



12TH Annual Scientific Meeting

Innovations in Dialysis Access Care: Moving the Field Forward

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Congenital Anomalies of Superior Vena Cava and Central Venous Catheterization

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UNIVERSITÀ DEGLI STUDI
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FACOLTÀ DI MEDICINA E CHIRURGIA

Congenital Anomalies of Superior Vena Cava and Central Venous Catheterization

- **Although rare, the presence of congenital anomalies of the SVC may determine failure and/or complications of CVC implantation.**
- **Thus, knowledge of normal vascular anatomy and its possible variants is crucial for the success of the procedure**

THE LEFT-SIDED SUPERIOR VENA CAVA

BY

MAURICE CAMPBELL AND D. C. DEUCHAR

From the Cardiac Department, Guy's Hospital, and the Institute of Cardiology

Received June 12, 1954

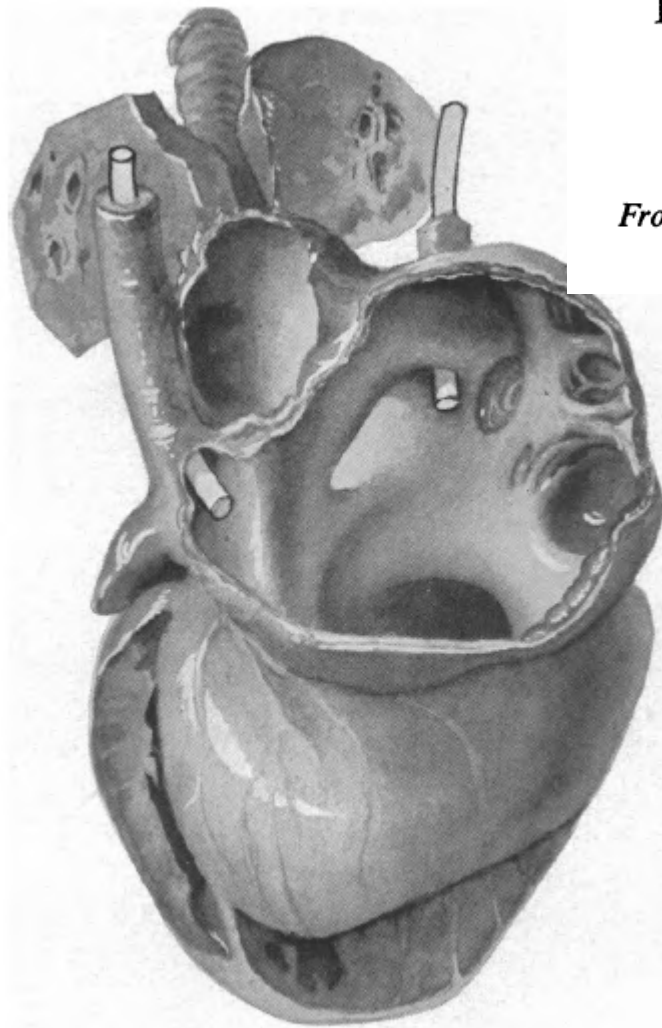


FIG. 4.—The heart with the atria opened and viewed from behind showing bilateral superior venæ cavæ, the right entering the right atrium normally and the left entering an unusually dilated coronary sinus that looks as if it was part of a very large right atrium. The left atrium is pushed upwards and to the left. There was a large atrial septal defect which no doubt further increased the size of the right atrium. Both atria communicated with their respective ventricles but the left was very much smaller than the right. Case 45, Brinton and Campbell (1953).

cm 1 2 3 4 5 6

ASDM Clinical Case Focus



Persistent Left Superior Vena Cava: Diagnosis and Implications for the Interventional Nephrologist

Haimanot Wasse

Semin Dial 2006; 19: 540–542

Renal Division, Department of Medicine, Emory University, Atlanta, Georgia

- **A PLSVC is the most common variant of anomalous venous entry into the heart.**
- **It should be considered in the setting of:**
 - **Difficult central vein catheterization**
 - **When a central venous catheter takes an unexpected left-mediastinal course**

Congenital anomalies of superior vena cava and their implications in central venous catheterization

Umberto G. Rossi¹, Paolo Rigamonti¹, Pierluca Torcia¹, Giovanni Mauri², Francesca Brunini³, Michele Rossi⁴, Maurizio Gallieni³, Maurizio Cariatì¹

PLSVC = Persistent left superior vena cava

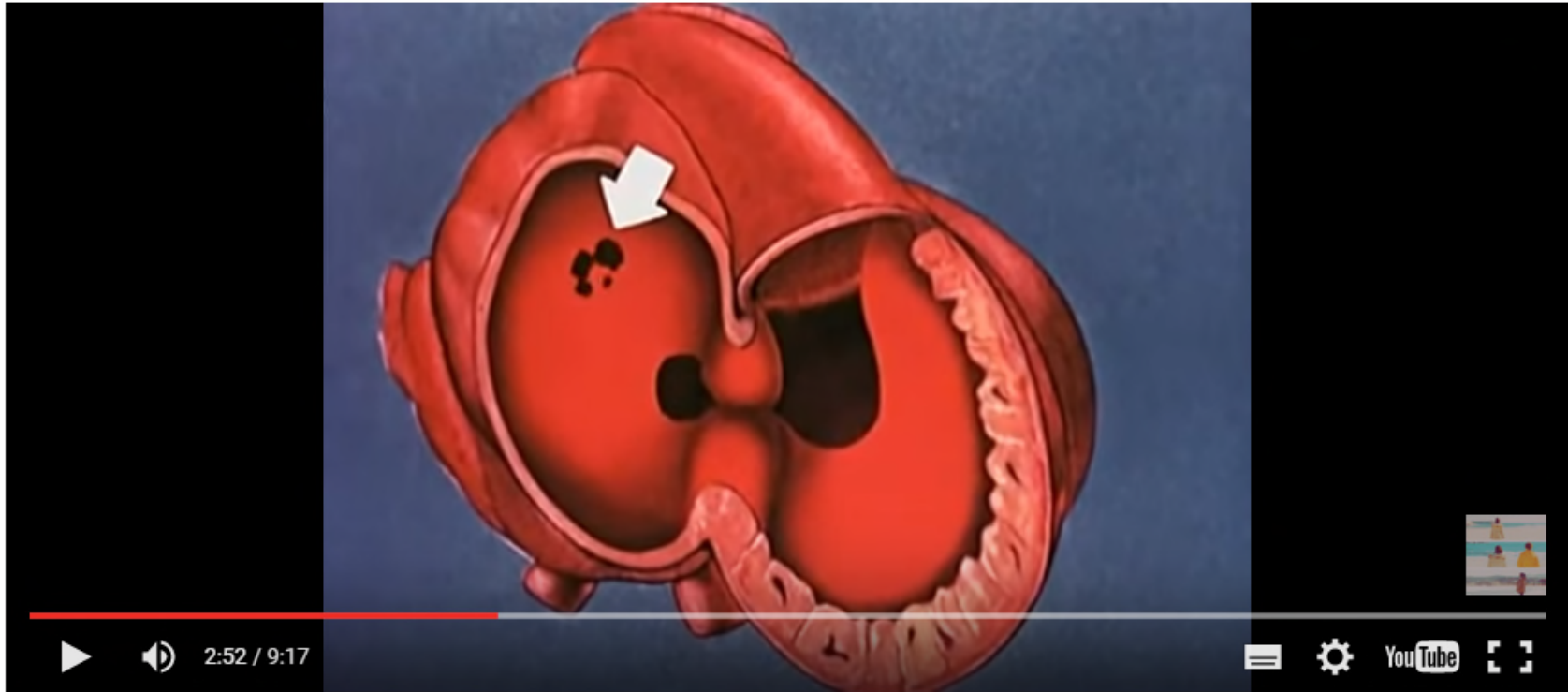
- **Embryology of the superior vena cava**
- **Incidence of PLSVC**
- **Diagnosis of PLSVC**
- **Clinical significance of PLSVC for CVC placement**

<https://meducation.net/resources/1750959-Cardiovascular-System-Heart-Embryology>

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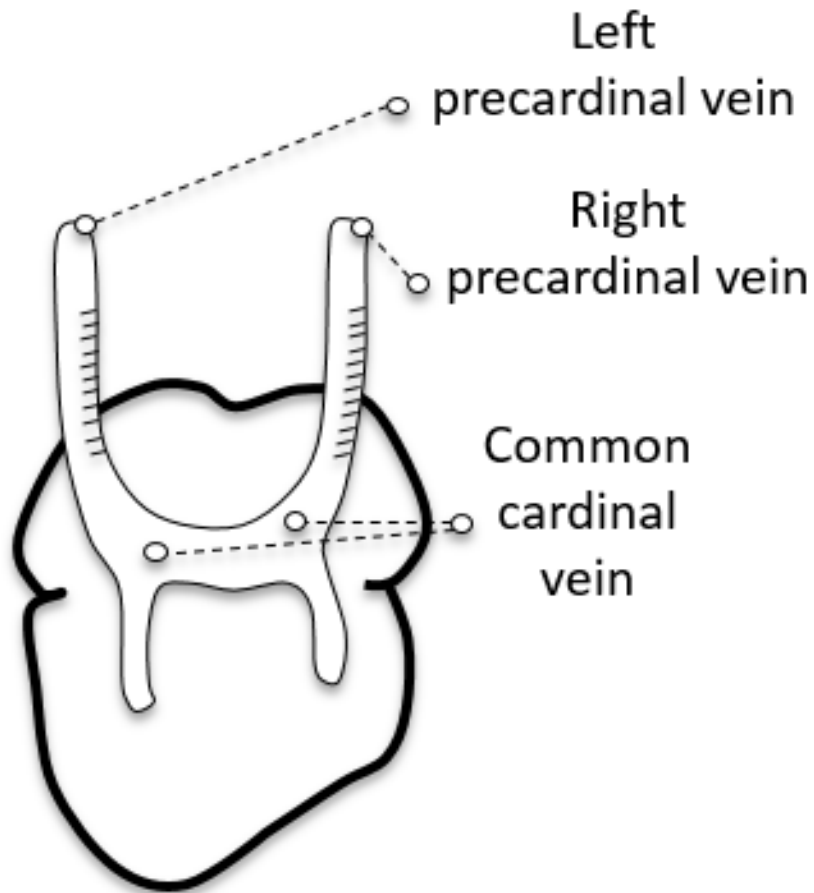
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Cardiovascular System - Heart Embryology

Embryology of the superior vena cava

Posterior view of the embryological vein system



5th week of gestation

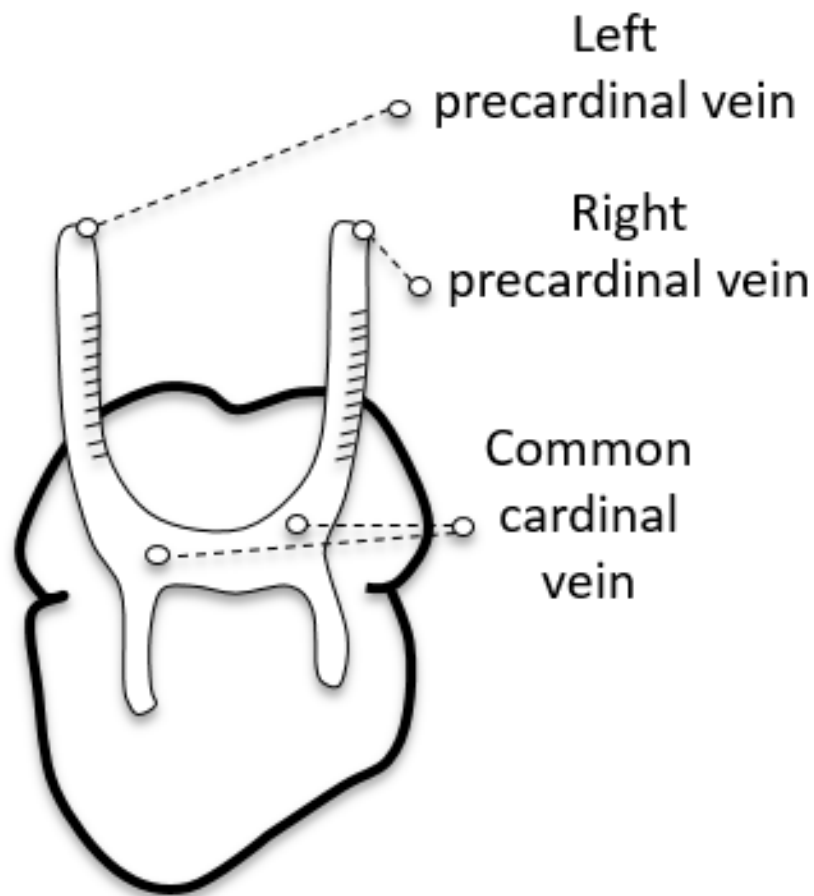
- During the fourth week of gestation, two major symmetrical vein systems, the right and the left precardinal veins, drain the upper portion of the embryo.
- Each precardinal vein drains into a common cardinal vein before entering the embryological heart.

Embryology of the superior vena cava

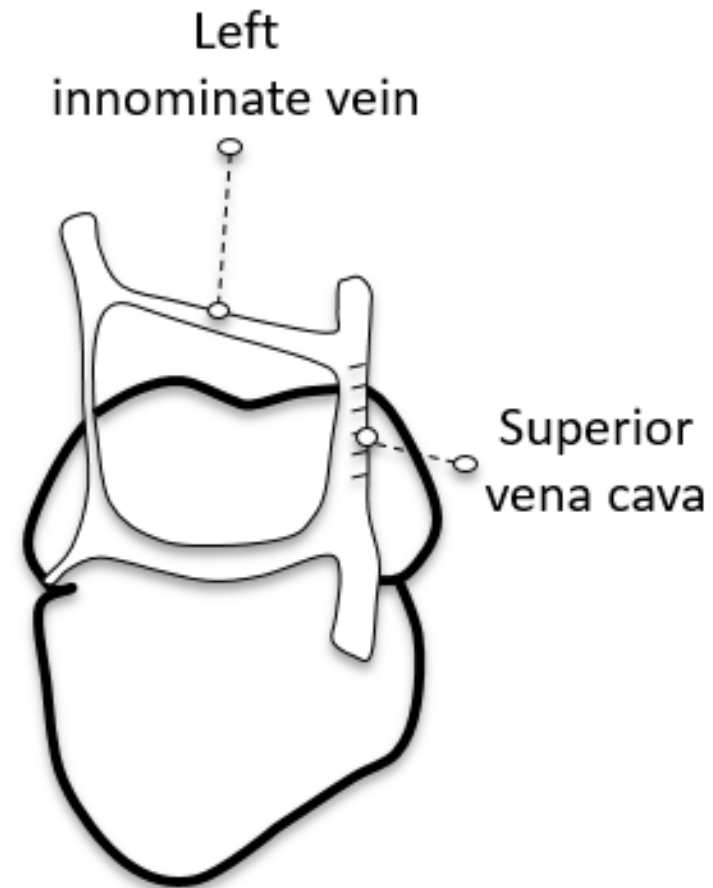
- During the fourth week of gestation, two major symmetrical vein systems, the right and the left precardinal veins, drain the upper portion of the embryo.
- Each precardinal vein drains into a common cardinal vein before entering the embryological heart.
- **By the eighth week of gestation, a vein anastomosis connects the two precardinal veins and the left common cardinal vein atrophies progressively until complete regression.**

Embryology of the superior vena cava

Posterior view of the embryological vein system



5th week of gestation



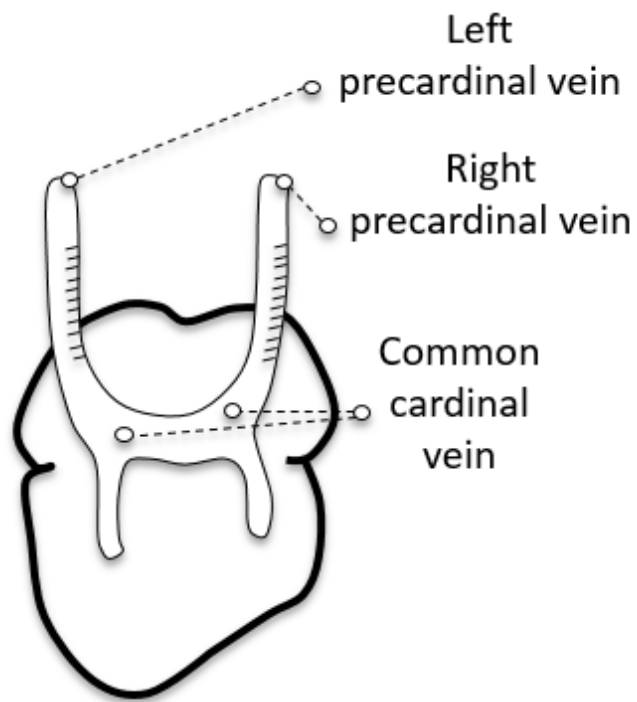
8th week of gestation

Embryology of the superior vena cava

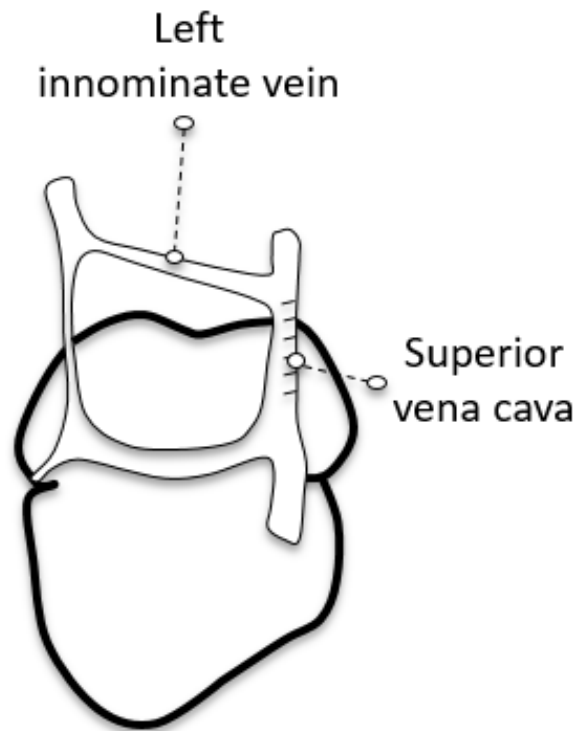
- During the fourth week of gestation, two major symmetrical vein systems, the right and the left precardinal veins, drain the upper portion of the embryo.
- Each precardinal vein drains into a common cardinal vein before entering the embryological heart.
- By the eighth week of gestation, a vein anastomosis connects the two precardinal veins and the left common cardinal vein atrophies progressively until complete regression.
- **With this regression, the anastomotic vein between the two precardinal veins becomes the left innominate (brachio-cephalic) vein and the right precardinal vein and the right common cardinal vein form the SVC.**

Embryology of the superior vena cava

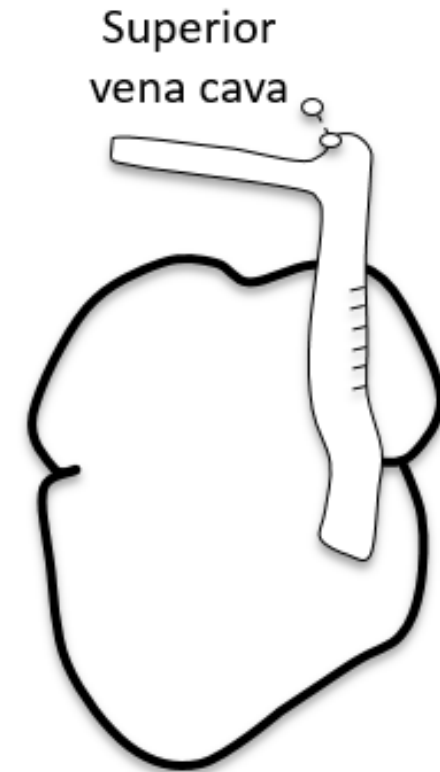
Posterior view of the embryological vein system



5th week



8th week



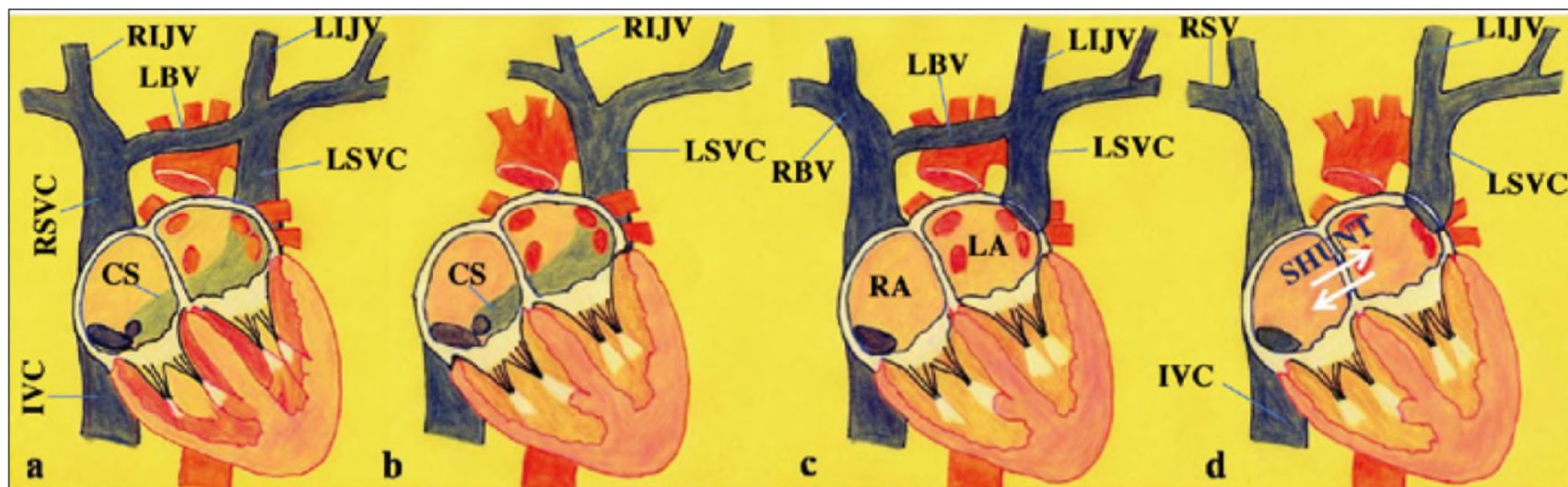
Normal

Embryology of the superior vena cava

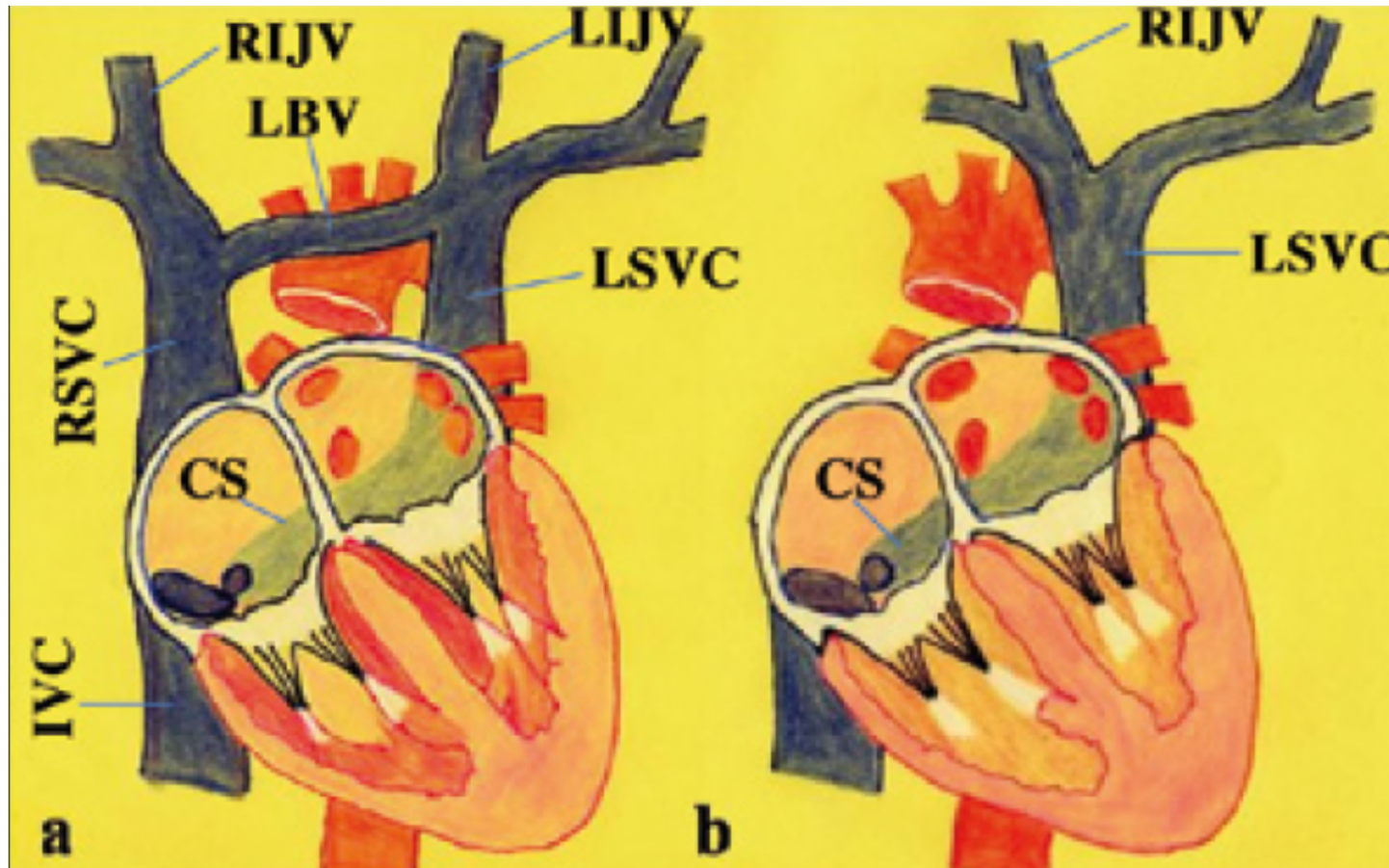
- **The failure of this normal regression can lead to the formation of a PLSVC, determining the clinical situation of double SVC.**
- **This PLSVC runs on the back of the left atrium, entering the right atrium through the orifice of an enlarged coronary sinus, although in about 10% of cases it drains in the left atrium.**
- **If the normally persistent right cardinal vein undergoes regression, then there is only an LSVC. This anatomical variant is the rarest, with a right innominate (brachio-cephalic) vein draining blood from the right to the left**

Persistent left superior vena cava: what the interventional nephrologist needs to know

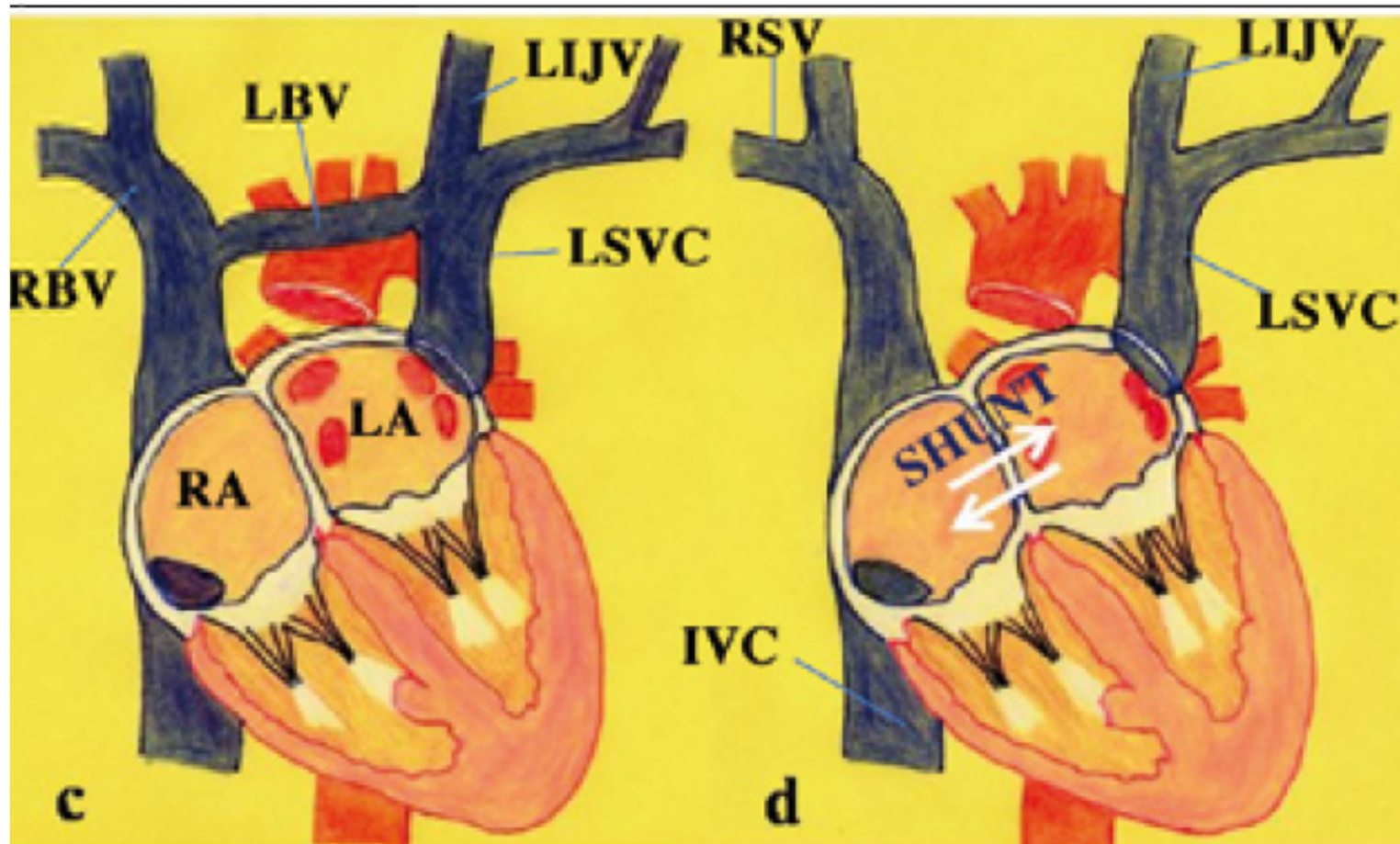
Antonio Granata¹, Simeone Andrulli², Fulvio Fiorini³, Francesco Logias⁴, Michele Figuera¹,
Renzo Mignani⁵, Antonio Basile¹, Carmelo Erio Fiore¹



Schematic drawing of variations of PLSVC



- a) LSVC drains via the coronary sinus (CS) into the right atrium (RA);
- b) Infrequently, the right superior vena cava (RSVC) may be absent. In this case, the CS is large because it receives blood from both the right and left upper parts of the body



- c) The CS is absent, the LSVC drains directly into the left atrium (LA), and the atrial septum has no defect.
- d) The LSVC connects to the LA, and there is a posterior atrial septal defect, which allows a predominant left-to-right atrial (RA) shunt.

Congenital anomalies of superior vena cava and their implications in central venous catheterization

Umberto G. Rossi¹, Paolo Rigamonti¹, Pierluca Torcia¹, Giovanni Mauri², Francesca Brunini³, Michele Rossi⁴, Maurizio Gallieni³, Maurizio Cariatì¹

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- Embryology of the superior vena cava
- **Incidence of PLSVC**
- Diagnosis of PLSVC
- Clinical significance of PLSVC for CVC placement

Congenital Anomalies of Superior Vena Cava Incidence

- The persistence of LSVC is described as the most common central venous anomaly.
- Its prevalence in the general population is estimated at **0.1-0.3%**.
- In patients with congenital heart disease, the reported prevalence ranges **between 2.1 and 5%**.

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Congenital Anomalies of Superior Vena Cava

Diagnosis

Radiologic signs suggestive of the presence of a PLSVC are:

- **Widening of the mediastinum**
- **An enlarged aortic shadow**
- **A paramediastinal bulge below the aortic arch on a posterior–anterior plain film**
- **Marked dilatation of the coronary sinus on echocardiography**
- **A leftward P axis with normal PR interval on electrocardiogram.**

Clinically, a left jugular vein distention may be noted.

Rossi U et al. J Vasc Access 2015; 6: 265-8

Wasse H. Semin Dial 2006; 19: 540–542

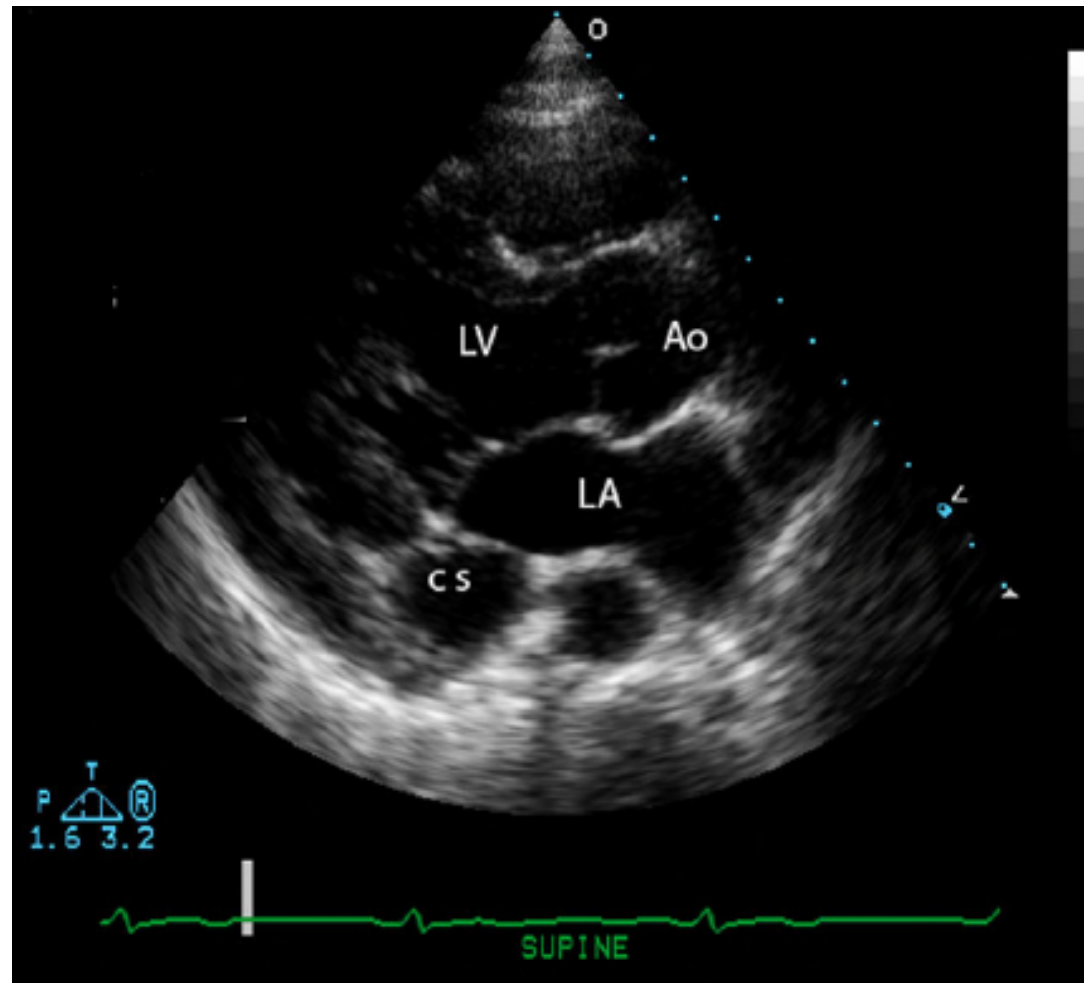
Congenital Anomalies of Superior Vena Cava

Diagnosis

- In about 90% of cases, the LSVC drains into the right atrium via the coronary sinus.
- In 10% of cases, the LSVC **drains into the left atrium**, causing right-to-left shunting, which may determine cyanosis and may be associated with cardiac malformations.
- Importantly, draining into the left atrium can be associated with hemodynamic instability, syncope, systemic emboli (thrombotic and infectious)
- More often, these anomalies are detected incidentally during imaging studies performed for other reasons

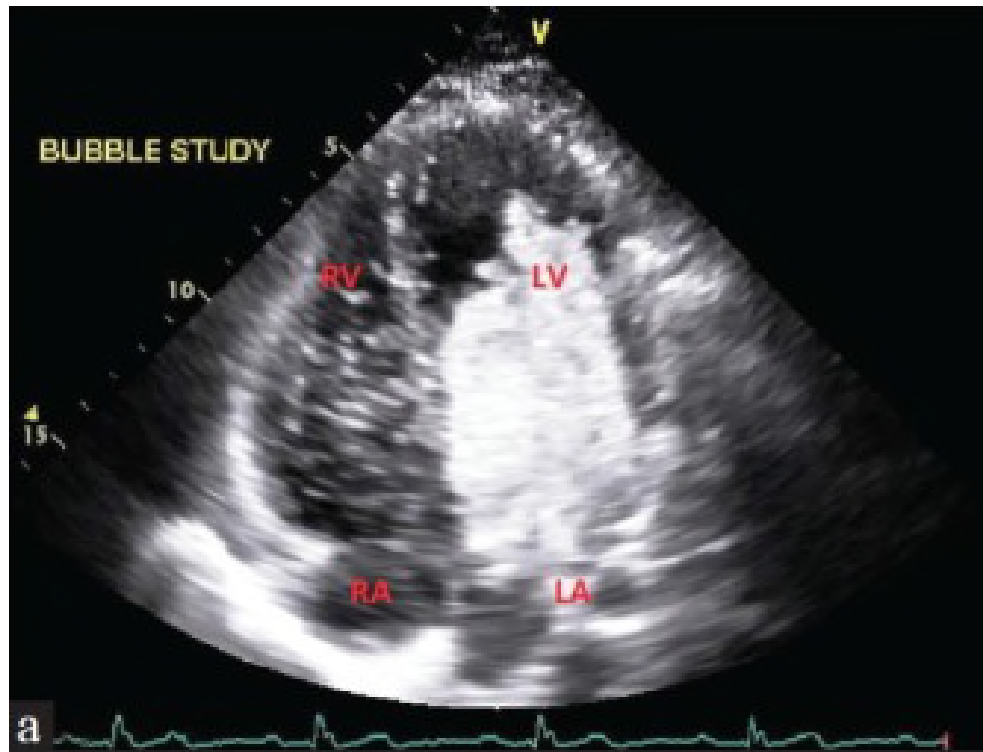
Congenital Anomalies of Superior Vena Cava Diagnosis

A dilated coronary sinus (CS) seen on echocardiography should raise the suspicion of PLSVC



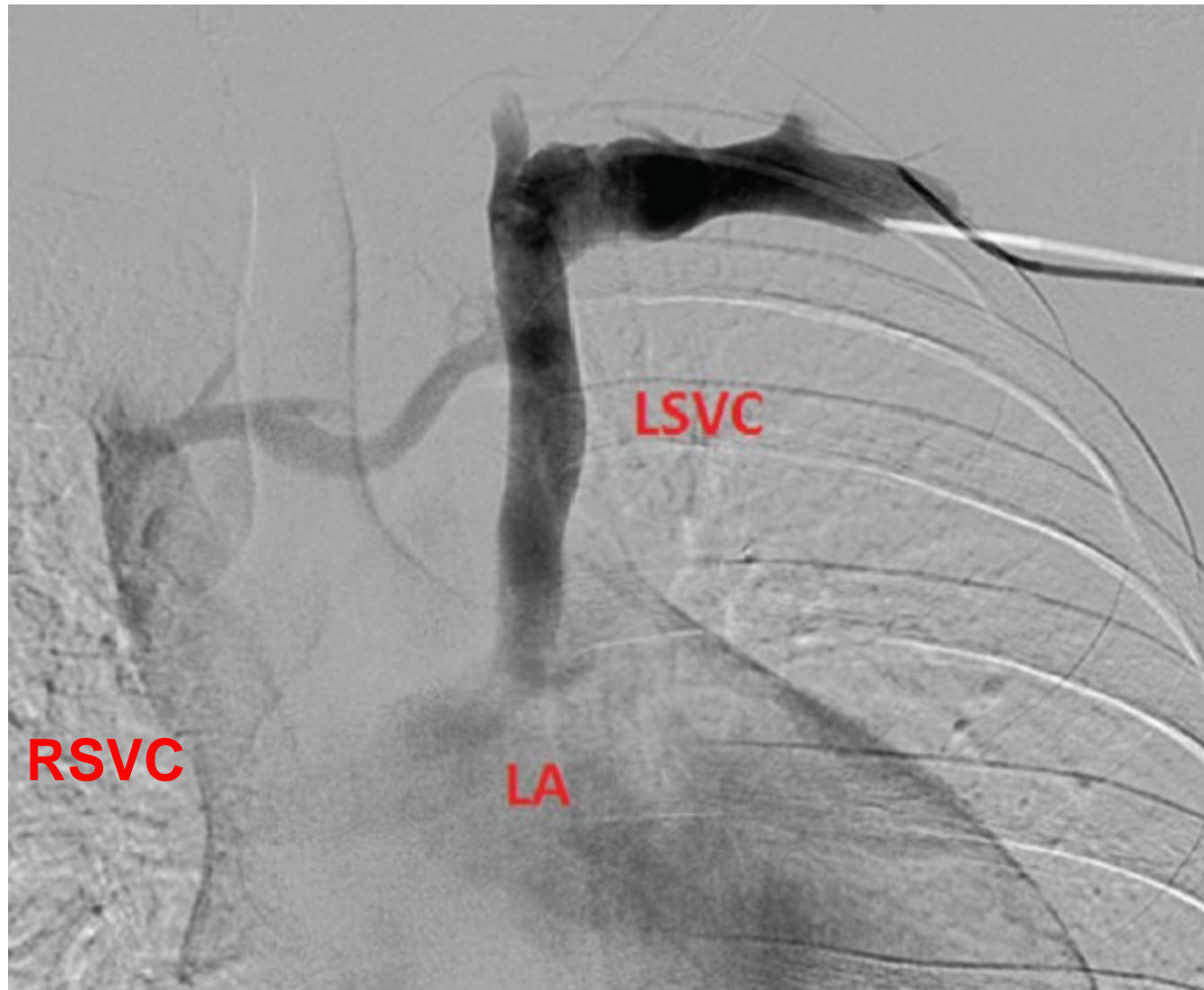
Congenital Anomalies of Superior Vena Cava Diagnosis

- **Contrast echocardiography is the main diagnostic test of PLSVC**



Echocardiogram showing a bolus of bubble into the left atrium (LA) and ventricle (LV) after peripheral injection (Tip: inject in the left arm).

Left arm venous angiogram showing LSVC draining into the left atrium (LA)



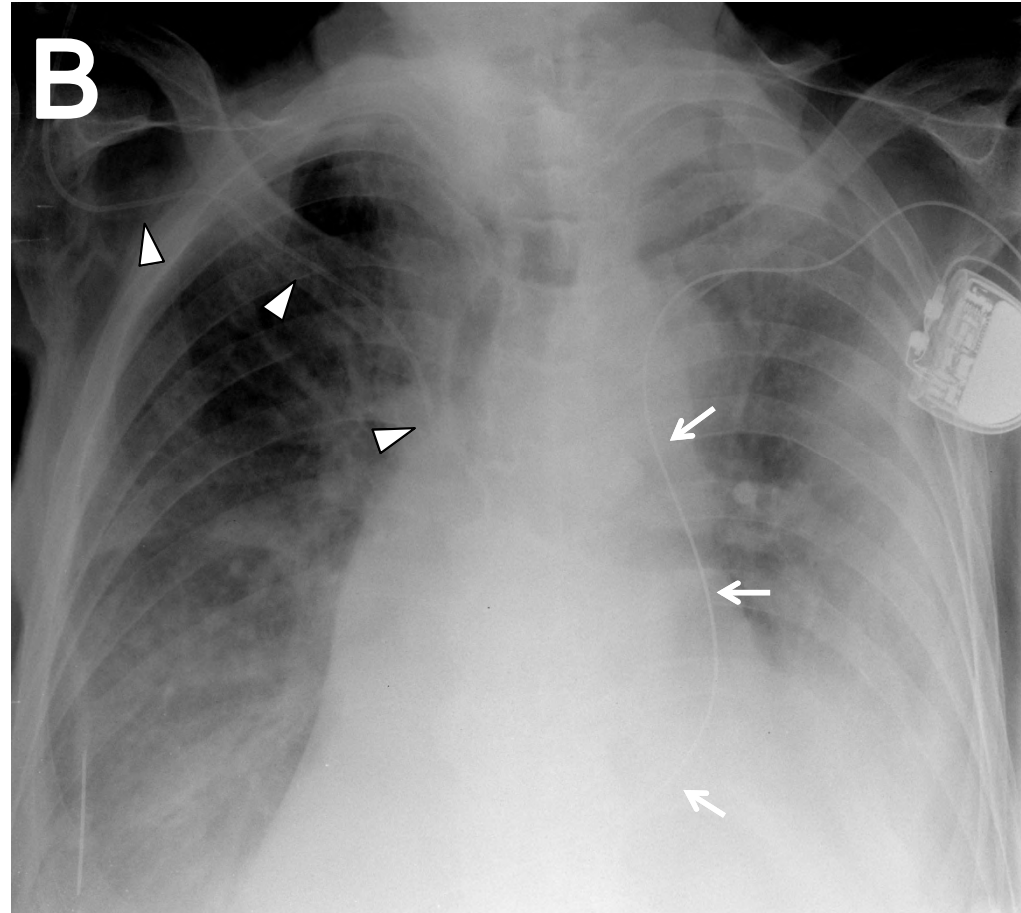
Gupta S. N Am J Med Sci. 2013; 5: 496–497

Chest venography can clarify PLSVC blood flow patterns



PLSVC (double superior vena cava)

Rossi U et al. J Vasc Access 2015



Right side CVC (arrowheads) and left side pacemaker (arrows).

CT scan: coronal multiplanar reconstruction



Rossi U et al. J Vasc Access 2015

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Clinical significance of PLSVC for CVC placement

Two different settings should be considered:

1. Patients with known congenital anomalies of the SVC
 - Accurate preoperative planning with imaging description of veins anomalies
 - Careful selection of the most adequate vascular access (avoid CVCs whenever possible, consider femoral access)
 - Use of ultrasound and fluoroscopic guidance by skilled operators.
2. Patients with no prior diagnosis of congenital anomalies of the SVC
 - Recognize the possibility of PLSVC during a procedure with unusual CVC positioning
 - Act consequently

Rossi U et al. J Vasc Access 2015; 6(4):265-8

Wasse H. Semin Dial 2006; 19: 540–542

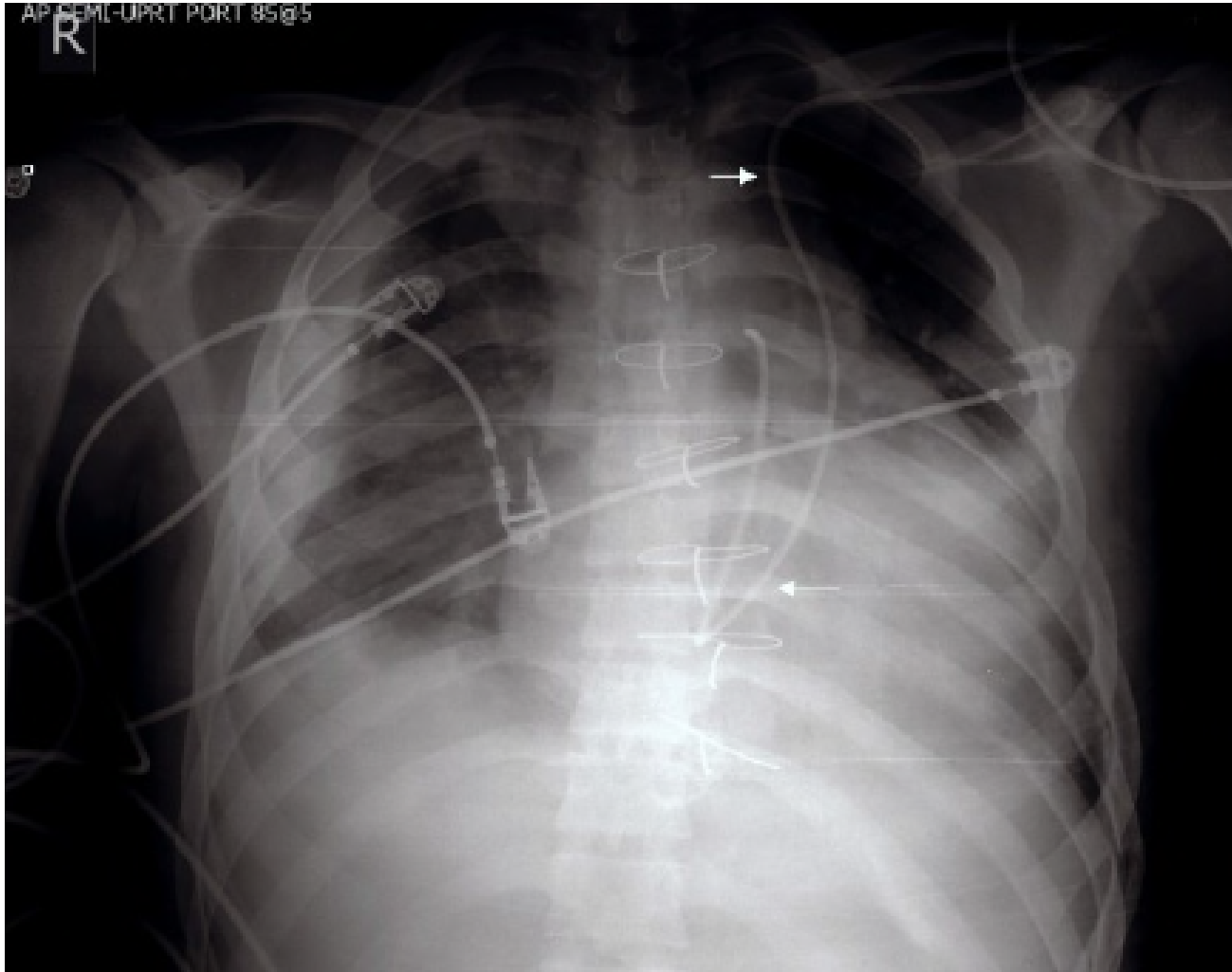
Clinical significance of PLSVC for CVC placement

- **In patients with no prior diagnosis of PLSVC, the most common situation is noticing, during an insertion procedure done under fluoroscopy, that the catheter follows an anomalous left-sided course.**
- **This may not only represent the presence of a PLSVC but also an intra-arterial catheter placement, as well as other possible tip displacements (pericardium, mediastinum pleural space, minor thoracic veins)**
- **In the absence of a right SVC, perforation of the right brachiocephalic (innominate) vein may be observed, while catheter tip manipulation in the coronary sinus may cause angina, arrhythmias, cardiac arrest, or coronary sinus thrombosis.**

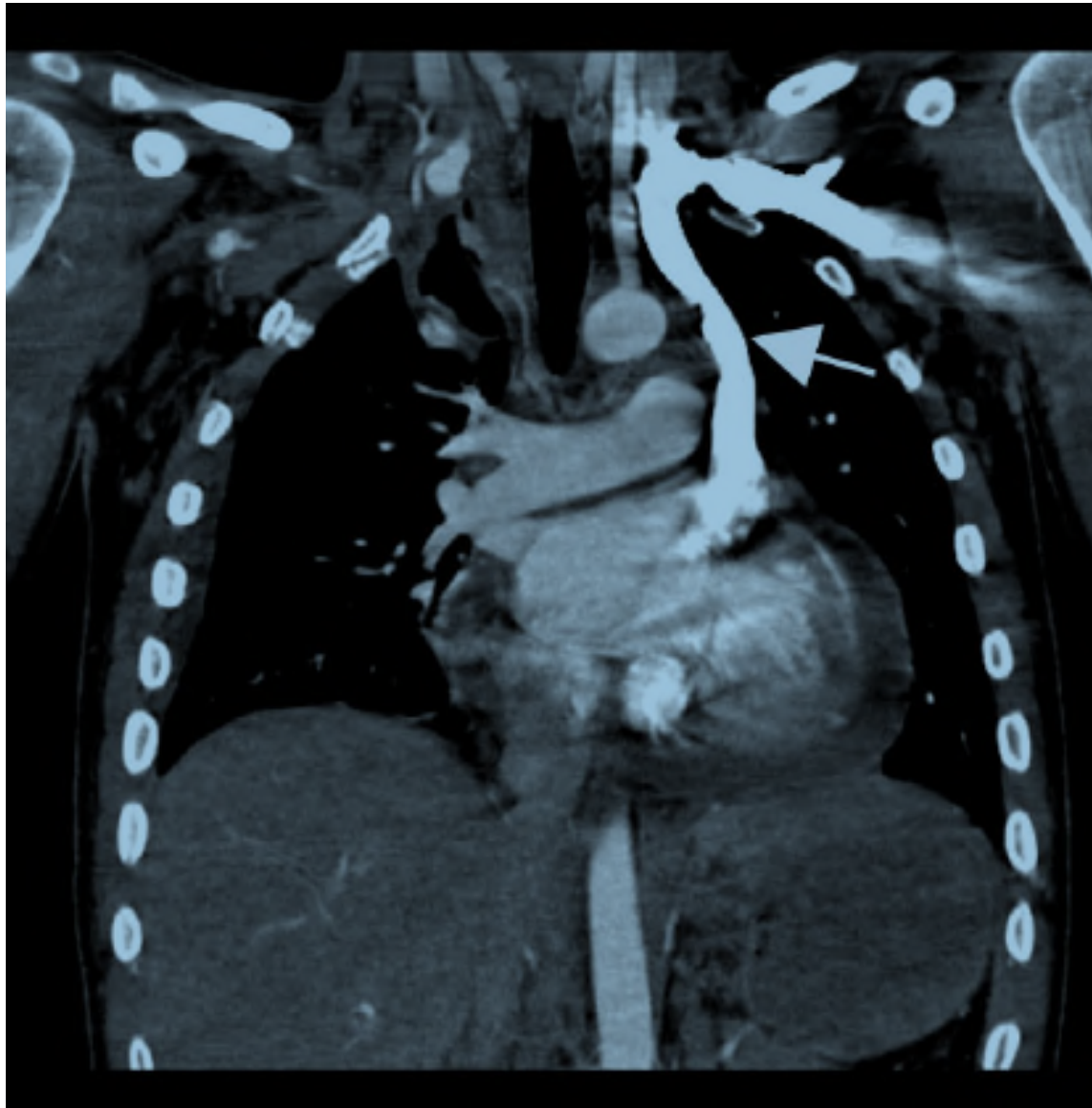
Rossi U et al. J Vasc Access 2015; 6(4):265-8

Wasse H. Semin Dial 2006; 19: 540–542

Unusual course of Swan-Ganz catheter (arrows)



Chest CT scan showing left sided SVC



Goyal S. Cardiovasc Ultrasound. 2008; 6: 50

Clinical significance of PLSVC for CVC placement

- **The presence of PLSVC may cause difficulties or possible complications during access to the right atrium, especially with a left internal jugular and a left subclavian approach, which are common sites of access for hemodialysis catheters or when placing pacemakers and Swan-Ganz catheters.**
- **Tip position: Wasse suggested that the tip should be located cranial to the PLSVC–coronary sinus junction in order to avoid coronary sinus thrombosis. This is in contrast with the guideline suggesting tip position of tunneled dialysis CVC to be positioned in the right atrium. PLSVC thrombosis may occur with both options. An AV access should be an absolute priority in this patient category**

Rossi U et al. J Vasc Access 2015; 6(4):265-8

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Conclusions

- **A persistent left superior vena cava (PLSVC) is a relatively rare but clinically relevant variant of venous return to the heart, which should be well known by the interventional nephrologist/radiologist**
- **Recognition during an interventional procedure of an unexpected course of the guidewire will indicate the possibility of performing a phlebography to confirm the possible diagnosis of congenital anomaly of SVC**
- **In patients with known anomalies of the SVC, pre-procedure imaging is warranted, in order to define the pattern of PSLVC**
- **Fluoroscopy is needed to adequately approach this clinical problem**



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