ABSTRACT

Introduction: Double superior vena cava with a Persistent Left Superior Vena Cava (PLSVC) is an uncommon abnormality but is the most common thoracic venous anomaly in the setting of SVC. It is estimated to occur in 0.3-0.5% of the general population and 3-10% of patients with other forms of congenital heart disease. The present study aimed to find the frequency of double superior vena cava, its embryological basis and to correlate its clinical significance.

Materials and methods: The present study, a descriptive study includes sixty cadavers allotted for first year MBBS students in Department of Anatomy at Velammal Medical College, Madurai during year 2013-2018. Superior vena cava along with other veins had been traced and studied in detail.

Results: Double superior vena cava was found in 2 cadavers out of 60 cadavers (3.33%). Persistent left superior vena cava opened into an enlarged coronary sinus that further drained into the right atrium in both the cases. Left brachiocephalic vein was present in one of them and left azygos vein ran upward along the left side of vertebral column and drained into PLSVC in the cadaver. There was no anastomosis between azygos and hemiazygos vein.

Conclusion: Persistent left superior vena cava (PLSVC) occurs as a result of failure of regression of left anterior cardinal and left common cardinal veins. The precise knowledge of double superior vena cava by surgeons, sonographers and interventional radiologists can prevent possible complications in routine clinical practice and during cardiopulmonary bypass.

Key words: Double superior vena cava, Persistent left superior vena cava, Left azygos vein, Anterior cardinal vein, Coronary sinus

INTRODUCTION:

The precise anatomical knowledge of vascular system anomalies is essential due to its significance in different developmental problems in adventant and injury in various invasive procedures. Normal anatomy describes the formation of a single superior venacava by the union of right and left brachiocephalic veins which drains the blood from upper part of the body. Superior vena cava anomalies are caused by variations in the development of the embryonic thoracic venous system. The anomalies that involve the superior vena cava include the persistent left superior vena cava with or without the right superior vena cava, absence of superior vena cava, hypoplasia of the superior vena cava, and abnormal opening of the superior vena cava into the left atrium, coronary sinus, azygous venous system, and the pulmonary vein.¹

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cardinal and left common cardinal veins. PLSVC may open into left atrium of the heart, coronary sinus, right superior vena cava is innominate vein. Persistent left superior vena cava is a rare venous malformation. There is limited number of cadaveric study on this anomaly.

The reason is that the double superior vena cava often associated with cardiovascular anomalies, allowing only the limited number of adult patients to survive. However, the developments of procedures for cardiovascular catheterization and clinical imaging examinations have increased the number of anomaly is being reported. It is estimated to occur in 0.3-0.5% of the general population and 3-10% of patients with other forms of congenital heart disease.3,4

Double superior vena cava is usually asymptomatic unless other cardiac anomalies exist. It is frequently associated with cardiac abnormalities like ventricular septal defect, atrial septal defect or endocardial cushion defect that have significant mortality and morbidity.5 PLSVC may also interfere and cause problems during various invasive procedures such as pacemaker implantation, central venous catheterisation, retrograde delivery of cardioplegia and retrograde left ventricular pacing.6,7,8

The aim of the present study is to find the frequency of double superior vena cava, its embryological basis and to correlate its clinical significance. The present study also compared the frequency of double superior vena cava reported in other similar studies.

MATERIALS AND METHODS:

This is a descriptive study which includes the cadavers allotted for first year MBBS students in Department of Anatomy at Velammal Medical College, Madurai during year 2013-2018. The mode of formation and drainage of persistent left superior vena cava along with other thoracic veins have been properly traced and studied in detail.

RESULTS

We found the presence of persistent left superior vena cava among 2 cadavers out of 60 cadavers (3.33%). Persistent left superior vena cava was observed in adult male cadaver aged 50 years and adult female cadaver aged 55 years.

Case 1- Adult male cadaver aged 50 years:

Persistent left superior vena cava was formed by the union of left internal jugular vein and left subclavian vein. Left brachiocephalic vein was absent. Persistent left superior vena cava coursed vertically down anterior to arch of aorta and left pulmonary trunk (Fig.1). When traced, it opened into an enlarged coronary sinus that further drained into the right atrium (Fig.2). There was no communication between the two vena cavae. The persistent left superior vena cava had the same length and caliber as compared to the right superior vena cava (Fig.1, Fig.2). A left azygos vein replaced the accessory azygos and the hemiazygos veins. It ran upward along the left side of vertebral column and arched over the root of the left lung to drain into PLSVC (Fig.3).

The azygos and the left azygos veins did not communicate with each other. Azygos system was made up of bilaterally symmetrical veins. Right superior vena cava was present as a continuation of right brachiocephalic vein which in turn formed by the union of right internal jugular vein and right subclavian vein. Right superior vena cava opened into right atrium and received the drainage of azygos vein (Fig.2). No other associated anomaly of heart was observed.
**Fig. 1:** Heart in situ showing double superior vena cavae without any communication between them. RSVC - right superior vena cava, PLSVC - persistent left superior vena cava, PT - pulmonary trunk.

**Fig. 2:** Posterior view of resected heart showing double azygos vein with double superior vena cavae. Right azygos draining into right superior vena cava and left azygos vein draining into persistent left superior vena cava. Persistent left superior vena opening into enlarged coronary sinus. RSVC - right superior vena cava, PLSVC - persistent left superior vena cava, RAV - right azygos vein, LAV - left azygos vein, CS - coronary sinus.

**Fig. 3:** Dissected thoracic cavity showing left azygos vein draining into persistent left superior vena cava. LAV - left azygos vein, PLSVC - persistent left superior vena cava.

**Fig. 4:** Posterior view of resected heart showing double superior vena cava with double azygos vein. Left brachiocephalic vein present which unite with right brachiocephalic vein to form right superior vena cava. Persistent left superior vena cava and left azygos vein are of reduced dimension as compared to right side. Coronary sinus are dilated into which persistent left superior vena cava drained. RSVC - right superior vena cava, PLSVC - persistent left superior vena cava, RBCV - right brachiocephalic vein, LBCV - left brachiocephalic vein, CS - coronary sinus.
present study. They classified the cases according to the presence / absence of pairing of azygous veins, as well as the presence / absence of anastomotic branch, into the following 4 types:

I) The pattern with an anastomotic branch between the right and left superior vena cava,
II) the pattern without any anastomotic branch,
III) the pattern showing the presence of the left superior vena cava alone and degeneration and disappearance of the right superior vena cava, and
IV) the pattern of the double superior vena cava with paired azygos veins. In the present study cases were of type IV.

During the fifth week of intrauterine life, in the human fetus, three pairs of major veins can be distinguished: the vitelline veins, carrying blood from the yolk sac to the sinus venosus; the umbilical veins, originating in the chorionic villi and carrying oxygenated blood to the embryo; and the cardinal veins, draining the body of the embryo.

The anterior and posterior cardinal veins join to form common cardinal veins and enter the right and left horns of the sinus venosus. Formation of the vena cava system is characterized by the appearance of anastomosis between the left and right sides in such a manner that the blood from the left side is directed to the right side. The left anterior cardinal vein above and the anastomosis between the anterior cardinal veins develops into the left brachiocephalic vein. Most of the blood from the left side of the head and the left upper extremity is thus directed to the right. The superior vena cava is
thus formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein. On the other hand, the left common cardinal vein and the distal part of the left horn become atretic and forms the ligament of Marshall or ligament of the left superior vena cava. The caudal part of anterior cardinal vein distal to the anastomosis obliterates. The failure of this normal regression i.e, persistence of left anterior cardinal vein and left common cardinal vein results in PLSVC.

During fifth to seventh week, a number of additional veins are formed: a) subcardinal vein, which mainly drain the kidneys b) sacrocardinal vein, which drain the lower extremity and c)supracardinal vein, which drain the body wall by way of the intercostal veins, taking over the functions of the posterior cardinal vein after its major obliteration. Azygos venous system develops from a pair of the right and left supracardinal veins. Both supracardinal veins communicate with the posterior cardinal veins to form the primordium of a pair of right and left azygos veins. Subsequently, right supracardinal together with a portion of posterior cardinal vein forms the azygos vein. On the other hand, left supracardinal vein forms hemiazygos vein which drains into azygos vein.13 The left azygos vein appears to be developed from the persistent proximal portion of the left posterior cardinal vein and the left supracardinal vein.12

There are differences in frequency of double superior vena cava showed by various studies. A persistent left superior vena cava is the most common anomaly of abnormal veins flowing into the heart, with a frequency of 0.3 - 0.5% in healthy individuals, and 2 - 4% in patients with congenital heart diseases.14,15,16 The frequency of double superior vena cava reported by various authors are as follows- 0.16% by Bergman RA, and 0.3% by M. Uemura.17 But AP. Gesase et al have shown highfrequency10 - 11% of the double superior vena cava but that was observed in congenital heart disease patients.18 The present study showed 2.22% of frequency of double superior vena cava.

PLSVC is asymptomatic and compatible with life unless the PLSVC opens into the left atrium or is accompanied by other congenital heart defects like atrial septal defect, ventricular septal defect and tetralogy of Fallot.19 Other congenital anomalies in the heart and an associated shunt between the right and the left cardiac system occurring should be considered from the presence of the persistent left superior vena cava.14 PLSVC may drain into left atrium in 7.5% cases which results in small right to left shunt. This has a minor haemo dynamic effect, mainly a variable degree of systemic cyanosis and may lead to clinical symptoms.20,21 PLSVC may also give rise to rhythm disturbances such as sinus node dysfunction and atrioventricular block. The arrhythmia problems may be related to the stretching of the conduction tissue caused by the enlargement of the coronary sinus.7,8 LSVC has been associated with an increased risk of arrhythmias, most commonly atrial fibrillation.22

The knowledge of existence of PLSVC during CT scan evaluation of the mediastinum is important because LSVC can be mistaken for a lymph node and result in false staging of lung cancer.23 Invasive procedures like cardiac pacemaker implantation, resynchronization therapy, radio frequency catheter ablation, internal jugular or subclavian vein catheter insertion require detailed echo graphic studies of this rare congenital anomaly to avoid further complication.6,7,20 Awareness of PLSVC is necessary for intensive care clinicians not to place catheters outside the venous circulation & to avoid complications like perforation, shock, cardiac arrest, cardiac tamponade and thrombosis.20
Central venous cannulation may result in unusual catheter positions and inadvertent coronary sinus cannulation may result in cardiac perforation. Cannulation of the heart having PLSVC for cardiopulmonary bypass may result in effective retrograde cardioplegia. Therefore during cardiac surgery, the presence of PLSVC with adequate development of left brachiocephalic vein is a relative contraindication to the administration of retrograde cardioplegia. It may be possible to clamp the PLSVC to avoid the cardioplegia solution from perfusing retrograde up the PLSVC and its branches. However, there is a possibility that there may be some steal of cardioplegia solution through an accessory vein. If the right superior vena cava is absent, all venous return from the upper body will drain through the PLSVC.

Hence, this may affect the use of the retrograde cardioplegia. In such a case, occlusion or ligation of the PLSVC would be fatal as this may result in cerebral congestion. During heart transplantation in a patient with PLSVC, the coronary sinus must be dissected carefully to permit re-anastomosis of PLSVC to right atrium. The dilated coronary venous sinus associated with PLSVC obscure contrast radiographic findings of the interatrial septum and the space between the ventricles on cardiac angiography.

Transthoracic echo cardio graphy including agitated saline infusion to both antecubital vein - CT or MRI are important non-invasive diagnostic tools for accurate diagnosis of this rare congenital venous malformation. However, the gold standard for the definitive diagnosis of double SVC is usually discovered with invasive angiography. The left SVC opening to the right atrium through the coronary sinus can be clearly shown by MPR and 3D VR obtained through MDCT. When the persistent left superior vena cava is ascertained by these diagnostic techniques, other associated congenital anomalies should be investigated so as to comprehend the risks related to catheterization and selection of a safe route should be considered.

**CONCLUSION:**

Although the double superior vena cava is a rare congenital anomaly, prior knowledge of it and the associated anomalies should be considered seriously by surgeons, sonographers and interventional radiologists. This can prevent possible complications in routine clinical practice and during cardiopulmonary bypass and minimize the diagnostic errors. As the literature available on this congenital anomaly is sparse, the present study can add to the existing knowledge of this congenital anomaly and stresses on its significant clinical implications, especially during central venous catheter placement or during surgical procedures in the chest.

**CONFLICTS OF INTEREST:**

The authors have none to declare.

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**REFERENCES:**


