Coronary sinus ostium in the lateral right atrium: Implications for the electrophysiologist

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Introduction
The coronary sinus (CS) is a critical structure that has helped electrophysiologists by providing information about epicardial activation along the mitral annulus. Additionally, the advent of biventricular pacing utilizes the CS venous system for lead placement. The CS collects the venous drainage of the atrial and ventricular venous branches and drains into the right atrium (RA). CS anomalies have been recognized, namely atresia of the CS ostium, unroofed CS, connection with the left atrium, and even hypoplasia of the CS.1 We report 3 cases of CS ostium opening in the midlateral RA. This anomaly may provide new insight into the development of the CS as well as provide an explanation for difficult cannulation of the CS.

Case report
Case 1
A 39-year-old man with a history of alcohol-related cardiomyopathy, left bundle branch block on electrocardiogram, and NYHA class III symptoms was referred for biventricular implantable cardioverter-defibrillator implantation. No evidence of coronary venous anomalies was suggested on echocardiography. After implantation of the right ventricle (RV) lead, left ventricle (LV) lead implantation was attempted. A deflectable sheath was advanced over a wire to the low RA. There, multiple attempts to engage the CS ostium were unsuccessful despite using an Amplatz left (AL I) coronary catheter and CS luminal catheter (St Jude Medical, Minnetonka, MN). Further, contrast injections in the low, septal RA did not identify the CS ostium. Given difficulty in accessing the CS, left coronary angiography was performed to assess the location of the CS ostium. During levophase, CS filling was noted and it extended along the right atrial floor and ultimately drained in the posterolateral RA (Figure 1A, with white arrow pointing to CS ostium).

After the CS ostium was identified, the deflectable sheath was retracted to the lateral RA. Through this sheath, an AL I catheter with Wholey wire and then a CS luminal catheter were unsuccessful in engaging the CS ostium. Finally, a Williams right coronary angiographic catheter successfully engaged the CS with appropriate wire advancement. The outer sheath was then advanced into the CS without difficulty. At this point, a CS venogram was obtained, which identified a suitable lateral venous branch. An LV lead was then advanced through the sheath into the CS and advanced to the lateral venous branch (Figure 1B).

Case 2
A 45-year-old man with recurrent, drug-refractory supraventricular tachycardia was referred for electrophysiology study and ablation. CS cannulation was attempted with deflectable decapolar and deflectable quadripolar catheters, without success. After repeated unsuccessful attempts, levophase coronary angiography revealed the presence of a CS that extended to the lateral RA (Figure 2). After this was noted, further attempts to cannulate the CS were abandoned. With catheters in the high RA, His bundle position, and RV apex, pacing maneuvers suggested the presence of a concealed left-sided accessory pathway. Further mapping was performed with an ablation catheter in the left atrium via a transseptal approach. Successful catheter ablation of the concealed accessory pathway was performed at the posterolateral mitral annulus.

Case 3
A 44-year-old man underwent electrophysiology study for recurrent supraventricular tachycardia. Attempts at cannulating the CS using 5F and 4F deflectable decapolar catheters were unsuccessful. The left coronary angiogram was performed to delineate the CS ostium. The levophase revealed a bulbous blunt end of the CS at the usual site of opening in the RA. Instead, the flow continued via a tributary along the right atrioventricular groove, which opened in the
midlateral RA (Video). Electrode catheters were positioned in the RA, His bundle region, and RV. The tachycardia was easily inducible and found to be typical atrioventricular (AV) nodal reentrant tachycardia. The blunt end of the CS was marked as the “CS os” region. Radiofrequency ablation of the slow pathway was performed successfully in the conventional manner.

Discussion
To our knowledge, only 1 other report in the literature notes a CS that courses along the RA floor and ultimately drains in the lateral RA. None of the patients described had any evidence of a persistent left superior vena cava (SVC) or other forms of congenital heart disease. The CS is known to have 5 general categories of abnormalities. This includes enlargement of the CS (with or without associated left-sided shunt), complete atresia of the CS, hypoplasia of the CS, unroofing of the CS, and CS ostium atresia. With CS ostium atresia, the CS ostium does not allow for venous blood to enter the RA, and blood usually enters the RA via small Thebesian veins or via a left-sided SVC, if present. Further, in 15% of patients, the anterior cardiac vein can drain directly to the RA and, rarely, the great cardiac vein may directly drain into the SVC. In the present case series, however, coronary angiography clearly demonstrated a CS that continues to the lateral RA before draining into the RA.

Multiple explanations may account for this CS anomaly. The CS develops from the primordial sinus venosus, which by 8 weeks of age has both a left and right horn. The left horn develops into the CS while the right horn becomes the “smooth part” of the RA (sinus venarum). In our cases, the CS continued along the floor of the RA to the lateral RA. Abnormal signaling between the 2 horns of the sinus venosus may have caused the continuation of the CS into the RA. Another explanation may involve the drainage of the anterior cardiac veins. The anterior cardiac veins drain venous blood from the RV. In approximately 60% of cases, the anterior cardiac veins drain into intramural channels in the RA parallel to the tricuspid annulus. These sinuses can measure 2–12 cm in length and 1–4 mm in diameter and have been dubbed the “right atrial coronary sinus.” One could hypothesize incorporation of the CS with this channel, with ultimate drainage in the lateral RA. Finally, Uemura and colleagues described the cardiac venous return in a series of 148 patients with isomeric atrial appendages. In patients with left atrial appendage isomerism, 10% did not exhibit a CS within the AV groove and had direct venous return to the atrium. This venous return was noted either to be a direct connection near the AV groove, to be a “crooked path” where a venous branch took an intramural course along the AV groove, or to drain at a distant site from the AV groove. Our cases, however, suggest continuous venous return via the CS in the typical left-sided AV groove that continues to the lateral RA. Finally, the described cases may represent a variant of CS atresia with well-formed collaterals, which then ultimately drain the lateral RA. Given a typical left-sided CS course, echocardiography or other imaging is unlikely to identify this anomaly.

During potential LV lead implant, an angulated coronary catheter (Williams right / Multipurpose / Hockey Stick) would be useful in wire cannulation of the anomalous CS ostium. Once wire cannulation is achieved, an LV delivery sheath can be advanced over the wire. Once a suitable LV

Figure 1  Coronary sinus ostium in the high lateral right atrium. A: During levophase of left coronary angiography, the coronary sinus was visualized and was noted to continue to the ostium located in high lateral right atrium (white arrow). B: Successful left ventricle lead placement after cannulation of the coronary sinus anomaly.
Figure 2  Levophase of coronary angiography in A: left anterior oblique (LAO) view and B: anteroposterior (AP) view, demonstrating the coronary sinus ostium in the posterolateral right atrium (black arrow).

branch is identified, a longer LV lead would be useful to traverse the RA course of this CS anomaly.

Although 3 cases are presented in this report, the actual incidence of this anomaly is likely rare, given the lack of this anomaly finding in prior autopsy series.7

Conclusion
This series demonstrates a unique CS anomaly with the CS ostium in the lateral RA and highlights the utility of coronary angiography for difficult CS cannulation.

Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at http://dx.doi.org/10.1016/j.hrcr.2017.07.017.

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