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Myosin Inhibitors Can Reduce Primary Mitral Regurgitation by Improving Systolic Anterior Motion of the Mitral Leaflets

Kaitlin Roehl, PA*, and Said Alsidawi, MD, FACC*

Department of Cardiovascular Diseases, Mayo Clinic, Scottsdale, AZ, USA

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Correspondence to
Said Alsidawi, MD, FACC
Department of Cardiovascular Diseases, Mayo Clinic, 13400 E. Shea Blvd., Scottsdale, AZ 85259, USA.
Email: Alsidawi.said@mayo.edu

A 74-year-old male with symptomatic hypertrophic obstructive cardiomyopathy (oHCM) and primary mitral regurgitation (MR) presented to our institution for a second opinion after he was recommended to have surgical myectomy and mitral valve repair. Transthoracic echocardiogram (TTE) showed ejection fraction 76%, septal thickness 18 mm, systolic anterior motion (SAM) of the mitral valve (**Figure 1A**), severe anteriorly directed MR (**Figure 1B**) and resting left ventricular outflow tract (LVOT) gradient of 100 mmHg (**Figure 1C**). A transesophageal echocardiogram was subsequently performed showing P2 scallop prolapse (arrow, **Figure 1D**, **Supplementary Videos 1** and **2**) and severe MR with a highly eccentric, anteriorly directed regurgitant jet (**Figure 1E**). There was SAM along with leaflet coaptation gap exacerbated by SAM (**Figure 1E**).

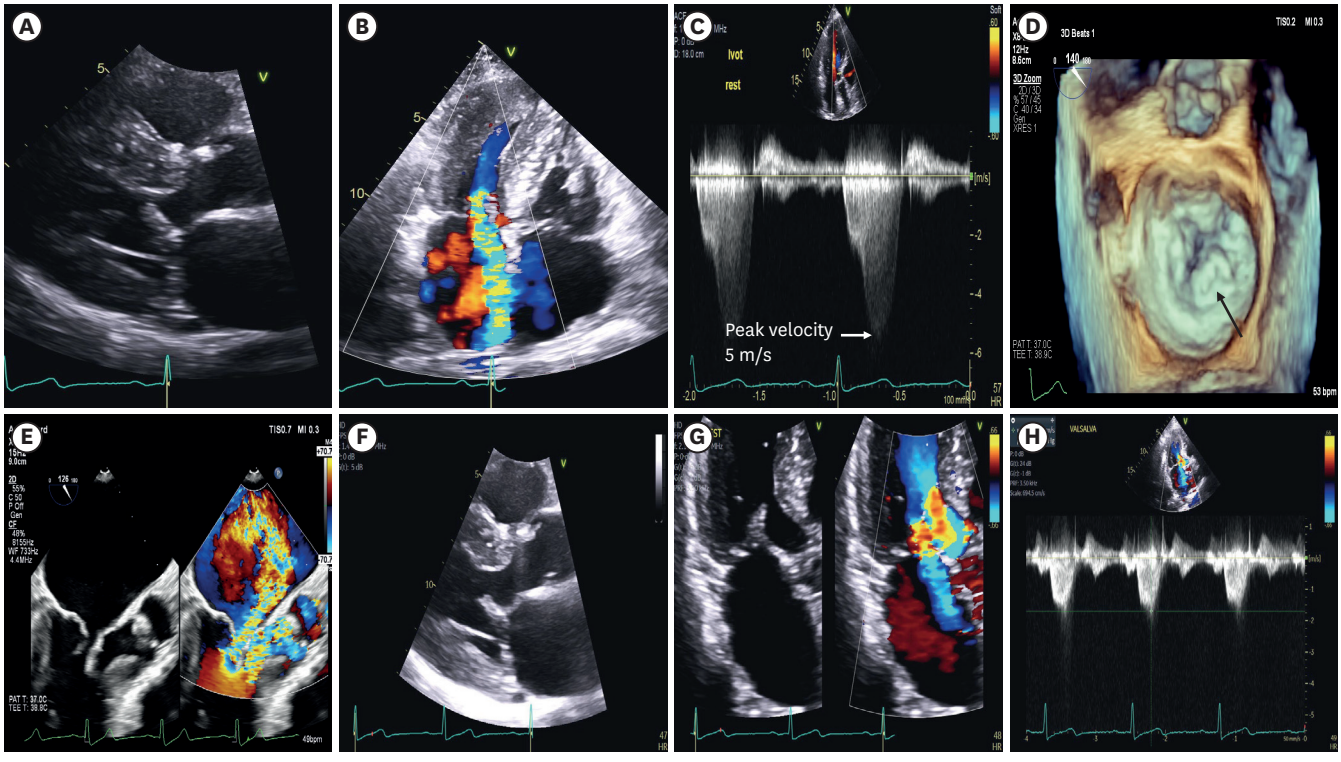

Figure 1. Images showing the improvement of MR on Mavacamten.
(A) Parasternal long axis image showing significant SAM. (B) Apical 4-chamber image showing severe anteriorly directed-MR. (C) Doppler image showing severe LVOT obstruction at rest of 5 m/s. (D) 3D TEE showing P2 scallop prolapse. (E) Image showing SAM along with leaflet coaptation gap exacerbated by SAM and severe, anteriorly directed MR jet. (F) Parasternal long axis image showing improvement of SAM on Mavacamten therapy. (G) Apical 4-chamber image with color compare showing mild-moderate residual MR on Mavacamten. (H) Doppler image of the LVOT showing no residual obstruction.
LVOT = left ventricular outflow tract; MR = mitral regurgitation; SAM = systolic anterior motion; 2D = two dimensional; 3D = three dimensional.

*Kaitlin Roehl and Said Alsidawi contributed equally to this work.

ORCID iDs
Kaitlin Roehl
https://orcid.org/0009-0001-6908-8608
Said Alsidawi
https://orcid.org/0000-0001-8750-7880

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Conflict of Interest
The authors have no financial conflicts of interest.

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The data generated in this study is available from the corresponding author upon reasonable request.

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The effective regurgitant orifice by proximal iso-velocity surface area was 0.44 cm^2.

The patients preferred medical therapy and it was felt that the degree of MR was exacerbated by SAM. Decision was made to initiate therapy with Mavacamten to relieve LVOT obstruction and SAM with re-evaluation of MR severity. After 4 weeks on Mavacamten, the patient had significant improvement in symptoms. He underwent TTE which showed an ejection fraction of 66%, significant improvement in SAM (**Figure 1F**), mild-moderate residual MR (**Figure 1G**) and no residual LVOT obstruction (**Figure 1H**).

Mitral valve regurgitation related to SAM in oHCM can further worsen when concomitant mitral valve prolapse is present with unusual jet direction (anterior in this case). We show for the first time that relieving LVOT obstruction and SAM using a myosin inhibitor can also improve mitral valve regurgitation in this population.

An informed consent was obtained from the patient to publish this work.

SUPPLEMENTARY MATERIALS

Supplementary Video 1

3D TEE of the mitral valve showing P2 scallop prolapse.

Supplementary Video 2

TrueView 3D image of the mitral valve showing P2 scallop prolapse.