Case Report

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Persistent Left Superior Vena Cava

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Abstract

Persistent left superior vena cava can be incidental, accidental, descriptive and investigative when the left anterior cardinal veins not obliterated. Dilated coronary sinus in the absence elevated right sided filling presence, coronary sinus opacification after right arm anticubital vein injection are the trinity for left sided superior vena cava. A case of persistent left superior vena cava in a 10 year girl with a normal right superior vena cava and a persistent bridging vein brachiocephalic vein leads to formation of superior vena cava duplication.

Keywords: Coronary Sinus; Echocardiography; Computed Tompgraphy; Vena Cava.

Introduction

Haemodynamically insignificant persistent left superior vena cava has 923 citation in the literature. It has an incidence of 0.3% to 0.5% of general population representing one in every 200 to 325 people and a prevalence of 6.1%. 4.3-11% of patients with congenital heart diseases. 15% patients catheterized for congenital heart disease have a PLSVC. PLSVC when present travels lateral to the aortic arch at an orthogonal angle, then travels inferiorly between the left atrial appendage and the left upper pulmonary vein where the ligament of Marshall (fibrosed Left SVC) should be. PLSVC has two types of connection to the heart. Either it drains into the right atrium via coronary sinus (90%), or it drains into the left atrium creating a right to left shunt of small magnitude(10%).

PLSVC is a venous anomaly of the thorax. It was first described by LECHAT in 1787. It may have variable communications to right superior vena cava or no communication with the RSVC i.e the innominate vein may be absent. The normal RSVC may be absent in 0.09 – 0.13% cases.

It is often suspected when, on echo there is a dilated coronary sinus with rapid blood flow, difficulty in reaching convenient pacing site when pacing leads are inserted from left side and rhythm disturbance due to SA node dysfunction or AV block. Bartram et al noticed 46% of congenital heart anomalies like ASD (16%), endocardial cushion defect (11%) and Tetralogy of Fallot (9%) associated with this defect. In 80-90% of cases a coexistent right superior vena cava is present which is usually smaller than normal. In less than 10% of cases persistent LSVC with unroofed coronary sinus drains into the left atrium or in a pulmonary vein leading to predisposition of cyanosis, paradoxical embolism and brain abscess. Higher incidence of conduction anomaly can occur with this malformation along with other congenital anomalies. PLSVC, with an aberrant right superior vena cava or absent / small left brachiocephalic vein (65%) and absent superior vena cava (10%) is frequently diagnosed incidentally during pacemaker implantation or central venous catheter insertion. PLSVC with absent RSVC interfere with the use of retrograde cardioplegia. Cannulation in a case of PLSVC can create disadvantages during an atrial septal defect repair [1-4].

Case Report

A 10 year old female presented with progressive shortness of breath and palpitation. She denied any syncopal attacks, chest pain or limb swelling. On examination a widely split second heart sound and a systolic murmur was noted. Her electrocardiogram was normal. Echocardiogram revealed a dilated coronary sinus of 25mm. Agitated saline injection into left antecubital vein showed dilated coronary sinus filled first then emptied into the right atrium. Cardiac computed tomography was performed and revealed a persistent Left SVC draining into the dilated coronary sinus. There was a small connecting vein between the right and left superior vena cavae. (The arterial saturation of the patient was 95% on room air. The anomalous vena cava was seen descending vertically along the left superior mediastinum and continued caudally posterior to the left atrial appendage and left atrium and anterior to left superior pulmonary vein and then entered the coronary sinus (vide images). Anterior posterior and oblique three dimension volume rendered images from a contrast enhanced computed tomographic scan demonstrated left SVC draining into the coronary sinus.

Discussion

X-Ray of the chest with a prior left sided central venous catheter placement is symbolic of persistent left SVC. It shows the catheter course from left subclavian/jugular vein to PLSVC then on left of Aortic arch to area of coronary sinus. A blood gas study compatible with venous/arterial blood may be useful to differentiate whether the catheter tip is in Coronary sinus/RA or into the LA, respectively. Apart from that obese patient, short neck, vasculopathy can be suggestive of PLSVC.

Associated finding with persistent LSVC are genetic (VACTERL, CHARGE and OPITZ G/BBB Syndrome), chromosomal (Trisomi 21, TURNER and microdiletion 22 q11.2), increased nucheal translucency, oesophageal atresia, cardiac heterotaxy, Tetralogy of Fallot, coarctation of aorta,

Modified, Adjusted Diagram of the Left superior Vena Cava

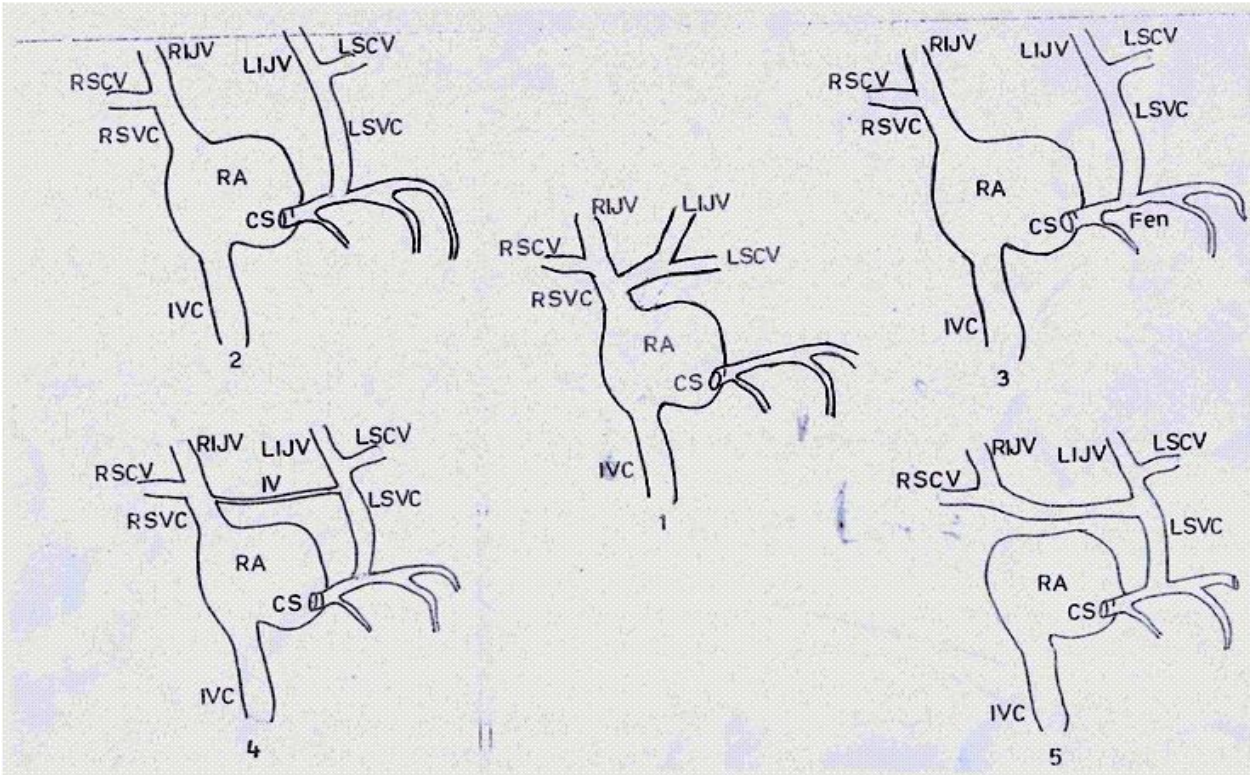

Fig. 1: Normal Figure: Coronary Sinus, Draining into Right Atrium, No Left Superior Vena Cava. **Fig. 2:** Persistent Left SVC draining through dilated Coronary Sinus into Right Atrium. **Fig. 3:** Persistent Left SVC draining through Fenestrated Coronary Sinus, No Left Brachiochephalic vein. **Fig. 4:** Persistent Left SVC connected to Right Superior Vena Cava by Innominate Vein and also to Coronary Sinus. **Fig. 5:** Persistent Left SVC draining into Right Atrium through Coronary Sinus. No Right Superior Vena Cava. Right Innominate vein joins to Left SVC through Left Innominate Vein. This is isolated Left Superior Vena Cava

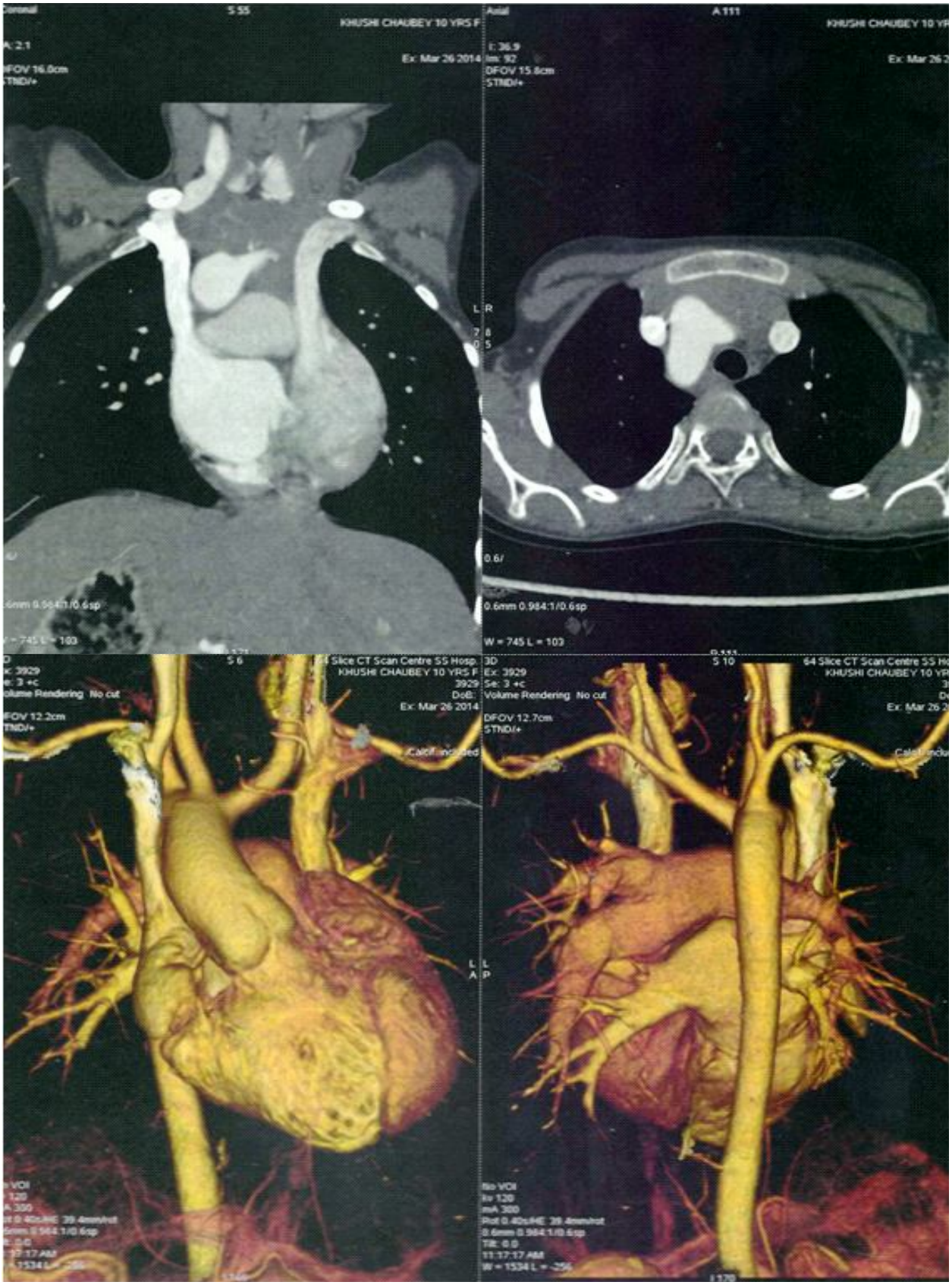

Fig. 1 A,B,C,D: Contrast enhanced CT images coronal (A), Axial (B) showing right and left superior vena cava but no innominate vein (C, D). Left superior vena cava is draining into coronary sinus

VSD, double aortic arch [5,8], bicuspid aortic valve and cortriatrium. Dextrocardia with tga, bilateral SVC and left atrial isomerism [11].

Persistent Left SVC is diagnosed indirectly by a recognition of a dilated coronary sinus in parasternal long axis view during echocardiography. It appears like a circular structure in the atrioventricular groove, located anterior to the posterior pericardium. In four chamber view with posterior angulation coronary sinus can be viewed in long axis passing behind the left atrium towards the right side. Contrast echocardiography from the left antecubital vein shows coronary sinus opacification with contrast and dilated coronary sinus in absence of elevated right atrial filling pressure, and, lastly normal sequence of opacification after right arm antecubital vein injection. In PLSVC the coronary sinus has oval shape with eccentricity index < 0.8 but symmetric distension of the vessel suggests elevated right atrial pressure. The echocardiographic findings [5] in left SVC are

1. Dilated coronary sinus in the absence of the elevated right side filling pressures.
2. Normal sequence of opacification after right arm antecubital vein injection of agitated saline.
3. Coronary sinus opacification by bubble study before the right atrium when injected in the left arm.
4. Presence of bilateral superior vena cava.
5. In 10% cases Right superior vena cava is smaller or absent.

Course and Tributaries of PLSVC

A. PLSVC Draining in Left Atrium (Unroofed Coronary Sinus)

1. Starts at junction of left subclavian and left internal jugular vein
2. Passes lateral to the aortic arch
3. Receives left superior intercostal vein
4. Courses anterior to the left hilum of lung.
5. Joined by hemiazygos vein
6. Passes between the left superior pulmonary vein posteriorly and the left atrial appendage anteriorly to become coronary sinus.
7. Opens into LA or some pulmonary vein.

B. PLSVC Draining into Right Atrium

1. Starts at junction of the left subclavian vein and left internal jugular.
2. Passes lateral to aortic arch
3. Receives the left superior intercostal vein
4. Courses anterior to the left hilum of lung.
5. Joined by hemiazygos vein
6. Crosses posterior wall of the left atrium and midline obliquely.
7. Receives great cardiac vein to become coronary sinus (usual).

Contrast enhanced computed tomography is the most easy and accurate imaging modality to diagnose and confirm the exact anatomy of PLSVC. It not only demonstrates the PLSVC, gives its exact course, size, drainage site but shows the anatomy, size and tributaries of RSVC. Other imaging modalities are contrast venography, cardiovascular magnetic resonance and foetal echocardiography showing coronary sinus enlargement and abnormal three vessel view. A venogram via the right internal jugular vein or the subclavian vein can show the absence of right superior vena cava and the brachiocephalic vein joining with the PLSVC that drained into the right atrium through the dilated coronary sinus [1,2]. Clinical problems associated with persistent Left SVC are rhythm disturbances, inability to insert central venous catheters, inability to insert transvenous pacing leads through the left internal jugular or left subclavian veins, and delivery of retrograde cardioplegia. The existence of acute angle between the coronary sinus and the tricuspid valve in persistent LSVC calls for long (85 cm) active fixation with wide loop technique during pacemaker implantation. Pacing through the tributaries of the coronary sinus in a patient with LSVC has been reported. Failure to drain the left superior vena cava during cardiopulmonary bypass could result in inadequate venous return to the pump, excessive blood return to the operative filed, unnecessary rewarning of the heart and residual intracardiac shunt due to poor visibility. A PLSVC needs drainage by a separate cannula. Either a tricaval cannulation is done or usual bicaval cannulation and a third cannula or suction for coronary sinus is used in right sided open heart surgical procedures. Delivery of retrograde cardioplegia is not feasible because of inability to obtain tight seal by the balloon of cardioplegia catheter, inadequate cardioplegia solution delivery to the myocardium and lastly fear of cerebral congestion. During PA catheterization a lack of progression of normal right heart waveform occurred in presence of high cardiac output when a pulmonary artery catheter (PAC) was incorrectly placed into the coronary sinus, is reported.

The embryological development of systemic and pulmonary veins is complex and subject to considerable variation. During normal development, the anterior cardinal veins, which drain the head, neck and arms unite with the posterior cardinal vein in the very early embryonic stage and enter the heart as the right and left horns of sinus venosus. With the exception that the cardinal veins of both i.e., right and left side drain into the right atrium, the cardinal venous system is bilaterally symmetrical at this stage. Most of the left sided cardinal system disappears leaving only the coronary sinus and a remnant of obliterated LSVC known as the ligament of Marshall. Simple failure of obliteration of the left anterior cardinal vein results in the persistence of LSVC[10]. Other variations are absence of RSVC (0.36%), drainage into LA with variable communications, creating a right to left shunt, absence of bridging innominate vein between PLSVC and Right SVC. The genetic culprit may be genes for left to right signalling [5-7].

When the right ventricle appears dilated and volume loaded with an apparently intact atrial septum on a conventional transthoracic echo view, it may be needed to look for a sinus venosus atrial septal defect. Diagnosis of PLSVC is usually incidental during retrograde cardiophegia in cardiac surgery, left subclavian vein cannulation for monitoring or therapeutic purposes, device implantation or cardiovascular imaging but transthoracic contrast echocardiography is the method of choice [9].

Abbreviations and ACRONYMS

RA	:	Right Atrium
LA	:	Left Atrium
CS	:	Coronary Sinus
LSVC	:	Left Superior Vena Cava
RSVC	:	Right Superior Vena Cava
PLSVC	:	Persistent Left Superior Vena Cava
IVC	:	Inferior Vena Cava
RIJV	:	Right Internal Jugular Vein
LIJV	:	Left Internal Jugular Vein
IV	:	Innominate Vein (Brachiocephalic Vein)
LSCV	:	Left Subclavian Vein
RSCV	:	Right Subclavian Vein
MDCT	:	Multi Detector CT Scan
TGA	:	Transposition of Great Arteries
VACTERL	:	Syndromic Anomalies of **V**ertebrae, **A**nus, **C**ardiac, **T**rachea, **E**sophagus, **R**enals - Radial, and **L**imb
CHARGE	:	**C**oloboma of eye, **H**eart defects, **A**tresia of nasal Choanae, **R**etardation of growth, **G**enital and urinary abnormalities, **E**ar abnormalities.

Conclusion

Persistent superior vena cava connection to the coronary sinus is often incidental but an important finding which helps in planning safe invasive procedures [11,12].

Recognition of persistent left superior vena cava with embryological variants is essential for physicians. These variations can be detected by a radiological picture of the heart showing straight borders on both sides and a broad pedicle. MDCT cardiac angiography is very useful to confirm the presence or absence of PLSVC. It shows the clear anatomy and sizes of both SVCs and also shows the bridging innominate vein. On transthoracic Echocardiography with Doppler, the large size of the coronary sinus with its ostium and unusually excessive venous drainage through it indicates the presence of a persistent left superior vena cava, draining into the coronary sinus. In the ICU setting when central line is in place through the left sided veins a skiagram of the chest along with a blood gas study compatible with venous blood may be enough to make the diagnosis of persistent left superior vena cava; others are Transesophageal echocardiography, MRI. Preoperative knowledge about PLSVC and other anatomical variants are necessary for the surgeon for planning of surgery for congenital cardiac lesions.

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