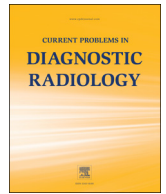




Current Problems in Diagnostic Radiology

journal homepage: www.cprjournal.com



Congenital Thoracic Venous Anomalies in Adults: Morphologic MR Imaging

Carina A. Ruano, MD^{a,*}, António Marinho-da-Silva, MD^b, Paulo Donato, PhD^{c,d}

^a Department of Radiology, Central Lisbon Hospital Centre, Lisbon, Portugal

^b Department of Cardiology, Coimbra Hospital and University Centre, Coimbra, Portugal

^c Department of Radiology, Coimbra Hospital and University Centre, Coimbra, Portugal

^d Faculty of Medicine, University of Coimbra, Coimbra, Portugal

Congenital anomalies of the thoracic veins are rare yet important developmental abnormalities, usually classified into systemic and pulmonary. They may be encountered incidentally; as such the radiologist must be aware of their imaging presentation and clinical relevance. Furthermore, to understand these anomalies, knowledge of the embryological development and of the normal anatomy of the thoracic veins is required. In the age of non-invasive imaging modalities, magnetic resonance is paramount for the characterization of these developmental abnormalities.

© 2015 Mosby, Inc. All rights reserved.

Introduction

Congenital anomalies of the thoracic veins may go unnoticed until adulthood, when they are diagnosed incidentally as part of an examination for a totally unrelated indication. Despite being frequently asymptomatic, these abnormalities require accurate characterization as they may be associated with increased morbidity and mortality and have important clinical or surgical implications.

Patients with congenital anomalies of the thoracic veins can undergo evaluation using catheter angiography, echocardiography, computed tomography (CT), or magnetic resonance (MR) imaging (MRI) and angiography (MRA). Though catheter angiography has played a major role in the characterization of these disorders in the past, it is currently reserved for endovascular interventions because of its invasiveness. Echocardiography, despite allowing a direct and noninvasive evaluation, is limited by a reduced field of view, which may prevent a complete evaluation. Although CT has excellent spatial resolution and anatomical definition, it has the disadvantage of using ionizing radiation and iodinated contrast medium. Conversely, MRI (and MRA) is an excellent imaging technique to characterize these anomalies, as it is a noninvasive method with high inherent soft tissue contrast that does not require ionizing radiation or iodinated contrast medium. Furthermore, it is capable of multiplanar imaging and acquiring multiple angiographic phases with a single intravenous bolus of gadolinium.

This article reviews the embryogenesis and normal imaging anatomy of the thoracic veins and illustrates through MRI the most common congenital thoracic venous anomalies (of both the systemic and pulmonary circulation) that can be diagnosed in the adult population. Congenital anomalies specific to the pediatric

population (namely total anomalous pulmonary venous return, which typically presents in the neonatal period) have not been discussed as they evade the focus of the article (Table).

MR Imaging

Role of MRI in Diagnostic Workup

Congenital anomalies of the thoracic veins are frequently diagnosed on echocardiography or CT scans. CT is usually performed in asymptomatic patients who present with an abnormal mediastinal contour at chest radiography. Not all of these patients require MR evaluation. Whether to proceed to MRI or MRA depends on the following factors: (1) if the patient is symptomatic or asymptomatic; (2) if the full extent of the abnormality is adequately evaluated by the initial study; (3) if the anomaly is commonly associated with other congenital abnormalities (namely congenital heart disease), and (4) if such potential anomalies can or cannot be documented on the initial study.¹

MRI is particularly valuable in patients in whom the use of iodinated intravenous contrast material is contraindicated. Other advantages of this technique include the lack of ionizing radiation (particularly useful in the evaluation of young adults) and the ability to characterize the thoracic vasculature and cardiac morphology and function with noncontrast techniques (invaluable in patients at risk for nephrogenic systemic fibrosis and with allergy to gadolinium). Furthermore, MRI allows simultaneous evaluation of vascular and cardiac developmental abnormalities, which frequently coexist in the same patient.^{1,2}

MRI and MRA Technique

A typical MRI-MRA protocol for the evaluation of congenital thoracic vascular abnormalities includes electrocardiogram-gated

* Reprint requests: Carina Alexandra Santos Ruano, MD, Department of Radiology, Hospital de Santo António dos Capuchos, Alameda de Santo António dos Capuchos, 1169-050 Lisboa, Portugal.

E-mail address: carinaasr@gmail.com (C.A. Ruano).

TABLE
Congenital thoracic venous anomalies

Congenital thoracic venous anomalies
Systemic venous anomalies
Persistent left superior vena cava (LSVC) Single LSVC terminating in the coronary sinus Single LSVC terminating in the left atrium Duplicated SVC connected by the brachiocephalic vein Duplicated SVC not connected by the brachiocephalic vein
Anomalies of the right superior vena cava (SVC) Absent right SVC with persistent LSVC Right SVC connecting to the left atrium Anomalous low insertion of the SVC Aneurysmatic dilatation of the SVC
Anomalies of the brachiocephalic vein (BCV) Retroaortic left BCV
Anomalies of the inferior vena cava (IVC) Azygos continuation of the IVC Hemiazygos continuation of the IVC Anomalous connection of the IVC to the coronary sinus Anomalous connection of the IVC to the left atrium Anomalous high insertion of the IVC to the right atrium
Anomalies of the azygos vein Absent azygos vein Azygos lobe
Pulmonary venous anomalies
Partial anomalous pulmonary venous return (PAPVR) Right veins (to the SVC, right atrium, or azygos vein) ⁺ Left veins (to the left brachiocephalic vein or coronary sinus) Right veins to the IVC (scimitar syndrome)
Total anomalous pulmonary venous return (TAPVR) [*] Supracardiac [*] Intracardiac [*] Infracardiac [*] Mixed [*]

Anomalies exclusive to and more frequently seen in the pediatric population are highlighted with * and +, respectively.

black-blood and bright-blood sequences in addition to an angiographic sequence.³

Both black-blood and bright-blood images can be obtained without intravenous administration of contrast material. Black-blood images (typically acquired with single-shot fast spin echo with double inversion recovery or half-Fourier single-shot turbo spin echo sequences) are useful to study the anatomy of the heart and mediastinum; cine bright-blood images (usually obtained using a 2-dimensional (2D) or 3D balanced steady-state free precession sequence) allow the assessment of the cardiac chambers and valvular function.^{3,4}

Angiographic techniques include 3D T1-weighted contrast-enhanced MRA, time of flight MRA, and phase contrast MRA, the latter 2 providing angiographic information in patients in whom intravenous administration of gadolinium is contraindicated. Gadolinium-enhanced MRA (non-electrocardiogram-gated 3D spoiled gradient recalled echo imaging technique) is extremely valuable in the assessment of the thoracic vascular structures, allowing dynamic imaging during the arterial and venous phases, of both the systemic and pulmonary circulation. Phase contrast MRA, also called velocity-encoded sequence, evaluates flow direction and velocity, allowing the quantification of shunts.²⁻⁴

Our standard protocol for evaluating thoracic venous anomalies consists of axial, sagittal, and coronal half-Fourier single-shot turbo spin echo sequences, steady-state free precession sequences to evaluate ventricular function, and 3D contrast-enhanced MRA. Contrast-enhanced MRA image is obtained after intravenous

injection of a double dose (0.20 mmol/kg of body weight) of gadolinium-containing contrast media, at an injection rate of 2 mL/s; the acquisition is initiated manually using care bolus tracking technique. The angiographic images are acquired in a sagittal or coronal plane, and subsequently reformatted in multiple planes and reconstructed as maximum intensity projection or volume-rendered images. 2D maximum intensity projection and 3D volume-rendered images provide excellent spatial detail; structural detail is better assessed with 2D multiplanar reconstructed images.

The radiologist should be familiarized with the patient's medical history and prior imaging studies as to better select the most appropriate imaging protocol. In our institution, the radiologists themselves perform the postprocessing techniques. Though time consuming, this stage is paramount for a complete understanding of the vascular system. Postprocessed images are also extremely useful to clinicians, especially when considering surgical procedures.

The MRI report should include information on vascular morphology (detailed description of the anomalous vessels and relation to relevant vascular structures), hemodynamic information (when relevant), and description of associated cardiac and thoracic abnormalities (when present).

Systemic Thoracic Venous Anomalies

Superior Vena Cava and Brachiocephalic Vein

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).

Embryologically, these veins derive from the paired anterior and common cardinal veins (with the common cardinal veins resulting from the union of the anterior and posterior cardinal veins, before entering the sinus venosus). By the seventh week of gestation, the anterior cardinal veins become connected by an anastomosis, which shunts blood from the left to right (Fig 2A).

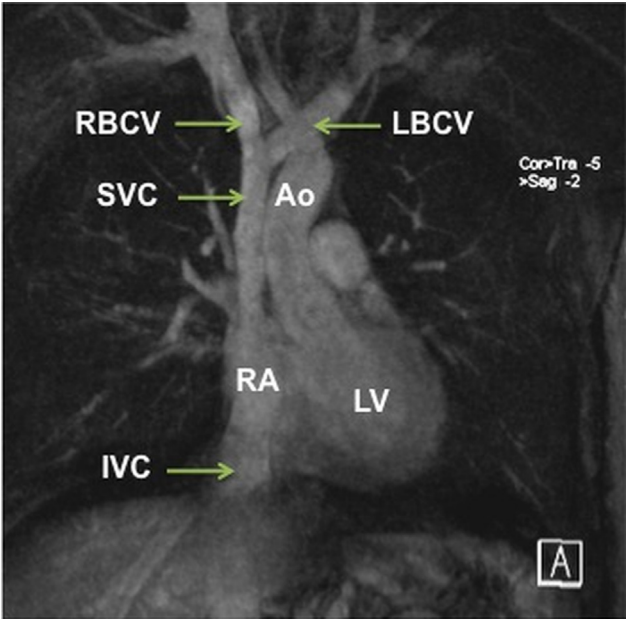


FIG 1. Normal anatomy of the systemic thoracic veins, illustrated with coronal gadolinium-enhanced 3D MRA image. Ao, aorta; IVC, inferior vena cava; LBCV, left brachiocephalic vein; LV, left ventricle; RA, right atrium; RBCV, right brachiocephalic vein; SVC, superior vena cava.

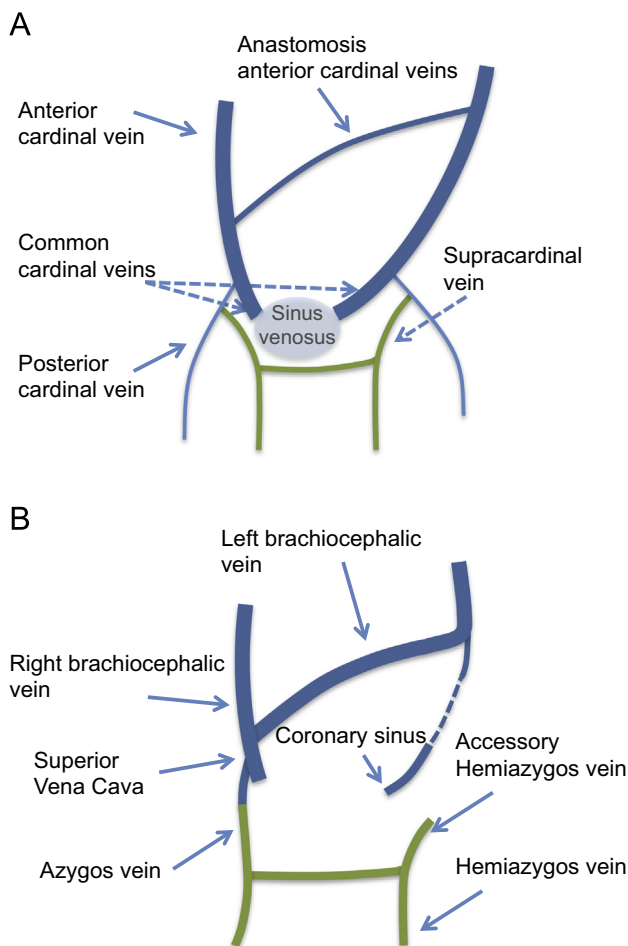


FIG 2. Embryological development of the systemic thoracic veins: (A) seventh week of gestation and (B) at term. Initially, the cardinal veins form the main venous drainage system of the embryo. The anterior cardinal veins (which drain the cephalic part of the embryo) and the posterior cardinal veins (which drain the rest of the embryo) join before entering the sinus venosus, forming the common cardinal veins. By the seventh week of gestation, the anterior cardinal veins become connected by an anastomosis, which posteriorly develops to be the LBCV. The right cardinal system originates the SVC, RBCV, and right internal jugular vein. The left anterior cardinal vein undergoes near-complete atrophy below the level of the anastomosis (forming the ligament of the left vena cava) and persists above this level as left SVC and internal jugular vein. The left common cardinal vein becomes the oblique vein of the left atrium, draining into the coronary sinus. RBCV, right brachiocephalic vein. Adapted from Sadler.⁶

Subsequently the right cardinal system develops into the SVC, right BCV, and right internal jugular vein. The left anterior cardinal vein undergoes near-complete atrophy below the level of the anastomosis, persisting above this level to originate the internal jugular vein and the left SCV. The left common cardinal vein becomes the oblique vein of the left atrium (vein of Marshall). The anastomosis between the anterior cardinal veins originates the left BCV (LBCV) (Fig 2B).^{3,5,6}

Anomalies

Persistent Left SVC. A persistent left SVC (LSVC) is described in 0.3% of the general population, with prevalence increasing to 4.3% in patients with congenital cardiac abnormalities.^{7,8} The commonly associated cardiovascular anomalies include septal defects, aortic coarctation, and anomalous pulmonary venous return.⁵ This anomaly is frequently discovered incidentally in asymptomatic patients. However, it is a relevant anatomical finding that requires clear documentation, as it can interfere in the placement of several devices such as central venous catheters, cardiac pacemaker, and

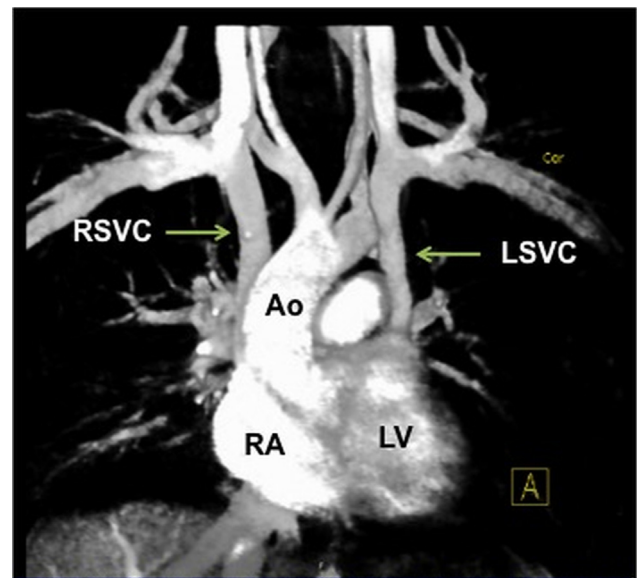


FIG 3. Persistent left superior vena cava (LSVC), with presence of the right superior vena cava (RSVC), illustrated with maximum intensity projection (MIP) image from MR angiographic data. Ao, aorta; LV, left ventricle; RA, right atrium.

defibrillator leads. It may also have surgical implications, namely in coronary artery bypass surgery.^{7,8}

Embryologically, this anomaly results from persistence of the left anterior cardinal vein. In most cases (82%–90%), a right SVC (usually of reduced caliber) is also present, resulting in a duplicated SVC (Fig 3). An anastomosis between the right and left SVC occurs in 25%–35% of these patients.⁹ In the remaining cases, involution of the right common cardinal vein and the central anterior cardinal vein, along with persistence of the left anterior cardinal vein and of the anastomosis between the right and left cardinal veins, results in absence of the right SVC with a mirror image of the thoracic systemic venous drainage (Fig 4).³

In most patients (92%), the LSVC drains into the right atrium through the coronary sinus, which enlarges to accommodate the blood flow (Figs 4, 5).^{7,9} Enlargement of the coronary sinus may potentially lead to left atrioventricular valve inflow obstruction,

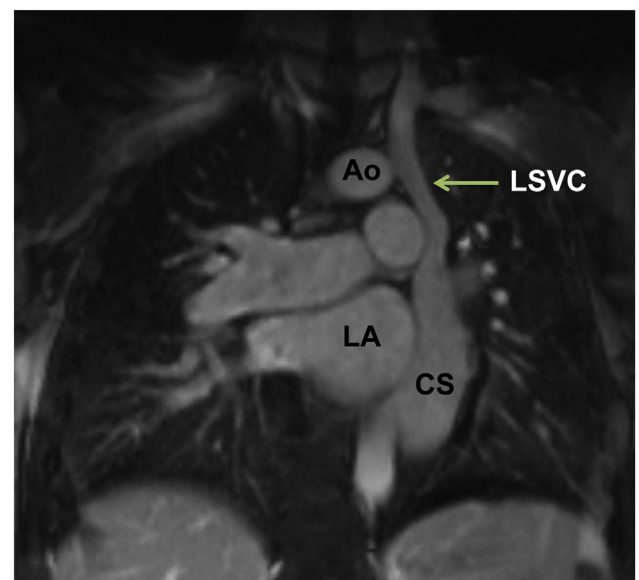


FIG 4. Persistent left superior vena cava (LSVC), draining to an enlarged coronary sinus (CS), with absence of the right superior vena cava, illustrated with steadystate free precession (SSFP) image in an oblique coronal plane. Ao, aorta; LA, left atrium.

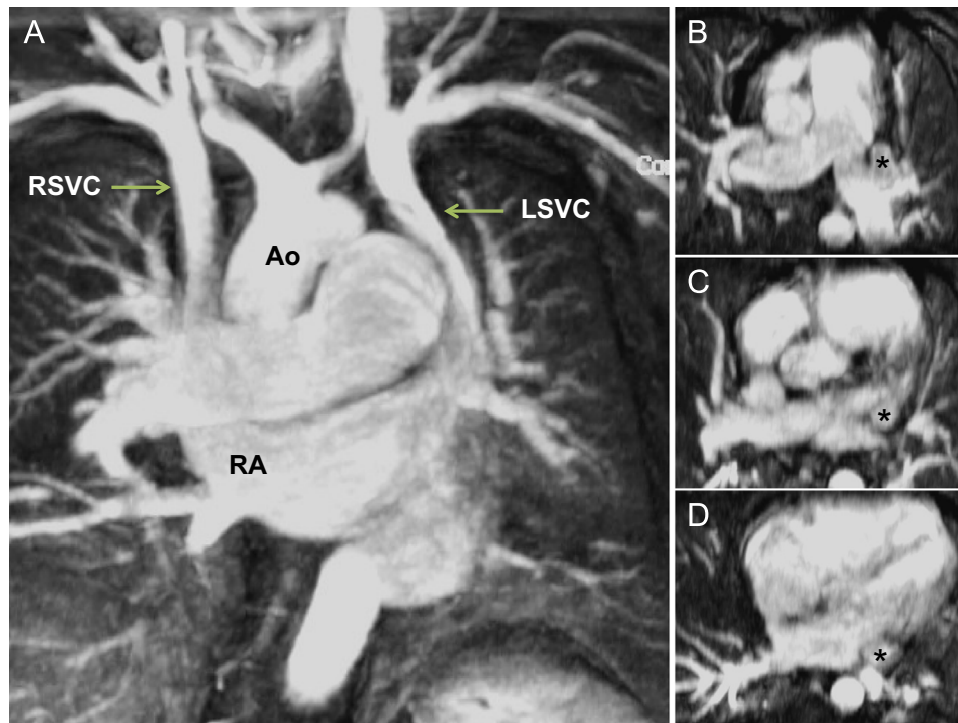


FIG 5. Persistent left superior vena cava (LSVC) with presence of the right superior vena cava (RSVC), illustrated with coronal (A) and axial (B–D) MIP images from MR angiographic data, showing the LSVC draining into an enlarged coronary sinus (*). Ao = aorta; RA, right atrium.

and subsequently to cardiac arrhythmias or sudden death.³ In the remaining cases, the LSVC terminates in the left atrium, resulting in a right to left shunt.⁷ The risk of congenital heart disease in such patients is considerably increased.¹

Anomalies of the Right SVC. Intrinsic anomalies of the right SVC are rare and consist of anomalous drainage of the SVC to the left atrium, low insertion of the SVC into the right atrium, and

aneurysmatic dilatation of the SVC. Drainage of the SVC into the left atrium results in a right to left shunt and can occur without any associated anomaly. Contrarily, a low insertion of the SVC into the right atrium is associated with complex congenital heart disease. An aneurysmatic dilatation of the SVC is frequently an incidental finding. However, this anomaly requires precise documentation as it can mimic a mediastinal mass in radiography and may be associated with increased risk of thrombosis, embolization, and SVC obstruction.^{10,11}

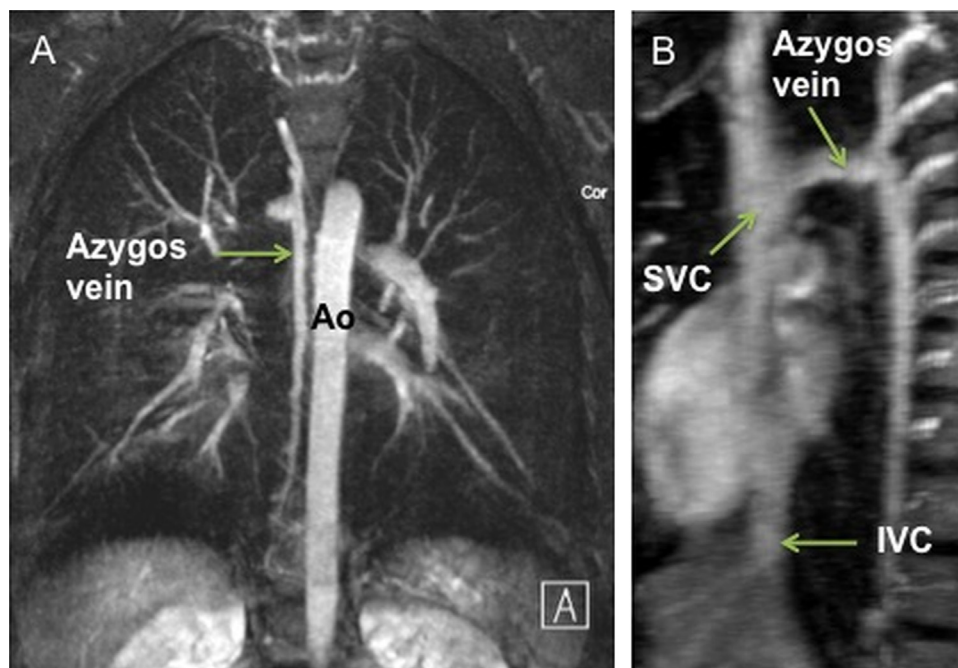


FIG 6. Normal anatomy of the azygos vein, illustrated with coronal (A) and sagittal (B) gadolinium-enhanced 3D MRA images. Ao, aorta; IVC, inferior vena cava; SVC, superior vena cava.

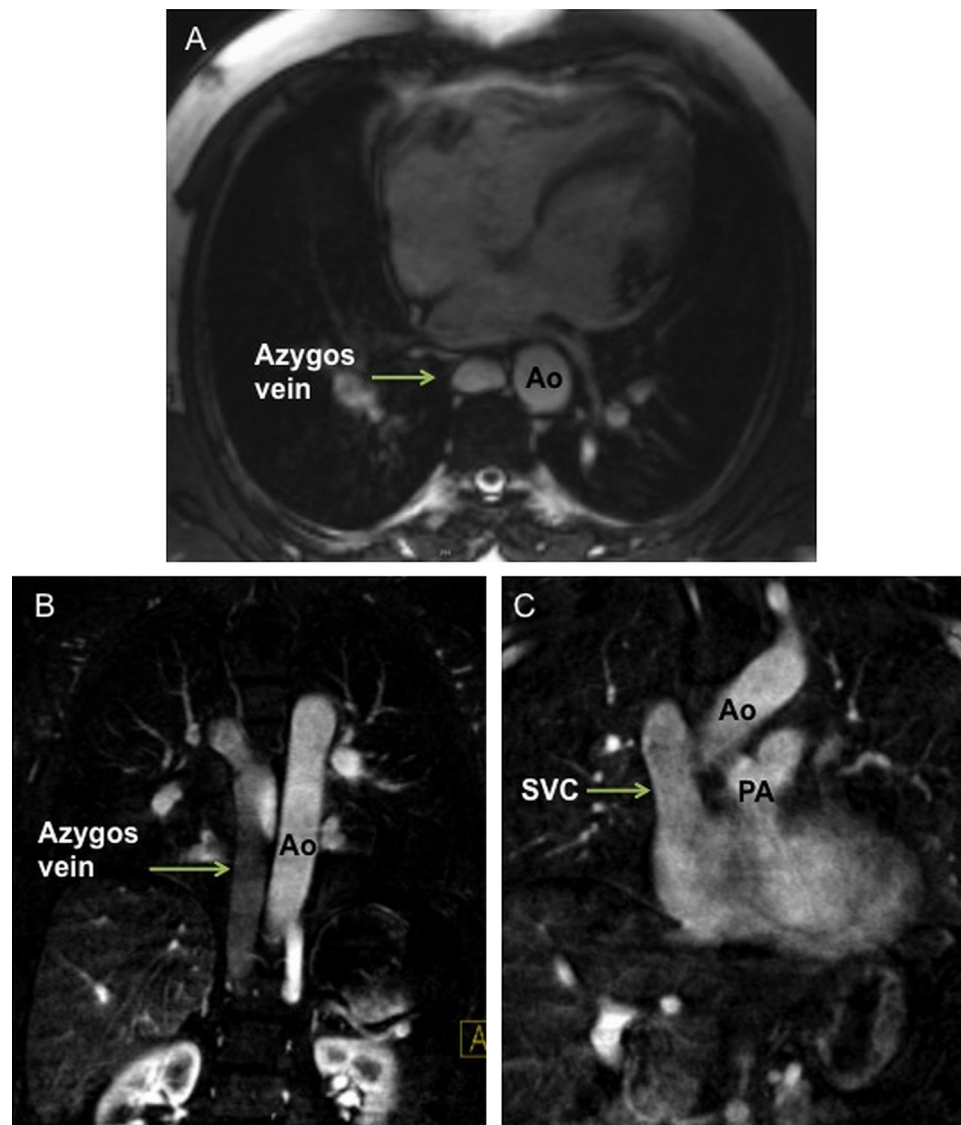


FIG 7. Azygos continuation of the inferior vena cava illustrated with 4-chamber view SSFP image (A) and coronal multiplanar reconstructed (MPR) images from MR angiographic data (B and C). (A and B) Enlarged azygos vein. (C) Absence of the hepatic segment of the IVC in a patient with corrected transposition of the great arteries. Ao, aorta; PA, pulmonary artery; SVC, superior vena cava.

Anomalies of the BCV. A retroaortic LBCV is usually seen in patients with congenital heart disease (prevalence of 0.5%-0.6%), being rarely described in patients without it (prevalence of 0.02%). Although a retroaortic LBCV alone does not cause physiologic or hemodynamic modifications, it is relevant because of the associated congenital heart anomalies (namely tetralogy of Fallot, pulmonary atresia, and truncus arteriosus) and its clinical and surgical implications (for instance, in the insertion of central venous pressure lines and pacemakers).^{3,12}

The precise embryology of this anomaly remains unknown, with 3 major hypotheses having been proposed: (1) the existence of 2 precardinal anastomoses, with regression of the upper one; (2) obstruction of the normal course of the LBCV with development of an alternative channel; (3) development of the precardinal anastomosis in any available pathway after the development of the aortic arch (this theory being supported by a known association with anomalies of the aortic arch and pulmonary artery, namely right-sided aortic arch and pulmonary trunk atresia).¹²

While the normal LBCV joins the right BCV anteriorly to the aorta, the retroaortic LBCV courses posteriorly to the ascending aorta, underneath the aortic arch, and anterior to the central main

and right pulmonary arteries, to join the RSVC caudal to the azygos vein. In isolated cross-sectional images, this anomaly may resemble a persistent LSVC or partial anomalous pulmonary venous return (PAPVR); however, tracing the vessel through sequential images provides the correct diagnosis.^{10,12}

Inferior Vena Cava and Azygos System

Anatomy and Embryology

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. 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 intrahepatic segment of the IVC.¹³

The azygos venous system initially derives from the paired supracardinal veins (Fig 2A). The azygos vein originates from the anastomosis between the cranial segments of both the right supracardinal and the posterior cardinal veins. The hemiazygos

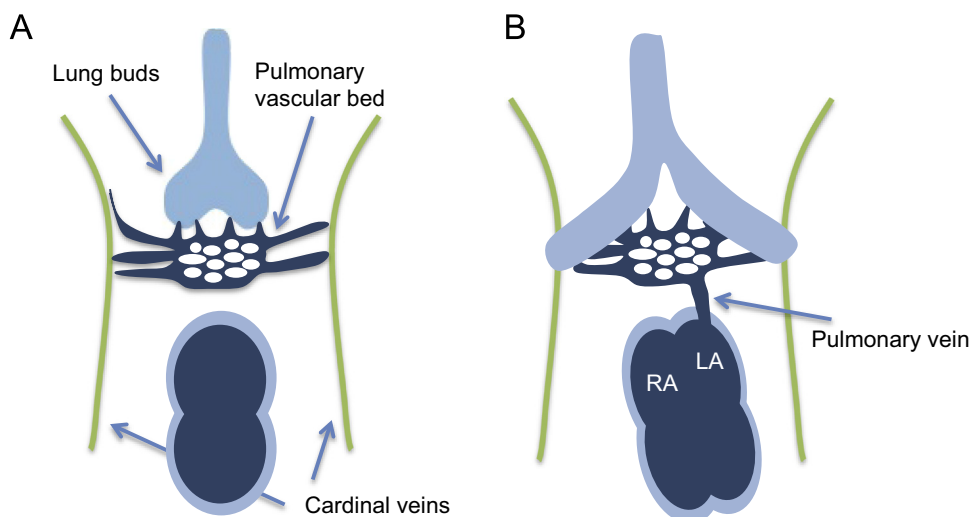


FIG 8. Embryological development of the pulmonary veins. Before the fifth week of gestation, the lung buds drain via a venous plexus into the cardinal veins (A). The common pulmonary vein arises from the left atrium, and at approximately the 28th day of gestation, it establishes a connection with the venous plexus (B). Connections to the systemic venous circulation begin to involute afterwards. LA, left atrium; RA, right atrium. Adapted from Sears et al.¹⁸

vein originates from the cranial part of the left supracardinal vein (Fig 2B). The hemiazygos vein (located on the left side of the spine) drains into the azygos vein (along the right side of the vertebral bodies), which in turn drains into the SVC (Fig 6).¹⁴

Anomalies

Anomalies of the IVC. The azygos continuation of the IVC (Fig 7), also described as the absence of the hepatic segment of the IVC with azygos continuation, occurs in 0.6% of the general population and results from failure to form the right subcardinal–hepatic anastomosis, with subsequent 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 into the SVC, with the hepatic veins draining to the right atrium through the suprahepatic IVC.¹³

The hemiazygos continuation of the IVC is a less frequently seen abnormality, with 3 possible paths of drainage being described: (1) through the azygos vein; (2) into a persistent LSV; and (3) into a normal right SVC (coursing through the accessory hemiazygos vein and LBCV).¹⁴

Although previously thought to be associated with severe congenital heart disease and heterotaxy syndrome,¹⁵ both these anomalies are being increasingly documented in asymptomatic patients since the advent of cross-sectional imaging. Awareness of these anomalies avoids misdiagnosing enlarged azygos or hemiazygos veins for retrocrural adenopathy.¹³

Other anomalies of the IVC are rarer than those cited previously and include anomalous connection of the IVC to the left atrium or coronary sinus and a high insertion of the IVC into the right atrium.¹

Anomalies of the Azygos Vein. Congenital absence of the azygos vein is uncommon, with very few cases reported in the literature. Imaging studies of patients with this anomaly demonstrate absence of the azygos vein, and subsequent enlargement of the hemiazygos, accessory hemiazygos, and left superior intercostal

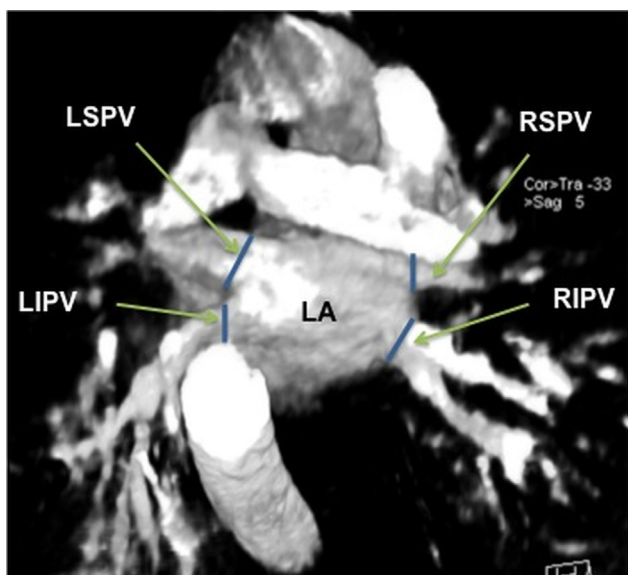


FIG 9. Normal anatomy of the pulmonary veins, illustrated with volume-rendered (VR) contrast-enhanced MRA, posterior view. LA, left atrium; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein.

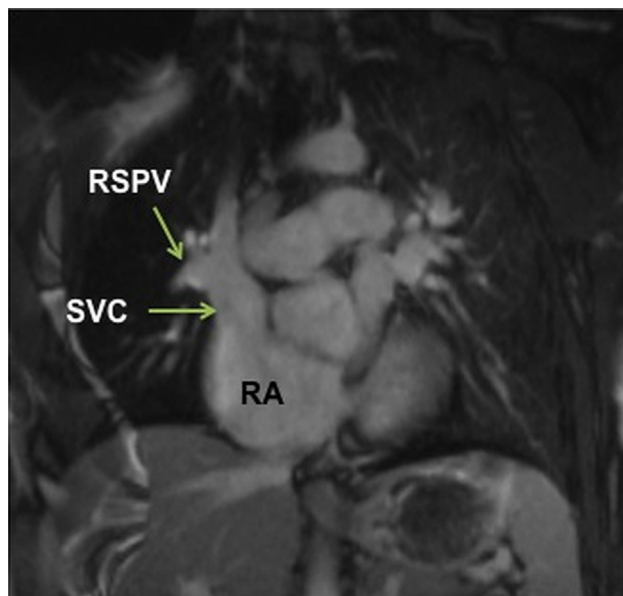


FIG 10. Anomalous pulmonary venous drainage of the right superior pulmonary vein (RSPV) into the superior vena cava (SVC), illustrated with SSFP image in an oblique coronal plane. RA, right atrium.

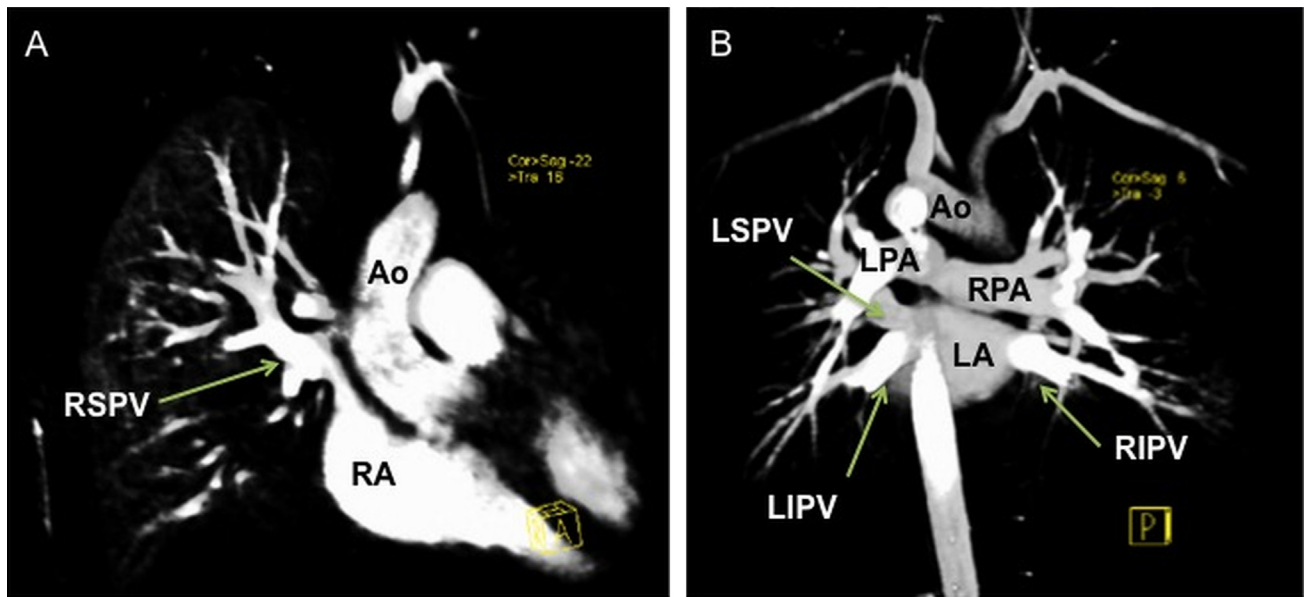


FIG 11. Anomalous pulmonary venous drainage of the right superior pulmonary vein (RSPV) into the right atrium (RA) (A), illustrated with MIP images from MR angiographic data. The right inferior pulmonary vein (RIPV) and left superior and left inferior pulmonary veins (LSPV and LIPV) drain into the left atrium (LA), as shown in (B) (posterior view). Ao, aorta; LPA, left pulmonary artery; RPA, right pulmonary artery.

veins. These enlarged vessels should not be misinterpreted as lymphadenopathy.^{14,16}

Pulmonary Venous Anomalies

Anatomy and Embryology

The lung buds originate from the ventral wall of the foregut at the fourth week of embryological development and initially drain into the systemic veins without having any connection with the heart (Fig 8A). Subsequently, the common pulmonary vein arises

from the left atrium, and at approximately day 28 of gestation, it establishes a connection with the pulmonary vascular bed. Once a direct connection with the heart is established, connections to the systemic veins begin to involute. Blood then drains from the developing lung into the common pulmonary vein (Fig 8B), which is then divided into 4 individual pulmonary veins and integrated in the left atrium.^{2,6,17}

In normal conditions, 4 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 9).¹⁹

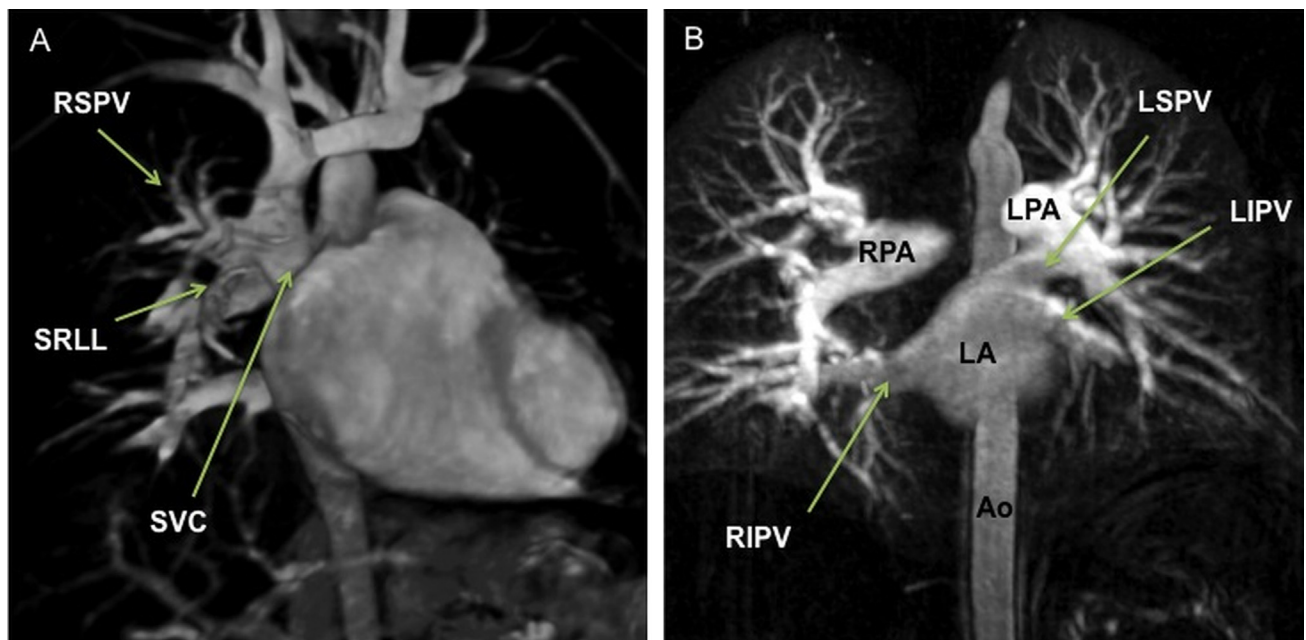


FIG 12. Anomalous drainage of the right superior pulmonary vein (RSPV) and of the superior vein of the right lower lobe (SRLL) into the superior vena cava (SVC), illustrated with VR (A) and MIP (B) contrast-enhanced MRA images. The right inferior pulmonary vein (RIPV) and left superior and inferior pulmonary veins (LSPV and LIPV) drain into the left atrium. Ao, aorta; LA, left atrium; LPA, left pulmonary artery; MIP, maximum intensity projection; RPA, right pulmonary artery; VR, volume-rendered.

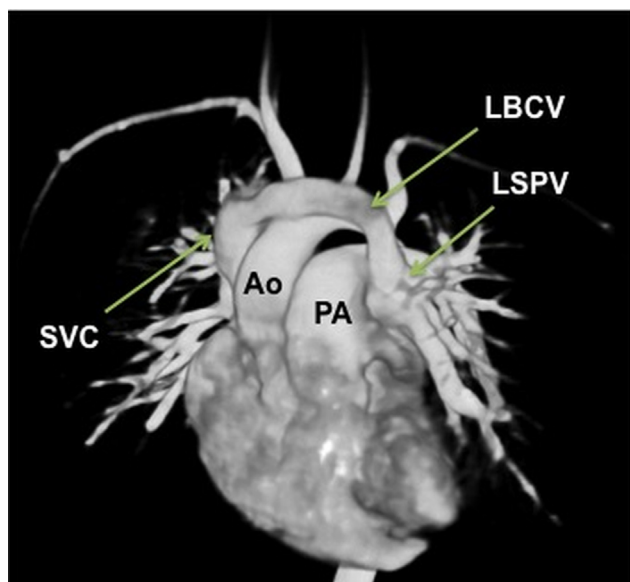


FIG 13. Anomalous drainage of the left superior pulmonary vein (LSPV) into the left brachiocephalic vein (LBCV), and then into the superior vena cava (SVC), illustrated with VR contrast-enhanced MRA image. Ao, aorta; PA, pulmonary artery.

Anomalies

Partial Anomalous Pulmonary Venous Return

PAPVR has a prevalence of 0.4%–0.7% in the general population.^{19,20} While pediatric patients with PAPVR are usually symptomatic, leading to an early diagnosis, asymptomatic patients usually remain undiagnosed until adulthood.²

PAPVR arises when, during the embryological development, one or more (but not all) of the pulmonary veins maintain systemic venous connection instead of connecting to the common pulmonary vein.²⁰ As a result, one or more pulmonary veins drain into a location other than the left atrium, that is, to the systemic veins (SVC, IVC, SCV, BCV, and azygos vein), right atrium, or coronary sinus.³

This anomaly results in a left to right shunt, with cardiopulmonary signs and symptoms depending on the magnitude of the shunt, on the hemodynamic changes created by the shunt, and on the presence of associated cardiac anomalies. The shunt is thought to become clinically significant when 50% or more of the pulmonary blood flow returns anomalously.⁴

The incidental detection of PAPVR requires detailed anatomical description of the anomaly, namely in patients undergoing lobectomy for a pulmonary neoplasm. When the anomalous vessel is located in a lobe other than the one affected by the neoplasm, some authors suggest that the PAPVR should be corrected during lobectomy to prevent right-sided heart failure caused by increased blood flow through the aberrant vein.^{21–23}

Right upper lobe PAPVR (Figs 10–12) is reported more frequently in children and consists of anomalous pulmonary drainage into the SVC, azygos vein, or the right atrium. A sinus venosus type of atrial septal defect must be sought whenever this type of PAPVR is diagnosed, as it is described in 80%–90% of patients with right-sided PAPVR.² This diagnosis is vital not only because it results in higher probability of hemodynamically significant shunting,²⁴ but also because an undiagnosed atrial septal defect may predispose the patient to paradoxical emboli.²⁵

Left pulmonary venous drainage to the LBCV (Fig 13) is frequently an incidental finding in adults without congenital heart disease.¹⁹ It consists of an aberrant vertical vessel that conducts blood in a cephalic direction, from the left upper lobe to the LBCV.² This anomalous vessel may simulate the appearance of a LSVC. These entities can be easily differentiated on cross-sectional imaging by the following features: (1) the LSVC can be followed inferiorly to the coronary sinus (which is usually dilated); in PAPVR, the intraparenchymal upper lobe vessels connect with the anomalous vein; (2) in patients with LSVC, 2 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; (3) LSVC conducts 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.^{2,24,26}

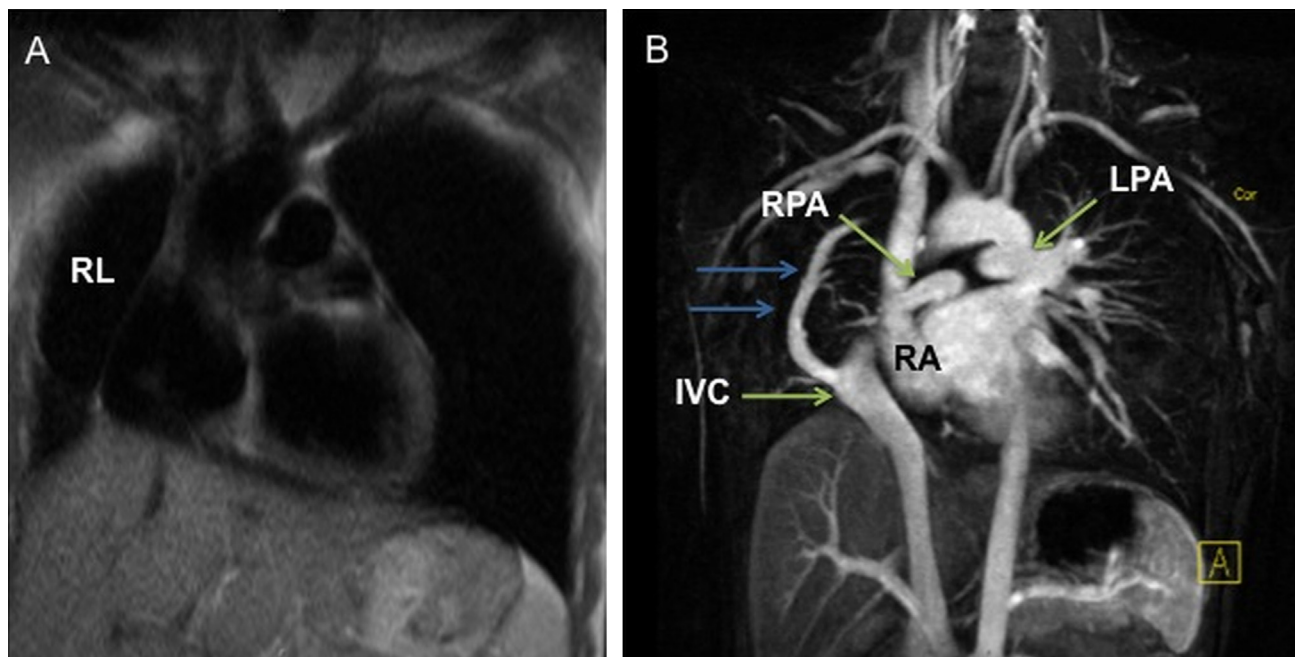


FIG 14. Scimitar syndrome, illustrated with coronal half-Fourier-acquisition single-shot turbo spin echo (HASTE) image (A) and gadolinium-enhanced 3D MRA image (B). Note the anomalous crescent-shaped vessel (blue arrows) draining the right lung into the inferior vena cava (IVC) and the hypoplasia of the right lung (RL) and the right pulmonary artery (RPA). The left pulmonary artery (LPA) has a normal caliber. This patient also has an anomalous high insertion of the IVC into the right atrium (RA).

Scimitar syndrome (Fig 14), also called venolobar or hypogenetic lung syndrome, is a complex and rare form of PAPVR that almost exclusively involves the right lung. It consists of anomalous drainage of a portion or of the entire lung into the systemic veins (most commonly to the infradiaphragmatic IVC), associated with right lung and pulmonary artery hypoplasia or aplasia and cardiac dextroposition.^{24,27} Less commonly, the anomalous vein may drain into the suprahepatic portion of the IVC, hepatic veins, portal vein, azygos veins, coronary sinus, or to the right atrium.²⁷ Systemic arterial blood supply to the right lower lung may also be identified, arising from branches of the abdominal aorta, an abnormality more commonly seen in the infantile form of the syndrome. The right lung may have abnormal lobation with only 2 lobes, mimicking the bronchial pattern of the left lung.² The designation of scimitar syndrome derives from the crescent-shaped abnormal pulmonary vein draining to the IVC, an appearance that resembles a scimitar, a curved Turkish sword.²⁸

Scimitar syndrome is usually divided into 3 main forms: (1) an infantile form with symptoms and pulmonary hypertension, (2) an “older” adult form, which is typically asymptomatic in infancy, and (3) a form with associated congenital cardiac anomalies.²⁹ The clinical presentation and prognosis are therefore variable, depending on the age of presentation, presence or absence of pulmonary arterial hypertension, and associated congenital cardiac abnormalities. While some patients present severe cardiorespiratory symptoms, large left to right shunts or cardiac anomalies requiring surgery, others may be asymptomatic or have only mild symptoms thereby requiring no therapy.¹⁰

Congenital anomalies that may be associated with scimitar syndrome include hypoplastic left heart, aortic coarctation, bronchogenic cysts, bronchopulmonary sequestration, horseshoe lung, pulmonary arteriovenous malformations, accessory diaphragm, and diaphragmatic hernia.^{3,28}

Conclusions

Congenital anomalies of the thoracic veins are rare and frequently silent. However, they may have important clinical implications and be associated with significant patient morbidity and mortality. As such, the radiologist should be aware of their imaging presentation and clinical relevance and should provide an accurate characterization of these abnormalities. For that intent, MRI is an excellent diagnostic modality.

References

- White CS, Baffa JM, Haney PJ, et al. MR imaging of congenital anomalies of the thoracic veins. *Radiographics* 1997;17:595–608.
- Katre R, Burns SK, Murillo H, et al. Anomalous pulmonary venous connections. *Semin Ultrasound CT MR* 2012;33(6):485–99.
- Hellinger JC, Daubert M, Lee EY, et al. Congenital thoracic vascular anomalies: Evaluation with state-of-the-art MR imaging and MDCT. *Radiol Clin North Am* 2011;49(5):969–96.
- Dillman JR, Yarram SG, Hernandez RJ. Imaging of pulmonary venous developmental anomalies. *Am J Roentgenol* 2009;192(5):1272–85.
- Cha EM, Khoury GH. Persistent left superior vena cava. Radiologic and clinical significance. *Radiology* 1972;103(2):375–81.
- Sadler TW. *Langman's Medical Embryology*. 9th ed. Baltimore: Lippincott Williams & Wilkins; 2003.
- Gonzalez-Juanatey C, Testa A, Vidan J, et al. Persistent left superior vena cava draining into the coronary sinus: Report of 10 cases and literature review. *Clin Cardiol* 2004;27(9):515–8.
- Biffi M, Boriani G, Frabetti L, et al. Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: A 10-year experience. *Chest* 2001;120(1):139–44.
- Pretorius PM, Gleeson FV. Case 74: Right-sided superior vena cava draining into left atrium in a patient with persistent left-sided superior vena cava. *Radiology* 2004;232(3):730–4.
- Demos TC, Posniak HV, Pierce KL, et al. Venous anomalies of the thorax. *Am J Roentgenol* 2004;182(5):1139–50.
- Cormier MG, Yedlicka JW, Gray RJ, et al. Congenital anomalies of the superior vena cava: A CT study. *Semin Roentgenol* 1989;24(2):77–83.
- Chen SJ, Liu KL, Chen HY, et al. Anomalous brachiocephalic vein: CT, embryology, and clinical implications. *Am J Roentgenol* 2005;184(4):1235–40.
- Bass JE, Redwine MD, Kramer LA, et al. Spectrum of congenital anomalies of the inferior vena cava: Cross-sectional imaging findings. *Radiographics* 2000;20(3):639–52.
- Dudiak CM, Olson MC, Posniak HV. CT evaluation of congenital and acquired abnormalities of the azygos system. *Radiographics* 1991;11(2):233–46.
- Applegate KE, Goske MJ, Pierce G, et al. Situs revisited: Imaging of the heterotaxy syndrome. *Radiographics* 1999;19(4):837–52.
- Hatfield MK, Vyborny CJ, MacMahon H, et al. Congenital absence of the azygos vein: A cause for “aortic nipple” enlargement. *Am J Roentgenol* 1987;149(2):273–4.
- Murillo H, Cutalo MJ, Jones RP, et al. Pulmonary circulation imaging: Embryology and normal anatomy. *Semin Ultrasound CT MR* 2012;33(6):473–84.
- Sears EH, Aliotta JM, Klinger JR. Partial anomalous pulmonary venous return presenting with adult-onset pulmonary hypertension. *Pulm Circ* 2012;2(2):250–5.
- Porres DV, Morenza OP, Pallisa E, et al. Learning from the pulmonary veins. *Radiographics* 2013;33(4):999–1022.
- Herlong JR, Jagers JJ, Ungerleider RM. Congenital Heart Surgery Nomenclature and Database Project: Pulmonary venous anomalies. *Ann Thorac Surg* 2000;69:S56–S69.
- Sakurai H, Kondo H, Sekiguchi A, et al. Left pneumonectomy for lung cancer after correction of contralateral partial anomalous pulmonary venous return. *Ann Thorac Surg* 2005;79(5):1778–80.
- Takamori S, Hayashi A, Nagamatsu Y, et al. Left partial anomalous pulmonary venous connection found during a lobectomy for lung cancer: Report of a case. *Surg Today* 1995;25(11):982–3.
- Sasaki H, Naka N, Kitahara N, et al. Right partial anomalous pulmonary venous connection found during lobectomy for coexisting lung cancer and tuberculosis: Report of a case. *Clin Lung Cancer* 2006;7(5):350–2.
- Maldonado JA, Henry T, Gutiérrez FR. Congenital thoracic vascular anomalies. *Radiol Clin North Am* 2010;48(1):85–115.
- Benson RE, Songrug T. CT appearance of persistent left superior vena cava, anomalous right superior pulmonary venous return into the right-sided superior vena cava and a sinus venosus-type atrial septal defect. *Br J Radiol* 2009;82(983):e235–9.
- Dillon EH, Camputaro C. Partial anomalous pulmonary venous drainage of the left upper lobe vs duplication of the superior vena cava: Distinction based on CT findings. *Am J Roentgenol* 1993;160(2):375–9.
- Woodring JH, Howard TA, Kanga JF. Congenital pulmonary venolobar syndrome revisited. *Radiographics* 1994;14(2):349–69.
- Gudjonsson U, Brown JW. Scimitar syndrome. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2006:56–62.
- Vida VL, Padalino MA, Boccuzzo G, et al. Scimitar syndrome: A European Congenital Heart Surgeons Association (ECHSA) multicentric study. *Circulation* 2010;122(12):1159–66.