

Persistent left superior vena cava and its clinical correlation - A cadaveric study



Sachendra Kumar Mittal¹, Rekha Parashar², Pankaj Kumar Singh³, Leena Jadon⁴

¹Assistant Professor, ²Associate Professor, ³Professor, Department of Anatomy, National Institute of Medical Sciences and Research, Jaipur, Rajasthan, India, ⁴Associate Professor, Department of Physiotherapy, Jaipur Hospital College of Physiotherapy, Rajasthan, India

Submission: 09-12-2020

Revision: 24-02-2021

Publication: 01-04-2021

ABSTRACT

Background: Presented is a case of persistent left superior vena cava draining into the right atrium through coronary sinus and finally opens into right atrium. Abnormalities of the vascular system are more commonly seen due to its importance in circulation. Persistent left superior vena cava is rare but important congenital vascular anomaly. It results when the left superior cardinal vein caudal to the innominate vein fails to regress. The venous anomaly of a persistent left superior vena cava (PLSVC) affects 0.3%–0.5% of the general population. Normally the superior vena cava is a single vascular structure formed by the union of right and left brachiocephalic veins which are in turn formed by the union of internal jugular and subclavian veins of corresponding side, draining the head and neck as well as the superior extremity. **Aims and Objective:** To evaluate the accuracy of persistent left superior vena cava and to find out the opening of PLSVC and formations of both SVC. **Materials and Methods:** During routine dissection of Thorax, we have opened the thoracic cage and take out the Heart. during that we found separate SVC and then we did the study on this PLSVC in the Department of Anatomy, Jaipur National University Institute for Medical Sciences and Research Centre (JNUIMSRC) Jaipur and National Institute of Medical Sciences and Research (NIMS & R). **Results:** We found persistent left superior vena cava in two cadavers out of 30 cadavers (6.66%) one was 64-year-old male cadaver and another 72-year-old male cadaver. Both the vena cavae were formed as of brachiocephalic veins of the corresponding side. The persistent left superior vena cava opened into the enlarged coronary sinus that drained into the right atrium between the opening of inferior vena cava and right atrio-ventricular orifice. **Conclusion:** It has important clinical implications in certain clinical interventions. It may complicate placement of cardiac catheters or pacemaker leads.

Key words: Persistent left superior vena cava; Coronary sinus; Superior vena cava; Right atrium

INTRODUCTION

Persistent left superior vena cava (PLSVC) is a rare venous abnormality. It is, however, the most common congenital anomaly of the thoracic venous system with a frequency of less than 0.5% of the general population and up to 10% of patients with congenital heart disease.¹⁻³ Normal anatomy describes the formation of a single superior vena cava by the union of right and left brachiocephalic veins which are in turn is formed by the union of corresponding internal jugular and subclavian

veins, draining the head and neck as well as the superior extremity.⁴ Double superior vena cava(SVC) with the persistent left superior vena cava(PLSVC) is a rare venous malformation. Patients with PLSVC may have other associated cardiac malformations such as atrial septal defect, ventricular septal defect or endocardial cushion defects.^{5,6} Presence of PLSVC may also interfere and cause problems during various invasive procedures such as pacemaker implantation, central venous catheterization, retrograde delivery of cardioplegia and retrograde left ventricular pacing.^{7,8}

Access this article online

Website:

<http://nepjol.info/index.php/AJMS>

DOI: 10.3126/ajms.v12i4.33321

E-ISSN: 2091-0576

P-ISSN: 2467-9100

Copyright (c) 2021 Asian Journal of Medical Sciences



This work is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

Address for Correspondence:

Dr. Rekha Parashar, Associate Professor, Department of Anatomy, National Institute of Medical Sciences and Research, Jaipur, Rajasthan, India, 302020, State Rajasthan, India. **Mobile:** +91-9649889333. **E-mail:** lineparashar1985@gmail.com

MATERIALS AND METHODS

During routine dissection in the department of Anatomy, JNUIMSRC, Jaipur from 2016 to 2019, and in the department of Anatomy, National Institute of Medical Sciences and Research (NIMS & R) from 2019 to 2020 we found in two cadavers (6.66%) among 30 cadavers double superior vena cava with persistent left superior vena cava (PLSVC) in a 64-year-old male cadaver and another 72 year old male cadaver there is no joining between left brachio-cephalic vein and right brachio-cephalic to forming a single superior vena cava, Both the vena cavae were formed as brachiocephalic veins of the corresponding sides in both the cadavers (Figure 1). The PLSVC was longer length compared to the superior vena cava in both the cadavers (Figure 2). When we traced, it opened into the enlarged coronary sinus that further drained into the right atrium between the opening of inferior venacava and right atrio-ventricular orifice in both cadavers (Figure 3). There was

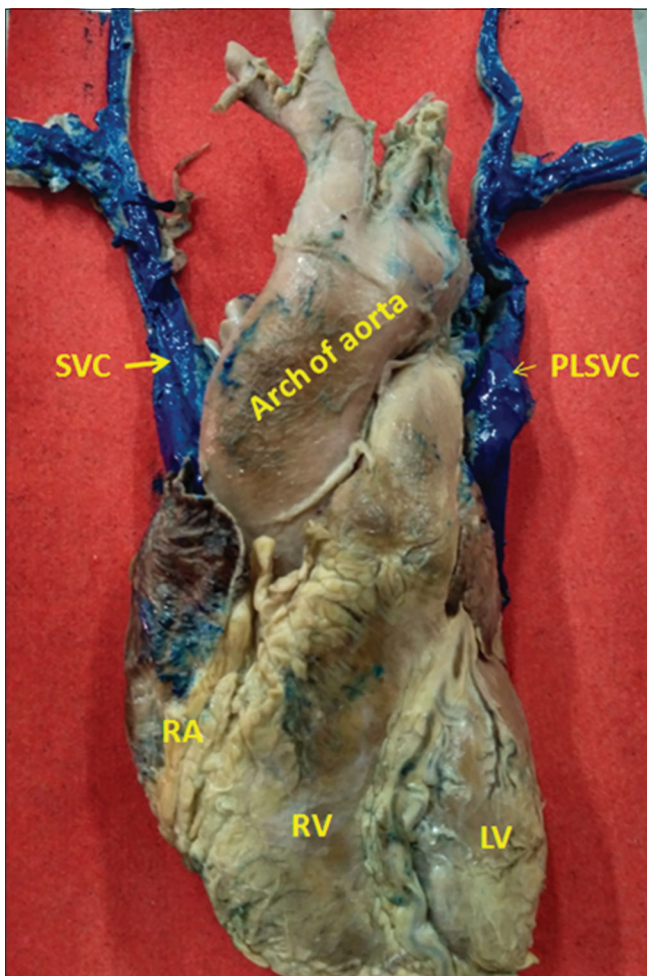


Figure 1: Showing persistent left superior vena cava (PLSVC) and superior vena cava (SVC) on Both side.the There is no communication between these two SVC and PLSVC veins. SVC- Superior venacava PLSVC- persistent left superior venacava RA- Right Atrium, RV –Right ventricle, LV- left ventricle

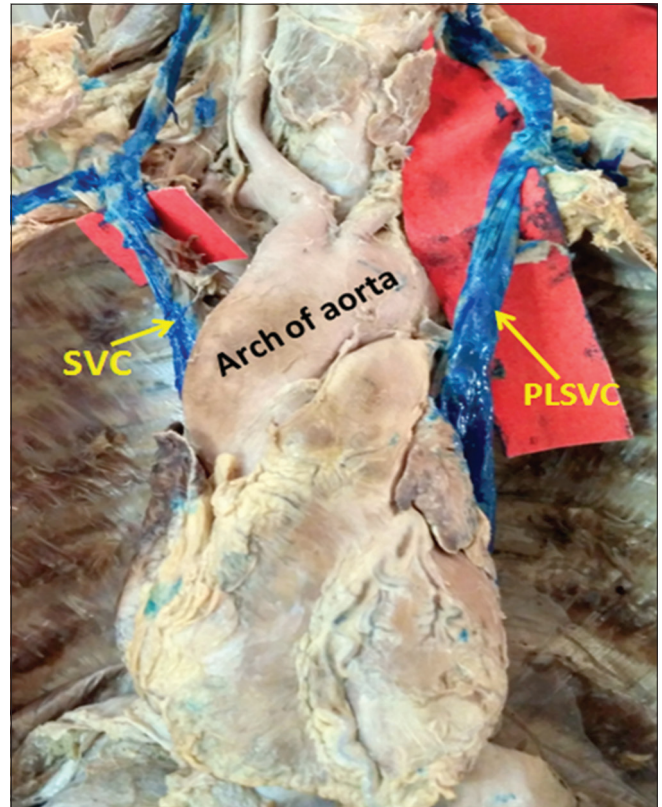


Figure 2: Showing persistent left superior vena cava (PLSVC). Both the vena cavae (SVC & PLSVC) were formed as brachiocephalic veins of the corresponding side. Thereis no communication between the two veins

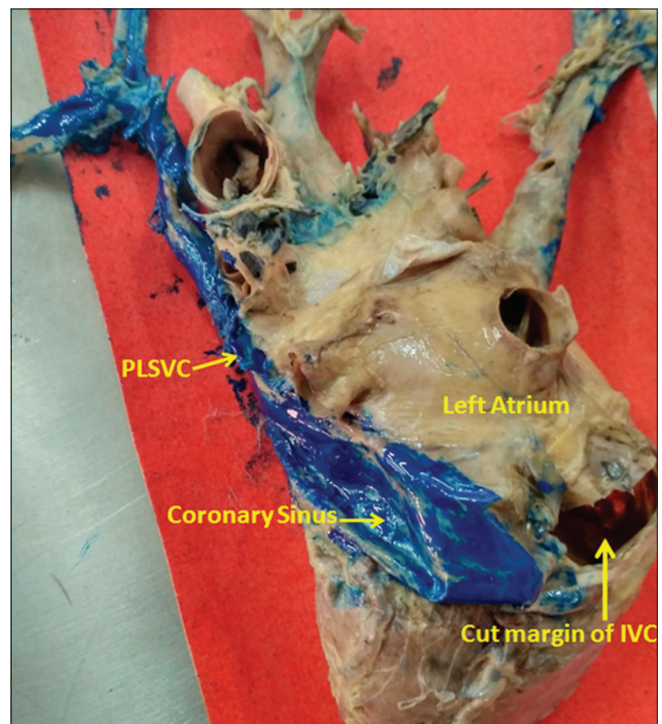


Figure 3: Showing the persistent left superior vena cava (PLSVC) opening into the enlarged coronary sinus that drains into the right atrium between the opening of inferior vena cava and right Atrioventricular orifice

no communication between the two vena cavae (Figure 3). The hemiazygos and accessory hemiazygos veins drained (normally) into the azygos vein which in turn drained into the right superior vena cava. No other associated variations were observed in both cadavers.

RESULT

We found persistent left superior vena cava in two cadavers out of 30 cadavers (6.66%) one was 64-year-old male cadaver and another 72-year-old male cadaver. Both the vena cavae were formed as of brachiocephalic veins of the corresponding side. The persistent left superior vena cava opened into the enlarged coronary sinus that drained into the right atrium between the opening of inferior vena cava and right atrio-ventricular orifice.

DISCUSSION

During the fifth week of intrauterine life, in the human fetus, three pairs of major veins can be distinguished: the vitelline veins, carrying blood from the yolk sac to the sinus venosus; the umbilical veins, originating in the chorionic villi and carrying oxygenated blood to the embryo; and the cardinal veins, draining the body of the embryo proper. The cardinal veins form the main venous drainage system of the embryo. This system consists of the anterior cardinal veins, which drain the cephalic part of the embryo, and the posterior cardinal veins, draining the remaining part of the body of the embryo. The anterior and posterior cardinal veins join to form common cardinal veins and enter the right and left horns of the sinus venosus. Formation of the vena cava system is characterized by the appearance of anastomoses between the left and right sides in such a manner that the blood from the left side is directed to the right side. The anastomosis between the anterior cardinal veins develops into the left brachiocephalic vein. Most of the blood from the left side of the head and the left upper extremity is thus directed to the right. The terminal portion of the left anterior cardinal vein entering into the left brachiocephalic vein is retained as the left superior intercostal vein. The superior vena cava is thus formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein. On the other hand, the left common cardinal vein and the distal part of the left horn become atretic and form the ligament of Marshall or ligament of the left superior vena cava. If this normal regression of the left cardinal vein fails to occur, it results in a PLSVC.⁹

The PLSVC is normal in some mammals but it is rare in man. Congenital abnormalities of the superior vena cava generally fall into one of two categories: anomalies of

position or anomalies of drainage. Anomalies of position, especially a PLSVC are far more frequent than those of drainage. A PLSVC in itself causes no hemodynamic disturbance.¹⁰

PLSVC is an uncommon and yet the most commonly reported thoracic venous abnormality. The frequency of a PLSVC is 0.3-0.5% among healthy individuals and as many as 10% of patients with congenital heart diseases.¹⁻³ There two types of PLSVC described in the literature. In 92% of cases, PLSVC Drains into to the right atrium via coronary sinus with no hemodynamically significant consequence and in 8% of cases, PLSVC drains into directly or through the pulmonary veins to the left atrium causing a right to left shunt.¹¹

The most common thoracic venous abnormality is the LPSVC draining into the coronary sinus in the presence of both left and right superior vena cavae. This anomaly is usually asymptomatic and does not require treatment unless accompanied by other cardiac anomalies.¹²

PLSVC drains into the right atrium *via* the coronary sinus in 92% of cases. but in the remainder of cases, it connects to the left atrium in such variants with absent or unroofed coronary sinus or normal coronary sinus and so creates a right-to-left shunt. Although the anomalies of systemic venous connection to the right atrium require no treatment when they occur alone, the PLSVC assumes particular significance when it communicates with the left atrium. Such patients usually present with cyanosis, polycythemia or clubbing.¹³

In the present study a double SVC with a PLSVC was observed in two cadaveric heart and there was no communication between the two-superior vena cavae unlike as reported previously. PLSVC may also give rise to rhythm disturbances such as sinus node dysfunction and atrioventricular block. These rhythm problems may be related to the stretching of the conduction tissue caused by the enlargement of the coronary sinus.³ It may also be associated with other malformations such as situs inversus or tetralogy of Fallot.⁹

During cardiac surgery, the presence of PLSVC would be a relative contraindication to the administration of retrograde cardioplegia. It may be possible to clamp the PLSVC to avoid the cardioplegia solution from perfusing retrograde up the PLSVC and its tributaries. However, there is a possibility that there may be some steal of cardioplegia solution through an accessory vein. Further, the coronary sinus catheter balloon may not be able to occlude the dilated coronary sinus. This may result in the failure of flow of cardioplegia solution to the myocardium. Thus,

the cardioplegia solution administered would largely be distributed to the left internal jugular and left sub-clavian veins, rather than the myocardium.¹⁴

CONCLUSION

Persistent left superior vena cava PLSVC is a rare congenital anomaly and. Therefore, for the clinicians should be alert about the possible existence of these venous anomalies, and cardiac abnormalities associated with it and their clinical consequences so as to prevent possible complications in routine clinical practice and during cardiopulmonary bypass surgeries. The other studies report that PLSVC can be found up to 10% cases. in our study on 30 cadavers, we found PLSVC in two male cadavers which is about 6.66%. This also to be added to the existing knowledge of these congenital abnormalities and stresses on the use of different diagnostic techniques for their accurate diagnosis. thereby avoiding further complications while planning different interventions.

ACKNOWLEDGEMENTS

We are grateful to the cadaver donors who have voluntarily donated the same for the purpose of medical research. The authors take this opportunity to thank Department of Anatomy of JNUIMSRC and NIMS & R for their whole hearted support for this study

REFERENCES

1. Akalin H, Uysalel A, Ozyurda U, Corapcioglu T, Eren NT, Emirogullari N, et al. The triad of persistent left superior vena cava connected to the coronary sinus, right superior vena cava draining into the left atrium, and atrial septal defect: report of a successful operation for a rare anomaly. *J Thorac Cardiovasc Surg.* 1987;94(1):151-153.
[https://doi.org/10.1016/S0022-5223\(19\)36333-0](https://doi.org/10.1016/S0022-5223(19)36333-0)
2. Nsah EN, Moore GW and Hutchins GM. Pathogenesis of persistent left superior vena cava with a coronary sinus connection. *Pediatr Pathol.* 1991;11(2):261-269.
<https://doi.org/10.3109/15513819109064763>
3. Goyal SK, Punnam SR, Verma G and Ruberg FL. Persistent left superior vena cava: a case report and review of literature. *Cardiovasc Ultrasound.* 2008;6:50.
<https://doi.org/10.1186/1476-7120-6-50>
4. Kotian SR, Souza AS, Ravichandran P, Bhat P and Hosapatna M. Double Superior venacava and its Associated Clinical Implications-A Case Report and Literature Review. *Nitte University Journal of Health Science.* 2015;5(1):75.
5. Erdogan M, Karakas P and Uygur F. Persistent left superior vena cava: the anatomical and surgical importance. *West Indian Med J.* 2007; 56: 72-76.
<https://doi.org/10.1590/S0043-31442007000100013>
6. Ying ZQ, Ma J, Xu G and Chen MY. Double superior vena cava with persistent left superior vena cava. *Intern Med.* 2008; 47: 679-680.
<https://doi.org/10.2169/internalmedicine.47.0840>
7. Biffi M, Boriani G, Frabetti L, Bronzetti G and Branzi A. Left superior vena cava persistence in patients undergoing pacemaker or cardioverter defibrillator implantation: a 10-year experience. *Chest.* 2001; 120:139-144.
<https://doi.org/10.1378/chest.120.1.139>
8. Chandra A and Reul GJ Jr. Persistent left superior vena cava. Discovered during placement of central venous catheter. *Tex Heart Inst J.* 1998; 25:90.
9. Sadler TW. *Langman's Medical Embryology.* 7th ed. Baltimore: Williams and Wilkins. 1995; 221-3.
10. Sharma OP and Senthila S. Left sided Superior Vena cava: (A case report and review of literature), *Asian Journal of Medical Sciences.* 2010;1(1): 18-19.
<https://doi.org/10.3126/ajms.v1i1.2508>
11. Ratliff HL, Yousufuddin M, Lieving WR, Watson BE, Malas A, Rosencrance G, et al. Persistent left superior vena cava: case reports and clinical implications. *Int J Cardiol.* 2006;113(2):242-246.
<https://doi.org/10.1016/j.ijcard.2005.08.067>
12. Park MK. *Pediatric cardiology for practitioners.* 4th ed. St. Louis: Mosby. 2002; 1411- 263.
13. Goyal SK, Punnam SR, Verma G and Ruberg FL. Persistent left superior vena cava: a case report and review of literature. *Cardiovascular ultrasound.* 2008;6(1):50.
<https://doi.org/10.1186/1476-7120-6-50>
14. Pavai J and Nayak S. A persistent left superior vena cava. *Singapore Med J.* 2007;48: 90-93.

Author's Contribution:

SK-Concept and design of the study; prepared first draft of manuscript; **SK**- Interpreted the results; reviewed the literature and manuscript preparation; **RP**-Concept, coordination, review of literature and manuscript preparation; **PK**- Statistically analysed and interpreted, preparation of manuscript and revision of the manuscript. **LJ**- Revision of the manuscript

Work attributed to: Department of Anatomy, Jaipur National University Institute for Medical Sciences And Research Centre (JNUIMSRC) Jaipur and National Institute of Medical Sciences and Research (NIMS & R).

Orcid ID:

Dr. Sachendra Kumar Mittal- <https://orcid.org/0000-0002-8691-1563>
 Dr. Rekha Parashar- <https://orcid.org/0000-0002-5192-1294>
 Dr. Pankaj Kumar Singh - <https://orcid.org/0000-0002-5716-2022>
 Dr. Leena Jadon - <https://orcid.org/0000-0002-2712-9002>

Source of Funding: None, **Conflict of Interest:** None