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CASE REPORT

INTERMEDIATE

CLINICAL CASE

# When Hindsight Is 20/20

## Stiff Left Atrium Syndrome Masquerading as Mitral Regurgitation

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**ABSTRACT**

Scarring from atrial ablation carries a risk of developing stiff left atrium syndrome, which can mimic mitral valve disease. We present a case of a 73-year-old man whose stiff left atrium syndrome came to light after percutaneous mitral valve repair. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:648–51) © 2019 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

In February 2018, a 73-year-old man presented with mitral regurgitation (MR) and New York Heart Association functional class III dyspnea. On examination, his chest was clear to auscultation and jugular venous pressure elevated to 7 cm above the sternal angle. Cardiac auscultation revealed a loud P2 and a grade 2/6 pansystolic murmur at the apex and left lower sternal border with no S3. Moderate bilateral lower extremity pitting edema was noted with no hepatosplenomegaly.

### LEARNING OBJECTIVES

- Physicians should be able to recognize the presentation of SLAS.
- Physicians should be able to form a differential diagnosis of exertional dyspnea after atrial ablation.

### MEDICAL HISTORY

The patient had a dual-chamber pacemaker inserted for sick sinus syndrome in 2009. He subsequently developed recurrent symptomatic paroxysmal atrial fibrillation (AF) and flutter. After failing to improve with medical therapy in June 2015, he underwent cryoballoon catheter ablation followed by endocardial radiofrequency (RF) ablation in July 2016 with pulmonary vein isolation as the first portion of a hybrid convergent ablation procedure. In August 2016, he underwent pericardioscopic epicardial RF ablation of the left atrium with pulmonary vein isolation. For this, 32 RF lesions were created and the left atrial appendage ligated. Despite this and multiple trials of oral antiarrhythmic agents, symptomatic atrial flutter with rapid ventricular rates recurred. In March 2017, the patient underwent a final RF ablation. In total, he underwent 4 AF ablation procedures over 3 years. Transesophageal echocardiography performed at the time of his last ablation revealed moderately severe (3+) MR and a large (1.1 cm) atrial septal defect (ASD)

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associated with a pulmonary-to-systemic flow ratio (Qp:Qs) of 1.5, suggesting significant left-to-right shunting.

## DIFFERENTIAL DIAGNOSIS

The differential diagnosis included pulmonary hypertension secondary to: MR, pulmonary vein stenosis, stiff left atrial syndrome (SLAS), and ASD.

## INVESTIGATIONS

In 2017, the patient underwent transcatheter ASD closure using a 35-mm Amplatzer PFO Occluder (St. Jude Medical, Saint Paul, Minnesota), with no symptomatic benefit. A cardiac computed tomography scan in May 2017 revealed severe biatrial enlargement with no pulmonary vein stenosis. Subsequent right and left heart catheterization revealed the following hemodynamic values: mean pulmonary capillary wedge pressure (PCWP) of 40 mm Hg (V waves to 45 mm Hg), pulmonary artery pressure of 100/37 mm Hg (mean 60 mm Hg), and left ventricular (LV) end-diastolic pressure of 23 mm Hg. LV angiography revealed 3+ MR with patent coronary arteries. Trans-esophageal echocardiography in October 2017 revealed an LV ejection fraction of 65%, two separate moderate jets of MR, with MR radii of 0.8 and 0.7 cm, respectively (**Figure 1A**, Video 1). The left upper pulmonary vein showed mild systolic flow reversal, and the right upper pulmonary vein showed systolic blunting. Repeat heart catheterization (December 2017) confirmed patent coronary arteries with normal LV systolic function and 3+ MR. The pulmonary artery pressure was markedly elevated at 86/20 mm Hg, and mean PCWP was 30 mm Hg with V waves to 35 mm Hg.

## MANAGEMENT

The etiology of the MR was deemed to be primary or at least "mixed" and not purely functional. The patient underwent percutaneous MitraClip (Abbott, Abbott Park, Illinois) repair of the MR. Trans-septal puncture was performed adjacent (superior and posterior) to the previous ASD closure. His initial mean left atrial (LA) pressure was 31 mm Hg with V waves to 49 mm Hg. A single MitraClip was placed across the A2/P2 mitral coaptation point, reducing the MR to trivial (1+) (**Figure 1B**, Video 2). Despite near elimination of MR, the patient's LA pressure and pulmonary vein inflow pattern remained unaltered. Right heart catheterization was repeated 1-day following the MitraClip and PCWP procedures, right atrial and LV pressures recorded at baseline, and after administration of intravenous dopamine, intra-arterial nitroglycerin, and inhaled nitric oxide (**Figure 2**). Mean PCWP and V waves were elevated at rest, increased significantly with dopamine, and decreased markedly with nitroglycerin. Nitric oxide had little effect on PCWP but resulted in a further reduction in right atrial pressure compared with nitroglycerin.

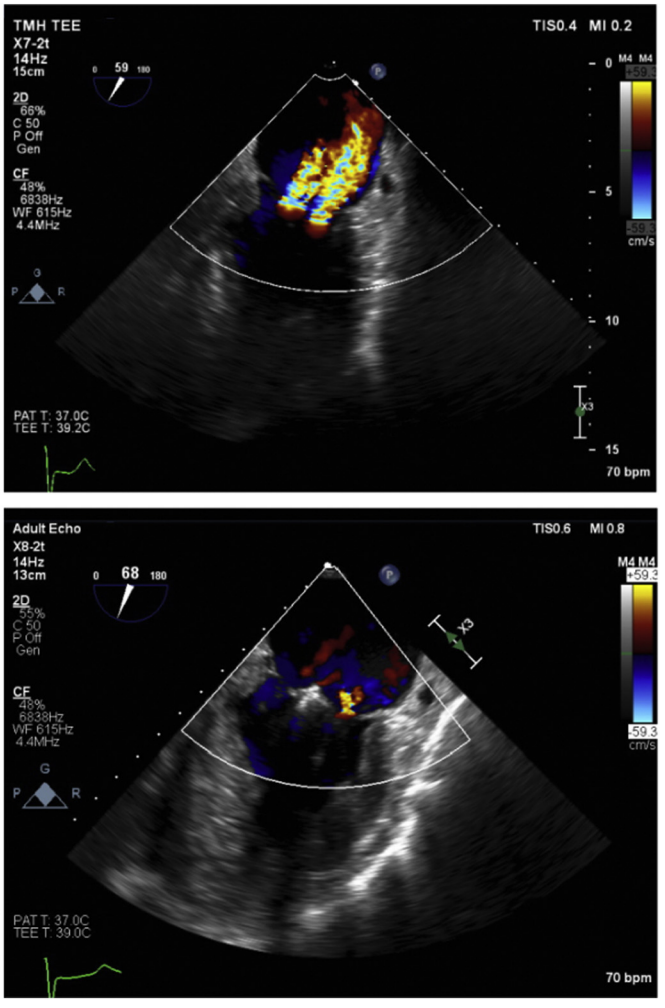
## DISCUSSION

During LV systole, the left atrium serves as a reservoir for pulmonary venous return and throughout LV diastole as a conduit from the

### ABBREVIATIONS AND ACRONYMS

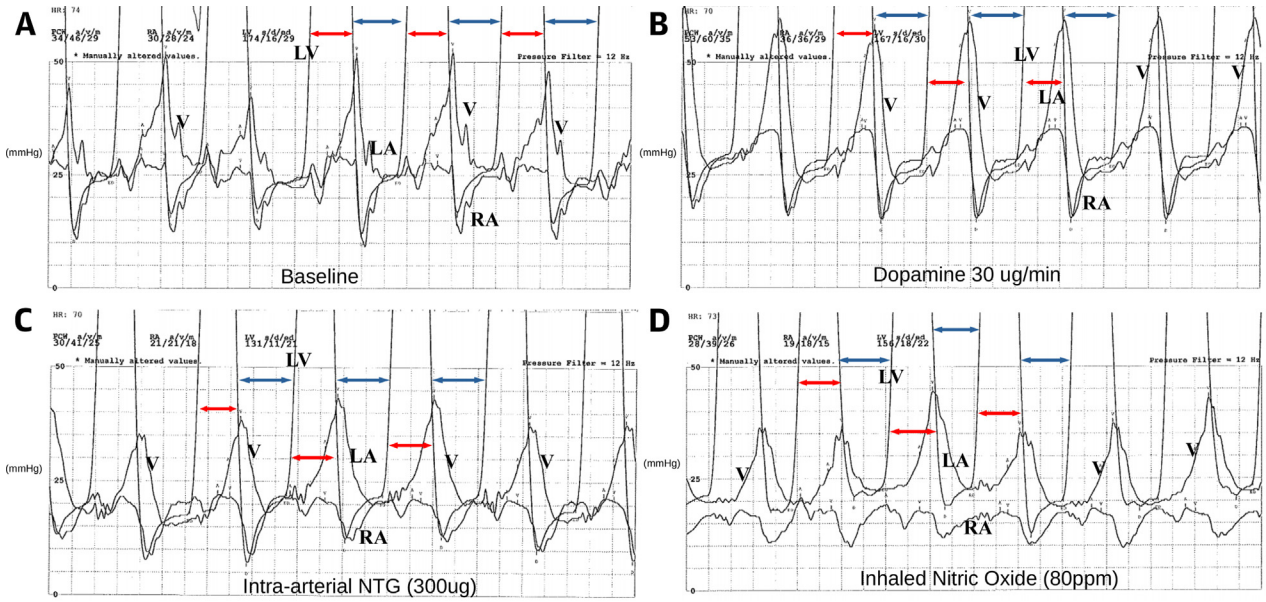
**AF** = atrial fibrillation
**ASD** = atrial septal defect
**LA** = left atrial
**LV** = left ventricular
**MR** = mitral regurgitation
**PCWP** = pulmonary capillary wedge pressure
**RF** = radiofrequency
**SLAS** = stiff left atrial syndrome

**FIGURE 1** Mitral Valve Color Doppler




Color Doppler of mitral valve and pulmonary vein inflow before **(A)** and after **(B)** MitraClip mitral valve repair. See Videos 1 and 2.

**FIGURE 2 Right Heart Catheterization Pressure Readings**




Pressure waveforms depicting left ventricular (LV), left atrial (LA), and right atrial (RA) pressures at **(A)** baseline and following administration of **(B)** intravenous dopamine, **(C)** intra-arterial nitroglycerin (NTG), and **(D)** inhaled nitric oxide. The **red arrows** indicate ventricular systole, and the **blue arrows** represent ventricular diastole. The LA pressure and its corresponding V waves (V) increased with intravenous dopamine and decreased with intra-arterial NTG. Inhaled nitric oxide had minimal effect on LA pressure but significantly decreased RA pressure.

pulmonary veins to the left ventricle (1). Not only does LA contraction augment end-diastolic LV filling, the left atrium itself serves as an important modulator of neurohormonal feedback systems that are activated by LA stretch (2). In most healthy adults, a loss of atrial contraction is tolerated with minimal symptoms, especially if heart rate is controlled. In older individuals, particularly those with a reduction in ventricular compliance, atrial contraction may be responsible for up to 30% of ventricular preload (3), and its loss is poorly tolerated. During LA relaxation, LA pressure initially decreases (x-descent), the mitral valve closes, and the atrium refills from both right ventricular contraction and passive filling from the pulmonary veins. This action generates an increase in LA pressure (V-wave). Patients with reduced LA compliance often have an accentuated V-wave due to the atrium's inability to accommodate atrial filling. Similarly, in a patient with significant MR, the additional regurgitant volume from the left ventricle increases the amplitude of the V wave.

Post-ablation pulmonary hypertension occurs in ~8% of cases and may be due to pulmonary vein stenosis, SLAS, and/or LV diastolic dysfunction unmasked by the procedure (4). Because our patient had both LA appendage and ASD closure devices, it is feasible that these devices may also have reduced atrial compliance. However, we consider this unlikely as recent studies show little remodeling or hemodynamic impact of LA appendage closure (5), and the deleterious effects of transcatheter ASD closure have been shown to resolve by 6 months' post-procedure (6).

The patient's dyspnea was potentially attributable to several etiologies. Hemodynamic values suggested pulmonary hypertension secondary to a left heart origin. The findings of elevated PCWP with prominent V waves and 3+ MR led to the initial impression that the patient's MR was a major contributor. However, his V waves were timed slightly later in diastole than typical of MR, did not resolve with valve repair, and his left ventricular end-diastolic pressure was lower than expected for severe MR. In retrospect,

SLAS rather than MR was the likely cause of our patient's presenting dyspnea and abnormal hemodynamic values. The final diagnosis of SLAS rested on the persistence of elevated LA pressure and prominent V waves, despite effective MR reduction. With MR, the pulmonary vein inflow pattern typically shows a late decrease in the S-wave on Doppler. In contrast, our patient exhibited early S-wave reversal, which is more consistent with reduced LA compliance. Finally, mitral valve repair did not alter early S-wave reversal. Similar to mitral stenosis, the pathogenesis of dyspnea from SLAS is pulmonary congestion due to elevated LA pressure. A longer diastolic filling time resulting from beta-blockers in combination with reduced LA preload brought on by nitrates and diuretics ultimately translated into symptomatic improvement in our patient.

Our case has some similarities to another noted following multiple AF ablations reported by Amjad et al. (7). As recently suggested by Packer (8), these cases remind us that the benefits of multiple catheter ablations for AF must be carefully weighed against potential deleterious effects, including impairment of the left atrium's reservoir, conduit, and transport functions.

## FOLLOW-UP

The patient was discharged on high-dose beta-blockers, diuretics, and long-acting oral nitrates. His dyspnea improved from New York Heart Association functional class III to I/II. He has since fared well on medical therapy with no further episodes of heart failure and/or hospital readmission.

## CONCLUSIONS

The prevalence of SLAS after AF ablation is unknown. Research by Gibson et al. (9) provides some insight into the epidemiology, but the condition is likely underdiagnosed as only persistently symptomatic patients undergo the comprehensive evaluation necessary for diagnosis. As AF ablation becomes more widely adopted, physicians need to be aware of this rare complication, its associated hemodynamic parameters, and optimal pharmacological therapies.

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**KEY WORDS** ablation, atrial fibrillation, mitral valve, pulmonary hypertension, valve repair

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