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CASE REPORT

ADVANCED

CLINICAL CASE

Valve-in-Mitral Annular Calcification Transcatheter Mitral Valve Replacement After Thrombosis of Extracardiac Valved Conduit

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ABSTRACT

We report a patient with severe mitral annular calcification, mitral stenosis/regurgitation, hypertrophic obstructive cardiomyopathy, and subaortic membrane treated with valved left atrium—left ventricle conduit, septal myectomy, and membrane resection. Subsequent thrombosis of the conduit prompted successful valve-in- mitral annular calcification transcatheter mitral valve replacement and laceration of the anterior mitral leaflet to prevent outflow obstruction. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1267–1273) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 72-year-old woman presented with New York Heart Association (NYHA) functional class III heart failure and exertional near-syncope due to severe mitral annular calcification (MAC) with a mean transmitral gradient of 12 mm Hg, orifice area of 1.2 cm^2 (Video 1), moderate mitral regurgitation, basal septal thickness of 22 mm, left ventricular outflow tract (LVOT) gradient of 70 mm Hg, and ejection fraction of 70%. Despite medical therapy with metoprolol, diltiazem, disopyramide, and diuretics, she had persistent symptoms. She was referred to a quaternary surgical center for surgical intervention with planned mitral valve replacement and septal myectomy. Intraoperatively, the surgical team had a strong concern about serious complications, given the significant degree of MAC. Therefore, the patient underwent: 1) construction of a conduit anastomosed to the atrial appendage and the left ventricular apex (26-mm Hemashield graft with a 25-mm Hancock bioprosthetic valve at the distal end of the conduit);

LEARNING OBJECTIVES

- To recognize valve-in-MAC TMVR as an emerging strategy to treat severe mitral stenosis in selected patients with calcific mitral valve disease.
- To understand the technical aspects of valve-in-MAC TMVR and laceration of the anterior mitral leaflet to prevent outflow obstruction in patients at risk for left ventricular outflow tract obstruction.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

INR = international normalized ratio

LAMPOON = laceration of the anterior mitral leaflet to prevent outflow obstruction

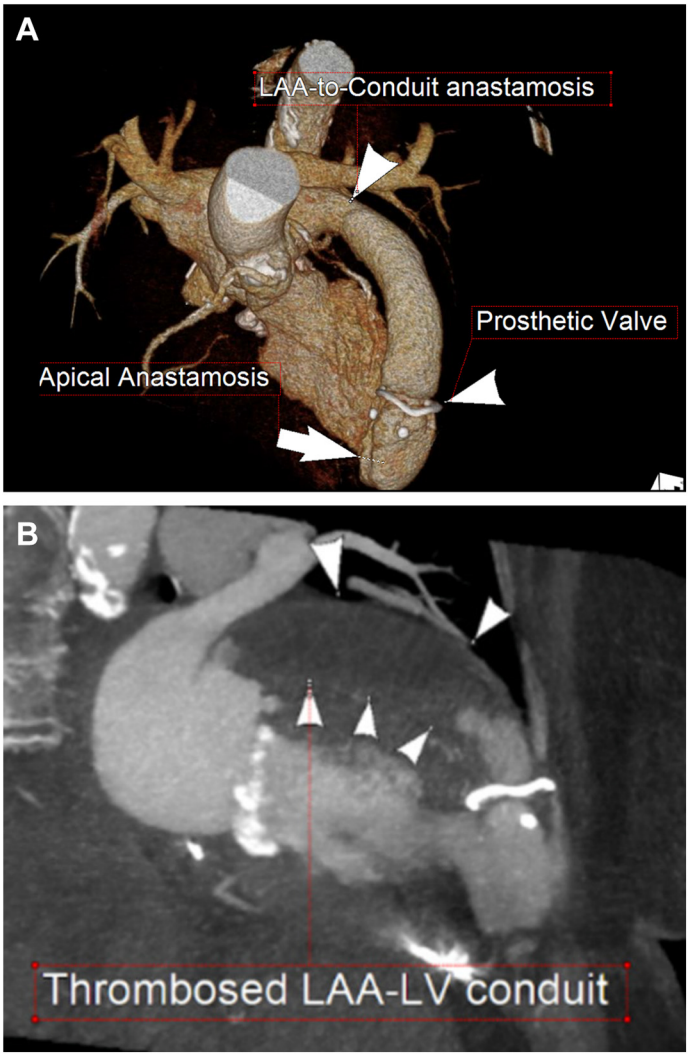
LVOT = left ventricular outflow tract

MAC = mitral annular calcification

TMVR = transcatheter mitral valve replacement

2) septal myectomy; and 3) resection of a subaortic membrane (Figure 1A). The patient experienced resolution of dyspnea and was given warfarin with a goal of 2.5 to 3.5 international normalized ratio (INR). Six months later, she experienced refractory symptomatic atrial fibrillation and required atrioventricular nodal ablation and implantation of a bi-ventricular implantable cardioverter defibrillator. During this time her INR was found to be intermittently subtherapeutic. Computed tomographic angiography (CTA) revealed thickening of the conduit valve leaflets, suggestive of partial leaflet thrombosis. She continued to take warfarin. Twelve months later, she experienced severe dyspnea and was rehospitalized.

FIGURE 1 Cardiac Computed Tomography



(A) Computed tomography volume rendering after surgical construction of a left atrium–left ventricle conduit. **(B)** Thrombosed valved left atrium-left ventricle conduit. LAA = left atrial appendage.

MEDICAL HISTORY

The patient had a long history of exertional dyspnea and pulmonary hypertension. Right heart catheterization before a cardiology consultation was significant for pulmonary artery pressure 82/30 mm Hg (mean 50 mm Hg), pulmonary capillary wedge pressure 18 mm Hg, Fick cardiac output 3.8 L/min, and pulmonary vascular resistance 8.4 WU. Pulmonary vasodilators were discontinued after the echocardiogram revealed structural heart disease as described above. The patient also had non-insulin-dependent diabetes, obesity, and hypertension.

DIFFERENTIAL DIAGNOSIS

The cause of her worsening heart failure was likely thrombosis of the valved conduit and uncorrected severe mitral valve obstruction. Contributing factors include recurrent atrial fibrillation and loss of atrial contraction.

INVESTIGATION

CTA confirmed complete thrombosis of the conduit (Figure 1B). The native mitral valve was obstructed, similarly to its preoperative condition (Figure 2). Basal septal thickness was 15 mm, and there was no residual LVOT gradient. An electrocardiogram displayed normal function of the BiV pacemaker with underlying atrial fibrillation. B-type natriuretic peptide was 224 pg/nL. INR preceding hospitalization was intermittently subtherapeutic.

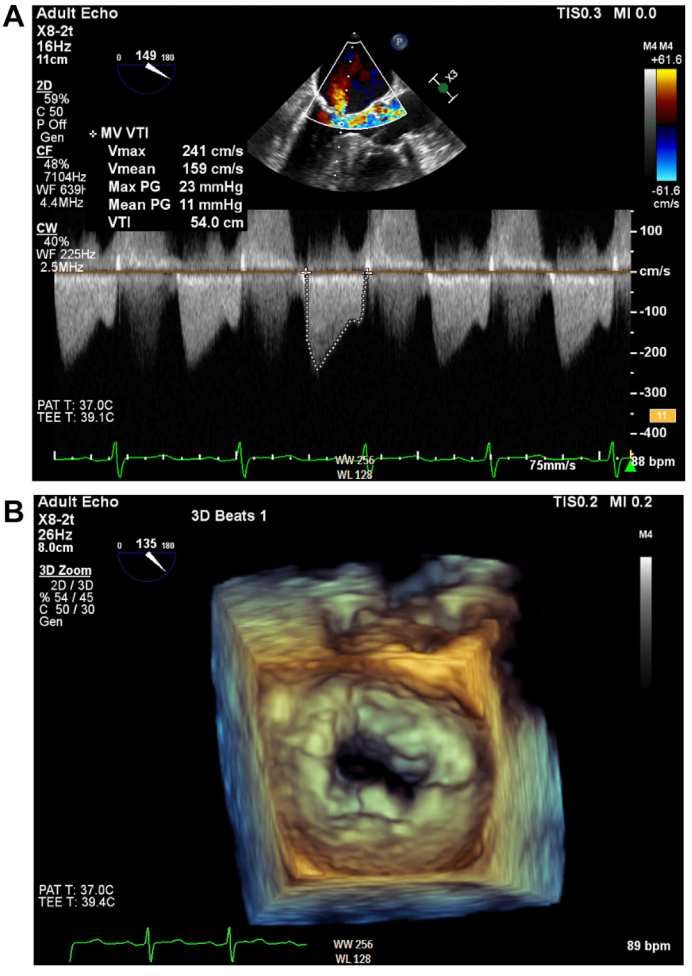
MANAGEMENT

A multidisciplinary heart team evaluation concluded that the patient was not a candidate for redo surgery. The CT MAC score was 9,[1] and the annular area within the calcific ring was 480 mm^2. Valve-in-MAC transcatheter mitral valve replacement (TMVR) was recommended. The predicted neo-LVOT according to CT planning software was 174 mm^2 (Figure 3), suggesting increased risk of LVOT obstruction after TMVR.[2] Alcohol ablation was not considered owing to mild thickness of the basal septum, so laceration of the anterior mitral leaflet to prevent outflow obstruction (LAMPOON) was planned before valve deployment.

A transvenous pacer and an intra-aortic balloon pump were placed. Inferior-posterior trans-septal puncture was performed under guidance from transesophageal echocardiography. Two steerable sheaths were placed in the LA through the same trans-septal access. Antegrade LAMPOON was performed, as previously described.[3] Through 1 of the steerable sheaths, a balloon-tipped catheter was passed across the mitral valve into the ascending aorta. Via femoral arterial access, a JR4 guiding catheter and 18/30 Ensnare were placed in the ascending aorta. An 0.014-inch 300-cm wire was advanced through the balloon-tipped catheter, snared, and externalized via the femoral artery. A JL 3.5 guide was advanced antegrade over this wire and used to position an 18/30 Ensnare in the LVOT. Through the other trans-septal sheath, a JR4 guiding catheter was directed toward the base of an A2 scallop. Through the JR4, an 0.014-inch Astato 20 wire insulated within a piggyback 0.035-inch converter microcatheter was used to traverse the base of the A2 scallop by advancing on 50W cut mode. The Astato wire was snared in the LVOT. The piggyback was withdrawn to create a 90° bend at the middle of the wire, then locked in place. Leaflet laceration was performed with retraction of the guide catheters while delivering 70 W in cut mode (Video 2).

Atrial septostomy was performed using a 14 × 40-mm balloon over an extra-small Safari wire positioned in the LV apex. A 29-mm Edwards Sapien Ultra valve was advanced across the septum and implanted in the MAC during rapid pacing (**Figure 4,** Video 3). Echocardiographic imaging after implantation demonstrated no transvalvular leak and a trivial paravalvular leak with a mean gradient of 2 mm Hg at 55 beats/min (**Figure 5,** Videos 4 and 5). The mean pulmonary arterial pressure decreased from 62 mm Hg to 42 mm Hg (**Figure 6**). The LVOT gradient was negligible.

FIGURE 2 Baseline Transesophageal Echocardiography



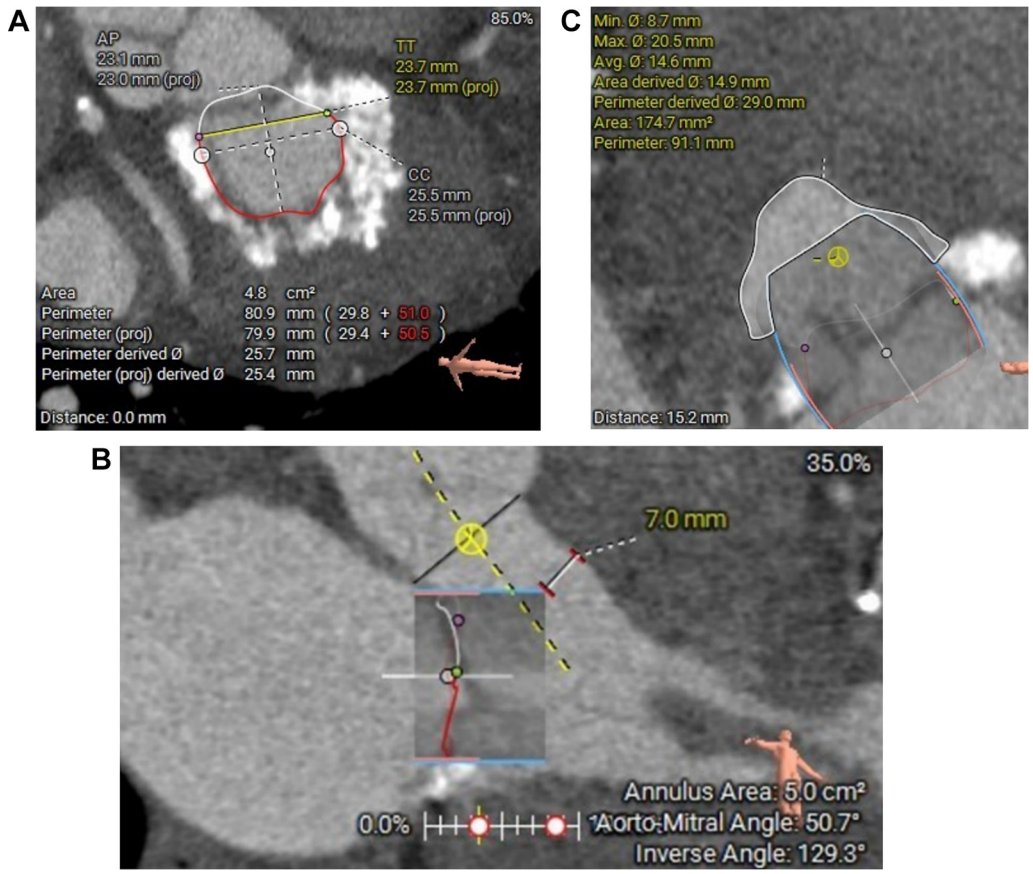
(A) Hemodynamic profile of native mitral valve demonstrating severe mitral stenosis and mild mitral regurgitation. **(B)** Three-dimensional echocardiography of severely calcified mitral annulus with reduced orifice area.

DISCUSSION

This patient presented with multiple structural abnormalities initially managed with a surgical approach after medical therapy had failed. In isolation, these conditions are commonly managed with either surgical or catheter-based approaches. Septal myectomy may be definitive therapy in patients with hypertrophic cardiomyopathy and LVOT obstruction, and the outcomes are superior to those of medical therapy.[4] In patients who are not surgical candidates, alcohol septal ablation is an alternative therapy but with a higher risk of residual LVOT gradient and need for repeated intervention.[5]

Surgery for severe calcific mitral valve disease in the setting of severe MAC carries higher rates of morbidity and mortality. Annular debridement may lead to atrioventricular groove disruption, left ventricular free wall rupture, and injury to the left circumflex coronary artery. Creation of a valved left atrium-left ventricle conduit, as was performed for this patient, has been used in selected cases as an alternative surgical approach.[6] At the time of the patient's presentation, catheter-based approaches

FIGURE 3 Pre-Procedural Computed Tomography for TMVR Planning



(A) Circumferential mitral annular calcification (MAC) with CT-based MAC score of 9. **(B and C)** Predicted neo-left ventricular outflow tract (LVOT) using CT postprocessing modeling software.

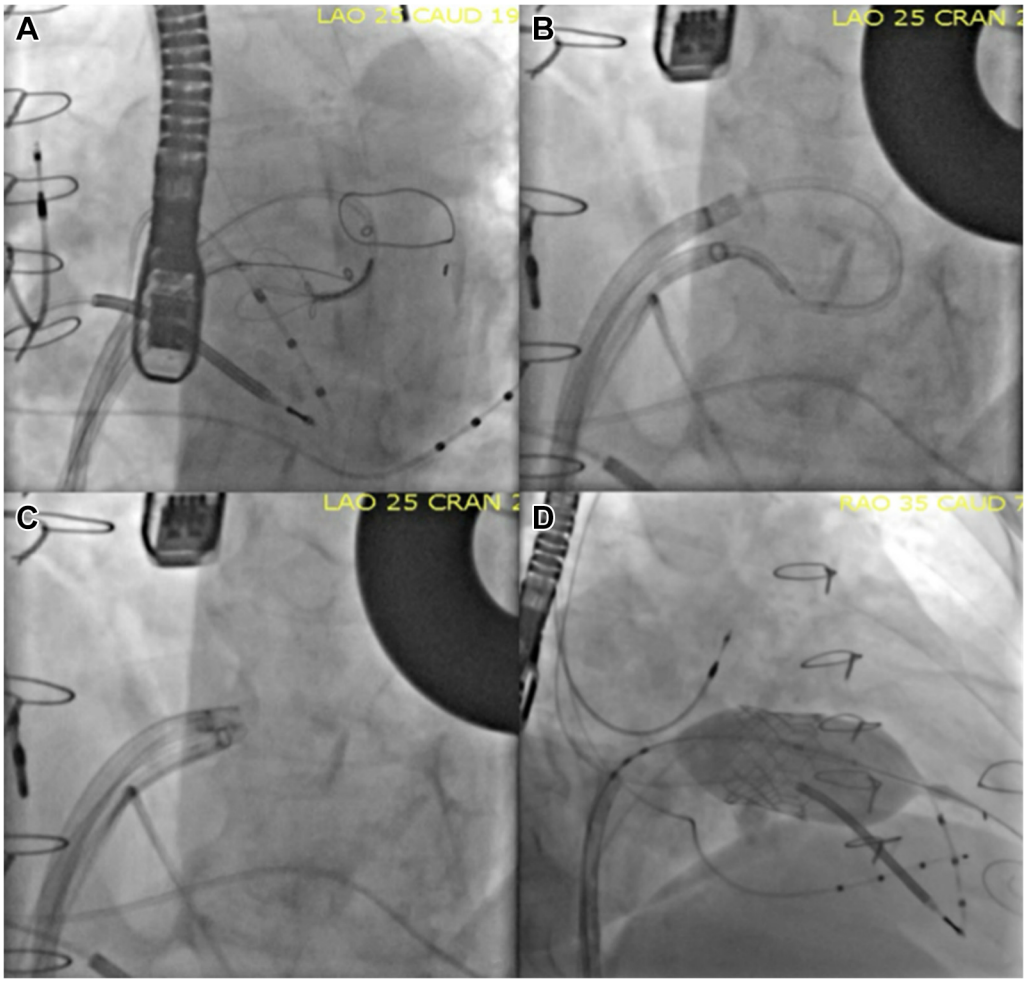
for calcific mitral stenosis were not readily available. As technology, procedural planning, and operator experience have developed, TMVR has emerged as a promising alternative to surgical replacement.

TMVR is a nonsurgical alternative for symptomatic patients with severe MAC and concomitant severe mitral valve dysfunction. When compared with valve-in-valve and valve-in-ring, valve-in-MAC TMVR carries the highest risk of mortality, stroke, LVOT obstruction, and valve embolization.[7,8] Mortality is driven by multiple factors, including implantation of a circular prosthesis into a saddle-shaped valve, which can cause paravalvular leaks, difficulty measuring the annular area in the presence of heavy calcification, lack of suitable anchoring. and interaction of the subvalvular apparatus with the LVOT. However, the most important predictor of early and late mortality is the interaction between the native valve apparatus and the LVOT causing obstruction.[7-9] Preprocedural planning with cardiac CT is paramount to identify patients at risk for LVOT obstruction needing septal modification or LAMPOON before valve implantation. Predicted neo-LVOT areas <190 mm^2 after implantation of the prosthesis are associated with higher rates of LVOT obstruction and subsequent mortality.[2,7] Techniques to increase the neo-LVOT area include contemporary septal reduction techniques or LAMPOON, which enhance the safety of TMVR.[10]

FOLLOW-UP

The patient's condition improved after valve implantation, and her hospital course was uneventful.

FIGURE 4 Transcatheter Mitral Valve Replacement Procedure



(A) Leaflet transversal and snaring. **(B)** Positioning of bend in Astato wire at leaflet base. **(C)** Post-leaflet laceration. **(D)** Edwards Sapien Ultra valve deployment in mitral position.

She had New York Heart Association functional class II dyspnea and normal prosthetic valve function at her 30-day follow-up visit.

CONCLUSIONS

Valve-in-MAC TMVR is an emerging strategy to treat high-risk patients with severe MAC and concomitant severe mitral valve disease.

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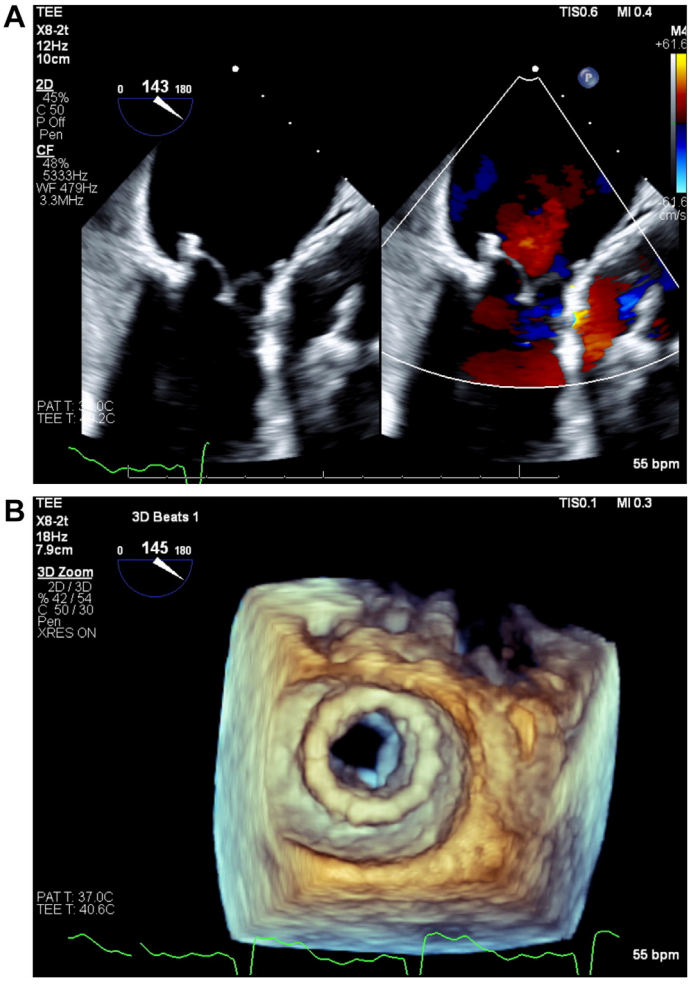
FIGURE 5 Intraprocedural Transesophageal Echocardiography



(A) Two-chamber view showing no transvalvular/trivial perivalvular leak. **(B)** Three-dimensional view showing Edwards Sapien Ultra valve in mitral position.

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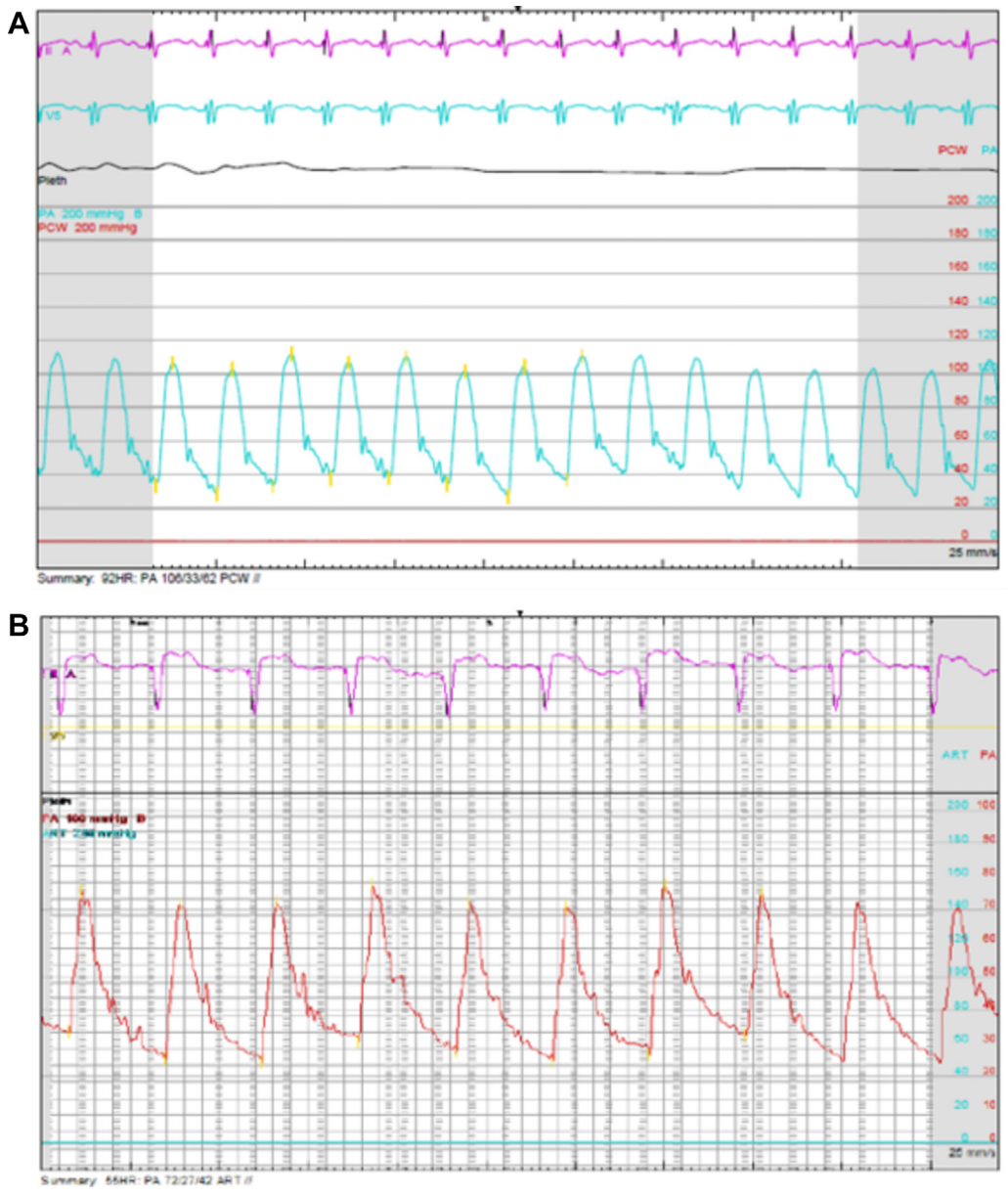
FIGURE 6 **Invasive Hemodynamics**



(A) Pulmonary arterial pressure (PA) 106/33/62 mm Hg before transcatheter mitral valve replacement (TMVR). **(B)** PA 72/27/42 mm Hg after TMVR.

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KEY WORDS computed tomography, echocardiography, hemodynamics, mitral valve, pulmonary hypertension, valve replacement

APPENDIX For supplemental videos, please see the online version of this paper.