

Pulmonary vein isolation for paroxysmal atrial fibrillation in a patient with stand-alone unroofed coronary sinus



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Introduction

Atrial arrhythmia is common in patients with an atrial septal defect (ASD).¹ Atrial fibrillation (AF) is the main cause of morbidity in older patients with ASD.² An unroofed coronary sinus (CS) is rare (in <1% of all types of ASD)³ and is often associated with persistent left superior vena cava (PLSVC) and other forms of complex congenital heart disease and heterotaxy syndromes. Unroofed CS rarely occurs alone. This is a unique report of pulmonary vein isolation (PVI) for paroxysmal AF in a patient with stand-alone unroofed CS.

Case report

A 74-year-old woman was admitted to our hospital to undergo catheter ablation for treatment of symptomatic paroxysmal AF. She had been anticoagulated for 6 years. Transthoracic echocardiography showed right atrial and ventricular dilation, pulmonary hypertension (estimated right ventricular systolic pressure 49 mm Hg), and a dilated CS in the parasternal long-axis view. Enhanced computed tomography (CT) revealed an unroofed CS (Figure 1A and 1B) and a grossly dilated pulmonary artery. There were no other congenital anomalies. The patient chose catheter ablation to control the rhythm, rather than surgical repair of the unroofed CS and the Maze procedure.

Written informed consent was obtained. Cardiac catheterization showed increased O₂ saturation in the CS (95.1% in the CS, and 83.4% in the right atrium), a pulmonary-to-systemic flow ratio (Qp/Qs) of 2.45, and elevated main pulmonary artery pressure of 33/19 mm Hg (mean 25 mm Hg). A duodecapolar catheter was placed in the CS via the jugular vein. Two long sheaths from the femoral vein were introduced into the left atrium (LA) through a single transseptal puncture under intracardiac

echocardiographic guidance instead of through access via the unroofed CS. Access via the unroofed CS would have been difficult because the CS opened into the left posterior LA. A transesophageal thermometer was inserted to avoid injury to the esophagus. The activated clotting time was controlled at approximately 300–350 seconds during the procedure. A 3.5-mm, open, irrigated-tip catheter (Navistar ThermoCool SF; Biosense Webster, Diamond Bar, CA) was used for mapping and ablation.

The patient underwent wide circumferential PVI guided by a 3-dimensional electroanatomic mapping system with CT integration (CARTO3; Biosense-Webster) (Figure 2A and 2B). Dormant pulmonary vein conduction was not observed. However, sustained AF was initiated by premature atrial contractions from the ostium of the CS after a bolus injection of adenosine triphosphate during continuous infusion of isoproterenol (Figure 3A). After ablation of non-pulmonary vein (PV) foci at the ostial region of the CS floor (Figure 3B), AF was not induced by programmed electrical stimulation or adenosine triphosphate infusion. The patient was asymptomatic for 6 months after the procedure, and Holter electrocardiography (3 and 6 months) showed a regular sinus rhythm. We recommend early surgical repair of unroofed CS.

Discussion

We performed circumferential PVI in a patient with a stand-alone unroofed CS via an atrial septal puncture. The approach to the pulmonary veins through the unroofed CS was impossible because the unroofed CS opened into the posterior LA below the left inferior PV. Successful PVI and non-PV foci ablation eliminated paroxysmal AF without antiarrhythmic drugs.

Unroofed CS, a rare type of ASD, is a direct communication between the CS and LA. Unroofed CS is classified into 4 subtypes, as follows: completely unroofed with PLSVC (type I); completely unroofed without PLSVC (type II); partially unroofed mid portion (type III); partially unroofed terminal portion (type IV).⁴ The present case was classified as type III with no other congenital heart disease.

Unroofed CS causes nonspecific clinical signs and symptoms. Transthoracic echocardiography is commonly used to

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KEY TEACHING POINTS

- An unroofed coronary sinus is a rare type of atrial septal defect.
- Cardiac computed tomography is useful for diagnosing unroofed coronary sinus.
- Most patients with unroofed coronary sinus and atrial fibrillation require surgical repair; however, some patients may benefit more from catheter ablation.
- Pulmonary vein isolation is the cornerstone for paroxysmal atrial fibrillation. A dilated coronary sinus due to unroofed coronary sinus might be the source of arrhythmogenesis in atrial fibrillation.

detect unroofed CS, but it cannot visualize posterior cardiac structures. Enhanced CT and magnetic resonance imaging are useful diagnostic modalities that allow anatomic and morphologic assessment of the posterior portions of the heart.^{5,6} In our case, enhanced cardiac CT findings eventually led to the diagnosis of unroofed CS.

Most patients with unroofed CS and AF are treated by surgical repair. Some elderly patients whose clinical symptoms appear to be mostly due to AF, however, benefit from

catheter ablations of the AF alone. The choice of approach for left atrial ablation depends on the type of unroofed CS. The approach to the LA through the unroofed CS is possible for types I and II. For types III and IV, however, a transeptal approach facilitates catheter manipulation.

Little is known about electrophysiological abnormalities causing AF in patients with an unroofed CS. Our experience suggested that the ostium of the dilated CS could be a nonpulmonary focus of AF to be examined and treated. PVI is the cornerstone of radiofrequency catheter ablation for paroxysmal AF, even in cases with congenital heart disease. In drug-refractory adult patients with AF and congenital heart disease, the success rate after PVI is similar to that in those with noncongenital structural heart disease.⁷ Catheter ablation sites in patients with unrepaired ASD are similar to those with normal septal anatomy.⁸ Nevertheless, the left superior vena cava can trigger AF.⁹ Furthermore, when patients develop recurrent AF after repair of an unroofed CS with a concomitant Maze procedure, catheter ablation of a dilated CS may be the solution because CS ablation is not included in the Cox–Maze IV lesion set.¹⁰

In conclusion, we successfully treated paroxysmal AF by PVI and CS ablation in an older patient with a stand-alone anomaly of unroofed CS. This case report has some limitations. First, AF recurrence may have been undetected, although this patient had highly symptomatic episodes of AF. Second, the 6-month follow-up might have been an inadequate length of time to draw accurate conclusions based on the results.

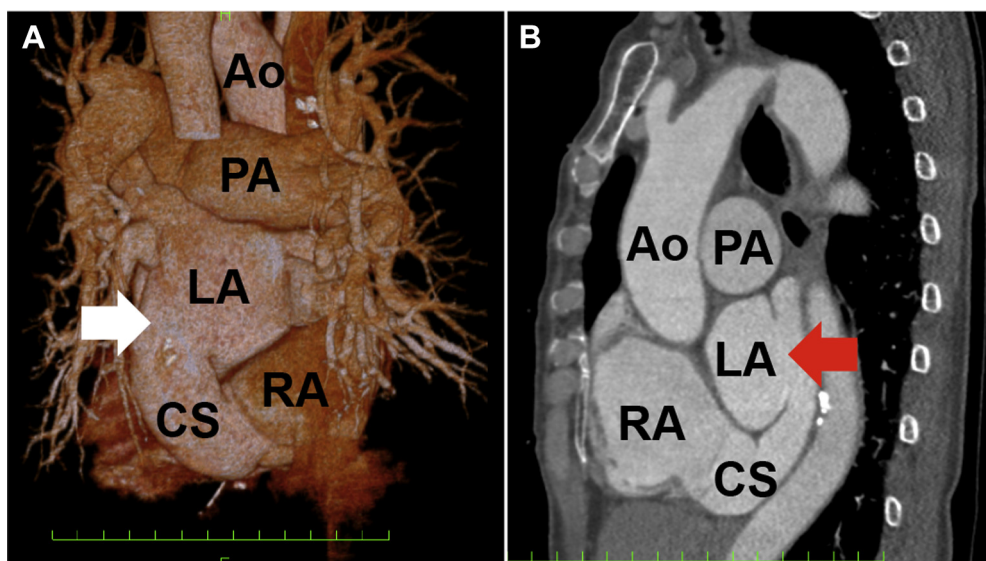


Figure 1 Computed tomographic coronary angiogram. **A:** Three-dimensional construction of the heart in the posteroanterior view. The coronary sinus connects the left inferoposterior site of the left atrium (*white arrow*). **B:** Sagittal view shows left-to-right communication through a defect in the unroofed coronary sinus (*red arrow*). Ao = aorta; CS = coronary sinus; LA = left atrium; PA = pulmonary vein; RA = right atrium.

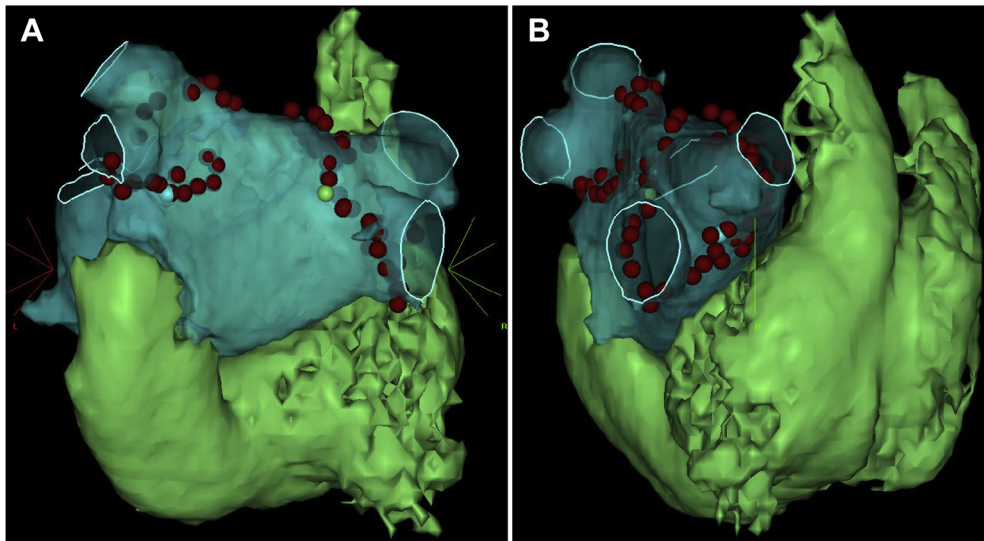


Figure 2 Three-dimensional electroanatomic map of the left and right atria: **A**: posteroanterior view; **B**: right-left view. The red dots represent ablation points, and the blue and green dots represent isolation points of the left and right pulmonary veins, respectively.

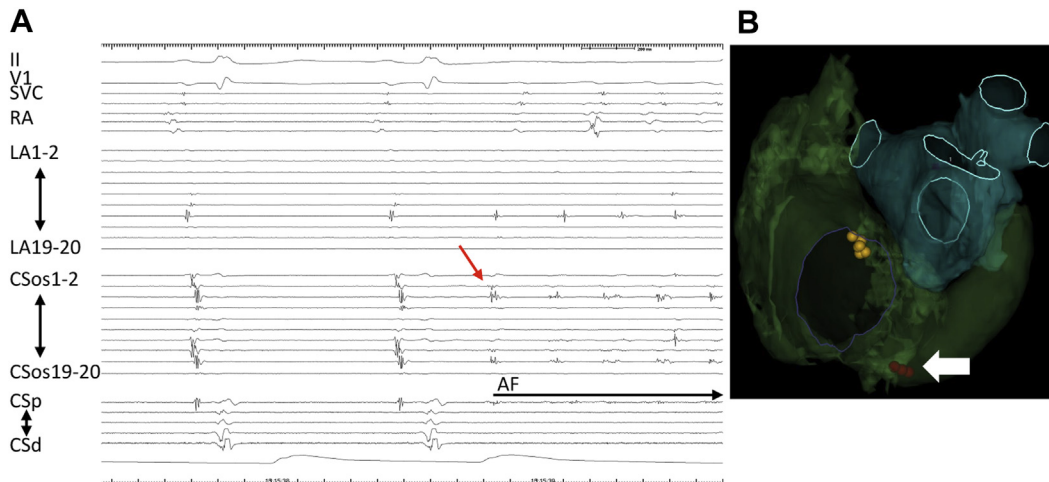


Figure 3 **A**: Sustained atrial fibrillation was initiated by atrial premature contraction recorded by a PentaRay NAV (Biosense-Webster, Diamond Bar, CA) catheter that was placed in the ostium of the coronary sinus (red arrow). A Lasso NAV (Biosense-Webster, Diamond Bar, CA) catheter was placed on the bottom of the left atrium. AF = atrial fibrillation; CS = coronary sinus; CSd = distal coronary sinus; CSos = ostium of the coronary sinus; CSp = paroxysmal coronary sinus; LA = left atrium; RA = right atrium; SVC = superior vena cava. **B**: Three-dimensional electroanatomic map of the left and right atria in the right anterior oblique projection. The ablation points were located on the floor of the coronary sinus ostium (white arrow). The yellow tags show the His bundle potential.

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