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VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

When Is Mitral Regurgitation Not Just Systolic?

A Case of Near-Continuous Mitral Regurgitation

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ABSTRACT

A 30-year-old man presented with symptomatic severe chronic aortic regurgitation, first-degree atrioventricular block, and near-continuous mitral regurgitation. Surgical intervention, including aortic root replacement and mitral valve repair, was successful, highlighting the close relationship between valvular diseases and emphasizing the importance of comprehensive assessment for optimal management. (JACC Case Rep. 2024;29:102533) © 2024 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 30-year-old male presented to the emergency department, Cleveland Clinic, Abu Dhabi with a 4-week history of chest pain, cough, and dyspnea. On examination, his heart rate was 90 beats/min, with wide pulse pressure of 104/45 mm Hg, elevated jugular venous pressure, bibasilar crackles, and a blowing 3/6 early diastolic decrescendo murmur at the left third intercostal space and a soft long systolic murmur at the apex. There were no characteristics to suggest Marfan syndrome or other connective tissue disorders.

TAKE-HOME MESSAGES

- The case highlights a rare situation where chronic severe AR combined with significant first-degree AV block led to near-continuous MR.
- It emphasizes the importance of prompt diagnosis and comprehensive management, including surgical intervention when possible, as well as exploring AV synchrony optimization to potentially improve heart function and future surgical outcomes.

PAST MEDICAL HISTORY

The patient denied any significant past medical history, allergies, or medication use.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included aortic regurgitation (AR). The early diastolic decrescendo murmur with wide pulse pressure was suggestive of severe AR. Although infective endocarditis can manifest with acute valvular regurgitation with nonspecific symptoms, the absence of such symptoms and signs

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ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

AV = atrioventricular

TEE = transesophageal echocardiogram

CTA = computed tomography angiography

LV = left ventricular

MR = mitral regurgitation

PMVC = premature mitral valve closure

on examination makes it less likely. Finally, a connective tissue disease such as Marfan syndrome can lead to aortic root dilation and subsequent AR.

INVESTIGATIONS

The electocardiogram revealed sinus rhythm with first-degree atrioventricular (AV) block and poor R-wave progression (**Figure 1**). Laboratory tests revealed mildly elevated troponin, mild renal impairment, and elevated B-type natriuretic peptide.

The transthoracic echocardiogram revealed severe AR with holodiastolic flow reversal in the abdominal aorta (**Figure 2**), a severely dilated left ventricle with a moderately reduced ejection fraction, moderate mitral regurgitation (MR), and moderate pulmonary hypertension.

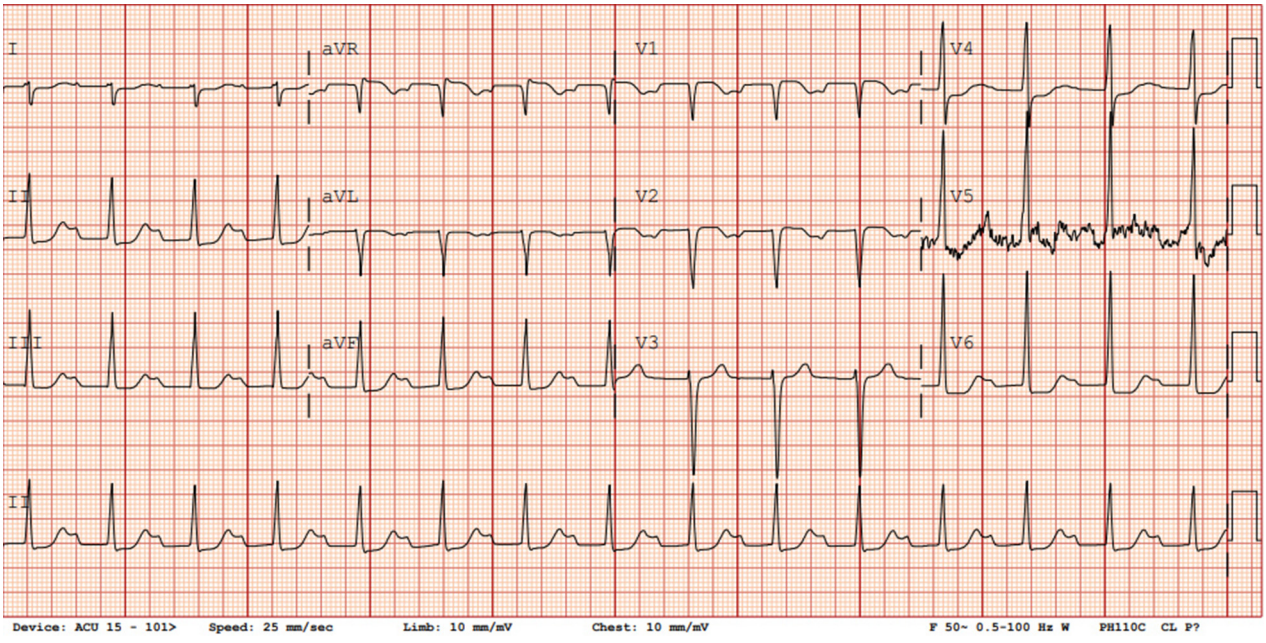
The transesophageal echocardiogram (TEE) confirmed chronic left ventricular (LV) enlargement with moderate to severe LV systolic dysfunction (Video 1), severe AR secondary to aneurysmal dilatation of the sinus of Valsalva (mainly left coronary sinus) (**Figure 3**, Videos 2 to 4), near-continuous MR (**Figures 4 and 5**, Videos 5 and 6) with exaggerated premature mitral valve closure (PMVC) (**Figure 6**), systolic blunting of all pulmonary veins (**Figure 7**), and moderate pulmonary hypertension.

The patient underwent computed tomography angiography (CTA) of the chest (**Figures 8, 9A, and 9B**), which confirmed aneurysmal dilatation of the aortic root and identified a narrow-neck pseudoaneurysm communicating with the left coronary sinus.

MANAGEMENT

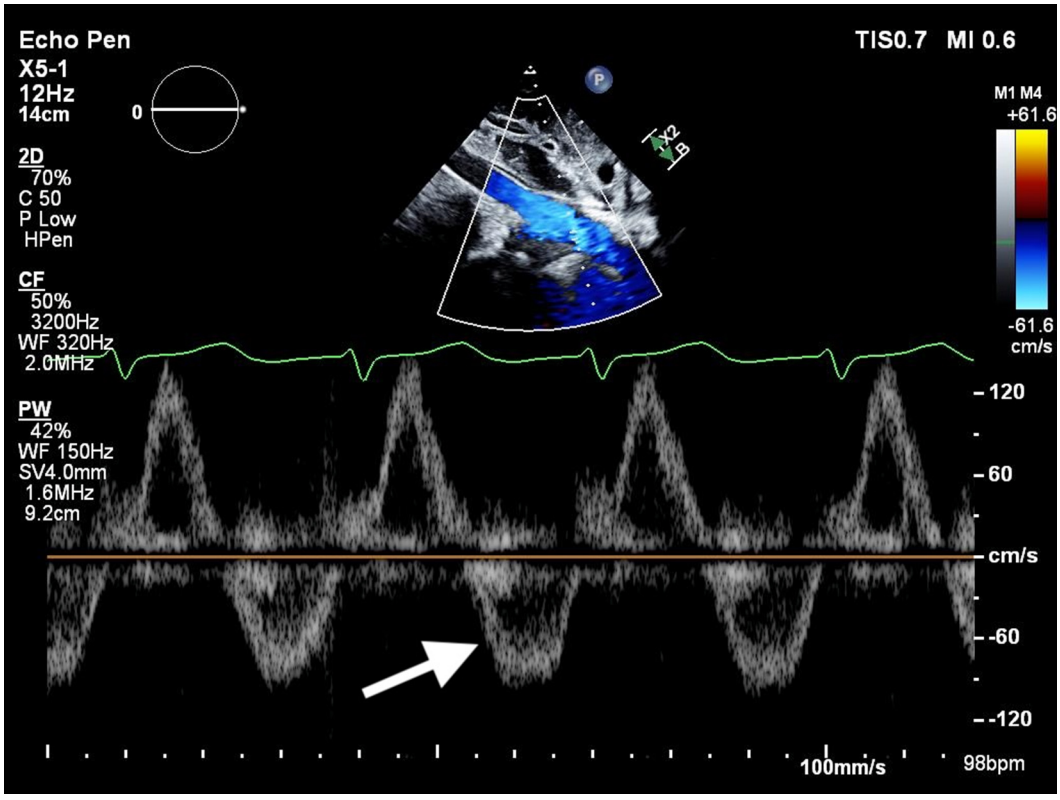
The patient was initially managed in the intensive care unit with intravenous diuretic agents. Intravenous dobutamine was initiated to treat his heart failure and to improve cardiac output and reduce afterload. Careful monitoring of fluid balance, renal function, and electrolyte levels was performed. After exclusion of underlying infection, the patient underwent successful aortic root replacement using a composite prosthesis, and mitral valve repair was performed. The pathology report revealed

FIGURE 1 Electrocardiogram



Electrocardiogram showing sinus rhythm, first-degree atrioventricular block, and poor R-wave progression in the anteroseptal leads.

FIGURE 2 **Pulsed-Wave Doppler Imaging of the Abdominal Aorta**



Significant holodiastolic flow reversal (arrow) noted in the pulse wave Doppler image of the abdominal aorta. CF = color flow; PW = pulsed wave; 2D = 2-dimensional.

inflammatory changes and fibromyxoid valvular degeneration of the aortic valve but no vegetations.

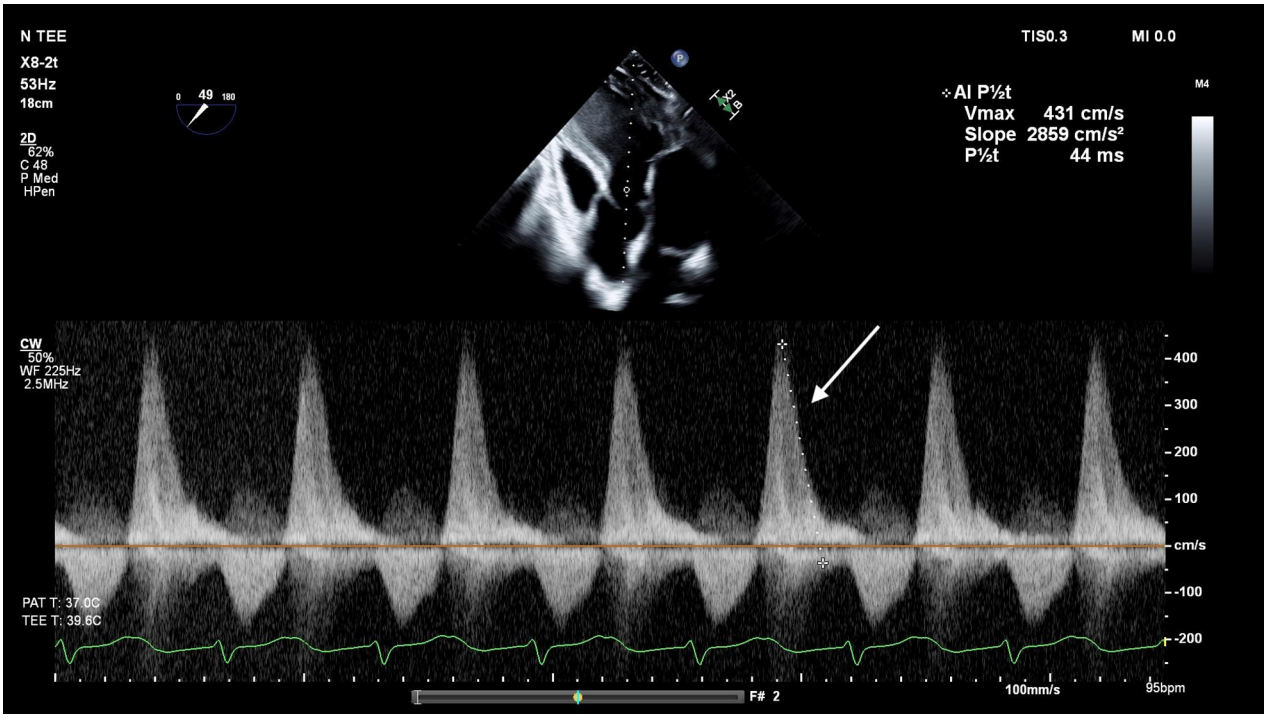
The patient had an uneventful hospital course and was discharged home 1 week postoperatively. He returned for an outpatient follow up appointment 2 weeks later and was doing remarkably well.

DISCUSSION

This case demonstrates the complex relationship between severe chronic AR and the development of near-continuous MR when combined with AV block. Echocardiographic findings revealed severe AR with aneurysmal dilatation of the aortic root and near-continuous MR caused by PMVC, thereby suggesting that a series of hemodynamic events led to significant left LV dysfunction. Acute AR typically causes diastolic MR resulting from a rapid rise in LV diastolic pressure, but the chronicity of AR in this case presents a unique scenario.[1,2]

The near-continuous nature of the MR in this patient is highly significant. Although diastolic MR is often associated with elevated LV filling pressures or AV dyssynchrony, such as in AV blocks of any degree, or the presence of restrictive cardiomyopathy or severe AR,[2-4] the marked PMVC observed here, driven by severe AR alone, would not typically lead to significant diastolic MR, especially in a long-term setting. However, when combined with significant first-degree AV block, it can result in striking, near-continuous MR, as seen in this case. The presence of first-degree AV block in this patient further

FIGURE 3 Continuous Wave Doppler Imaging of the Aortic Valve



The continuous wave Doppler image of the aortic valve from the deep gastric view demonstrating severe aortic regurgitation (arrow) with a steep deceleration slope. TEE = transesophageal echocardiography; other abbreviations as in Figure 2.

exacerbated the diastolic MR by impairing LV filling and increasing filling pressures.

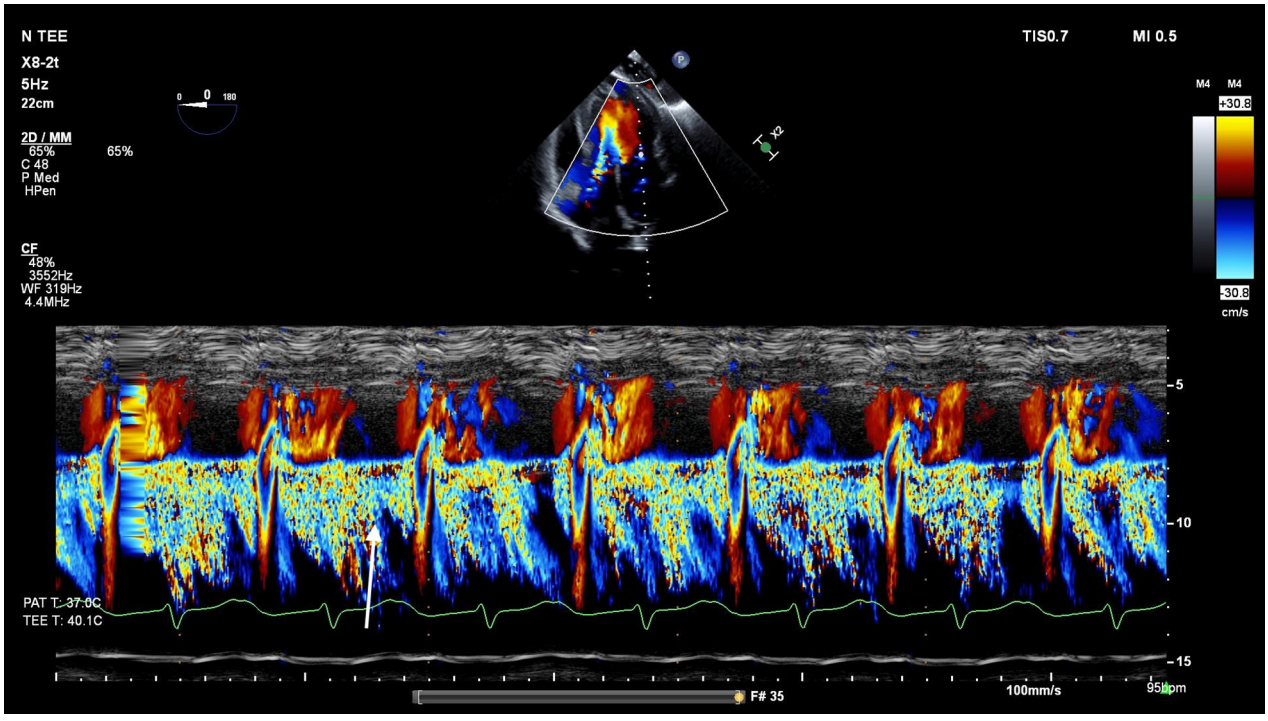
Although diastolic MR secondary to AV block alone is often harmless, near-continuous MR with severe LV dysfunction and AR, as seen in this case, stresses the importance of maintaining proper AV synchrony for optimal LV filling. Albeit not used in this case, dual-chamber pacing at a shorter AV interval in patients with significant first-degree AV block and severe AR theoretically could improve LV filling dynamics by optimizing the timing of atrial and ventricular contractions, extending the effective filling period, and potentially reducing or eliminating diastolic MR.[4]

The successful surgical intervention, involving aortic root and mitral valve repair, demonstrates the importance of a comprehensive approach to valvular heart disease. The use of advanced imaging modalities, such as TEE and CTA, was integral in determining the valvular anatomy and guiding surgical planning. The patient's uneventful postoperative course and subsequent improvement in cardiac function highlight the potential for excellent outcomes in young patients with severe valvular disease when diagnosed and managed promptly.

CONCLUSIONS

This case highlights the rare but significant association of near-continuous MR with severe chronic AR and significant first-degree AV block. These factors together can lead to severe LV dysfunction, thus emphasizing the need for prompt recognition and comprehensive management. The successful surgical intervention in this young patient illustrates the potential for excellent outcomes when a

FIGURE 4 Transgastric Color M-Mode Imaging



Color M-mode recording of flow through the mitral valve from a transgastric view. Mitral regurgitation is evident throughout the majority of the cardiac cycle, including most of diastole (arrow) and systole. Abbreviations as in Figures 2 and 3.

multidisciplinary approach is used, incorporating advanced imaging and timely surgical repair. Optimizing AV synchrony, although not used here, potentially through interventions such as dual-chamber pacing, may be considered in patients with severe valvular disease who are not immediate surgical candidates, to improve their overall cardiac function and potentially enhance surgical outcomes when surgery becomes feasible.

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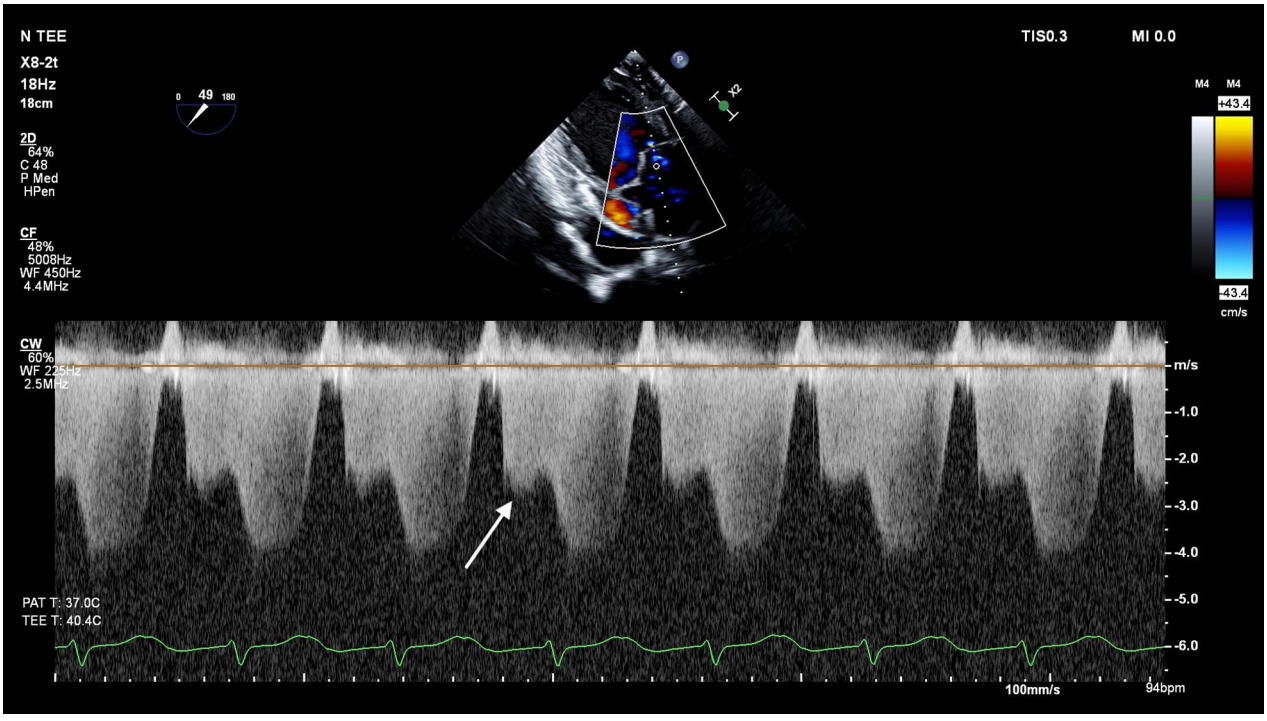
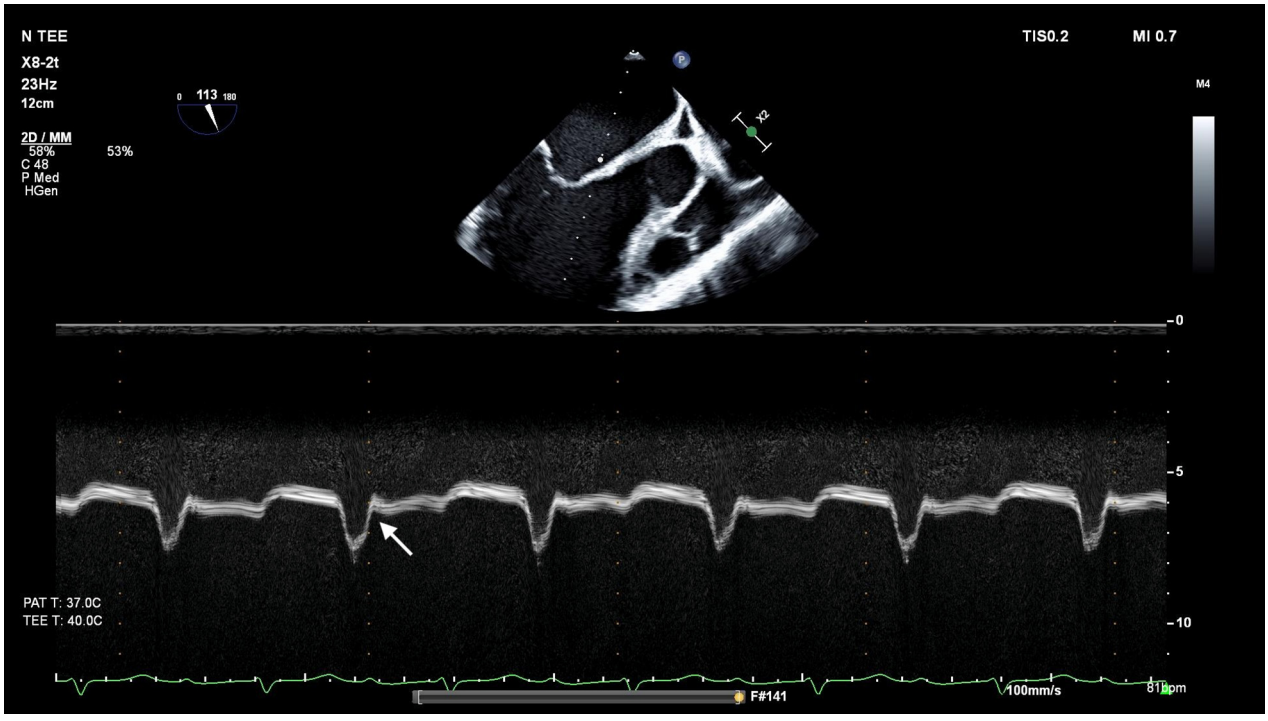
FIGURE 5 **Transmitral Continuous Wave Doppler Imaging**



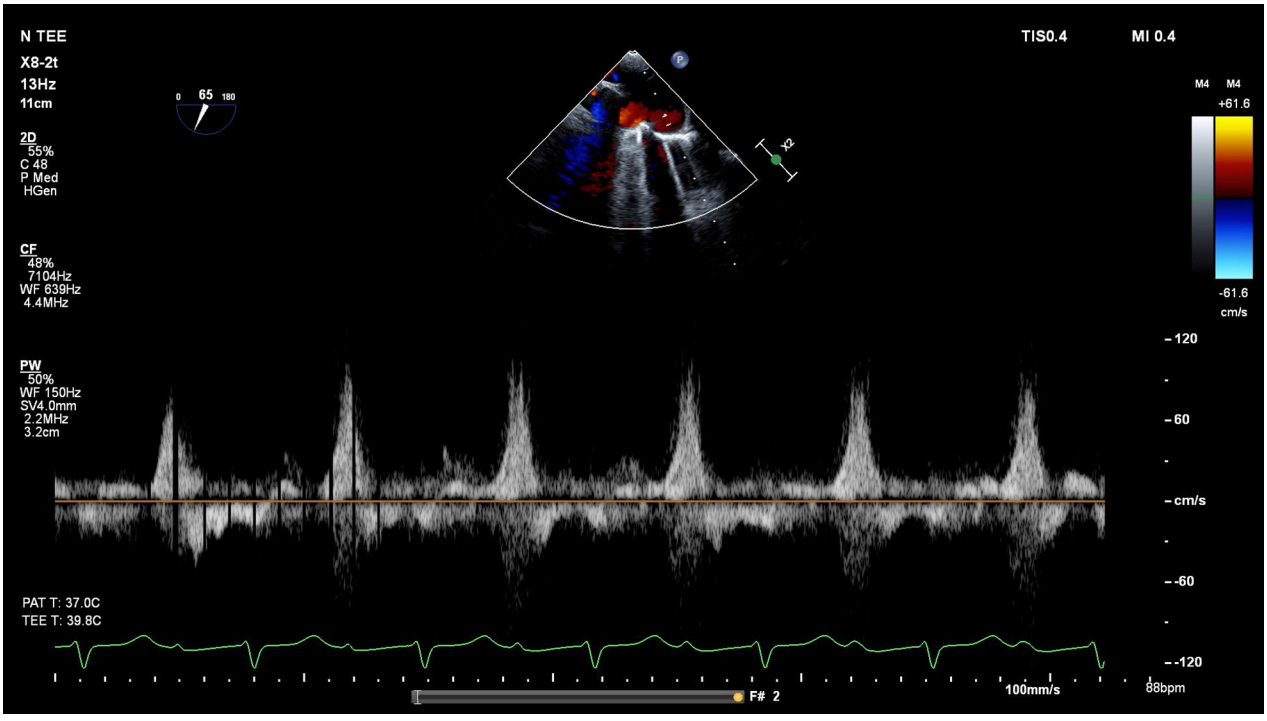
Continuous wave Doppler recording of transmitral flow from a transgastric view. A slightly lower-velocity signal of diastolic mitral regurgitation (arrow) precedes systolic regurgitant signal. Abbreviations as in Figures 2 and 3.

FIGURE 6 **M-Mode Mitral Valve Imaging**



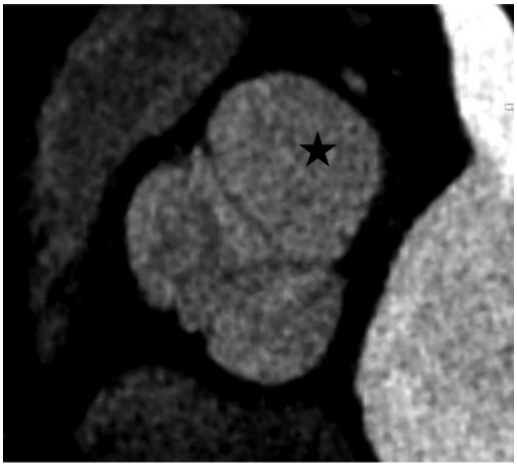
M-Mode echocardiogram of the mitral valve demonstrating significant premature mitral valve closure (arrow). Abbreviations as in Figures 2 and 3.

FIGURE 7 Pulmonary Vein Pulsed Wave Doppler Imaging



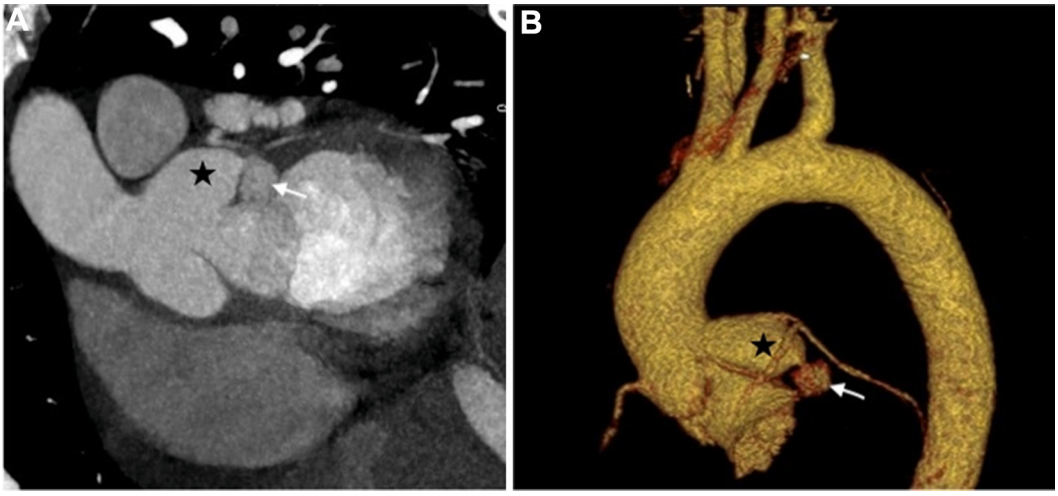
Pulsed wave Doppler of pulmonary venous flow patterns showing blunted systolic flow in all 4 pulmonary veins. Abbreviations as in Figures 2 and 3.

FIGURE 8 Computed Tomography Angiography



Contrast-enhanced gated computed tomography angiography in a zoomed short-axis view at the level of the sinus of Valsalva demonstrates aneurysmal dilatation of the left coronary sinus (star) compared with the other 2 coronary sinuses.

FIGURE 9 **Contrast-Enhanced 2-Chamber View**



(A) Contrast-enhanced 2-chamber view cardiac image and (B) volume rendering reconstructed image illustrate aneurysmal dilatation of the left coronary sinus compared with the right (star). There is narrow neck inferior outpouching (arrow) communicating with the left sinus compatible with pseudoaneurysm.

REFERENCES

1. Downes TR, Nomeir AM, Hackshaw BT, Kellam LJ, Watts LE, Little WC. Diastolic mitral regurgitation in acute but not chronic aortic regurgitation: implications regarding the mechanism of mitral closure. *Am Heart J*. 1989;117(5):1106-1112.

2. Lancellotti P, Tribouilloy C, Hagendorff A, et al, European Association of Echocardiography. European Association of Echocardiography recommendations for the assessment of valvular regurgitation. Part 1: aortic and pulmonary regurgitation (native valve disease). *Eur J Echocardiogr*. 2010;11(3):223-244.

3. Rajesh G, Raju D, Krishnan MN. 'Continuously regurgitating mitral valve': systolic and diastolic mitral regurgitation in a case of severe aortic regurgitation and complete heart block. *Heart Asia*. 2013;5(1):172-173.

4. Agmon Y, Freeman WK, Oh JK, Seward JB. Diastolic mitral regurgitation. *Circulation*. 1999;99(21):e13.

KEY WORDS aortic valve, electrophysiology, mitral valve

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