Venous Anomalies of the Thorax

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Learning objectives

• To recognize the normal anatomy, normal variants, and congenital anomalies of the thoracic systemic veins and the pulmonary veins

• Discuss the applications of multi-detector row CT and MRI in imaging of the thoracic systemic veins and the pulmonary veins

Background

Venous anomalies of the thorax can involve systemic or pulmonary veins and range from an isolated incidental finding to component of a more complex anomalies. Correct diagnosis can avoid unnecessary additional studies and may help taking some decisions like the precise display of venous anatomy prior radiofrequency ablation of arrhythmogenic pulmonary vein foci. Many anomalies of these veins may be revealed by radiography but most of them are seen on CT or MR. The CT and MR appearances of some anomalies are illustrated in this review which is divided into three sections; the first two sections, the Superior Vena Cava and the Azygos System, and the third address the pulmonary veins.

The thoracic venous anomalies are the result of complex variations in the persistence and regression of segments of three sets of veins during the first 2 months of fetal development: the umbilical, vitelline and cardinal venous systems; all three drain into the sinus venosus, which forms part of the right atrium on the right and the coronary sinus on the left.

The azygos vein is considered to derive from the upper right cardinal vein, the azygos arch from an upper segment of the right posterior cardinal vein, and the hemiazygos vein from the upper left cardinal vein. The intermediate segment of the right cardinal vein joins the IVC and azygos or hemiazygos veins, normally it regresses, but if the suprarenal segment of the IVC fails to develop, it persists, resulting in azygos or hemiazygos continuation.

During first two months of gestation, the pulmonary venous blood drains via the splanchnic plexus into the primordium of the systemic venous system while an outpouching from the primitive left atrium forms a common pulmonary vein. When the lung buds fuse with the common pulmonary vein, the splanchnic pulmonary connections are obliterated and leave four independent pulmonary veins directly entering the left atrium. When there is a failure of connection between the primitive pulmonary splanchnic plexus and the common pulmonary vein appears the anomalous pulmonary venous drainage.
Imaging findings OR Procedure details

SUPERIOR VENA CAVA

The proximal superior vena cava (SVC) is formed by the confluence of the right and left brachiocephalic veins on the right side of the superior mediastinum and then extends caudally entering the right atrium. The right anterior cardinal vein and the common cardinal vein becomes the SVC. If the left cardinal vein does not become occluded, there will be a left superior vena cava draining through the coronary sinus in the right atrium.

A persistent left SVC is an incidental finding in less than 0.5% of the general population but occurs in approximately 4% of patients with congenital heart disease, being part of a duplicated SVC in 82-90% of cases. Double SVC is a rare congenital anomaly, with a frequency between 0.3-1.3%, in which the individual has both the right and left superior vena cava draining to the right atrium (Figures 2 and 3). The frequency is higher (11%) in congenital heart disease.

In the absence of congenital heart disease, a left SVC descends lateral to the aortic arch and anterior to the hilium and enters the pericardium in the posterior atrioventricular groove and almost always drains into the coronary sinus; drainage to the left atrium is associated congenital heart disease and it is called Raghib syndrome.

Patients with a left SVC draining into a coronary sinus that has a narrowed ostium have presented difficulties in introducing IV lines and pacemaker or defibrillator leads.

The left innominate vein persists in 35% of patients with a left SVC (Figure 4). In 65%, the left brachiocephalic vein is absent (Figures 2 and 3) or small. 20% of people with a persistent left SVC have a communication between the left superior intercostal vein, the hemiazygos vein and the cava, producing a left hemiazygos arch analogous to the right-sided azygos arch.

There are no clinical signs or symptoms to suggest the presence of a left sided SVC. A left SVC does not affect the prognosis of the patient except in very rare cases where it drains into the left atrium, and so produces a right-to-left shunt causing difficulties to the surgeon in the correction of associated cardiac condition.

A left SVC can be detected if a dilatated coronary sinus, a focal widening of the left mediastinum or if an IV catheter on the left side is seen on a chest radiograph.
AZYGOS AND HEMIAZIGOS SYSTEM

The azygos system is a paired paravertebral venous pathway in the posterior thorax. The azygos vein originates at the junction of the right ascending lumbar and subcostal veins, then it enters the thorax through the aortic hiatus, and ascends anterolateral to the vertebrae; at T5-T6, it arches ventrally just cephalad to the right main bronchus and drains into the SVC or more infrequent into the right brachiocephalic vein, right subclavian vein, intrapericardial SVC, or right atrium.

The hemiazygos vein originates at the junction of the left ascending lumbar and left subcostal veins and often receives tributaries from the left renal vein and inferior vena cava (IVC). The hemiazygos vein ascends next anterolateral to the thoracic vertebrae and at T8-T9 crosses dorsal to aorta to join the azygos vein. The accessory hemiazygos vein extends cephalad in a left paravertebral position and communicates with the azygos vein at different levels.

Intercostal veins and mediastinal tributaries drain into the azygos, hemiazygos, and accessory hemiazygos veins. The right superior intercostal vein joins the azygos vein just proximal to the arch over the right main bronchus. The left superior intercostal vein communicates with the accessory hemiazygos vein in 75% of patients; it arches ventrally and drains into the left brachiocephalic vein (Figure 10, 11, 12 and 13). This vein seen adjacent to the aortic knob on frontal chest radiographs is termed the "aortic nipple." The right and left supreme intercostals veins empty into the brachiocephalic veins and communicate with the superior intercostals veins.

The interruption of the IVC with azygos (Figure 14, 15 and 16) or hemiazygos (Figure 17) continuation may be isolated or associated with other anomalies, with an incidence of 0.2-1.3% . Azygos continuation is common in patients with polysplenia (left isomerism). Other associated anomalies are abnormal abdominal situs, left or duplicated IVC and azygos lobe.

The imaging features include dilatation of the azygos vein, azygos arch (Figure 14), and SVC caused by increased flow. The hepatic veins drain into the right atrium via the suprahepatic IVC. The hepatic segment of the IVC is absent or hypoplastic.

Hemiazygos continuation of a left-sided IVC has several variations; in the first, the hemiazygos drains into the azygos vein at T8-T9 (Figure 17), where the findings are similar to azygos continuation with enlargement of the distal azygos and hemiazygos vein; in the second route, the blood flows from the hemiazygos to the accessory hemiazygos to a left SVC and into the coronary sinus; and in the third route, the
hemiazygos vein drains to the accessory hemiazygos vein, left superior intercostals vein, and left brachiocephalic vein into a normal right SVC.

PULMONARY VEINS ANOMALIES

Normally the four pulmonary veins drain into the left atrium. The right superior vein drains the upper and middle lobes, the left superior vein drains the left upper lobe and lingula, and the right and left inferior veins drain the lower lobes. The superior veins are anterior and caudal to the pulmonary arteries.

The variation in number of vein have become important and increasingly recognized because of the use of CT to depict pulmonary vein anatomy in patients with atrial #brillation who are treated by radiofrequency ablation of arrhythmogenic foci located near pulmonary vein ostia. The most common variation is three right veins, with the third vein draining the right middle lobe (Figure 18 and 19).

Anomalous pulmonary venous drainage can be partial or total. It results in a left-to-right shunt as pulmonary venous blood #ows into the right side of the heart or systemic veins. The total type needs a right-to-left shunt (septal defect or patent ductus arteriosus).

Partial anomalous pulmonary venous return (PAPVR) has an incidence approximately of 0.5%. It involves the right lung more frequently and is more often hemodynamically signi#cant when associated with congenital heart disease (Figure 25, 26 and 27) or scimitar syndrome (Figure 28, 29 and 30). Anomalous right lung veins can drain into the SVC (Figure 31, 32 and 33), azygos vein, right atrium, coronary sinus, or IVC.

Anomalous veins of the left lung often drains the left upper lobe; they continue cephalad as a vertical vein that joins the left brachiocephalic vein (Figure 25). It is almost always an incidental finding, but it may be suspected by an abnormal IV catheter and confirmed by CT. Conventional radiographs do not distinguish a persistent left SVC from an APVDLUL so careful CT analysis is required. Normally the only vessel ventral to the left main bronchus is the left superior pulmonary vein. With PAPVR, the normal left superior pulmonary vein is absent so there is no vessel ventral to the bronchus and the pulmonary veins of the left upper lobe that can be followed to the anomalous vein in the aortopulmonary window.

A PAPVR draining into the IVC, portal, hepatic, or other veins below the diaphragm in association with hypoplasia of the right lung has been called the scimitar, venolobar, or hypogenetic lung syndrome (Figure 28, 29 and 30). It is associated with atrial septal
defect, systemic blood supply to the lung, extralobar sequestration, horseshoe lung, and pulmonary arteriovenous malformation.

The findings on chest radiography of the anomalous vein draining into the IVC can be seen as a curved opacity in addition to a small right lung and dextroversion. Other anomalous right pulmonary veins that drains to the left atrium have been called "pseudoscimitar syndrome."

Total anomalous pulmonary venous return (TAPVR) has been classified into four types:

- Supracardiac to the left brachiocephalic vein, right SVC, or azygos vein. The drainage to the left brachiocephalic vein is the most common. This results in the "snowman" sign (the dilated vertical vein on the left and the large superior vena cava on the right form the head of the snowman, and the body of the snowman is formed by the heart).
- Cardiac to the coronary sinus or right atrium.
- Infracardiac to the portal vein, ductus venosus, or right atrium. It is less common, often results in early and severe congestive heart failure. The diagnosis on CT angiography and MRI have been shown to provide accurate anatomic information for preoperative planning.
- Mixed

It always have an atrial septal duct or patent foramen ovale, increased pulmonary vascularity. TAPVR is common with asplenia (right isomerism).

Images for this section:
**Fig. 1:** DUPLICATION OF SVC
**Fig. 2:** 3D CT images shows the catheter (arrows) in a left SVC.
Fig. 3: 30-year-old man with duplicated superior vena cava (SVC). Curve multiplanar CT reconstruction shows persistent left superior vena cava lateral to aortic arch on left side.
**Fig. 4:** 37 year-old female with a duplicated SVC with a left SVC passing under the aortic arch demonstrated in MIP images.
Fig. 5: 37 year-old female with a duplicated SVC with a left SVC passing under the aortic arch demonstrated in 3D CT images.
Fig. 6: 54 years-old male patient with a left SVC without right SVC in a coronal CT.
**Fig. 7**: 54 years-old male patient with a left SVC without right SVC in a MIP reconstruction.
Fig. 8: 54 years-old male patient with a left SVC without right SVC in a 3D images.
**Fig. 9:** The left superior intercostal vein communicates with the accessory hemiazygos vein in 75% of patients; it arches ventrally and drains into the left brachiocephalic vein. This vein seen adjacent to the aortic knob on frontal chest radiographs is termed the "aortic nipple."
**Fig. 10:** CT scan demonstrates the left superior intercostal vein (yellow arrow)
**Fig. 11:** MPR CT scan demonstrates the left superior intercostal vein (yellow arrow) draining into the left brachiocephalic vein (gross yellow arrow), accessory hemiazygos vein (black arrow).

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Fig. 12: Volume rendering demonstrating the accessory hemizygos vein (yellow arrows) draining into the left brachiocephalic vein.
Fig. 13: 3D images demonstrating the accessory hemizygos vein (yellow arrows) draining into the left brachiocephalic vein.
Fig. 14: Congenital interruption of the IVC with continuation of the azygos vein. The azygos arch is enlarged (yellow arrow) as it drains into the SVC.
Fig. 15: Congenital interruption of the IVC with continuation of the azygos vein. Series of CT images showing enlarged azygos vein paralleling the aorta (yellow arrow).
Fig. 16: Congenital interruption of the IVC with continuation of the azygos vein. Series of CT images showing enlarged azygos vein paralleling the aorta (yellow arrow).
**Fig. 17:** Duplication of the IVC with hemiazygos continuation of the left IVC (yellow arrows). The hemiazygos vein drains into the azygos vein in the lower thorax.
Fig. 18: Supernumerary pulmonary veins are frequently seen, the most common of which is a separate right middle pulmonary vein that drains the middle lobe of the lung. If unrecognized, it is at risk of trauma during the ablation procedure and later of pulmonary vein stenosis. Modification of ablation technique may be required in the presence of a right middle vein, because a figure-of-eight ablation is not possible, and the rim of intervening tissue between veins may not be adequate to stably support the ablation catheter. The ostial diameter of a right middle pulmonary vein (mean, 9.9 ± 1.9 mm) is smaller than that of other veins. Posterior view of CT volume-rendered image show three right pulmonary veins. Third vein (arrow) drains right middle lobe.
**Fig. 19**: 3D endocardial view show three right pulmonary veins. Third vein (arrow) drains right middle lobe.
**Fig. 20:** Supernumerary pulmonary vein. Posterior oblique views of CT volume-rendered demonstrating anomalous insertion of a supernumerary pulmonary vein (arrows) in the right posterior left atrium wall.

**Fig. 21:** Supernumerary pulmonary vein. Posterior oblique views of CT volume-rendered demonstrating anomalous insertion of a supernumerary pulmonary vein (arrows) in the right posterior left atrium wall.

**Fig. 22:** Supernumerary pulmonary vein. Oblique MIP view demonstrating anomalous insertion of a supernumerary pulmonary vein (arrows) in the right posterior left atrium wall.
**Fig. 23:** Common ostia. Posterior view of CT volume-rendered image show a left common pulmonary vein ostia (arrows). In general, common ostia are more frequently observed on the left side, whereas the presence of additional veins and early branching is described more often for the right-sided veins. Furthermore, the right-sided venous drainage is also more variable than the left-sided drainage.
**Fig. 24:** Common ostia. 3D endocardial view show a left common pulmonary vein ostia (arrows).
Fig. 25: 17 years- old patient with anomalous drainage of the superior pulmonary veins into a colector vein in the mediastinum which drains into the left brachiocephalic vein.
Fig. 26: 17 years-old patient with anomalous drainage of the superior pulmonary veins into a colector vein in the mediastinum which drains into the left brachiocephalic vein. This patient also have a bivalve aortic valvula....
Fig. 27: ... and a membrane in the outlet tract of the left ventricle.
**Fig. 28:** Chest radiograph showing a vascular opacity draining from the right lower zone to above the diaphragm - scimitar syndrome (yellow) arrow.
Fig. 29: 3D MIP images generated from a MRI multiphase angiogram provides an overall analysis of the scimitar vein (white arrows) draining into the IVC above the diaphragm without stenosis.
Fig. 30: 3D MIP images generated from a MRI multiphase angiogram provides an overall analysis of the scimitar vein (white arrows) draining into the IVC above the diaphragm without stenosis.
**Fig. 31:** Anomalous pulmonary vein from the right upper lobe in a 40 year-old man. Axial CT image shows a connection from an anomalous right upper lobe vein (yellow arrow) to the superior vena cava.
**Fig. 32:** Anomalous pulmonary vein from the right upper lobe in a 40 year-old man. Coronal Multiplanar show anomalous drainage of a right upper lobe veins into the superior vena cava (yellow arrow).
**Fig. 33:** Anomalous pulmonary vein from the right upper lobe in a 40 year-old man. 3D image show anomalous drainage of a right upper lobe veins into the superior vena cava (yellow arrow).
Conclusion

The catheter is in a vertical left mediastinal vein:

Chances are....

- Left partial anomalous pulmonary venous drainage (PAPVD) draining to the left brachiocephalic vein? or,
- Persistence of a left SVC with or without a coexisting right superior vena cava? or,
- The left superior intercostal vein communicates with the accessory hemiazygous vein draining into the left brachiocephalic vein.

Venous anomalies of the thorax are a frequent finding on imaging studies, sometimes just like the unusual course of a catheter or as a subtle finding. These anomalies usually do not cause symptoms but in some cases it can lead to complications like difficulties in introducing pacemaker or defibrillator leads or surgical ligation of a single left SVC. Diagnosing some of these anomalies may help making the diagnosis of other malformations like PAPVR from the right upper lobe can lead to the diagnosis of an atrial septal defect that predisposes the patient to paradoxical emboli.

The use of MDCT and MRI that allows multiplanar reconstructions and 3D images, radiologist can afford better evaluation of the vascular anatomy of the thorax, differentiating normal and pathologic variants. Knowledge of this anomalies helps displaying the precise anatomy and a correct diagnosis before procedures like radiofrequency ablation of arrhythmogenic pulmonary vein foci is being performed with increasing frequency.

Images for this section:
**Fig. 34:** Unusual position of a left subclavian IV catheter (arrows) on radiograph. Where is the left catheter?
Left Partial anomalous pulmonary venous drainage draining to the left brachiocephalic vein

Right SVC

Left SVC

SVC

Left superior intercostal vein

Accessory hemiazygos vein

Azygos
Fig. 35: VENOUS ANOMALIES OF THE THORAX

Fig. 36: Careful analysis of axial CT images is required; Normally one vessel is seen anterior to the left main bronchus (the left superior pulmonary vein/white arrow).
**Fig. 37:** Two vessels are seen anterior to the left main bronchus in persistent left superior vena cava (Left SVC/yellow arrow and the left superior pulmonary vein/white arrow).
Fig. 38: Whereas none of those are seen there with anomalous left upper lobe venous drainage.
Fig. 2: 3D CT images shows the catheter (arrows) in a left SVC.
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