

## Case Report

# When the Left is left!

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### ABSTRACT

Persistent left superior vena cava is an uncommon vascular anomaly; however it is the most common anomaly of the thoracic venous system. It may stand alone or associated with other congenital heart diseases and even other extracardiac anomalies. It is due to a lack of regression and adsorption of the left anterior cardinal vein. The persistence of this vessel renders a left subclavian approach for interventions on the right heart a challenge. It may be responsible for arrhythmias. We present a report of a persistent left superior vena cava draining into the coronary sinus with a coexisting normal right superior vena cava. Keeping in mind its widespread implications on cardiac procedures and a causative factor of cardiac disturbances we have considered its course, embryological source and clinical significance.

**KEYWORDS:** Anterior Cardinal Vein, Persistent Left Superior Vena Cava, Congenital Heart Disease, Dilated Coronary Sinus, Atrial Fibrillation.

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## INTRODUCTION

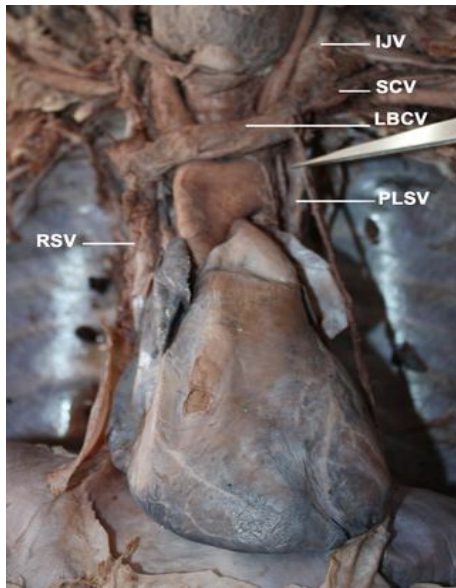
Formation of blood vessels takes place at around the 17<sup>th</sup> day of intrauterine life within the yolk sac. At about day 21, blood islands within the yolk sac may be observed. Central parts of the islands host formed elements, while the outer layers transform into tubular structures resulting in angiogenesis and thereby blood vessels [1]. Developing veins form three main systems, umbilical, vitelline and cardinal veins which carry blood into the sinus venosus. Due to the complicated process of development and regression of cardinal veins several anomalies may result. Persistent left superior vena cava results from disturbances in process of obliteration of left anterior cardinal vein what leads to its patency. As it has no impact on hemodynamics it is picked up during interventions when its

presence may be marked by complications or it may be directly responsible for arrhythmias.

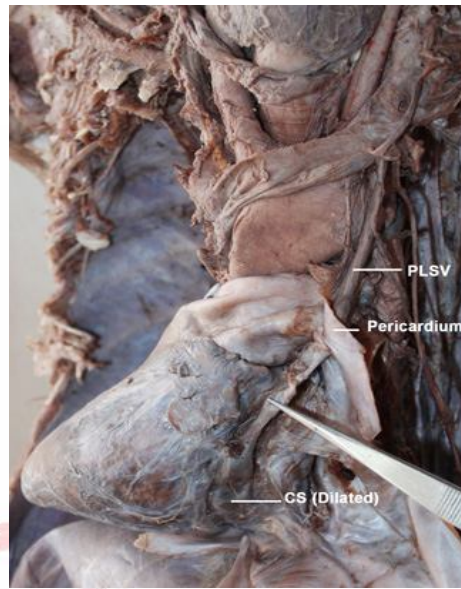
## CASE REPORT

During a routine dissection for the first year medical students of the Amrita School of Medicine, a Persistent Left Superior Vena Cava was noted with a coexisting normal right superior vena cava (Figure 1). The left superior vena cava extended from the first left intercostal space, running a straight vertical course anterior and to left of the aortic arch and pulmonary trunk. It pierced the pericardium at the third intercostal space and entered the markedly dilated coronary sinus at its left end (Figure 2). The dimensions of the right and left superior vena cava are stated in Table 1.

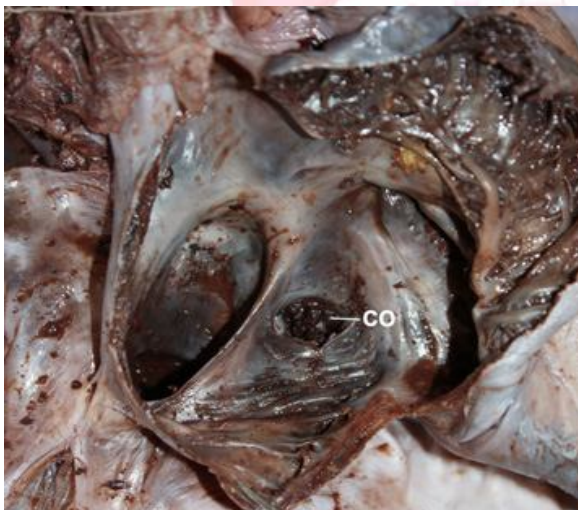
**Fig. 1:** Normal right superior vena cava (RSV) and persistent left superior vena cava (PLSV), Internal jugular vein (IJV), Subclavian vein (SCV), Left brachiocephalic vein (LBCV).



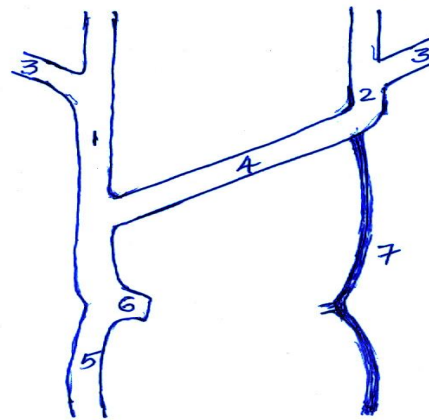
**Fig. 2:** Course of PLSV and its termination into Coronary sinus (CO).



**Fig. 3:** Interior of right atrium with enlarged coronary ostium (CO).



**Fig. 4:** Schematic diagram showing the development and fate of cardinal veins.



- 1- Right anterior cardinal vein
- 2- Left anterior cardinal vein
- 3- Subclavian vein
- 4- Oblique channel
- 5- Posterior cardinal vein
- 6- Right common cardinal vein
- 7- Involuting left cardinal veins caudal to the transverse Anastomosis.

The coronary sinus opened into the right atrium and a coronary ostium of diameter of 2 cm was noted (Figure 3). No other anomalies were observed.

**Table 1:** Parameters of right and left superior vena cava.

Parameters	Right SVC	Left SVC
Total Length	5.5 cm	9.5cm
Extrapericardial length	4.5cm	5.5cm
Intrapericardial length	1cm	4cm
Circumference	4.5cm	1cm

**DISCUSSION**

Resulting from persistence of the left anterior cardinal vein, left superior vena cava is the most common systemic venous anomaly seen in about 0.5% of the general population [2]. But it may be encountered in as much 10% of patients with congenital heart disease, especially cardiac transposition, bicuspid aortic valve, septal defects and TOF [3]. It is associated with oesophageal atresia among the extracardiac lesions.

In 82% of the patients with persistent left superior vena cava, a right sided superior vena cava is also present. Persistence of left superior vena cava with, absent right superior vena cava is much rarer with an occurrence of 0.09 – 0.13% of patients with congenital heart disease [4]. A dilated coronary sinus in the absence of right atrial enlargement on echocardiography should alert the physician about the possibility of a persistent left superior vena cava.

It was first described by Mc Cotter in 1916[5]. Edwards and Du Shane in 1960 have done exhaustive work on this anomaly [6].

During the 5<sup>th</sup> week of intrauterine development, three pairs of major veins can be distinguished – umbilical, vitelline and cardinal veins. The cardinal veins are somatic and appear as symmetrical structures. They are named as anterior cardinal draining the cephalad of the embryo and posterior cardinal veins draining the caudal part of the embryo. The anterior and posterior cardinal veins of each side unite to form the common cardinal vein or the duct of Cuvier which open into the right and left horns of the sinus venosus. An oblique anastomotic channel develops between the anterior cardinal veins causing a left to right shunt. The left anterior cardinal vein caudal to the channel disappears except at its cranial most part. The degenerated part forms a fibrotic cord called the left vena cava or the fold of Marshall. The seventh intersegmental vein joins the anterior cardinal vein cranial to the anastomotic channel and forms the subclavian vein.

The schematic diagram (Figure 4) represents the formation and fate of venous channels. The failure of involution of the left anterior cardinal vein to produce the ligament of Marshall results in the persistent left superior vena cava [7] as has happened in this instance. This drains into the coronary sinus and subsequently into the right atrium in 98% cases producing no significant hemodynamic changes but in the remaining 2% cases it drains into the left atrium through a deroofed coronary sinus producing a significant right to left shunt [8] resulting in paradoxical thromboembolism, air and septic embolism [9].

Although a left superior vena cava draining into

the coronary sinus has no adverse hemodynamic effects, it has immense impact on procedures which require upper limb venous access. For the interventional cardiologist challenges have been widely reported during transvenous pacing for bradycardia [10] and advanced device implantation [11].

For the critical care personnel, central venous cannulation may result in unusual catheter positions [12] and inadvertent coronary sinus cannulation may result in cardiac perforation [13]. Cannulation of the heart for cardiopulmonary bypass may result in retrograde cardioplegia [14]. Left superior vena cava has been associated with an increased risk of arrhythmias especially atrial fibrillation due to the enlarged coronary sinus stimulating the adjacent atrioventricular node [15]. The presence of dilated coronary sinus without any right atrial pathology should prompt the clinician to search for a persistent left superior vena cava and its presence should in turn be a clue to other congenital anomalies.

## CONCLUSION AND CLINICAL SIGNIFICANCE

Persistence of left superior vena cava may be missed as it causes no hemodynamic challenges but it may pose a problem during interventions and surgical procedures and may cause arrhythmias. Hence its awareness is of prime importance.

**Conflicts of Interests: None**

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