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Gajendra Agrawal *

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Review Article

Persistent Left Superior Vena Cava Draining into Left Atrium: A Brief Review

Anuj Chaturvedi [1], Gajendra Agrawal [2*]

[1] (General Medicine), Senior Resident (Cardiology), JNMC, DMIHER.

[2] (Cardiology), Professor and Head of the Department of Cardiology, JNMC, DMIHER.

***Correspondence Author:** Gajendra Agrawal, MBBS, MD, DNB(Cardiology), Professor and Head of the Department of Cardiology, JNMC, DMIHER.

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Abstract

Persistent left SVC (LSVC) is a relatively rare vena caval anomaly that can be seen in 0.3% of asymptomatic healthy individuals but increases in prevalence with presence of other congenital cardiac anomalies [1]. Association of other anomalies and symptomatic disease may require surgical intervention. Intracardiac and extracardiac approaches are adopted for rerouting the PLSVC flux. Knowledge of PLSVC is necessary for certain invasive procedures to avoid complications during such interventions. In this review article, we briefly discuss the current evidence with LSVC draining in to LA along with various surgical approaches.

Keywords: LSVC; UCSS; Heterotaxy; Surgical intervention; TAPVC

Introduction

Superior vena cava (SVC) anomalies are detected incidentally while performing some cardiac procedures. Persistent left SVC (LSVC) is a relatively rare vena caval anomaly that can be seen in 0.3% of asymptomatic healthy individuals but increases in prevalence with presence of other congenital cardiac anomalies [1]. Draining of LSVC in left atrium (LA) in absence of coronary sinus (CS) can be seen in unroofed coronary sinus syndrome (UCSS) and heterotaxy syndrome [2,3]. Echocardiography aids in the diagnosis of LSVC with identification of LSVC along suprasternal axis without dilated coronary sinus [4]. Surgical management is necessary with adoption of intra- and extra-cardiac techniques depending on the anatomy, age, associated anomalies, and cardiomyopathies [5]. Here, we briefly discuss the current evidence with LSVC draining in to LA along with various surgical approaches.

The Persistent LVSC: UCSS and Heterotaxy syndrome

The UCSS is characterized by partial or complete absence of CS. It is categorized as type I, II and III as total absence, partial absence with one or more anomalies in midportion and partial form of outlet, respectively [4]. The complete absence of coronary sinus is a part of Raghib's syndrome with LSVC draining in to the upper left LA and coronary sinus type atrial septal defect (ASD) [6]. Majority of cases (80% - 90%) are associated with absence of innominate vein [7]. Other anomalies can be associated with UCSS such as tetralogy of Fallot, double outlet right ventricles, etc [6,7]. The heterotaxy syndrome (HS) is associated with abnormal distribution of internal thoracic and abdominal organs along the left to right axis of the body. Isomerism of atrial appendages seen in HS indicates same morphology of atrial appendages as either atrium. These abnormalities are frequently associated with persistent LSVC draining into LA [5]. Compared to left atrial appendage isomerism, extracardiac total anomalous pulmonary venous connection (TAPVC) is common in right atrial appendage isomerism with universal absence of coronary sinus. In left atrial appendage isomerism, the most common anomaly is discontinuation of intrahepatic inferior vena cava with continuation of azygous/hemiazygous vein. Thus, LSVC drains into the coronary sinus in these cases [3,8]. These differences are essential to understand for optimal surgical approaches in presence of other anomalies and cardiomyopathies.

Diagnosis of LSVC

Echocardiography (ECHO) is the first diagnostic modality. Visualization of LSVC along suprasternal axis without dilated coronary sinus suggests persistent LSVC. In case of doubts, air-bubble ECHO study can detect bubble in LA before right atrium (RA) if LSVC is draining into LA. Furthermore, computed tomography with contrast and magnetic resonance imaging helps in delineation of cardiac anatomy along complete identification of other vascular and cardiac anomalies [9].

Current Evidence with persistent LSVC draining in LA

Table 1 [10-17] highlights the reports of LSVC draining in LA along with surgical approaches as described in the reports. Persistent LSVC (PLSVC) is detected incidentally. It can be commonly associated with atrial septal defect (ASD) as seen in majority of reports. PLSVC drainage is commonly in RA (nearly 80-90% cases) but LA drainage is also seen (10-20% cases). LA drainage can occur in LA appendage, left pulmonary veins or via coronary sinus [18]. The LA drainage may result in right – left shunt. Enlargement of CS is seen in most cases but may not reach to level of aneurysm formation.

Generally, PLSVC may not be symptomatic as most cases are detected incidentally. Clinical significance lies in knowing its presence especially when performing the central venous catheter insertion, during cardiac resynchronization therapy and pacemaker implantation [19,20].

Depending on presence of other anomalies, surgical techniques may vary. In the unroofed CS, two-patch repair technique involves rerouting of the flux from the LSVC to the interatrial septum plane followed by closure of ASD. In single patch repair, flux is directed to tricuspid valve and closure of ASD is done. Two patch repair technique for reconstruction of CS involves rerouting of the flux from the LSVC to the tricuspid valve followed by ASD closure. In the extracardiac approach, hypoplastic innominate veing can be expanded using the autologous pericardial patch along with ligation of LSVC drainage in LA roof. In absence of innominate vein, LSVC disconnected from the LA and can be reconnected with RA appendage or RSVC behind the aorta [5].

Author (year)	Age/gender	Finding	Surgical technique
Meadows and Sharp (1965) [10]	36 years / Male	PLSVC draining in LA Right to left shunt without peripheral desaturation Coarctation of the aorta	-
Soward et al. (1986) [11]	32 years / Female	PLSVC draining in LA LV outflow tract obstruction ASD Incidental detection during catheterization from left arm	-
Komai et al. (1996) [12]	4 years / Female	PLSVC draining in LA (upper left quadrant) ASD Absent innominate vein	Construction of internal conduit with inverted flap of LA appendage ASD closure with autologous pericardium
Ramos et al. (2005) [13]	1 month / Male	Complete ASD PLSVC draining in LA Situs solitus Absent RSVC	PTFE conduit: IV and the RA appendage
Raj et al. (2010) [14]	76 years / Male	PLSVC draining in LA Unroofed CS ASD Pulmonary Hypertension	-
Tobbia et al. (2013) [15]	40 years / Male	PLSVC draining in LA (diagnosed on MRI angiography) Right to left vascular shunt Normal ventricular function	-
Zhong et al. (2015) [16]	4 years / Male	PLSVC draining in LA without unroofed CS Dextrocardia Dilated RA, and RV ASD	Extracardiac conduit using polytetrafluoroethylene (PTFE) graft: connect LSVC and RA appendage Closure of ASD with autologous pericardium Three years follow-up without complications
Bisoti et al. (2017) [17]	43 years / Male	PLSVC lateral to LA dilated CS Severe Aortic stenosis Absent RSVC	-

Table 1: Current reports with PLSVC draining in LA (This table is the sole creator of the author)

Conclusion

Persistent left superior vena cava draining in to left atrium is a rare and incidental finding. Majority of cases remain asymptomatic. Association of other anomalies and symptomatic disease may require surgical intervention. Intracardiac and extracardiac approaches are adopted for rerouting the PLSVC flux. Knowledge of PLSVC is necessary for certain invasive procedures to avoid complications during such interventions.

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