

Right Heart Catheterization Through Persistent Left Superior Vena Cava, an Extremely Rare Procedure and Review of Current Literature



Abdel Rahman Al Emam¹, Ahmed Sami Abuzaid^{2*}, Mohammad Chamsi-Pasha³ and Nattapong Sricharoen⁴

¹University of Nebraska Medical Center, Department of Cardiology, 982265 Nebraska Medical Center, Omaha, NE 68198-2265, USA; ²Sidney Kimmel Medical College at Thomas Jefferson University/Christiana Care Health System, Cardiology department, Heart and Vascular Institute, Delaware, Newark, USA; ³University of Nebraska Medical Center, Department of Cardiology; ⁴University of Nebraska Medical Center, Department of Cardiology, USA

Abstract: Persistent left superior vena cava (PLSVC) is encountered occasionally during angiographic procedures. It usually coexists with right superior vena cava and drains to the right atrium through the coronary sinus, but multiple variations are described. Although PLSVC is extensively reported in the literature, there are very few articles addressing right heart catheterization in patients with isolated PLSVC. We present a patient with absent right superior vena cava and PLSVC draining to a dilated coronary sinus diagnosed during right heart catheterization in the setting of pulmonary hypertension. We were able to safely complete the procedure through the right internal jugular vein. Transthoracic echocardiography and chest CT scan were consistent with this finding. Although clinically silent most of the time, undiagnosed PLSVC can lead to catastrophic consequences when the patient undergoes invasive procedures. If PLSVC is suspected, the anatomy of the thoracic venous system must be identified before invasive cardiac procedures.

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INTRODUCTION

Persistent left superior vena cava is the most common congenital anomaly of the systemic venous return. Although clinically silent most of the time, undiagnosed PLSVC can lead to catastrophic consequences when the patient undergoes invasive procedures. We present a patient who underwent successful right heart catheterization through the right internal jugular vein, persistent left superior vena cava and dilated coronary sinus.

OUR CASE

A 52-year-old female patient with hepatitis C related cirrhosis, non-insulin dependent diabetes mellitus and hypertension presented to our emergency department with shortness of breath. Her transthoracic echocardiography showed normal left and right ventricular function, moderately dilated right ventricle, and elevated pulmonary artery systolic pressure at 75 mmHg. We proceeded with right heart catheterization for further evaluation of pulmonary hypertension through the right internal jugular vein. However, after getting access, we noticed that the micro-puncture wire went to

the left side of the chest (Fig. 1). After a venous puncture was confirmed by oxygen saturation, we inserted the micro-puncture sheath and injected contrast which revealed persistent left superior vena cava (PLSVC) draining to a dilated coronary sinus (Fig. 2) in the absence of right superior vena cava. We advanced the Swan Ganz catheter through the PLSVC and the coronary sinus (Fig. 3) and obtained right atrial (6 mmHg), right ventricular (76/11 mmHg), pulmonary artery (78/33 with a mean of 50 mmHg) and wedge (12 mmHg) pressures. Pulmonary vascular resistance was elevated at 4.8 wood units. Results were consistent with pulmonary arterial hypertension due to portal hypertension and chronic liver disease (Group 1 associated PAH). A chest CT scan showed again the absence of right superior vena cava and a PLSVC that drains to the right atrium through a dilated coronary sinus (Fig. 4A and B). Retrospective review of our echocardiography showed dilated coronary sinus with instant opacification with agitated saline contrast injection into left arm vein (Fig. 5A and B), consistent with persistent left superior vena cava. The patient was started on sildenafil and 2 month follow-up echocardiogram showed improving pulmonary artery systolic pressure to 55 mm Hg.

DISCUSSION

Persistent left superior vena cava (PLSVC) is the most common congenital anomaly of the systemic venous return

*Address correspondence to this author at the University of Nebraska Medical Center, Department of Cardiology, 982265 Nebraska Medical Center, Omaha, NE 68198-2265, USA; Tel: (402) 559-9252; Fax: (402) 559-8355; E-mail: emammd@gmail.com

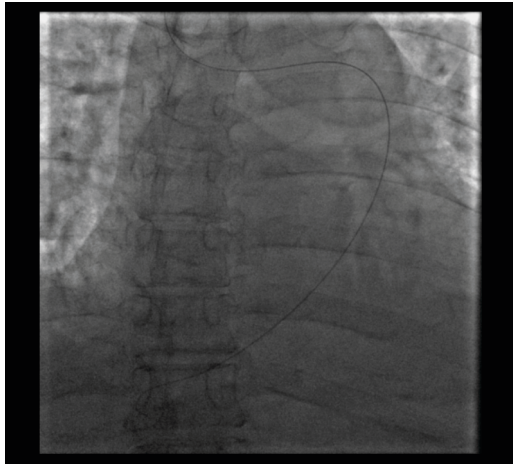


Fig. (1). Right heart catheterization showing after accessing the right internal jugular vein, micro-puncture wire went to the left side of the chest.

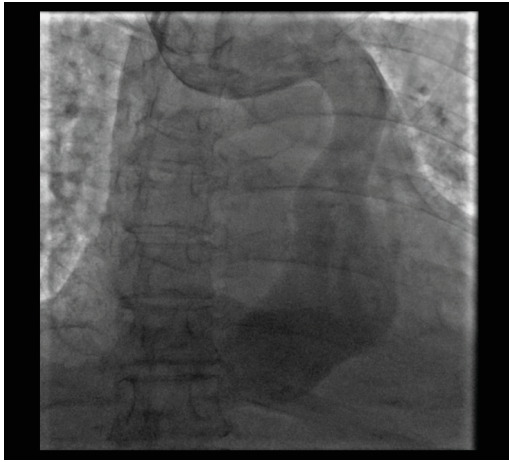


Fig. (2). Venography showed which revealed persistent left superior vena cava (PLSVC) draining to a dilated coronary sinus.

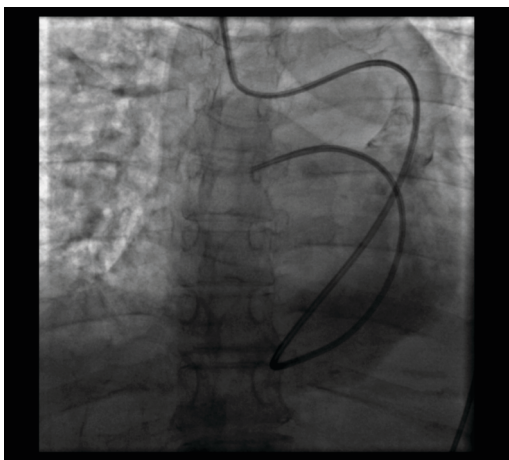
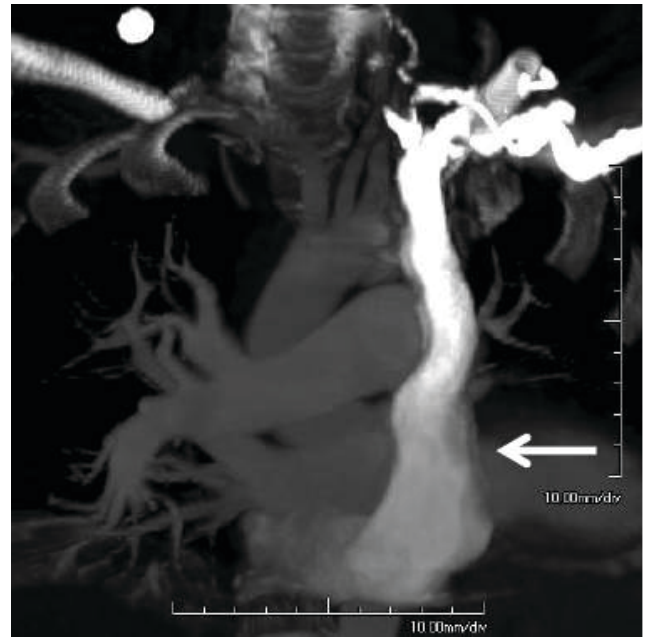
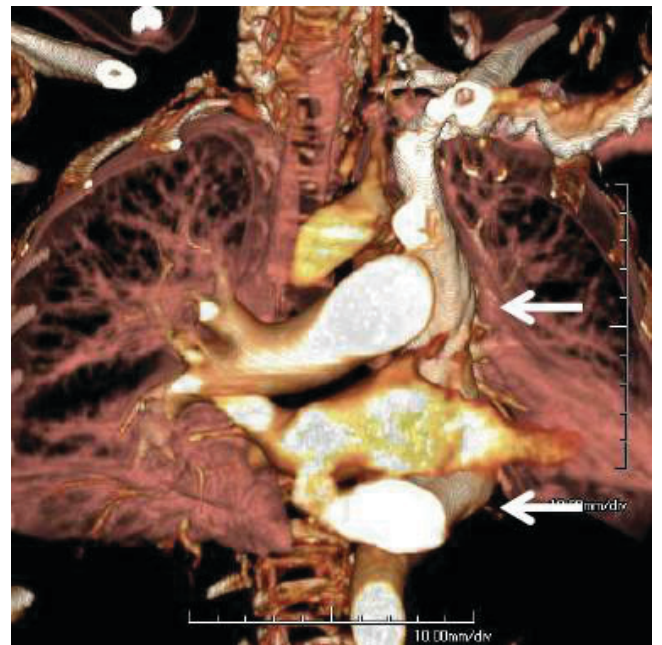


Fig. (3). Advancing the Swan Ganz catheter through the PLSVC and the coronary sinus.



4A



4B

Fig. (4). A and B: Chest CT scan showing the absence of right superior vena cava and a PLSVC (arrows) that drains to the right atrium through a dilated coronary sinus.

[1]. Although there is a debate about who was the first to report this anomaly, it was first reviewed in depth by Marshall in 1850 [2]. PLSVC results from the failure of regression of the caudal part of the left arterial cardiac vein [3]. It is really hard to estimate the prevalence of PLSVC as it is frequently asymptomatic, but available literature suggests a

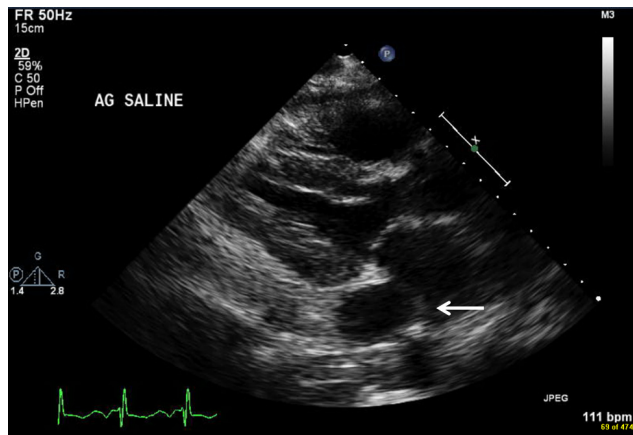


Fig. (5A). Transthoracic echocardiography (Parasternal Long Axis view) showing dilated coronary sinus (arrow) on echocardiography.

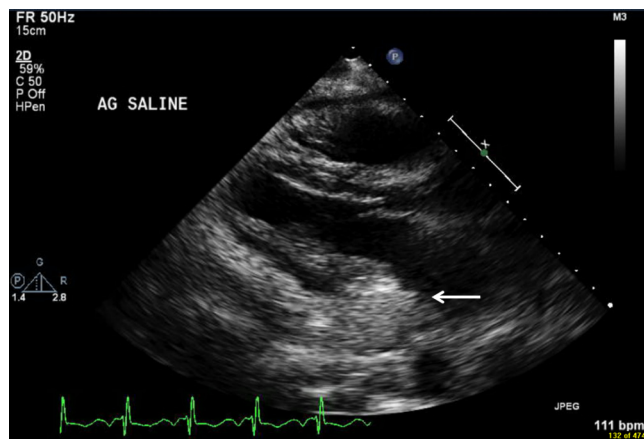


Fig. (5B). Transthoracic echocardiography (Parasternal Long Axis view) showing Instant opacification of the coronary sinus (arrow) with agitated saline contrast injection into left arm vein.

prevalence of 0.1-0.5 % [1]. In patients with other congenital anomalies, however, its prevalence may be up to 12.9% [1].

PLSVC coexists with right superior vena cava in up to 90% of the time, in such scenarios [1], the left innominate vein may be completely absent in up to approximately 65% of such cases [4]. In about 10% of cases, the PLSVC will be responsible for the venous drainage of the upper part of the body. PLSVC drains to the right atrium through a coronary sinus (CS) in about 80-92% of cases, while in the remaining 8–20% the LSVC drains into the left atrium and creates a right to-left shunt [1].

The most common cardiac anomalies associated with PLSVC are ventricular septal defects, atrial septal defects and pulmonary stenosis [5], while the most common associated non-cardiac anomaly is esophageal atresia [1]. Multi-organ congenital anomalies have also been reported, including VACTERL (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities or trisomy 21 [6].

In the majority of patients, PLSVC is clinically silent as it drains to the right atrium. Furthermore, the presence of right superior vena cava makes it even less likely to be inci-

dentally discovered during central venous catheterization through the right internal jugular or subclavian veins. Most of the time PLSVC is diagnosed at the time of central venous catheterization (either from the left side, or, in the case of absent RSVC, from the right side), chest imaging for other indications (e.g. dyspnea) and cardiac procedures. Our case was unique that it was diagnosed during right heart catheterization. In the minority of patients where the PLSVC drains to the left atrium, the patient will have right to left shunt. The hemodynamic consequences of this shunt will depend on the existence of an associated RSVC and communication through the left innominate vein between both SVCs. Patients with significant shunts may have cyanosis and clubbing [1, 3]. These patients are also more prone for paradoxical systemic embolization, either thrombotic or air embolization [2]. In cases of double SVC when PLSVC drains to the left atrium, there are very few reported cases of left to right shunt [1]. Rarely, patients present with arrhythmias secondary to persistent left sided fetal pacemaker tissue or stretching of the atrioventricular node or bundle of His [7-10]. Although clinically silent most of the time, undiagnosed PLSVC can lead to catastrophic consequences when the patient undergoes invasive procedures or cardioplegia [11, 12]. With caution, we demonstrated that right heart catheterization through a PLSVC draining to a dilated CS can be safely performed. Also, there have been reports in the literature that dialysis catheters and pacemaker wires can be inserted with no complications. However, special instruments may be needed during pacemaker implantation [13].

Diagnosis modalities include: Chest X ray can show widening of the left superior mediastinum, but this finding is nonspecific. It may also show malpositioning of the central venous catheters. Transthoracic echocardiography (TTE) with contrast is noninvasive, cost effective and radially available method to diagnose PLSVC. With contrast in injection through the left upper extremity, opacification of the CS before the right atrium confirms the diagnosis of PLSVC. It usually shows dilated CS. However, the latter is a nonspecific sign as it can be seen in isolated atresia or stenosis of the CS, partial anomalous hepatic venous connection to CS, partially unroofed CS, anomalous pulmonary venous return to CS, coronary arterio-venous fistula, and aneurysm of the CS [1, 8]. On the other hand Computed Tomography (CT) scan without contrast may be helpful but for definitive diagnosis by CT intravenous contrast use is mandatory. Multi-planar reformatting at workstations offers an excellent opportunity to produce venography like images and helps in the assessment of the anomaly [1]. Magnetic Resonance Imaging (MRI) Although expensive and time consuming, MRI with or without contrast can confirm the diagnosis. MRI also supplies precise demonstration of the venous anomaly, flow direction, its drainage and associated cardiovascular malformations [1, 9, 10]. Ultimately Conventional venography has been the gold standard for diagnosis, however, the wide spread use of CT scan decreased the need for this invasive procedure.

CONCLUSION

Although clinically silent most of the time, undiagnosed PLSVC can lead to catastrophic consequences when the patient undergoes invasive procedures. If PLSVC is suspected,

the anatomy of the thoracic venous system must be identified before invasive cardiac procedures.

CONFLICT OF INTEREST

The authors confirm that this article content has no conflict of interest.

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Declared none.

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