Congenital anomalies of the thoracic veins in adults: morphologic evaluation with magnetic resonance imaging

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Learning objectives

- To review the embryological development and the anatomy of the thoracic veins.
- To illustrate, with magnetic resonance imaging, the most frequent congenital anomalies of the thoracic veins diagnosed in the adult population, of both the systemic and pulmonary circulation.
Background

Thoracic venous abnormalities can be classified into systemic and pulmonary.

While systemic venous anomalies are frequently an incidental finding, pulmonary venous abnormalities are usually symptomatic and associated with congenital cardiac anomalies.

Though angiography has played a major role in the characterization of these disorders in the past, it has currently been replaced by computed tomography (CT) and magnetic resonance (MR) angiography, being presently reserved for endovascular interventions.

While CT has excellent spatial resolution and anatomic definition, it has the disadvantage of using ionizing radiation and iodinated contrast medium.

MR is a non-invasive method, capable of multiplanar imaging and of acquiring multiple imaging phases with a single intravenous bolus of gadolinium. As it lacks ionizing radiation, it is particularly valuable in the evaluation of pediatric patients and young adults.
1) **Systemic venous anomalies**

**Anatomy and Embryology**

Normal thoracic systemic venous anatomy includes bilateral subclavian veins (SCV) and bilateral brachiocephalic veins (BCV) draining to the superior vena cava (SVC), which in turn drains to the right atrium (Fig. 1 on page 9).

Embryologically, these veins derive from the paired anterior and common cardinal veins (Fig. 2 on page 9). The right cardinal system develops into the SVC, right BCV (RBCV) and right internal jugular vein. The left anterior cardinal vein undergoes near complete atrophy below the level of the communicating vein (originating the ligament of the left vena cava). Above this level it will persist as internal jugular vein, which will join the left SCV from the developing upper limb. The left common cardinal vein will become the oblique vein of the left atrium (vein of Marshall) and will drain into the coronary sinus of the right atrium. The communicating vein, which anastomoses the anterior cardinal veins, will originate the left BCV (LBCV).

The azygos venous system initially derives from the paired supracardinal veins. The azygos vein originates from the anastomosis between the cranial segments of the right posterior cardinal vein and the right supracardinal vein, with the cranial part of the left supracardinal vein becoming the hemiazygos vein (Fig. 2 on page 9). The hemiazygos vein (located on the left side of the spine) will drain into the azygos vein (along the right side of the vertebral bodies), which in turn will drain into the SVC (Fig. 3 on page 10).

The inferior vena cava (IVC) is formed by a series of complex developmental stages involving the fusion of multiple segments of the vitelline, posterior cardinal, supracardinal, and subcardinal veins. The suprahepatic segment of the IVC originates from the proximal portion of the right vitelline vein, which is the precursor of the hepatic veins. The suprarenal segment of the IVC derives from the right subcardinal vein, which fuses with the right vitelline vein dorsal to the developing liver, forming the intra-hepatic segment of the IVC.

Anomalies of the systemic thoracic veins are rare, and may be asymptomatic. However, clear documentation of these anomalies is paramount, namely because they can interfere
in the placement of several devices (for instance central venous catheters, cardiac pacemaker and defibrillator leads).

These anomalies may occur as isolated findings or associated with congenital heart disease and arrhythmias.

Persistent left superior vena cava

A persistent left superior vena cava (LSVC) is described in 0.1% to 0.5% of the general population, frequently appearing as an isolated finding. However, almost 40% of these patients will have associated cardiac anomalies, such as atrial septal defects, bicuspid aortic valve, and aortic coarctation.

Embryologically, this anomaly results from persistence of the left anterior cardinal vein.

In the majority of cases, a right SVC (usually of reduced caliber) is also present (Fig. 4 on page 11 a). Anastomosis between the right and left SVC occurs in 35% of cases.

In the remaining cases, involution of the right cardinal venous system, along with persistence of the communicating vein and the left anterior cardinal vein, will result in absence of the right SVC, with mirror image of the thoracic systemic venous drainage (Fig. 4 on page 11 b).

In most patients (92%) the LSVC will drain through the vein of Marshal into the coronary sinus, which will enlarge to accommodate the blood flow. Enlargement of the coronary sinus may potentially lead to left atrioventricular valve inflow obstruction, and subsequently to cardiac arrhythmias and/or sudden death.

In the remaining cases, the LSVC terminates in the left atrium, resulting in a right-to-left shunt.

As a persistent LSVC courses laterally to the aortic arch, it may be mistaken for left upper lobe partial anomalous pulmonary venous return (see PAPVR section).

Azygos/Hemiazygos continuation of the Inferior Vena Cava

The azygos continuation of the inferior vena cava (IVC), also named absence of the hepatic segment of the IVC with azygos continuation, is a relatively common thoracic venous anomaly, with a prevalence of 0.6% amongst the general population.
This anomaly results from failure to form the right subcardinal-hepatic anastomosis, with atrophy of the suprarenal part of the right subcardinal vein.

With interruption of the intrahepatic portion of the IVC, blood is directed to a large azygos vein (which ascends along the right margin of the vertebral bodies) into the SVC (Fig. 5 on page 11), with the hepatic veins draining to the right atrium through the suprahepatic IVC.

The hemiazygos continuation of IVC is a less frequent abnormality, with three possible paths of drainage being described:

- Through the azygos vein;
- Into a persistent LSVC;
- Into a normal right SVC (coursing through the accessory hemiazygos vein, left superior intercostal vein and left brachiocephalic vein).

The dilated hemiazygos vein is seen along the left margin of the spine.

Both azygos and hemiazygos continuation of the IVC may be associated with other malformations, such as heterotaxic syndromes and persistent left SVC.

2) Pulmonary venous anomalies

Anatomy and Embryology

In the human embryo, the primordial lung bud initially drains into the cardinal venous system, without having any connection with the heart (Fig. 6 on page 12 a). By days 32-33, the common pulmonary vein arises from the left atrium, establishing a connection with the pulmonary vascular bed. Once a direct connection with the heart is established, connections to the cardinal venous system begin to involute, with blood draining from the developing lung to the common pulmonary vein (Fig. 6 on page 12 b) and then to the left atrium through four individual pulmonary veins. Finally, the common pulmonary vein incorporates into the left atrium and the four pulmonary veins connect directly to the heart (Fig. 7 on page 13).

In normal conditions, four pulmonary veins carrying oxygenated blood drain into the left atrium. The right superior pulmonary vein drains the right upper and middle lobes, the left superior pulmonary vein drains the left upper lobe and lingula, and the right and left inferior pulmonary veins drain the lower lobes (Fig. 7 on page 13).
Partial anomalous pulmonary venous return

Partial anomalous pulmonary venous return (PAPVR) has a prevalence of 0.4-0.7% in the general population, with prevalence abruptly rising in patients with atrial septal defects (10-15% in ostium secundum type and nearly 100% in sinus venosus type).

PAPVR arises when, during the embryologic development, one or more (but not all) of the pulmonary veins maintain systemic venous connection, instead of connecting to the common pulmonary vein. As a result, one or more pulmonary veins will drain to a location other than the left atrium - to the systemic veins (SVC, IVC, SCV, BCV, azygos vein), right atrium or coronary sinus.

This anomaly results in left-to-right shunt, with partial admixture of deoxygenated and oxygenated blood. However this shunt is usually hemodynamically insignificant, with most patients being asymptomatic or mildly symptomatic. As such, it is commonly detected incidentally on imaging studies.

As the above described systemic thoracic anomalies, PAPVR may be associated with other congenital heart diseases, namely atrial, ventricular or atrioventricular septal defects, tetralogy of Fallot and aortic coarctation.

Three types of PAPVR are described, with right-side PAPVR being more frequent than the left.

1. Right upper lobe venous drainage into the low SVC or superior cavoatrial junction (Fig. 8 on page 13, Fig. 9 on page 14, Fig. 10 on page 15, Fig. 11 on page 16)

   - The most frequent type of PAPVR.
   - When this type of PAPVR is diagnosed, a sinus venosus type of atrial septal defect must be sought, as it is present in up to 90% of patients with PAPVR of the right upper lobe. This diagnosis is vital, not only because it results in higher probability of hemodynamically significant shunting, but also because an undiagnosed atrial septal defect predisposes the patient to paradoxical emboli.

2. Left pulmonary venous drainage to the LBCV (Fig. 12 on page 16)

   - Most common type of PAPVR in patients without heart disease.
   - May simulate the appearance of a LSVC. These two entities may be distinguishable by the following features:
• LSVC can be followed inferiorly to the coronary sinus (which is usually dilated); In PAPVR the intraparenchymal upper lobe vessels can be seen to connect with the anomalous vein.
• In patients with LSVC two vessels are seen anterior to the left main bronchus - the normal left superior pulmonary vein and the LSVC. In patients with PAPVR no vessel is seen anterior to the bronchus.
• LSVC conduits blood caudally from the left subclavian and jugular veins into the right atrium; In PAPVR the abnormal vein conducts blood cranially from the left upper lobe to the LBCV.

3. Anomalous drainage from the right lung to the IVC with an intact atrial septum (Fig. 13 on page 17)

• This type of PAPVR may be associated with complex pulmonary anomalies in the spectrum of congenital pulmonary venolobar syndrome.

Scimitar syndrome (Fig. 13 on page 17)

Scimitar syndrome, also named venolobar syndrome, is a rare form of PAPVR, almost always involving the right lung. It consists of middle or lower lobe anomalous drainage to the systemic veins, associated with hypogenetic lung (congenital anomaly combining pulmonary hypoplasia with decreased vascularity on the involved side and cardiac dextroposition).

Though the anomalous vein most frequently drains to the IVC below the right hemidiaphragm, it may rarely drain into the suprahepatic portion of the IVC, to the hepatic veins, to the portal vein, to the azygos veins, to the coronary sinus or to the right atrium.

Associated anomalies include congenital heart disease, bronchogenic cysts, horseshoe lung, pulmonary arteriovenous malformations, accessory diaphragm and congenital diaphragmatic hernia.

The designation of Scimitar syndrome derives from the curvilinear appearance of the pulmonary vein draining to the IVC, as this appearance resembles a scimitar, a curved turkish sword.
Fig. 1: Normal anatomy of the systemic thoracic veins, illustrated at gadolinium-enhanced 3D MR angiography with coronal maximum-intensity projection (MIP) image. SVC = superior vena cava; IVC = inferior vena cava; RBCV = right brachiocephalic vein; LBCV = left brachiocephalic vein.

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Fig. 2: Subsequent stages of the embryological development of the systemic thoracic veins.

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**Fig. 3:** Normal anatomy of the azygos venous system, illustrated at gadolinium-enhanced 3D MR angiography with coronal (a) and sagittal (c) maximum-intensity projection (MIP) images. SVC = superior vena cava.

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**Fig. 4:** Volume rendered (VR) contrast-enhanced MR angiographic image (a) shows persistent left superior vena cava (LSVC), with presence of the right superior vena cava (RSVC). Gadolinium-enhanced 3D MR angiography coronal image (b) shows persistent LSVC with absence of the RSVC.

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Fig. 5: Azygos continuation of the inferior vena cava in a patient with corrected transposition of the great arteries, illustrated by a true fast imaging with steady-state precession (true FISP) 4 chamber-view image (a) and multiplanar reconstructed (MPR) image from MR angiographic data (b).

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Fig. 6: Subsequent stages of the embryological development of the pulmonary veins. RA = right atrium; LA = left atrium.

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Fig. 7: Normal anatomy of the pulmonary veins, illustrated at VR contrast-enhanced MR angiographic image (posterior view). RSPV = right superior pulmonary vein; LSPV = left superior pulmonary vein; RIPV = right inferior pulmonary vein; LIPV = left inferior pulmonary vein; LA = left atrium.

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**Fig. 8:** Anomalous pulmonary venous drainage of the right superior pulmonary vein (RSPV) into the superior vena cava (SVC), illustrated with gadolinium-enhanced 3D MR angiography coronal image.

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Fig. 9: VR contrast-enhanced MR angiographic images illustrate anomalous pulmonary venous drainage of the right superior pulmonary vein (RSPV) into the right atrium (RA) (a). The right inferior pulmonary vein (RIPV), left superior and left inferior pulmonary veins (LSPV, LIPV) drain into the left atrium (LA), as shown in b) (posterior view). This patient also had a persistent left superior vena cava and aortic coarctation (not shown).

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Fig. 10: VR (a) and MIP (b) contrast-enhanced MR angiographic images illustrate anomalous drainage from the right upper lobe (RUL) and superior segment of the right

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lower lobe (SRLL) into the superior vena cava (SVC). Right inferior pulmonary vein (RIPV), left superior and inferior pulmonary veins (LSPV, LIPV) drain into the left atrium.

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**Fig. 11:** Veno-venous fistula (blue arrows) between the superior vena cava (SVC) and the right superior pulmonary vein (RSPV), which normally drains into the left atrium (LA), illustrated with coronal (a) and axial (b) MIP images from MR angiographic data.

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Fig. 12: VR contrast-enhanced MR angiographic image illustrates anomalous drainage of the left upper lobe pulmonary vein (LULPV) into the left brachiocephalic vein (LBCV), and then into the superior vena cava (SVC).

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**Fig. 13:** Coronal half-fourier acquisition single-shot turbo spin-echo (HASTE) image and MIP contrast-enhanced MR angiographic image illustrate anomalous pulmonary venous drainage (blue arrows) of the right lung into the inferior vena cava (IVC), with hypoplasia of the right lung (RL) and of the right pulmonary artery (RPA). LPA = Left pulmonary artery.

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Conclusion

Though congenital anomalies of the thoracic veins are rare, they represent important developmental abnormalities. Knowledge of the embryological development and of the normal anatomy of the thoracic veins is paramount to understand and recognize congenital thoracic venous anomalies.


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