“PERSISTANT LEFT SUPERIOR VENA CAVA”
EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE
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ABSTRACT
Persistent left superior vena cava is rare but an important congenital vascular anomaly. When superior cardinal vein caudal to innominate vein fails to regress PLSVC occurs. It is observed in isolation but associated with other cardiovascular abnormalities including atrial septal defect, bicuspid aortic valve, and coarctation of aorta, coronary sinus ostial atresia, and corteatrium. The PLSVC can render access to right slide of heart challenging via the left subclavian approach, which is utilized using pacemaker and Swiz-Ganz catheters. Incidental notation of dilated coronary sinus on echocardiography raises the suspicion of PLSVC. The diagnosis is confirmed by saline contrast echocardiography.

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Introduction
Persistent left superior vena cava (PLSVC) is an infrequent vascular anomaly, However it is the most conjoint congenital anomaly of thoracic venous system. It results when the left superior cardinal vein caudal to innominate vein fails to relapse [1, 2]. PLSVC is detected when cardiovascular imaging is, performed for dissimilar reasons. The most common subtype of PLSVC results in presence of both left and right SVC [3]. A bridging innominate vein may or may not be present. PLSVC is associated with absence of innominate vein in 65% cases. Most rarely the caudal right superior cardinal vein regresses, leading to an absent right SVC with PLSVC. In this case the left SVC revenues. The superior vena cava helps to return the deoxygenated blood from upper half of the body systemic circulation to the right atrium of heart. The SVC is formed by left and right brachiocephalic vein and the azygos vein. It is located in the anterior right superior mediastinum. Persistent left superior vena cava all the blood from cranial aspect of body. Through the coronary sinus the PLSVC drains into right atrium and is of no hemodynamic consequence. In the remaining case, it may drain into left atrium resulting in right to left shunt [1, 4].

The left sided superior vena cava is the most common congenital venous anomalies in the chest. They are asymptomatic and the presence of the vessel is identified by CT scan. Left sided SVC can result in right to left shunt in minority of the cases [5, 6]. In right to left shunt, because of the direct drainage in the left atrium (8%) doesn’t cause cyanosis, since it only gutters the left upper limb and left side of head and neck. Diagnosis of PLSVC is done by unusual course of catheter on chest X-Ray. The PLSVC can resent technical difficulties during intravascular procedures such as Swan- Ganz catheterization and insertion of pacing system or during cardiac surgery [3, 4].

Incidence
PLSVC is the most congenital thoracic venous anomaly with an incidence of 0.3% - 0.5% in general population and 5% of those with congenital heart disease [2].

Ontogenesis for normal development of superior vena cava
The development of superior vena cava if similar to the development of inferior vena cava, but is formed somewhat later. The left and right brachioccephalic veins forms the structure of superior vena cava which is also referred as innominate vein., receives blood from the eyes, upper limbs, neck, and behind the lower border of the first right costal cartilage[6,7]. In 5th week fetus, the common cardinal veins, umbilical vein, and the vitelline vein drain into sinus venosus. The major venous drainage for the embryo is common cardinal vein whereas the anterior cardinal veins carrying blood from cephalic part of the embryo and the posterior cardinal vein from caudal part to embryo [7, 8]. The yolk sac and the sinus venosus are connected by vitelline vein. Umbilical veins are the source for carrying oxygenated blood from the placenta for the fetus. In 8th week a large anastomosis (derived from thymic and thyroid veins) channels the blood from the superior (left anterior) cardinal vein towards the right, which gives rise to the future left brachiocephalic venous trunk. The anterior cardinal vein after anastomosis becomes internal jugular veins [8]. The anterior veins of the mandible give rise to external jugular vein. At the upper limb venous plexus fuses to form the subclavian vein. The latter originally opens at the posterior cardinal vein but the heart shifts somewhat caudally in its development, and the subclavian vein finally shifts to open into the anterior caval vein. Below the anastomosis, the left anterior cardinal vein loses its connection with the left common cardinal vein [8, 9]. The part that persists is a short segment which forms the left superior intercostal vein.
The coronary sinus venosus is formed by the left common cardinal vein which persists as a very short segment [9, 10]. The right common cardinal vein and the proximal part of the right anterior cardinal vein finally form the superior vena cava. Malfunctions are rare whereas the abnormal pulmonary venous return drains at either the superior vena cava or the right atrium. The left SVC is formed by the left anterior cardinal vein and common cardinal vein whereas the right anterior cardinal vein and the common cardinal vein forms the SVC [10]. At the 8th week of embryo the formation of the ligament of Marshall Process gets completed by the degeneration of the left superior vena cava. The failure of the degeneration of left posterior cardinal vein results in insistent left superior vena cava [11, 12,13].
Both the pairs of veins form the left and right common
cardinal veins before entering the embryological heart. The
left common cardinal vein continues to form coronary sinus
and oblique vein of the left atrium. An anastomosis is formed
between right and left cardinal veins resulting in the
innominate (brachiocephalic) vein, during the 8th week of
gestation [25,26]. The internal jugular vein is formed by the
cephalic portion of superior cardinal veins. The normal right
sided superior vena cava is formed by the caudal portion of
right superior veins whereas the portion of left superior
cardinal vein innominate vein normally regresses to become
“ligament of Marshall” .If this normal regression fails to
occur, a persistent left sided vascular structure that drains into
the coronary sinus, results in PLSVC [27, 28]. The innominate
vein may or may not degenerate in these cases leading to
disparities in anatomy. The most conjoint subtype of PLSVC
results in the presence in presence of both left and right
superior vena cava. A bridging innominate vein may or may
not be present. PLSVC is associated with absence of
innominate vein 65% cases [29, 30]. The regression of
caudal right superior cardinal vein leads to the absence of right
SVC with PLSVC. In this case, the left SVC returns blood from
cardinal aspect of the body. Variation is also mentioned in the
insertion of left SVC [31, 32, 33]. In most of the individuals
(80- 90%) the persistent left superior vena cava gutters into
the right atrium by the coronary sinus and is of no
hemodynamic consequence. In the remaining cases it may
drain in left atrium resulting in right to left sided shunt [34,
35]. Diagnosis is usually made by incident finding during
cardiovascular imaging or surgery. In the case of PLSVC an
unusual course of catheter on chest X-ray (Swan-Ganz
catheter via left subclavian) is done. The diluted coronary
sinus is revealed by transthoracic echocardiography and the
diagnosis is confirmed by the use of saline constant
echocardiography [36,37]. The diluted coronary sinus causes
PLSVC and it is also caused by other etiologies including
erected right atrial pressure, coronary arterio-venous fistula,
partial anomalous pulmonary venous return, or an “unroofed”
coronary sinus giving shunt flow between the left atrium and
coronary sinus. The following diagnostic criteria can be used
along with the echocardiography: 1) In the absence of evident
of elevated right sided pressures the diluted coronary sinus is
present on the two-dimensional echocardiography; 2)
transesophageal echocardiography and radionuclide
angiography have also been used for diagnosis. Patient with
PLSVC can have cardiac anomalies such atrial septal defect,
bicuspid aortic valve, coarctation of aorta, coronary sinus ostial
atresia, and core tritriatum [38,39]. The presence of associated
anomalies is more common with concomitant absence of right
SVC. The PLSVC is associated with anatomical and
architectural abnormalities of the sinus node and conduction
tissues. Both sinus and AV node can have persistent fetal
dispersion in central fibrous body in subjects with PLSVC
[39]. Left subclavian is used for access to right side of the
heart or pulmonary vasculature. As Swan-Ganz catheter
placement is performed without imaging it remains
challenging in many circumstances. When permanent
pacemaker or catheter has been inserted via PLSVC it leads to
serious complications such as arrhythmia, cardiogenic shock,
cardiac tamponade, and coronary sinus thrombosis.

Now a day’s improvement in catheter type and techniques
permitted successful placement of right atria and right
ventricular leads for dual-chamber pacing [39,40]. In addition
to this cardiac resynchronization therapy for advanced chronic
heart failure requires the placement of third pacing lead in left
poster lateral vein of the heart. In PLSVC several operations
have successfully placed a cardiac resynchronization system
and a lead via coronary sinus [40,41]. During cardiac surgery,
the presence of PLSVC is a relative contraindication to the
administration of retrograde cardioplegia. It is possible to
clamp the PLSVC to prevent the cardioplegia solution from
perfusing ebbing up the PLSVC and its subdivisions with
inadequate myocardial protection. However, there is a
probability that there may be some steal of cardioplegia
solution through an accessory vein. The coronary sinus must
be dismembered carefully to permit reanastomosis of PLSVC
to right atrium during transplantation in a patient with PLSVC
[41].

Conclusion

There is a possibility of PLSVC when dilated coronary
sinus is present. The diagnosis is confirmed by saline contrast
echocardiography. Whenever a catheter or guide wire is
inserted via left subclavian vein takes an unusual left sided
downward course, the critical care physicians or cardiologist
should consider the presence the PLSVC. A PLSVC certainly
presents technical difficulties with right heart entree via the
left subclavian, but does not impede insertion of catheters;
whereas the additional accompanying risks should be
discussed with the patient if the diagnosis of PLSVC is
already well-known.

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