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CASE REPORT

ADVANCED

CLINICAL CASE

Intractable Heart Failure From a Severe Mitral Paravalvular Leak in Surgically Created Bipartite Left Atrium

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ABSTRACT

We describe a 38-year-old Middle Eastern woman with a long history of multiple cardiac surgical procedures for mitral valve disease who presented with intractable heart failure from severe mitral paravalvular regurgitation requiring multiple medical admissions since 2019. She was deemed a very high surgical risk and was treated successfully with a percutaneous technique. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1542–1547)

INTRODUCTION

A paravalvular leak (PVL) is regurgitation between the sewing ring of the prosthetic valve and the native heart tissue, the annulus. It can result from improper sealing in the case of a calcified annulus or weak tissues from inflammation or infection. The estimated incidence of mitral valve (MV) PVL is approximately 7% to 17%. Although most PVLs are asymptomatic, 1% to 5% of patients have serious clinical consequences, which may consist of hemolytic anemia, infective endocarditis, or heart failure.[1] Currently, the gold standard treatment is redo cardiac surgery involving either repair of the leak or repeat replacement of the valve. In very high risk surgical cases, transcatheter techniques have been used to address PVL.[2,3]

LEARNING OBJECTIVES

- To be able to make a differential diagnosis of severe PVL with multimodality imaging.
- To understand the role of device closure in management.

HISTORY OF PRESENTATION

A 38-year-old Middle Eastern woman with a long history of multiple cardiac surgical procedures for MV disease presented with dyspnea on mild exertion, easy fatigability, and palpitations of 2 months' duration.

PAST HISTORY

Her clinical history was relevant for multiple cardiac surgical procedures since 2002: MV and tricuspid valve repair in 2002 for severe MV and tricuspid regurgitation at the age of 20 years; MV replacements (MVRs) with a 23-mm St Jude Medical prosthesis in 2016 and 2018; and in 2019, MVR with a 25-mm mechanical prosthesis after she presented with acute heart failure and an echocardiogram revealed dehiscence of the of prosthetic valve sewing ring along the medial aspect with severe PVL. The MVR is 2019 was her fourth cardiac surgery with her third MVR. Because the mitral annulus was weak and torn, a neoannulus was created in the left atrium (LA), and

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the mechanical prosthesis was positioned in the LA below the entrance of the pulmonary veins. All her medical care had been in the Middle East, and her medical records were not available. Two months after her fourth MV intervention, class III symptoms developed, and she was hospitalized multiple times for recurrent heart failure. She came to us for further evaluation and for a possible fifth cardiac surgery or cardiac transplantation.

DIFFERENTIAL DIAGNOSIS

Pulmonary venous obstruction was the initial differential diagnosis considered on the basis of the valve position in the LA. Cardiac computed tomography (CT) clearly revealed the valve dehiscence with normal flowing pulmonary veins, and this was further confirmed with transesophageal echocardiography (TEE).

INVESTIGATIONS

TEE (**Figure 1,** Video 1) revealed a structurally normally functioning MV prosthesis located in the middle of the LA, above the fossa ovalis, below the right pulmonary venous entrance medially, and just below the left pulmonary venous entrance laterally. The LA was divided into a small superior pulmonary venous chamber above and a ventricularized LA below the valve. There was paravalvular dehiscence of 12 × 8 mm along the posterolateral aspect of the sewing ring with severe paravalvular regurgitation, a proximal isovelocity surface area effective regurgitant orifice area of 0.88 cm^2, and regurgitation volume of 88 mL. The left and right ventricular functions were normal. She underwent cardiac CT for better definition of the anatomy with a plan for potential percutaneous intervention. CT confirmed the TEE findings (**Figures 2 and 3,** Video 2).

MANAGEMENT

Her EuroSCORE was 18%, indicating a very high surgical risk, and hence it was decided to address the MV PVL by using a transcatheter technique. The procedure was performed with the patient under general anesthesia, and access to the left ventricular apex was obtained with a small left anterolateral thoracotomy. A 30-cm 10-F Cook sheath was placed in the left ventricle through an apical puncture. Using a telescoped 5-F Judkins right diagnostic catheter, the PVL defect was crossed with a hydrophilic angled Terumo guidewire that was parked in the right superior pulmonary vein. This was then exchanged with a 0.038-inch Amplatz Super Stiff guidewire (Boston Scientific), and the Cook sheath was negotiated carefully into the superiorly placed "pulmonary venous LA chamber." A 14/5-mm Amplatzer Vascular Plug III (Abbott) was deployed, resulting in almost complete closure of the PVL (**Figures 4, 5A to 5C, and 6A and 6B,** Videos 3A to 3C, 4A and 4B, and 5), thus leaving only a small regurgitant jet. The apical puncture site was closed by preplaced purse-string sutures. She made an excellent recovery and was discharged in stable condition. She remains symptom free since discharge.

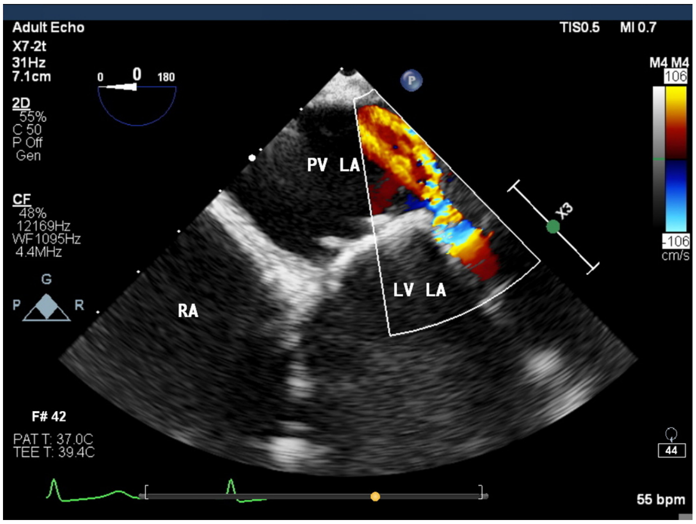
DISCUSSION

Transcatheter techniques have been increasingly used to address PVLs. Two large case series demonstrated a technical success rate ranging from 77% to 86 %, a clinical success rate of almost 90%, and a major adverse event rate of <10% at 30 days.[2,3] Three

ABBREVIATIONS AND ACRONYMS

CT = computed tomography
LA = left atrium
MV = mitral valve
MVR = mitral valve replacement
PVL = paravalvular leak
TEE = transesophageal echocardiography

FIGURE 1 Transesophageal Echocardiogram, 2-Chamber View With Color Flow, Showing Paravalvular Regurgitation



LV LA = left ventricular left atrium; PV LA = pulmonary venous left atrium; RA = right atrium.

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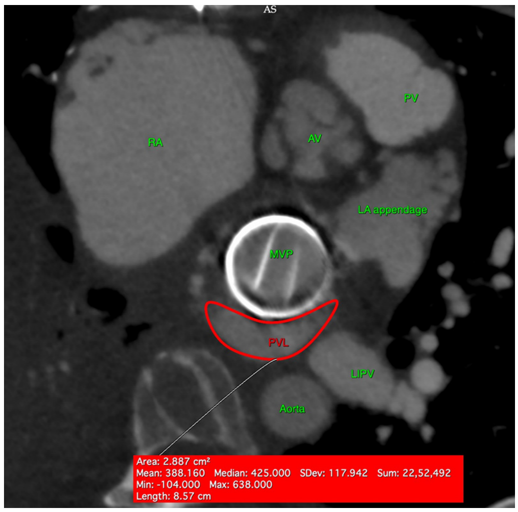
FIGURE 2 Cardiac Computed Tomography, Short-Axis View, Showing Prosthetic Mitral Valve With PVL

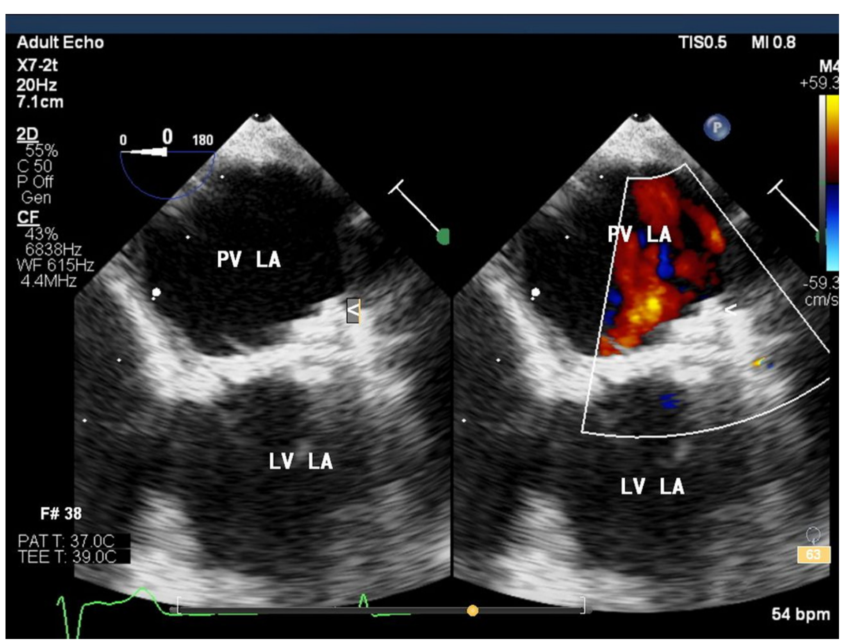


AS = atrial septum; AV = aortic valve; LA = left atrial; LIPV = left inferior pulmonary vein; MVP = mitral valve prosthesis; PV = pulmonary valve; PVL = paravalvular leak; RA = right atrium.

FIGURE 3 Cardiac Computed Tomography, Sagittal View, Showing Prosthetic Mitral Valve in the Mid-Left Atrial Cavity With Paravalvular Leak Around the Posterolateral Wall

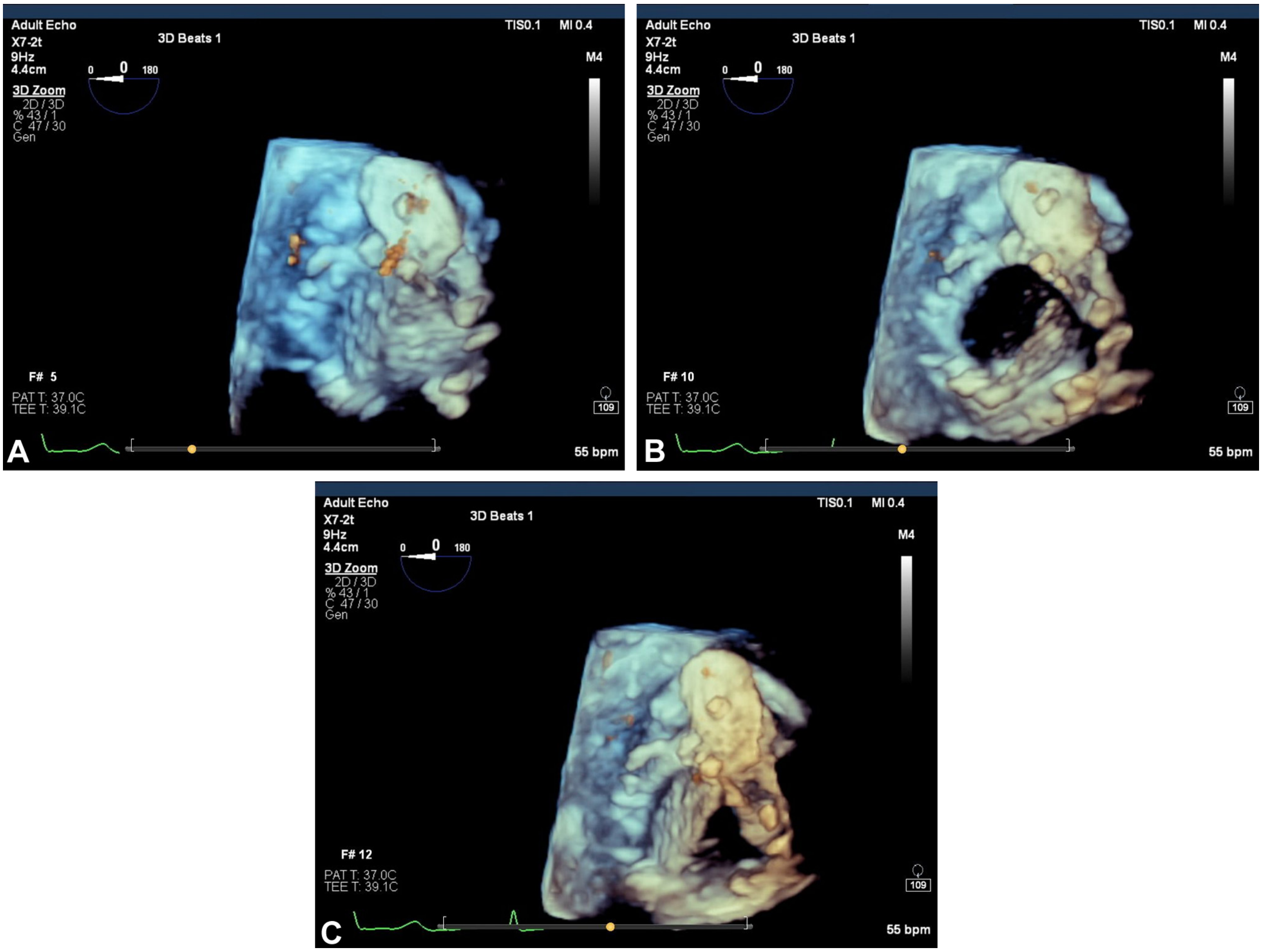
AV = aortic valve; IVS = interventricular septum; LA = left atrium; LV = left ventricle; MV = mitral valve; RV = right ventricle.

FIGURE 4 Postprocedure Transesophageal Echocardiogram, 2-Chamber View Without and With Color Flow, Showing the Amplatzer Vascular Plug Sealing the Leak



The **arrowhead** indicates the Amplatzer Vascular Plug III (Abbott). Abbreviations as in Figure 1.

FIGURE 5 **Postprocedure Transesophageal Echocardiograms, Left Atrial View**



(A to C) Postprocedure 3-dimensional transesophageal echocardiogram, left atrial view, showing the Amplatzer Vascular Plug III (Abbott) with a normally functioning mitral valve.

approaches can be used: retrograde transapical, antegrade transseptal, or retrograde aortic.

In this case, the PVL was approached through the transapical route because the traditional transvenous transseptal approach will lead to the "ventricularized left atrial chamber," given that her MV prosthesis was in the middle of the bipartite LA created during the fourth valve replacement where the fossa ovalis was below the MV prosthesis. The transfemoral arterial approach with or without an arteriovenous loop[2-5] is frequently used when the transvenous approach is not feasible or difficult.

Transapical transcatheter PVL closure has increased in popularity.[6] Although a transaortic retrograde technique was possible, we believed that the small supravalvular left atrial chamber posed a risk of lesser control of a long retroaortic sheath, whereas the transapical approach with a short sheath offers more direct and controlled manipulation, positioning, and release of the plug. This case report emphasizes that severely symptomatic refractory heart failure can be a class 1 indication for closure of a PVL in patients with high surgical risk, given the fast and complete clinical recovery after closure of the defect in our patient.

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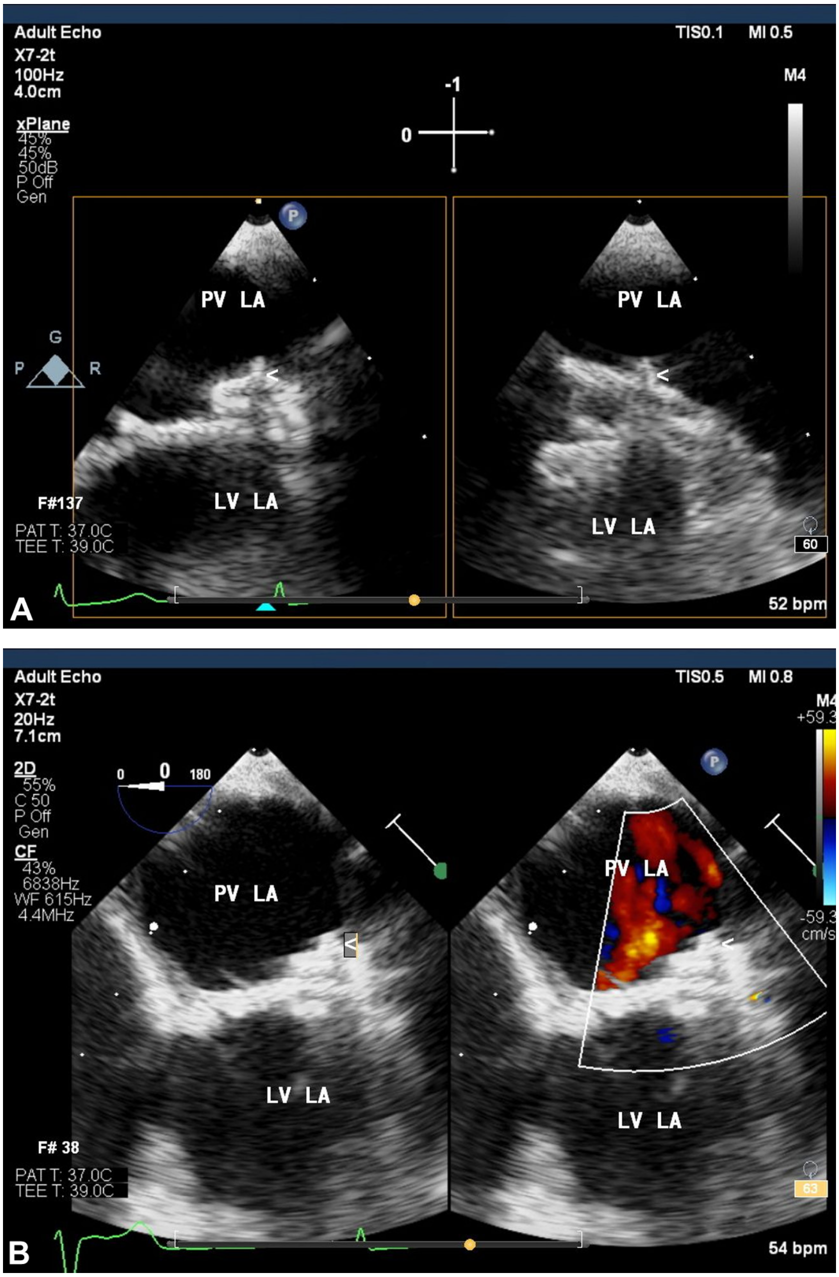
FIGURE 6 **Postprocedure Transesophageal Echocardiograms, Biplane View**



(A and B) Postprocedure transesophageal echocardiogram, biplane view, showing the Amplatzer Vascular Plug III (Abbott) sealing the leak. The **arrowhead** indicates the Amplatzer Vascular Plug III. Abbreviations as in Figure 1.

Furthermore, it demonstrates the feasibility and the efficacy of transcatheter PVL closure even in extremely rare and challenging anatomical scenarios, thereby avoiding the risks inherent in redo cardiac surgery. To the best of our knowledge, this is the first case where a PVL in a very unusual location was treated successfully and managed by a plug in a patient who was a very high surgical risk.

FOLLOW-UP

PVL closure led to a spectacular hemodynamic improvement in the patient. She became asymptomatic during the recovery period and was doing well at her 1-, 3-, and 6-month follow-ups.

CONCLUSIONS

Multimodality imaging solved the puzzle in this clinical case scenario and also guided the transcatheter management option.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS paravalvular leak, surgical bipartite left atrium, transapical device closure

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