A 21-year-old white male athlete was admitted for syncope. ECG and telemetry showed sinus arrhythmia and sinus bradycardia; at this time, he was asymptomatic. A chest radiograph showed unremarkable findings (Figure 1). During his transthoracic echocardiogram, a massive coronary sinus (area, 3.9 cm²) was identified (Figure 2). Because of suspicion of a persistent left superior vena cava (PLSVC), an agitated saline (“bubble”) study was immediately performed. After injection of agitated saline through his left antecubital vein, the dilated coronary sinus was opacified before the right atrium and ventricle, consistent with PLSVC (Figure 3 and Movie I in the online-only Data Supplement).

PLSVC, a congenital anomaly of the thoracic venous system, is present in 0.4% of the general population.1 When the left superior cardinal vein fails to regress to form the ligament of Marshall, a persistent left-sided vasculature system drains to the coronary sinus, and thus PLSVC occurs. In 90% of these cases, bilateral superior vena cava (SVC) is present. However, concomitant agenesis of the right-sided SVC is rare. To evaluate the potential existence of the right SVC, a subsequent injection of agitated saline was given from the right antecubital vein. The dilated coronary sinus, right atrium, and right ventricle were opacified sequentially, consistent with agenesis of the right SVC (Movie II in the online-only Data Supplement). The presence of PLSVC and the absence of the right SVC combined become a very rare anomaly, namely isolated PLSVC. Coexistent agenesis of the right SVC renders the coronary sinus further dilated because of increased venous return directly to the coronary sinus. Computerized tomography angiography (Figure 4) supported the findings of isolated PLSVC and identified no other anomalies. The patient was discharged with an event monitor. The results of a 30-day event monitor showed sinus arrhythmia and Mobitz type I atrioventricular block, and the patient remained asymptomatic at the last follow-up.
Although the isolated PLSVC drains to the coronary sinus and right atrium and is of little hemodynamic consequence, it is frequently associated with other anomalies. Isolated PLSVC is of diagnostic importance. It is noteworthy that a transthoracic echocardiogram combined with agitated saline injections through left and right antecubital veins provides a simple but elegant study for a rapid bedside diagnosis.\(^2,3\) The role of noninvasive approaches in the diagnosis of PLSVC was summarized in a previous study.\(^3\) Isolated PLSVC has practical clinical implications during central vein catheter placement, cardiac pacemaker lead insertion, cardiac resynchronization therapy (left ventricle pacing lead positioned in the left posterolateral vein of the heart), cardiopulmonary bypass surgery (retrograde cardioplegia through the PLSVC causing ineffective myocardial perfusion), and heart transplantation. The ligament of Marshall is known to be associated with the genesis of atrial arrhythmias.\(^4\) Both PLSVC and agenesis of the right SVC are related to anatomic abnormality and cardiac electric instability of the sinus node and conduction system.\(^5,6\) The dilated coronary sinus can stretch and fragmentize the conduction tissue.

**Disclosures**

None.

**References**

Concomitant Persistent Left Superior Vena Cava and Agenesis of Right Superior Vena Cava: A Rare Congenital Anomaly
Wei Xiong and Chanjuan Shi

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