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.[1] Other uncommon complications that can occur subacutely include pulmonary embolism, pulmonary vein stenosis,[2] and stiff left atrial syndrome.[3] 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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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 transseptal 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 dypsnea 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,[4,5] though up to 26% can persist beyond this time.[6] 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.[7] 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.[8] 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

	Pressure (mm Hg)				O_2 saturations (%)	
	Pre-closure		Post-closure			
Chamber	Systolic/diastolic	Mean	Systolic/diastolic	Mean	Pre-closure	Post-closure
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