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Case Report

Double superior vena cava incidentally found during permanent catheter placement: A case report and literature review ☆,☆☆

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ABSTRACT

A devastating admission to a tertiary hospital intensive care unit led to the discovery of a case of congenital duplicated superior vena cava (DSVC). Following the insertion of a temporary left internal jugular vein catheter, a subsequent chest X-ray indicated that the left catheter was not reaching the right heart. This resulted in a chest computed tomography scan with contrast, which confirmed the diagnosis of DSVC. The asymptomatic nature of these conditions reduces their prevalence. To date, there is no documented history of DSVC patients in Palestine. Therefore, it is important to report such a case to highlight the needs for larger studies in the future that aim to define the characteristics and varying features of patients with DSVC and the associated clinical findings.

Introduction

The superior vena cava (SVC) is the largest thoracic vein, responsible for carrying deoxygenated blood from the upper part of the body, passing through the brachiocephalic veins and ending at the right atrium of the heart [1,2]. However, anatomical variations of the SVC, such as duplicated superior vena cava (DSVC), are noteworthy due to their rarity and potential complications during medical interventions.

Abbreviation: SVC, superior vena cava; DSVC, double superior vena cava; PLSVC, persistent left superior vena cava; CHD, congenital heart disease; RSVC, right superior vena cava; ABG, Arterial blood gases; ICU, intensive care unit; CV, cardinal veins; LBCV, left brachiocephalic vein; RBCV, right brachiocephalic vein; JV, jagular vein; SV, subclavian vein; CS, coronary sinus; GI, gastrointestinal; PPI, proton pump inhibitors; IV, intravenous.

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DSVC is mostly asymptomatic and often discovered incidentally. Consequently, its exact frequency remains unknown. However, the prevalence of DSVC ranges from 0.3% to 2% in the general population, rising to 10%-11% in patients with other congenital heart diseases (CHD) [3–5].

The aetiology of DSVC arises from the embryonic failure to regress the proximal part of the left anterior common cardinal vein during the thoracic venous system's development in the fifth week of the embryonic period. This failure results in the persistence of the left SVC alongside the normal right SVC, leading to the formation of a double SVC [5,6], as shown in Figure 1.

DSVC can occur independently or in conjunction with other congenital anomalies such as horseshoe kidneys, and heart defects such as transposition of great vessels, double coronary sinus, and dextrocardia [7]. Figure 2 illustrates the different anatomical types of double SVC.

The majority of patients with DSVC are asymptomatic and unexpectedly encountered during imaging or procedures such as permanent catheter placement. Despite being silent, DSVC can complicate intravascular procedures such as hemodialysis catheterization [8], central line placement [9], and pacemaker implantation [10]. As a result, it is critical to understand this anomaly and its potential medical consequences.

In this report, we aim to present a rare clinical scenario of DSVC during the insertion of a permanent catheter for an elderly female. By reporting this case, we seek to increase knowledge about DSVC and probable challenges during invasive interventions, ultimately leading to better patient clinical care and outcome.

Case presentation

The patient, an 86-year-old female, has a history of chronic kidney disease, with a baseline creatinine of 2.5-3.4 mg/dL (reference range: 0.6-1.1 mg/dL), asthma, valvular heart disease, and a history of rectal carcinoma. She underwent colonic polypectomy in addition to laparoscopic cholecystectomy 4 years ago. She takes nifedipine and hydralazine on a regular basis for hypertension, as well as symbicort and ipratromium promide for asthma. The patient is known to be a nonsmoker with no known drug or food allergies.

After her relatives noticed her sleepiness, they admitted the patient to the hospital. After initial evaluation, the patient was found to have O2 saturation of 93% on room air, electrocardiography (ECG) showed atrial fibrillation, brain computed tomography (CT) scan came back free, chest X-ray manifested bilateral pleural effusion and laboratory findings revealed hyponatremia along with hypochloremic metabolic alkalosis. The patient was admitted to the ward and given intravenous fluids after nephrology consultation along with bisoprolol and enoxaparin for atrial fibrillation.

After performing ultrasound-guided pleurocentesis on the left side, the procedure became complicated due to pneumothorax, necessitating the insertion of a chest tube. Then, the patient had an echocardiogram, which showed left ventricular hypertrophy and dilatation, pulmonary artery systolic pressure of 81 mmHg, mild aortic stenosis, moderate mitral regurgitation, tricuspid regurgitation, an ejection fraction of 25%, and 5 centimetres of left pleural effusion. Several days later, the patient had improvement in laboratory findings with respect to sodium, chloride, pH, and bicarbonate level. The neurology team assumed that this episode of sleepiness was related to hyponatremia as a result of furosemide overuse. The neurologists decided to start the patient on normal saline at a rate of 50 cc per hour after blood transfusion as hemoglobin level was 7.6 and complete blood count showed normochromic anemia. They had 2 possible differentials of anemia, either anemia of chronic kidney disease or anemia due to upper gastrointestinal bleeding (GI). Therefore, enoxaparin was stopped. The patient was given therapeutic proton pump inhibitors, and the surgical team was consulted, and after a detailed assessment, upper GI bleeding was ruled out.

The patient suddenly developed shortness of breath and tachypnea on room air, with a respiratory rate of 30-33 and an O2 saturation of 93%-94%. On physical examination, there was bilateral diffuse wheezes. A chest X-ray was done and excluded pneumothorax. The diagnosis was either asthma exacerbation or transfusion-associated circulatory overload. After a pulmonology consultation, the patient started on hydrocortisone along with several trials of nebulizers (salbutamol, budesonide, ipratropium bromide), magnesium sulphate, and furosemide. Initially, the patient improved mildly, but 5 hours later she became sleepy with a decreased level of consciousness. We performed arterial blood gases (ABGs) and found that the patient's pH was 7.42 (reference range: 7.35-7.45), her PaCO2 was 47 mmHg (reference range: 35-45 mm Hg), her PaO2 was 58 mmHg (reference range: 75-100 mm Hg), and her HCO3 was 30 mEq/L (reference range: 22-26 mEq/L). Upon admission to the ICU, the patient was intubated and deeply sedated. We inserted a temporary left internal jugular vein catheter, but a chest X-ray showed that the left catheter was not reaching the right atrium. Consequently, we performed a CT scan with intravenous (IV) contrast, which identified DSVC as a congenital anomaly. The catheter was functioning, and the patient underwent 3 hemodialysis sessions using it with improvement in her consciousness, creatinine, and blood urea nitrogen levels.

Discussion

The embryonic development of the cardiovascular system involves the formation and regression of cardinal veins, ultimately giving rise to SVC through the fusion of the right common cardinal vein and right anterior cardinal vein. Anomalies in this process can result in a variety of anatomical variations such as DSVC, which occurs in approximately 0.3% to 2% of the general population. DSVC is often associated with other congenital heart diseases, further complicating its clinical significance [2,11].

The cardinal veins (CVs) play a vital role in draining the embryonic body, while the anterior and posterior ones drain the cranial and caudal parts of the body, respectively. Both veins empty into the common CVs, which then enter the sinus vein of the embryonic heart [11].

Between the seventh and 10th weeks of gestation, the proximal left anterior CV regresses and disconnects from the sinus venosus. This causes a shunt to the right anterior cardinal

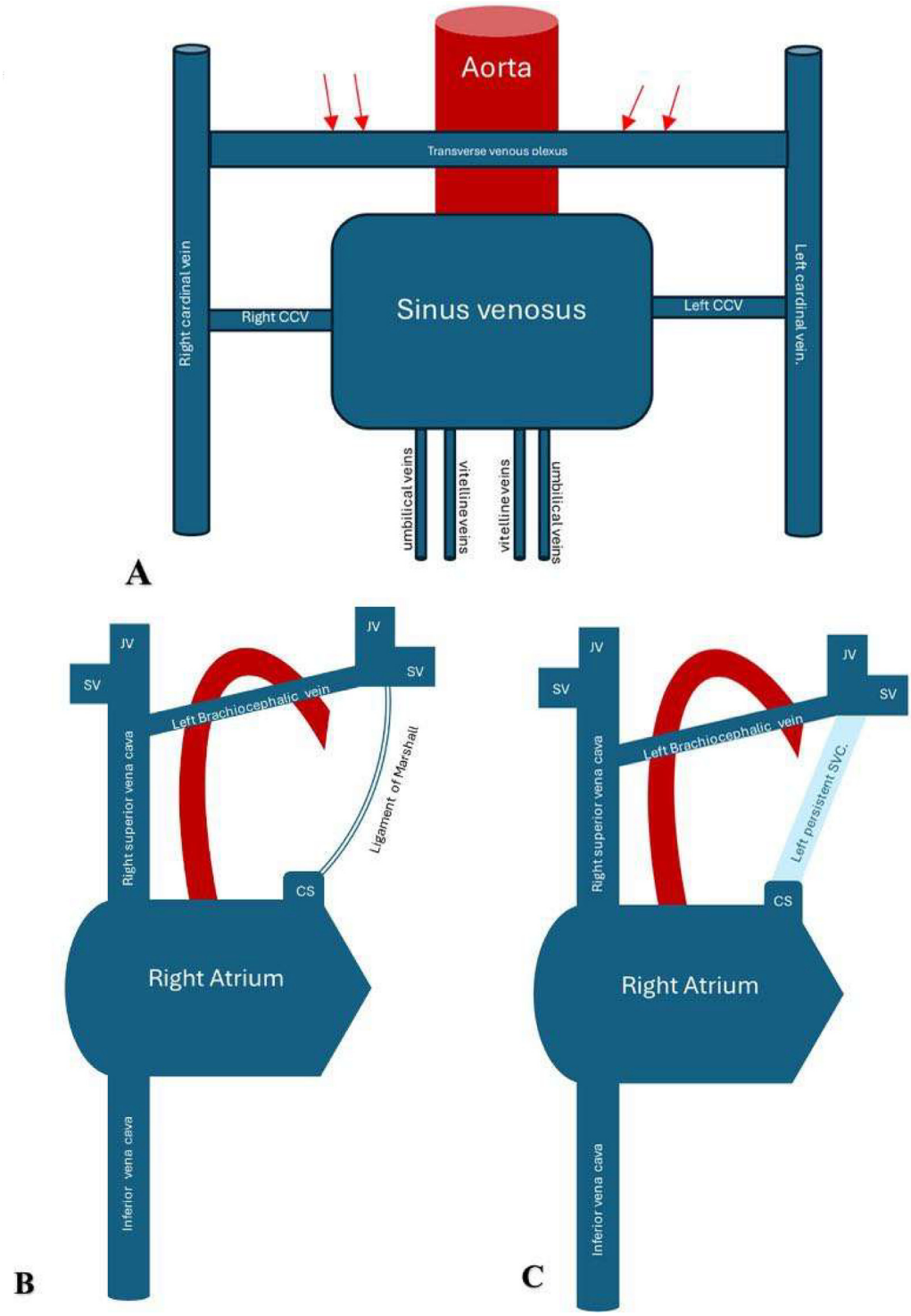

Fig. 1 – The embryological development of DSVC (A) Transverse anastomosis (red arrow) between right and left anterior cardinal veins. (B) Normally, the proximal part of left anterior cardinal vein regresses and forms the ligament of Marshall. (C) If failure of this obliteration occurs, this leads to persistent left SVC occur.

vein, which forms the left brachiocephalic vein. Meanwhile, the right anterior cardinal vein transforms into the right brachiocephalic vein. Meanwhile, the segment between the junction of the left and right brachiocephalic veins and the right atrium develops into the superior vena cava (SVC) [11]. The persistence of the left anterior cardinal vein leads to the formation of the left superior vena cava [2].

There are 4 types of DSVC based on certain anatomical features. These include whether the left brachiocephalic vein (LBCV) is present or not, whether there is an anastomotic vein

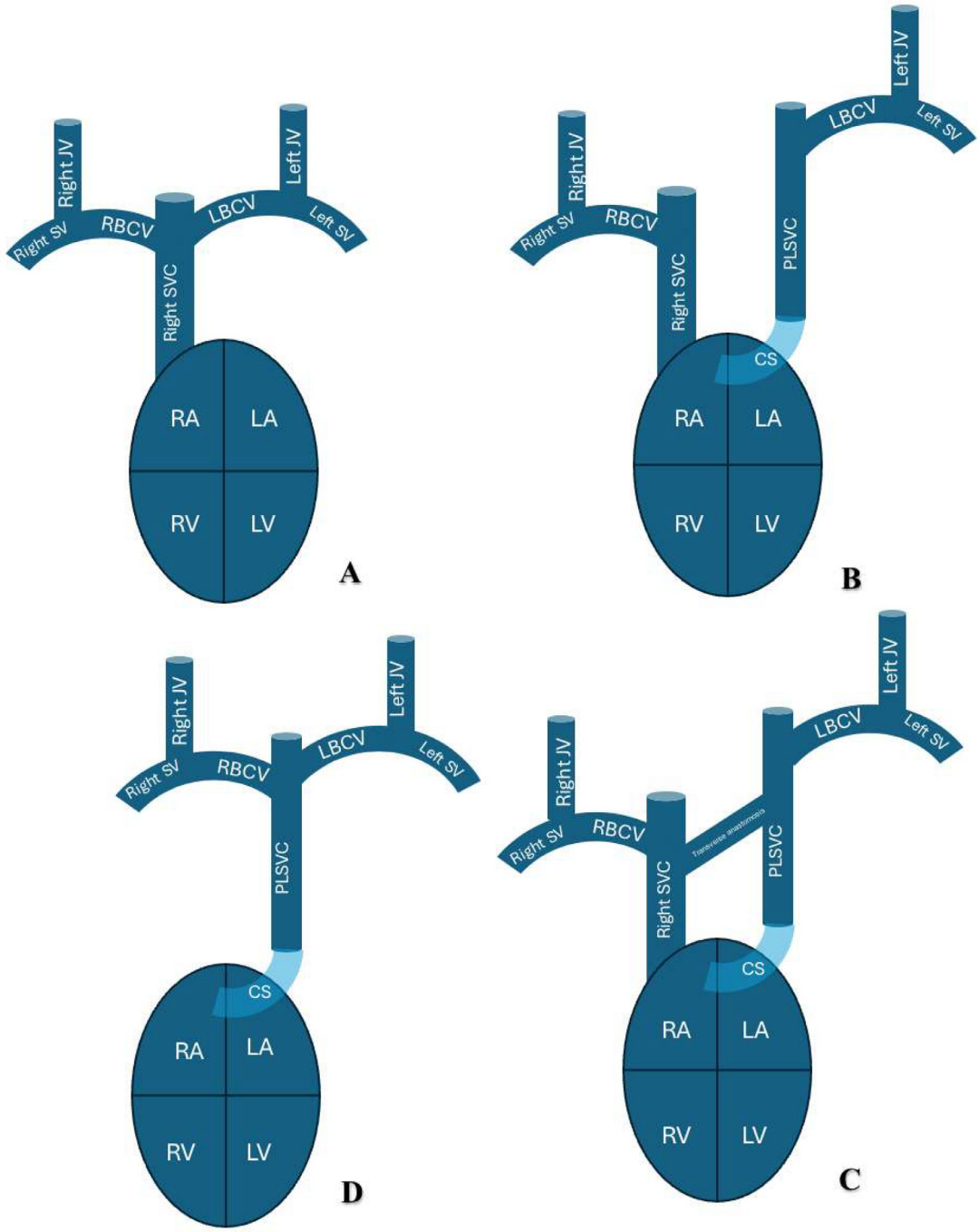

Fig. 2 – Anatomical types of PLSVC. (A) Normal, right sided SVC. (B) Double SVC, without any anastomosis. (C) Double SVC, with transverse anastomosis between both venae cavae. (D) Left SVC, with absent of right SVC.

bridging the bilateral superior vena cava (SVC), and how the left superior vena cava drains. These types are as follows:

- Type I: DSVC without LBCV, with LSVC drainage into the right atrium via the coronary sinus.
- Type II: DSVC with LBCV, with LSVC drainage into the right atrium via the coronary sinus.
- Type III: DSVC without LBCV, where the LSVC drains into the right atrium via an anastomotic vein bridging the LSVC and RSVC.
- Type IV: DSVC with LBCV, where the LSVC drains into the right atrium via an anastomotic vein bridging the LSVC and RSVC [2].

Type I DSVC represents the most frequently encountered subtype, accounting for 51.6% of cases. In 80% to 90% of cases involving persistent left superior vena cava (PLSVC), it is concomitant with a right superior vena cava, resulting in the presence of a double SVC. Conversely, in 10% of cases, an isolated left SVC is present due to involution of the right cardinal vein.

In the majority of patients (85.2% of type I and type II cases), the predominant drainage pattern involves the left SVC draining into the right atrium via the coronary sinus [12].

Even a rarer anomaly involves the persistence of a left SVC directly connecting to the left atrium, occurring in approximately 7.5% of cases of left SVC. This anomaly results in a small right-to-left shunt which may lead to systemic cyanosis of varying degrees [13].

Persistent left superior vena cava (PLSVC) is not merely an anatomical anomaly, as the clinical impact of persistent left superior vena cava (PLSVC) depends on both the site of venous drainage and the presence of associated anomalies. In cases where PLSVC is unaccompanied by cardiac abnormalities, it is often asymptomatic and discovered incidentally. Conversely, when PLSVC drains into the right atrium, the coronary sinus (CS) frequently becomes enlarged which can lead to mechanical compression, fragmentation and stretching of the conduction tissue of the atrioventricular node and His bundle, increasing the risk of arrhythmias such as atrial or ventricular fibrillation [13,14]. This coronary sinus dilation can also exert pressure on the left atrium, leading to decreased cardiac output. Moreover, the proximity of a dilated CS to the mitral valve can pose challenges during mitral valve surgery, increasing the risk of intraoperative complications. PLSVC plays a prominent role in over 50% of patients in the induction and maintenance of atrial fibrillation (AF). Thus, preradiofrequency catheter ablation imaging in AF patients is crucial, not only for evaluating pulmonary venous anatomy but also for detecting PLSVC. In case PLSVC is the trigger or driver for AF, it may be eliminated surgically [13,14]. Also, double SVC can pose a challenge to certain producers such as venous catheterization, cardiac resynchronization therapy lead placement, and pacemaker implantation, as it can introduce significant challenges. In pacemaker implantation, the tortuous course of PLSVC may make it difficult to properly position the electrode. Additionally, performing CVC insertion without fluoroscopic guidance increases the risk of serious complications, including angina, hypotension, and even cardiac perforation [1,13].

The occurrence of double SVC is significantly more common in individuals with congenital heart conditions (10% to 11%) compared to the general population (0.3% to 2%) and several congenital cardiac defects have been associated with PLSVC, including shunt lesions, conotruncal malformations, left-sided obstructive lesions, right-sided lesions, aortic arch anomalies, and cervical arch anomalies. More complex malformations, such as Shone's syndrome and Raghib syndrome, as well as systemic anomalies like VACTERL association and esophageal atresia, have also been observed in conjunction with PLSVC. Subaortic stenosis in patients with isolated PLSVC is rarely diagnosed in otherwise healthy adults. Recent evidence suggests that this condition may arise later in life as an acquired lesion due to the morphology of the left ventricular outflow tract (LVOT), which can generate turbulent blood flow and subsequent endothelial damage. This damage promotes fibromuscular proliferation, ultimately leading to the formation of a subaortic membrane. Studies show a higher prevalence of conditions like coarctation of the aorta, subaortic stenosis, and mitral stenosis in patients with PLSVC, suggesting that PLSVC may interfere with normal left ventricular development and contribute to inflow and outflow tract obstructions. Additionally, PLSVC may play a role in the development of secondary subaortic stenosis following cardiac surgeries for other anomalies. Despite this, subaortic stenosis diagnosed in adulthood generally progresses slowly and is often associated with mild aortic regurgitation. Therefore, it is important to investigate other potential congenital abnormalities using echocardiography, CT scan or MRI modalities [1,3,13]. Management of a double superior vena cava (DSVC) largely depends on the presence of symptoms and associated conditions. Asymptomatic DSVC typically requires no immediate intervention but may necessitate periodic monitoring, especially when identified incidentally during procedures. Routine follow-up ensures that any potential complications, such as arrhythmias or vascular obstructions, are detected early. Symptomatic DSVC, treatment is generally aimed at addressing related arrhythmias, which may include pharmacological management or pacemaker implantation. If there are significant shunts or congenital heart defects associated with DSVC, surgical intervention may be required. Various surgical techniques have been developed to address persistent left superior vena cava (PLSVC). These techniques include ligation of the Left SVC. This approach effectively occludes the intracardiac shunt. However, it poses significant risks unless there is sufficient collateral circulation, especially in the head and neck, to maintain adequate venous return to the heart. Secondly, redirecting blood flow. This method involves guiding blood flow from the left SVC into the right atrium, thereby facilitating normal circulation. Thirdly, reimplantation of the left SVC. It's often favored in surgical practice, this technique involves repositioning the left SVC into the right atrium, pulmonary artery, or superior vena cava. It is particularly beneficial when there is a concern that an intra-atrial baffle may impede systemic or pulmonary venous return due to the orientation of the vein's orifices [15].

Double superior vena cava (DSVC) can be an incidental finding during surgery, imaging, or autopsy, but invasive angiography is considered the most reliable method for a definitive diagnosis. CT is a useful and precise imaging tool for evaluating DSVC, offering a wide view and allowing analysis from multiple angles, with advanced postprocessing techniques that enhance data usage. On cross-sectional images, a left superior vena cava (LSVC) appears as a round nodule on the left side of the aortic arch, which can sometimes be confused with swollen lymph nodes. Coronal images can confirm the presence of the LSVC and show nearby structures and its anatomical path, including how it drains in certain patients. While nonenhanced CT can detect an LSVC, small LSVCs may be overlooked, especially if they drain into the right superior vena cava (RSVC) through a connecting vein, as the LSVC may collapse in such cases. For patients with unclear drainage sites on nonenhanced CT or those requiring further treatment, contrast-enhanced CT or angiography is needed. However, CT angiography can sometimes miss the drainage patterns of a bilateral SVC and may not clearly show the direction of blood flow, with the timing of scans after contrast injection impacting accuracy. Other imaging methods, like echocardiography and cardiovascular magnetic resonance imaging (CMR),

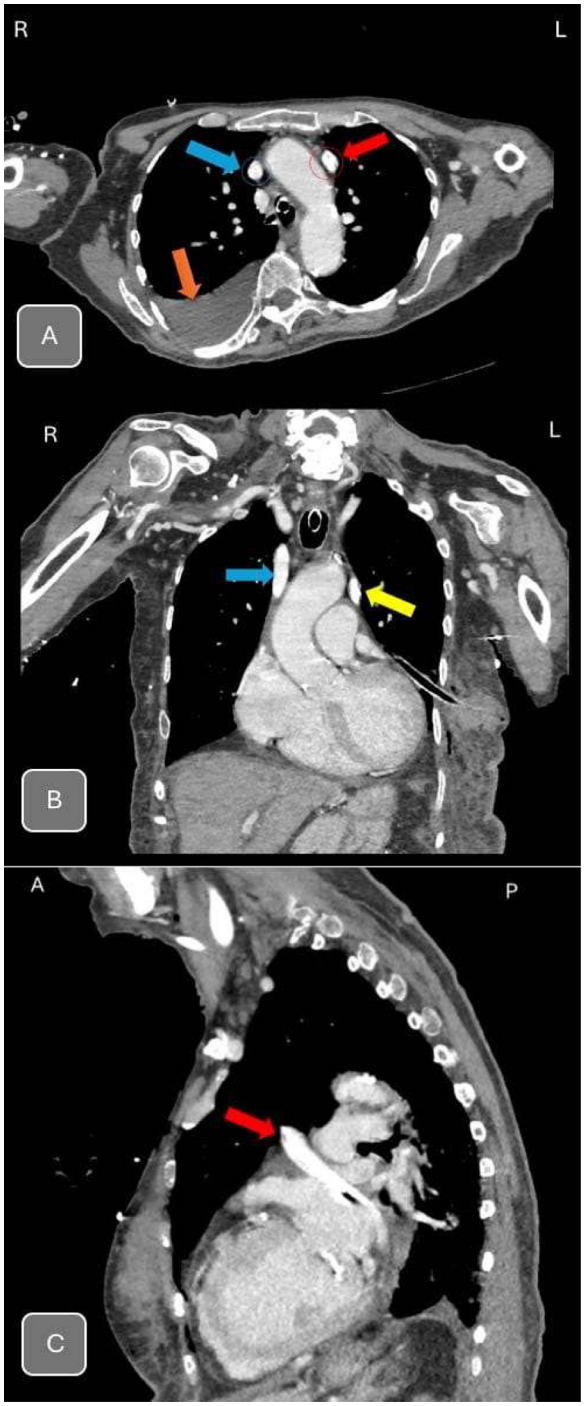

Fig. 3 – Several views of CT scan with IV contrast. (A) Axial chest CT scan with IV contrast (venous phase) show Double SVC, right sided SVC (blue arrow), Left SVC (red arrow), and right pleural effusion is also noted. (B) Coronal chest CT scan with IV contrast (venous phase) show Double SVC, right sided SVC (blue arrow), and Left SVC (yellow arrow). (C) Sagittal chest CT scan with IV contrast (venous phase), Left SVC is draining into coronary sinus with a central line (red arrow), its distal tip in mid left SVC (away from coronary sinus).

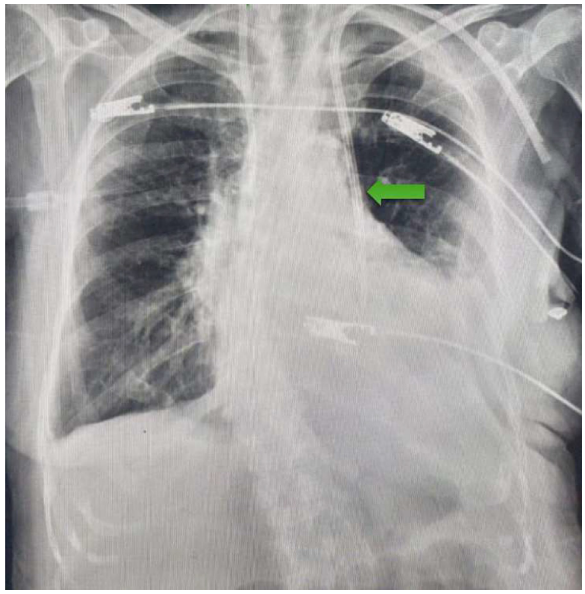

Fig. 4 – Chest X-ray show right sided central line (in the right SVC), Left perm Cath in the left sided SVC (green arrow). in addition to, Blunting of both costophrenic angles (more on the left side).

can also be used to examine the LSVC. Echocardiography is readily available and can be done at the bedside, but its accuracy can be limited by the patient's body structure. CMR is a more advanced imaging technique that provides detailed information about heart structure and function but is less commonly used than echocardiography or CT due to its complexity and cost [1,2].

Conclusion

The embryonic development of the cardiovascular system involves complex processes of vein formation and regression, leading to the establishment of SVC and its variations. Anomalies such as DSVC can occur due to disruptions in these developmental pathways, impacting approximately 0.3% to 2% of the general population.

Persistence of left SVC is mostly asymptomatic and harmless but potential complications include: systemic cyanosis and rhythm disturbances like sinus node dysfunction and atrioventricular block. These complications can pose challenges during certain medical procedures.

Detection of DSVC often occurs inadvertently during surgery, imaging, or autopsy, with invasive angiography serving as the gold standard for diagnosis. Given the association of DSVC with congenital heart diseases, comprehensive investigations using echocardiogram, CT, or MRI are crucial to identify potential coexisting defects. Understanding these complexities is essential for accurate diagnosis, treatment, and management of patients with DSVC and related conditions (Figures 3 and 4).

Ethics approval

An Najah National University does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article. A copy of the approved consent according to An Najah National University Hospital guidelines could be submitted to the editor-in -chief, if it's necessary.

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