Available online at www.elixirpublishers.com (Elixir International Journal)

Embryology

Elixir Embryology 100 (2016) 43454-43457

ISSN: 2229-712X

"PERSISTANT LEFT SUPERIOR VENA CAVA" EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

Ganesh Elumalai and Mouna Arumugam

Department of Embryology, College of Medicine, Texila American University, South America.

ARTICLE INFO

Article history:

Received: 29 September 2016;

Received in revised form:

02 November 2016;

Accepted: 07 November 2016;

Keywords

Right superior venacava,
Left superior vena cava,
Right anterior cardinal vein,
Left anterior cardinal vein,
Right superior cardinal vein,
Left superior cardinal vein.

ABSTRACT

Persistent left superior vena cava is rare but an important congenital vascular anomaly. When superior cardinal vein caudal to innominate vein fails to regresses PLSVC occurs. It is observed in isolation but associated with other cardiovascular abnormalities including atrial septal defect, biscuspid aortic valve, and coarctation of aorta, coronary sinus ostial atresia, and cortriatratium. 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.

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 sided 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 brachiocephalic 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 5^{th} week fetus, the common cardinal veins, umblical vein, and the vitelline vein drain into sinous 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. Umblical veins are the source for carrying oxygenated blood from the placenta for the fetus. In 8^{th} week a large anastomosis (derived from thymic and thyroid veins) channels the blood from the superior (left anterior) cardial 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 caudal 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.

Tele: +91-7708905280
E-mail address: mouna51297@gmail.com

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].

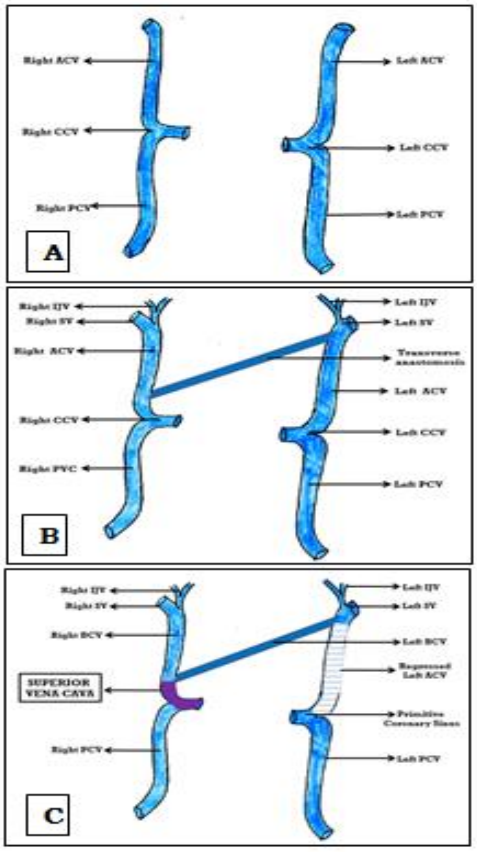

Fig 1.Schematic representation shows [A] The anterior cardinal vein(ACV) and posterior cardinal vein(PCV) opens into common cardinal vein(CCV) [B] Transverse anastomosis develops between right and left anterior cardinal vein(ACV) and internal jugular vein(IJV), subclavian vein(SV) appears and develops at either side of right and left anterior cardinal vein. [C] Transverse anastomosis develops as left brachiocephalic vein (BCV) and the left anterior cardinal vein regress (ACV) and superior vena cava (SVC) is developed between the left brachiocephalic vein and common cardinal vein (CCV).

Ontogenesis for the Persistent of left superior vena cava

A left sided SVC forms when the left anterior cardinal vein is not shattered during normal fetal development. The persistent lift sided superior vena cava passes lateral to the aortic arch before rejoining the circulatory system and anterior to the left hilum [14,15,16]. Right sided SVC and LSVC co-exist in 80% - 90% of the case.

LSVC drain into dilated coronary sinus, but it can also drain less frequently into left atrium [17,18]. The cardinal veins on the right and the left side drain into right atrium where the cardinal venous system is bilateral at this stage [19,20].

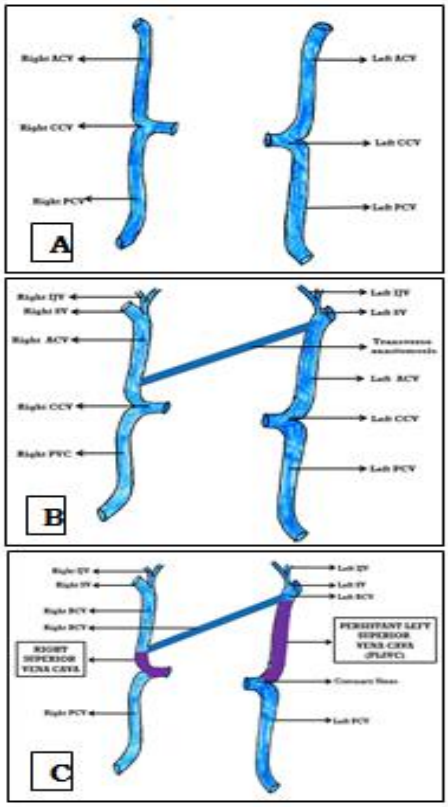

Fig 2. Schematic representation shows the anterior cardinal vein and the transverse anastomosis develops as right brachiocephalic vein (BCV). The left anterior cardinal vein (ACV) persists as left superior vena cava (SVC). The left superior vena cava opens into coronary sinus in the case of persistent left superior vena cava.

Coronary sinus is absent and persistent LSVC drains directly into the atrium. Individuals with PLSVC possess a normal SVC whereas the congenital anomalies of superior vena cava are discovered during central catheter insertion, pacemaker electrode placement, and cardiopulmonary bypass surgery[21,22].

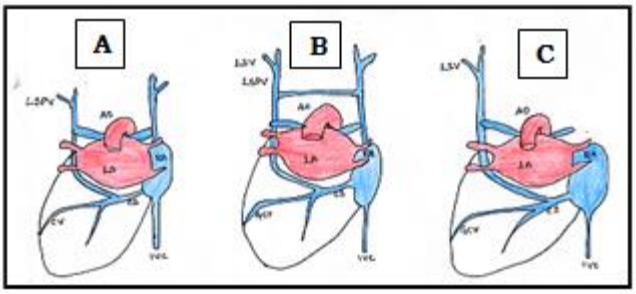

Fig-3: Schematic representation shows [A] An abnormal double Superior vena cava with single transverse anastomosis between right and left anterior cardinal veins (SV) below the descending aorta. [B] Abnormal double Superior vena cava with double transverse anastomosis between right and left anterior cardinal veins, one above and the other below the arch of aorta. [C] Abnormal double Superior vena cava with single transverse anastomosis, which drains into the coronary sinus.

Discussion

Persistent left superior vena cava is the common congenital thoracic venous anomaly. The thoracic embryonic venous system is of two large veins (the superior cardinal veins) which return blood from cardinal aspect of embryo and the other one is_inferior cardinal vein which returns blood from caudal aspect[23,24].

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 dilated coronary sinus is revealed by transthoracic echocardiography and the diagnosis is confirmed by the the use of saline constant echocardiography [36,37]. The dilated coronary sinus causes PLSVC and it is also caused by other etiologies including elevated 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 dilated coronary sinus is present on the two-dimensional echocardiography; 2) enhancement of dilated coronary sinus before the right atrium after contrast material infusion into left arm vein; 3) normal transit of contrast injected from the right arm. Diagnosis can also be done by Multislice computed tomography or magnetic resonance venography [38]. Single or multiplane 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, coartation 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 abnormlities 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 PSLVC. 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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