
Anomalies and Malformations of the Pulmonary Circulation

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Abstract

Congenital anomalies of the pulmonary circulation can be seen as isolated anomalies or as part of more complex malformations affecting numerous organs and systems, in particular the lungs, heart, and great vessels. These malformations may affect the pulmonary arteries, pulmonary veins, the aorta, and the intrathoracic systemic veins. With its superb spatial and temporal resolution, contrast-enhanced multidetector CT has become the imaging modality of choice for the evaluation of these entities, allowing imaging of the heart, vasculature, and lungs in one single examination.

1 Introduction

Congenital anomalies of the pulmonary circulation can be seen as isolated anomalies or as part of more complex malformations affecting numerous organs and systems, in particular the lungs, heart, and the great vessels. These malformations may affect the pulmonary arteries, pulmonary veins, the aorta, and the intrathoracic systemic veins. With its superb spatial and temporal resolution, contrast-enhanced multidetector CT has become the imaging modality of choice for the evaluation of these entities,

allowing imaging of the heart, vasculature, and lungs in one single examination.

2 Congenital Anomalies of the Pulmonary Arteries

There are four pulmonary artery congenital abnormalities that can be seen as isolated anomalies or associated with other cardiovascular and tracheobronchial anomalies: (1) congenital interruption of the pulmonary artery, (2) peripheral pulmonary artery stenosis, (3) idiopathic aneurysm of the pulmonary artery, and (4) aberrant origin of the left pulmonary artery. Conotruncal abnormalities (tetralogy of Fallot, transposition of the great arteries, interrupted aortic arch, aortopulmonary window) comprise a diverse group of complex congenital heart defects affecting the great vessels, and outflow tracts of the heart

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(embryonic conus arteriosus and truncus arteriosus) are more in the realm of congenital cardiac pathology and will not be discussed here.

3 Congenital Interruption of the Pulmonary Artery

Unilateral absence of a pulmonary artery may be seen associated with congenital heart disease, such as septal defects, truncus arteriosus, and tetralogy of Fallot, or as an isolated anomaly. This condition was previously known as congenital absence of the pulmonary artery, but the term “interruption” is preferred, since only the mediastinal segment of the right or left pulmonary artery is absent, whereas the more peripheral intrapulmonary network is present. The affected lung receives oxygenated blood and vascular supply through collateral systemic arteries such as the bronchial arteries, intercostal arteries, internal mammary artery, and a patent ductus arteriosus. The interrupted pulmonary artery usually lies on the opposite side of the aortic arch (Figs. 1 and 2). The right pulmonary artery is more commonly affected in the isolated form, with congenital absence or interruption of the left more often seen in patients with other congenital anomalies like tetralogy of Fallot (Davis 2000). Women are affected more often than men (3:2), with those patients with associated congenital heart disease manifesting earlier in life. A patent ductus arteriosus is commonly present ipsilateral to the absent pulmonary artery (Apostolopouou et al. 2002). Patients with the isolated form usually present with nonspecific clinical manifestations such as dyspnea (40%), recurrent pulmonary infection (37%), or hemoptysis (20%), and nearly half of affected patients (44%) will have arterial pulmonary hypertension (Harjel et al. 2002). It has been postulated that a unilateral interruption of the pulmonary artery results from the proximal sixth aortic arch in the embryo which gives rise to the extraparenchymal pulmonary artery with persistent connection of the intra-



Fig. 1 Interrupted right pulmonary artery in an 11-year-old female with a normal left-side aortic arch. Contrast-enhanced CT shows only a left pulmonary artery arising from the pulmonary trunk and slightly prominent bronchial arteries in the right hilum (*arrows*)

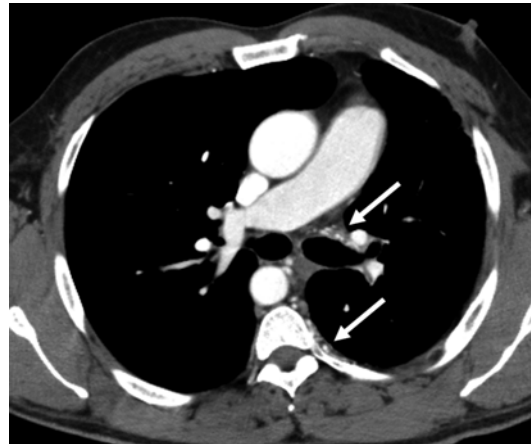


Fig. 2 Congenital interruption of the left pulmonary artery in a 48-year-old male with right-side aortic arch. Contrast-enhanced CT shows absence of the left pulmonary artery, a right-side descending thoracic aorta, and prominent systemic arterial circulation in the left hilum, mediastinum, and left-side chest wall (*arrows*)

pulmonary arteries to the distal sixth aortic arch which originates the ductus arteriosus (Apostolopouou et al. 2002). On imaging exams the affected lung is usually smaller, which is evidenced by shifting of the heart and mediastinum, and elevation of the hemidiaphragm to the affected side (Fig. 3). There is complete absence of the mediastinal segment of the pulmonary artery with a smaller ipsilateral hilum and a prominent contralateral hilum

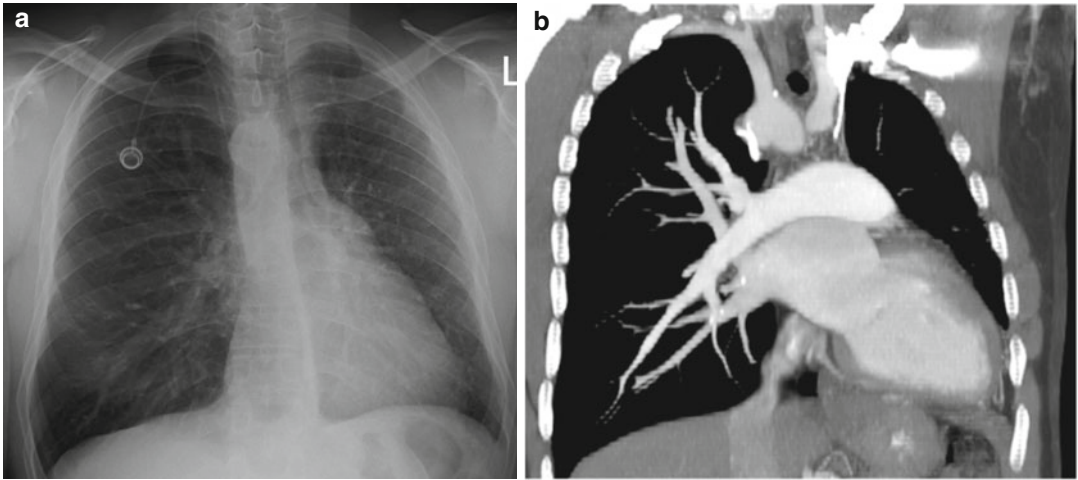


Fig. 3 Left-side cardiac and mediastinal shifting in a patient with congenital interruption of the left-side pulmonary artery. (a) Chest X-ray shows a right-side aortic arch and diminished volume in the left hemithorax. (b)

Contrast-enhanced CT, MIP coronal reconstruction reveals a mildly enlarged single right pulmonary artery with leftward deviation of the heart with a smaller left hemithorax

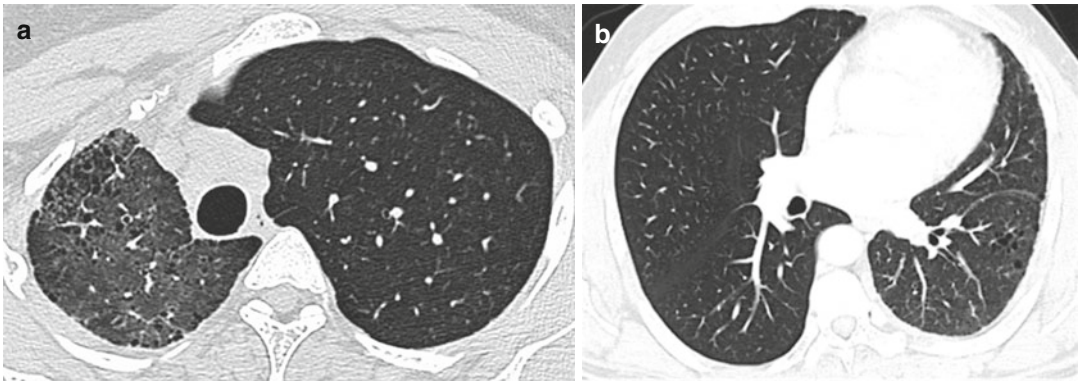


Fig. 4 (a) High-resolution CT in a female patient with congenital interruption of the right pulmonary artery. Axial image at the level of the upper lobes demonstrates numerous cysts in the right lung. (b) Lung window axial

image at the level of the lower lobes in a patient with congenital interruption of the left pulmonary artery shows small pulmonary cysts in the left lung

which receives the entire output of the right ventricle and pulmonary trunk. The extrapleural space is prominent, and collateral circulation may be also appreciated in the chest wall. On high-resolution CT, the affected lung dem-

onstrates reticular opacities, septal thickening, cystic lung changes/honeycombing, and bronchial dilation similar to interstitial pulmonary fibrosis (Fig. 4) (Ryu et al. 2004; Sakai et al. 2002).

4 Peripheral Pulmonary Artery Stenosis

Pulmonary artery stenosis is commonly associated with other complex congenital heart diseases, in particular tetralogy of Fallot, transposition of the great vessels, atrial septal defect, and ventricular septal defect. Isolated congenital peripheral pulmonary stenosis or pulmonary branch stenosis is less common. Three conditions classically associated with peripheral pulmonary artery stenosis are in utero exposure to rubella virus (post-rubella or maternal rubella syndrome), Noonan's syndrome, and Williams' syndrome. More recently Alagille's syndrome, cutis laxa syndrome, and Ehlers-Danlos syndrome have also been reported to be also associated with congenital pulmonary artery branch stenosis (Kamath et al. 2004; Wahab et al. 2003). Rarely, it can present as an idiopathic isolated condition in the adult not associated with any systemic condition or known syndrome (Kreutzer et al. 1996; Shaj et al. 2000). Depending on the degree and extension of stenosis, affected patients may be asymptomatic or present with arterial pulmonary hypertension. Physical signs and symptoms may include dyspnea, fatigue, chest pain, palpitations, right ventricular heart failure, hepatomegaly, and peripheral edema. The mor-

phology of peripheral pulmonary artery stenosis is quite variable and can present as single narrowing of the pulmonary trunk, right or left pulmonary artery (type I), bifurcation of the pulmonary trunk extending to the proximal right or left pulmonary arteries (type II), multiple peripheral pulmonary arteries without abnormality of the central vasculature (type III), or a combination of involvement of the central and peripheral vessels (type IV) (Gray et al. 1963). Differential diagnosis includes changes from systemic vasculitis such as Behçet's disease or Takayasu's arteritis (Toledano et al. 2011). In cases of vasculitis, there is associated abnormal thickening of the arterial walls, not encountered in congenital causes (Fig. 5).

5 Idiopathic Aneurysm of the Main Pulmonary Artery

Idiopathic dilation of the main pulmonary artery or pulmonary trunk with or without the central pulmonary arteries is defined as an abnormal dilation that occurs in the absence of a preexistent cardiac or pulmonary disease such as pulmonary valve stenosis, pulmonary arterial hypertension, syphilis, or connective tissue disorder.

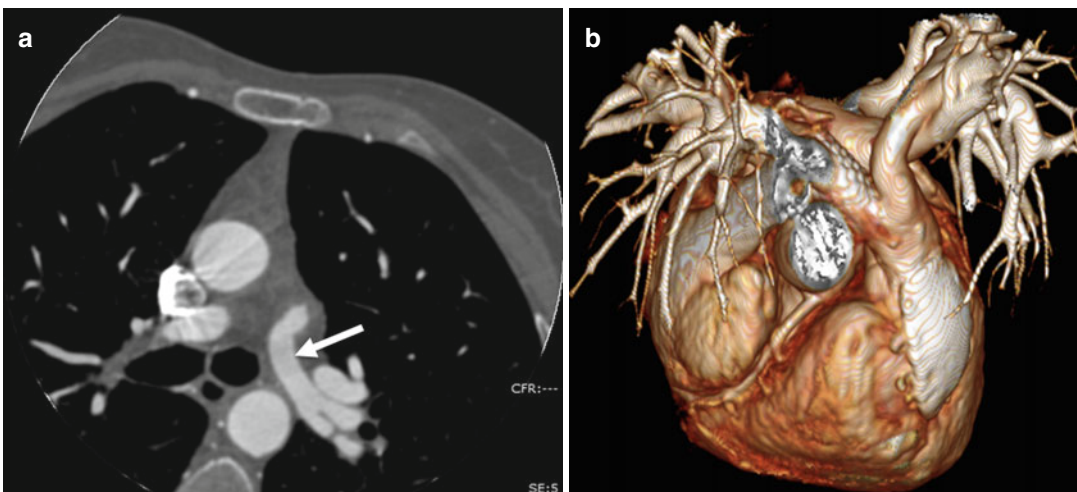


Fig. 5 Branch pulmonary artery stenosis in a female patient with maternal rubella syndrome. (a) Contrast-enhanced axial CT. (b) Volume-rendered 3-D reconstruction.

tion. CT demonstrates a long segment stenosis of the left pulmonary artery

der. Idiopathic pulmonary artery aneurysms are rare, typically asymptomatic and incidentally encountered. Little is known about the natural history of this condition, and reported outcomes range from asymptomatic cases to rupture and sudden death (van Rens et al. 2000; Nair and Cobanoglu 2001; Seguchi et al. 2011; Andrews et al. 1993). These aneurysms can grow very large, up to 12 cm in diameter, producing mass effect on adjacent structures such as the trachea or laryngeal nerve. Cystic medial degeneration of the arterial wall is the most common pathologic abnormality reported (Deb et al. 2005). The diagnosis of this condition is established by exclusion of diseases that induce enlargement of the pulmo-

nary arteries. The following diagnostic criteria have been proposed: (1) simple dilatation of the pulmonary trunk, (2) absence of intra- or extra-cardiac shunt, (3) absence of chronic cardiopulmonary disease, (4) absence of arterial wall disease, and (5) normal pressure in the right ventricle and pulmonary artery (Niida et al. 2011). Even though the pulmonary arterial system is a low pressure entity, surgery is advocated for aneurysms with a diameter of 6 cm or greater to prevent potential complications, such as rupture, dissection, and cardiac death, and is also recommended in patients experiencing symptoms such as tracheobronchial compression or chest pain (Fig. 6) (Kharge et al. 2013).

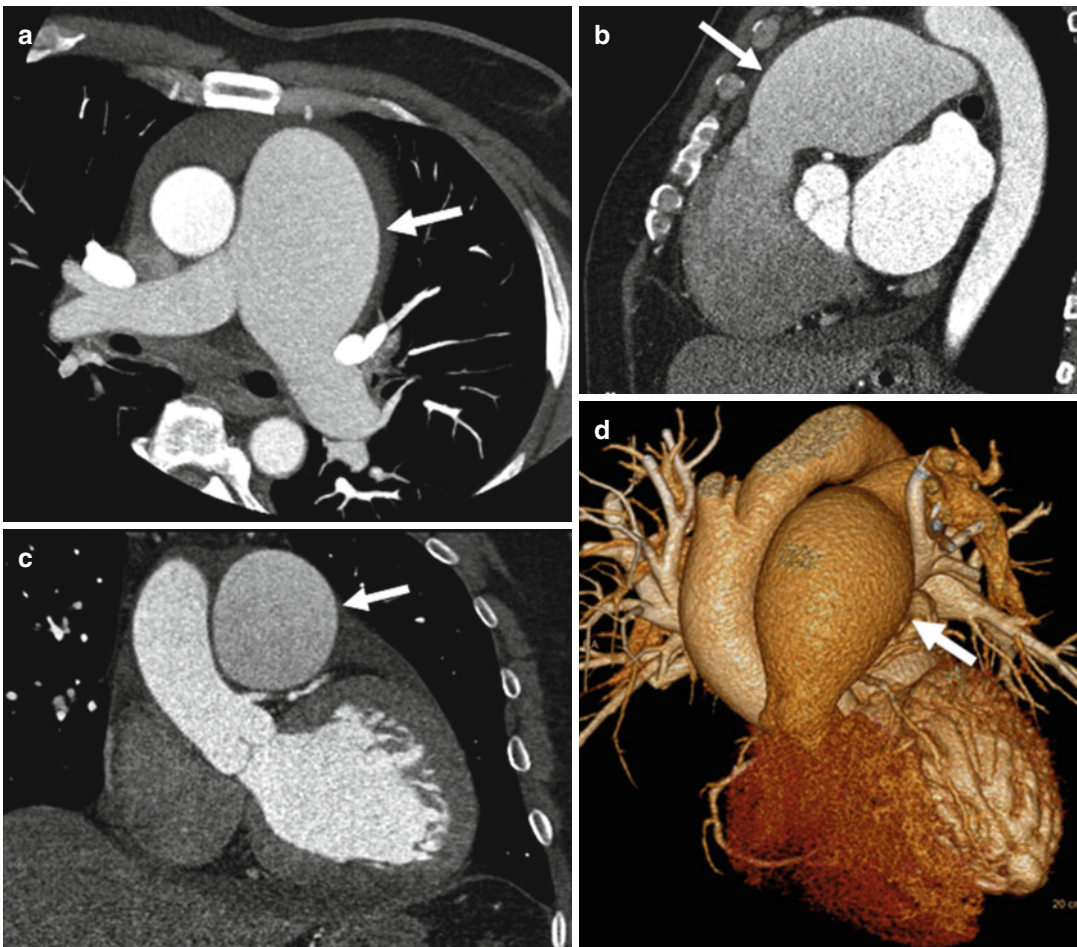


Fig. 6 Idiopathic aneurysm of the main pulmonary artery. (a–d) Contrast-enhanced cardiac-gated CT with axial, sagittal, coronal, and 3-D reconstruction. There is a large aneurysmal dilation of the pulmonary trunk and to a lesser

extent of the central pulmonary arteries in this patient with no evidence of cardiopulmonary disease, systemic, or known arterial wall disorder (arrows)

6 Aberrant Origin of the Left Pulmonary Artery (Pulmonary Artery Sling)

This rare condition is characterized by an abnormal origin of the left pulmonary artery from the posterior aspect of the right pulmonary artery to the right of the trachea and immediately above the right mainstem bronchus with a course between the trachea and the esophagus on its way to the left pulmonary

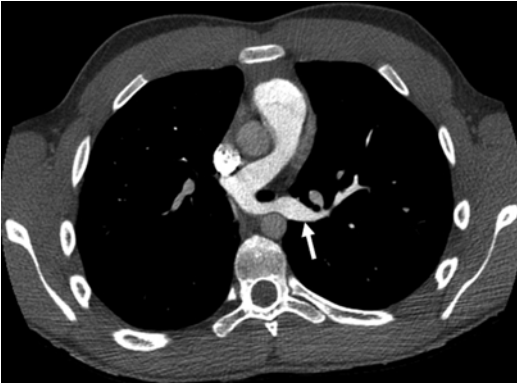


Fig. 7 Pulmonary artery sling in a young adult female patient. Contrast-enhanced CT, axial image at the level of the pulmonary trunk demonstrates aberrant origin of the left pulmonary artery from the right pulmonary artery, with the aberrant artery passing between the trachea and the esophagus (*arrow*)

hilum (Fig. 7). The exact prevalence of this condition is not known, but is considered a rare anomaly with a male to female ratio of 3:2. The majority of patients are symptomatic during the first year of life (90%), mainly with respiratory symptoms to include stridor, wheezing, and recurrent respiratory infection. Tracheobronchial anomalies are common (40%), including tracheal stenosis, abnormal branching pattern with the right tracheal bronchus, and complete cartilaginous tracheal rings, an association known as the “ring-sling” complex (Berdon et al. 1984). Underdevelopment of the right lung presenting as either agenesis or hypoplasia can be seen in up to 22% of cases (Chen et al. 2007). Major cardiovascular anomalies including atrial septal defect, ventricular septal defect, patent ductus arteriosus, aortic coarctation, and tetralogy of Fallot are found in 30% of affected patients (Gikonyo et al. 1989). Occasionally asymptomatic individuals are diagnosed later in life (Collins et al. 2008). Cross-sectional imaging, in particular multidetector CT, offers accurate anatomic information on both the vascular and tracheobronchial abnormalities of these patients, allowing appropriate surgical planning (Fig. 8) (Chen et al. 2007; Lee et al. 2001; Kagadis et al. 2007).

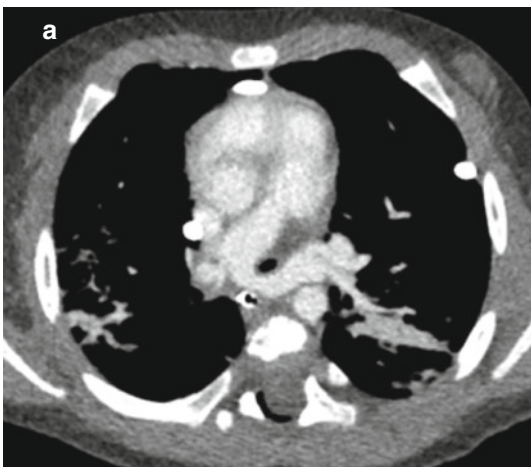


Fig. 8 Pulmonary artery sling in a four-month-old male with history of recurrent respiratory symptoms. (a) Cardiac CT axial image depicts the abnormal origin of the



left pulmonary artery from the right. (b) Volume-rendered 3-D reconstruction demonstrates a long segment tracheal stenosis

7 Developmental Anomalies of Pulmonary Veins

Developmental pulmonary venous anomalies are predominantly comprised of anomalous venous connections which result in an extracardiac left-to-right shunt, as pulmonary venous blood flows directly into the right side of the heart or into the systemic veins. In partial anomalous pulmonary venous return (PAPVR), up to three pulmonary veins drain into the systemic circulation, while in total anomalous pulmonary venous return (TAPVR), all four pulmonary veins drain into the systemic circulation. To be compatible with life, TAPVR requires a right-to-left shunt via a cardiac septal defect or a patent ductus arteriosus. TAPVR has been classified into four groups: supracardiac, cardiac and infracardiac, and mixed. Pathogenesis of this entity is closely related to the complex embryology.

8 Embryology

Three basic systems of venous drainage, the cardinal, umbilicovitelline, and omphalomesenteric systems, are seen in a developing embryo. The cardinal veins eventually differentiate into the superior vena cava (SVC) and coronary sinus, whereas the umbilicovitelline system develops into the inferior vena cava (IVC), ductus venosus, and portal vein. The common pulmonary vein (CPV) develops from the primordial left atrium and extends toward the lung buds. At approximately day 28 of gestation, the CPV joins the pulmonary portion of the splanchnic plexus, allowing flow of pulmonary blood into the heart. Over time, obliteration of pulmonary-splanchnic connections occurs with incorporation of CPV into the left atrial wall, leaving four independent pulmonary veins directly entering the left atrium. Early atresia or malposition of the CPV results in anomalous pulmonary venous return, with one or more pulmonary veins draining into the systemic venous circulation. Persistence of the cardinal veins and umbilicovitelline veins results in TAPVR (Total Anomalous Pulmonary Venous Return). It is hypothesized that “unroofing” of

the right superior pulmonary vein into the SVC during development produces a combination of anomalous return and a distinct type of atrial septal defect, the sinus venosus atrial septal defect (SVASD). Failure of resorption of membrane between the CPV and primordial left atrium results in cor triatriatum (Katre et al. 2012).

9 Partial Anomalous Pulmonary Venous Return

Classically, a partial anomalous pulmonary venous connection has been described as presenting in childhood, most frequently on the right side and associated with SVASD. The overall incidence of PAPVR is approximately 0.5% (Van Meter et al. 1990; Posniak et al. 1993; Dillon and Camputaro 1993). Significant variation is seen in the epidemiology of PAPVR in pediatric and adult populations. In pediatric patients who are symptomatic, the diagnosis is usually made earlier secondary to significant hemodynamic changes, while the condition remains undiagnosed until the adulthood in asymptomatic patients. In adults, frequently PAPVR is an incidental finding and only a few adult patients present with symptoms (Ho et al. 2009; Oliver et al. 2002). In pediatric studies (<18 years old), PAPVR has been reported to be twice as common in males as females and more frequently arises from the right (90%) more often than the left upper lobe (LUL) (10%). In addition, right upper lobe (RUL) PAPVR is associated with an SVASD in 80–90% which is located high and posterior in the septum near the SVC orifice, while ostium secundum atrial septal defect (OSASD) is seen in 10–15% of cases (Davia et al. 1973). PAPVR is more often hemodynamically significant when associated with congenital heart disease or scimitar syndrome. The “scimitar syndrome” is a complex form of PAPVR in which the anomalous pulmonary veins may drain into the supra- or infradiaphragmatic IVC or rarely into the hepatic or portal veins. It is also known as “hypogenetic lung syndrome” or “venobar syndrome” to emphasize that this anomaly is not simply a variant of pulmonary venous return, but it is rather more extensive mal-

formation. Scimitar syndrome is essentially an anomaly of the right lung. The anomalous right pulmonary vein commonly drains the entire right lung but rarely may drain only the middle and lower lobes and usually descends in a cephalad-to-caudal direction toward the diaphragm with a crescentic shape. The radiographic appearance of this anomalous vein resembles a “curved Turkish sword” or “scimitar,” from which the name has been derived (Vida et al. 2010; Gudjonsson and Brown 2006). Cardiopulmonary signs and symptoms depend upon the hemodynamic changes secondary to a left-to-right shunt and volume of the shunted blood, as well as whether PAPVR is associated with an atrial septal defect (ASD) or other congenital heart diseases. The volume of the shunt is calculated as a ratio between pulmonary blood flow (Q_p) and systemic blood flow (Q_s), at a given point of time. If the volume of the shunt is significant, that is, if the $Q_p:Q_s$ is more than 1.5–1.7, surgical intervention is usually needed to prevent development of pulmonary hypertension (Greene and Miller 1986). Commonly encountered signs and symptoms include dyspnea, orthopnea, fatigue, chest pain, palpitations, tachycardia, and peripheral edema. Electrocardiographic abnormalities include abnormal P wave, nonspecific ST segment changes, left axis deviation, right axis deviation, sinus tachycardia, sinus bradycardia, and other arrhythmias.

10 Imaging Findings

Left upper lobe PAPVR is characterized by an aberrant draining vein which usually conducts blood cranially from the left upper lobe to the left innominate vein which eventually drains into the normally positioned superior vena cava on the right side of the aortic arch. This vein is called the “vertical vein,” which is visualized as an anomalous vascular structure in the left para-aortic region or prevascular space where only mediastinal fat is normally present. A similar finding is observed in patients with persistent left-sided SVC (PLSVC); however a few differences exist between the vertical vein and PLSVC. In PLSVC, an aberrant vein usually con-

ducts blood caudally from the left subclavian and jugular veins to the right atrium via the coronary sinus. In LUL PAPVR, a normal to large left innominate vein is seen in the expected position (Adler and Silverman 1973; Oropeza et al. 1970). However in PLSVC, left innominate vein is often absent or small in caliber. In PLSVC, up to 20% of cases left innominate vein can be of normal size (Winter 1954). Therefore, although the absence of a left innominate vein is a reliable indicator of PLSVC, visualization of this vein does not aid in differentiation. The coronary sinus appears normal in LUL PAPVR but is usually enlarged in left-sided SVC, as it receives the blood flow from the left subclavian and jugular veins. Another differentiating point is the number of veins anterior to the left mainstem bronchus. Normally, only one vessel, which is the left superior pulmonary vein, is present in this location. In PLSVC, two vessels are present anterior to the left mainstem bronchus, specifically the left superior pulmonary vein and the more medially located left superior vena cava. In LUL PAPVR, no vessel is present in this location (Fig. 9).

Right upper lobe PAPVR is frequently more severe and diagnosed early in the pediatric population. It tends to be associated with other congenital malformations including a distinct type of ASD called as sinus venosus type ASD which is seen along the most posterior and superior aspect of the interatrial septum, usually at the level of SVC (Haramati et al. 2003). This is thought to be due to “unroofing” of the right superior pulmonary vein and the SVC during development. Anomalous right upper lobe pulmonary vein may connect to the superior vena cava, azygos vein, or right atrium directly (Herlong et al. 2000). This can be a subtle finding on CT, and unless anatomy of the region is closely scrutinized, this anomaly may remain undetected (Fig. 10).

The “scimitar syndrome” is commonly associated with right lung hypoplasia, dextroposition of the heart, hypoplasia of the right pulmonary artery, systemic arterial blood supply to the right lower lung from the branches of infradiaphragmatic aorta, lung sequestration, and sinus venosus or septum secundum atrial septal defects. The right lung may have abnormal lobation (fre-

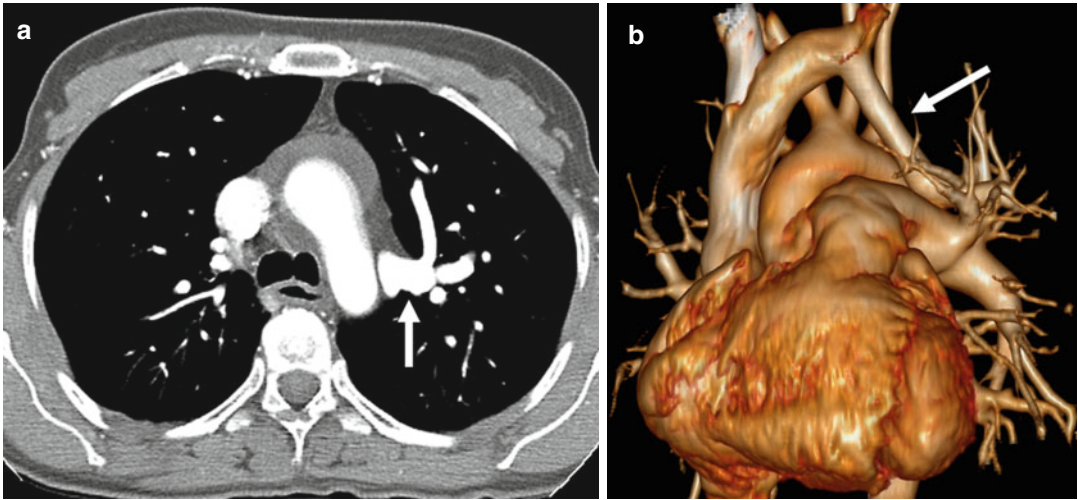


Fig. 9 Left upper lobe partial anomalous pulmonary venous return. (a) Contrast-enhanced CT, axial image at the level of the aortic arch. (b) CT and volume-rendered

3-D reconstruction. Images show an abnormal left upper lobe pulmonary vein draining to a left-side vertical vein (arrow)

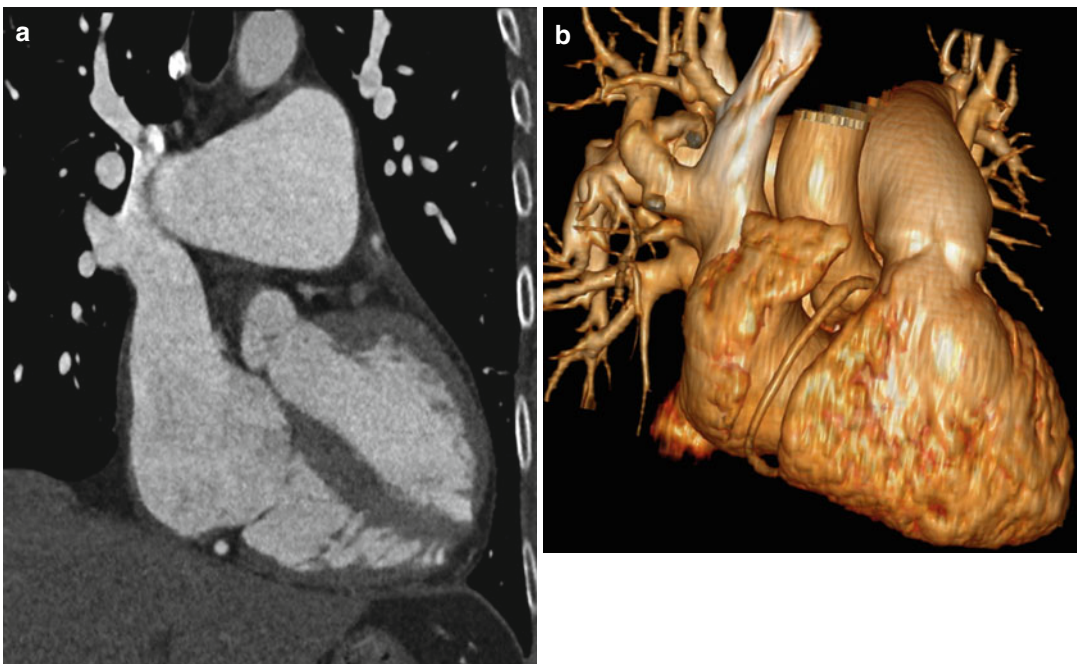


Fig. 10 Right upper lobe partial anomalous pulmonary venous return. (a) Contrast-enhanced CT with coronal reconstruction. (b) Volume-rendered 3-D reconstruction.

There is abnormal drainage of the right upper lobe pulmonary veins to the distal superior vena cava

quently only two lobes) with a bronchial branching pattern that mimics the left lung (Gudjonsson and Brown 2006; Korkmaz et al. 2011; Konen

et al. 2003). In most patients, right pulmonary hypoplasia causes a marked mediastinal shift and cardiac dextroposition. In severe cases, the entire

heart can be seen in the right hemithorax. Systemic arterial flow to the hypoplastic right lung originates directly from the upper segment of the abdominal aorta, and it is more common in the infantile form than in the adult form. A small pulmonary artery may also be present. Associated cardiac anomalies such as hypoplastic left heart, coarctation of the aorta, patent ductus arteriosus, tetralogy of Fallot, and persistent left superior vena cava can be seen. Overall, 19–31% of patients with scimitar syndrome have associated cardiac anomalies (Fig. 11) (Alsoufi et al. 2007).

“Pseudoscimitar syndrome” or a “meandering right pulmonary vein” (MRPV), the entity described by Goodman and colleagues, is closely related to scimitar syndrome in which an anomalous right pulmonary vein courses circuitously through the right lung, seen as scimitar sign on the chest radiograph, and drains into the left atrium rather than into the IVC or systemic circulation. MRPV can occur with or without other features of the classic scimitar syndrome. While most cases involve the right pulmonary veins, cases of anomalous

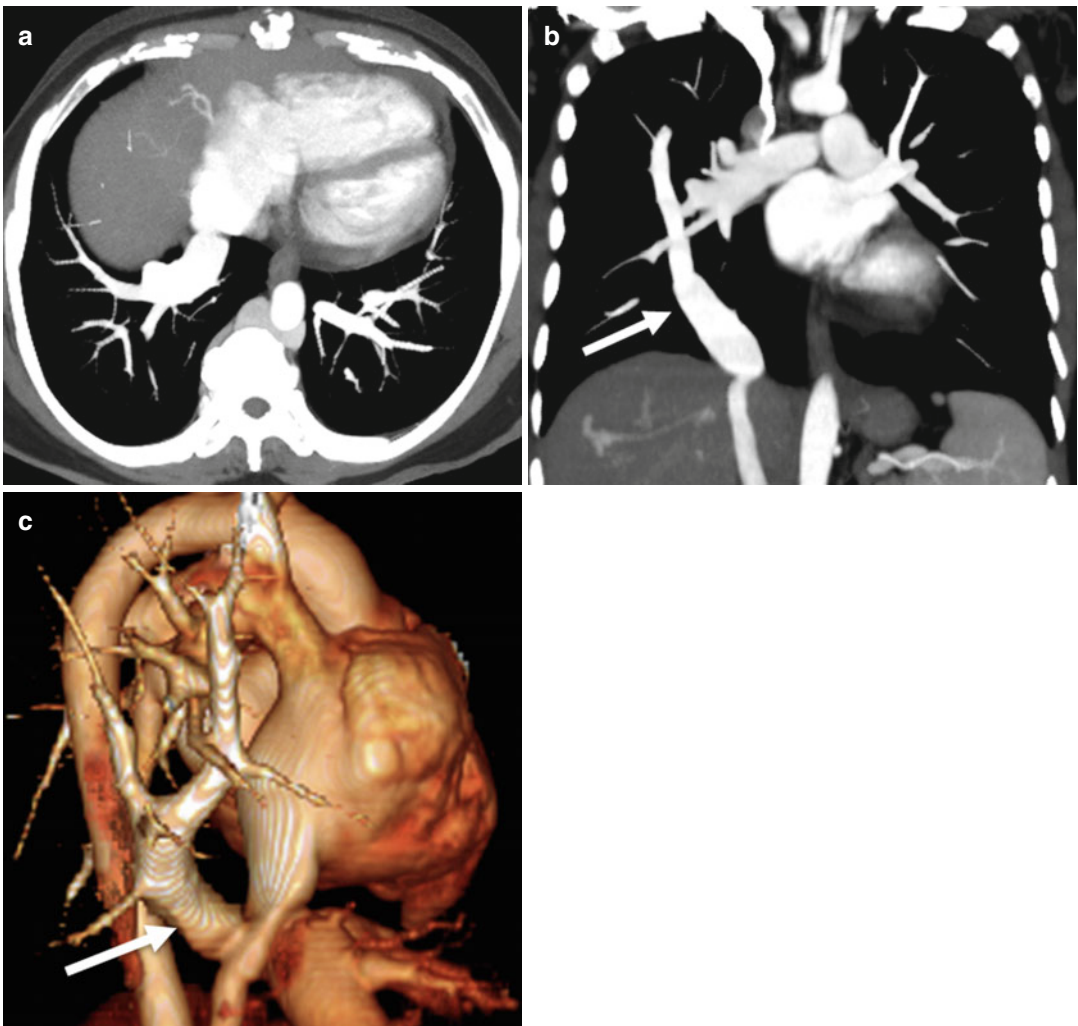


Fig. 11 Scimitar syndrome. (a) Contrast-enhanced CT, axial MIP. (b) Coronal MIP. (c) Volume-rendered 3-D reconstruction. There is a large anomalous vein (arrows)

collecting the entire venous drainage of the right lung draining to the suprahepatic inferior vena cava immediately above the diaphragm

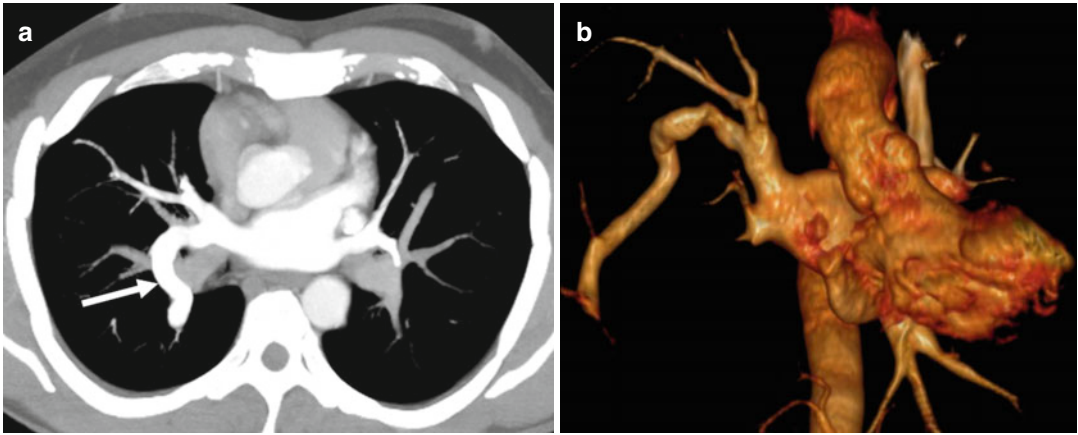


Fig. 12 Pseudoscimitar. (a, b) Contrast-enhanced CT, axial MIP, and volume-rendered 3-D reconstruction. There is an elongated and tortuous pulmonary vein in the right mid lung that terminates normally in the left atrium (*arrow*)

right and left pulmonary veins have been described (Goodman et al. 1972; Collins et al. 1982). The scimitar sign is not always present. It is important to distinguish between scimitar syndrome and MRPV. Scimitar syndrome results in a left-to-right shunt, which can lead to cyanosis and may require surgical correction. Consequently the patients are often symptomatic and present at a young age. In contrast, there is no left-to-right shunt in MRPV. In addition, some of the reported cases of meandering pulmonary veins connect to the left atrium and IVC simultaneously, also called as scimitar variant (Takeda et al. 1994). Contrast-enhanced multidetector computed tomography (MDCT) is the modality of choice for diagnosis of anomalous pulmonary venous connections, considered as “one stop shop” imaging. It allows rapid acquisition of data with high spatial resolution and wide anatomic coverage with a high sensitivity and specificity that approaches 100% (Fig. 12) (Kim et al. 2000).

11 Cor Triatriatum

Cor triatriatum (CTT) is a rare congenital heart anomaly (0.1%) in which left atrium is divided into two distinct anterior and posterior chambers by a fibromuscular membrane. Failure of

resorption of a membrane between common pulmonary vein and primordial left atrium results in this anomaly. The posterior chamber receives blood flow from the pulmonary veins, whereas the anterior chamber delivers blood to the mitral valve. The location of the atrial membrane differentiates cor triatriatum from a supralvalvular mitral ring. CTT is differentiated from supralvalvular mitral ring by the position of the left atrial appendage (LAA). In CTT, the LAA is part of the anterior (mitral valve) atrial chamber, whereas the LAA is part of the posterior (pulmonary vein) atrial chamber in patient with a supralvalvular ring (Bezzin et al. 2014). Although symptoms are usually encountered in infancy, this entity may rarely present in adulthood when the membrane contains multiple fenestrations. The most common presenting symptoms in adults are dyspnea, hemoptysis, and orthopnea, which mimic mitral stenosis at presentation (Su et al. 2008). Cor triatriatum is frequently associated with other cardiovascular abnormalities. In adults, the most common associated abnormalities are atrial secundum septal defect, mitral regurgitation, and the presence of PLSVC. Diagnosis is usually established by echocardiography; however MDCT or MRI is more specific in evaluating the size and number of fenestrations and to find associated congenital cardiovascular anomalies (Fig. 13).

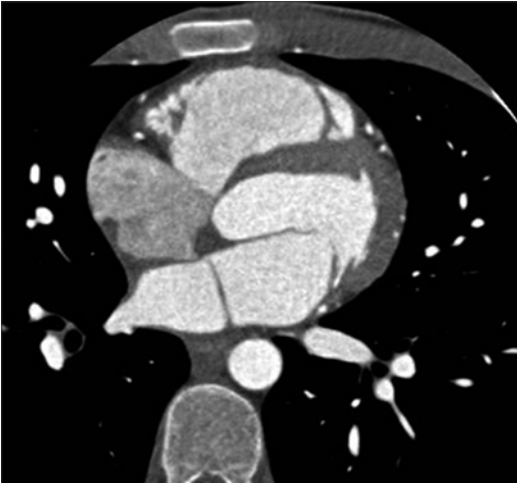


Fig. 13 Cor triatriatum in a 24-year-old male. Contrast-enhanced CT shows a thin membrane dividing the left atrium in two distinct chambers

12 Total Anomalous Pulmonary Venous Connection

Total anomalous pulmonary venous connection is a rare form of congenital heart disease accounting for 1.5–3% of congenital heart diseases in which all pulmonary veins connect to the systemic veins, right atrium, coronary sinus, or IVC. Depending on the drainage site of the pulmonary veins, the defect may be divided into the following four types: supracardiac (50%), intracardiac (25%), infracardiac (20%), and mixed type (5%). This condition is a cause of neonatal cyanosis. Approximately 1/3 of patients with TAPVC have other associated cardiac lesions including single ventricle, atrioventricular septal defect, transposition of the great arteries, hypoplastic left heart syndrome, or patent ductus arteriosus. Type 1 TAPVC (supracardiac connection) is the most common subtype in which connection to the left innominate vein is found (Karamlou et al. 2007). Other less common supracardiac venous connections include SVC and azygos veins. Anomalous connection to the right superior vena cava is much less frequent, but when present it is often associated with heterotaxy syndrome (polysplenia/asplenia, Ivemark syndrome). Pulmonary venous obstruction (PVO) is less common in this type. On chest radiograph,

the classic “snowman” or “figure of 8” appearance is seen secondary to widening of the mediastinum due to dilated SVC and innominate vein. But this diagnostic cardiac configuration might not manifest during neonatal and infantile periods until there is a physiological decrease in pulmonary vascular resistance type 2 TAPVC (cardiac connection) in which all four pulmonary veins connect at the level of the coronary sinus or in the posterior wall of right atrium near the midatrial septum (Shen et al. 2013). The anomalous veins may connect via a short channel or multiple openings to the right atrium. The coronary sinus ostium is markedly enlarged although normal in position. The infracardiac type, type 3, is the most common type of pulmonary venous obstruction, and this obstruction results in severe pulmonary congestion and pulmonary hypertension. The obstruction is either within the anomalous connecting vein or at its connection to the systemic circulation. The pulmonary veins from both sides converge behind the left atrium and form a common vertical descending vein, which courses anterior to the esophagus and traverses the diaphragm at the esophageal hiatus. This vertical descending vein may join the portal venous system (80–90%, in which case obstruction is almost always present since the blood passes through the hepatic sinusoids), either at the splenic or splenic-superior mesenteric venous confluence (Shen et al. 2013). Occasionally, the vertical vein may connect directly to the ductus venosus, hepatic veins, or inferior vena cava. Chest radiograph typically demonstrates passive vascular engorgement or pulmonary edema or both and absence of cardiomegaly. The last subtype is the mixed pattern representing about 2–10% of cases. In this form, pulmonary veins drain to at least two different locations, including a brachiocephalic vein, SVC, azygos vein, coronary sinus, right atrium, or below the diaphragm. The most common pattern of mixed obstruction is drainage of a vertical vein to the left innominate vein and drainage of the right lung either via the right atrium or the coronary sinus. This pattern of anomalous venous connection is generally associated with other major cardiac lesions. Transthoracic echocardiography is the first-line

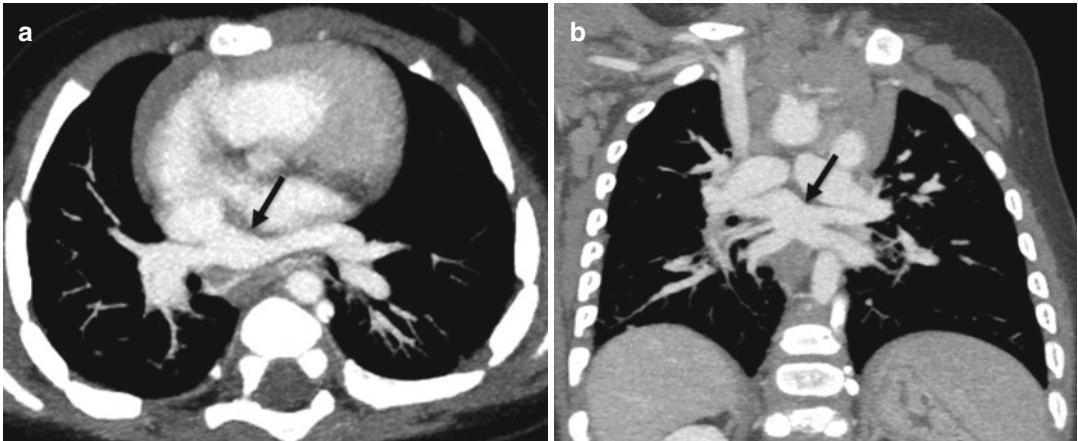


Fig. 14 Total anomalous pulmonary venous return in a seven-month-old boy. **(a)** Contrast-enhanced CT, axial MIP. **(b)** Coronal reconstruction. All bilateral pulmonary

veins are seen draining into the posterior aspect of the right atrium (*arrows*)

study for assessing children with suspected TAPVC. However, cross-sectional imaging is most useful in evaluation of patients with mixed TAPVC and PAPVC. Detailed 3-D and volume-rendered capacity of MDCT and MRI are a unique contribution to both diagnosis and treatment (Fig. 14).

13 Pulmonary Arteriovenous Malformation

Pulmonary arteriovenous malformation (PAVM) is defined as a direct communication between a peripheral pulmonary artery and a peripheral pulmonary vein without an intervening capillary bed. This is probably the most common anomaly of the pulmonary vasculature and commonly associated with hereditary hemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu disease (80%), an autosomal dominant hereditary condition with variable penetrance, clinically characterized by epistaxis (80%), mucocutaneous telangiectasia (70%), and visceral arteriovenous malformations (20–50%). All patients with possible or confirmed HHT and family members should be screened for PAVM, since pulmonary arteriovenous anomalies are so frequent in this population (45%), with 60% of affected individuals having multiple lesions (Gossage and Kanj

1998; Khurshid and Downie 2002; Jaskolka et al. 2004). Contrast echocardiography is commonly used as the first imaging modality of choice for screening, but CT is considered the gold standard for the diagnosis of this condition. Current technology with MDCT allows detection of small PAVM even on non-contrast examination (Faugham et al. 2009). On CT, PAVM appears as dilated feeding and draining intrapulmonary vessels which present as irregular tubular densities in communication with “nodules” or serpiginous masses, more commonly seen in the lower lobes. Smaller lesions are better appreciated on thick slab images or maximum intensity projection (MIP) with multiplanar reconstruction. As for other vascular pathologies, contrast-enhanced CT can better demonstrate the vascular nature of this condition, but contrast is not always required (Fig. 15).

13.1 Systemic Venous Anomalies

As opposed to pulmonary venous abnormalities, systemic venous anomalies are typically incidental findings. The incidence of systemic venous anomalies of the thorax is quite rare, but these anomalies are more frequently encountered in patients with congenital heart disease (CHD) as opposed to the general population. These anoma-

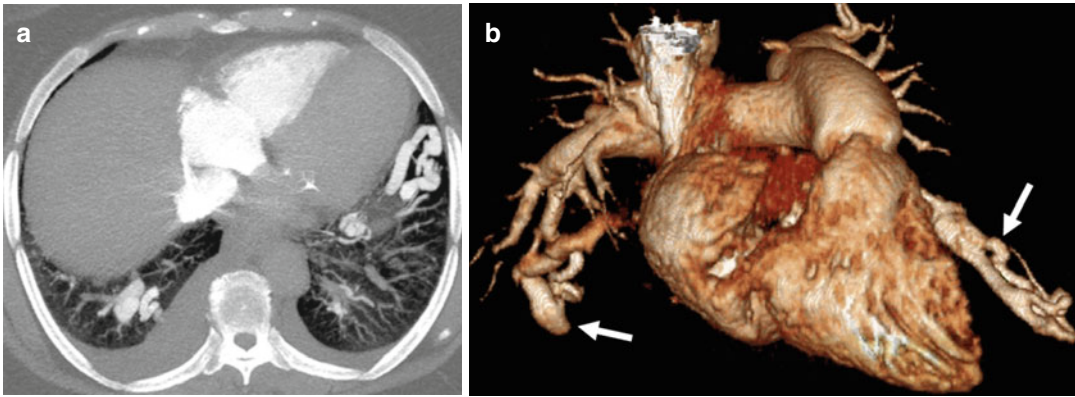


Fig. 15 Bilateral pulmonary arteriovenous malformations in a patient with HHT. **(a)** Contrast-enhanced CT, axial MIP. **(b)** Volume-rendered 3-D reconstruction. Dilated and

tortuous vessels with abnormal communication between arteries and veins are appreciated in the bilateral lower lobes (*arrows*), as well as bilateral pleural effusions

lies can be especially important in patients with CHD with regard to selection of a surgical approach and also have implications relative to the venous cannulation for cardiac bypass during surgery; thus they should be purposely sought for in these patients (Corno et al. 2013). These systemic venous anomalies in the thorax are the result of complex variations in the persistence and/or regression of segments of three sets of veins during the first 2 months of fetal development including the umbilical, vitelline, and cardinal systems. Anomalies most frequently encountered include those related to the superior vena cava (SVC) and azygos system although left brachiocephalic vein and intrathoracic IVC anomalies are also encountered (Webb et al. 1982).

13.2 SVC Anomalies

13.2.1 Left-Sided SVC/Double SVC

In normal cases, the right anterior and common cardinal veins form a right-sided SVC, and the left anterior cardinal vein regresses. If there is persistence of both anterior cardinal veins, a double SVC is encountered, and if the right anterior cardinal vein regresses, a left-sided SVC will be the only large vessel draining into the right side

of the heart (Webb et al. 1982; Demos et al. 2004).

Persistent LSVC is the most common variant of systemic venous drainage which is discovered as an incidental finding in 0.3–0.5% of the normal population and seen in up to 4–5% of patients with congenital heart disease (Webb et al. 1982; Lawler et al. 2002; Godwin and Chen 1986; Biffi et al. 2001; Piciucchi et al. 2014). In the vast majority of cases (82–90%), the left SVC is a component of a duplicated system (double SVC). Sixty-five percent of these cases will have absence of the left brachiocephalic vein (LBCV). The right and left SVC will lie in the same relative position on either side of the mediastinum and be similar in caliber, as opposed to cases with persistence of the LBCV, where the right SVC will typically be larger than the left (Webb et al. 1982; Godwin and Chen 1986). As an isolated anomaly in the absence of CHD, the left SVC will almost always drain through the oblique vein of Marshall which extends behind the left atrium, into the coronary sinus and then into the right atrium (Lawler et al. 2002). In these cases, the coronary sinus is typically enlarged, and if encountered during contrast-enhanced CT, the vessel may be densely opacified if IV contrast is injected into the left arm (Demos et al. 2004). PLSVC is readily recognized on CT and MR of

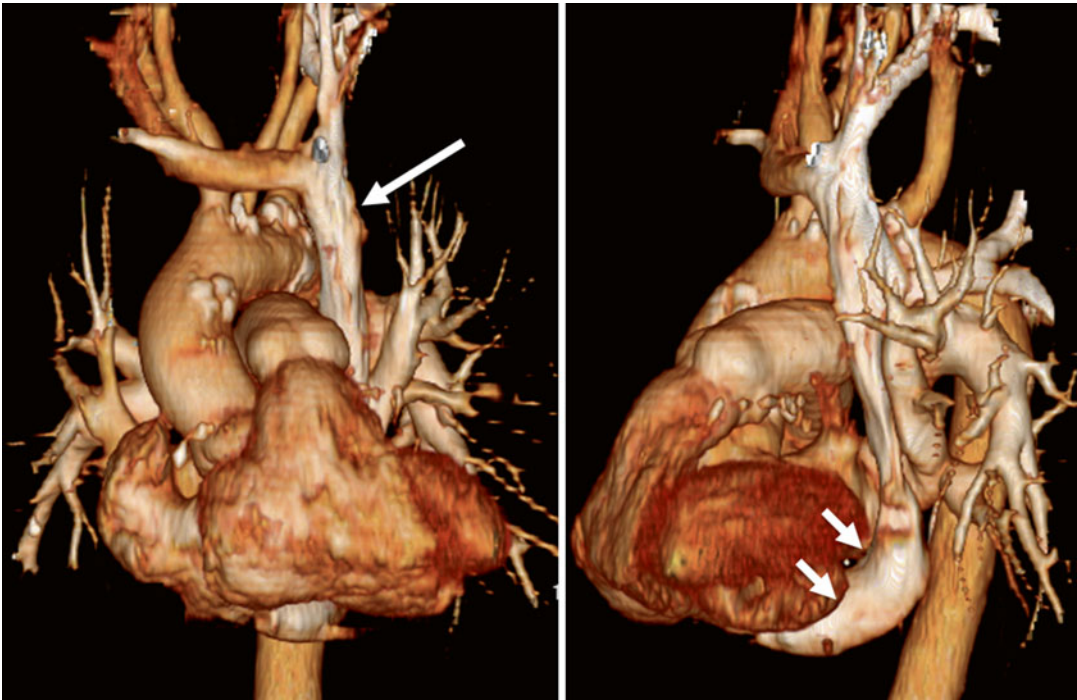


Fig. 16 Left-side superior vena cava. Volume-rendered 3-D reconstructions show a single left-side superior vena cava (*long arrow*) ending in a dilated coronary sinus (*short arrows*)

the chest and may be seen, often retrospectively, as focal widening of the mediastinum on chest X-ray along the superior mediastinal border. However, this anomaly is most often recognized on chest X-ray when the vessel is traversed by an indwelling catheter (Fig. 16). In patients with a solitary left SVC, the condition can simulate an abnormal aorta, although this vascular variant is not clinically significant unless there is narrowing of the ostium of the coronary sinus, which can lead to difficulties placing IV lines and pacemaker/defibrillator leads (Demos et al. 2004; Lawler et al. 2002; Biffi et al. 2001). Additionally, atresia of the coronary sinus may be encountered in these patients. Although occluded, the coronary sinus, by way of collateral vessels, still receives the total venous output of the coronary veins. The coronary venous drainage to the heart, however, is then retrograde through the left SVC to the LBCV. If this is unrecognized, ligation of the LSVC as part of a cardiac surgical procedure has led to acute coronary venous hypertension and myocardial ischemia. Drainage of a PLSVC

into the left atrium instead of the CS is rarely encountered and occurs in association with many types of CHD, but is rare if the heart is normal (Fig. 17) (Demos et al. 2004).

13.2.2 Right-Sided SVC

Isolated anomalies of a right-sided SVC are rare. The vessel may insert low into the right atrium, may drain into the left atrium, or may be aneurysmal, typically congenital in nature (Demos et al. 2004).

13.3 Azygos System Anomalies

The azygos system consists of paired paravertebral venous structures in the posterior thorax including the azygos and hemiazygos veins with the intercostal and mediastinal tributaries draining into the azygos, hemiazygos, and accessory hemiazygos veins. The azygos vein originates at the junction of the right ascending lumbar and subcostal veins, enters the chest at the aortic hiatus,



Fig. 17 Left superior vena cava draining into the left atrium in a patient with a duplicated vena cava. Contrast-enhanced CT, coronal reconstruction shows the right superior vena cava terminating in the right atrium and the left superior vena cava terminating in the left atrium

and ascends along the right anterolateral surface of the thoracic vertebrae. At T5–T6, the vessel arches anteriorly, just cephalad to the right mainstem bronchus, and drains into the SVC, or rarely the right brachiocephalic vein (BCV), right subclavian vein (SCV), intrapericardial SVC, or right atrium. Similar to the azygos vein, 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 IVC. The right superior intercostal vein drains the right second through fourth intercostal spaces and joins the azygos just proximal to the arch. The hemiazygos ascends along the left anterolateral aspect of the thoracic vertebral column and at T8–T9 and crosses dorsal to the descending thoracic aorta to join the azygos vein. From this point, the accessory hemiazygos vein extends further cephalad in the left paravertebral location and may communicate with the azygos vein at different levels. The left superior intercostal vein drains the left second through fourth intercostal spaces and communicates with the accessory hemiazygos in 75% of patients, then arches ventrally adjacent to the aortic arch and drains into the left BCV. This can be seen on frontal CXR en face, known as the aortic nipple (Demos et al. 2004).

The right and left supreme intercostal veins drain the first intercostal spaces and usually empty into the left BCV, although they may communicate with the corresponding superior intercostal veins.

The embryology of the azygos-hemiazygos venous system is not entirely clear. The azygos venous system develops on the basis of multiple transformations of the subcardinal veins with the azygos vein considered to derive from the upper right supracardinal vein, the arch from the upper segment of right posterior cardinal vein, and the hemiazygos from the upper left supracardinal vein (Piciocchi et al. 2014). The left superior intercostal vein and accessory hemiazygos vein are derived from the left posterior cardinal vein, and this vein simultaneously forms the upper part of the azygos vein. The part connecting the azygos to the hemiazygos vein is the remainder of the anastomosis between the left and right posterior cardinal veins.

13.4 Anomalies

13.4.1 Absent Azygos Vein

Total absence of the azygos vein occurs rarely when the right cranial aspect of the supracardinal vein fails to develop. Drainage of the right and left intercostal vein in this case happens through hemiazygos and accessory hemiazygos veins where the accessory hemiazygos vein converges with the left BCV through the left supreme intercostal vein. Resultant/expected enlargement of the hemiazygos, accessory hemiazygos, and left superior intercostal veins will be seen in these patients with no clinical symptoms encountered (Demos et al. 2004).

13.4.2 Azygos/Hemiazygos Lobe

Anomalous course of the azygos vein in the right lung apex gives rise to azygos lobe in 0.4–1% of the population, encountered on about 0.4% of chest radiographs and 1% of anatomic specimens. It occurs when the right posterior cardinal vein, one of the precursors of the azygos vein,

fails to migrate over the apex of the lung and penetrates it instead, carrying along pleural layers that entrap a portion of the right upper lobe (Demos et al. 2004; Mata et al. 1991; Kim et al. 1995). This anomaly is readily identifiable on imaging studies, typically plain radiographs and CT, as the abnormally located vessel indents the lung and overlying visceral and parietal pleural layers, resulting in four layers of pleura that form a “mesentery-like” structure, the meso-azygos, which contains the azygos vein, seen coursing through the right upper lobe, located more cephalad than normal. The enclosed lung between the fissure and the mediastinum is not an independent segment as its bronchial and arterial supplies arise from the apical or posterior segments of the right upper lobe (Mata et al. 1991). The fissure is visible as a fine line with right convexity on chest X-ray with the azygos vein visible as a tear-shaped opacity in the lowermost part of the fissure. The azygos arch is not visible at its usual location at the right tracheobronchial angle (Kim et al. 1995). Much less commonly seen is a left azygos or hemiazygos lobe caused by malpositioned left superior intercostal vein draining into the left BCV (Fig. 18).



Fig. 18 Azygos fissure in the right upper lobe. The azygos vein is seen as a linear or tubular density in the right upper lobe lateral to the trachea

13.4.3 Azygos and Hemiazygos Continuation of the IVC

Embryogenesis of the IVC is one of the most complicated sequences as several vessels must develop and regress in turn for it to form normally. In utero, the vessels that form the azygos and hemiazygos veins normally communicate with the suprarenal IVC, but this communication breaks down in the typical scenario. If it does not, azygos or hemiazygos continuation of the IVC is said to be present. These lesions may be isolated or associated with other congenital anomalies. The incidence in patients with CHD is 0.2–1.3% (Demos et al. 2004).

13.4.4 Azygos Continuation

Azygos continuation of the IVC (ACIVC) is a sporadic congenital anomaly characterized by agenesis of the hepatic tract of the IVC and posterior redirection of blood through the enlarged azygos vein that courses the right edge of the spine and drains into the SVC. This anomaly is common in patients with polysplenia (left isomerism) and rarely seen in patients with asplenia (right isomerism). Other associated anomalies include abnormal abdominal situs and left or duplicated IVC. At imaging, dilation of the azygos vein, azygos arch, and SVC is seen secondary to increased flow, and the hepatic veins drain into the right atrium via a suprahepatic IVC. The hepatic segment of the IVC is absent or hypoplastic, and this condition must be documented to exclude other causes of an enlarged azygos vein. Azygos continuation has also been reported in association with azygos lobe (Fig. 19) (Demos et al. 2004).

13.4.5 Hemiazygos Continuation

Hemiazygos continuation of the IVC (HAIVC) is an even rarer anomaly where the large venous blood vessel courses the left side of the spine. In some cases, the enlarged HV drains into the SVC. Hemiazygos continuation of a left-sided IVC has several variations including three possible routes for blood in the hemiazygos vein to reach the right atrium. Most commonly, the hemiazygos vein drains into the azygos vein at T8–T9 (in this case the findings at more cephalad levels are simi-

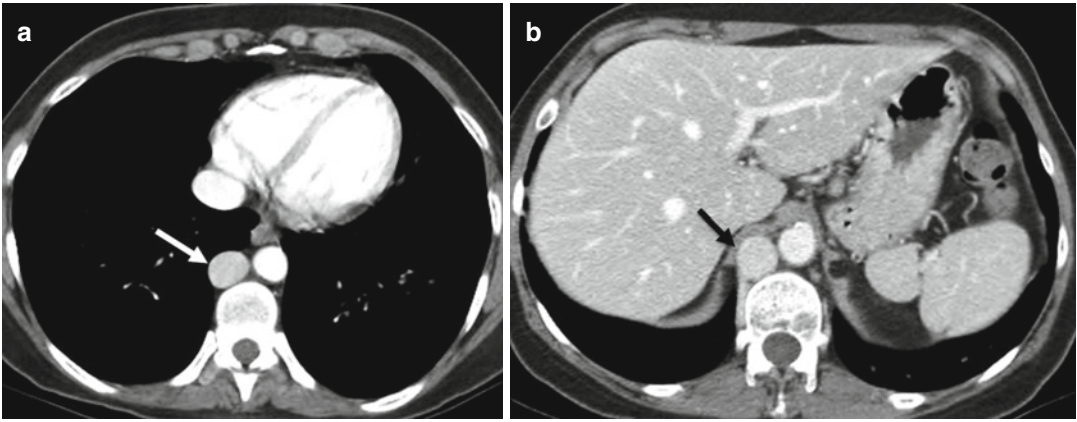


Fig. 19 Azygos continuation of the inferior vena cava. Contrast-enhanced CT axial images in the chest (a) and abdomen (b) show the dilated azygos vein to the right of the aorta and anterior to the spine (arrows). Note the absence of the intrahepatic segment of the IVC in the upper abdomen

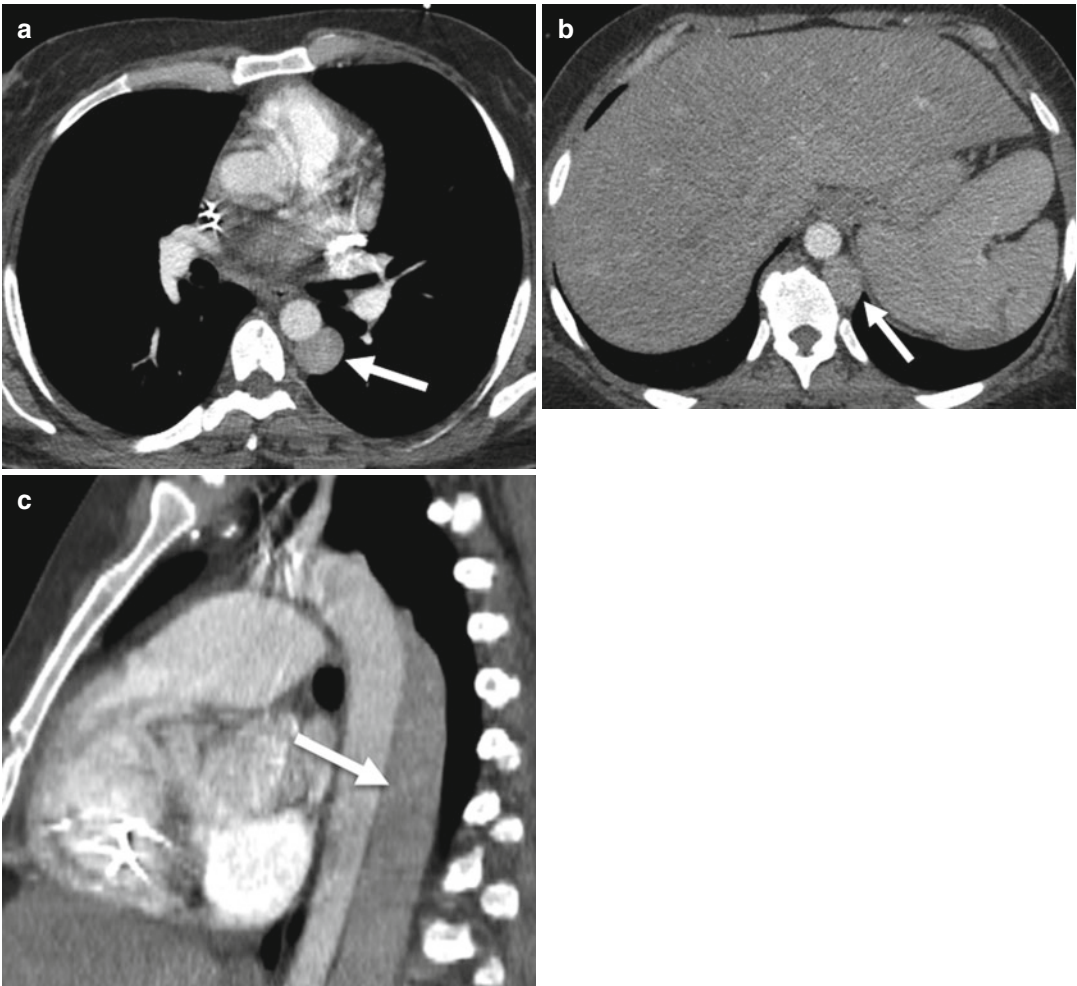


Fig. 20 Intrahepatic interruption of the inferior vena cava with hemiazygos continuation. (a, b) Contrast-enhanced CT axial images at the level of the lower thoracic region and upper abdomen demonstrate a dilated hemiazygos vein posterolateral to the descending aorta (arrows). (c) Left sagittal reconstruction shows the hemiazygos vein behind the descending thoracic aorta

lar to azygos continuation with enlargement of the distal azygos, and hemiazygos vein is also enlarged). Second, a persistent LSVC with blood flowing from the hemiazygos into the accessory hemiazygos vein and left SVC and then into the coronary sinus, all of which are dilated, and finally may see hemiazygos vein drain into the accessory hemiazygos vein, left superior intercostal vein, and left BCV into a normal right SVC (Fig. 20) (Demos et al. 2004; Kim et al. 1995; Bass et al. 2000).

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