

## Incidentally Detected “Silent” Vena Cava Superior Anomalies

Keser Nurgül,<sup>1,2</sup> Dilmen Serkan,<sup>1</sup> Işılak Zafer,<sup>1</sup> Uzun Mehmet,<sup>1</sup> Hayıroğlu İlker Mert<sup>1</sup> and Orhan Ahmet Lütfullah<sup>1</sup>

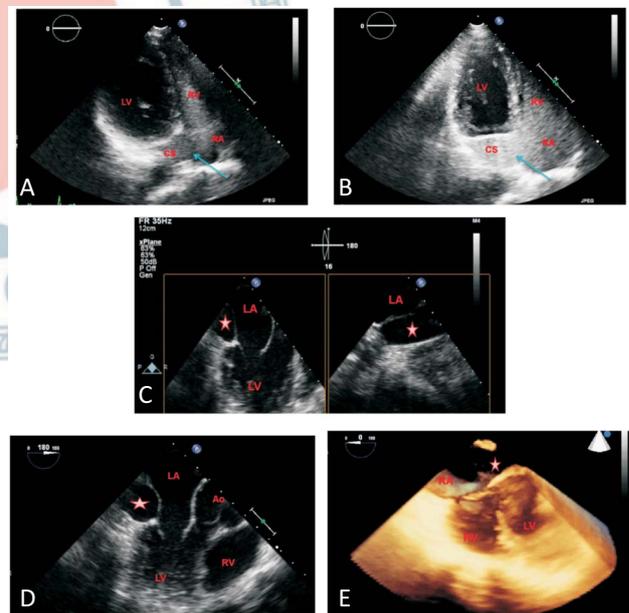
### INTRODUCTION

Presently described is the case of a 27-year-old asymptomatic man diagnosed incidentally with persistent left superior vena cava (PLSVC) with an absent right superior vena cava (aRSVC). It was observed that the coronary sinus (CS) was almost 2 cm in diameter, and an agitated saline injection (SI) administered to the antecubital veins in both arms led to the diagnosis of the combined anomaly, which was confirmed with transesophageal echocardiography (TEE) and a thorax computed tomography (CT) imaging with contrast.

### CASE

A 27-year-old man with no complaints or known disease presented at the outpatient clinic for a checkup program. The physical evaluation, routine laboratory tests, and electrocardiography results were completely normal. Transthoracic echocardiography (TTE) results were also normal, but revealed a dilated CS (1.8 cm) in the left parasternal and modified apical 4-chamber views, which prompted the use of contrast echocardiography with an SI. A standard injection from the left antecubital vein (LAV) showed fast filling of the CS followed by right atrial (RA) filling as well as the presence of a PLSVC (Figure 1A). Since the CS was so dilated, the examination was also performed from the right antecubital vein (RAV), where contrast appeared slowly in the CS and RA (Figure 1B). TEE was performed for further evaluation. The di-

lated CS could be visualized from the mid-esophageal 2C view (Figures 1C-E) and filling was confirmed after a contrast injection from the LAV (Figure 2A). The bicaval position revealed the aRSVC (Figure 2B). Contrast failed to fill the usual location of the RSVC. Although PLSVC is generally noticed at the left atrial appendage border, TEE did not offer clear differentiation. The patient was referred for a thorax CT with contrast imaging, which confirmed the aRSVC with PLSVC and an opening into the CS, resulting in extreme dilatation (Figures 2C, 2D). When CS dilatation is seen on TTE, SI from the LAV is generally performed; however, the need to perform the test in both arms should be kept in mind so as not to omit any other coexisting abnormalities. This case re-

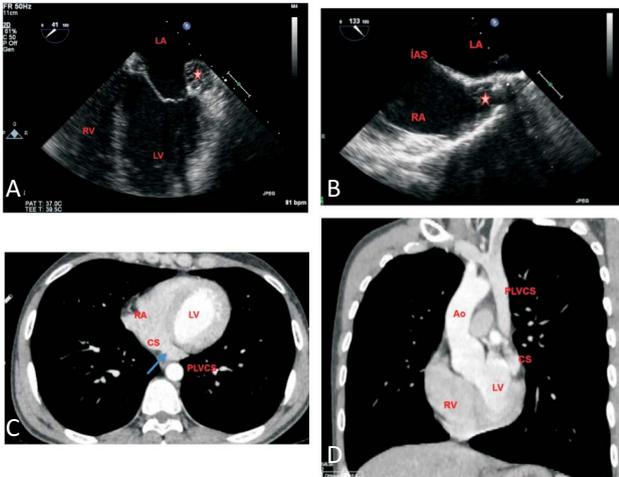


**Figure 1.** (A) A standard injection from the left antecubital vein (LAV) showed fast filling of the coronary sinus (CS) followed by right atrial (RA) filling, as well as the presence of a persistent left superior vena cava (PLSVC). (B) Contrast injected from the right antecubital vein (RAV) appeared slowly in the coronary sinus (CS) and right atrium (RA). (C, D, E) The dilated coronary sinus (CS) could be visualized from the mid-esophageal 2C view of transesophageal echocardiography (TEE).

Received: February 24, 2019 Accepted: May 27, 2019

<sup>1</sup>Department of Cardiology, Istanbul Sultan Abdülhamit Han Research and Training Hospital; <sup>2</sup>Faculty of Medicine, University of Sakarya.

Corresponding author: Dr. Mert İlker Hayıroğlu, Department of Cardiology, Istanbul Sultan Abdülhamit Han Research and Training Hospital, Üsküdar; Istanbul, Turkey. E-mail: mertilkerh@yahoo.com



**Figures 2.** (A) Filling was confirmed after a contrast injection from the left antecubital vein (LAV). (B) The bi-caval position revealed the absence of the right superior vena cava (aRSVC). (C, D) Thorax computed tomography (CT) with contrast imaging that confirmed the absence of the right superior vena cava (aRSVC) with persistent left superior vena cava (PLSVC) and an opening into the coronary sinus (CS), resulting in extreme dilatation.

port illustrates the need for a detailed examination to identify any coexisting anomalies in the presence of a dilated CS in order to avoid potential future complications, such as in the event of an emergent surgical or interventional procedure using the central venous route, like catheter placement or pacemaker implantation.

## DISCUSSION

PLSVC is one type of congenital venous development anomaly. It has been reported to have an incidence of 0.3% to 0.5% in the general population.<sup>1</sup> In patients with coexisting congenital heart diseases (CHD), the incidence of PLSVC may reach 4.3%.<sup>2</sup> Failure of the left anterior cardinal vein to close during embryogenesis results in PLSVC, which generally drains into the CS. PLSVC with aRSVC occurs when the right anterior cardinal vein is obliterated, a rare abnormality observed in 10% to 20% of PLSVC patients.<sup>2</sup> In patients with CHD, the incidence of PLSVC with aRSVC has been reported to be around 0.09% to 0.13%.<sup>1</sup> Bartram et al.<sup>3</sup> reported that the most common forms of CHD in which these combined venous anomalies were seen were various types of atrial septal defects, endocardial cushion defects, and tetralogy of Fallot. Although generally asymp-

tomatic, 36% of cases of PLSVC with aRSVC have various rhythm disorders that may necessitate close follow-up.<sup>1,3,4</sup> The possible underlying reason for some of these rhythm problems might be the stretching of the impulse formation and transmission routes by a dilated CS in some cases, while in others it might be related to the left pacemaker area. While the cells of the right pacemaker area form a sinoatrial node (SN) during embryogenesis, the fate of those on the left is as yet unknown.<sup>2,5</sup>

In various studies,<sup>2,5</sup> dysfunction of a SN has been observed in patients with aRSVC. In a case series presented by Morgan et al.,<sup>6</sup> sick sinus syndrome and atrial fibrillation originating from PLSVC were reported to be the main heart rhythm problems in these patients. Another important concern is if the PLSVC drains into the LA, as seen in almost 10% of PLSVC cases, as it creates a partial anomalous systemic venous return with right-to-left shunting that can result in cyanosis and paradox embolism.<sup>3,5</sup>

The classic finding leading to the suspicion of PLVCS is the presence of a dilated CS observed on TTE. This necessitates agitated saline infusion into the LAV, which first appears in the CS, followed by filling of the RA if PLSVC exists.<sup>2</sup> Besides, agitated saline injection has an important role in identifying congenital anomalies.<sup>7</sup> If aRSVC accompanies the PLSVC, then the contrast flows into the CS when injected from both arms, as seen in our case. Since the role of TEE in the identification of the coexistence of venous anomalies is well defined, TEE imaging was used in this case to supplement the diagnosis.<sup>8</sup> Mid-esophageal 2 and 4C views with slight retroflexing of the probe clearly revealed the dilated CS. Bicaval views exposed the aRSVC. However, the PLSVC couldn't be depicted easily, possibly due to the enlarged CS. To evaluate the location of the PLSVC and in order not to miss any other anomaly, a thorax CT was performed. This is among the various recommended imaging modalities, which also includes venous angiography and magnetic resonance imaging.<sup>3</sup> CT images demonstrated the opening of the PLSVC into the CS nicely without any other additional coexisting congenital anomalies. Patients with these anomalies are frequently asymptomatic and therefore, the diagnosis is often made incidentally when interventional procedures are performed via the central venous route, such as a central venous catheter insertion or pacemaker implantation, which

may lead to catastrophic complications.<sup>3</sup> A dilated CS should bring the possibility of PLSVC to mind, and a large dilatation should suggest that there may be accompanying aRSVC, which warrants SI from both arms. If TEE is suboptimal, thorax CT may provide additional information.

### LEARNING POINTS

1. The classic finding leading to the suspicion of PLVCS is the presence of a dilated CS observed on TTE. This necessitates agitated saline infusion into the LAV, which first appears in the CS, followed by filling of the RA if PLSVC exists.
2. Patients with these anomalies are frequently asymptomatic, the diagnosis is often made incidentally when interventional procedures are performed via the central venous route, such as a central venous catheter insertion or pacemaker implantation, which may lead to catastrophic complications.
3. Our case illustrates the need for a detailed examination to identify any coexisting anomalies in the presence of a dilated CS in order to avoid potential future complications.

### ACKNOWLEDGEMENT

None.

### DECLARATION OF INTEREST

The authors declare that there are no conflicts of interest.

### REFERENCES

1. Martinez-Villar M, Gran F, Ferrer Q, et al. Persistent left superior vena cava with absent right superior vena cava. *Rev Esp Cardiol (Engl Ed)* 2016;69:220-1.
2. Yilmaz AT, Arslan M, Demirkiliç U, et al. Partially unroofed coronary sinus syndrome with persistent left superior vena cava, absent right superior vena cava and right-sided pericardial defect. *Eur J Cardiothorac Surg* 1996;10:1027-9.
3. Pálincás A, Nagy E, Forster T, et al. A case of absent right and persistent left superior vena cava. *Cardiovasc Ultrasound* 2006;26:6.
4. Sheik AS, Mazhar S. Persistent left superior vena cava with absent right superior vena cava: review of the literature and clinical implications. *Echocardiography* 2014;31:674-9.
5. Kusaka Y, Sawai T, Nakahira J, et al. Persistent left superior vena cava with absent right superior vena cava detected during emergent coronary artery bypass grafting surgery. *JA Clin Rep* 2015; 1:2.
6. Morgan DR, Hanratty CG, Dixon LJ, et al. Anomalies of cardiac venous drainage associated with abnormalities of cardiac conduction system. *Europace* 2002;4:281-7.
7. Liao CS, Hsiao JK, Chu IT. Pulmonary arteriovenous fistula with pulmonary hypertension - to close or not to close? *Acta Cardiol Sin* 2016;32:116-9.
8. Konecky N, Freedberg RS, McCauley D, et al. Absent right and persistent left superior vena cava without other congenital anomaly: a rare combination diagnosed by transesophageal echocardiography. *J Am Soc Echocardiogr* 1995;8:761-6.