



## “LEFT SUPERIOR VENA CAVA” EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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### ABSTRACT

The left superior vena cava (LSVC) is the most frequent abnormality in the general population and upto 12.9% of patients with the systemic venous return, with the frequency of 0.1%-0.5% in the congenital heart disease. The persistence of LSVC results from the failure of the involution of the left anterior cardinal vein. Right SVC and LSVC coexist together in 80%-90% of cases. LSVC usually drains into a delicate coronary sinus, but it can also drains into less frequently into the left atrium and thus responsible for the right to left shunt or even left to right shunt with important clinical implications.

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### Introduction

The first description of left superior vena cava (LSVC) was reported by LE CAT in 1738. LSVC is not a rare congenital abnormality of the systemic venous returns that can have various presentations and can be isolated or associated with congenital heart disease. The persistence of the LSVC results from the failure of involution of the left anterior cardinal vein (CV). In the past, this anomaly was usually detected incidentally during cardiac surgery or autopsy (CHA And KHOURY 1972). In many cases, LSVC has no consequence, but this variant is important to know in case of pacemaker insertion, central venous catheter positioning or cardiac surgery. LSVC is usually draining into the dilated coronary sinus (CS), but it can also drain less frequent into the left atrium and being responsible for the right to left shunt or even left to right shunt with important medical implications (Mustafa yurtdas, Musa sahin,). The right cardinal vein also forms a superior vena cava (SVC) on the right side and failure in the formation of left brachiocephalic vein (Thierry Couvreur MD and Beniot Ghaye MD). Left superior vena cava is a rare anomaly. They also have some clinical importance. Moreover, left superior vena cava is surgically important in the presence of congenital heart disease. They are due to the variations in the development of embryonic venous system. The superior vena cava duplication is a rare anomaly with a rate in the general population of 0.3%. The majority of the cases are asymptomatic and diagnosed incidentally by imaging alone for another lesson. Angiogram was done to show a good sized cephalic vein which is wide open and mild to moderate stenosis at subclavian area. When this condition exists, the caliber of right vessel is normal, but it may be smaller than unusual due to the low or reduced blood volume [1-3].

The most frequent double vena cava presents with a normal right superior vena cava anatomy and the left superior vena cava enters into the coronary sinus and subsequently draining

into the right atrium. Sometimes, there may be transverse anastomosis between the paired cavas [4]. In absence of these, the right and left half of head and the upper trunk drains back to the heart independently. The coronary sinus is large which approaches the caliber of the cava that enters it. When an anastomosis does exist between the two cavas, it may be oriented inferiorly towards the left or directed transversely. In some cases, additional cross communication exists in the form of irregular branching veins; small slender, plexiform arrangement represents the persistent vessels that normally form the left innominate vein [5].

### Incidence

The incidence of left superior vena cava in general population is 0.3-0.5% and ~5% in those with congenital heart disease, it is only seen in isolation in 10% of the cases since the vast majority is accompanied by a normal right superior vena cava which is termed as SVC duplication. Congenital heart disease is present in 4.4% of the patients with CHD.

### Ontogenesis of normal development of superior vena cava (Fig-2):

In the fifth week development of the venous system in the embryo, three pairs of major veins can be distinguished. They are: The vitalize veins, carries blood from yolk sac to the sinus venosus. The umbilical veins, which originates in the chorionic villi, carrying oxygenated blood to the embryo. The cardinal veins, drains the body of the embryo proper [8-19].

### Cardinal veins

The formation of vena cava system is characterized by the appearance of anastomoses between the left and right in such a manner that blood from the right side is channeled to the right side. The anastomosis between the cardinal vein leads to the development of the left brachiocephalic vein. The most blood the left side of the head and the left upper extremity is then channeled to the right side of the head. The terminal portion of the left posterior cardinal vein, that enters into the left brachiocephalic veins retained as a small vessel, the left

superior intercostal vein. This vessel receives blood from second and brachiocephalic veins retained as a small vessel, the left superior intercostal vein. This vessel receives blood from second and third intercostal spaces. The superior vena cava is formed by the right common cardinal vein and proximal portion of the right anterior cardinal vein. (Mary G.Cormier, Joseph.W Yedlicka)

The anastomosis between the sub cardinal veins forms the left renal vein. The left sub cardinal vein disappears and the distal portion remains as gonadal vein. Then the right sub cranial vein becomes the main drainage channel that develops into the renal development of inferior vena cava (Richard J.Gray, Rogelio Moncada). The anastomosis between the sarcocardinal vein forms left common iliac vein. When the renal segment of the inferior vena cava connects with the hepatic system, the inferior vena cava is complete [20] The fourth to eleventh intercostal veins empty into the right supracardinal vein from the azygous vein, with the portion of the posterior cardinal vein forms the azygous vein. On the left fourth and seventh intercostal veins enters into the left supracardinal vein, then it is known as the homozygous vein, that empties into the azygous vein [21].

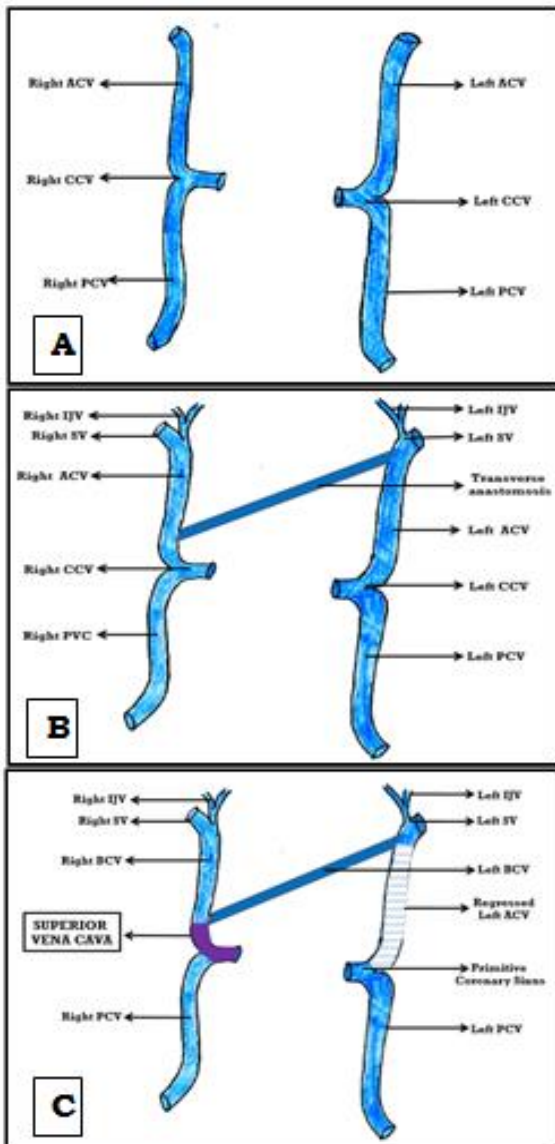


Fig 1: Schematic representation of normal development of Right superior Vena cava.

**Ontogenesis for the abnormal left superior vena cava**

During the development of right atrium, the sinu atrial differentiates the sinus venosus and atrial chamber. The right and the left cardiac veins drains the cardiac region into the respective ducts of cuvier ,that drains into the sinus venosus.it eventually absorbed into the structure of the right atrium with the bridging between the anterior cardinal veins which forms the left innominate vein. the right duct of cuvier and the right cardinal vein from the superior vena cava ,whereas the left cardinal vein typically obliterates. If the occlusion of the left cardinal vein fails to occur then that vessel persists as the "LEFT SVC" which drains into the CS through the vein of marshal. (R.B.Irwin, M.Greaves and M.Schmitt) (Fig-2).

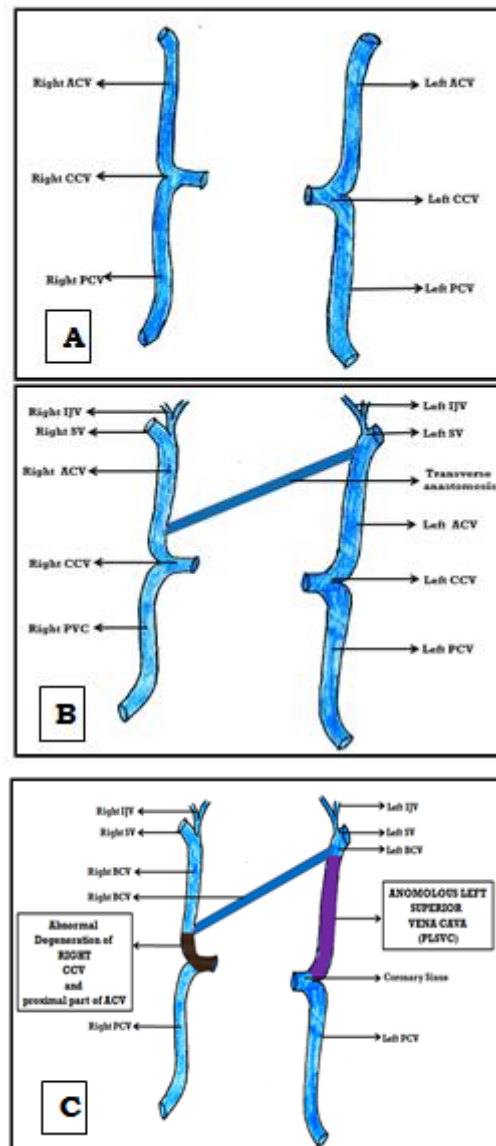


Fig 2: Schematic representation of abnormal development of Left superior Vena cava.

**Discussion**

Left superior vena cava is a congenital venous anomaly that occurs in 0.5% of general population, 0.3% of the healthy population and 4.3% of the patients with congenital heart diseases. In 82% of the patients with L SVC, the right superior vena cava is also present. Persistence of L SVC with the absent right superior vena cava usually occurs only 0.09-0.13% of the patients with congenital heart disease. [22]

This anomaly is frequently associated with situs inversus. Left SVC with other anomalies and structures in the left

atrioventricular groove may be overlooked if they are not carefully and approximately evolved. [23]

The persistent of the left cardinal vein results in the L SVC. This by itself does not produce any physiological derangements but it is associated with other congenital heart diseases such as septal defects, situs inversus, tetralogy of Fallot's etc...Left SVC drains into the right atrium via the coronary sinus venosus or coronary sinus and is asymptomatic [24]. They are considered to be anomaly of coronary sinus. 8% of L SVC drain into left atrium and causes right to left shunt and may present with unexplained cyanosis and clubbing. It is a problem to keep blood out of the field during cardio pulmonary by-pass and thus the left SVC may need to be ligated or separately canulated to avoid its venous return causing distension of right heart. Also it is a contra indication to retrograde cardio plegia. Other problems may include arrhythmia cardiac arrest and coronary sinus thrombosis [25]. It can cause difficult left sided central insertion of pulmonary artery catheter or pacing wire attributable to orientation. The clinical signs include Jugular venous distension on the left side, abnormal left Jugular venous wave form due to direct transmission of left atrial contraction, atrial pressure and non-atrial blood gas analysis.[26]

Conventional chest x-ray usually display widening of aortic shadow, paramedian bulge along left heart border and crescentic vascular shadow projecting from left upper border of aortic arch to the middle 3rd of the clavicle. [27]

Other diagnostic tools are multi slice CT scan or dual source CT-scan transthoracic-echo-cardiography and venography with fluoroscopy. Safest diagnostic option in transthoracic echocardiography with an agitated saline micro bubble contrast media which has the advantages of being non-invasive, without radiation effect and causing haemodynamic instability in a sick person compared with contrast imaging [28]. It also demonstrates draining of contrast into the coronary sinus. It is likely to be associated with heterotrophy discordance or malposition of thoraco abdominal organs and vessels, complex congenital heart anomalies and extra cardiac defects or defect involving mid-line structures.[29] It could be associated with interruption of inferior vena-cava and splenic anomalies. It can present like left sided Para-cardiac mass due to dilated coronary sinus. The diagnosis is usually confirmed on multi-slice row detected CT scans [30].

### Conclusion

Anomalies of superior vena cava are frequently found as incidental findings on cross sectional imaging and are occasionally associated with important clinical sequelae and echocardiography [31]. Cardiac imaging specialists should be aware of their imaging features and associations to differentiate the persistent left superior vena cava from the partial anomalous pulmonary venous drainage of the left upper lobe. If the latter is present and the right heart dilation or strain is evident. Additionally right sided venous drainage abnormalities should be actively searched for and excluded. A rare variant of L SVC with an intra-atrial course has been identified and that may become increasingly reported as a use of advanced cardiac imaging techniques becomes more widespread. (R.B.Irwin, M.Greaves and M.Schmitt) The majority of the cases are incidentally diagnosed on imaging for other reasons, which alerts the physician of other congenital abnormalities that need further work up [31]. However these venous abnormalities should be recognized, as they have significant clinical implications, especially during central venous catheter placement,

radiofrequency ablation, pacemaker insertion or coronary artery bypass graft.

However, continued documentation of such anomalies is clinically important, and it remains to be important in medical field [32].

### References

- [1] Ajit Kumar, E.Ganesh, T.Malarvani, Manish Kr. Singh. Bilateral supernumerary heads of biceps brachii. *Int J Anat Res.* 2014; 2(4):650-52.
- [2] Ajit Kumar, Ganesh Elumalai, Malarvani Thangamani, Nirmala Palayathan, Manish Kr Singh. A Rare Variation in Facial Artery and Its Implications in Facial Surgery: Case Report. *Journal of Surgery.*2014; 2(5): 68-71.
- [3] Arat N, Sokmen Y, Golbasi Z. Persistent left SVC with absent Right SVC & coronary sinus dilatation mimicking a paracardiac mass. *Image in cardiovascular medicine* 2007; 34: 492-493.
- [4] Soward A, Ten cate F, Fioretti P, Roeelands J, Serruys PW. An Persistent left SVC draining into left atrium. *Cardiology* 1986; 73: 368-371.
- [5] Lonquist PA, Olsson GL. Persistent left SVC – an unusual location of central venous catheter in children. *Intensive care Med.* 1997; 17: 497500.
- [6] Azocar RJ, Narang P, Talmor D, Klisbon S, Kayaar AM. Persistent Left SVC identified after cannulation of right subclavian vein. *Anaesthes Analog* 2002; 95:305-307.
- [7] Brzezinski M, Keller R, Grichnik KP, Swaminathan M. Persistent Left SVC in a patient with a history of TOF. *Anaesthesia Aalog* 2005; 100: 1269-1270.
- [8] Higgs AG, Paris s, Potter F. Discovery of left sided SVC during central venous catheterization. *Brit J anesthesia* 1998; 81: 260-261.
- [9] Ganesh Elumalai, Sushma Chodisetty. Anomalous “Mutilated Common Trunk” Aortic Arch Embryological Basis and its Clinical Significance. *Texila International Journal of Basic Medical Science.* 2016; 1(1): 1-9.
- [10] Ganesh Elumalai, Emad Abdulrahim Ezzeddin. “The sudden soul reaper” - hypertrophic cardiomyopathy – its embryological basis. *Elixir Embryology.* 2016; 99: 43284-43288.
- [11] Ganesh Elumalai, Muziwandile Bayede Mdletshe. “Arteria lusoria”- aberrant right subclavian artery embryological basis and its clinical significance. *Elixir Embryology.* 2016; 99: 43289-43292.
- [12] Ganesh Elumalai, Sushma Chodisetty, Pavan Kumar D.2016. Ganesh Elumalai et al Classification of Type - I and Type - II “Branching Patterns of the Left Arch Aorta”. *Imperial Journal of Interdisciplinary Research.* 2(9): 161-181.
- [13] Ganesh E, Sushma C. The deer horn aortic arches” embryological basis and surgical implications. *Anatomy Journal of Africa.*2016; 5(2): 746 – 759.
- [14] Ganesh Elumalai, Sushma Chodisetty. Teratological Effects of High Dose Progesterone on Neural Tube Development in Chick Embryos. *Elixir Gynaecology.* 2016; 97: 42085-42089.
- [15] Ganesh Elumalai, Sushma Chodisetty. “The True Silent Killers” - Bovine and Truncus Bicaroticus Aortic Arches its Embryological Basis and Surgical Implications. *Elixir Physio. & Anatomy.* 2016; 97: 42246-42252.
- Ganesh Elumalai, Sushma Chodisetty, Bridget Omo Usen and Rozminabanu Daud Patel. “Patent Ductus Caroticus” - Embryological Basis and its Clinical significance. *Elixir Physio. & Anatomy.* 2016; 98: 42439-42442.
- [16]

- [17] Ganesh Elumalai, Sushma Chodisetty, Eliza Arineta Oudith and Rozminabanu Daud Patel. Common anomalies origin of left vertebral artery and its embryological basis. *Elixir Embryology*. 2016; 99: 43225-43229.
- [18] Ganesh Elumalai, Sushma Chodisetty, Sanjoy Sanyal. Common Nasal Anomalies and Its Implications on Intubation in Head and Neck Surgeries. *Journal of Surgery*. 2016; 4 (4): 81-84.
- [19] Ganesh Elumalai, Malarvani Thangamani, Sanjoy Sanyal, Palani Kanagarajan. Deficient sacral hiatus cause mechanical low back pain: a radiological study. *Int J Anat Res*. 2016; 4(1):1758-64.
- [20] Moonley IP, Synder CL, Holder TM. An absent right & persistent left SVC in an infant requiring extracorporeal membrane oxygenation therapy: *J Paed surg*1993; 28: 1733-1734.
- [21] Ghadiali N, Teo LM, Sheath K. Bed side confirmation of a persistent left SVC based on aberrantly positioned central venous catheter on chest radiograph. *Brit J Anaesthesia*. *Brit J anaesthesia* 2005; 96: 53-56.
- [22] Alharthi M, Mookadam F, Collins J, Chandrasekharan K, Scott L, Tajik J. Extracardiac venous heterotaxy syndrome. Complete noninvasive diagnosis by multimodality imaging *Circulation* 2008; 117: 498-503.
- [23] Dwight T. Absence of the inferior vena cava below the diaphragm. *Anat Physiol*. 1901;35:7-20.
- [24] Rose V; Izukawa T, Moses C. Syndromes of asplenia & Poly splenia a review of cardiac & noncardiac malformation in 60 cases with special reference to diagnosis and prognosis. *Brit Heart J* 1975; 37: 840-852.
- [25] Abadir S, Bouzguenda I, Boudjemline Y et al. (2007) Percutaneous occlusion of a left superior vena cava draining into the left atrium: two case reports. *Arch Mal Coeur Vaiss* 100:470-473
- [26] Ancel P, Villemain F (1908) Sur la persistance de la veine cave supérieure gauche chez l'homme. *J de l'anat* 44:46-62
- [27] Bjerregaard P, Laursen HB (1980) Persistent left superior vena cava. Incidence, associated congenital heart defects and frontal plane P-wave axis in a paediatric population with congenital heart disease. *Acta Paediatr Scand* 69:105-108.
- [28] Blank E, Zuberbuhler JR (1968) Left to right shunt through a left atrial left superior vena cava. *Am J Roentgenol Radium Ther Nucl Med* 103:87-92
- [29] Bonnet D (2003) Anomalies du retour veineux systémique. *Encycl Méd Chir (Elsevier, Paris), Radiodiagnostic- Coeur-Poumon* 32-016-A-15:1-4
- [30] Bousuges A, Ambrosi P, Gainnier M et al. (1997) Left-sided superior vena cava: diagnosis by magnetic resonance imaging. *Int Care Med* 23:702-703.
- [31] Brickner ME, Eichhorn EJ, Netto D et al. (1990) Left-sided inferior vena cava draining into the coronary sinus via persistent left superior vena cava: case report and review of the literature. *Cathet Cardiovasc Diagn* 20:189-192.
- [32] Buirski G, Jordan SC, Joffe HS et al. (1986) Superior vena caval abnormalities: their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. *Clin Radiol* 37:131-138.
- [34] Bunker PC, Neufeld DA, Moore JC et al. (1981) Persistent left superior vena cava and associated structural and functional considerations. *Angiology* 32:601-608.