Persistent left superior vena cava (PLSVC) is the most common thoracic systemic venous drainage anomaly, occurring in 0.5% of the general population and in 10% of patients with other congenital cardiac anomalies. Anomalous connection of the right superior vena cava (RSVC) to the left atrium (LA) is extremely rare.

**Case History**

A 40-year-old man presented with history of recent-onset dyspnea, light-headedness, and presyncope on exertion. Physical examination revealed a baseline pulse rate of 42 bpm, blood pressure of 110/70 mm Hg with the absence of postural hypotension, cyanosis, clubbing, and room air oxygen saturation of 96%. Cardiovascular system examination was unremarkable. ECG showed sinus bradycardia, and chronotropic incompetence was demonstrated on an exercise test. A 24-hour ECG Holter showed significant sinus pauses and confirmed the diagnosis of symptomatic sinus node dysfunction, so a permanent pacemaker implantation was planned. A left upper-limb venogram showed the PLSVC draining into the right atrium (RA) via a hugely dilated coronary sinus (CS; Figure 1A and Movie I in the online-only Data Supplement). Because lead placement via the PLSVC is technically challenging, the procedure was planned via the RSVC. The ventricular pacing lead introduced through RSVC, however, went inadvertently into the left ventricle, with left ventricular pacing spikes on the surface ECG. Backflush of contrast in the right side of the heart. Backflush of contrast in the right superior pulmonary vein also could be seen (Figure 3A and 3B and Movies II and III in the online-only Data Supplement). This confirmed the anomalous drainage of the RSVC into the LA and the absence of any atrial septal defect because contrast did not enter the RA. This RSVC anomaly on hindsight was missed on transthoracic echocardiography, which otherwise showed a structurally normal heart. After confirmation of anomalous drainage of the RSVC to the LA, the procedure was switched back through the challenging PLSVC approach.

The ventricular lead was maneuvered to form a loop on the RA free wall and, with the help of reshaped stylet (L-shaped configuration), was advanced through the tricuspid orifice toward the interventricular septum. The lead was then prolapsed and positioned at the lower part of the interventricular septum, and an active fixation helix was deployed. The atrial lead was similarly positioned and screwed to the RA free wall (Figure 4A and 4B), and a dual-chamber permanent pacemaker was successfully implemented. Chest computed tomography angiogram (Figures 5A, 5B, and 6 and Movie IV in the online-only Data Supplement) confirmed drainage of the RSVC into the LA, whereas the PLSVC with a pacemaker lead in situ was seen draining into the RA via the CS.

This is an extremely rare thoracic systemic venous anomaly of the RSVC draining into the LA and PLSVC draining into the RA with symptomatic sinus node dysfunction presenting as a challenging surprise during pacemaker lead placement.

**Discussion**

During the fourth week of gestation, a pair of anterior cardinal veins form that drain the cephalic part of the embryo and a pair of posterior cardinal veins form that drain the rest. Anterior and posterior cardinal veins join to form a short common cardinal vein (duct of Cuvier), which opens into the respective horns of sinus venosus. The proximal portions of the right anterior cardinal veins and right common cardinal vein form the RSVC. The left anterior cardinal veins normally obliterate to form the ligament of Marshall, whereas the left horn of the sinus venosus and adjacent portion of the left common cardinal vein form the CS. Persistence of the left anterior cardinal vein forms the left SVC, which always continues with the CS as the left common cardinal vein that is part of both the CS and left SVC. Flow of PLSVC blood into the CS results in its enlargement, and a dilated CS is often the first clue for the diagnosis of PLSVC during echocardiography. In 82% to 90% of cases, PLSVC is associated with bilateral SVC; isolated PLSVC is occasionally seen in association with situs inversus or dextrocardia. In the absence of any other cardiac malformation, PLSVC to the RA is an incidental finding, but it
may have an impact on procedures that require left upper-limb venous access such as transvenous pacemaker lead implantation. Similarly, cannulation of the heart for cardiopulmonary bypass may result in ineffective retrograde cardioplegia. The earliest reported case in the English literature of LA drainage of the RSVC was described by Wood, and at least 18 additional cases have been published in English since then. It is believed that a deficiency of the wall between the RSVC and right superior pulmonary vein results in shunting of RSVC blood into the LA via the LA orifice of the right superior pulmonary vein, and if this shunt predominates in fetal life, blood flow to the RA through the RA orifice of the SVC will be diminished or completely eliminated, leading to SVC orifice atresia and eventually RSVC draining only into the LA. A patient with RSVC draining into the LA may present with symptoms of hypoxemia, decreased effort tolerance, cyanosis, and clubbing, depending on degree of right-to-left shunt, which was absent in our case; as greater volume of venous blood probably returns by way of the inferior vena cava and PLSVC into the RA and was normally oxygenated, thus minimizing the degree of right to left shunting. Abnormal development of the right horn of the sinus venosus and right anterior cardinal vein may jeopardize the normal development of the sinoatrial node and result in abnormal sinus node function. In the presence of PLSVC, placing a stable pacemaker lead in the RV through the CS can be difficult because the tip of the lead is deflected away from the tricuspid orifice. Reshaping the stylet tip into 3- to 4-cm-wide closed pigtail loop with U- or L-shaped bends, depending on the geometry and shape of heart, may help in maneuvering of the lead into the RV. In the absence of symptoms of right-to-left shunt in the present case, surgical diversion of RSVC blood into the RA was deferred.

Disclosures
None.

References
Figure 3. A, Anteroposterior projection of a right superior vena cava (RSVC) venogram showing contrast injected into the RSVC seemingly flowing into the left atrium (LA) and left ventricle (LV). Backflush of contrast into the right superior pulmonary vein (RSPV) can be seen. Contrast has not entered the right side of the heart. B, An RSVC venogram in the left anterior oblique projection showing contrast injected into RSVC entering the posterior chambers of the heart, ie, the LA and LV. RA indicates right atrium; and RV, right ventricle.

Figure 4. A, Fluoroscopy in the right anterior oblique projection showing the ventricular lead introduced through the persistent left superior vena cava and coronary sinus entering the right atrium (RA) and forming a loop on the RA free wall before entering the right ventricle (RV). The atrial lead was screwed to the RA free wall. B, Fluoroscopy in the left anterior oblique projection showing the direction of the ventricular lead, from the posterior to the anterior and then leftward, almost on the same horizontal plane.
Figure 5. A, Chest computed tomography (CT) angiogram, volume-rendered technique, showing a right superior vena cava (SVC) draining into the left atrium near the opening of the right superior pulmonary vein. B, Chest CT angiogram, volume-rendered technique, showing persistent left SVC entering the right ventricle via the coronary sinus with the pacemaker lead in situ.

Figure 6. Chest computed tomography angiogram, maximum-intensity projection, showing the right superior vena cava (RSVC) draining into the left atrium (LA). Reflux of contrast from the LA into the RSVC can be seen.
Bilateral Superior Venae Cavae With Crisscross Atrial Drainage
Sushil P. Tripathi, Ashish A. Nabar, Prafulla G. Kerkar, Hemant B. Telkar and Ashlesha S. Udare

Circulation. 2015;132:e365-e368
doi: 10.1161/CIRCULATIONAHA.115.018898

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2015 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/132/23/e365

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2015/12/07/CIRCULATIONAHA.115.018898.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/
Movie Legend

**Movie 1.** LSVC venogram in Left anterior oblique (LAO) projection showing contrast injected in LSVC entering RA via dilated CS. A pacing leads passed via RSVC seen entering LA and LV while pacing catheter passed via IVC seen entering CS. Best viewed with Windows Media Player.

**Movie 2.** Anteroposterior (AP) projection; RSVC venogram showing contrast injected in RSVC flowing seemingly into LA and LV, also backflush of contrast into RSPV can be seen. Contrast has not entered right side of the heart. Best viewed with Windows Media Player.

**Movie 3.** RSVC venogram in LAO projection showing contrast injected in RSVC entering the posterior chambers of the heart; LA and LV. Contrast has not entered in RA or RV, confirming LA drainage of right SVC. Best viewed with Windows Media Player.

**Movie 4.** Axial CECT images post intravenous contrast injection from lower limb showing right SVC draining into left atrium and persistent LSVC opening into RA via coronary sinus. Best viewed with Windows Media Player.