

# Iatrogenic atrial septal defect with right-to-left shunt following atrial fibrillation ablation in a patient with arrhythmogenic right ventricular cardiomyopathy



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## Introduction

Dyspnea following catheter ablation for atrial fibrillation (AF) is a symptom that always requires further investigation. The most common causes acutely include pulmonary congestion from volume overload, aspiration, and, less commonly, phrenic nerve injury.<sup>1</sup> Other uncommon complications that can occur subacutely include pulmonary embolism, pulmonary vein stenosis,<sup>2</sup> and stiff left atrial syndrome.<sup>3</sup> We describe a rare case of acute dyspnea and hypoxemia owing to a persistent right-to-left shunt across a residual iatrogenic atrial septal defect (ASD) in a patient with arrhythmogenic right ventricular cardiomyopathy (ARVC).

## Case report

A 71-year-old man presented for catheter ablation of symptomatic persistent AF. He had a history of ARVC requiring an implantable cardioverter-defibrillator and 2 prior ablation procedures for sustained ventricular tachycardia.

Although he was 2 years free of ventricular tachycardia following his last ablation procedure, he remained highly symptomatic from recurrent persistent AF. Direct current cardioversion resulted in symptomatic improvement, but sinus rhythm was unable to be maintained despite sotalol therapy, and amiodarone was poorly tolerated. He agreed to undergo AF ablation in order to achieve durable symptomatic relief.

His preprocedural transthoracic echocardiogram (TTE) demonstrated normal left ventricular size and systolic function (left ventricular ejection fraction 65%) and a severely enlarged right ventricle (RV) with severely impaired systolic function. There was severe tricuspid

regurgitation (TR) owing to annular dilatation and failure of leaflet coaptation. The right atrium (RA) was severely dilated with leftward bowing of the interatrial septum.

The patient was placed under general anesthesia. During the procedure, access to the left atrium (LA) was performed via 2 separate transseptal punctures (TSPs) under fluoroscopic and intracardiac echocardiographic guidance. The initial puncture was performed with an 8.5F Swartz braided SL1 transseptal sheath (St. Jude Medical, St. Paul, MN) using an 89-cm BRK-1 transseptal needle (St. Jude Medical) to allow passage of a circular mapping catheter. A second, separate TSP was performed with an 8.5F (11F outer diameter) Agilis medium curl steerable introducer sheath (St. Jude Medical) and a 98-cm BRK-1 transseptal needle to allow passage of the ablation catheter. Extensive areas of low voltage (< 0.5 mV) were seen throughout the LA, suggestive of diffuse atrial fibrosis. Pulmonary vein isolation was achieved, with demonstrable pulmonary vein entry and exit block following restoration of sinus rhythm with direct current cardioversion.

After catheter and sheath removal, systemic oxygen saturation decreased to 87%. As this was attributed to volume overload, the patient was prescribed furosemide and supplemental oxygen. By the following day he had clinically improved. He maintained oxygen saturations of 94% on room air and was therefore discharged home.

Over the next month he continued to suffer marked exertional dyspnea, fatigue, poor exercise tolerance, and weight loss. He was noted to be hypoxic on multiple occasions, with room air oxygen saturations ranging from 70% to 90%, which did not improve with supplemental oxygen. Peripheral cyanosis was noted on clinical examination. He remained in an atrial and ventricular paced rhythm with no further AF episodes on device interrogation. A computed tomography angiogram excluded pulmonary edema, pulmonary embolism, and pulmonary vein stenosis.

Repeat TTE identified a residual iatrogenic ASD with an associated right-to-left shunt. Pulmonary artery (PA) systolic pressures could not be reliably estimated on this or previous studies owing to the severe TR. The patient underwent a right heart catheterization, which demonstrated an elevated mean

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**KEYWORDS** Arrhythmogenic right ventricular cardiomyopathy; Arrhythmogenic right ventricular dysplasia; Atrial fibrillation; Atrial septal defect; Cyanosis; Pulmonary vein isolation; Right-to-left shunt; Transseptal puncture  
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## KEY TEACHING POINTS

- Transseptal puncture results in the formation of an iatrogenic atrial septal defect (ASD). In patients with right side–dominant cardiac pathologies (such as arrhythmogenic right ventricular cardiomyopathy), the transeptal pressure gradient from the right to left atrium promotes the formation of a right-to-left shunt across this ASD.
- A right-to-left shunt across an iatrogenic ASD should be considered in these patients who present with dyspnea following their procedure, particularly if hypoxemic and cyanotic.
- Potential strategies to minimize the risk of this complication in this population include avoiding “double sheathing” a single transeptal puncture, avoiding the use of larger sheaths, and assessing a right-to-left shunt at the conclusion of the case with echocardiography.
- Patients who remain symptomatic of a right-to-left shunt should undergo ASD closure.

RA pressure of 11 mm Hg and low RV and PA pressures (RV: 14/6 [mean 8] mm Hg, PA: 11/3 [mean 7] mm Hg). The mean pulmonary capillary wedge pressure was low at 4 mm Hg. Saturation runs confirmed a right-to-left shunt with a low pulmonary capillary wedge oxygen saturation of 83% (Table 1).

A transesophageal echocardiogram (TEE) confirmed persistence of 2 small ASDs in close proximity, resulting in a combined complex encompassing both defects measuring 7 mm at its maximal diameter. There was a continuous right-to-left shunt across this complex on color flow Doppler imaging throughout the entire cardiac cycle (Figure 1A and B, Supplemental Video).

The patient remained highly symptomatic despite cessation of furosemide in an attempt to raise LA filling pressures. The decision was therefore made to proceed to percutaneous closure of the ASD complex under fluoroscopic and TEE

guidance. While the sizing balloon was insufflated across the ASD and flow occlusion was confirmed on color Doppler (Figure 2A), the systemic oxygen saturation increased from 82% to 96%. The combined size of the ASD complex was confirmed as 6–7 mm on fluoroscopy measurement and a 10-mm Amplatzer septal occluder (St. Jude Medical) was deployed, resulting in occlusion of both defects (Figures 1C and 2B). No significant residual interatrial shunt was seen on TEE or contrast fluoroscopy, and repeat right heart catheterization revealed elevation in right-sided filling pressures and normalization of systemic arterial oxygenation (Table 1).

At 3 months post-procedure, the patient had no further resting dyspnea and had improved exercise tolerance. His oxygen saturation remained stable (97%) on ambulation.

## Discussion

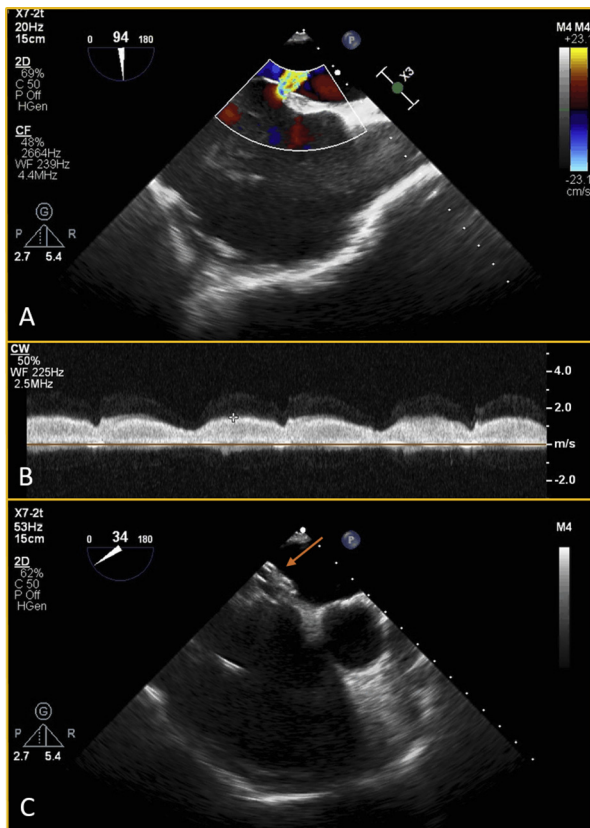
Catheter ablation for AF requires access to the LA via TSP. TSP is preferred even in patients with pre-existing patent foramina ovale, as the former provides better access to the posterior structures (eg, pulmonary veins) targeted during this procedure. As such, an iatrogenic ASD is present in nearly all patients post-procedure.

The majority of iatrogenic ASDs spontaneously close by 12 months,<sup>4,5</sup> though up to 26% can persist beyond this time.<sup>6</sup> As LA pressure tends to be greater than RA pressure, the majority of iatrogenic ASDs (up to 90%) are associated with some degree of left-to-right shunting when assessed on echocardiography in the first 6 months, although this is rarely clinically significant.<sup>7</sup> Right-to-left shunting can occur during early ventricular systole (or during a Valsalva maneuver or coughing) when the RA pressure transiently exceeds LA pressure.<sup>8</sup> Although this can give rise to paradoxical embolism, the momentary shunt reversal is not significant enough to cause hypoxemia.

The presence of a persistent right-to-left shunt requires the maintenance of a pressure gradient from the RA to LA throughout the cardiac cycle. In the absence of pulmonary hypertension, this phenomenon is rarely encountered, as it is conditional on the presence of isolated dysfunction of the right-sided cardiac chambers in patients with relatively preserved left-sided cardiac function. This prototypically occurs in ARVC, as suffered by our patient. However, other

**Table 1** Right heart catheterization values pre- and post-atrial septal defect closure, showing elevation of right-sided filling pressures and normalization of systemic oxygen saturations post-closure

Chamber	Pressure (mm Hg)				O <sub>2</sub> saturations (%)	
	Pre-closure		Post-closure		Pre-closure	Post-closure
	Systolic/diastolic	Mean	Systolic/diastolic	Mean		
Right atrium	-	11	-	16	52.1	-
Right ventricle	11/4	7	20/13	13	52.5	
Pulmonary artery	11/5	7	23/11	16	55.6	
Pulmonary capillary wedge (pre-closure)/direct left atrium (post-closure)	-	4	-	10	83.0	98.2



**Figure 1** A, B: Color flow (A) and continuous wave Doppler (B) during transesophageal echocardiography demonstrating right-to-left shunting across the iatrogenic atrial septal defect (ASD), continuous throughout the cardiac cycle. C: Final position of ASD occlusion (orange arrow) device across the interatrial septum.

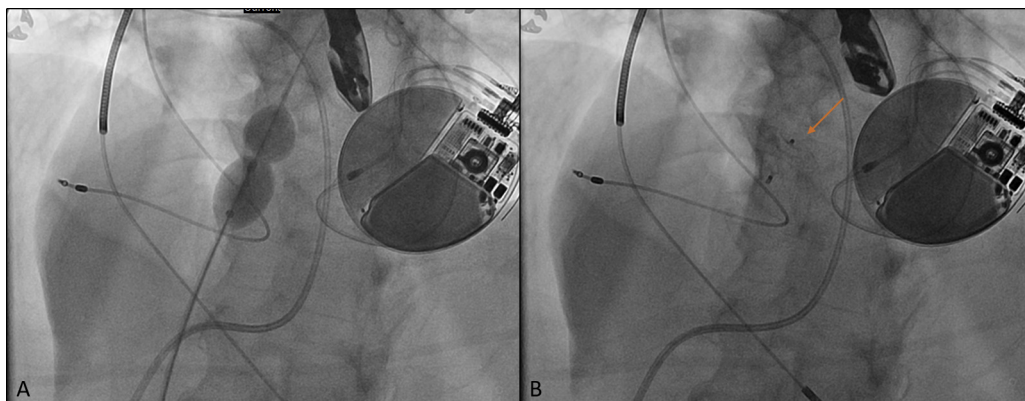
conditions that can present with RV-dominant disease include sarcoidosis,<sup>9</sup> carcinoid heart disease,<sup>10</sup> a large isolated RV infarct,<sup>11</sup> or adult congenital heart disease.<sup>12</sup> Tellingly, the only other reported case of a symptomatic right-to-left shunt owing to an iatrogenic ASD following AF ablation also occurred in a patient with ARVC, although this patient also had contributory pulmonary hypertension.<sup>13</sup>

In our patient, the elevated RA pressure was maintained by severe TR, while the severe RV systolic impairment and diuresis ensured a low LA pressure. The right-to-left shunt was therefore maintained by the RA-to-LA transeptal pressure gradient ( $11 - 4 = 7$  mm Hg), as demonstrated during the right heart catheterization study (Table 1).

Importantly, a right-to-left shunt through an iatrogenic ASD will not occur in the more common setting of cardiomyopathies with biventricular involvement (such as dilated cardiomyopathy), as the increase in RA pressure will be balanced by a proportional increase in LA pressure. In the AATAC trial, 102 patients with congestive cardiac failure underwent pulmonary vein isolation without any reports of residual shunts.<sup>14</sup>

Awareness of this complication is important, as it is predictable and therefore should be factored into procedural planning and the patient discussion of risks and benefits prior to considering a LA ablation procedure. In patients with ARVC, there is an unexpectedly high rate of AF (up to 24%) likely owing not only to chronic right atrial stretch but also to a diffuse biatrial myopathy, as was evident from the diffuse low voltage throughout the LA in our patient. Despite the apparent right-sided dominant pathology, many of these patients may benefit from LA procedures requiring TSP. Therefore, the rate of this complication may be underestimated in this population.<sup>15</sup>

We suggest the following preventative measures in these patients: (1) performing 2 separate TSPs rather than “double sheathing” a single puncture,<sup>5</sup> (2) performing radiofrequency ablation rather than laser or cryoablation owing to the need for a larger (15F) transeptal sheath size<sup>6,7</sup> in the latter cases, and (3) using TEE or intracardiac echocardiography guidance. We suggest performing color flow Doppler imaging of the iatrogenic ASD at the end of the procedure by withdrawing the transeptal sheaths into the RA while maintaining LA access with an 0.035-inch wire in the left upper pulmonary vein. If a right-to-left shunt is observed, it would be reasonable to perform a right heart catheterization study in order to directly measure pressure and O<sub>2</sub> saturations in both atria. If hypoxemia owing to a right-to-left shunt is



**Figure 2** A: Right anterior oblique 30° left anterior oblique 30° fluoroscopy view demonstrating insufflation of the sizing balloon across the atrial septal defect (ASD). This resulted in normalization of systemic oxygen saturation. B: Final position of ASD occluder (orange arrow).

confirmed, consideration for either immediate percutaneous ASD closure<sup>13</sup> or close follow-up should be determined based on clinical symptoms.

If a patient is minimally symptomatic, clinical surveillance with periodic TTEs to assess for spontaneous iatrogenic ASD closure would be reasonable, as the presence of a septal closure device makes subsequent LA access more challenging in the event of recurrent arrhythmias. That said, it is not known whether the presence of a persistent right-to-left shunt reduces the rate of spontaneous iatrogenic ASD closure. Therefore it is important to ensure that all involved caregivers have a low threshold to reinvestigate the patient in the case of worsening symptoms or cyanosis.

## Conclusion

Symptomatic hypoxemia owing to a persistent right-to-left shunt across an iatrogenic ASD is a rare but predictable complication of AF ablation in patients with RV-dominant diseases such as ARVC. In symptomatic cases, ASD closure should be performed.

## Appendix Supplementary data

Supplementary data associated with this article can be found in the online version at <https://doi.org/10.1016/j.hrcre.2017.11.013>.

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