

Isolated Persistent Left Superior Vena Cava: A Case Report and its Clinical Implications

Abstract

The venous anomaly of a persistent left superior vena cava (PLSVC) affects 0.3%–0.5% of the general population. PLSVC with absent right superior vena cava, also termed as “isolated PLSVC,” is an extremely rare venous anomaly. Almost half of the patients with isolated PLSVC have cardiac anomalies in the form of atrial septal defect, endocardial cushion defects, or tetralogy of Fallot. Isolated PLSVC is usually innocuous. Its discovery, however, has important clinical implications. It can pose clinical difficulties with central venous access, cardiothoracic surgeries, and pacemaker implantation. When it drains to the left atrium, it may create a right to left shunt. In this case report, we present the incidental finding of isolated PLSVC in a patient who underwent aortic valve replacement. Awareness about this condition and its variations is important to avoid complications.

Keywords: *Absent right superior vena cava, embryology, persistent left superior vena cava, transesophageal echocardiography*

Introduction

The congenital variations in the venous system are no exception in the gamut of anatomical malformations of various organs in the human body. The venous anomaly of a persistent left superior vena cava (PLSVC) is the most common congenital malformation of the thoracic venous system. It affects 0.3%–0.5% of the general population, and this incidence is up to 10% of those with congenital heart disease (CHD).^[1,2] “Isolated PLSVC” is an extremely rare venous anomaly. It occurs in 0.09%–0.13% of patients who have congenital heart defects with visceratrial situs solitus.^[3] It can be associated with situs inversus. Almost half of the patients with isolated PLSVC have cardiac anomalies in the form of atrial septal defect (ASD), endocardial cushion defects, or tetralogy of Fallot (TOF).^[4] Isolated PLSVC is usually innocuous; however, it can pose clinical difficulties with central venous access, cardiothoracic surgeries, and pacemaker implantation. When it drains to the left atrium (LA), it may create a right to left shunt.

Case Report

A 43-year-old man was referred for valvular surgery. Clinical examination findings were

consistent with severe aortic stenosis (AS). On auscultation, a mid-systolic ejection murmur was heard best over the “aortic area” with radiation into the right neck. Echocardiography revealed severe rheumatic AS and good biventricular function. Chest radiograph showed clear lung fields. The biochemical investigations were within normal limits.

The patient was premedicated the previous night, and anesthesia was induced with midazolam 0.1 mg/kg and fentanyl 10 µg/kg. The trachea was intubated with vecuronium bromide 0.1 mg/kg. After induction of general anesthesia, a central venous catheter (CVC) was inserted through right internal jugular vein (RIJV) under ultrasound guidance. Standard monitoring parameters were used along with transesophageal echocardiograph (TEE). Intraoperative TEE findings were consistent with the preoperative echocardiography findings. However, bicaval TEE view showed neither the distal end of CVC nor the right superior vena cava (RSVC) [Figure 1]. With curiosity as we searched for the left superior vena cava (LSVC) on TEE, we found a dilated coronary sinus (CS) [Video 1]. On agitated saline injection into the RIJV

**Samarjit Bisoyi,
Usha Jagannathan,
Anjan Kumar Dash,
Sabyasachi
Tripathy,
Raghunath
Mohapatra¹,
Naba Kumar
Pattnaik¹,
Satyajit Sahu¹,
Debashish Nayak¹**

*From the Department of Cardiac Anesthesiology,
¹Department of Cardiothoracic and Vascular Surgery, Apollo Hospitals, Bhubaneswar, Odisha, India*

Address for correspondence:

*Dr. Samarjit Bisoyi, Plot No. 360/7, Sishu Vihar, Patia, Bhubaneswar - 751 024, Odisha, India.
E-mail: dr_sbisoyi@rediffmail.com*

Videos Available on:
www.annals.in

Access this article online

Website: www.annals.in

DOI: 10.4103/0971-9784.197847

Quick Response Code:



How to cite this article: Bisoyi S, Jagannathan U, Dash AK, Tripathy S, Mohapatra R, Pattnaik NK, *et al*. Isolated persistent left superior vena cava: A case report and its clinical implications. *Ann Card Anaesth* 2017;20:104-7.

Received: November, 2016. **Accepted:** November, 2016.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

through the CVC, the flow of echo contrast was visible in the dilated CS [Video 2]. The flow of the contrast was first noted in the LSVC [Figure 2], CS, and then in the right atrium (RA). Figure 2 (TEE mid-esophageal view) shows the echo contrast in the LSVC lying just lateral to the LA. During surgery, it was observed that the RSVC was absent [Figure 3]. The LSVC joined by the right brachiocephalic vein was large and prominent [Figure 4]. The aortic valve replacement was performed with a two-stage venous cannula through the RA for the venous drainage to the cardiopulmonary bypass reservoir. The bypass duration was 87 min, and aortic cross-clamp duration was 67 min. The postoperative course was uneventful. In the postoperative chest X-ray [Figure 5], the course of CVC was from RIJV through the innominate vein to the LSVC.

Discussion

Isolated PLSVC is a very rare venous malformation. PLSVC affects 0.3%–0.5% of the general population and in up to 10% of those with CHD. Isolated "PLSVC" occurs in only 0.09%–0.13% of patients who have CHD. Almost

half of the patients with isolated PLSVC have cardiac anomalies in the form of ASDs, endocardial cushion defects, or TOF.

Developmental anomaly

During normal development of the RA, the sinoatrial (SA) chamber differentiates into the sinus venosus and the atrial chamber. The right and left anterior cardinal veins drain the cranial regions into the respective ducts of Cuvier, which in turn drain into the sinus venosus. The sinus venosus is eventually absorbed into the structure of the RA, with the bridging connection between the anterior cardinal veins forming the left innominate vein. The right cardinal vein and right duct of Cuvier form the SVC, whereas the left cardinal vein typically obliterates leaving the CS and the ligament of Marshall [Figure 6]. Failure of the closure of the left anterior cardinal vein results in PLSVC.^[5] In general, PLSVC is associated with RSVC and drains into the RA through a dilated CS. When developmental arrest occurs at an earlier stage, the CS is absent and the PLSVC drains into the LA. Either isolated or associated

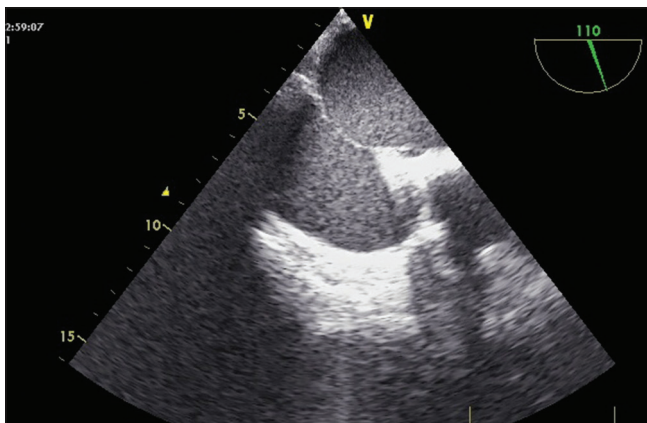


Figure 1: Bicaudal view on transesophageal echocardiograph showing absent right superior vena cava

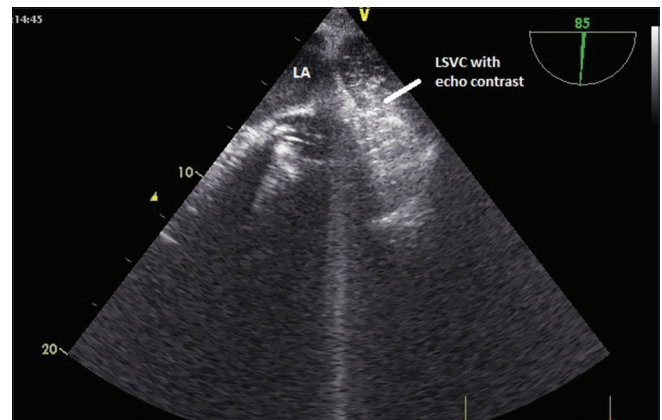


Figure 2: Mid-esophageal view on transesophageal echocardiograph. Echo contrast in the left superior vena cava located lateral to the left atrium, after injection of agitated saline into the RIJV. RIJV: Right internal jugular vein

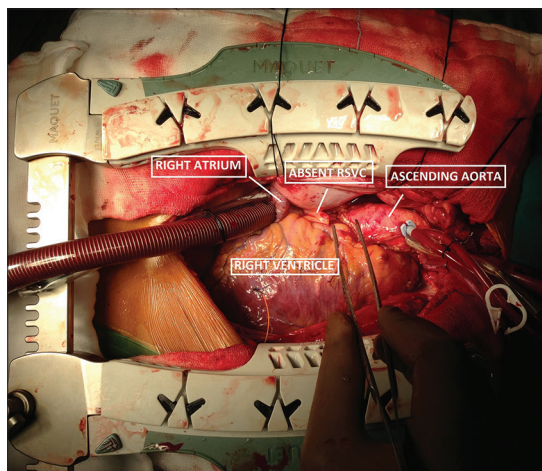


Figure 3: Surgical field showing absent right superior vena cava

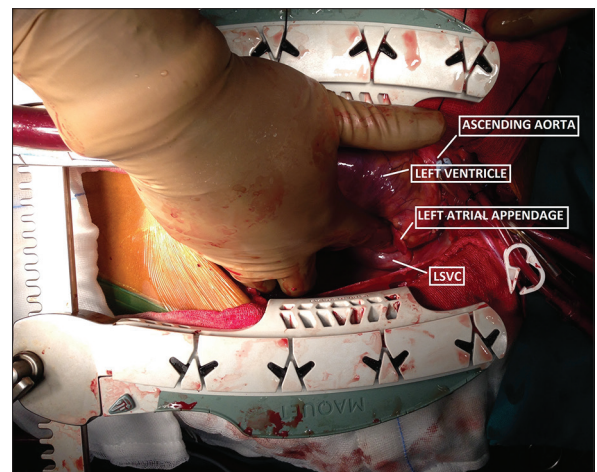


Figure 4: The heart is turned to right. Left superior vena cava in the operative field

with RSVC, this venous malformation itself causes no hemodynamic disturbance and is diagnosed incidentally. When the isolated LSVC is encountered, dextrocardia or complete situs inversus may be present.

Course of left superior vena cava

The LSVC usually descends vertically, anterior, and to the left of the aortic arch and main pulmonary artery [Figure 7]. It runs adjacent to the LA before turning medially, piercing the pericardium to run in the posterior atrioventricular (AV)

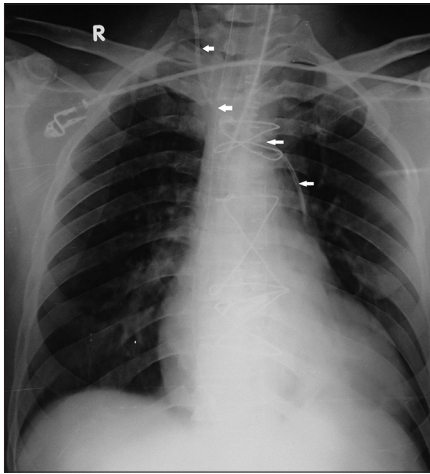


Figure 5: Chest X-ray showing the course (arrows) of central venous catheter

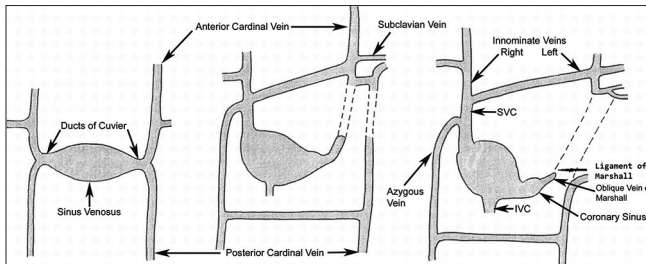


Figure 6: Development of vena cavae

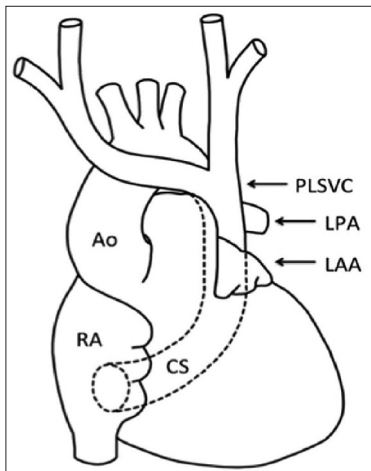


Figure 7: Diagram shows the course of isolated persistent left superior vena cava

groove.^[6] In about 90% of cases, it drains into the CS; alternative sites include the inferior vena cava, hepatic vein, and LA. The entry into LA is invariably associated with an ASD.^[7,8]

Clinical implications

- Central venous catheterization may be difficult due to relatively tortuous course [Figure 5]. Manipulation of guide wire (due to the proximity to CS) can cause hemodynamic instability, arrhythmias, perforation of heart, and tamponade^[9]
- Placement of pacemaker leads and pulmonary artery catheter can be arduous due to the same reason.^[10] During leads placement for a permanent pacemaker in such cases, the left subclavian vein is the preferred route. There is an acute angle between the CS ostium and the tricuspid valve; therefore, the lead should be looped in the RA to enter the right ventricle^[11]
- Cardiac surgeries involving cannulation of SVC for cardiopulmonary bypass require isolated LSVC cannulation whenever it is detected. However, it does not allow the use of retrograde cardioplegia as it may be ineffective. It may be possible to clamp the PLSVC to prevent the cardioplegia solution from perfusing retrograde up the PLSVC and its branches with inadequate myocardial protection^[12]
- Arrhythmias and conduction abnormalities are common in the presence of PLSVC. This could be due to a dilated CS which stretches the AV nodal tissue and predisposes to reentrant tachycardia. The other reason could be due to the SA node dysfunction. The specialized pacemaker cells which eventually evolve to form the SA node originate in the sinus venosus.^[13] Abnormal development of the right horn of the sinus venosus and right superior cardinal vein may jeopardize the normal development of the SA node and result in abnormal sinus node function^[14-16]
- Associated congenital cardiac anomalies should be searched for in patients with PLSVC. There is a high incidence of ventricular septal defect, ASD, endocardial cushion defect, CS ostial atresia, coarctation of aorta, cor triatriatum, and TOF in patients with isolated PLSVC. In patients with PLSVC Postema *et al.* found, confirmed syndromes were present in 42% of patients including VACTERL association (vertebral defects, anal atresia, cardiac malformations, tracheoesophageal fistula with esophageal atresia, radial and renal dysplasia, and limb anomalies), trisomy 21, 22q11, and CHARGE association (coloboma, heart defects, atresia of choanae, retardation, genital and ear anomalies)^[4,17]
- Right to left (R→L) shunt may occur when a PLSVC drains into the LA either directly or through an unroofed CS. This increases the risk of paradoxical embolism. Drugs administered intravenously (IV) bypass the right heart and reach the systemic circulation directly in the presence of isolated PLSVC. In 10% of patients,

a PLSVC may drain into LA.^[18] A large R→L shunt requires surgical correction

- ECG may show left axis deviation of the *P* wave with shortened PR interval. Chest X-ray may show a crescent-shaped shadow of the PLSVC at the aortic knuckle or left upper mediastinum. TEE shows a large CS (normal diameter of the CS <1 cm).^[19] The mid-esophageal view on TEE shows the PLSVC near the LA appendage and left pulmonary vein. In the bicaval view, the absence of RSVC is evident
- IV-agitated saline injection in the upper limb normally opacifies the RA. In isolated PLSVC, as in our case, the CS opacifies first before the RA
- Cardiac surgeries involving opening of right heart chamber require draining of the isolated PLSVC by a separate venous cannula.

Isolated PLSVC is a very rare condition. It is usually asymptomatic and is an incidental finding on either imaging or during surgery. Its discovery, however, has important clinical implications. Clinicians in many fields should be well aware of its variations and management techniques to avoid complications. The presence of a dilated CS should always alert us to search for LSVC.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Danielpour PJ, Aalberg JK, El-Ramey M, Sivina M, Wodnicki H. Persistent left superior vena cava: An incidental finding during central venous catheterization-a case report. *Vasc Endovascular Surg* 2005;39:109-11.
2. Edwards J, Du Shane J. Thoracic venous anomalies. *Arch Pathol* 1950;49:514-37.
3. Soward A, ten Cate F, Fioretti P, Roelandt J, Serruys PW. An elusive persistent left superior vena cava draining into left atrium. *Cardiology* 1986;73:368-71.
4. Bartram U, Van Praagh S, Levine JC, Hines M, Bensky AS, Van Praagh R. Absent right superior vena cava in visceratrial situs solitus. *Am J Cardiol* 1997;80:175-83.
5. Fischer DR, Zuberbuhler JR. Anomalous systemic venous return. In: Anderson RH, Baker E, McCartney RF, editors. *Paediatric Cardiology*. New York: Churchill Livingstone; 2002. p. 851-65.
6. Cormier MG, Yedlicka JW, Gray RJ, Moncada R. Congenital anomalies of the superior vena cava: A CT study. *Semin Roentgenol* 1989;24:77-83.
7. Campbell M, Deuchar DC. The left-sided superior vena cava. *Br Heart J* 1954;16:423-39.
8. Raghbi G, Ruttenberg HD, Anderson RC, Amplatz K, Adams P Jr, Edwards JE. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus; a developmental complex. *Circulation* 1965;31:906-18.
9. Azocar RJ, Narang P, Talmor D, Lisbon A, Kaynar AM. Persistent left superior vena cava identified after cannulation of the right subclavian vein. *Anesth Analg* 2002;95:305-7.
10. Recupero A, Pugliatti P, Rizzo F, Carerj S, Cavalli G, de Gregorio C, *et al.* Persistent left-sided superior vena cava: Integrated noninvasive diagnosis. *Echocardiography* 2007;24:982-6.
11. Hsiao HC, Kong CW, Wang JJ, Chan WL, Wang SP, Chang MS, *et al.* Right ventricular electrode lead implantation via a persistent left superior vena cava. An improved technique. *Angiology* 1997;48:919-23.
12. Oosawa M, Sakai A, Abe M, Hanayama N, Lin ZB, Kodera K. Repeat open heart surgery in a case associated with persistent left superior vena cava: A method of simple occlusion of L-SVC using an alternative extra-pericardial approach and retrograde cardioplegia. *Kyobu Geka* 1995;48:741-4.
13. Brooks CM, Lu HH. *The Sino-atrial Pacemaker of the Heart*. Springfield, Illinois: Charles C. Thomas; 1972.
14. Camm AJ, Dymond D, Spurrell RA. Sinus node dysfunction associated with absence of right superior vena cava. *Br Heart J* 1979;41:504-7.
15. Morgan DR, Hanratty CG, Dixon LJ, Trimble M, O'Keefe DB. Anomalies of cardiac venous drainage associated with abnormalities of cardiac conduction system. *Europace* 2002;4:281-7.
16. Lenox CC, Hashida Y, Anderson RH, Hubbard JD. Conduction tissue anomalies in absence of the right superior caval vein. *Int J Cardiol* 1985;8:251-60.
17. Postema PG, Rammeloo LA, van Litsenburg R, Rothuis EG, Hruđa J. Left superior vena cava in pediatric cardiology associated with extra-cardiac anomalies. *Int J Cardiol* 2008;123:302-6.
18. Uçar O, Pasaoglu L, Çiçekçioğlu H, Vural M, Kocaoglu I, Aydogdu S. Persistent left superior vena cava with absent right superior vena cava: A case report and review of the literature. *Cardiovasc J Afr* 2010;21:164-6.
19. Chan KL, Abdulla A. Images in cardiology. Giant coronary sinus and absent right superior vena cava. *Heart* 2000;83:704.