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# A Case of Congenital Coronary Sinus Ostial Atresia with Persistent Left Superior Vena Cava

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## CASE REPORT

# A Case of Congenital Coronary Sinus Ostial Atresia with Persistent Left Superior Vena Cava

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**Introduction:** The coronary sinus is responsible for venous drainage of the heart. Congenital anomalies of the coronary sinus are rare and typically associated with other anomalies, such as atrial septal defects or other congenital heart malformations.

**Clinical Findings:** We present a case of incidentally found atresia of the coronary sinus ostium with associated persistent left superior vena cava. This case involved a 52-year-old patient who initially presented with a supraventricular tachycardia and elevated troponin.

**Clinical Course:** The patient underwent cardiac workup with a coronary computed tomographic angiography scan. This scan showed no obstructive coronary artery disease, but did show congenital atresia of the coronary sinus and a persistent left superior vena cava.

**Conclusions:** This case shows that coronary computed tomographic angiography is an excellent modality for identifying anomalous coronary venous return and congenital abnormalities of the coronary sinus.

**Keywords:** coronary sinus, coronary circulation, coronary artery disease, coronary disease

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A 52-year-old patient presented with symptomatic, new-onset narrow complex tachycardia with heart rates up to 230 beats per minute. Their past medical history included hypothyroidism with replacement therapy and no prior cardiac history.

In the exam, the patient was hemodynamically stable without evidence of volume overload. The patient was initially treated with adenosine but did not respond. Electrical cardioversion was also unsuccessful. The patient finally responded to intravenous beta blockers and amiodarone, which improved their heart rate to the low 100s. The patient's bloodwork showed an elevated troponin T with a peak of 0.6 ng/mL (normal range <0.01 ng/mL). An echocardiogram showed normal biventricular systolic function, moderate left atrial enlargement, and no valvular abnormalities.

Given the elevated troponin, we pursued a coronary computed tomographic angiography scan. This scan showed no obstructive coronary artery disease, but revealed a persistent left superior vena cava (SVC) connecting into the left atrium (Figure 1). The coronary sinus ostia was atretic, and coronary venous drainage occurred via anomalous cardiac venous connections to the left atrium. Ultimately, we deemed the hemodynamic significance of this anomaly as minimal because the patient had no evidence of hypoxia to suggest a large right-to-left shunt.

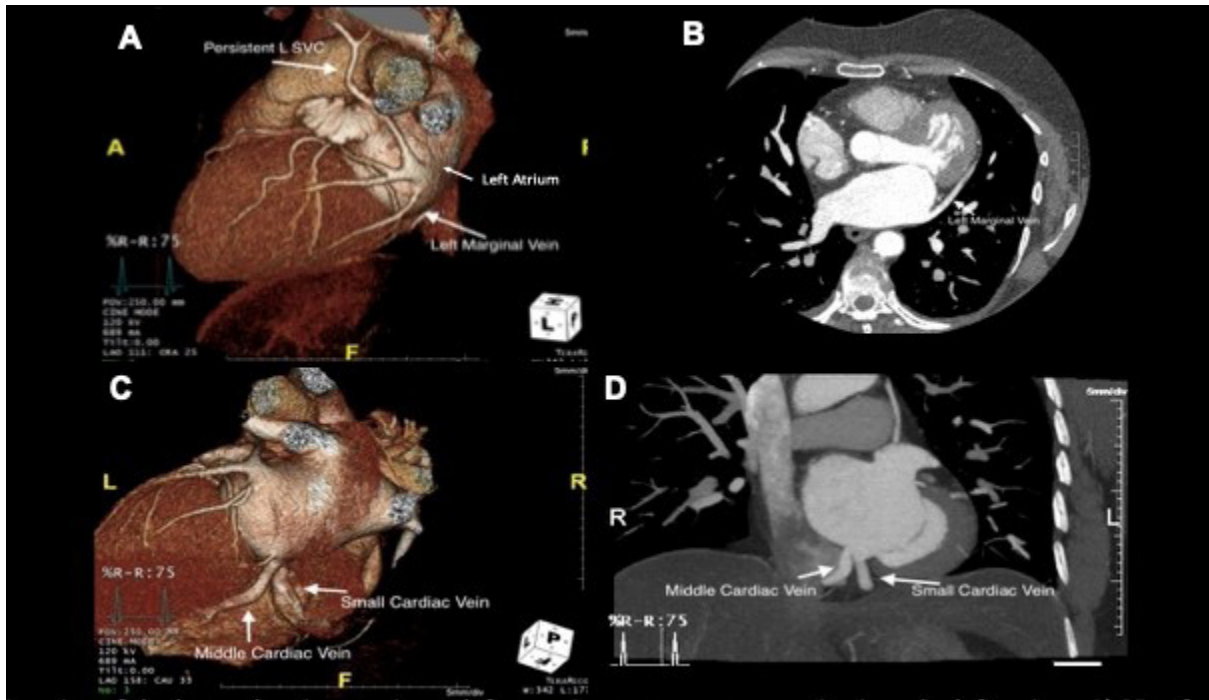
Subsequently, the patient underwent successful ablation of a concealed left lateral accessory pathway for an atrioventricular nodal re-entry tachycardia. Since the ablation, the patient has done well.

## DISCUSSION

Typically, the coronary sinus is formed by the connection between the great cardiac vein and the vein of Marshall in the posterior atrioventricular groove.<sup>1</sup>

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**Figure 1.** Imaging of the Heart Showing the Persistent Left Superior Vena Cava Connecting into the Left Atrium. A, 3-dimensional reformat showing the cardiac veins and persistent left superior vena cava connecting into the left atrium. B, Axial view showing the left marginal vein connecting into the left atrium. C, 3-dimensional reformat of the cardiac veins connecting into the left atrium. D, Axial view of the cardiac veins connecting into the left atrium. L SVC, left superior vena cava.

In atresia of the coronary sinus ostium, the coronary sinus is present, but it ends as a blind pouch or severe stenosis.<sup>2</sup> The difference between coronary sinus atresia and the absence of the coronary sinus can be challenging to determine and is usually diagnosed on autopsy. However, the clinical implications of these conditions are similar.<sup>3</sup>

As a result of coronary sinus ostial atresia or absence, coronary venous blood return occurs via communication with the left atrium and can often include a persistent left SVC.<sup>2</sup> In these rare cases, the persistent left SVC can carry blood from the coronary venous system via a bridging vein to the right SVC and then into the right atrium.<sup>2</sup> Cases of coronary sinus ostial atresia without a persistent left SVC have venous drainage into the left atrium via other cardiac veins, such as the great cardiac vein.<sup>2</sup> Ultimately, this anomaly leads to a small right-to-left shunt, though typically not with high enough volume to be clinically or hemodynamically significant.<sup>2</sup> As a result, the finding is typically incidental and management is conservative.

This anomaly can have important implications, especially for electrophysiology procedures that may use the coronary sinus. In rare cases, anomalous coronary venous return can result in

the development of a variety of signs related to right-to-left shunting, including the potential for embolic events, unexplained cerebral abscess, and important implications for electrophysiologic procedures that use the coronary sinus for pacing, mapping, or ablation.<sup>1</sup>

### Learning Points

Clinicians should be familiar with identifying a persistent left SVC (Figure 1). In addition, this case shows that coronary computed tomographic angiography is an excellent modality for identifying anomalous coronary venous return and congenital abnormalities of the coronary sinus.

**Conflicts of Interest:** None

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