DOI: 10.7860/JCDR/2016/17785.7821

Case Report

Anatomy Section

Left-Sided Persistent Superior Vena Cava with Superior Hemiazygos Arch

LOKADOLALU CHANDRACHARYA PRASANNA[1], HUBAN R. THOMAS[2], ASWIN DAS[3], RAKESH KUMAR[4]

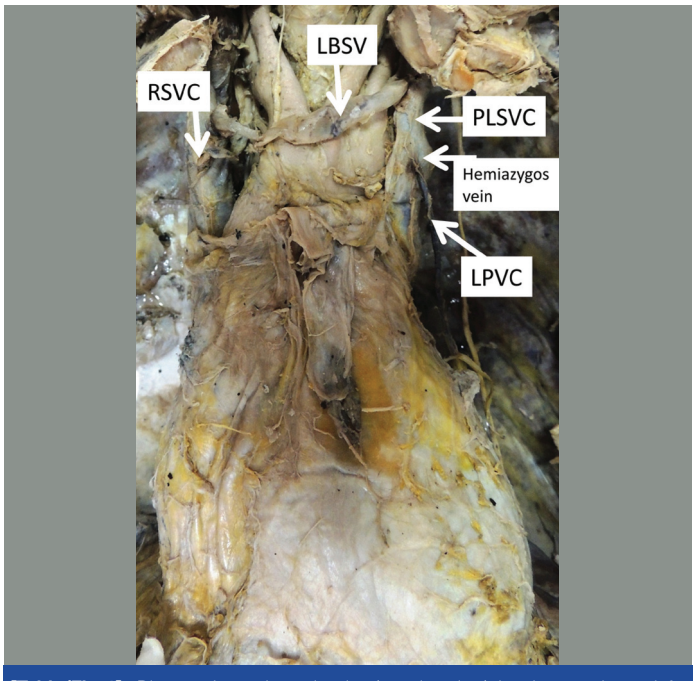
ABSTRACT

Persistent Left-side Superior Vena Cava (PLSVC) is the congenital central venous anomaly draining into the right atrium in 82-90% of cases via coronary sinus produce no haemodynamically significant consequences. In few cases it may drain into the left atrium directly or through the pulmonary veins resulting in right to left shunt. During routine undergraduate dissection, we found a PLSVC formed by the union of left subclavian and left internal jugular veins behind the left sternoclavicular joint was terminated into the right atrium through a larger coronary sinus ostium. Before its termination, PLSVC received a left hemiazygos vein formed by the continuation of the superior and inferior hemiazygos veins. A larger but thin communicating vein was noted between the right superior vena cava and PLSVC. Prior knowledge about such variations is essential in all the intervention procedures on right atrium through the left subclavian approach and also like in our case, the larger coronary sinus ostium if found during transthoracic echocardiography should be considered as an indication for the diagnosis of PLSVC.

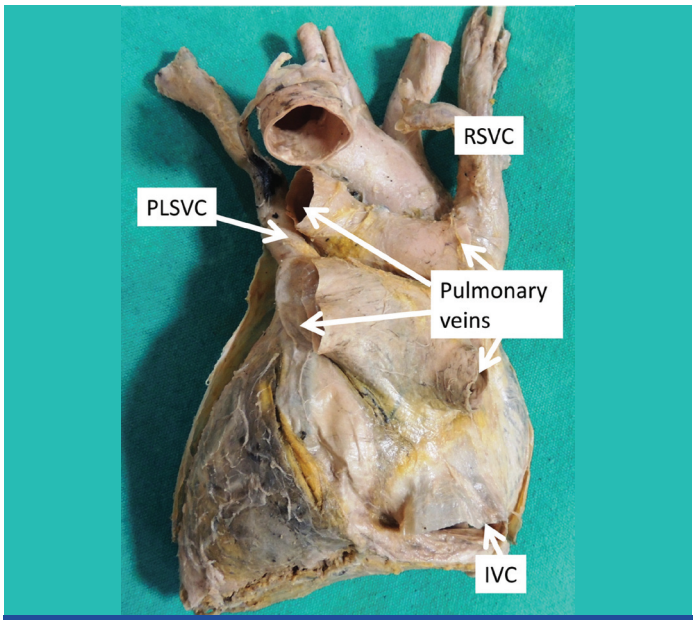
Keywords: Anteriorcardinal veins, Coronary ostium, Double superior vena cava

CASE REPORT

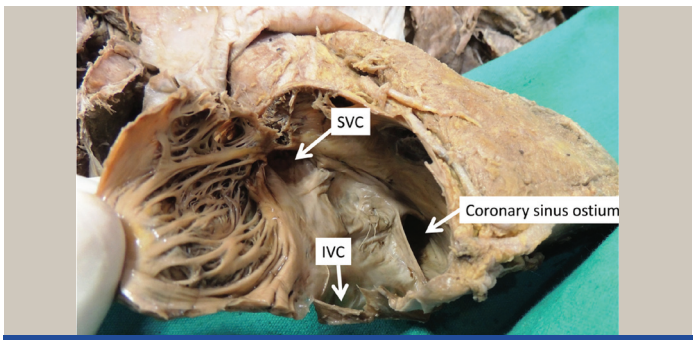
During routine cadaveric dissection for undergraduates in the Department of Anatomy, we encountered numerous systemic vascular anomalies in the thoracic cavity of an adult. We found a PLSVC formed by the union of left side internal jugular and subclavian veins behind the left sternoclavicular joint. It descends vertically, anterior and to the left side of the aortic arch and pulmonary trunk before its entry into the fibrous pericardium. Its caliber was approximaletly same as that of normal SVC on the right side [Table/Fig-1&2]. PLSVC then received a smaller Left Pericardiophrenic Vein (LPCPV) and a larger LPICV in the form of an arch over the root of the left lung. Inside the pericardium, PLSVC open into the coronary sinus after the latter has been formed by the greater cardiac vein and oblique vein of left atrium. Ultimately the coronary sinus terminate into the right atrium.Upon dissecting the interior of the right atrium, we noticed a larger coronary sinus ostium [Table/Fig-3] measuring 3.1 cm vertically and 2.6 cm from the sides, which is approximately twice the size of the superior



[Table/Fig-1]: Dissected specimen in situ (anterior view)showing persistent left-sided superior vena cava(PLSVC) receiving left superior intercostal vein (LSICV) as hemiazygos arch. Left brachiocephalic vein (LBSV) connecting PLSVC and normal right-side superior vena cava(RSVC).



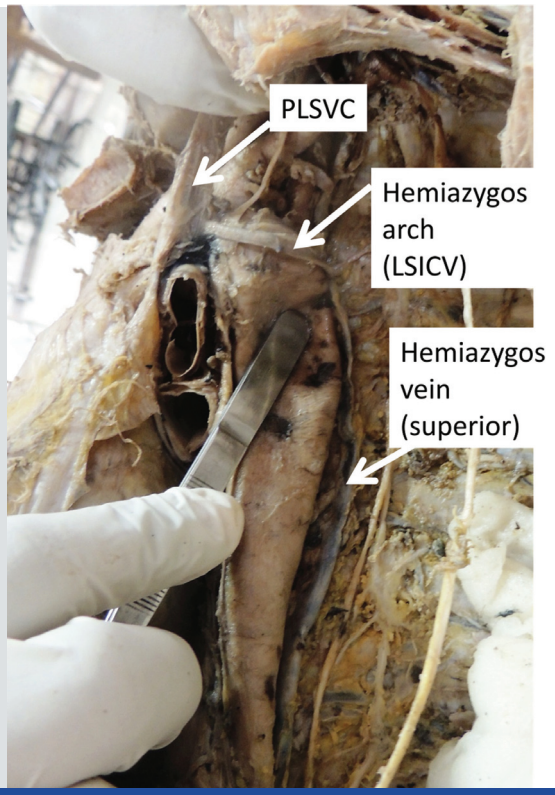
[Table/Fig-2]: Heart specimen (posterior view) showing the termination of PLSVC in the coronary sinus.



[Table/Fig-3]: Dissected specimen (interior of right atrium) showing larger coronary ostium. Compare the diameter of superior vena cava(SVC) and inferior vena cava (IVC).

vena cava and no other anomalies in other chambers of the heart. No septal defects and stenotic larger vessels were found.Though, a connecting vein was found between the two vena cavae representing the left brachiocephalic vein, its caliber was same as that of vena cavae and was very thin. No other anomalies were noted in major arteries except the left vertebral artery arising from the arch of aorta.

LPICV, when traced down continued with the accessory (superior) hemiazygos vein. Both superior and inferior hemiazygos veins continuous with each other without any change in their caliber throughout the posterior mediastinum [Table/Fig-4]. Also, both hemiazygos veins communicate with the azygos vein as two small venous channels in front of the body of 8th thoracic vertebrae.



[Table/Fig-4]: Oblique view of the thoracic cavity (thoracic aorta was retracted) to show the continuity of accessory and hemiazygos veins, which in turn continue with left superior intercostal vein (LSICV) as hemiazygos arch.

DISCUSSION

Congenital abnormalities of Superior Vena Cava (SVC) noted till date are double SVC, persistent left SVC, and a single left-sided SVC. Prevalence of Persistent Left-Sided SVC (PLSVC) is around 0.3-0.5% in general population and 4.3-11% in patients with congenital heart diseases [1,2]. Failure of normal regression of the left anterior cardinal vein caudal to the innominate (brachiocephalic) vein results in the PLSVC, draining the venous blood into right atrium through enlarged coronary sinus (in 92% cases) via vein of Marshall [3]. In 40% of cases, PLSVC can have associated cardiac anomalies like septal defects, coarctation of aorta, coronary ostium atresia, cortriatriatum and pulmonary stenosis [4].

In general Left Posterior Intercostal Vein (LPICV) forms a tributary of the left brachiocephalic vein and in 20% LPICV forms the hemiazygos arch and drain into the PLSVC [4].

Paired anterior cardinal veins drain the venous blood from the head and neck regions of the corresponding sides into the future right atrium. During 8th week of intra uterine life an oblique communication was established between the right and left indicating the future left brachiocephalic (innominate) vein. Fate of these anterior cardinal veins was different on both the sides. The caudal part of the right anterior cardinal vein forms the normal right-sided superior vena cava, while the part of the left anterior cardinal vein caudal to the brachiocephalic vein normally regress to form ligament of Marshall. Failure of this regression results in PLSVC that empties into coronary sinus [5,6].

The hemiazygos veins drains usually into the azygos system but in few it may directly open into the SVC or into the left brachiocephalic vein. In our case, a continuous hemiazygos channel comprising superior and inferior hemiazygos veins run in the posterior mediastinum with a small horizontal communications with the azygos vein at 8th thoracic vertebrae.

Variations reported about the PLSVC includes the presence or absence of connecting innominate vein between the right SVC and PLSVC, isolated left SVC, PLSVC with partial anomalous pulmonary venous return (PAPVR). Also, variations have been reported in the mode of termination into right or left atria. The former case is of no haemodynamic effects but the latter results in right to left sided shunt [3].

PLSVC in 90% of cases open into the right atrium via coronary sinus but the most dreaded complication is the opening of PLSVC into left atrium (in 10%). In the latter case, PLSVC lies between the left atrial appendage and the left pair of pulmonary vein giving an impression of lymphadenopathy in imaging, posing difficulty in central venous catheterization and also in systemic embolization of a thrombus or air [1].

The larger coronary ostium in our case could be due to PLSVC as the findings suggestive of elevated right atrial pressure, coronary atrio-ventricular fistula, and PAPVR could not be made out [3].

Available literatures correlate the finding of PLSVC shows other cardiac congenital anomalies in 40% of cases and those are more often if PLSVC with absent right side SVC [1,2]. In our case none of the anomalies were noted except the larger coronary ostium.

PLSVC has been associated with various practical considerations: 1) Abnormalities in both nodal and conducting tissues of the heart because of the dispersion of these tissues in the central fibrous body; 2) Placement of cardiac pacemakers and ICDs leading to arrhythmias, cardiogenic shock and coronary sinus thrombosis; 3) Accessing the right side heart and pulmonary vasculature through the left brachiocephalic vein as it is extremely thin in our case leading to major haemorrhage even in trivial trauma; 4) Posing problems in administering the cardioplegic solution in cardiac surgery because the uncontrolled blood flow in PLSVC into the coronary sinus would warm the heart if cardioplegic arrest is carried out. Finally, in cardiac transplantation in a patient with PLSVC, the coronary sinus must be isolated with caution to allow reanastomosis of PLSVC to right atrium [3,4,7].

The left superior intercostal vein (LSICV) drains the left second to fourth intercostal space terminating into the left brachiocephalic vein. In our case, the LSICV communicate with the accessory hemiazygos vein (which is common in 75% of patients). It then arches anterior to the aortic arch (appear as “aortic nipple” in chest x-ray) forming a left azygos arch and empty into PLSVC [6]. Azygos and hemiazygos continuation of Inferior Vena Cava (IVC) are associated with 0.2-1.3% of cardiac anomalies, polysplenia, abdominal situs, and left or duplicated inferior vena cava. No such anomalies were noted in our case [4,5,7].

CONCLUSION

Though detected incidentally and with no significant haemodynamics effects, knowledge of this variant is important in catheter placement when left subclavian vein approach is used to access the right atrium. Also, PLSVC can complicate placement of permanent pacemaker and implantable cardioverter defibrillator (ICD). Larger/dilated coronary sinus in transthoracic echocardiography should be considered as an indication for the diagnosis of PLSVC.

ETHICS STATEMENT

The study was performed in a manner to confirm with the Helsinki Declaration of 1975, as revised in 2000 and 2008 concerning Human and Animal Rights. Also, the authors followed the policy concerning informed consent as shown on publishing group.

REFERENCES

[1] Burney K, Young H, Barnard SA, McCoubrie P, Darby B. CT appearances of congential and acquired abnormalities of the superior vena cava. *Clinical Radiology*. 2007;62:837-42.

[2] Kim H, Kim JH, Lee H. Persistent left superior vena cava: diagnosed by bedside echocardiography in a liver transplant patient: a case report. *Korean J Anesthesiol*. 2014; 67(6):429–32.

[3] Goyal SK, Punnam SR, Verma G, Ruberg FL. Persistent left superior vena cava: a case report and review of literature. *Cardiovascular Ultrasound*. 2008;6:50.

[4] Singh B, Ramsaroop L, Maharaj J, Reddi A. Case of double superior vena cava. *Clinical Anatomy*. 2005;18:366–69.

[5] Irwin RB, Greaves M, Schmitt M. Left superior vena cava: revisited. *European Heart Journal – Cardiovascular Imaging*. 2012;13:284–91.

[6] Kurtoglua E, Cakinb O, Akcayc S, Akturkd E, Korkmaza H. Persistent left superior vena cava draining into the coronary sinus. *Cardiol Res*. 2011;2:249–52.

[7] Demos TC, Posniak HV, Pierce KL, Olson MC, Muscato M. Venous anomalies of the thorax. *AJR*. 2004;182:1139–50.

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Anatomy, Kasturba Medical College, Manipal University, Manipal, India.
2. Senior Grade Lecturer, Department of Anatomy, Kasturba Medical College, Manipal University, Manipal, India.
3. Postgraduate Scholar, Department of Anatomy, Kasturba Medical College, Manipal University, Manipal, India.
4. Postgraduate Scholar, Department of Anatomy, Kasturba Medical College, Manipal University, Manipal, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Lokadolalu Chandracharya Prasanna,
Associate Professor, Department of Anatomy, Kasturba Medical College, Manipal University,
Manipal-576104, India.
E-mail: anatomylcp@yahoo.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Nov 11, 2015**
Date of Peer Review: **Jan 07, 2016**
Date of Acceptance: **Feb 29, 2016**
Date of Publishing: **May 01, 2016**