SPECIAL ARTICLE

Three-Dimensional Imaging and Dynamic Modeling of Systolic Anterior Motion of the Mitral Valve

Alan Vainrib, MD, Daniele Massera, MD, MSc, Mark V. Sherrid, MD, Daniel G. Swistel, MD, Daniel Bamira, MD, Homam Ibrahim, MD, Cezar Staniloae, MD, Mathew R. Williams, MD, and Muhamed Saric, MD, PhD, *New York, New York*

Left ventricular outflow tract (LVOT) obstruction in hypertrophic cardiomyopathy (HCM) is often caused by systolic anterior motion (SAM) of the mitral valve caused by the interplay between increased left ventricular (LV) wall thickness and an abnormal mitral valve anatomy and geometry. Three-dimensional (3D) echocardiographic imaging of the mitral valve has revolutionized the practice of cardiology, paving the way for new methods to see and treat valvular heart disease. Here we present the novel and incremental value of 3D transesophageal echocardiography (TEE) of SAM visualization. This review first provides step-by-step instructions on acquiring and optimizing 3D TEE imaging of SAM. It then describes the unique and novel findings using standard 3D TEE rendering as well as dynamic mitral valve modeling of SAM from 3D data sets, which can provide a more detailed visualization of SAM features. The findings include double-orifice LVOT caused by the residual leaflet, the dolphin smile phenomenon, and delineation of SAM width. Finally, the review discusses the essential role of 3D TEE imaging for preprocedural assessment and intraprocedural guidance of surgical and novel percutaneous treatments of SAM. (J Am Soc Echocardiogr 2021;34:89-96.)

Keywords: Mitral valve, 3D echocardiography, Hypertrophic cardiomyopathy, Systolic anterior motion (SAM)

Anatomic alterations of the mitral valve (MV) apparatus are part of the pathologic process of hypertrophic cardiomyopathy (HCM); in recent years their important contribution to the obstruction process has been identified.[1] The most salient and common features are (1) elongated MV leaflets, (2) anterior and basal displaced papillary muscles, particularly a small anterior head of the anterolateral papillary muscle, and (3) shortened thickened chordae or a papillary muscle head that inserts into the midanterior MV leaflet. All these abnormalities contribute to mitral leaflet slack (decreased restraint to anterior motion) and leaflet protrusion into the left ventricular (LV) outflow tract (LVOT) where they then are predisposed to be swept into the septum.[2] A more profound abnormality is an large anomalous papillary muscle that inserts directly into the middle of the anterior mitral leaflet.[3] This can obstruct directly because of apposition of this papillary muscle and the septum, apical of the coaptation point but also because it can tent the leaflet anteriorly and predispose to systolic anterior motion (SAM). This pathology is important to recognize because it often requires extended myectomy to the papillary muscle level often accompanied by resection (if feasible) or thinning of the anomalous muscle (if not).[1] This review focuses on three-dimensional (3D) imaging of abnormalities of the MV leaflets and unique modeling of this pathology. Abnormalities of the papillary muscles, also uniquely demonstrated on 3D echo, are described in detail elsewhere.[4]

The appearance of elongated MV leaflets in systole has been described as a "nightcap" MV, with the protrusion height measuring approximately 26 mm above the plane of the mitral annulus compared with approximately 13 mm in normal hearts.[5] This protrusion into the LVOT places the leaflets in the ejection flow where drag catches them and pushes them from behind and subsequently into the interventricular septum. When the posterior mitral leaflet is not as long or as mobile as the anterior mitral leaflet, a mitral coaptation gap is created that leads to significant mitral regurgitation.[4,5]

While many aspects of the mitral-septal contact and LVOT obstruction have been characterized by M mode and two-dimensional (2D) echocardiography, there is a paucity of data on direct 3D echocardiographic visualization of this process (Table 1). In this paper, we describe methods to acquire and optimize 3D transesophageal echocardiography (TEE) data sets for SAM visualization, 3D SAM modeling and quantification, and 3D SAM width measurement in preparation for surgical or percutaneous SAM repair.[4]

3D TEE IMAGE ACQUISITION OF SAM

Three-dimensional echocardiographic imaging of SAM is best obtained using 3D TEE rather than transthoracic echocardiography due to the posterior position of the MV and subvalvular apparatus adjacent to the esophagus.

From the Leon H. Charney Division of Cardiology, Echocardiography Laboratory, Hypertrophic Cardiomyopathy Program (A.V., D.M., M.V.S., D.B., H.I., C.S., M.R.W., M.S.); and Department of Cardiothoracic Surgery (D.G.S., M.R.W.), New York University Langone Health, New York, New York.

A.V. is a consultant for Micro Interventional Devices. M.S. is a member of the Philips, Boston Scientific, and Medtronic speakers' bureaus and a member of Siemens advisory board.

Luigi P. Badano, MD, PhD, FASE, served as guest editor for this report.

Reprint requests: Muhamed Saric, MD, PhD, Director, Noninvasive Cardiology, Professor of Medicine, Noninvasive Cardiology, New York University, 560 First Avenue, New York, NY 10016 (E-mail: *muhamed.saric@nyumc.org*).

0894-7317/$36.00

https://doi.org/10.1016/j.echo.2020.08.019

Abbreviations

2D = Two-dimensional

3D = Three-dimensional

CT = Computed tomography

CW = Continuous wave

DO = Double orifice

HCM = Hypertrophic cardiomyopathy

LV = Left ventricular

LVOT = Left ventricular outflow tract

MV = Mitral valve

ROI = Region of interest

SAM = Systolic anterior motion

SAV = Short-axis view

TEE = Transesophageal echocardiography

Step 1: Region of Interest (ROI) Selection

Three-dimensional TEE acquisition of SAM begins with 3D zoom ROI placement on the MV, as previously described in comprehensive TEE guidelines published by the American Society of Echocardiography.[10] Due to elongation of the MV leaflets, it is important to place the 3D ROI deeper within the LV than typically performed for 3D imaging of the en face view of the MV. To ensure adequate frame rate, the 3D ROI sector should be as narrow as possible while still including the entire MV. The 3D sector typically will not include papillary muscles as these would block the view of the MV within the LVOT.

Step 2: Azimuth Plane Adjustment

To image the LVOT adequately, the 3D ROI must include a wider azimuth plane to ensure the entire LVOT is enclosed (Figure 1A).

Step 3: 3D Image Acquisition

Once an adequate ROI is selected, 3D acquisition is initiated. Single-beat imaging is preferred to avoid the stitching artifact typically associated with multibeat reconstruction.

Step 4: 3D TEE Image Display

The initial 3D TEE image is subsequently tilted upward to reveal its LV side. Alternatively, one can use an instantaneous directional 3D plane cropping tool available on many 3D TEE ultrasound systems (such as QuickVue from Philips Ultrasound, Bothell, WA; D'art from Siemens Healthineers, Mountainview, CA; and 2-Click Crop from GE Healthcare, Chicago, IL).

Step 5: Z-Plane Rotation

Subsequently, the image is then rotated in the *Z* axis to position the aortic valve on top. (Figure 1B, Supplemental Video 1, available at www.onlinejase.com). This "looking up" at the valve view offers unique perspective on the residual leaflet that is completely opaque to the typical "surgeon's view" looking down on the valve. The structures and pathology described here cannot be seen from the left atrial looking-down view.

Step 6: Dynamic Range and Gain Adjustment

As the anterior mitral leaflet is a thin structure, controlling gain and dynamic range is critical to ensure that SAM is well visualized by avoiding dropouts. Systolic anterior motion and "double orifice" (DO) LVOT (DO-LVOT) can be well demonstrated, and this 3D TEE view mimics true anatomic views of the MV from its LV aspect (Figure 2, Supplemental Video 2, available at www.onlinejase.com).

Step 7: Quantitative Analysis

Such analysis may then be performed directly on the 3D data set to measure a variety of LVOT and MV parameters including the width of SAM within the LVOT (Figure 3, Supplemental Video 3, available at www.onlinejase.com).

Novel 3D Imaging of SAM: DO-LVOT

Prior to 3D echocardiographic imaging, echocardiographers relied on 2D short-axis views (SAV) and long-axis views of the MV to evaluate the extent of SAM. However, obtaining accurate on-axis SAV images of SAM is difficult at best and DO-LVOT is not typically apparent on SAV. Three-dimensional echocardiography has revolutionized imaging of the MV by allowing real-time direct visualization of the MV leaflets, subvalvular apparatus, and LVOT to illuminate the dynamic interplay between these structures in HCM.[11]

The 3D en face view of the LVOT from the LV perspective provides a unique vantage point to reveal the extent of systolic mitral-septal contact within the LVOT along the medial-lateral plane. The leaflet that protrudes past the coaptation plane is the residual leaflet. The residual leaflet is a unique feature of obstructive HCM because it is not bounded by the LV-left atrial pressure difference and thus is not held in place by that pressure difference. Instead the residual leaflet has LV flow on both sides and thus readily moves with the intraventricular flow. Systolic anterior motion often begins even before ejection starts, because the thin flexible residual leaflet moves even in low-velocity flow.[5,12] Typically, the presence of the anterior mitral leaflet or residual leaflet within the LVOT making contact with the interventricular septum (mitral-septal contact) during systole produces a characteristic DO appearance of the LVOT (Figure 4, Supplemental Video 4, available at www.onlinejase.com).

This 3D TEE en face view of LVOT can be obtained using standard commercially available 3D TEE ultrasound systems in real time. In our

Table 1 Historical perspective

Year	Authors	Description	Reference
1960	Braunwald *et al.*	Frist description of HCM (then referred as idiopathic hypertrophic subaortic stenosis)	6
1961	Björk *et al.*	First description of MV apparatus abnormalities in HCM	7
1967	Shah *et al.*	First description of SAM on M-mode echocardiography	8
2000	Sherrid *et al.*	Detailed description of flow drag effect vs Venturi effect explanation of SAM mechanism	9
2016	Sherrid *et al.*	Detailed description of MV abnormalities in HCM	1

HIGHLIGHTS

- 3D dynamic modeling can provide incremental value for visualization of SAM.
- 3D SAM imaging novel findings: double-orifice LVOT, dolphin smile, and SAM width.
- 3D TEE imaging is critical for surgical and percutaneous SAM therapies.

laboratory, we have also been able to reproduce the equivalent en face LVOT view using electrocardiogram-gated computed tomography (CT; Figure 5, Supplemental Video 5, available at www.onlinejase.com). However, CT-based imaging is not commonly done in HCM and requires specialized postprocessing software (such as 3Mensio, Pie Medical Imaging, Maastricht, Netherlands).

3D VISUALIZATION OF SAM WIDTH

Three-dimensional TEE imaging easily demonstrates that there is a variation in the extent or width for which the MV protrudes into the LVOT in systole, which is not easily seen by other imaging techniques. In some patients, only a narrow portion of the MV obstructs the LVOT; in others the width is greater. Narrow SAM occupies less space in the outflow tract and for any given LVOT area would be expected to cause less obstruction and lower instantaneous LVOT gradients. This provides a unique window into the pathophysiology of SAM and is important in planning for surgical and percutaneous SAM repair as described below.

3D Modeling of SAM: Dolphin Smile Phenomenon

In addition to enhanced SAM visualization and analysis using 3D echocardiography, recent advances in digital MV leaflet tracking along with dynamic 3D machine learning–driven MV modeling algorithms provide a novel method for qualitative and quantitative SAM analysis. Modeling algorithms may use either single-frame (static model) or frame-by-frame (dynamic model) format. By simplifying the 3D anatomy these models emphasize the key anatomic and functional features of a cardiac structure. When applied to 3D MV data of SAM, one can generate never before seen snapshots of the MV in HCM as well as key quantitative LVOT and MV parameters.

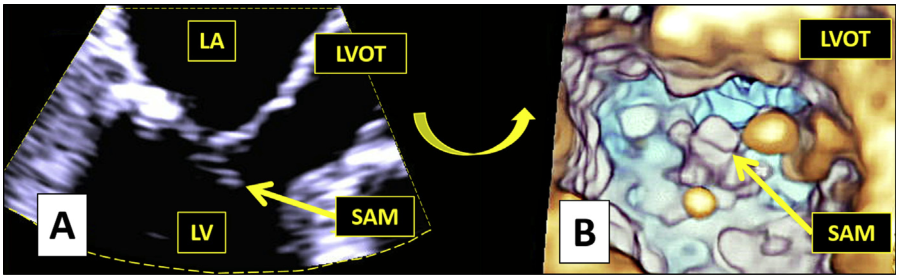

Figure 1 3D SAM ROI acquisition. **(A)** 2D TEE in midesophageal long-axis view. The ROI for 3D zoom is represented by the *dotted yellow line*. Selecting the proper ROI is the critical first step for creating an optimal 3D image of SAM (*yellow arrow*). Note adequate depth to capture SAM and adequate azimuth width to capture full LVOT. **(B)** 3D of SAM and LVOT from LV perspective. Note the 3D image has been tilted upward to better reveal SAM and LVOT. *Ao*, Aorta, *LA*, left atrium; *LV*, left ventricle.

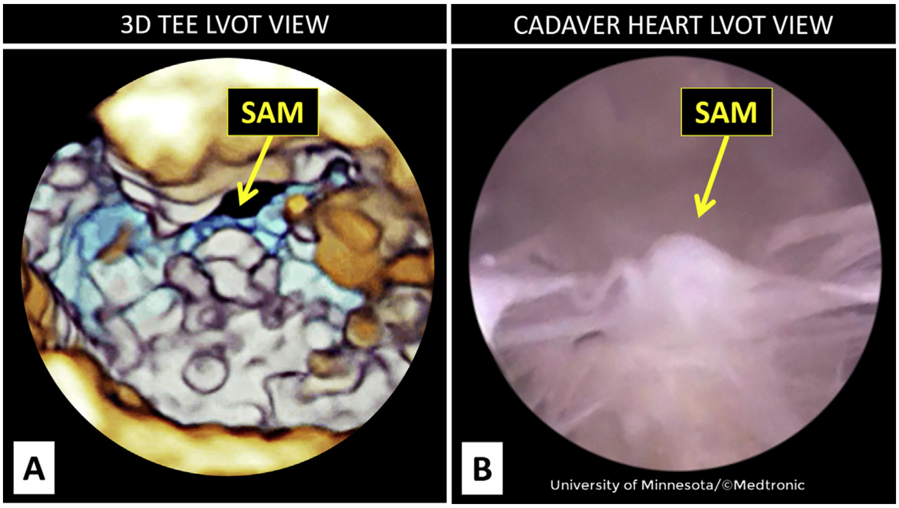

Figure 2 SAM on cadaver heart and 3D TEE. **(A)** 3D TEE demonstrating LVOT view with SAM. **(B)** Cadaver heart from LV perspective demonstrating SAM. Obtained with permission from Visible Heart Lab website/University of Minnesota/Medtronic.

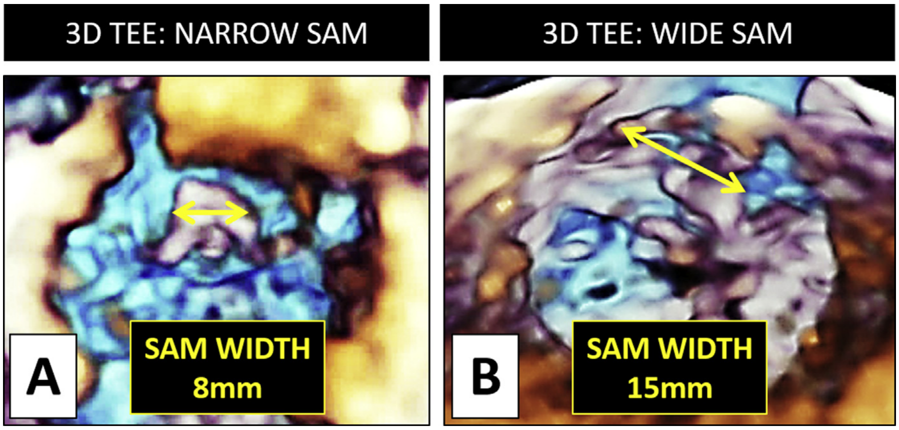

Figure 3 3D TEE SAM width. **(A)** 3D TEE in SAM LVOT view demonstrating narrow SAM width of 8 mm. **(B)** 3D TEE in SAM LVOT view demonstrating wide SAM width of 15 mm.

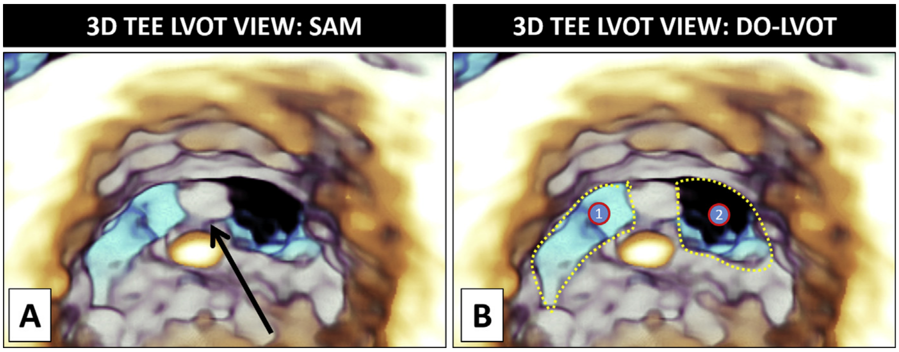

Figure 4 DO-LVOT on 3D TEE. **(A and B)** 3D of SAM in midsystole, viewed looking up from the LV toward the MV and LVOT. The DO-LVOT on 3D TEE is demonstrated; the individual orifices (1 and 2) are highlighted using *dashed yellow lines*. The *black arrow* points to the SAM. This en face view of the MV contrasts with the typical surgeon's view of the MV from the left atrium, looking down the SAM shown here, and cannot be visualized from this conventional 3D view.

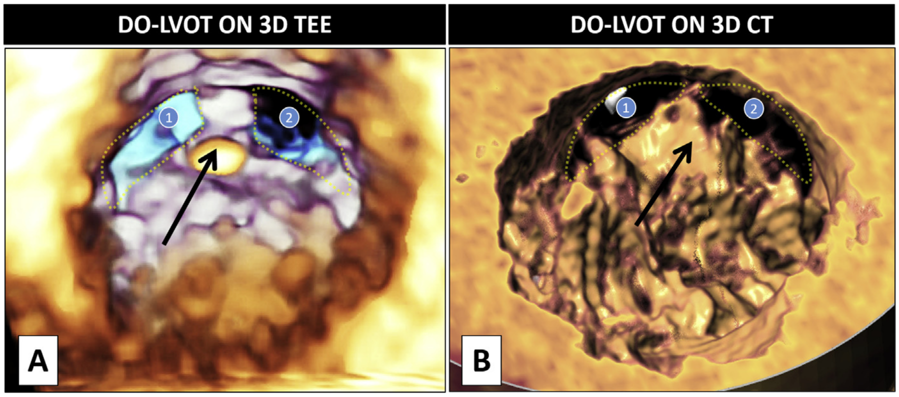

Figure 5 DO- LVOT, equivalent 3D TEE and 3D CT views. **(A)** 3D of SAM with a DO-LVOT on 3D TEE. Individual orifices (1 and 2) are highlighted using *dashed yellow lines*. **(B)** 3D of SAM with a DO-LVOT on 3D CT. Individual orifices (1 and 2) are highlighted using *dashed yellow lines*. The *black arrow* points to SAM within LVOT.

UNIQUE OVERALL VIEWS

Using these models, one may view SAM from an en face view or from a side/oblique perspective throughout the cardiac cycle (Figure 6, Supplemental Video 6, available at www.onlinejase.com).

Systolic Views

During systole, 3D modeling of SAM anatomy demonstrates what we named the "dolphin smile" phenomenon. Two components give rise to this phenomenon: (1) the excessively long MV leaflets protruding into the LVOT appear as the dolphin's snout and (2) the

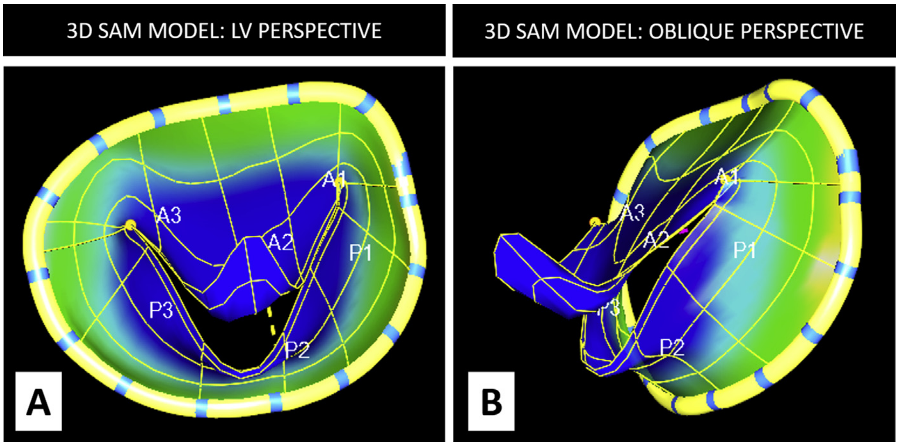

Figure 6 3D SAM dynamic modeling. **(A)** 3D SAM model from LV/en face perspective. **(B)** 3D SAM model from oblique perspective. This technology, available commercially using Esie Valves software (Siemens Healthineers), avoids 3D dropout and side lobe artifact and creates static and dynamic models of SAM.

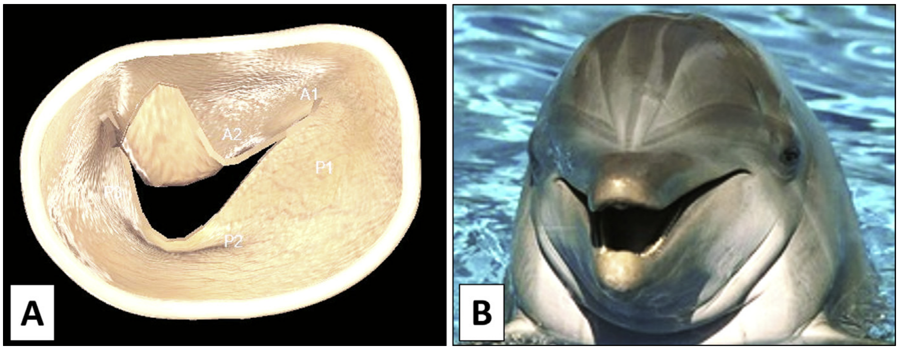

Figure 7 Dolphin smile phenomenon. **(A)** 3D SAM model from LV/en face perspective demonstrating the dolphin smile phenomenon. **(B)** Image depicting a characteristic dolphin smile.

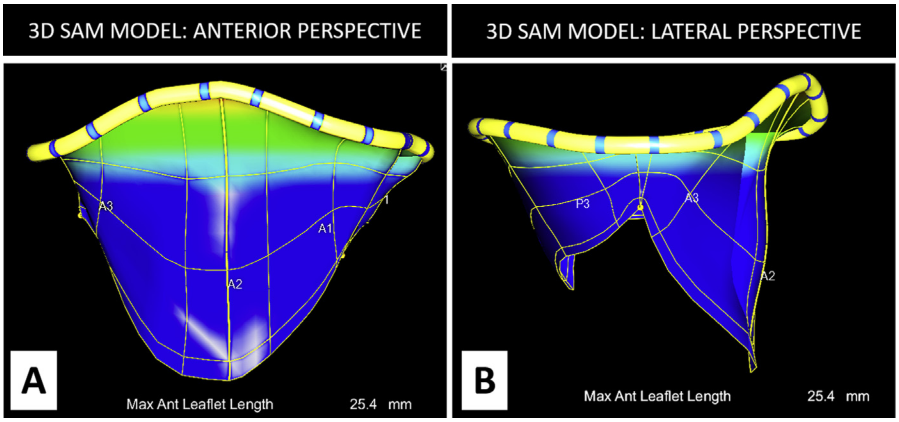

Figure 8 3D SAM model: automated anterior mitral leaflet length. **(A)** 3D SAM model demonstrating automated measurement of anterior mitral leaflet length from an anterior perspective. **(B)** 3D SAM model demonstrating automated measurement of anterior mitral leaflet length from a lateral perspective.

resulting mitral regurgitant orifice appears as the dolphin's mouth (Figure 7).

Diastolic Views

Mitral valve models may be useful in providing automated measurements of mitral leaflet lengths (Figure 8). The MV model may also be helpful in determining candidacy for surgical or percutaneous edge-to-edge repair by providing a diastolic MV area (Figure 9); a diastolic MV area of <4.0 cm^2 is considered to be a relative contraindication.[13] At present, the use of percutaneous edge-to-edge techniques to treat SAM is considered experimental. We have employed it in a highly selective manner when patients cannot have surgery because of frailty yet have both severe SAM and severe intrinsic mitral pathology that

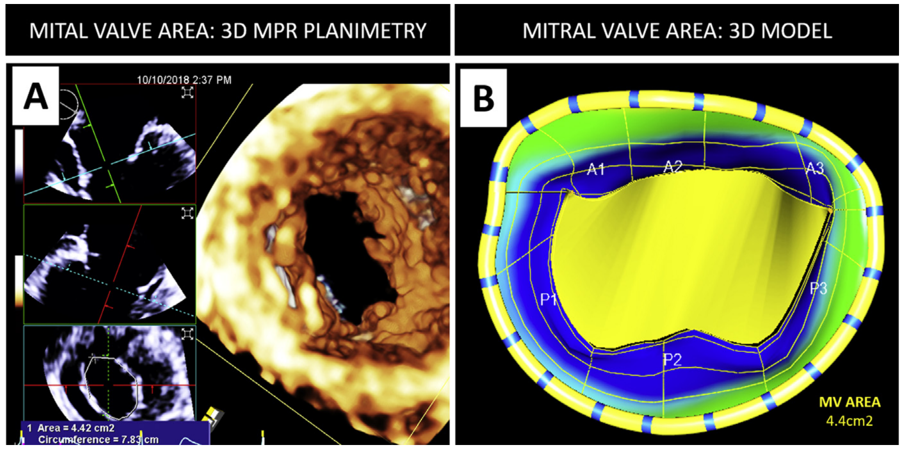

Figure 9 Diastolic MV area modeling. **(A)** Measurement of diastolic MV area prior to MV clipping using 3D TEE multiplanar reconstruction (MPR) planimetry. **(B)** Measurement of diastolic MV area prior to MV clipping using 3D MV modeling.

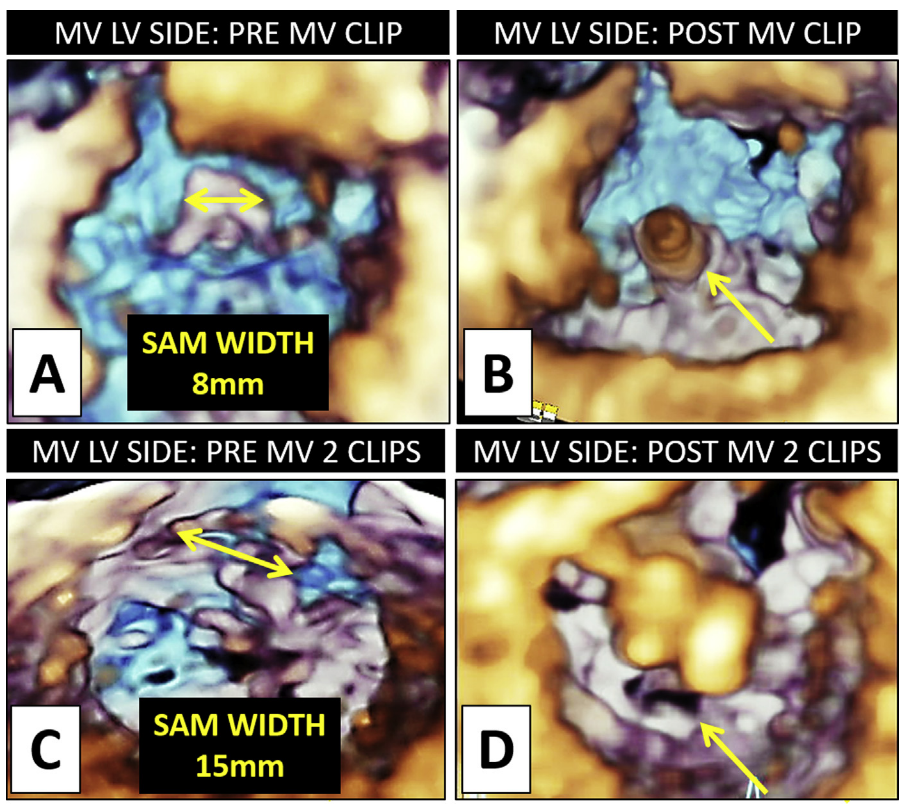

Figure 10 SAM width: number of MitraClips. **(A and B)** Measurement of SAM width of only 8 mm **(A)** and subsequent successful alleviation of SAM using one MitraClip **(B)**. **(C and D)** Measurement of SAM width of 15 mm **(C)** and subsequent successful alleviation of SAM using two MitraClips **(D)**.

would not likely respond to alcohol ablation, that is, concomitant mitral prolapse. It is also a therapeutic option when patients have unsuitable coronary anatomy for ablation.

3D SAM WIDTH: IMPLICATIONS FOR PERCUTANEOUS SAM REPAIR

Surgical repair of SAM with septal myectomy is the mainstay for treatment of obstructive HCM and has been performed for >50 years. In recent years the development of HCM centers of excellence have allowed a concentration of surgical expertise and experience, such that in-hospital mortality for myectomy at such centers averages 0.6%. Myectomy can be combined with native MV modifying techniques, such as horizontal anterior mitral leaflet plication, residual leaflet excision (also called ReLex), and resection/release of the papillary muscles or chordae.[1,4,14-16] Mitral valve replacement can be performed if repair is not possible because of severe calcification of either the leaflets or the annulus; however, if the valve is not calcified a native mitral sparing operation is greatly preferred to

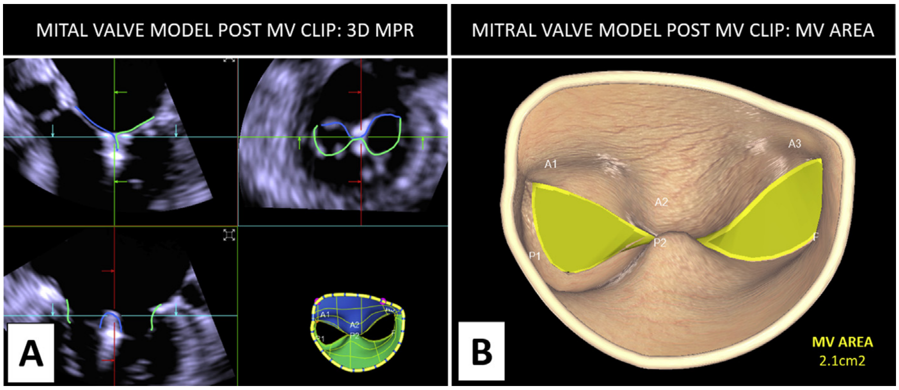

Figure 11 3D MV modeling after MV clipping. **(A)** After percutaneous MV clipping is performed, an MV model is generated using 3D TEE and edited on 3D multiplanar reconstruction (MPR). A DO MV is well visualized.[18] **(B)** 3D surgeon's view is shown using 3D MV modeling after percutaneous MV clipping. Diastolic MV area after MV clipping is measured at 2.1 cm^2.

avoid prosthetic replacement and its attendant morbidity. Although surgical SAM repair has an excellent success rate with low perioperative morbidity and mortality, there is a subset of patients who have prohibitive surgical risk. Alcohol septal ablation is an option for these patients; however, this treatment does not address MV pathology, may have limited success rates, and has a higher risk of causing heart block than surgery.

Percutaneous edge-to-edge repair is a novel approach to treating SAM. It utilizes a transcatheter mitral clip to connect the anterior and posterior mitral leaflets to provide restraint to the anterior leaflet and reduce the amount of mitral tissue protruding into the LVOT during systole. There is limited experience with this technology as a therapy for SAM.[17] In published reports, typically only one clip is placed for this scenario. However, SAM width of the LVOT determined by 3D TEE may be a key anatomic parameter in determining whether one or more clips are needed for successful percutaneous edge-to-edge relief of SAM (Figure 10, Supplemental Video 7, available at www.onlinejase.com). Given limited experience with this approach with a small number of patients the appropriate selection of those likely to benefit from the one- versus two-clip approach is incompletely resolved. Therefore, further clinical studies may elucidate the optimal (single vs multiple clip) approach to SAM treatment as well as the role of 3D SAM width in severity of degree of LVOT obstruction.

Three-dimensional TEE imaging is essential for procedural guidance and evaluation of procedural success. Ideally, the percutaneous SAM repair should eliminate LVOT obstruction by using the posterior leaflet to provide restraint to the anterior leaflet, thus preventing SAM (while avoiding iatrogenic MV stenosis). Occasionally SAM persists even after clip deployment; we refer to that phenomenon as "clip SAM," which can be a catastrophic complication. This occurs because the clip does not provide enough restraint and the anterior leaflet and the apparatus are pushed into the septum, occupying an even larger area of the outflow tract than initially. It is possible that modeling of the outflow tract and the MV as described here might predict the likelihood of clip SAM, prompting variation in technique or intelligence to avoid clipping entirely. A crucial variable that may be difficult to predict is the extent of leaflet slack. Excessive slack unrelieved by the clip can be lethal. Three-dimensional MV modeling may be applied after MV clipping to determine residual diastolic MV area (Figure 11, Supplemental Video 8, available at www.onlinejase.com).

CONCLUSION

Three-dimensional imaging of the MV enables an entirely new method of visualization of SAM. Data sets obtained by 3D TEE can be used to generate unique models of MV anatomy and pathophysiology in obstructive HCM. This novel perspective of 3D TEE visualization has implications not only for elucidation of pathologic mechanisms but also for applications in surgical and percutaneous therapy.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.echo.2020.08.019.

REFERENCES

1. Sherrid MV, Balaram S, Kim B, Axel L, Swistel DG. The mitral valve in obstructive hypertrophic cardiomyopathy: a test in context. J Am Coll Cardiol 2016;67:1846-58.
2. Cape EG, Simons D, Jimoh A, Weyman AE, Yoganathan AP, Levine RA. Chordal geometry determines the shape and extent of systolic anterior mitral motion: in vitro studies. J Am Coll Cardiol 1989;13:1438-48.
3. Klues HG, Maron BJ, Dollar AL, Roberts WC. Diversity of structural mitral valve alterations in hypertrophic cardiomyopathy. Circulation 1992;85:1651-60.
4. Nampiaparampil RG, Swistel DG, Schlame M, Saric M, Sherrid MV. Intraoperative two- and three-dimensional transesophageal echocardiography in combined myectomy-mitral operations for hypertrophic cardiomyopathy. J Am Soc Echocardiogr 2018;31:275-88.
5. Ro R, Halpern D, Sahn DJ, Homel P, Arabadjian M, Lopresto C, et al. Vector flow mapping in obstructive hypertrophic cardiomyopathy to assess the relationship of early systolic left ventricular flow and the mitral valve. J Am Coll Cardiol 2014;64:1984-95.
6. Braunwald E, Morrow AG, Cornell WP, Aygen MM, Hilbish TF. Idiopathic hypertrophic subaortic stenosis: clinical, hemodynamic and angiographic manifestations. Am J Med 1960;29:924-45.
7. Björk VO, Hultquist G, Lodin H. Subaortic stenosis produced by an abnormally placed anterior mitral leaflet. J Thorac Cardiovasc Surg 1961;41:659-69.

8. Shah PM, Gramiak R, Kramer DH. Ultrasound localization of left ventricular outflow obstruction in hypertrophic obstructive cardiomyopathy. Circulation 1969;40:3-11.
9. Sherrid MV, Gunsburg DZ, Moldenhauer S, Pearle G. Systolic anterior motion begins at low left ventricular outflow tract velocity in obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol 2000;36:1344-54.
10. Hahn RT, Abraham T, Adams MS, Bruce CJ, Glas KE, Lang RM, et al. Guidelines for performing a comprehensive transesophageal echocardiographic examination: recommendations from the American Society of Echocardiography and the Society of Cardiovascular Anesthesiologists. J Am Soc Echocardiogr 2013;26:921-64.
11. Jafar N, Moses MJ, Benenstein RJ, Vainrib AF, Slater JN, Tran HA, et al. 3D transesophageal echocardiography and radiography of mitral valve prostheses and repairs. Echocardiography 2017;34:1687-701.
12. Jiang L, Levine RA, King ME, Weyman AE. An integrated mechanism for systolic anterior motion of the mitral valve in hypertrophic cardiomyopathy based on echocardiographic observations. Am Heart J 1987;113:633-44.
13. Mauri L, Garg P, Massaro JM, Foster E, Glower D, Mehoudar P, et al. The EVEREST II Trial: design and rationale for a randomized study of the evalve mitraclip system compared with mitral valve surgery for mitral regurgitation. Am Heart J 2010;160:23-9.
14. Balaram SK, Ross RE, Sherrid MV, Schwartz GS, Hillel Z, Winson G, et al. Role of mitral valve plication in the surgical management of hypertrophic cardiomyopathy. Ann Thorac Surg 2012;94:1990-7. discussion 7-8.
15. Swistel DG, Sherrid MV. The surgical management of obstructive hypertrophic cardiomyopathy: the RPR procedure-resection, plication, release. Ann Cardiothorac Surg 2017;6:423-5.
16. Halpern DG, Swistel DG, Po JR, Joshi R, Winson G, Arabadjian M, et al. Echocardiography before and after resect-plicate-release surgical myectomy for obstructive hypertrophic cardiomyopathy. J Am Soc Echocardiogr 2015;28:1318-28.
17. Sorajja P, Pedersen WA, Bae R, Lesser JR, Jay D, Lin D, et al. First experience with percutaneous mitral valve plication as primary therapy for symptomatic obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol 2016;67:2811-8.
18. Vainrib AF, Loulmet DF, Williams MR, Saric M. Tale of 2 orifices. Circ Cardiovasc Imaging 2019;12:e008372.