

Persistent Left Superior Vena Cava The Anatomical and Surgical Importance

M Erdođan¹, P Karaka^{o2}, F Uygur¹, B Me^{oe}1, B Yamak¹, MG Bozkir²

ABSTRACT

Objective: Persistence of the left superior vena cava (PLSVC) is a congenital anomaly resulting from failure of degeneration of the left cardinal vein. The prevalence of this anatomical variant is approximately 0.3% in the general population. The aim of this study was to determine the prevalence of the persistence of the left superior vena cava in order to avoid potential complications.

Subjects and methods: The study was conducted during a five-year period in 152 patients (64 males, 88 females) aged 1–65 years who underwent cardiac surgery for congenital heart diseases.

Results: The persistence of the left superior vena cava was found in 5 of 152 patients (3.3%). All persistent left superior vena cavae opened into the coronary sinus. Furthermore, the coronary sinus was enlarged in all patients. The right superior vena cava was absent in one of the five cases (20%) in this study.

Conclusion: This anatomical variant must be recognized to avoid the potential complications.

Vena Cava Superior Izquierda Persistente Importancia Anatómica y Quirúrgica

M Erdođan¹, P Karaka^{o2}, F Uygur¹, B Me^{oe}1, B Yamak¹, MG Bozkir²

RESUMEN

Objetivo: La persistencia de la vena cava superior (PVCSI) es una anomalía congénita que se presenta como resultado de un fallo degenerativo en la vena cardinal izquierda. La prevalencia de esta variante anatómica es aproximadamente 0.3% en la población general. El objetivo de este estudio es determinar la prevalencia de la persistencia de de la vena cava superior izquierda a fin de evitar complicaciones potenciales.

Sujetos y métodos: El estudio fue realizado durante un periodo de cinco años en 152 pacientes (64 varones, 88 hembras) de 1–65 años, que fueran sometidos a cirugía cardíaca debido a enfermedades cardíacas congénitas.

Resultados: Se halló persistencia de la vena cava superior izquierda en 5 de 152 pacientes (3.3%). Todas las venas cavas superiores izquierdas desembocan en el seno coronario. Además, el seno coronario de todos los pacientes presentaba un ensanchamiento. No había vena cava superior en uno de los cinco casos (20%) de este estudio.

Conclusión: Esta variante anatómica tiene que ser reconocida a fin de evitar potenciales complicaciones.

West Indian Med J 2007; 56 (1): 72

INTRODUCTION

Persistence of the left superior vena cava (PLSVC) has an incidence of 0.3–0.5% in the general population. In the presence of other congenital heart disease, the incidence of

PLSVC increases to 3–10% (1–11). It is frequently associated with ventricular septal defect (VSD), atrial septal defect (ASD), coarctation of the aorta and mitral atresia (12–15). Due to associated diseases, rhythm disturbances of cardiac impulse formation and conduction could be seen (9,10). Precise anatomical knowledge of the great vessels of the neck and thorax is essential for safe anaesthesia, intensive care practice, pacemaker implantation and cardiac surgery. Therefore, this anatomical variant must be recognized to

From: Department of Cardiovascular Surgery¹, Sani Konukođlu Hospital, Gaziantep and Department of Anatomy², Faculty of Medicine, Çukurova University, Adana, Turkey.

Correspondence: Dr P Karaka^o, Department of Anatomy, Faculty of Medicine, Çukurova University, Adana, Turkey.

avoid the potential complications such as those associated with placement of central catheters and cardiac surgery.

The aim of the study was to investigate the anatomy of the superior caval veins so as to determine the prevalence of the common anatomic variant of caval venous system – persistence of the left superior caval vein – in order to avoid the complications and risks to patients during surgery.

SUBJECTS AND METHOD

This study was carried out between May 1999 and December 2004 in patients who underwent cardiac surgery for congenital heart diseases. During this period, 152 patients underwent open heart surgery for congenital heart defects. Sixty-four of the 152 patients (42%) were male and 88 (58%) were female. The mean age was 24.57 ± 15.01 (range 1–65 years). The pathologies were secundum ASD in 84 patients and VSD in 34 patients. The rest are listed in Table 1. The diagnoses of

Table 1: Details of the cardiac defects (n= 152)

Defect	Number of Patients
ASD secundum	81
ASD secundum + PDA	3
High venous ASD + PAPVDA	15
VSD	33
VSD + AI + MI	1
TOF	3
ASD + VSD	3
ASD + PS	4
ASD + MS	2
VSD + PS	5
PS	1
VSD + AS	1

ASD = atrial septal defect, VSD = ventricular septal defect, PS = pulmonary stenosis, TOF = tetralogy of fallot, AI = aortic insufficiency, MI = mitral insufficiency, AS = aortic stenosis, MS = mitral stenosis, PAPVDA = partial abnormal pulmonary venous drainage anomaly, PDA = patent ductus arteriosus.

all the patients were made by echocardiography and cardiac catheterization. Patients who were above 35 years of age underwent coronary angiography before the operation. All of the operations were performed with median sternotomy and cardiopulmonary bypass with membrane oxygenator. During the operations, all of the 152 patients were carefully examined for anomalies of systemic venous drainage which may cause complications during the cannulations for cardiopulmonary bypass. Absence of the left brachiocephalic vein was considered an early clue to the presence of a persistent left superior vena cava which will be found in the area of the Marshall ligament, anterior to the left pulmonary artery and veins (Fig. 1).

RESULTS

During five years, persistence of the left superior vena cava (PLSVC) was encountered in 5 of 152 patients (3.3%). All persistent left *venae cavae superiores* emptied into the

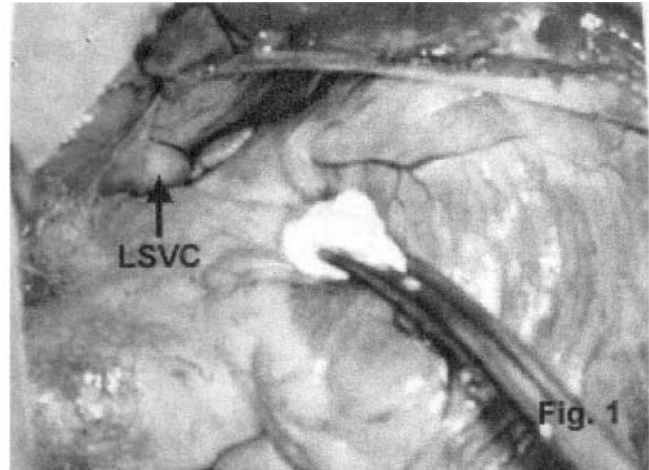


Fig. 1: Operative view through a median sternotomy of a left superior caval vein (LSCV) entering the pericardial cavity between left atrial appendage and the pulmonary trunk.

coronary sinus. The coronary sinus was enlarged in these cases. Moreover, the right superior vena cava was absent in 20% of the patients (1/5 patients). If the persistent left superior vena cava was very large or larger than the right or represented a single superior vena cava, it was drained separately with an appropriately sized cannula introduced into the coronary sinus through the right atrium or through the superior vena cava itself. When the left superior vena cava was small (especially when there was a left brachiocephalic vein between the two superior venae cavae), it was temporarily occluded and cannulated only if the venous pressure rose to over 30 mm Hg. The PLSVC was cannulated in two of the five patients. The cannula was inserted via the coronary sinus orifice in one patient. In the other patient, the left superior vena cava was directly cannulated. In three patients, the PLSVC was clamped and the pressure before the clamp was checked continuously. The details of the operations are shown in Table 2.

Table 2: Operative details (n= 152)

Operative Details	Number of patients
ASD secundum	
Primary closure	68
Closure with patch	16
Persistent Left VCS	5
Canulation	2
Clamping	3
Associated procedures	
AVR + MVR	1
MVR	1
PDA ligation	3
Subvalvular Membrane Resection	1

ASD = atrial septal defect, VCS = vena cava superior, AVR = aortic valve replacement, MVR = mitral valve replacement, PDA = patent ductus arteriosus.

The early postoperative period was uneventful. There was no mortality and morbidity during the hospital stay. All patients were discharged on the 6th postoperative day. During the late postoperative period, one patient who underwent primary closure of secundum ASD developed constrictive pericarditis. This patient had pericardiectomy as a second operation. No cardiac rhythm problem was observed within the perioperative and postoperative period.

DISCUSSION

The persistence of the left superior vena cava (PLSVC) is a congenital anomaly resulting from failure of degeneration of the left cardinal vein. While the embryo grows, new organs appear and persist and others are transient and disappear. The development of the great systemic veins is a complex process and of clinical importance. In a 4 mm embryo, three main groups of veins are seen. These include the omphalo-mesenteric veins, the umbilical veins and the common cardinal veins. The anterior and posterior cardinal veins join to form common cardinal veins and enter the right and left horns of the sinus venosus. Due to the rightward direction of blood flow, the right horn of the sinus venosus develops. Additionally, the left common cardinal veins and the distal part of the left horn become atretic and is designated the ligament of Marshall or ligament of the left superior vena cava. The failure of the left anterior cardinal vein to obliterate results in PLSVC (9,12,16–18). This vein generally drains to the coronary sinus and then into the right atrium (Fig. 2a and 2b). The incidence of the persistence of the left

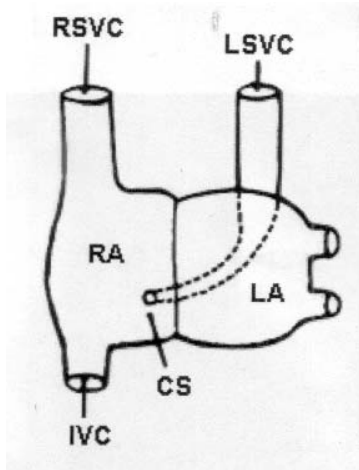


Fig. 2a: Persistent left superior vena cava (LSVC) draining into the right atrium (RA) via the coronary sinus (CS). The inferior vena cava (IVC) and right superior vena cava (RSVC) join the right atrium normally; LA = Left atrium.

superior vena cava is approximately 0.3% in the general population and in 3% to 10% of patients have additional cardiac defects (1–11,19).

The PLSVC is normal in some mammals but it is rare in man (16,17). Other much rarer anomalies including ab-

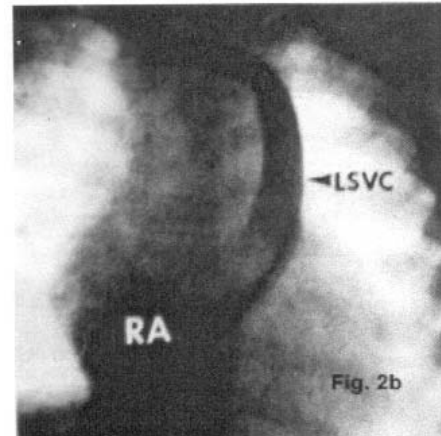


Fig. 2b: Left superior vena cava (LSVC) draining into the coronary sinus; RA = Right atrium.

sence of right superior vena cava or even the complete lack of superior caval veins could be seen (4,5,10,15,19–23).

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 is far more frequent than those of drainage. A PLSVC in itself causes no haemodynamic disturbance because it harmlessly drains into the right atrium via the coronary sinus. A PLSVC draining into the coronary sinus is not only the most common thoracic venous anomaly but also the most frequent cause of enlargement of the coronary sinus (5, 9, 12, 24, 25). In the majority of cases, there is a right superior vena cava also, but frequently there is hypoplasia or agenesis of the left innominate vein. In general, the size of the two venae cavae is complementary; the larger the left superior vena cava, the smaller the right. Sometimes there is atresia of the right superior vena cava. In this instance, the anomaly is of major importance as the PLSVC is the sole route of venous return from the upper body (26). Not so harmless are the congenital atrioventricular conduction defects and arrhythmias that may occur in this setting (9, 10, 14). Moreover, PLSVC with absence of the right superior vena cava complicates pacemaker lead implantation via the transvenous approach (10). In the cases reported herein, the right superior vena cava was absent only in one patient and no complications were observed.

Persistent left superior vena cava is joined to the right superior caval vein by an innominate vein in about sixty per cent of cases (10, 27). In this arrangement, the PLSVC can simply be clamped or ligated to avoid flooding the field when the heart is opened.

In this study, a similar prevalence of the PLSVC was observed when compared to the general population (5 of 152 patients, 3.3%). Zerbe *et al* (4) reported 4 patients of 661 with PLSVC whereas Biffi and his colleagues (9) reported 6 of 1250 patients with this anatomical variant. In addition, the right superior vena cava was absent in 20% of our patients (1/5 patients) compared to an average 10% in other reports

(3, 5, 10, 23). This difference may be due to the low prevalence of this variant in each study.

The anatomy of the upper caval veins is important for pacemaker implantation and for cardiac surgery. The PLSVC is often found during surgery or catheterization due to the low frequency of presence of some diagnostic signs on the conventional chest X-rays (11, 24, 28, 29). Some researchers have proposed some diagnostic features which include the widening of the aortic shadow, paramedian bulging and a paramedian strip or crescent along the left heart border on chest X-ray (14, 30). The shadow of the PLSVC may also be seen along the left upper border of the mediastinum (10, 28). Moreover, it was reported that when the chest radiograph shows the central venous catheter passing along the border of the left heart and good blood return through the catheter then a PLSVC would be suspected (12). It is also possible to diagnose PLSVC by angiography, echocardiography, computed tomography and magnetic resonance imaging (4, 5, 11, 31–33). Diagnosis of PLSVC by echocardiography has 100% specificity and 96% sensitivity. In our cases, the diagnosis of PLSVC was also made by echocardiography.

This 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 (9, 13, 14, 18, 28, 32, 34–37). 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 (38). Such patients usually present with cyanosis, polycythaemia or clubbing, although some have no clinical findings (14, 32, 38). The cyanotic cases have the risk of paradoxical embolus. Cerebral emboli and abscess have been reported in these patients (7, 38). Intracardiac rerouting technique has been traditional and the common approach to correct this right-to-left shunt. The proximity of the pulmonary veins to the orifice of the left superior vena cava makes this technique a difficult approach. Therefore, extracardiac methods and acute ligation may be also preferred (36, 39).

Detailed study of the anatomy of the venous system is essential before cavopulmonary procedures such as Glenn anastomosis, bidirectional cavopulmonary connection or Fontan-type procedure to determine the anatomical variants of the venous system (37). Moreover, these data will be useful for safe anaesthetic and intensive care practice. According to this analysis, the clinician may plan different surgical approaches. Furthermore, it was reported that the intensive care clinicians should be aware of the presence of a left superior vena cava in order not to place catheters outside the venous circulation and to avoid complications like perforation, shock, cardiac arrest, cardiac tamponade and thrombosis (4, 6, 12, 28).

In summary, the diagnosis of PLSVC should not be left to chance. The anatomy of the systemic venous return should

be known precisely for procedures such as anaesthesia, placement of catheters and cardiac surgery in order to avoid complications.

REFERENCES

1. Saunders JM. Bilateral superior vena cava. *Anat Rec* 1946; **94**: 657–9.
2. Campbell M, Deuchar DC. The left-sided superior vena cava. *Br Heart J* 1954; **16**: 423.
3. Camm AJ, Dymond D, Spurrell RA. Sinus node dysfunction associated with absence of right superior vena cava. *Br Heart J* 1979; **41**: 504–7.
4. Zerbe F, Bornakowski J, Sarnowski W. Pacemaker electrode implantation in patients with persistent left superior vena cava. *Br Heart J* 1992; **67**: 65–6.
5. Bartram U, Van Praagh S, Levine JC, Hines M, Bensky AS, Van Praagh R. Absent right superior vena cava in viscerotransposed situs solitus. *Am J Cardiol* 1997; **80**: 175–83.
6. Cron TA, Buser PT, Osswald S. Placement of a temporary pacemaker electrode through a persistent left superior vena cava. *Intensive Care Med* 1998; **24**: 639–40.
7. Chandra A, Reul GJ Jr. Persistent left superior vena cava. Discovered during placement of central venous catheter. *Tex Heart Inst J* 1998; **25**: 90.
8. Birnie D, Tang AS. Permanent pacing from a left ventricular vein in a patient with persistent left and absent right superior vena cava. *Pacing Clin Electrophysiol* 2000; **23**: 2135–7.
9. Biffi M, Boriani G, Frabetti L, Bronzetti G, Branzi A. Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation. *Chest* 2001; **120**: 139–44.
10. Corbisiero R, DeVita M, Dennis C. Pacemaker implantation in a patient with persistent left superior vena cava and absent right superior vena cava. *J Intervent Card Electrophysiol* 2003; **9**: 35–7.
11. Lappegard KT, Prytz JF, Haug B. Pacemaker implantation in patients with persistent left superior vena cava. *Heart Vessels* 2004; **19**: 153–4.
12. Boussuges A, Ambrosi P, Gainnier M, Quenee V, Sainty JM. Left-sided superior vena cava: diagnosis by magnetic resonance imaging. *Intensive Care Med* 1997; **23**: 702–3.
13. Brunelli F, Amaducci A, Mhagna Z, Troise G, Quaini E. Uncommon presentation and surgical correction of unroofed coronary sinus syndrome. *Ann Thorac Surg* 2003; **76**: 619–21.
14. Higgs AG, Paris S, Potter F. Discovery of left-sided superior vena cava during central venous catheterization. *Br J Anaesth* 1998; **81**: 260–1.
15. Karnegis JN, Wang Y, Winchell P, Edwards JE. Persistent left superior vena cava, fibrous remnant of the right superior vena cava and ventricular septal defect. *Am J Cardiol* 1964; **14**: 573–7.
16. Netter FH. Embryology. In: Yonkman FF (ed), *The CIBA collection of medical illustrations, Volume 5 – Heart*. The Case-Hoyt Copr, Rochester, NY; 1981. 128–30.
17. Williams PL, Warwick R, Dyson M, Bannister LH. (eds) *Embryology*. In: *Gray's Anatomy*. 37th Ed. Churchill Livingstone; 1989. p. 219–25.
18. Jha NK, Gogna A, Tan TH, Wong KY, Shankar S. Atresia of coronary sinus ostium with retrograde drainage via persistent left superior vena cava. *Ann Thorac Surg* 2003; **76**: 2091–2.
19. Barbaix E, Kerckaert I, D'Herde K, Depreter M, Espeel M. Simultaneous occurrence of a thyromediastinal muscle, a truncus bicarotico-brachialis and a left superior vena cava. *Clin Anat* 2003; **16**: 176–81.
20. Lenox CC, Zuberbuhler JR, Park SC, Neches WH, Mathews RA, Fricker FJ et al. Absent right superior vena cava with persistent left superior vena cava: implications and management. *Am J Cardiol* 1980; **45**: 117–22.
21. Hussain SA, Chakravarty S, Chaikhouni A, Smith JR. Congenital absence of superior vena cava: unusual anomaly of superior systemic veins complicating pacemaker placement. *Pacing Clin Electrophysiol* 1981; **4**: 328–32.
22. Pugliese P, Murzi B, Aliboni M, Eufrates S. Absent right superior vena cava with persistent left superior vena cava: clinical and surgical considerations. *J Cardiovasc Surg* 1984; **25**: 134–7.

23. Eckart RE, Leitch WS, Shry EA, Krasuski RA, Lane MJ, Leclerc KM. Utility of magnetic resonance imaging in cardiac venous anatomic variants. *Cardiovasc Intervent Radiol* 2003; **26**: 309–11.
24. Azocar RJ, Narang P, Talmor D, Lisbon A, Kaynar M. Persistent left superior vena cava identified after cannulation of the right subclavian vein. *Anesth Analg* 2002; **95**: 305–7.
25. Dibardino DJ, Fraser CD Jr, Dickerson HA, Heinle JS, McKenzie ED, Kung G. Left ventricular inflow obstruction associated with persistent left superior vena cava and dilated coronary sinus. *J Thorac Cardiovasc Surg* 2004; **127**: 959–62.
26. Del Campo C. Persistent left superior vena cava. *Tex Heart Inst J* 1998; **25**: 161.
27. Moss AJ, Adams FH, Emmanoulides GC. *Heart Disease In Infants, Children and Adolescents*. 5th ed. 1995. p. 875–8.
28. Schummer W, Schummer C, Fröber R. Persistent left superior vena cava and central venous catheter position: clinical impact illustrated by four cases. *Surg Radiol Anat* 2003; **25**: 315–21.
29. Kamola PA, Seidner DL. Peripherally inserted central catheter malposition in a persistent left superior vena cava. *J Infus Nurs* 2004; **27**: 181–4.
30. Cha EM, Khoury GA. Persistent left superior vena cava. *Radiology* 1972; **103**: 375–81.
31. Hibi N, Fukui Y, Nishimura K, Miwa A, Kambe T, Sakamoto N. Cross-sectional echocardiographic study on persistent left superior vena cava. *Am Heart J* 1980; **100**: 69–76.
32. Miraldi F, di Gioia CR, Proietti P, De Santis M, d'Amati G, Gallo P. Cardinal vein isomerism: an embryological hypothesis to explain a persistent left superior vena cava draining into the roof of the left atrium in the absence of coronary sinus and atrial septal defect. *Cardiovas Pathol* 2002; **11**: 149–52.
33. Krasemann T, Kehl G, Vogt J, Asfour B. Unusual systemic venous return with complete absence of the superior caval veins. *Pediatr Cardiol* 2003; **24**: 397–9.
34. Wiles HB. Two cases of left superior vena cava draining directly to a left atrium with a normal coronary sinus. *Br Heart J* 1991; **65**: 158–60.
35. Adatia I, Gittenberger-de Groot AC. Unroofed coronary sinus and coronary sinus orifice atresia. Implications for management of complex congenital heart disease. *J Am Coll Cardiol* 1995; **25**: 948–53.
36. Reddy VM, McElhinney DB, Hanley FL. Correction of left superior vena cava draining to the left atrium using extracardiac techniques. *Ann Thorac Surg* 1997; **63**: 1800–02.
37. Filippini LHPM, Ovaert C, Nykanen DG, Freedom RM. Reopening of persistent left superior caval vein after bidirectional cavopulmonary connections. *Heart* 1998; **79**: 509–12.
38. Geggel RL, Perry SB, Blume ED, Baker CM. Left superior vena cava connection to unroofed coronary sinus associated with positional cyanosis: successful transcatheter treatment using gianturco-grifka vascular occlusion device. *Cathet Cardiovasc Intervent* 1999; **48**: 369–73.
39. Zimand S, Benjamin P, Frand M, Mishaly D, Smolinsky AK, Hegesh J. Left Superior Vena Cava to the Left Atrium: Do we have to change the traditional approach? *Ann Thorac Surg* 1999; **68**: 1869–72.