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CASE REPORT

INTERMEDIATE

CLINICAL CASE

An Unusual Cause of Shortness of Breath

Pulmonary Vein Stenosis After Surgical Mitral Valve Replacement

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ABSTRACT

A 79-year-old man with prior bioprosthetic mitral valve replacement presented with progressive shortness of breath and was found to have right upper pulmonary vein stenosis and paravalvular leak diagnosed with the use of multimodal imaging. The patient underwent balloon angioplasty, stenting of the pulmonary vein, and paravalvular leak closure with ultimate resolution of symptoms. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:533–537)

HISTORY OF PRESENTATION

A 79-year-old man presented with 1 year of progressive dyspnea on exertion (DOE) and worsening lower-extremity edema. The patient's medical history was notable for chronic diastolic heart failure, malignancies in remission (prostate cancer, breast cancer), hyperlipidemia, coronary artery disease status post-bypass grafting and percutaneous coronary intervention, and an extensive history of mitral valve dysfunction. The patient's valvulopathy was diagnosed at the age of 73 years when the patient began to experience progressive DOE. He was found to have a flail medial scallop of the posterior mitral valve leaflet due to a ruptured chorda tendinea. He underwent mitral valve repair with a 32-mm Cosgrove annuloplasty ring with concomitant coronary bypass grafting of the first diagonal branch. He initially did well after surgery, but 4 years later he had recurrence of DOE with functional decline. Transthoracic echocardiography (TTE) showed severe mitral regurgitation, and he underwent bioprosthetic mitral valve replacement with a 29-mm St Jude Epic valve. He improved after surgery, but again reported DOE after a few months, with transesophageal echocardiography (TEE) demonstrating severe lateral paravalvular leak (PVL). Transcatheter PVL closure was performed with 12-mm and 10-mm Amplatzer vascular plugs (AVP2s); however, intraprocedural TEE demonstrated residual moderate PVL. The patient was referred to cardiothoracic surgery and underwent reoperative bioprosthetic mitral valve replacement via right thoracotomy with a 31-mm Epic valve and removal of the

LEARNING OBJECTIVES

- To learn the etiology and epidemiology of primary and acquired pulmonary vein stenosis.
- To understand the diagnosis and treatment of pulmonary vein stenosis through multimodal imaging strategies and advanced percutaneous procedures.

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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

AF = atrial fibrillation

CCTA = cardiac computed tomographic angiography

CT = computed tomography

DOE = dyspnea on exertion

PV = pulmonary vein

PVL = paravalvular leak

PVS = pulmonary vein stenosis

RUPV = right upper pulmonary vein

TEE = transesophageal echocardiography

TTE = transthoracic echocardiography

V/Q = ventilation-perfusion

AVP2s. Several months later, he again reported DOE with TTE evidence of moderate PVL and was referred to our clinic for PVL closure.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes valvulopathy, coronary artery disease, heart failure, and anemia.

INVESTIGATIONS

Initial labs were notable for hemoglobin 8.2 g/dL, white blood cell count 5.22 × 10^3/μL, platelet count 117 × 10^3/μL, creatinine 0.97 mg/dL, and total bilirubin 0.7 mg/dL. TEE demonstrated a well seated bioprosthetic mitral valve with adequate leaflet excursion, and moderate PVL consisting of 2 jets (3-dimensional vena contracta areas 8 mm^2 and 14 mm^2) (Video 1). A severely stenosed right upper pulmonary vein (RUPV) with a peak gradient of 24 mm Hg and an inflow of 0.5 cm^2 (Video 2, **Figure 1**) was visualized. Cardiac computed tomographic angiography (CCTA) confirmed >70% proximal pulmonary vein stenosis (PVS) with vessel narrowing to a diameter of 3 × 2 mm (**Figure 2**). Given only moderate PVL, a ventilation-perfusion (V/Q) study was performed to determine the clinical significance of the PVS (**Figure 3**). It demonstrated 80% of total perfusion to the left lung, indicating considerable hypoperfusion of the right lung.

MANAGEMENT

The patient was admitted for percutaneous stenting of the RUPV and concurrent closure of mitral PVL via the right femoral vein. After transseptal puncture, the left atrial pressure was measured as 14 mm Hg. Percutaneous transluminal angioplasty of the RUPV was performed with a 5 × 40 mm Armada balloon, and a 10 × 29 mm Omnilink stent was deployed. Pulmonary vein angiography demonstrated an improvement in RUPV flow after stenting (Videos 3 and 4). Resolution of the pressure gradient was demonstrated by TEE and on catheter pullback from the RUPV into the left atrium. PVL closure of the larger, medial jet was performed with an 8-mm AVP2. After AVP2 placement, intraprocedural TEE demonstrated elimination of the medial jet (Video 5).

DISCUSSION

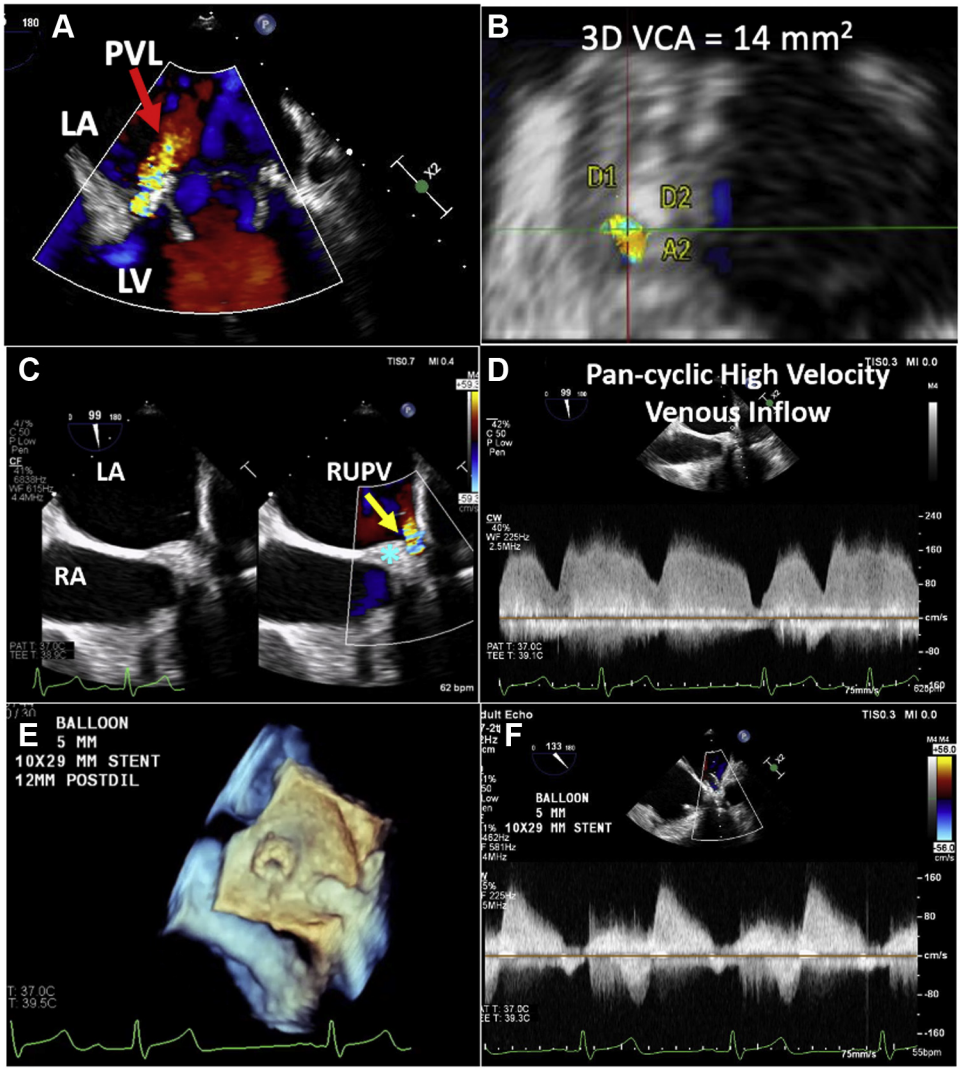
The diagnosis of PVS can be challenging owing to nonspecific symptomatology. Primary PVS is most commonly linked to congenital heart disease and developmental pathologies including total anomalous pulmonary venous return, septal defects, and transposition of the great vessels. Acquired PVS, however, is often secondary to neoplasms, inflammatory conditions, atrial fibrillation (AF) ablation, and cardiopulmonary bypass venting.[1] Acquired PVS has increased in incidence over the past two decades with increased performance of AF ablation.[2] In this population, the mechanism for vein obstruction seems to be inflammation and scarring from radiofrequency ablation. With surgical mitral valve replacement, the RUPV is a common site for cardiopulmonary bypass venting. On removal of the vent, the vein tissue is sutured, which can create a stenotic defect.[3] In our patient, this was likely the etiology of the RUPV stenosis along with the possible contribution of 3 previous distinct openings/closures of the atrioventricular groove.

PVS causes symptoms when the vein caliber is significantly stenosed (>50%) increasing lobar wedge pressure, or when the lung perfusion is decreased by more than 20%.[4] Both of these parameters were identified in our patient. Clinical manifestations can include progressive DOE, cough, pleuritic chest pain, and hemoptysis. TEE has emerged as a vital tool in the initial diagnosis, although a staged approach starting with TEE and adding CCTA has been shown to optimize fidelity by directly depicting vessel diameter.[5] In significant PVS, quantitative pulmonary flow V/Q scans can demonstrate perfusion defects of affected lung segments, providing the best correlation of anatomic pathology with resultant physiology.

In addition to RUPV stenosis, PVL likely contributed to our patient's clinical presentation. This is particularly true in an elderly patient with diastolic heart failure and likely reduced left atrial compliance after multiple open heart surgeries. On the other hand, the left atrial pressure was only mildly elevated before PVL closure. Because it was challenging to exclude that the PVL contributed to the patient's symptoms, we ultimately decided to treat the predominant PVL jet with a closure device. However, PVS remains the likely predominant etiology of our patient's symptoms owing to the marked abnormality of the V/Q scan. This case highlights the importance of considering PVS in the differential diagnosis of a patient with shortness of breath after cardiac surgery. It also underscores the importance and utility of multimodality testing in making the diagnosis of PVS.

Over the past decade the management of patients with PVS has primarily relied on transcatheter PV angioplasty with or without stenting.

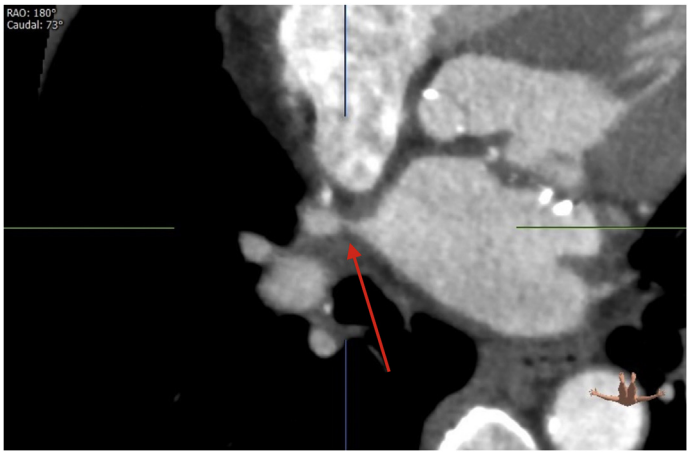
FIGURE 1 **Echocardiographic Images**



TEE demonstrating **(A)** mitral valve PVL with a **(B)** 3D VCA of 14 mm^2. **(C)** TEE demonstrating RUPV stenosis caused by surgical material **(blue asterisk)**. **(D)** Continuous-flow Doppler demonstrating high-velocity RUPV venous inflow with a peak gradient of 24 mm Hg and an inflow dimension of 0.5 cm^2. **(E)** TEE 3D reconstruction after RUPV stent placement with **(F)** continuous-flow Doppler demonstrating improvement in venous inflow. 3D = 3-dimensional; LA = left atrium; LV = left ventricle; RA = right atrium; TEE = transesophageal echocardiography; PVL = paravalvular leak; RUPV = right upper pulmonary vein; VCA = vena contracta area.

Polytetrafluoroethylene-covered stents have been used in congenital and acquired PV stenosis. Several studies have demonstrated that balloon-expandable covered stents may have better success in comparison to bare expandable stents.[6] Given increased rates of restenosis and reintervention as high as 40%, drug-eluting stents can be an attractive option as they can prevent intimal hyperplasia.[7] In cases where large-diameter stents can be implanted, as demonstrated in our patient, in-stent restenosis is less of a concern. Additionally, there have been several surgical procedures described to treat PVS, including reimplantation with direct anastomosis, stenotic excision, and "patchplasty."[8,9]

FIGURE 2 Diagnostic Cardiac Computed Tomography Angiography



Cardiac computed tomographic angiography demonstrating stenosis of the right upper pulmonary vein (**red arrow**).

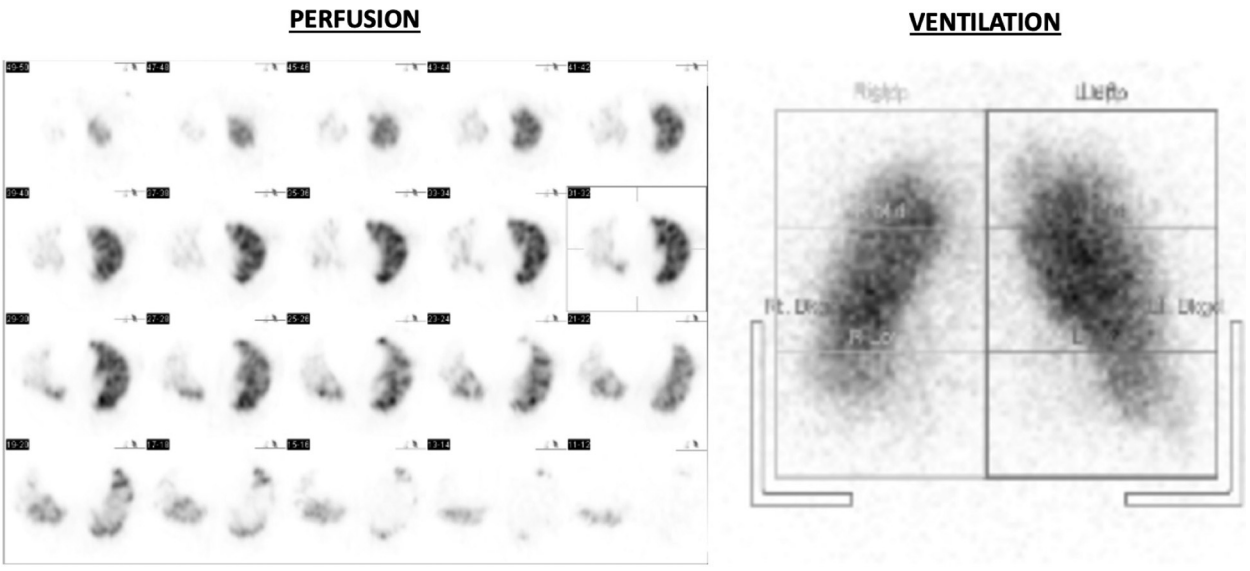
FOLLOW-UP

The patient had a notable improvement in dyspnea and exercise tolerance after pulmonary vein stent placement and PVL closure. CCTA and TTE at 90 days demonstrated a patent RUPV stent and trivial mitral valve PVL (Figure 4). Three years after the procedure, he has had no recurrence of symptoms.

CONCLUSIONS

Diagnosis of PVS requires a high level of clinical suspicion, but should be considered when evaluating shortness of breath in a post-cardiopulmonary bypass patient or a patient with multiple atrioventricular groove interventions. In addition to TEE, anatomic evaluation should also include CCTA, and V/Q scan should be considered to aid with clinical correlation. Finally, percutaneous placement of bare metal stents ≥10 mm provide a minimally invasive and effective treatment strategy for iatrogenic PVS.

FIGURE 3 Ventilation-Perfusion Scan



Ventilation-perfusion scan demonstrating perfusion defects of affected right lung segments.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr Hahn has received speaker fees from Abbott Structural, Edwards Lifesciences, and Philips Healthcare; has institutional educational and consulting contracts for which she receives no direct compensation with Abbott Structural, Boston Scientific, Edwards Lifesciences, and Medtronic; has equity with Navigate; and is the Chief Scientific Officer for the Echocardiography Core Laboratory at the Cardiovascular Research Foundation for multiple industry-sponsored trials, for which she receives no direct industry compensation. Dr Sommer has received institutional funding to Columbia University Irving Medical Center from Boston Scientific and W.l. Gore and Associates. Dr Vahl has received institutional funding to Columbia University Irving Medical Center from Boston Scientific, Edwards Lifesciences, JenaValve, Medtronic, and Siemens Healthineers; and has received personal consulting fees from Abbott Vascular, Boston Scientific, and Siemens Healthineers. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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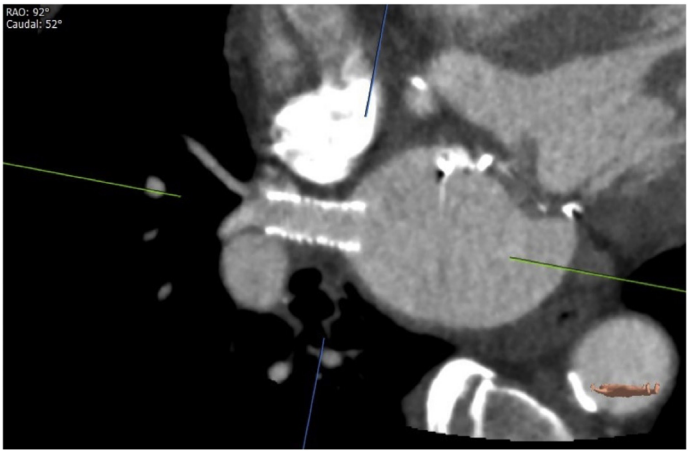
FIGURE 4 Post-Intervention Cardiac Computed Tomography Angiography



90-day postprocedural cardiac computed tomographic angiography demonstrating patent right upper pulmonary vein stent.

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KEY WORDS pulmonary vein stenosis, iatrogenic complication, paravalvular leak

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