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# Atrial Fibrillation Ablation in a Patient With SV ASD and PAPVR Preceding Transcatheter Septal Closure

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## ABSTRACT

Atrial fibrillation (AF) is common in adults with unrepaired atrial septal defects (ASDs). Sinus venosus (SV) ASDs associated with partial anomalous pulmonary venous return (PAPVR) are traditionally managed surgically. We report the first AF catheter ablation in a patient with SV ASD and PAPVR preceding transcatheter ASD repair with a covered stent. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;15:101862) © 2023 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 56 year-old woman presented with symptomatic paroxysmal atrial fibrillation (AF).

## PAST MEDICAL HISTORY

Her medical history was notable for hypertension.

### LEARNING OBJECTIVES

- To recognize AF as highly prevalent in adults with unrepaired ASDs and appreciate that ASD closure alone, although it is definitive treatment, is often insufficient to eliminate tachyarrhythmia.
- To show the first-in-human AF catheter ablation in a patient with SV ASD and PAPVR before transcutaneous SV ASD closure.

## INVESTIGATIONS

Cardiac evaluation, including transesophageal echocardiography, cardiac computed tomography, and right-sided heart catheterization, revealed a 4.9-mm sinus venosus (SV) atrial septal defect (ASD) with a left-to-right shunt, as well as an anomalous right superior pulmonary vein (RSPV) draining into the superior vena cava (SVC), biatrial and right ventricular dilation, and pulmonary hypertension (**Figures 1A to 1C and 2A and 2B**). Increased oxygen saturations were also noted in the right atrium (RA) and pulmonary artery.

## MANAGEMENT

The patient was evaluated by a multidisciplinary team, including a pediatric interventional cardiologist, a pediatric cardiac surgeon, an adult congenital heart specialist, and an electrophysiologist. In this

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**ABBREVIATIONS AND ACRONYMS**

**AF** = atrial fibrillation

**ASD** = atrial septal defect

**LA** = left atrium

**OS** = ostium secundum

**PAPVR** = partial anomalous pulmonary venous return

**RA** = right atrium

**RSPV** = right superior pulmonary vein

**SV** = sinus venosus

**SVC** = superior vena cava

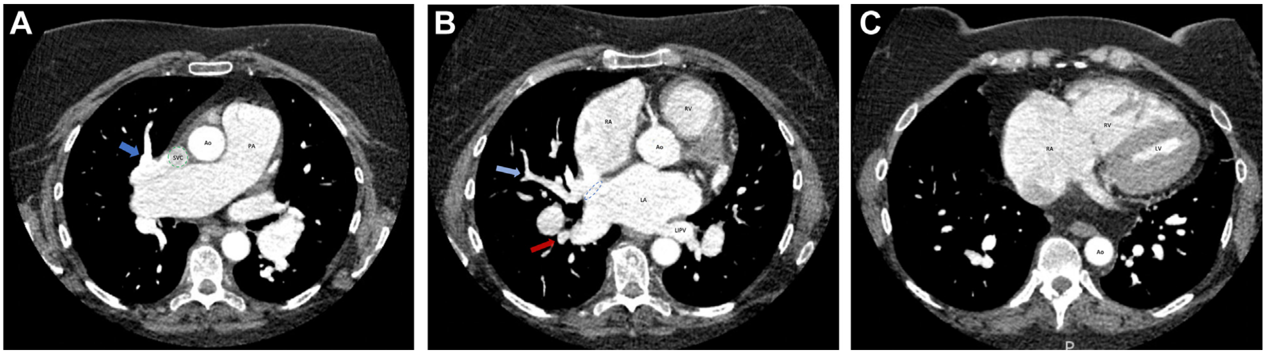
patient with pulmonary hypertension and atrial remodeling with consequential arrhythmia development, all ultimately related to her underlying ASD, closure of the SV ASD was believed to represent definitive management. Fortunately, she was deemed a candidate for transcatheter SV ASD repair. Because a covered stent repair would prohibit access to the RA-SVC junction and could complicate transseptal access to the left atrium (LA) endocardium, AF catheter ablation was planned before transcutaneous septal closure.

An RA-SVC electroanatomic map was first generated using a Pentaray high-density mapping catheter and the Carto 3 mapping system (Biosense Webster, Inc). Low-voltage myocardium was identified in the RA-SVC junction extending to the SVC. No voltage was identified in the anomalous RSPV (Figure 3A). Given the known role of SVC and the pulmonary veins as AF triggers, the RSPV-SVC communication, and future inaccessibility to this region after ASD closure with a covered stent, the decision was made to proceed with SVC isolation in conjunction with pulmonary vein isolation. Transseptal access was obtained in the fossa ovalis by using an NRG Transseptal Needle (Baylis Medical, Inc). An LA electroanatomic map was generated using the high-density mapping catheter (Figure 3B). A 28-mm cryoballoon catheter (Medtronic, Inc) was used to isolate the anatomically normal pulmonary veins. Cryoablation of the RSPV-SVC-LA junction was performed from the LA with selective engagement of the superior and inferior branches of the RSPV. Absence of phrenic nerve injury was confirmed with high-output pacing during cryoablations of the right-sided veins. Esophageal temperature was monitored during energy delivery. After cryoablation of all pulmonary veins, a region of viable myocardium remained in the anterolateral RA-SVC junction. After confirming absence of phrenic nerve capture, radiofrequency energy was delivered using an irrigated contact-force sensing ablation catheter (SmartTouch) to complete isolation of the SVC (Figures 4A and 4B). Entrance and exit blocks were confirmed for each pulmonary vein and the SVC before and after adenosine administration.

## DISCUSSION

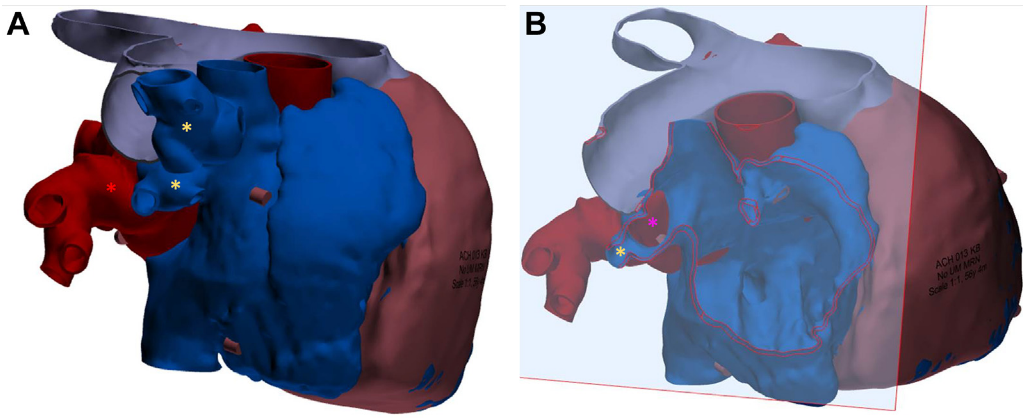
ASDs are among the most common adult congenital heart defects, with a prevalence of 0.88 per 1,000 adults. There are 3 main types of ASDs: ostium primum, ostium secundum (OS), and SV. OS ASDs account for 80% of ASDs and result from a deficiency of tissue at the level of the fossa ovalis. SV ASDs denote a tissue defect in the common wall between the SVC and the right-sided pulmonary veins; these defects represent 4% to 11% of ASDs and are almost always associated with partial anomalous pulmonary venous return (PAPVR).[1] According to the 2018 American Heart Association/American College of Cardiology guidelines for the management of adults with congenital heart disease, patients with SV ASDs

**FIGURE 1** Computed Tomography Imaging of PAVPR and SV ASD




**(A)** Superior branch of the right superior pulmonary vein **(blue arrow)** draining at the junction of the superior vena cava (SVC) **(green circle)** and right atrium (RA). The pulmonary artery (PA) is also notably enlarged compared with the aorta (Ao), consistent with pulmonary hypertension. **(B)** Inferior branch of the right superior pulmonary vein **(blue arrow)** draining into the right atrium at the level of the sinus venosus atrial septal defect **(dotted blue oval),** with the right inferior pulmonary vein **(red arrow)** draining into the left atrium (LA). **(C)** Markedly enlarged right atrium and right ventricle (RV). LIPV = left inferior pulmonary vein; LV = left ventricle.

**FIGURE 2** 3-Dimensional Modeling (Mimics Innovation Suite) of Partial Anomalous Pulmonary Venous Return and Sinus Venosus Atrial Septal Defect

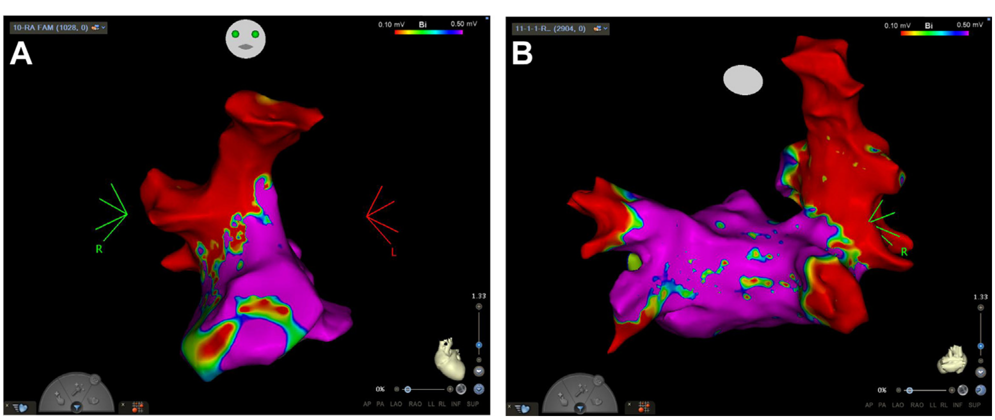



**(A)** Anomalous branches of the right superior pulmonary vein **(yellow asterisk)** draining into the superior vena cava, with the right inferior pulmonary vein **(red asterisk)** draining into the left atrium. **(B)** Cross-sectional view of the right atrium visualizing part of the anomalous right superior pulmonary vein **(yellow asterisk)** draining into the cavoatrial junction with a view of the sinus venosus atrial septal defect **(pink asterisk).**

whose defect causes impaired functional capacity, right atrial and/or right ventricular enlargement, and a left-to-right shunt leading to physiologic sequelae, notably without signs of elevated pulmonary vascular resistance concerning for existing or impending Eisenmenger syndrome, should undergo septal closure. Whereas OS ASDs are commonly repaired through transcatheter device closure, ostium primum and SV ASDs have traditionally been managed surgically. In recent years, transcatheter closure of SV ASDs with a covered stent has emerged as an alternative to surgery in a substantial portion of cases.[2,3]

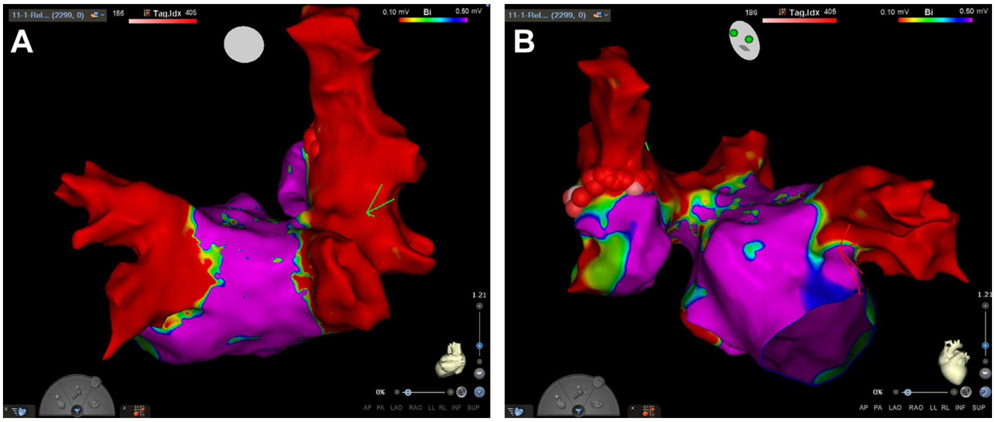
The prevalence of AF in adults with an unrepaired ASD is higher than in the general population, approximately 20% to 50% of adults over the age of 40 years.[4] Of note, most investigations of AF and

**FIGURE 3** Electroanatomic Mapping Featuring Partial Anomalous Pulmonary Venous Return, Pre-Ablation




**(A)** Anteroposterior cranial view: right atrium-superior vena cava and anomalous right superior pulmonary vein voltage mapping at baseline. **(B)** Posteroanterior view: left atrium, superior vena cava, and pulmonary veins, including the anomalous right superior pulmonary vein voltage mapping at baseline. L = left; R = right.

**FIGURE 4 Electroanatomic Mapping Featuring Partial Anomalous Pulmonary Venous Return, Post-Ablation**




**(A)** Posteroanterior view: left atrium voltage mapping after cryoablations. **(B)** Left anterior oblique cranial view: superior vena cava isolation voltage mapping with radiofrequency energy ablations **(red dots).**

ASDs predominantly feature patients with OS ASD, given its comparatively higher prevalence. ASD closure, either surgical or percutaneous, may result in reverse atrial modeling, both structurally and electrically.[4] However, ASD repair alone is often insufficient to eliminate pre-existing tachyarrhythmia[5]; similarly, AF ablation in isolation is unlikely to control arrhythmia in these patients.

Patients who undergo catheter ablation before OS ASD device closure have significantly higher AF-free survival rates post-ASD repair as compared with patients who have not undergone preclosure ablation.[6-8] Furthermore, the incidence of new onset AF post-transcatheter ASD closure is relatively low, particularly in patients who are non-elderly patients.[9] This finding suggests a strategy of ablation preceding ASD repair.

PAPVR is an uncommon, although likely underdiagnosed, cause of right chamber enlargement. There are only a few case reports of patients with PAPVR who have undergone AF catheter ablation, but this appears to be feasible and effective.[10] To our knowledge, our case is the first report of a patient with SV ASD with PAPVR who underwent AF catheter ablation as a staged procedure before transcatheter SV ASD repair.

## FOLLOW-UP

The patient tolerated the procedure well, without complications. Five months later, she underwent successful SV ASD closure with a covered stent. She has done well, with no symptoms to suggest recurrence of AF.

## CONCLUSIONS

We describe the first-in-human case of AF catheter ablation in a patient with SV ASD and PAPVR as a staged procedure before percutaneous septal closure. Our strategy may represent a paradigm shift in how we approach AF in SV ASD and PAPVR without need for open heart surgery.

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**KEY WORDS** atrial fibrillation, congenital heart disease, partial anomalous pulmonary venous return, sinus venosus atrial septal defect