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Review Article

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Persistent Left Superior Vena Cava (Plsvc) Beyond Anatomy: A Cardiologist's Guide

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ABSTRACT

Background: The persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly, occurring in 0.3–0.5% of the general population and in 10% of patients with congenital heart disease (CHD). Historically considered benign, PLSVC is increasingly recognized for its clinical significance in the field of interventional cardiology, electrophysiology, and surgery.

Objectives: The present narrative review combines available evidence regarding PLSVC, focusing on its embryology, anatomical variants, diagnostic challenges and tailored management strategies to address clinical uncertainties and improve procedural outcomes

Key Points

Embryology and Anatomy: PLSVC occurs because of the absence of regression of the left anterior cardinal vein, in most cases being linked with chromosomal defects (e.g., trisomy 21) and neural crest cell migration defects. Three subtypes occur: Bilateral SVC (with/without innominate vein) and isolated PLSVC.

Clinical Significance: Left atrial drainage (8 to 10% of the cases) leads to paradoxical embolism, stroke, or cyanosis. Unknown PLSVC renders central venous catheterization, pacemaker placement, and catheter ablation technically difficult due to the anomalous anatomy. Associated arrhythmias, stroke, and CHD must be diagnosed with high suspicion.

Diagnosis: Echocardiography with bubble study, CT/MRI angiography plays a critical role in the diagnosis of route and concomitant CHD.

Management: Surgery/percutaneous correction (e.g., stent placement, venous rerouting) is critical for patients with right-to-left shunting/symptoms, but symptom-free patients may not necessarily undergo intervention.

Conclusion: The clinician needs to understand the risk for right-to-left shunting of PLSVC, procedural risk during placement of the catheter or device, and its association with arrhythmias and stroke. Common in high-risk patients (e.g., in congenital heart disorders, unexplained hypoxia). Early detection by advanced imaging and forward procedural planning is key in reducing complications and optimizing patient outcomes.

Keywords: Persistent Left Superior Vena Cava, Congenital Heart Disease, Anatomical Variation, Paradoxical Embolism, Multimodal Imaging, Interventional Cardiology

Introduction

The inability of the anterior cardinal veins to regress during embryological development is known to cause persistent left superior vena cava (PLSVC) [1]. PLSVC is the most prevalent anomaly of the thoracic venous system, affecting 0.3 % of the general population with a much higher percentage in patients with congenital heart defects [2]. Historically, it was considered an asymptomatic benign vascular anomaly and discovered incidentally. But with advancement in the field of cardiology, serious implications of this anatomical variation are faced by the surgeons during interventional and device-based therapies [3,4].

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Understanding of PLSVC is important for the safe and effective central venous catheterization, pacemaker or ICD implantation, and cardiac procedures. Undiagnosed PLSVC may cause procedural difficulties and iatrogenic complications like arrhythmias [5]. Furthermore, its association with CHD, conduction defects, and paradoxical embolism highlights the need for thorough awareness [6]. Recognition of this anomaly is crucial, particularly in preoperative planning and intraoperative decision making.

Despite the advancement in imaging modalities like MRI, CT angiography, and endocardiography that have improved the detection, there is a serious lack of standardized diagnostic procedures, management guidelines, and risk stratification strategies [7]. Patients with other cardiac conditions are at higher risk of complications.

The majority of the literature is scattered in the form of case reports and clinical settings. This review aims to connect the up-to-date research with anatomical, diagnostic, and procedural knowledge to close the information gaps and to give cardiologists a clear and useful evidence–based framework for identifying and treating PLSVC in various clinical settings.

Embryology and Anatomy

During the 8th week of embryonic development, the failure of the anterior cardinal vein to regresses and redistribute the venous flow to right superior vena cava forms PLSVC [1,6]. It is frequently associated with chromosomal and genetic abnormalities like trisomies 18 and 21, Turner syndrome, VACTERL, CHARGE, and 22q11 deletion. Research shows that even in isolated cases of PLSVC there is a 7-12.5% possibility of chromosomal abnormality [2,8]. Its association with Waardenburg syndrome suggests that a possible disturbance in neural crest cell migration may impact the mesoderm–derived vasculature during development [9]. There is no current evidence of precise gene or signaling pathways that may cause PLSVC.

Anatomically the anomaly is subdivided by the presence or absence of the innominate (left brachiocephalic) vein. In bilateral PLSVC cases, this vein is absent 65%of the time indicating a lack of a typical venous bridge [10]. PLSVC drains into the right atrium through the dilated coronary sinus in 92% cases. About 8-10% of the time PLSVC drains into the left atrium due to atrial isomerism or an unroofed coronary sinus. [1,11]. This may cause a right–to-left shunt and increase the risk of systemic embolism. These variations which are sometimes described as Raghib syndrome, usually have more serious clinical implications [12]. The main anatomical subtypes of PLSVC are:

- Bilateral SVC with the innominate vein
- Bilateral SVC without the innominate vein
- Isolated PLSVC (RSVC absent) [13].

Each of these combinations needs a unique procedural approach and planning for the intervention. Different imaging modalities like MRI, CT angiography, and Echocardiography help in identifying the variant and subsequent pre-planning of the procedure. Cardiologists and cardiac surgeons can anticipate difficulties and prevent complications, and customize the procedural approach with comprehensive knowledge of the encountered variation. This emphasizes the significance of thorough preoperative imaging and standardized classification schemes, which currently need more research and clinical consensus.

Epidemiology And Clinical Association

PLSVC occurs in 0.3-0.5% of the general population and this occurrence is much more common in patients with congenital heart diseases [14]. Ebstein anomaly is and endocardial cushion defect with a prevalence of less than 1%, this endocardial defect is also associated with a Persistent left sided Superior vena cava [15].

Hemodynamic and physiological Implications

In cases of stroke after investigations it was found out that a PLSVC had created a paradoxical pathway for the embolus because of the bypassing of the lungs by right to left shunt offered by the PLSVC, the clot instead of going to the lungs went straight to the brain causing a stroke [16].

Electrophysiologic and Arrhythmic Consideration

Although Persistent Left Superior Vena Cava (PLSVC) is frequently asymptomatic, its existence—especially when linked with coronary sinus agenesis or drainage into the left atrium—can have significant effects on electrophysiology. Structural changes may result in the compression of the atrioventricular node or His bundle, potentially leading to the emergence of atrial or ventricular arrhythmias. Moreover, right atrial enlargement, as observed in this instance, may further heighten the risk of arrhythmias.

In cases of complex congenital heart disease and remaining shunts, PLSVC should be acknowledged as a possible source of arrhythmias, even in the lack of obvious clinical signs. This consideration becomes particularly important during dynamic phases such as the postpartum period, where hemodynamic changes can reveal hidden conduction abnormalities [17].

Diagnostic Modalities

The diagnostic evaluation of Persistent Left Superior Vena Cava (PLSVC) relies on high-grade clinical suspicion and multimodal imaging in a strategic approach. Although frequently identified incidentally—and most prominently during the placement of a device or during the placement of a central line—PLSVC has several characteristic appearances on imaging modalities that, in appropriate interpretation, establish the diagnosis and its clinical relevance [18].

Transthoracic Echocardiography (TTE)

TTE is often the first imaging modality to create suspicion for PLSVC. An enlarged coronary sinus (CS), especially in the context of normal right atrial pressure or no discernible structural CS abnormality, is the classic finding. Diagnostic confirmation is obtained through a bubble study: contrast in the form of agitated saline in the left antecubital vein opacified the CS before the right atrium if the PLSVC joins the CS. If the PLSVC directly joins the left atrium (LA), the contrast avoids the right heart altogether and reaches the LA first, indicating a right-to-left shunt [19,20].

Transesophageal Echocardiography (TEE)

TEE provides high spatial resolution, in particular for imaging posterior cardiac structures such as the coronary sinus. It is able to delineate:

Premature contrast opacification of the LA in the case of aberrant PLSVC drainage,
Enlargement or agenesis of the coronary sinus,
Associated anomalies such as unroofed coronary sinus, atrial septal defect (ASD), or partial anomalous pulmonary venous return (PAPVR) [6].

TEE is especially valuable in planning for surgery or electrophysiological study, where specific localisation of structures is desirable [21].

Contrast Echocardiography (Bubble Study)
This examination, conducted at either TTE or TEE, uses agitated saline injected in the both the right and the left arms to dynamically evaluate venous return. Early visualization of contrast on the left before visualization on the right determines PLSVC drainage to the LA or CS, respectively, according to the flow direction of the contrast tract. This is especially useful in situations where CS anatomy is in question or right-to-left shunting is expected [18,22].

Cardiac Computed Tomography (CT Angiography)
CT cardiac imaging is the established noninvasive standard for the diagnosis of PLSVC and evaluation of associated anomalies. It defines the origin, course, and drainage site of the PLSVC—to the CS, LA, or systematic veins. CT also works well for the diagnosis of coronary sinus agenesis, bridging veins, or complex venous anomalies such as Raghib syndrome. Three-dimensional reconstruction is also particularly valuable in preprocedural planning [17,20].

Cardiac Magnetic Resonance Imaging (MRI)
The cardiac MRI, in particular with contrast-enhanced or phase-contrast sequences, is a non-radiation technique appropriate for patients with renal insufficiency or pediatric patients. The technique provides good visualization of the venous tract and is able to assess the shunt volume and the direction if the PLSVC opens into the LA. The MRI also helps in the delineation of the associated CHD, especially in heterotaxy syndromes or complex anatomy [19,21].

Invasive Catheter Angiography (Venography)
While less frequently needed as an individual diagnostic testing modality, catheter-based venography is the absolute standard in the situation where the non-invasive imaging modalities are indefinite. PLSVC is usually found by accident during central line placement or electrophysiological studies, where the catheters take the unusual course. It permits in-time flow visualizing and venous mapping [18,22].

Intraoperative or Procedural Detection
PLSVC can be found incidentally during:
Pacemaker or cardiac resynchronization therapy (CRT) lead placements, where the lead trajectory deviates from expected anatomy and requires fluoroscopic or venographic confirmation,
Cardiac surgery, especially if retrograde cardioplegia delivery is not effective or if coronary sinus cannulation is challenging, for suspicion of unusual venous return [17].

Implication of Cardiac Device Implant
In cases of Atrial fibrillation when catheter ablation is required for its treatment, a PLSVC causes problems in visualization of the right atrium and right atrial appendage through CT scan, the usual atrial puncture cannot be performed because of the abnormal anatomy caused by PLSVC. This creates procedural difficulties and may require intracardiac echocardiography to properly visualize and plan management for Atrial fibrillation [23]. Sometimes it is asymptomatic throughout life and may present with conduction anomalies like heart blocks, arrhythmias imaging in these cases reveals occluded coronary sinus which means that there is no connection between Left atrium and right atrium [24].

Interventional and Surgical Challenges
Cardiac pacemaker placement possesses another challenge in cases of PLSVC where conventional methods provide no significance, deviation must be done from the norms, the usual transvenous lead cannot be placed because of the occluded Coronary Sinus and distorted anatomy so another approach like Sternotomy is performed. Post-op complications in these cases include death, heart transplants and atrial fibrillation [25].

PLSVC Draining into the Left Atrium: A Clinical Dilemma
A clinically significant right-to-left shunt is formed by direct drainage into the left atrium (LA) in rare cases. On the other hand, the persistent left superior vena cava (PLSVC) typically leads the blood to the right atrium through the coronary sinus. Such a condition can be a source of symptoms like paradoxical embolism, cryptogenic stroke, cyanosis, or systemic desaturation, especially in the case of children or adults having congenital heart disease that has been corrected [6,24]. The PLSVC-LA case only occurs once and is usually an incidental finding during imaging for unexplained hypoxia or as a part of preoperative assessment. Al-Muhaya et al. have provided examples of two pediatric cases with PLSVC that drain into the LA. Among the investigations, only contrast echocardiography and CT angiography have been used to establish the diagnosis, and the only clinical feature has been chronic desaturation [3].

Furthermore, Duong et al. have presented a case of a 26-year-old lady with exertional cyanosis and exhaustion. Cardiac MRI and angiography were thus employed to depict PLSVC-to-LA drainage [4].

Serious health problems of a clinical nature might accompany this abnormality. Such symptoms are cyanosis and hypoxemia, which are generally seen if no other structural defects in the heart are present [4,19]. In certain cases of paradoxical embolism and stroke that were not expected, the incidents happened at such high-risk periods as pregnancy. A case was presented of a patient who had a double outlet right ventricle repair and an uncorrected PLSVC draining into the LA. The patient subsequently had a stroke during pregnancy [5]. This condition may even cause the continuous manifestation of symptoms or at least the surgical failure if there is no treatment for it.

Management Strategies and Follow-Up
Most patients of persistent left superior vena cava (PLSVC) that leak into the coronary sinus are asymptomatic and do not require surgical correction [6]. Before conducting invasive operations

such as cardiothoracic surgery, pacemaker installation, or central line insertion, it is of great importance to recognize the abnormality to avoid technical problems and possible misplacement of the device [26].

To deal with the very rare case of this variation, methods both surgical and interventional have been described. For instance, to redirect the PLSVC flow, Altin et al. have come up with a unique surgical procedure by which the left atrial appendage was freed and anastomosed to the right atrium. This technique was applied to a 3-year-old child, and the cyanosis was eliminated; also, the child was not affected in any way after the operation [27]. In a percutaneous process, Duong et al. have indicated that a covered stent is positioned at the PLSVC orifice; thus, the blood that comes from the LA is rerouted to the systemic venous circuit. This operation has further confirmed, at the one-year follow-up the stent is open and the symptoms are completely managed [20]. Surgical closure of the PLSVC is an option that can be done with success in cases where sufficient right-sided venous return is proven before surgery, and also if the individual possesses the bridging innominate vein [28].

Patients with surgically or percutaneously treated have positive results. The intracardiac rerouting method has shown excellent oxygenation results with minimal complications [8]. Results of the percutaneous covered stent approach have been very positive, especially in older patients, where open surgery might be more dangerous [20]. Surgical ligation is still a possible method in some anatomical configurations if it is done with caution to avoid venous congestion [28].

The existence of a bridge vein, the exact drainage architecture of PLSVC, the accompanying congenital cardiac abnormalities, and institutional competence are the elements that decide the extent to which surgery or transcatheter repair is the best choice. To help with detailed preoperative planning and avoid unexpected situations during the surgery, the use of advanced imaging modalities, e.g., cardiac MRI, CT angiography, and contrast echocardiography, is very important [6,20,28]. Patients with this rare but greatly severe abnormality can have an optimistic long-term outcome if the diagnosis is made at the right time and anatomically correct treatments are implemented [29].

Regardless of the procedure chosen, follow-up over a longer time is essential. When the operation is being performed and the surgeon is handling the area near the atrial septum or the coronary sinus, the patient is at risk of developing some arrhythmias, like atrial fibrillation, for instance [27]. Those patients who have cardiac implanted electronic devices and these are inserted through a PLSVC have to be under close observation since they are at risk of venous stenosis, lead dislodgement, or the problem of pacing failure [27]. Regular check-ups together with oxygen saturation, echocardiography, and ECG will make sure that the hemodynamic stability is there and will identify any problems at an early stage [26,28].

Conclusion

Persistent Left Superior vena cava (PLSVC), often a silent variant of the normally occurring right sided Superior vena cava is of utmost importance for healthcare professionals. Although its prevalence is particularly low in the normal population, those with congenital heart disease carry an increased risk of this anomaly.

PLSVC is hard to visualize by our conventional imaging techniques and is often missed on screening of the patients. Advanced imaging techniques like CT/MRI and intraoperative imaging modalities are required to visualize it.

Since the anatomy of the normal heart is distorted by the PLSVC, the coronary sinus is often atretic and incidence of stroke is these patients is very high because of formation of a right to left shunt bypassing the lungs. Furthermore because of the distorted anatomy patients are predisposed to arrhythmias most commonly Atrial fibrillation. To cure Atrial fibrillation usually a catheter ablation is performed which cannot be performed in these cases because the transvenous lead cannot reach the left atrium. Special types of catheters and right sided access is required in this case to curb this problem.

Management of a PLSVC is often surgical with percutaneous treatment showing the best results, that being said patients are still at risk of developing arrhythmias and continuous cardiac monitoring is essential.

Moving forward awareness about this disease is necessary to be given to the general population so that it can be diagnosed and appropriately managed early on without causing much problems in the future. Better screening methods and imaging modalities are essential for diagnosis and their availability should be made certain at all primary health care centers so that the patients can avail them without losing much time in the process.

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