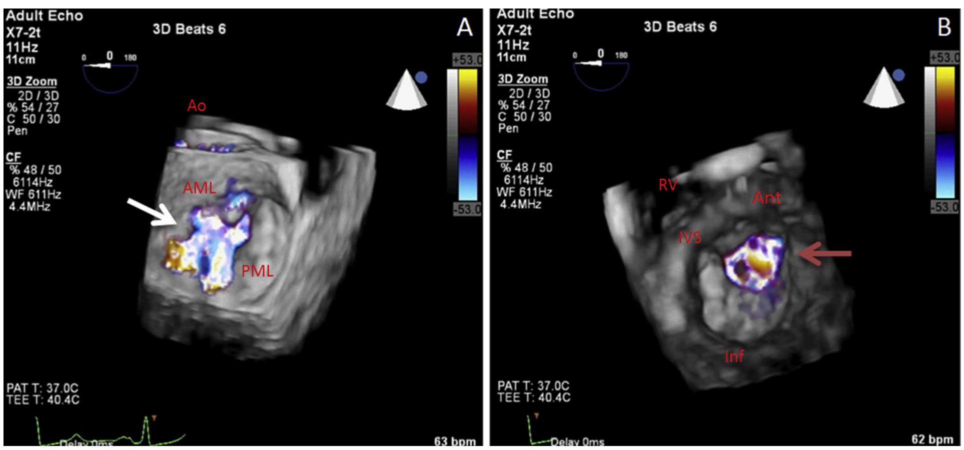

Figure 4 Three-dimensional multibeat color acquisition can be extremely useful to elucidate or confirm preoperative findings. **(A)**, oriented from the left atrial side, shows a severe broad jet of mitral regurgitation *(white arrow)* directed posteriorly and laterally, suggesting an etiology secondary to SAM. **(B)**, oriented from the LV side, reveals turbulence *(red arrow)* as blood is ejected through the LVOT during systole. *Ant*, Anterior left ventricle; *Ao*, ascending aorta; *Inf*, inferior left ventricle; *IVS*, interventricular septum; *PML*, posterior mitral leaflet; *RV*, right ventricle.

coaptation point, because that is the part that is caught by flow and pushed into the outflow tract. In a previous study, the AML was 34, 30, and 24 mm in patients with obstructed HCM, those with nonobstructed HCM, and those without HCM, respectively.[29] Three-dimensional TEE allows precise measurements, allowing one to identify AML lengths at A2, as well as anywhere along A1:A3 (Figure 6). As defined in this previous study, leaflets >16 mm/m^2 are elongated in HCM and associated with obstruction.

In the recent past, when a long leaflet participated in the pathophysiology of SAM, horizontal plication mattress sutures were placed.[20,25] Patients who were selected to undergo plication had longer AMLs (32 vs 28 mm).[34] A long redundant leaflet can thus be shortened and stiffened, decreasing the predilection for SAM and maintaining intact coaptation with the posterior leaflet.

MR often is present in patients with HCM. Three-dimensional TEE is an excellent modality to evaluate the mitral valve, particularly in identifying segmental mitral valve disease.[35] Careful assessment of the mitral valve and regurgitant jet will determine whether the etiology is secondary to SAM, structural mitral valve abnormalities, nonphysiologic loading conditions, or a combination of the three. MR due to SAM will be abolished or markedly reduced by correction of the outflow tract obstruction and the SAM. Significant intrinsic mitral valve pathology such as prolapse may warrant a concomitant repair. Infrequently, mitral valve replacement is deemed appropriate, usually because of extensive calcification.

Residual Leaflet. Additionally, there may be a residual part of the AML that extends past the point of coaptation, termed a residual leaflet.[14] This portion may become patulous and thinned, does not participate in coaptation, and can protrude into the LVOT (Figure 7, Video 1; available at www.onlinejase.com). For the last four years, we have applied a more nuanced approach to redundant leaflets. Mitral-septal contact can occur in two different ways: first, there may be billowing of the central portion of the leaflet because of excess tissue in the horizontal axis of the leaflet, leading to a "cowl" appearence of the anterior leaflet; or, second, and more commonly, there may be increased tissue in the vertical axis and excessive excursion of the leading edge, the *residual leaflet.* Differentiation of the two during preoperative echocardiography is now considered to be instrumental in planning the surgical approach to the mitral contribution of obstruction. The first type, with horizontal redundancy, lends itself to repair with a horizontal plication, described above. For the second type, patients with very long discrete residual leaflets that extend past the coaptation point of the mitral valve and protrude into the left ventricle, we now perform a direct residual leaflet excision termed "ReLex."[14] In making this decision, the length of the coaptation

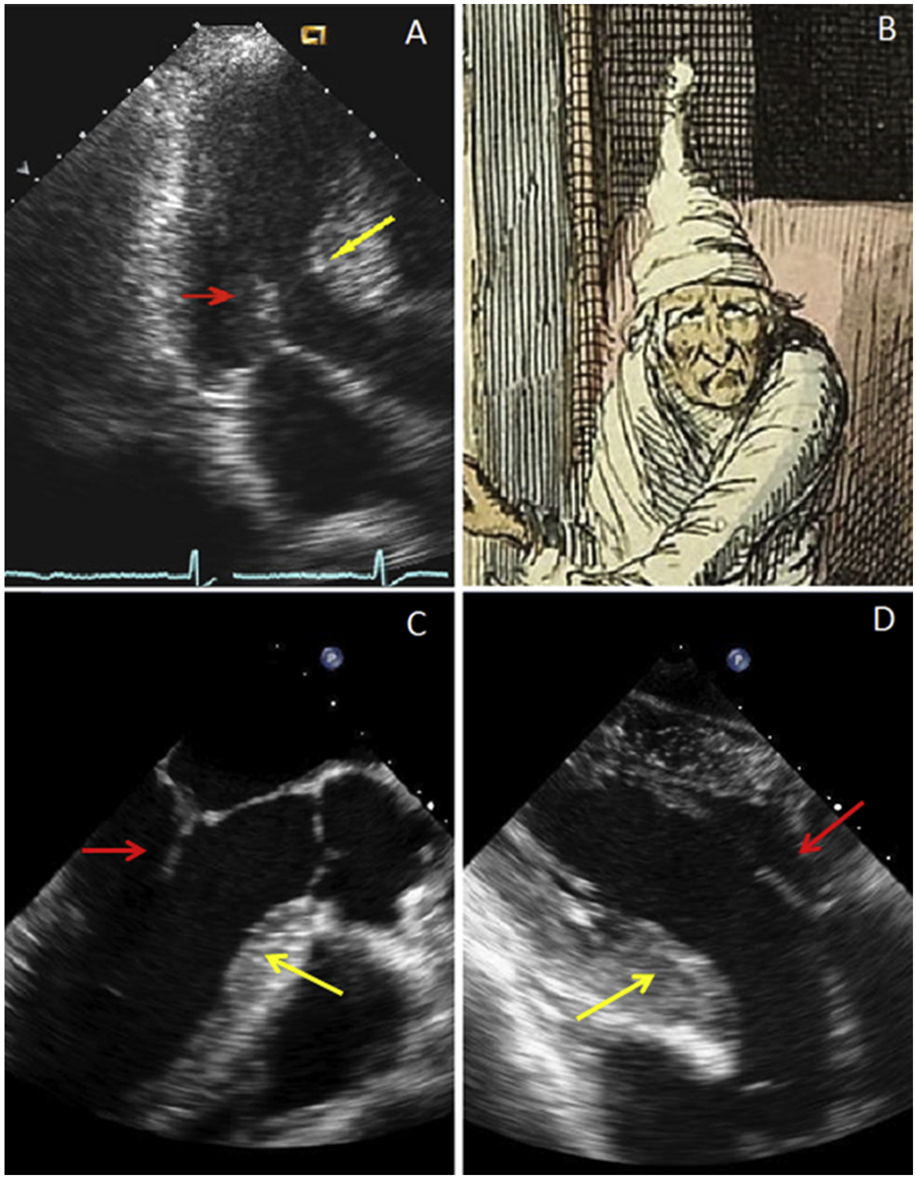

Figure 5 **(A)** Transthoracic echocardiographic apical three-chamber view. The redundant elongated AML (*red arrow)* appears as a "nightcap" mitral valve, protruding into the LVOT. **(B)** An elderly gentleman wears an old-fashioned nightcap, a typical appearance of the elongated leaflets seen in many HCM patients. **(C)** A greatly protruding mitral anterior leaflet on transesophageal echocardiographic midesophageal long-axis view. **(D)** Protruding leaflet on the transgastric long-axis view. The protruding elongated slack valve is subject to early systolic interventricular flow and is swept into the septum (septum shown with a *yellow arrow*).

zone is carefully measured, along with the length of excessive leading edge. In general, ≥1 cm of coaptation length must remain after excision to maintain competence, so only the length of the protruding leaflet in excess of the 1 cm of coaptation zone can be considered safe for excision. At times, a portion of the 1-cm coaptation area protrudes into the LVOT, but that area is not safely available for excision as it is necessary for the maintenance of competence. Additionally, excessive chordal tissue must not be confused with residual leaflet, as this may suggest that more tissue can be considered for resection than in actuality. These measurements are best accomplished during TEE just before surgery.

At surgical inspection, once the myectomy is accomplished, visualization of the mitral valve and supporting apparatus is greatly enhanced. If there is a significant residual leaflet, it invariably is attached to a number of loose, curlicued chords that are very myxomatous in appearance and can be identified within the collapsed heart (Figure 8). These chords are too long to prevent prolapse. Nerve hooks are used to lift the anterior leaflet into the LVOT, and the leading edge of A2 is visualized. The section attached to the redundant chordal tissue, usually anywhere from 2–5 mm, can be safely excised as determined by the preoperative echocardiographic measurements. This is performed only if the chordal attachments to the leaflet margin on either side of the residual leaflet are deemed competent enough to prevent leaflet flail when the redundant tip is excised. Competence results from a combination of maintenance of appropriate supporting chords and an adequate coaptation zone; both are necessary. This approach is attractive because it addresses directly an aspect of the primary pathology, which is the protruding

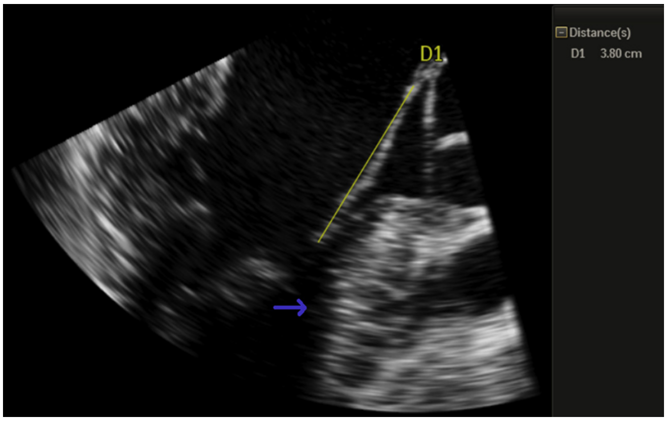

Figure 6 As seen in Figure 1, one can identify A2:P2 (or any plane along the annulus) by shifting the green plane. A precise measurement of AML length can then be made by zooming in on the green plane. The anterior septum *(blue arrow)* is also visualized.

residual leaflet that is caught by the flow and actually comes into apposition with the septum.

Edge-to-edge repair also has a place in the surgical management of obstruction in HCM, most notably in patients with minimal septal thickness.[36] We usually restrict this technique to older patients presenting for surgical management, as we are uncertain as to the long-term effects of this repair over a period of decades. In situations in which the body of the leaflet does not lend itself to plication or the leading edge to excision, edge-to-edge repair can be useful, either through the aortotomy or a counter incision in the left atrium.[36,37] Although we have done both, we prefer the left atrial approach, which allows the additional placement of an oversized mitral ring to stabilize the edge-to-edge suture. The need for the supporting ring remains controversial. Quite rarely, when overzealous residual leaflet excision has occurred with subsequent central mitral insufficiency, edge-to-edge repair has been an effective solution.

Subvalvular Apparatus

A thorough analysis of the subvalvular structures—the papillary muscles and chordae tendineae—is vital for determining the surgical plan.

Papillary Muscles. Patients with HCM may have abnormalities of the papillary muscles that significantly contribute to obstruction.[38] The two most common features noted are (1) anterior and basal displacement of the anterolateral papillary muscle and (2) abnormal attachments of the anterolateral papillary muscle to the anterolateral wall near the A1 scallop (Figure 9).[14] Both of these abnormalities predispose to SAM by dint of their anterior displacement of the mitral valve, by increasing the overlap between outflow and the mitral valve. Flow strikes the anteriorly displaced mitral leaflets from behind and sweeps them toward the septum.[29,30] In addition, anterior displacement of the papillary muscles decreases the posterior restraint on the leaflets predisposing to the slack that allows SAM.[39,40]

Release of the abnormal connections to the ventricular wall and thinning of the hypertrophied papillary muscle result in a posterior drop of the mitral leaflets away from the LVOT. Release is thus a key component of the operation.[16,19,25-27,34]

A less common but very important pathology to recognize is direct insertion of an anomalous head of the anterolateral papillary muscle into the middle of the anterior leaflet without intervening chordae.[14,41] These anomalous papillary muscles may cause obstruction in two ways. First, the anomalous muscle may impact the septum with each systole and cause obstruction below the level of the mitral leaflets. This is thus a form of midventricular obstruction. Second, they may bring the mitral leaflets anteriorly and into the ejection flow stream, causing SAM. The successful surgical approach to these anomalous papillary muscles requires an extended myectomy down to the level of the origin of the papillary muscles. When there is adequate support of the leaflet margin by normal chordae, these papillary muscles are completely resected. If there is no alternative support of the leaflet margin, the anomalous papillary muscles can be thinned along their longitudinal axis.[14,34] Anomalous papillary muscles (and chordae) that contribute to SAM and obstruct primarily may be identified on TTE.[14] However, they are more precisely identified on 3D TEE, particularly when visualizing the mitral valve en face, from the left ventricle, looking up at the outflow tract. Three-dimensional TEE can locate both direct insertion of papillary muscles into the mitral valve leaflets as well as accessory papillary muscle heads (Figure 10, Videos 2 and 3; available at www.onlinejase.com).

Papillary muscle thickening at the mid-LV level may contribute to mid-LV obstruction.[42] Normal papillary muscle diameter in the short axis is 0.7 ± 0.2 cm, with hypertrophy defined as >1.1 cm.[38,40,43] In mid-LV obstruction, not only are the mid-LV segments thickened, but the LV cavity is small, and they contract around the thickened papillary muscles, with resultant complete emptying of the mid left ventricle.[44] Importantly, patients with mid-LV obstruction and an akinetic apical chamber may not have elevated interventricular Doppler velocities, even though they have profound obstruction.[42] This is because there may be a complete cessation of ejection flow here, because of complete obstruction at the mid left ventricle. This is manifest as a midsystolic signal void on Doppler in the mid left ventricle (Figure 11). The complete cessation of flow is similar to the midsystolic drop seen in the "lobster claw abnormality" of the pulsed-wave Doppler signal in LVOT obstruction, except that the nadir of the drop in velocities in mid-LV obstruction is more severe, and flow often completely stops.

Chordae Tendineae. Chordal attachments to the mitral valve maintain coaptation between the two leaflets. However, fibrotic, retracted secondary chordae that insert into the middle of the AML can elevate the AML and predispose to SAM, prevent coaptation, and result in significant MR. Three-dimensional echocardiography is instrumental in determining the exact location of these chords.[45] Visualizing the mitral valve from the LV side will help differentiate the abnormal connections that tether the valve and others that are important in maintaining mitral valve integrity. Surgical release of the leaflet from these secondary chords will help decrease SAM, improve coaptation, and reduce MR.[46] The usual 3D surgeon's-eye view, looking down on the mitral orifice from the left atrium, is not as helpful in obstructive HCM, except to visualize the size of the MR regurgitant orifice; however, the view from above the mitral valve rarely contributes to an understanding of SAM pathophysiology.

By reconstructing a 3D zoom (or full-volume) multigated set, we can crop the data set in different planes to visualize the valve and chordal attachments. Depending on where these chordal attachments are, and where the valve is being tented and tethered, the goal is to release them to not only improve coaptation but to also relieve obstruction set up by valve prepositioning into the LVOT.

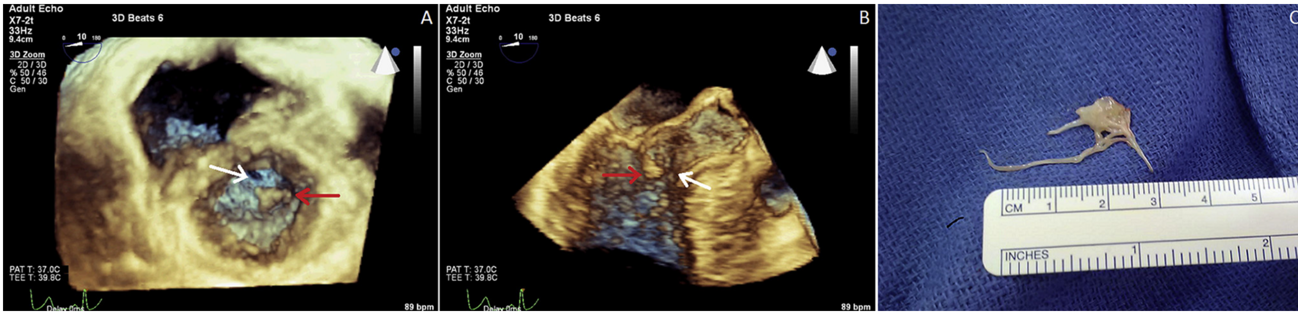

Figure 7 Short-axis 3D view looking up from the left ventricle toward the mitral valve and LVOT. The residual leaflet *(red arrow)* can be clearly visualized in this view protruding into the LVOT *(white arrow)*. Three-dimensional TEE locates it on the lateral side of A2 **(A)**. By performing a plane crop of the residual leaflet along A2:P2, we can visualize it in the midesophageal long-axis view as well **(B)**. With care, it may be safely excised to alleviate obstruction, as shown in **(C)**.

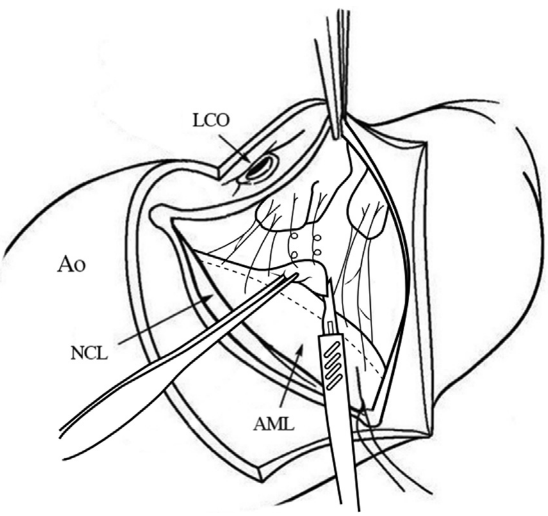

Figure 8 Residual leaflet excision (ReLex). Surgical excision of the residual leaflet is added to myectomy when the anterior leaflet is very elongated and contributes to SAM and when the residual portion does not contribute to coaptation. Adequate chordal support on either side of the residual leaflet is essential before contemplating this excision. Invariably, curlicued chordae attach to the free margin of the residual leaflet and aid in its identification. The *dashed line* indicates the free margin of the posterior leaflet, under the anterior leaflet. *Ao*, Ascending aorta; *LCO*, left coronary ostium; *NCL*, noncoronary leaflet.

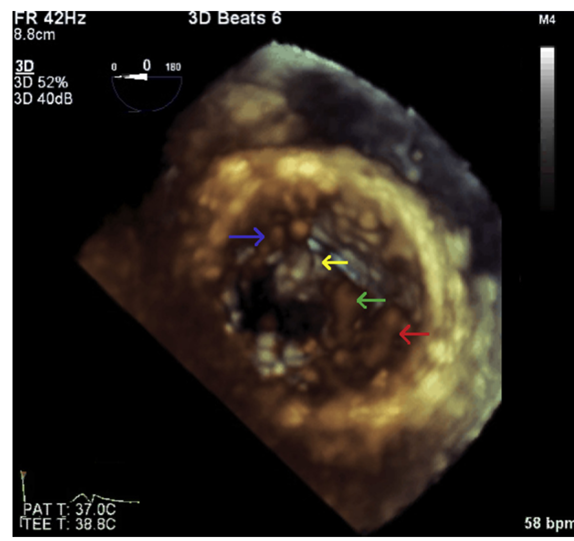

Figure 9 One can often visualize the papillary muscles from the left ventricle looking up at the mitral valve. Patients with HCM may, in addition to anterolateral *(red arrow)* and posteromedial *(blue arrow)* papillary muscles, have accessory papillary muscle heads *(green arrow)*, which may contribute to LVOT obstruction. Release of abnormal attachments and thinning of the papillary muscles to position the mitral valve *(yellow arrow)* more posteriorly is a critical component of combined myectomy-mitral operations.[16,19,25-27,34]

There are a number of ways to assess the valve and subvalvular apparatus using the 3D volume set. Among the most informative, the anterior leaflet can be visualized from the LV side, looking up at the outflow tract. The anomalous secondary chord appears as an abnormal nubbin (Figures 12 and 13).

Alternatively, we can visualize the mitral valve from the side in a midesophageal long-axis view and crop in and out of plane sequentially from A1:P1 to A3:P3 until the tented area is no longer seen. Once the chordal attachment or tethering is removed, we know the previous crop must have the culprit chordal attachment. Bringing this frame back into view and reorienting the data set to look at it from the LV side shows where on the anterior leaflet the chord inserts.

Release and resection of abnormal thickened chordae and anomalous papillary muscles and mitral shortening are particularly important when the interventricular septum is not very thickened (≤18 mm). In these cases, abnormalities of the mitral valve are necessarily more important as causing obstruction than the modest septal thickening, and thus mitral repair assumes a greater importance, because the myectomy will be limited to only a few millimeters.[14,46] In the past, mitral valve replacement was recommended for these patients. Now, with pre–cardiopulmonary bypass echocardiography, the essential abnormalities of the mitral valve can be identified ahead of time, and with judicious surgical technique, the native mitral valve can be saved. This is always preferred because of the known long-term adverse consequences of mitral valve replacement.

LVOT Obstruction

Patients undergoing surgical intervention for HCM have LVOT obstruction either at rest or with provocation. TEE can help elucidate factors contributing to SAM and LVOT obstruction, which include the

Figure 10 Obstructing papillary muscle and chordal resection; surgical specimen. In this patient, preoperative echocardiography demonstrated an anterior anomalous papillary muscle head and thickened chordae. The papillary muscle impacted the septum and obstructed below the mitral valve tips, and the chordae tented the mitral valve anteriorly into the outflow tract, predisposing to SAM. With care, the anomalous papillary muscle was resected, and the chordae were disconnected from the anterior leaflet.

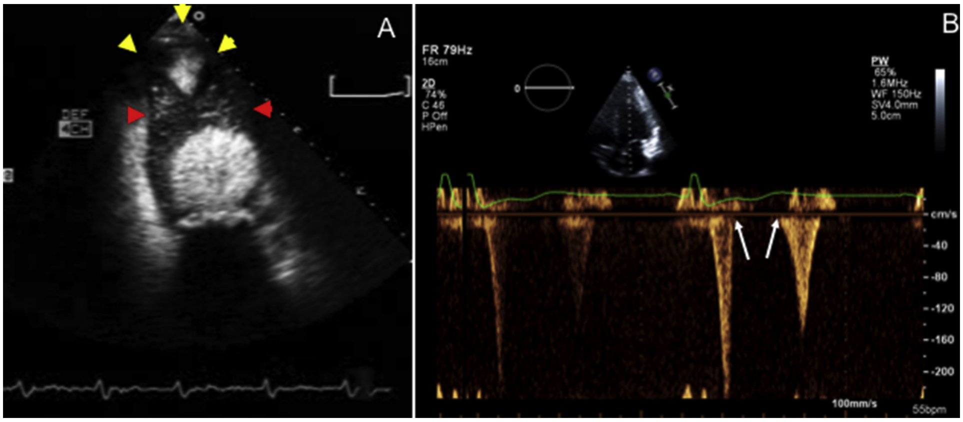

Figure 11 TTE in severe mid-LV obstruction and an apical akinetic aneurysm. **(A)** A very early systolic apical four-chamber view with contrast in the left ventricle. *Red arrowheads* show complete systolic emptying of the mid left ventricle with severe obstruction at that level, and *yellow arrowheads* show the apical akinetic aneurysm. **(B)** A pulsed-wave Doppler tracing at the level of the obstruction with a clear signal void *(white arrows)* between the initial systolic ejection flow, which is prematurely truncated, and paradoxical diastolic flow out of the apical aneurysm during diastole. Note that though there is very severe obstruction, the peak systolic velocities are modestly elevated at 2.8 m/sec. This is because there is no flow in midsystole because of the complete mid-LV obstruction. Because of the low velocities, such patients may be clinically misdiagnosed as nonobstructed if this pathophysiology is not appreciated.[42]

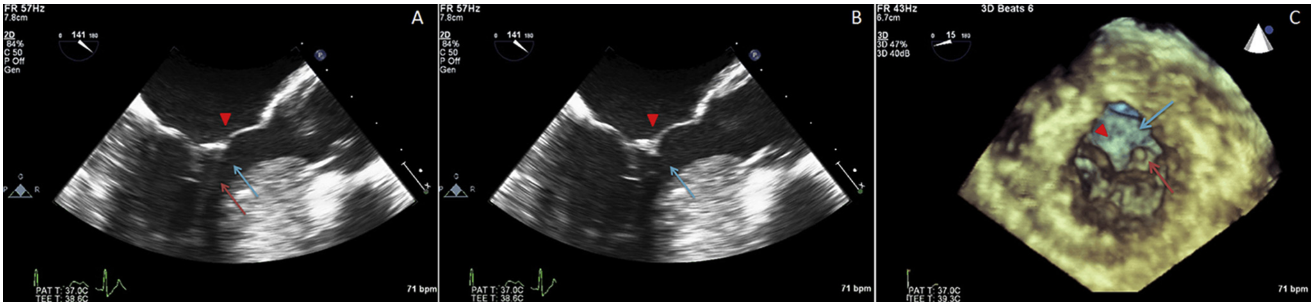

Figure 12 Patients with HCM may have retracted fibrotic chordae, which tent up the AML and predispose to SAM by positioning it into the ejection stream **(A)**. As systole progresses, the anterior leaflet *(red arrowhead)* is pushed into the LVOT *(blue arrow)*, narrowing it further **(B)**. Three-dimensional echocardiography can precisely determine the location of chordal attachment on the medial side of A1 and lateral side of A2 *(red arrow)* and guide the release portion of the myectomy-mitral operation **(C)**.

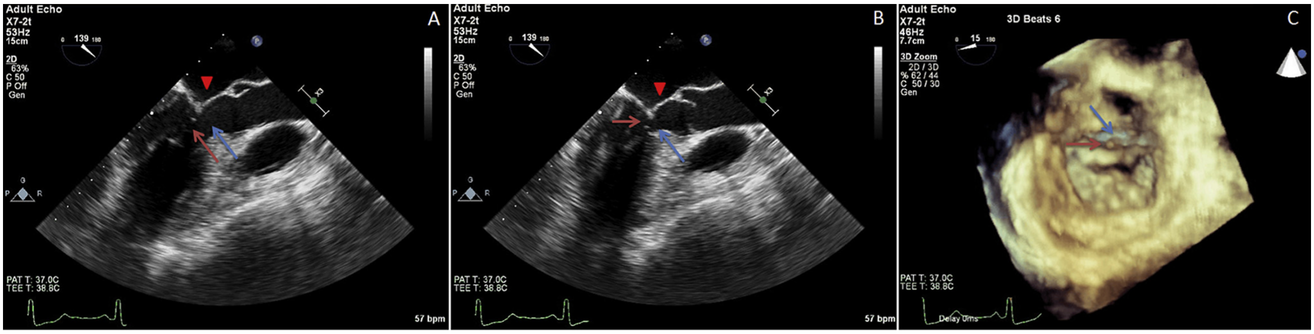

Figure 13 A different patient with retracted fibrotic chordae tendineae *(red arrow)* **(A)**, shifting the AML *(red arrowhead)* into the LVOT *(blue arrow)* **(B)**. The chordal attachment is seen on the medial side of A2 **(C)**, whereas in the previous figure chordal attachments were prominent on the medial side of A1 and lateral side of A2.

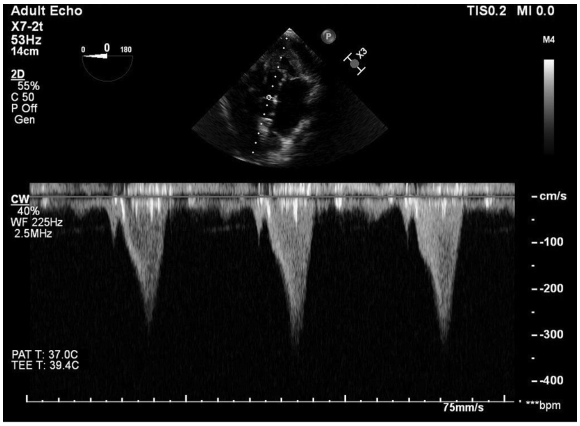

Figure 14 Continuous-wave Doppler through the LVOT in the deep transgastric view may show a classic "dagger-shaped" profile in patients with HCM, confirming dynamic obstruction. The Doppler tracing is concave to the left because the orifice narrows over time as the mitral valve is pushed further into the septum by the pressure gradient.

aforementioned septal hypertrophy, elongated leaflets, abnormal papillary muscles, and tethering of the valve by thickened shortened chordae. Obstruction may or may not be present at the time of surgery depending on loading conditions and whether obstruction was at rest or latent preoperatively.[47] Occasionally, if there is ambiguity about the level of obstruction or its cause before cardiopulmonary bypass, dobutamine can be administered transiently.

Color flow Doppler can confirm areas of flow convergence as blood travels toward the area of narrowing and turbulence as it is ejected past the narrowing. The extent of the obstruction is quantified with continuous-wave Doppler through the LVOT best obtained from the deep transgastric view to allow parallel orientation of the Doppler beam (Figure 14). Patients with HCM may have both mid-LV obstruction from complete systolic emptying at that level around thickened papillary muscles and also LVOT obstruction due to SAM.[42,44] Also, caution must be exercised in patients who have other pathology (e.g., aortic stenosis, subvalvular or supravalvular stenosis) to tease out precisely where the high velocity flow is occurring. Pulsed Doppler and color flow may assist in these efforts.

The transesophageal echocardiographic imaging protocol we perform to assess patients with obstructive HCM is summarized in Figure 15.

POSTOPERATIVE EXAMINATION

After surgical intervention, the patient is weaned off cardiopulmonary bypass, and repeat TEE is performed before decannulation.

Interventricular Septum

Assessment of residual septal thickness is essential in postmyectomy patients. The plane of resection can be determined in a two-dimensional view by obtaining the midesophageal long axis and rotating the probe clockwise and counterclockwise to visualize the operative thinning of the interventricular septum. Septal thickness can be compared with the preoperative measurement, ensuring that an adequate but not excessive myectomy was performed. Additional planes, including the coronal plane, may be useful to delineate the location of any residual septal tissue that may require removal.

The infrequent occurrence of a ventricular septal defect should be detected immediately after circulation is reestablished by careful inspection of the whole septum with color Doppler, with the cannulae in situ, as this can be corrected surgically either immediately, or later as a planned second procedure. Assessment is performed in both midesophageal long-axis and transgastric short-axis views. Transection of a septal perforator can cause a thin diastolic jet into the left ventricle that should not be confused with a ventricular septal defect (Figures 16-19, Videos 4-7; available at www.onlinejase.com). Such transection is a benign finding without sequelae.

Mitral Valve

The mitral valve is assessed to ensure resolution of SAM, reduction of MR, and improvement of coaptation. Anterior leaflet length can be quantified and compared with preoperative values, showing a decrease in length when residual leaflet excision or horizontal plication has been performed. Halpern *et al.*[34] showed with TTE that patients who underwent plication had decreased AML length by 16% (from 32 to 27 mm), residual leaflet length by 33%, and protrusion height by 24%. TEE before and after successful myectomy and mitral shortening shows elimination of the SAM, resolved LVOT obstruction, and excellent coaptation of the mitral valve leaflets (Figure 20, Videos 8 and 9; available at www.onlinejase.com).

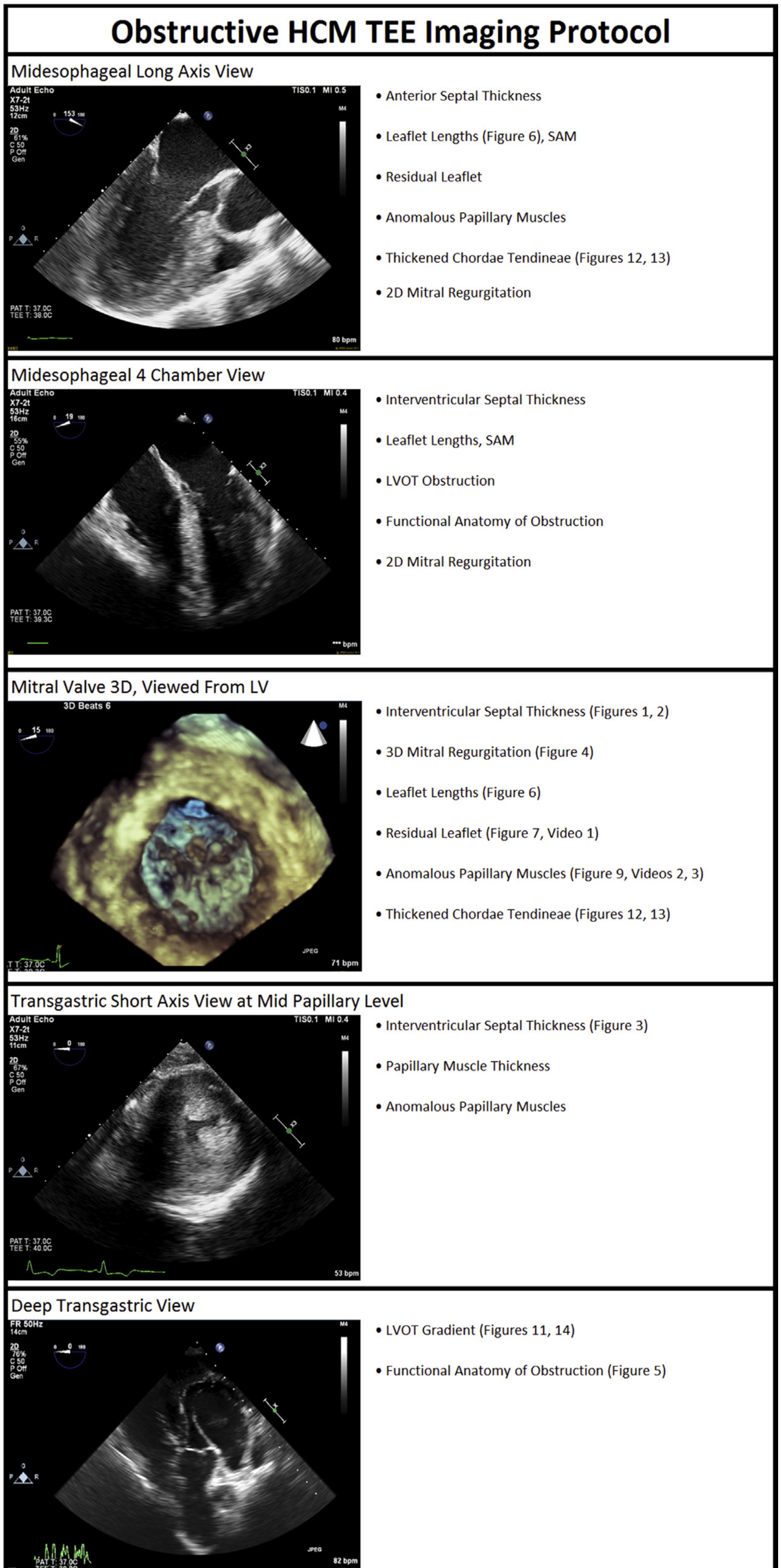

Figure 15 We recommend a systematic assessment to define the anatomic pathology and pathophysiology of patients with HCM. Listed above are the order of TEE views we use and pertinent features to evaluate in each of the views. *2D*, Two-dimensional.

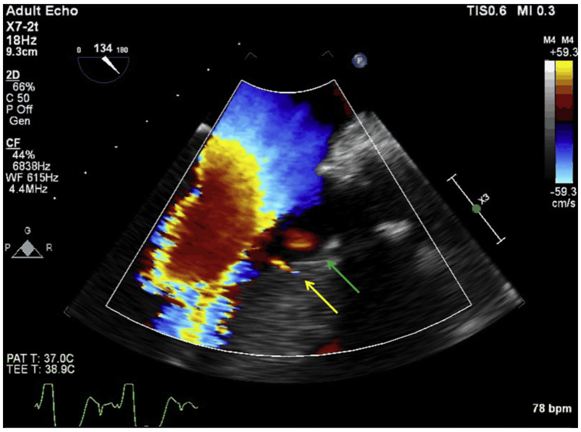

Figure 16 TEE of transected septal perforator. Mid-esophageal long axis view. The patient has two diastolic jets after undergoing septal myectomy and aortic valve replacement: a transected septal perforator jet *(yellow arrow)* and a paravalvular leak *(green arrow)*. The septal perforator jet flows only in diastole and does not extend into the right ventricle. See Video 4.

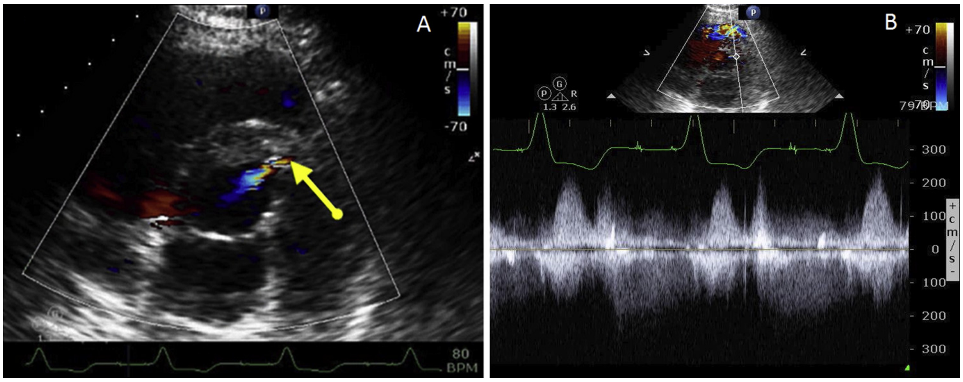

Figure 17 The septal perforator jet (*yellow arrow*) is present on the postoperative TTE, short-axis view, level of the outflow tract. **(A)** and can be recognized by its diastolic flow velocities, similar to coronary flow velocities, that have peak velocity of about 1 m/sec **(B)**. No intervention is needed, and the diastolic flow usually stops by 6 months after surgery. The aortic paravalvular jet noted in Figure 16 has resolved.

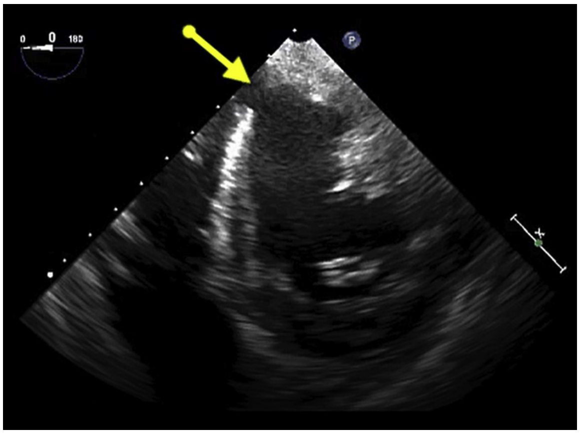

Figure 18 A ventricular septal defect (VSD; *yellow arrow)* is visualized in the midinferoseptal myocardium after surgical intervention. It is important to inspect the entire septum after the pump run, before decannulation. The VSD was closed during a second operation with patches on both the LV and right ventricular side of the septum, and the patient had a New York Heart Association functional class I outcome.

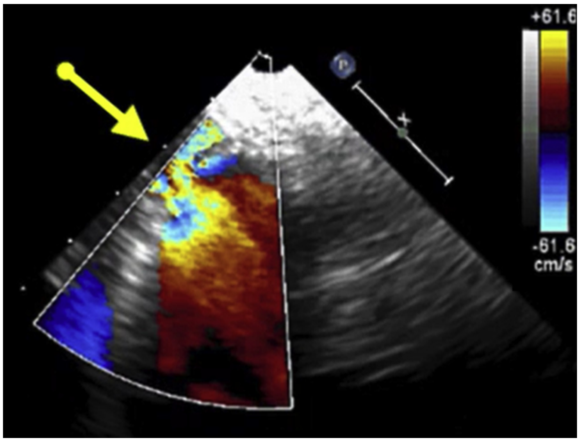

Figure 19 Prominent color flow *(yellow arrow)* can be visualized with left-to-right shunting through the ventricular septal defect.

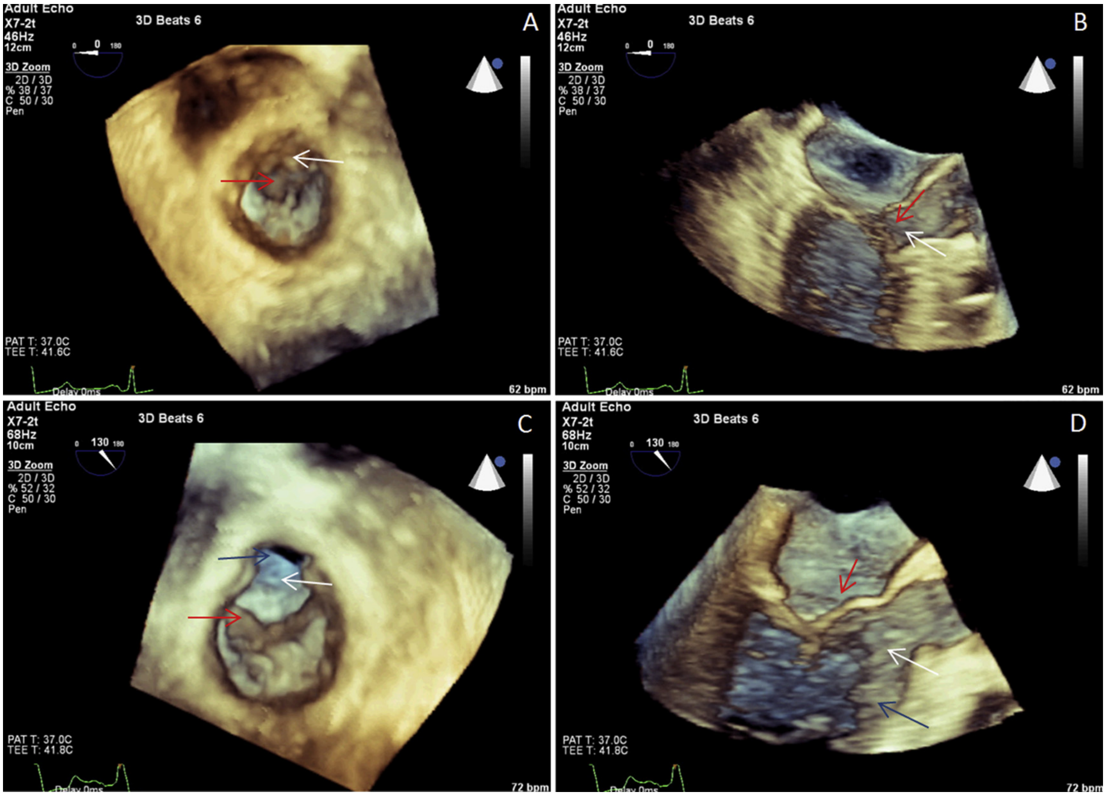

Figure 20 This patient underwent a successful combined myectomy-mitral shortening operation with an extended myectomy, residual leaflet excision, and chordal release. Preoperatively, the patient had significant obstruction of the LVOT *(white arrows)* exacerbated by SAM of the AML *(red arrows)* **(A, B)**. After the myectomy-mitral shortening operation **(C, D)**, the obstruction was alleviated and MR secondary to SAM resolved. The deep extended myectomy is well shown *(blue arrows)*.

Subvalvular Apparatus

The subvalvular apparatus is evaluated to confirm a successful release of the mitral valve leaflets. Resection of the papillary muscles may be visualized, particularly in the transgastric midpapillary short-axis view. In the midesophageal long-axis view, specific chordae tendineae and papillary muscle heads, which formerly retracted the anterior leaflet, should no longer be seen (Figure 10). Release of papillary muscles results in coaptation of the mitral valve more posteriorly in the LV cavity.[34]

Removal of chordae integral to mitral valve coaptation or excessive papillary muscle release could result in new or worsened MR because of prolapse or flail leaflets, which should always be excluded during postmyectomy TEE.

LVOT Obstruction

Peak gradient through the LVOT is again assessed and compared with preoperative values. In the presence of a significant gradient, one must again look for location and etiology to determine if further intervention is warranted.[32,48]

After initial postbypass assessment, dobutamine is initiated at 10 μg/kg/min, in a dose adequate to increase heart rate, and a repeat assessment is performed, paying close attention to the presence of SAM, change in MR, and any increase in peak gradient. This is necessary to ensure that the patient no longer has latent obstruction and can withstand exercise after discharge. If residual obstruction is present at rest or after dobutamine with LVOT velocities >3 m/sec or moderate or severe MR, then reinstituting cardiopulmonary bypass and additional surgical repair is warranted.[26,49] In our experience, this occurs approximately 2% of the time.

CONCLUSIONS

Obstructive HCM has increasingly become a treatable condition with good long-term prognosis. Surgery is indicated in patients with symptoms refractory to medications or those who are unable to tolerate side effects. This guide serves as a way to systematically evaluate these patients using two-dimensional and 3D TEE to determine the best surgical approach. We describe HCM surgery as applied in combined myectomy and mitral operations at our institution because we believe it is appropriate in carefully selected patients to correct all of HCM pathophysiology at the initial operation.[14-21,25-27,34]

Transesophageal echocardiographic assessment of the LV septum, mitral valve, and subvalvular apparatus is vital to determine the pathophysiology of LV obstruction and MR. Components of the operation are selectively implemented on the basis of preoperative TEE. Postoperative TEE is critical to evaluate for significant residual LV obstruction or MR and dictate whether further intervention is required. TEE is an invaluable tool in the operative management of patients with HCM.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.echo.2017.11.016.

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