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Original Article

Persistent left superior vena cava frequency in congenital heart surgery and its effect on surgical strategy

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Abstract

Aim: Persistent left superior vena cava encompasses a wide range of systemic venous anomalies. Persistent left superior vena cava is relatively infrequent and, under normal circumstances, asymptomatic but may be of clinical importance in cardiac surgery. This study aims to investigate the effect of persistent left superior vena cava on surgical planning.

Material and Methods: We included a total of 525 consecutive patients (310 males, 215 females) who underwent open-heart surgery for congenital heart disease. The association of persistent left superior vena cava with congenital anomalies and surgical approaches were recorded. Demographic data and outcome data were retrospectively analyzed.

Results: Persistent left superior vena cava was observed in 28 (5.3%) patients. The drainage point of PLSVC was coronary sinus in 23 (82.1%) patients, left atrium in 5 (17.9%) patients. The mean age and weight of the patients were 11.5 months (range, one day to 18 years), 9.3 kg (range, 0.5-65 kg), respectively. Persistent left superior vena cava was obtained in 11 (39.4%) patients with echocardiography, 12 (%42.8) patients with cardiac catheterization and tomography imaging, and 5 (17.8%) patients during surgery. Surgical management of the PLSVC included of temporary occlusion in 17 (60.7%) patients, direct cannulation in 6 (21.5%) patients, Glenn shunt in 2 (7.1%) patients, intracardiac rerouting in 2 (7.1%) patients and ligation in 1 (3.6%) patient. No operative morbidity and complication associated with persistent left superior vena cava were seen.

Conclusions: Consequently, persistent left superior vena cava is relatively infrequent, but the surgical team should be aware of this anomaly, its draining points, and possible complications, and it must be kept in mind simple and effective solutions about persistent left superior vena cava.

Keywords: Coronary sinus; left atrium; persistent left superior vena cava; surgery

INTRODUCTION

Persistent left superior vena cava (PLSVC) constitute the most variable class of congenital cardiovascular abnormalities. Although PLSVC can be detected incidentally, it can be seen more frequently as a component of a complex anomaly in congenital heart disease (1-4). These anomalies are relatively rare, and although they are observed in the general population at an average rate of 0.5 %, it is more frequently seen at 0.2-11 % in the subpopulation with congenital heart disease (4-8). It is mostly anatomically benign but can be problematic during the repair of congenital heart defects (5). It is necessary to know the details of the patient's cardiovascular anatomy to perform any planned surgical procedure more safely and accurately. In this study, we investigated the incidence of PLSVC, the effect of PLSVC on surgical strategy, and the management of the PLSVC in decision-making for surgical planning at the perioperative course.

MATERIALS and METHODS

A total of 525 consecutive pediatric patients undergoing cardiac surgery for congenital heart disease between February 2017 and March 2020 were retrospectively evaluated. Data including demographic characteristics, cardiac diagnoses, imaging methods, perioperative situation, and outcomes were collected. Patients greater than 18 years were excluded from the study. Preoperative assessment of congenital heart disease included transthoracic echocardiography in all patients. In some patients, the other diagnostic imaging techniques used to confirm the diagnosis of congenital heart defects, such as computed tomography angiography (CTA) and conventional angiography, were recorded. All patients with PLSVC detected by imaging methods were listed. Also, intraoperative clinical findings and intraoperative management of PLSVC were listed.

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Considering the pathophysiology of the primary cardiac defect, the PLSVC drainage point, and the presence of intercaval communication, we applied five different surgical methods in 28 patients with PLSVC. These techniques are temporary snaring (n=17), selective cannulation (n=6), intracardiac rerouting (n=2), bilateral bidirectional Glenn shunt (n=2), and Ligation (n=1). Temporary snaring was done with umbilical tape. A polytetrafluoroethylene patch (Dacron,C.R. Bard, Haverhill, Pennsylvania) was used in all patients in the rerouting procedure. L-shaped venous cannula was used for selective cannulation.

Our study was conducted in accordance with the principles of the Declaration of Helsinki. All image data used in this study were obtained from the clinical routine at our institution. All patients signed the informed consent. Data were collected and analyzed retrospectively. The institutional ethics committee approved our study design (Decision date: 27.04.2020, Decision no:2020/5-15).

SPSS Statistics (version 22; IBM Corp.) was used for statistical analyses. Descriptive statistics, percentages, and counts for all demographic variables were calculated.

RESULTS

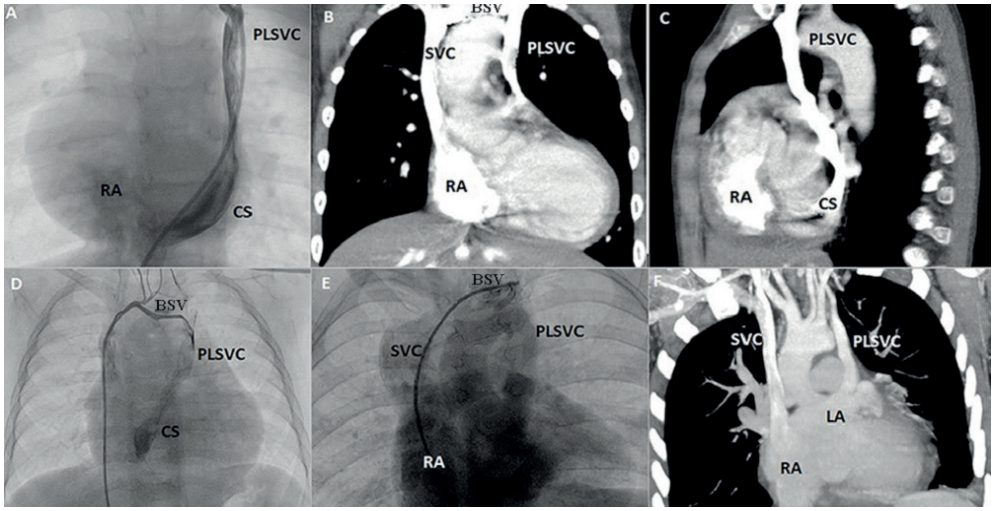
Persistent left superior vena cava (SVC) was reported in 28 patients (5.3%). The mean age was 11.5 months (range, 1 day to 18 years). The mean weight of the patients was 9.3 kg (range, 0.5 to 65 kg). The drainage point of PLSVC was coronary sinus in 23 (82.1%) patients (Figure 1), left atrium in 5 (17.9%) patients. In these five patients with LA connection (Figure 2,3), partially unroofed coronary sinus in 2 (7.2 %) cases (Figure 2), absence of coronary sinus in 1 case (3.6%), and left isomerism in one case (3.6%) was observed. The various congenital defects in patients with PLSVC were shown in Table 1. The anatomic characteristics of PLSVC were listed in Table 2.

Table 1.Congenital heart diseases in patients with PLSVC

	n	Type of congenital heart diseases
PLSVC-coronary sinus connection (n=24)	8	Ventricular septal defect (VSD)
Observed: 4.5 %	6	Complete AVSD
Literature: 0.3-10 %	3	Atrial septal defect
	2	Single ventricular heart DORV+Large VSD+Pulmonary stenosis RV hypoplasia+AVSD
	2	PAPVD
	2	Tetralogy of Fallot
	1	Intracardiac TAPCV
PLSVC-LA connection (n=4)	1	Complete AVSD (Partial unroofed CS)
Observed: 0.76 %	1	Transitional AVSD (Left isomerism)
Literature:0.3-4.3 %	1	Common atrium (Raghib's syndrome)*
	1	Atrial septal defect (Partial unroofed CS)
	1	Isolated
PLSVC (n=28)		
Observed: 5.3 %		
Literature:0.2-11 %		

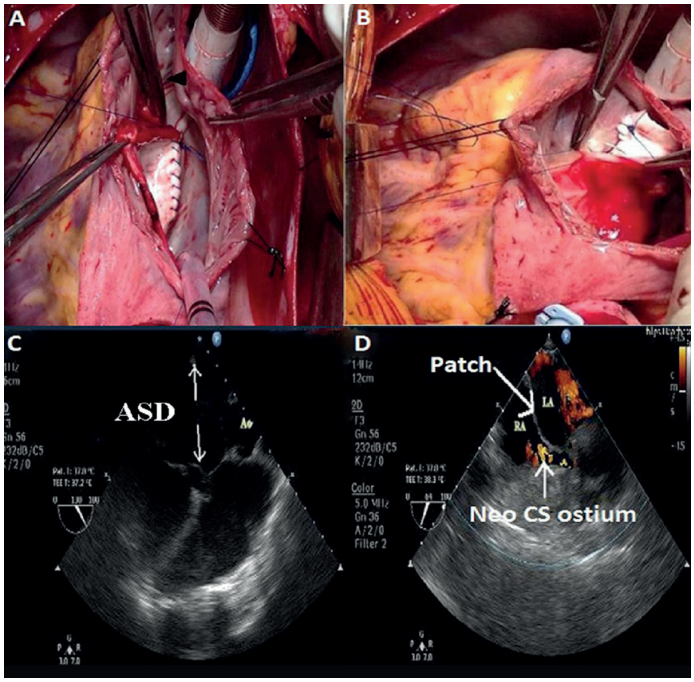
AVSD: Atrioventricular septal defect: DORV: double outlet right ventricle, IVC: inferior vena cava, LA: left atrium, PAPVD; partial anomalous pulmonary venous connection SVC: superior vena cava, TAPVC: total anomalous pulmonary venous connection.
***ASD+Absence of coronary sinus+ PLSVC-LA connection**

The diagnoses of PLSVC were obtained by CT angiography, conventional angiography, and intraoperative view in addition to echocardiography. All diagnostic techniques for PLSVC were listed in Table 2. Although 24 of 28 patients in the patient group were diagnosed with PLSVC in the preoperative period, we defined four patients intraoperatively. The PLSVC was draining into the coronary sinus in these four patients, and the primary diagnosis was a ventricular septal defect.



BSV:brachiocephalic vein, CS:coronary sinus, PLSVC:persistent left superior vena cava, SVC;superior vena cava

Figure 1. Angiography and CT images of PLSVC. PLSVC is draining into the right atrium via the coronary sinus, with the absence of brachiocephalic vein (A), thin calibrated brachiocephalic vein (D), and enlarged brachiocephalic vein (B), PLSVC with the presence of brachiocephalic vein draining into the right atrium via the coronary sinus (C,E), PLSVC with absent of brachiocephalic vein draining into the left atrium roof (F)



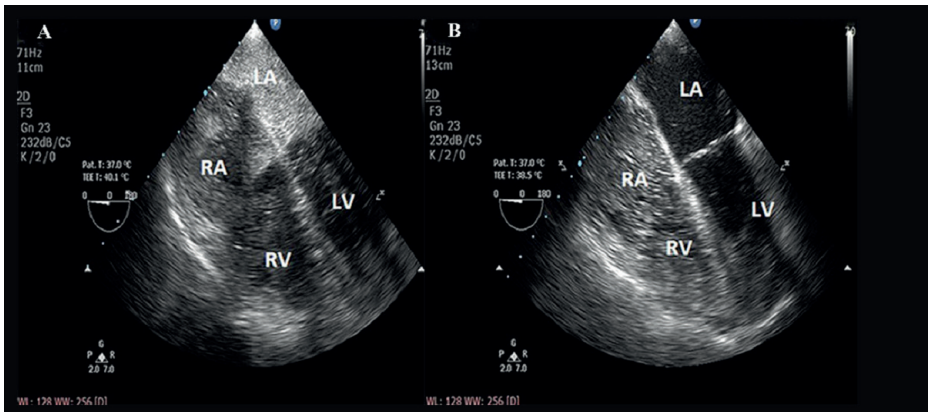
TEE: transesophageal echocardiography

Figure 2. Common atrium (Raghip's syndrome) A clamp tip (black arrowhead) is seen inside the PTFE baffle orifice, which allows PLSVC venous blood to be directed into the right atrium under the pericardial patch (A). ASD was closed over the PTFE graft with an autologous pericardial patch, with the PTFE baffle orifice remaining in the right atrium (B), Preoperative TEE (C), Postoperative TEE (D)

Table 2. Anatomic characteristics of patients of with PLSVC (n=28)

Type of venous anomalous	n
Congenital heart defect	
Biventricular heart	26
Single Ventricular Heart	2
Isolated (PLSVC-LA connection)	1
Arcus aorta (Right/Left)	22/6
Heterotaxia	
Left isomerism	1
Unroofed coronary sinus	2
Absence of coronary sinus	1
SVC	
Present	25
Absent	3
Intercaval communication (BSV)	
Normal	15
Hypoplastic	8
Absent	5
Point of drainage	
Coronary sinus	
Left atrium	
Diagnostic tools	
Echocardiography	12
CT	8
Angiography	4
Intraoperative	4
Total	28

BSV: brachiocephalic vein, CT: computed tomography, IVC: inferior vena cava, LA: left atrium, PLSVC: persistent left superior vena cava, SVC: superior vena cava



LA: Left atrium, LV: left ventricle, RV: Right ventricle, RA: Right atrium, TEE: transesophageal echocardiography

Figure 3. PLSVC-left atrial roof connection. Images showing that bubble contrast injected into the jugular vein and contrast enhancement in the left atrium (A, preoperative) and right atrium (B, postoperative)

Table 3. Intraoperative management of patients with PLSVC

Drainage	Absence of SVC	Intercaval communication	n	Management
Coronary sinus n=23	-	Normal	14	Snaring
	-	Hypoplastic	3	Selective cannulation
			3	Snaring
	2	Absent	3	Selective cannulation
Left atrium n=5	-	Normal	1	Ligation
	1	Hypoplastic	2	Intracardiac rerouting
	-	Absent	2	Bilateral Glenn shunt

Intraoperative management of the PLSVC included temporary occlusion in 17 (60.7%) patients, direct cannulation in 6 (21.5%) patients, Glenn shunt in 2 (7.1%) patients, intracardiac rerouting in 2 (7.1%) patients, and ligation in 1 (3,6%) patient (Table 3). According to drainage point, intraoperative management of the PLSVC absence of SVC and intercaval communication was listed in Table 3. The overall incidence of PLSVC included in this report has been compared with other works in the literature.

DISCUSSION

Persistent left SVC was seen in 0.5 % of the general population and 3-10 % associated with congenital heart disease (5-8). In the literature, the most common malformations associated with PLSVC are Ventricular septal defect (35%), Atrial septal defect (19%), Bicuspid aortic valve (15%), Tetralogy of Fallot (12%), Persistent ductus arteriosus (11%), Coarctation of the aorta (11%) has been reported (9). The presence of PLSVC should be investigated in these disease groups. These anomalies associated with congenital heart defects can play an essential role during the reconstruction of the cardiac defect, bidirectional cava-pulmonary shunt procedure (bilateral Glenn shunt), and cannulation strategies in cardiac surgery. Awareness and knowledge of PLSVC during the preoperative period have become essential for surgical planning. Every pediatric cardiac surgeon should well know the anatomical localization of the PLSVC. A PLSVC missed preoperatively should be routinely evaluated in every patient after the pericardium is opened. Although there are many anatomical variations, the PLSVC courses downward in front of the aortic arch and left pulmonary artery. Then, it is directed towards the coronary sinus between the left pulmonary veins and the left atrial appendage. We routinely examine this area intraoperatively in each case. We diagnosed 4 of 28 patients in our patient group by performing the intraoperative evaluation according to the anatomical localization of the PLSVC.

Most cases with PLSVC have a right SVC, but the left brachiocephalic vein does not exist. In bilateral SVC, 65% of patients have a greater left SVC (10-12). In 17% of PLSVC, there is no right SVC (8, 10-12). Persistent left SVC drains to coronary sinus (CS) at 92% of cases, and the rest of them drain to the left atrium (LA). Rarely, the persistent left superior vena cava drains into the left atrium via an unroofed coronary sinus or a direct connection to the left atrium. In our study, rates of drainage to the coronary sinus and left atrium were 82.1% and 17.9%, respectively.

Clinically PLSVC is usually associated with other cardiac abnormalities and drainage points (CS or LA), such as partial or complete atrioventricular septal defect, tetralogy of Fallot, ventricular septal defect, double outlet right ventricles, and heterotaxia syndrome, among others (13). Due to right to left shunt, desaturation may occur when a PLSVC drains into the LA either directly or through an unroofed CS (14). This desaturation is mild as room air oxygen saturation is between 85-90 %. Besides desaturation, there are risks about; cerebral embolization, brain abscess, and stroke (12-15).

The decision-making process to manage the determined PLSVC during the surgery depends upon different issues. Some questions must be answered by the surgical team while planning the cardiac procedure. The first issue is when to cannulate the PLSVC. The anatomy of the left brachiocephalic vein and the point of drainage of PLSVC is vital in the decision-making process of congenital cardiac surgery. In assessment after snaring the PLSVC, if the venous pressure of PLSVC exceeds 15 mm/Hg in the proximal site of occlusion must be cannulated (16). If the PLSVC is not cannulated and snared, venous pressure increases, and brain perfusion pressure decreases. As a result, it will cause cerebral congestion. There is no need for cannulation if a left brachiocephalic vein is at nearly the same dimension as PLSVC. Snaring of SVC for occlusion is adequate instead of LSVC cannulation whenever the presence of the appropriate size of the left innominate vein is detected. In addition, the surgeon must occlude with snaring the PLSVC in the intrapericardial segment due to the hemizygos vein. Otherwise, snare of the hemiazygos vein stays outside of the pericardial cavity, drains to the right atrial cavity, and deteriorates the surgeon's vision intraoperatively. If PLSVC is detected after initiation of CPB and placement of the cross (X) clamp, there are three options. Surgery can be continued, but if a right atriotomy has been performed, the operation time will be prolonged because the surgical area will be bloody. The second option, PLSVC can be occluded temporarily with snaring after X-clamp. However, due to insufficient venous filling, the procedure is complicated, and the risk of injury is high. The third option is to cannulate the coronary sinus through the right atriotomy.

The second issue is a discussion about the drainage point of the PLSVC. Persistent left SVC mostly drains to coronary sinus/right atrium, so there is no need for further intervention. However, PLSVC can drain to the left atrium via a partial defect of the coronary sinus or directly to the roof of the left atrium. Though rare, there is a risk of paradoxical embolism due to left atrium drainage of PLSVC. During the drug administered intravenously, embolism of air and clot may lead to problems as a heart attack or stroke. The third important issue is the close proximity of the left phrenic nerve to PLSVC. Major complication as left phrenic nerve palsy may occur during exploration of PLSVC. Thus, during the exploration of PLSVC, electrocautery should be avoided.

The fourth issue is retrograde cardioplegia applications. Retrograde cardioplegia is not effective in patients whose PLSVC is draining to the coronary sinus. Because all the cardioplegia solution mixes with systemic venous blood, solution concentration decreases, and the protective effect of cardioplegia may be lacking and lead to inadequate myocardial protection. PLSVC should be clamped to prevent the cardioplegia solution during retrograde cardioplegia to direct upwards to PLSVC and its branches.

The last issue about the presence of PLSVC is catheterization procedures. Central venous catheterization and placement of pacemaker leads can be arduous due to the relatively tortuous course (16). Manipulation of the guidewire (due to the proximity to the coronary sinus) may cause hemodynamic instability, arrhythmias, perforation of the heart, and tamponade (17).

The anatomy of the left brachiocephalic vein and the point of drainage of PLSVC is an essential point in the decision-making process of congenital cardiac surgery. The presence of right to left shunt requires surgical correction. These patients seen frequently desaturated because of venous blood draining to the left atrium (18). In the presence of PLSVC, this desaturation is mild, as the oxygen saturation of the patients in the room air is between 85-90%. Besides the desaturation, there is a risk about; cerebral embolization, brain abscess, and stroke (18,19). Many techniques are described in the literature. The persistent left SVC drainage to the left atrium can be accomplished through extracardiac or intracardiac techniques (20,21). Several surgical procedures to correct this anomaly have been reported, including ligation of the left SVC, intra-atrial redirection of flow from the left SVC to the right atrium, and reimplantation of the left SVC into the right atrium, pulmonary artery, or SVC (20, 21). In our study, in 5 cases with PLSVC-LA connection, two patients had the absence of left brachiocephalic vein, and the PLSVC was separated from the left atrium and anastomosed to the left pulmonary artery (Glenn shunt). In 2 other cases with ASD and common atrium, the PTFE patch was rerouted the PLSVC into the right atrium, and atrial septal defects were closed. In isolated case with normal left brachiocephalic vein, ligation was performed because of failed percutaneous intervention and right femoral arterial emboli due to coil.

LIMITATIONS

There are several limitations to our study. The patient number was limited, and this was a retrospective observation in one single hospital. The study focused on only the short-term outcomes, and the patients' inclusion criteria for the study population included only patients who underwent surgery for congenital heart disease. Despite these drawbacks, we still believe that our findings were relevant.

CONCLUSION

Consequently, PLSVC is relatively infrequent and asymptomatic under normal circumstances but can be crucial in the setting of congenital cardiac surgery. In the case of lack of knowledge about these anomalies may be problematic in the operative field, and in the intensive care period, the healing may be interrupted. In our cases, there was no complication during the operation or in the postoperative course. Knowledge of systemic venous anatomy before cardiac intervention is vital for the safe execution of any cardiac procedure. Before planning any procedure for the patient, the cardiac anatomy must be explained in detail. So the planned surgical procedure can be done more safely and accurately.

Competing Interests: The authors declare that they have no competing interest.

Financial Disclosure: There are no financial supports.

Ethical Approval: The institutional ethics committee approved our study design (Decision date: 27.04.2020, Decision no:2020/5-15).

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