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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Transient Severe Functional Mitral Valve Regurgitation

An Extremely Rare Cause of Recurrent Pulmonary Edema

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ABSTRACT

A 79-year-old woman presented with recurrent pulmonary edema. Extensive testing spanning 5 admissions showed only mild mitral regurgitation (MR). A transthoracic echocardiogram with the patient in the supine position and passive leg raise showed severe MR. This suggested transient severe MR. She underwent mitral valve replacement and had an uneventful postoperative course without recurrence of symptoms. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;16:101884) © 2023 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 79-year-old woman presented to the emergency department for substernal chest pain, diaphoresis, and shortness of breath for 3 days. This was 1 of her many hospitalizations for recurrent shortness of breath over 6 months. Her symptoms occurred when lying flat. She started to sleep upright to avoid symptoms. On initial presentation, her vital signs were remarkable for a blood pressure of 162/81 mm Hg, a heart rate of 77 beats/min, and a respiratory rate of 16 breaths/min on room air. Her physical examination was unremarkable, with normal breath sounds. Cardiac examination revealed normal rate and regular rhythm with no murmurs or gallops. Her lower extremities were without edema. Although this was her initial presentation, she had 4 more presentations with similar symptoms over a span of nearly 6 months.

LEARNING OBJECTIVES

- To better understand how to diagnose and manage transient severe MR.
- To understand the mechanism by which transient severe MR causes recurrent pulmonary edema, chest pain, and syncope.

PAST MEDICAL HISTORY

She had a history of hypertension and hyperlipidemia.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included acute coronary syndrome, heart failure, or flash pulmonary edema.

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ABBREVIATIONS AND ACRONYMS

ECG = electrocardiogram

LBBB = left bundle branch block

MR = mitral regurgitation

NIPPV = noninvasive positive pressure ventilation

TEE = transesophageal echocardiogram

TTE = transthoracic echocardiogram

INVESTIGATIONS

Given the patient's presenting symptoms, an extensive work-up was initiated. The complete blood count and basic metabolic panels were unremarkable. Her initial high-sensitivity troponin T value was 56 ng/L (reference, <14 ng/L), with a repeat value of 56 ng/L. The initial electrocardiogram (ECG) showed a new left bundle branch block (LBBB) (**Figure 1**). She experienced another episode of chest pain with shortness of breath and hypoxia associated with transient hypotension and worsening ST-segment and T-wave abnormalities (**Figure 2**). The initial transthoracic echocardiogram (TTE) showed a preserved ejection fraction of 65% without wall motion abnormalities and moderate mitral regurgitation (MR) (Videos 1 and 2). She underwent diuresis, with improvement in symptoms. Coronary angiography with hemodynamic assessment revealed no obstruction but demonstrated a myocardial bridge in the mid-left anterior descending artery. The hemodynamic assessment revealed normal biventricular filling pressures with a preserved cardiac index. Cardiac magnetic resonance revealed preserved biventricular function, no late gadolinium enhancement, and mild to moderate MR. Symptoms resolved, and she was discharged in stable condition with follow-up.

Unfortunately, she presented 4 hours after discharge with significant shortness of breath, chest pain, and hypotension. Initial examination revealed a holosystolic murmur. The ECG showed the known LBBB with no new changes. The initial chest radiograph showed pulmonary edema. She required noninvasive positive pressure ventilation (NIPPV) and underwent diuresis, with improvement in symptoms. She underwent a transesophageal echocardiogram (TEE), which showed only mild MR. A stress TTE showed a left ventricular outflow tract gradient of 5 mm Hg at rest and 10 mm Hg post-stress. The degree of MR was mild both at rest and after stress. She was discharged on as-needed diuretic therapy with follow-up.

She had recurrence of mild shortness of breath as an outpatient, but the condition resolved with as-needed diuretic therapy. She was re-evaluated for routine follow-up and was noted to have high blood pressure and episodic palpitations. She was started on diltiazem and underwent 14-day ambulatory cardiac monitoring, which showed asymptomatic paroxysmal episodes of atrial fibrillation and intermittent LBBB.

She presented 3 months later because of syncope and shortness of breath. The syncopal event was believed to be vasodepressor in nature, but given the intermittent LBBB, a high-grade atrioventricular block could not be excluded. She underwent dual-

FIGURE 1 Initial Electrocardiogram on Presentation

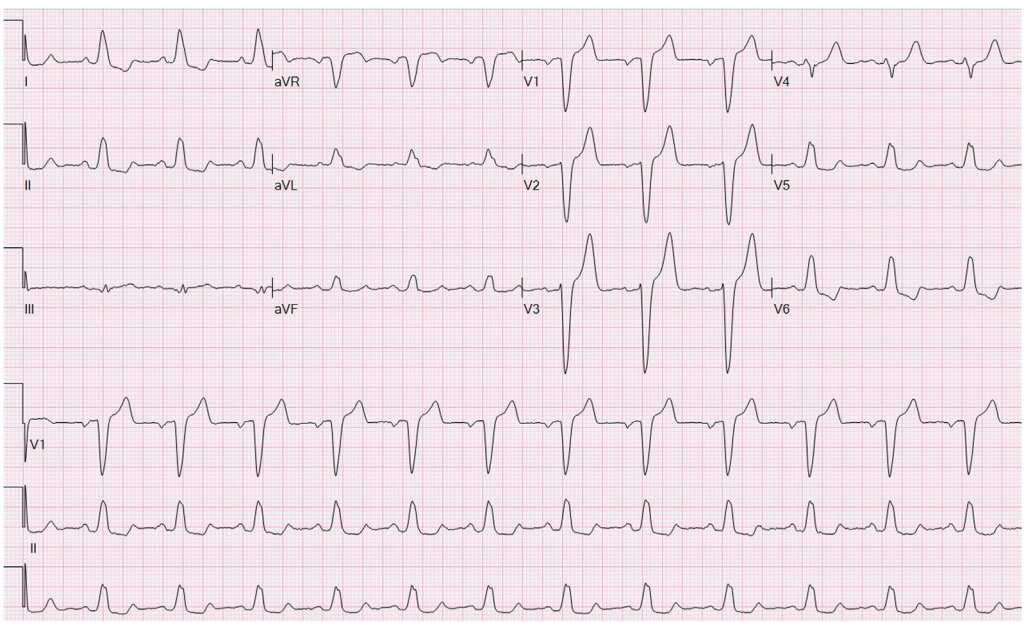
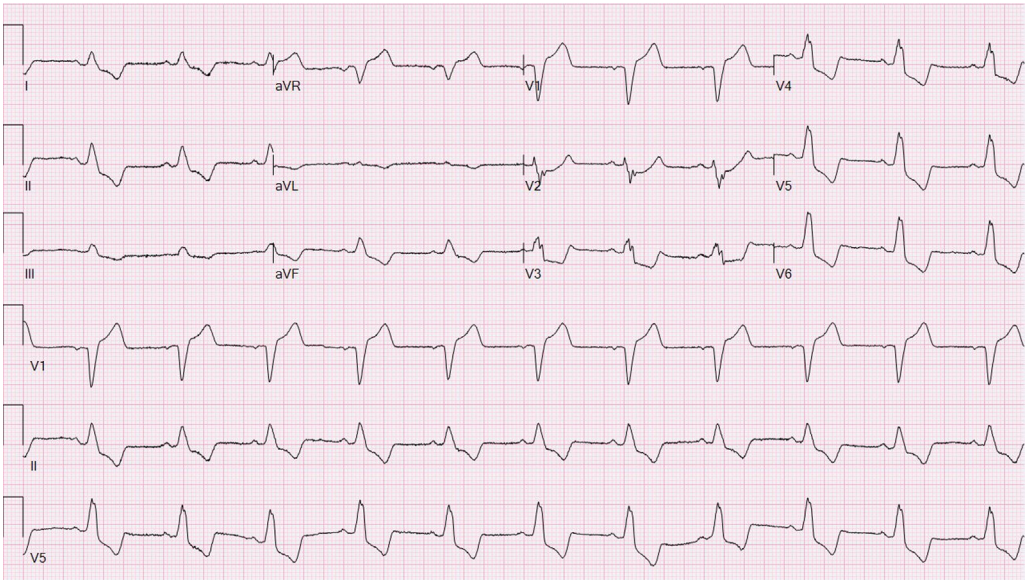


FIGURE 2 **Electrocardiographic Changes With Clinical Change**



chamber pacemaker implantation. However, she was readmitted a week later for syncope and shortness of breath despite the permanent pacemaker. The recurrence of symptoms prompted further work-up. Repeat coronary angiography was unremarkable once again. The working diagnosis was intermittent coronary vasospasm. She was discharged on verapamil, 40 mg twice daily.

A month later, she was admitted as a result of flash pulmonary edema requiring NIPPV and diuresis. The possibility of transient severe MR was considered. One of the proposed mechanisms of this rare disease is increased loading conditions causing worsening MR.[1] A bedside TTE was performed with positional changes and passive leg raise. The baseline TTE showed preserved biventricular function. In the upright position, MR was noted to be mild to moderate in severity with a normal pulmonary vein Doppler waveform (Video 3, **Figure 3**). The patient was placed supine, and a passive leg raise maneuver was performed. This resulted in loss of mitral valve leaflet coaptation and a broad jet of severe MR with systolic flow reversal in the right superior pulmonary vein (Video 4, **Figure 4**). Quantitatively, the effective regurgitant orifice was 0.6 cm^2, and the regurgitant volume was 121 mL (**Figure 5**). She became severely short of breath with hypoxia. Placing the patient in a seated position with NIPPV resolved her symptoms. The severity of MR also decreased to mild to moderate (Video 5). The MR was classified as secondary type 1 according to the Carpentier functional classification.

MANAGEMENT

Mitral valve replacement and repair were considered (surgical and transcatheter edge-to-edge repair), but because the mitral valve was morphologically normal during resting conditions on multimodality imaging, a repair would be of limited benefit. Moreover, patients described in the previous reports in the literature underwent valve replacement; therefore, this procedure was deemed to be the preferred option.[1] Intraoperative TEE confirmed worsening severity from mild to severe MR with passive leg raise while the patient was intubated (Videos 6 and 7). The patient underwent a 27-mm Mosaic porcine mitral valve replacement.

Epicardial left ventricular lead placement was also performed (without connection to the existing pacemaker generator) given the presence of intermittent LBBB in the event the patient's symptoms did not resolve with mitral valve replacement alone. Surgical pulmonary vein isolation and left atrial appendage excision with 4-0 polypropylene (Prolene, Ethicon) oversewn in 2 layers were performed for management of atrial fibrillation.

FIGURE 3 Right Superior Pulmonary Vein Doppler Echocardiogram Upright

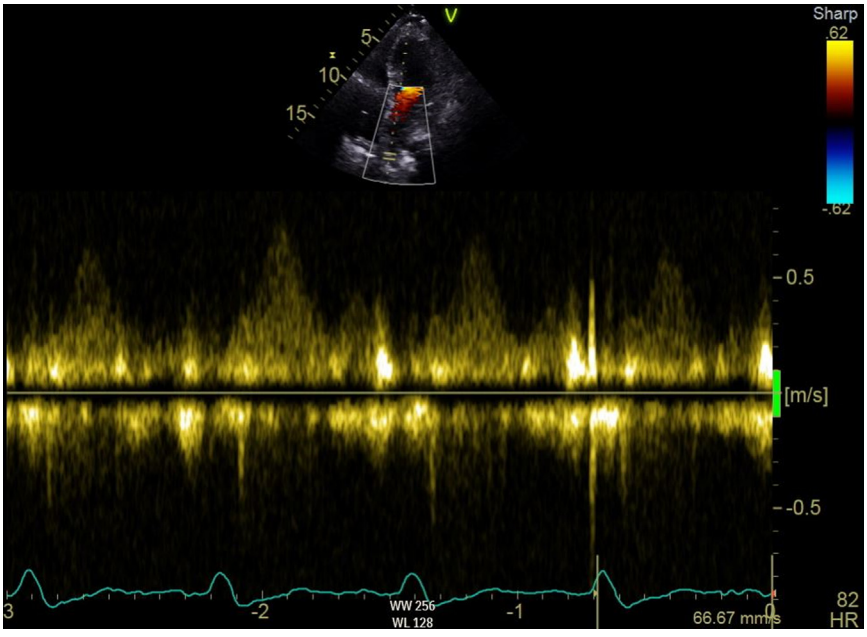


FIGURE 4 Right Superior Pulmonary Vein Doppler Echocardiogram After Leg Raise

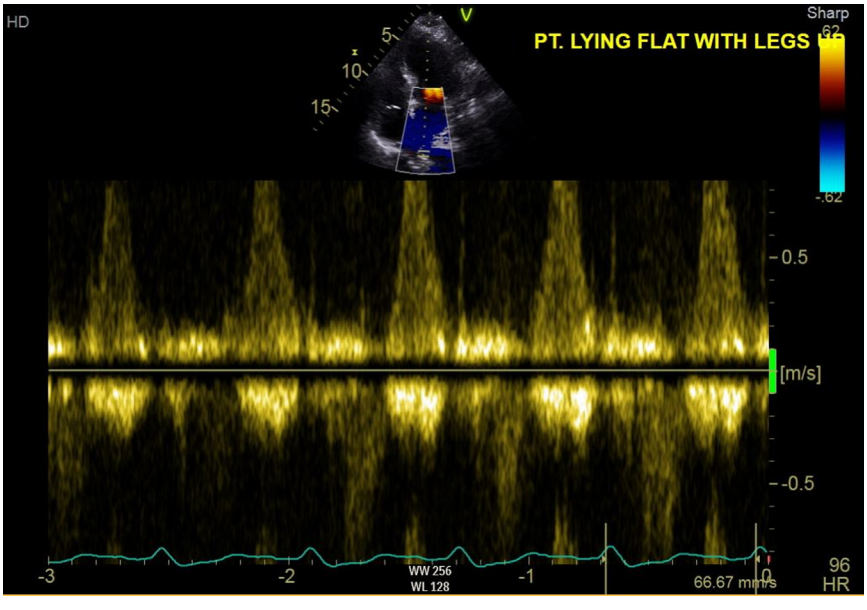
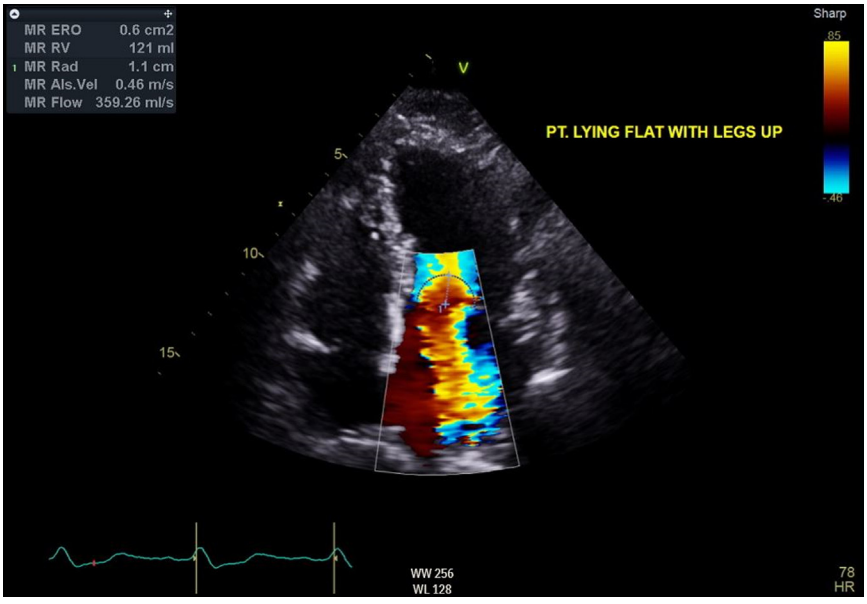


FIGURE 5 **Quantitative Assessment of the MR**



Als Vel = aliasing velocity; ERO = effective regurgitant orifice; MR = mitral regurgitation; Rad = radius; RV = regurgitant volume; other abbreviations as in Figure 4.

DISCUSSION

There are no guidelines to direct evaluation and management of this rare disease.[2] Current descriptions of transient MR are found in case reports and case series.[1,3] Patients with transient MR typically present with recurrent unexplained episodes of acute pulmonary edema in the setting of normal left ventricular function without coronary artery disease. Most patients in published case series were postmenopausal women.[1] Three types of transient MR were identified. Type 1 stemmed from a rate-dependent LBBB with an associated loss of left ventricular synchrony. Type 2 was reproduced by maneuvers that increased left-sided preload. Type 3 occurred with coronary vasospasm and was reproduced by the administration of methergine.[1,4] Our case is most like type 2, but it also demonstrated a unique positional component contributing to worsening symptomatic MR.

FOLLOW-UP

The patient's postoperative course was unremarkable. She was discharged home shortly after surgery. She has had no recurrent hospitalizations and reported complete resolution of symptoms. A follow-up TTE 1 month after surgery showed a well-seated bioprosthetic mitral valve without stenosis or regurgitation. The mean gradient was 7.3 mm Hg at 80 beats/min.

CONCLUSIONS

Transient severe MR is a challenging clinical entity to diagnose and can affect an individual's quality of life. It is important for physicians to consider transient MR in patients with recurrent unexplained dyspnea. Provocative echocardiographic assessments can help with early identification and disease management.

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KEY WORDS positional mitral regurgitation, recurrent pulmonary edema, transient mitral regurgitation

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