

# Venoatrial Abnormalities

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## Abstract

Congenital anomalies of thoracic veins can be subdivided into systemic and pulmonary anomalies. This group of pathologies is not uncommon. The most common systemic thoracic venous anomalies are: persistent left superior vena cava, retroaortic left brachiocephalic vein, and azygos continuation of inferior vena cava. The pulmonary venous anomalies can be separated into partial and total anomalous returns. They have a broad spectrum of presentations and can be associated with other congenital heart malformations. Most systemic venous anomalies are incidental findings whereas pulmonary venous malformations can have a relatively benign course like partial anomalous pulmonary venous return to a more severe critical disease like an obstructed pulmonary venous return. Also, total anomalous pulmonary venous return induces cyanosis and should be corrected surgically as soon as possible. Although echocardiography remains the initial noninvasive imaging modality, an accurate characterization and diagnosis of these anomalies can be obtained by CT or MR imaging. However, MRI stays the cross-section modality of choice because, in addition to providing a comprehensive assessment of the cardiac anatomy, it can accurately quantify shunts, ventricular size, and function.

## Keywords

Pulmonary • Systemic • Thoracic congenital vein

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1 Introduction

Congenital anomalies of the thoracic veins may go unnoticed until adulthood. Most of the time, they are recognized incidentally as part of an examination for a totally unrelated indication. However, these abnormalities require characterization as they may be associated with other malformations and have important clinical or surgical implications.

The chapter reviews and illustrates the normal and abnormal anatomy/venoatrial connections of the thoracic veins, which are separated into systemic and pulmonary anomalies. It also covers the postsurgical sequelae and imaging strategies.

1.1 Imaging Strategies

Patients with congenital anomalies of the thoracic veins can be evaluated using catheter angiography, echocardiography, computed tomography (CT), or magnetic resonance (MR) imaging. Despite the fact that echocardiography permits a noninvasive evaluation, this modality of investigation has limitations such as poor acoustic window and poor depiction of extravascular structures. In the past, catheter angiography played a major role, but nowadays it is currently reserved for vascular interventions and hemodynamic evaluations. Presently, the anomalies are assessed using CT or MRI with well-known advantages and limitations for each modality (Türkvtan et al. 2017; Hellinger et al. 2011; Ruano et al. 2015; White 2000; Rahmani and White 2008; White et al. 1997; Mueller et al. 2015).

Multidetector contrast-enhanced CT with reconstructions can be realized with or without cardiac gating. Acquisitions without gating permit a good evaluation of both systemic and pulmonary thoracic veins anatomy. However, the intracardiac anatomy, for example, associated with atrial (ASD) or ventricular (VSD) septal defects, is less precise on CT. The counterpart of that technique is the higher radiation dose.

MRI evaluation of the thoracic veins can be done with different sequences, with and without gadolinium (Table 1). MRI is a good imaging modality to assess both the venous anatomy and

Table 1 Institutional cardiovascular MRI protocol for thoracic venous assessment

1. Localizers through the thorax images
2. Anatomic examination with half Fournier shot turbo spin echo (HASTE) or balanced steady state free precession (SSFP) according to heart rate in axial and coronal plane of the thorax
3. Four-chamber SSFP cine stack views including the atrial septum
4. Short axis SSFP cine stack views including the atrial septum
5. Multiplanar oblique SSFP cine views on the abnormal veins (perpendicular views on each selected structure)
6. Qp/Qs evaluation by flow mapping on:
(a) Left pulmonary artery
(b) Right pulmonary artery
(c) Main pulmonary artery
(d) Ascending aorta
7. Gadolinium enhanced 3D MRA acquisition of the thorax in coronal plane, with multiple phases acquisition

the associated cardiac anomalies. By both cardiac function and flow assessment, MRI can precise the pulmonic-to-systemic flow ratio (QP/QS) and the eventual effect of the anomalies on the cardiac chambers size and function. In addition to be time consuming, MRI have well-known contraindications that preclude that technique (Mueller et al. 2015).

2 Systemic Thoracic Venous Anomalies

2.1 Normal Anatomy

Normal systemic thoracic venous anatomy for the upper part of the body consists of bilateral subclavian and brachiocephalic veins draining into the right atrium via the right superior vena cava (RSVC). The RSVC courses along the right mediastinum. The azygos vein, its major tributary, travels along the right anterior borders of the thoracic vertebrae up to the level of the carina and then arches over the right tracheobronchial angle to drain posteriorly into the RSVC. The coronary sinus travels in the left atrioventricular groove, receives mainly the great cardiac veins, and drains into the right atrium. The inferior vena

cava (IVC) receives systemic venous drainage from the legs, retroperitoneal viscera, and the hepatic circulation, and drains into the inferior surface of the right atrium.

## 2.2 Congenital Anomalies

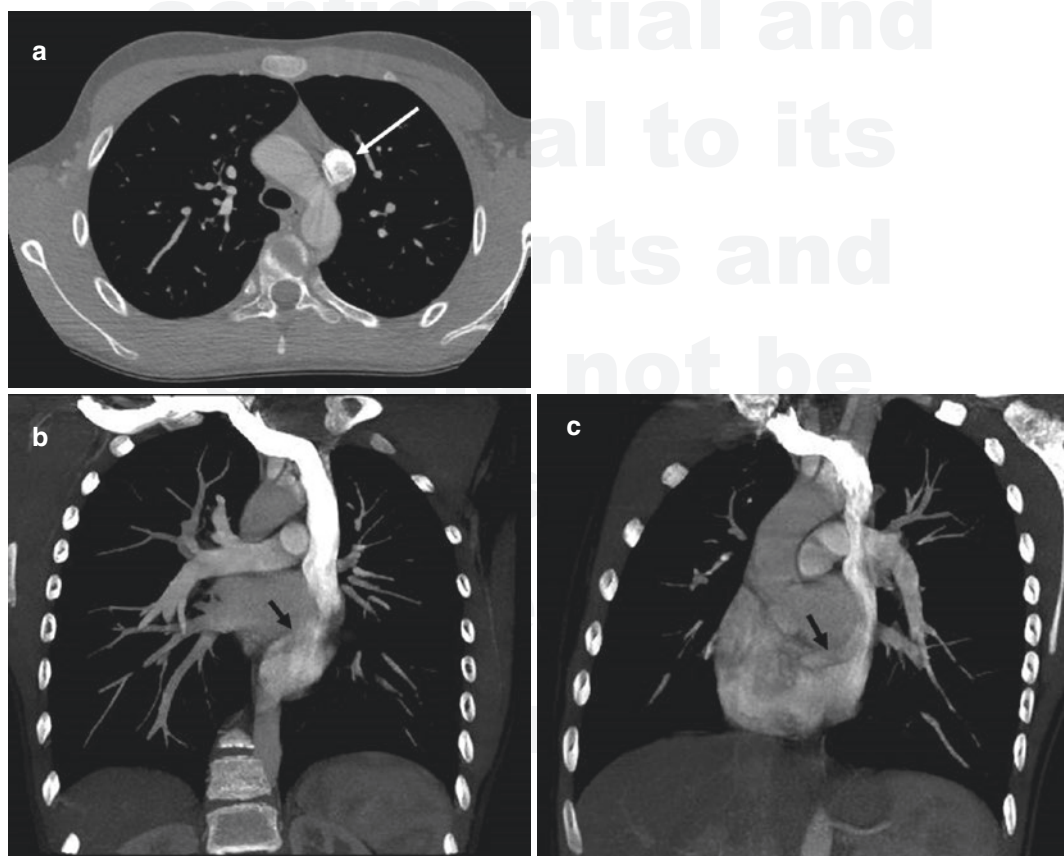
Many anomalies of the systemic veins are asymptomatic but can co-exist with other more serious congenital heart diseases (CHD).

### 2.2.1 Persistent Left Superior Vena Cava (LSVC)

A persistent LSVC, the most common congenital thoracic venous anomaly, is described in 0.3% of the general population with prevalence increasing to 4.3% in patients with congenital heart dis-

ease (Gonzalez-Juanatey et al. 2004; Biffi et al. 2001). The commonly associated anomalies include septal defects, aortic coarctation, and anomalous pulmonary venous return (Cha and Khoury 1972). It is frequently an incidental finding in an asymptomatic patient. Documentation of the anatomy is necessary because it can interfere with the placement of several devices such as central venous catheters, cardiac pacemakers, and defibrillator leads. It may have some surgical implications, namely artery bypass surgery (Gonzalez-Juanatey et al. 2004; Biffi et al. 2001).

In most patients (92%), the LSVC drains normally into the right atrium through an enlarged coronary sinus (Gonzalez-Juanatey et al. 2004). No RSVC is detected in 10–18% of cases (mirror image) (Fig. 1). A dilated coronary sinus, especially with an absent RSVC,

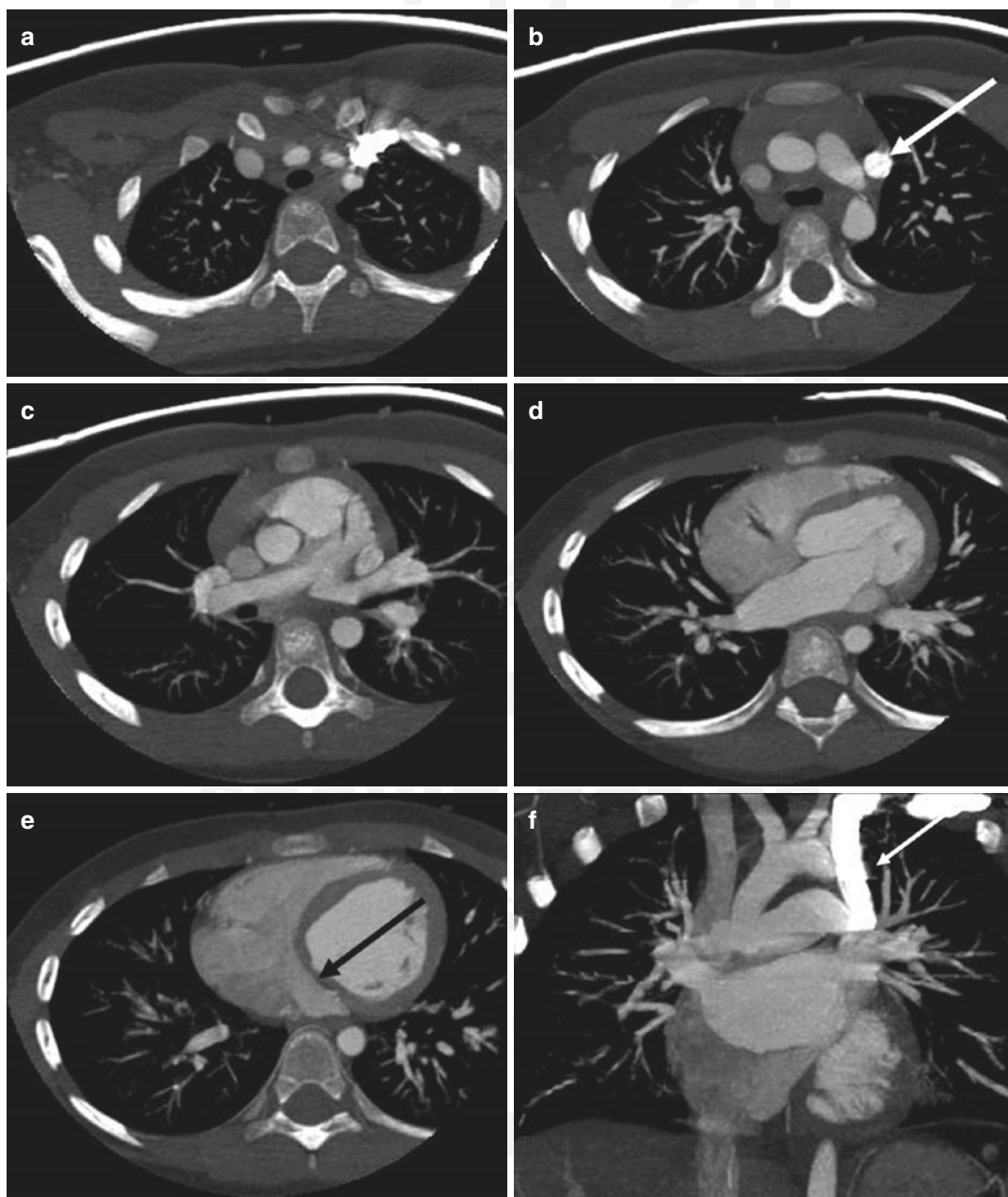


**Fig. 1** Persistent left superior vena cava (mirror image) at CT angiography. (a) Axial, (b) coronal oblique, and (c) sagittal oblique thin maximum intensity projection images

demonstrate a persistent left superior vena cava (white arrow) draining into an enlarged coronary sinus (black arrows) and an absence of right superior vena cava

can cause stretching of the atrioventricular node and the bundle of His; cardiac arrhythmias have been reported (James et al. 1976). In 65% of patients with persistent LSVC, the left bra-

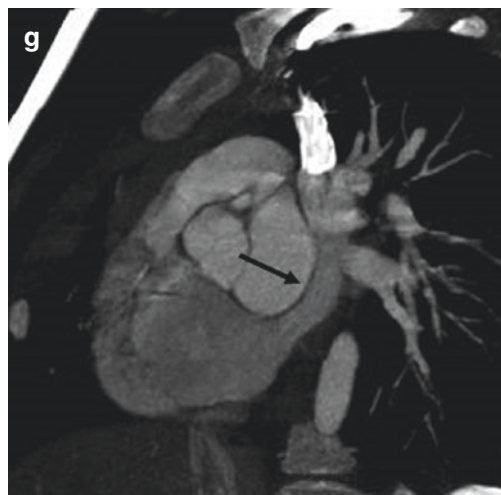
chiocephalic vein is absent (Fig. 2). Even if both vena cava are present, the caliber of the RSVC is reduced (Pretorius and Gleeson 2004). When isolated, the finding of LSVC has no



**Fig. 2** Persistent left superior vena cava at CT angiography. (a)–(e) Axial thin maximum intensity projection (MIP), (f) coronal MIP, and (g) sagittal oblique MIP images demonstrate a persistent left superior vena cava

(white arrows) draining into an enlarged coronary sinus (black arrows), a right superior vena cava, and an absence of left brachiocephalic vein



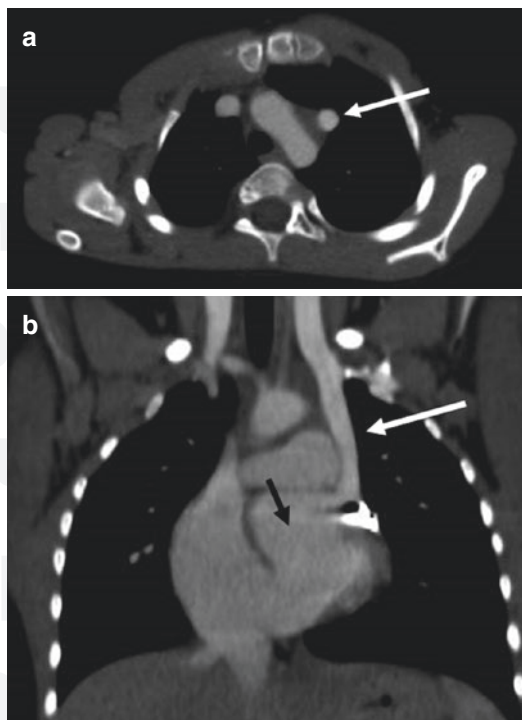


**Fig. 2** (continued)

physiologic consequence and is detected incidentally.

Drainage of LSVC into the left atrium is rare but creates a right-to-left shunt and is associated with significant increased incidence of CHD. Patients are usually asymptomatic but are at risk of right-to-left shunting complications including paradoxical emboli, brain abscess, strokes, and death (Geva and Van Praagh 2008a).

The unroofed coronary sinus can be included in this category because it is almost always associated with a LSVC. In this entity, the common wall between the left atrium and the coronary sinus is partially or completely absent (Fig. 3). The orifice of the unroofed coronary sinus is enlarged and functions like an interatrial communication (Raghib syndrome). Such a left-to-right shunt results in systemic arterial desaturation created by the mixing of LSVC blood with pulmonary venous flow blood to the left atrium. The degree of arterial desaturation is proportional to the net right-to-left shunt, which, in turn, depends on the amount of systemic venous blood carried by the LSVC and the proportion of systemic venous flow that crosses the atrial septum and reaches the pulmonary circulation. Patients with this anomaly are at risk of right-to-left shunting complications like patients with LSVC draining into the left atrium (Geva and Van Praagh 2008a). Treatment of unroofed coronary sinus and LSVC



**Fig. 3** Totally unroofed coronary sinus and persistent left superior vena cava at CT angiography. (a) Axial and (b) coronal thin maximum intensity projection images demonstrate an absence of wall between the coronary sinus and the left atrium (black arrow), a persistent left superior vena cava (white arrows), a right superior vena cava, and an absence of left brachiocephalic vein. The orifice of the coronary sinus is enlarged

into the left atrium, if needed, is surgical or through percutaneous correction. The type of treatment is based on the anatomy and associated abnormalities. Few postsurgical complications are described, usually either stenosis or residual shunt. Given the limited number of published cases, no standardized posttreatment follow-up recommendations are available.

To summarize, the most frequent conditions associated with a LSVC are:

- Single LSVC draining into the coronary sinus (mirror image)
- Single LSVC draining into the left atrium
- Dual SVC with or without left brachiocephalic vein
- Dual SVC or single LSVC with unroofed coronary sinus

### 2.2.2 Anomalies of Right SVC (RSVC)

Isolated anomalies of RSVC are rare. RSVC can drain into the left atrium, have low insertion into the right atrium, or be congenitally dilated (Cormier et al. 1989; Freedom et al. 1982; Modry et al. 1980; Park et al. 1983). Drainage of RSVC into the left atrium results in a right-to-left shunt and can occur without any associated anomalies. Accordingly, patients are at risk of right-to-left shunting complications. An aneurysmal dilatation is an incidental finding and a low insertion is associated to complex CHD.

### 2.2.3 Anomaly of Left Brachiocephalic Vein (LBCV)

Normally, the LBCV joins the RSVC anteriorly to the aorta. With retroaortic LBVC, the vein courses posteriorly to the ascending aorta, underneath the aortic arch, and anteriorly to the main pulmonary artery (Fig. 4). The prevalence of this finding is 0.5–0.6%, and is usually associated with CHD such as Tetralogy of Fallot, truncus arteriosus (Cormier et al. 1989). When isolated, this anomaly does not cause physiologic modifications, but can have some clinical implications at the time of insertion of central venous lines and pacemakers (Hellinger et al. 2011).

### 2.2.4 Anomalies of IVC

The azygos continuation of the IVC, also described as the absence of hepatic segment

of the IVC with azygos continuation, occurs in 0.6% of the general population (Ruano et al. 2015) (Fig. 5). The hepatic veins drain directly into the right atrium. This entity is a classical finding in heterotaxia syndrome with polysplenia (Geva and Van Praagh 2008a). Rarely, the continuation is through the hemiazygos vein with three possible paths of drainage: (1) the azygos vein, (2) a LSVC, and (3) a RSVC (coursing through the accessory hemiazygos vein and LBCV) (Dudiak et al. 1991) (Fig. 6). These venous anomalies do not have any consequences on physiologic circulation but may have clinical implications such as IVC filter placement. Since the advent of cross-sectional imaging, anomalies of IVC are being increasingly found in asymptomatic patient.

Other anomalies like bilateral IVC and IVC drainage into the left atrium are rare.

## 2.3 Postsurgical Sequelae

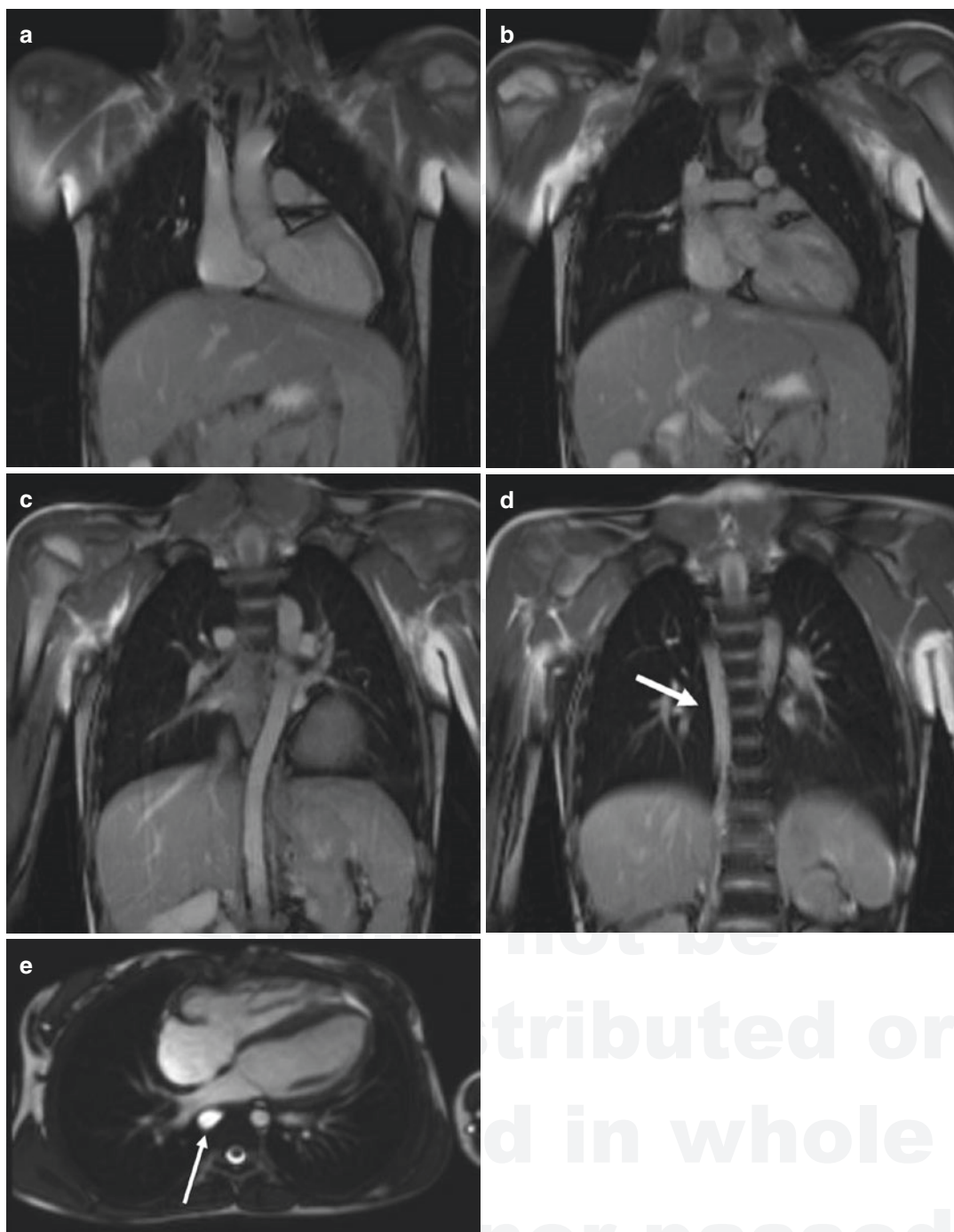
### 2.3.1 Atrial Switch

Prior to the establishment of the arterial switch procedure to treat patients with d-transposition of the great arteries (D-TGA), many patients have been palliated using a Mustard or Senning procedure (atrial switch) (Fig. 7). These surgeries consist of rerouting systemic and pulmonary venous return to the ventricles that were associated with the appropriate great vessels. So, oxygenated blood from the pulmonary veins is redirected to right ventricle, aorta, and systemic circulation. Conversely, the systemic blood from the systemic veins is redirected to left ventricle, pulmonary arteries, and lungs for oxygenation. Persistent and progressive functional cardiac abnormalities have long been recognized following repair and included: residual intra-atrial shunts, caval and pulmonary venous obstructions, right ventricular dysfunction, tricuspid regurgitation, and arrhythmia (Wernovsky 2008).

Residual intra-atrial baffle shunts occur most commonly at the superior right atrial baffle suture lines and may cause either



**Fig. 4** Retroaortic left brachiocephalic vein at CT angiography. Coronal maximum intensity projection image demonstrates the left brachiocephalic vein (black arrow) projective under the aortic arch



**Fig. 5** Interruption of inferior vena cava with azygos continuation (*white arrows*) on (a)–(d) coronal TruFISP and (e) on four-chamber view cine TruFISP MRI sequence images

systemic-to-pulmonary or pulmonary-to-systemic venous shunting. Although trivial leaks have been observed at late postoperative angi-

ography in 10–20% of patients, significant leaks requiring re-intervention have been uncommon (1–2%) (Wernovsky 2008).

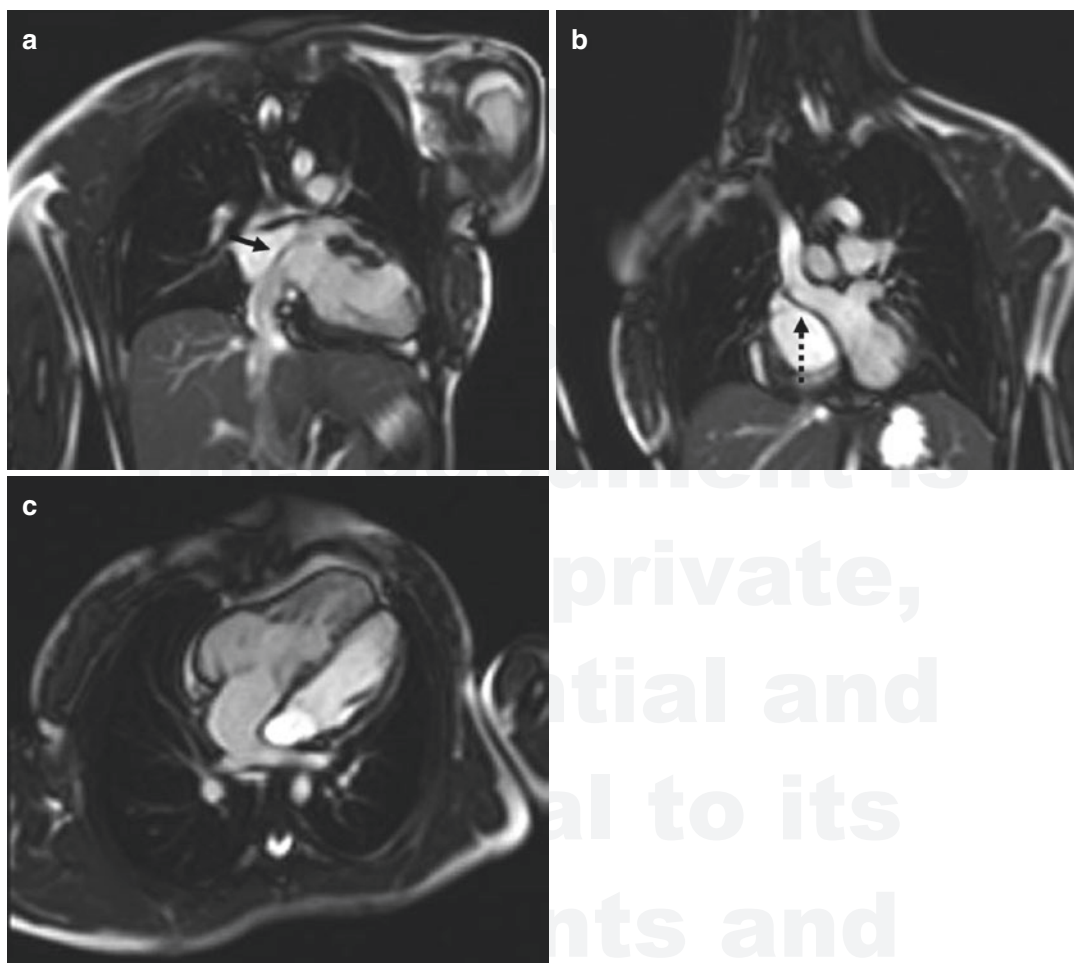


**Fig. 6** (a)–(c) Interruption of inferior vena cava with hemiazygos continuation (white arrow) and drainage into a left superior vena cava (white dotted arrow) on coronal MR angiography

Obstruction of superior vena cava pathway appears postoperatively in 5–10%. The location of obstruction is distal to the superior vena cava entrance, at the site of excision of the superior remnant of the atrial septum (Fig. 8). Despite

severe obstruction, patients can be asymptomatic because of decompression of the RSVC by the azygos/hemiazygos system. Obstruction of IVC is a serious but infrequent complication (1%) (Wernovsky 2008).





**Fig. 7** Atrial switch (Mustard procedure) for D-transposition of great arteries at cardiac MRI. (a), (b) Coronal oblique cine TruFISP MRI sequence images demonstrate drainage of right superior (black dotted

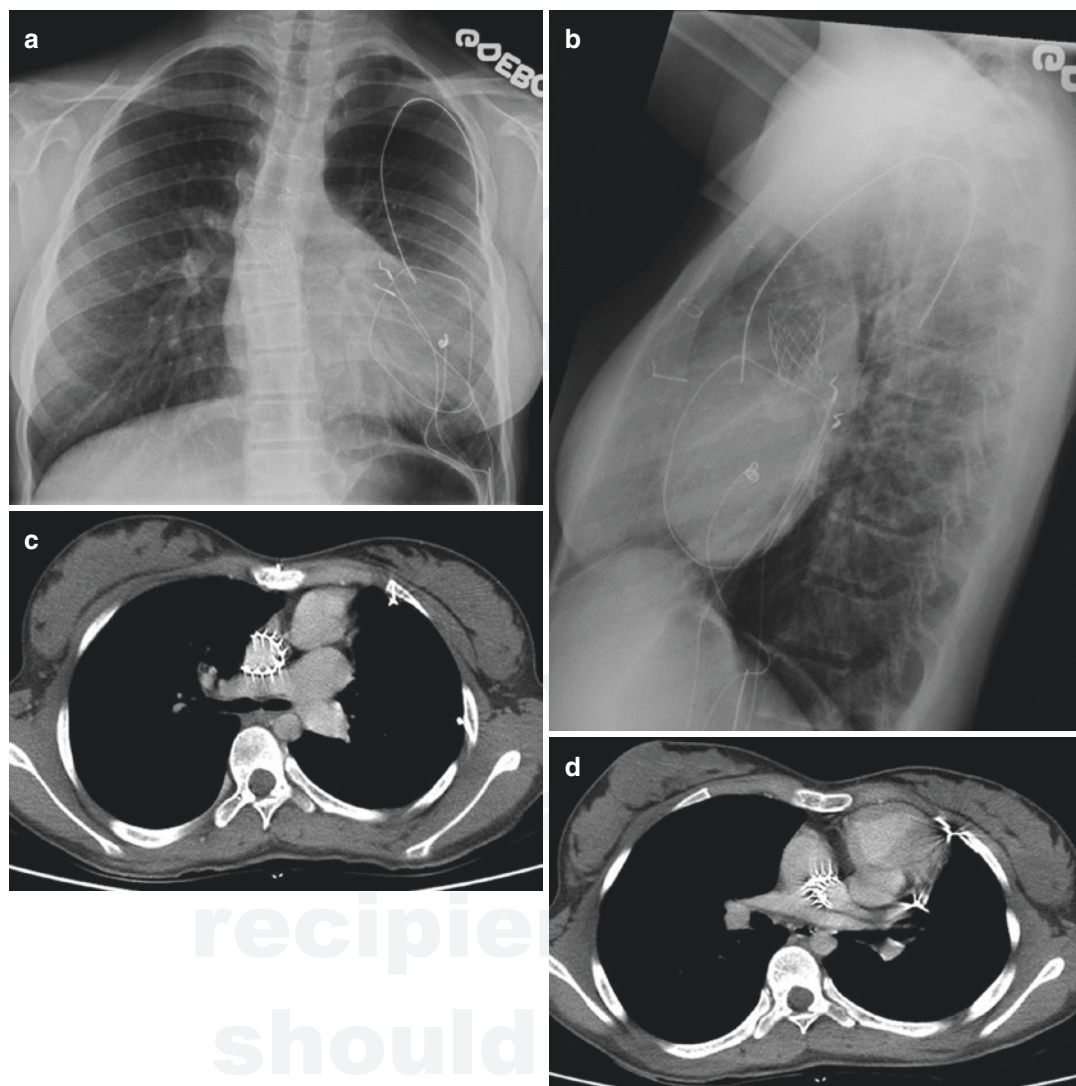
arrow) and inferior (black arrow) vena cava into the left atrium. (c) Four-chamber view cine TruFISP MRI sequence image shows drainage of pulmonary veins into the right atrium and right ventricular hypertrophy

### 3 Pulmonary Venous Anomalies

#### 3.1 Normal Anatomy

In normal conditions, two superior and two inferior pulmonary veins drain into the left atrium. The right superior pulmonary vein drains the right upper and the middle lobes, the left superior drains the left upper lobe including the lingula, and the inferior pulmonary veins drain their respective lower lobes (Porres et al. 2013).

Almost 25% of the population has a deviation from the normal pulmonary venous drainage: common vein, accessory vein, or early branching vein (Türkvtan et al. 2017). Common pulmonary vein occurs when superior and inferior veins join proximally to the left atrium. By definition, a distance of 0.5 cm or larger between the virtual border of the left atrium and the pulmonary venous bifurcation is necessary for diagnosis. This is more common on the left side (14%) than on the right side (2%). Another variation is accessory vein defined by extra-veins draining



**Fig. 8** Atrial switch (Mustard procedure) for D-transposition of great arteries and stent placement for obstruction of superior vena cava pathway on (a) frontal

and (b) lateral chest radiographs; and on (c) and (d) axial contrast-enhanced CT images. Note the presence of epicardial leads of a pacemaker for arrhythmia

into the left atrium. The most frequent is separate drainage of the right middle lobe or superior segment of the right upper lobe with typically narrower atriopulmonary venous junction (Kaseno et al. 2008). Early branching is defined as bifurcation of the pulmonary vein in two or more separate branches within 1 cm of the origin of the left atrium.

All of these variations are incidental findings without any functional repercussion. However, these variations are important to describe in

patient undergoing interventions on the pulmonary veins, such as radiofrequency catheter ablation for atrial fibrillation (Kaseno et al. 2008).

## 3.2 Congenital Anomalies

### 3.2.1 Partial Anomalous Pulmonary Venous Return (PAPVR)

The prevalence of PAPVR in general population is 0.4–0.7% (Porres et al. 2013; Herlong

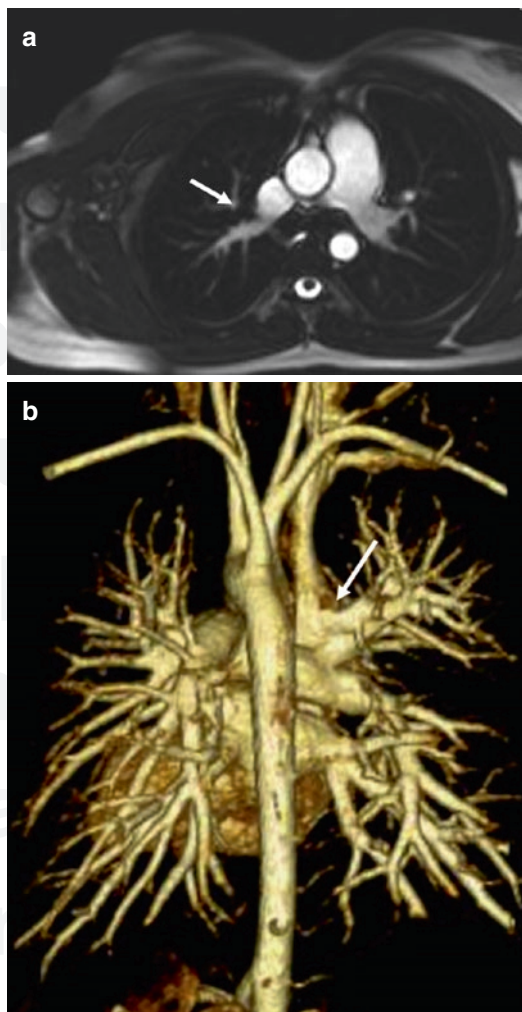
et al. 2000). By definition, at least one, but not all pulmonary veins, drains outside of the left atrium. This anomaly creates a left-to-right shunt similar to atrial septal defect. The majority of patients are asymptomatic during infancy and childhood. The signs and symptoms depend on the magnitude of the shunt, on the hemodynamic change created by the shunt, and on the presence of associated cardiac anomalies (Ruano et al. 2015). A pulmonary-to-systemic flow ratio (QP:QS) of 1.5:1 generally requires surgical repair with low morbidity and mortality. Imaging findings on chest radiography will depend on the configuration of anomalous drainage and the degree of left-to-right shunting.

#### (a) Right Upper Lobe PAPVR

The right PAPVR is twice more frequent than the left. In this condition, the right superior pulmonary vein drains into the RSVC, the azygos vein, the right atrium, or the coronary sinus (Fig. 9). The most common type of PAPVR is right upper lobe drainage into the RSVC below the azygos vein through one large or two or three smaller veins (Geva and Van Praagh 2008b) (Fig. 10). The anomalous pulmonary venous return always involves the right superior pulmonary lobe but the middle pulmonary lobe can also be affected. Usually, the vein from the right lower lobe drains normally into the left atrium. A sinus venosus type of atrial septal defect is highly associated with right upper lobe PAPVR in pediatric population (80–90%) whereas a recent study in adults by Ho et al. showed a moderate association of 47% (Katre et al. 2012; Ho et al. 2009). The cephalic position of sinus venosus atrial septal defect is difficult to diagnose at transthoracic echocardiography. CT angiography and MR imaging are excellent modalities to demonstrate this anatomic region.

#### (b) Left Upper Lobe PAPVR

This anomaly represents the second most frequent type of PAPVR. On adult CT chest examinations, it is the most frequently detected anomalous pulmonary venous connection (Haramati et al. 2003). It is charac-

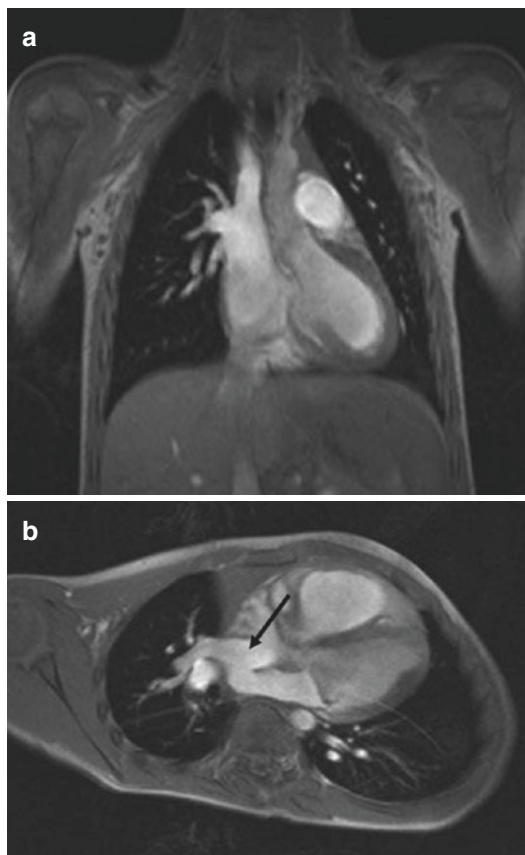


**Fig. 9** Right superior anomalous pulmonary venous return at cardiac MRI. (a) Axial view image obtained from cine FLASH MRI sequence and (b) posterior volume rendering image demonstrate an anomalous drainage of right superior pulmonary vein into the right superior vena cava (white arrows)

terized by an aberrant vertical vessel that conducts blood in a cephalic direction from the left superior pulmonary vein to the LBCV which eventually drains into the normal RSVC (Fig. 11). The anomalous pulmonary venous return always involves the left superior pulmonary lobe or the entire left lung.

The anomalous vessel (the vertical vein) can be confused with the appearance of the LSVC. Both entities can be differentiated on





**Fig. 10** Right superior anomalous pulmonary venous return at MRI. (a) Coronal oblique view and (b) axial oblique view images obtained from cine FLASH MRI sequence demonstrate an anomalous drainage of right superior pulmonary vein through two veins into the right superior vena cava (RSVC) and a sinus venosus type of atrial septal defect (*black arrow*). The anomalous drainage involves the right superior and middle pulmonary lobes. Note the drainage into RSVC is below the azygos vein

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 intra-parenchymal upper lobe vessels connect with the anomalous vein; (2) 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; (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 (Ruano et al. 2015; Katre et al. 2012; Maldonado et al. 2010; Dillon and Camputarò 1993).

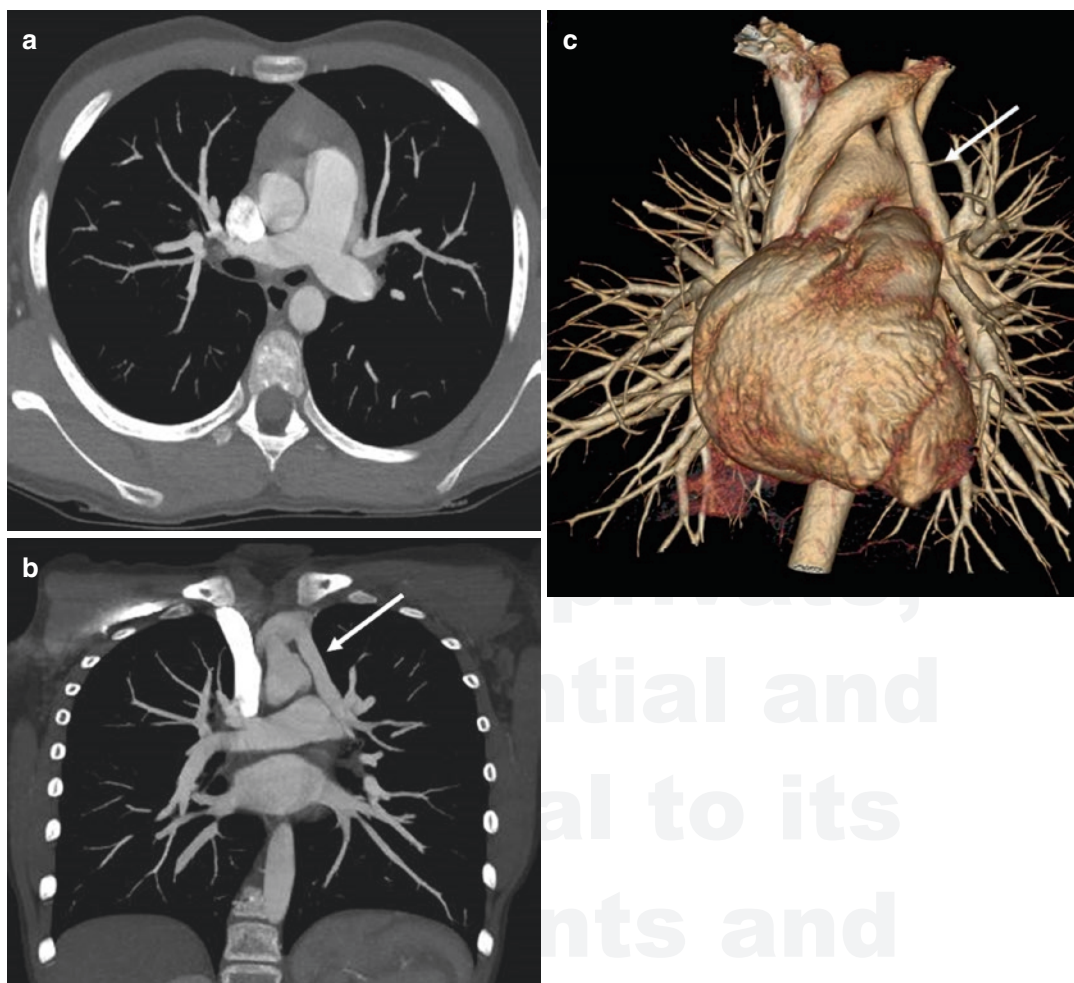
### (c) Scimitar Syndrome

Scimitar syndrome or pulmonary venobar syndrome is a rare and complex form of PAPVR that almost exclusively involves the right lung (Türkvan et al. 2017). This anomaly is characterized by an anomalous pulmonary vein draining a portion or the entire right pulmonary venous flow to the supradiaphragmatic or infradiaphragmatic IVC. The crescent-shaped abnormal vein descends in a caudal direction toward the diaphragm. The radiologic appearance of this anomalous vein resembles a curved Turkish sword or “scimitar” from which the name of this condition is derived (Gudjonsson and Brown 2006). The anomalous right pulmonary vein usually drains the entire right lung but rarely may drain only the middle and lower lobes (Katre et al. 2012). It is associated most of the time with hypoplasia of the right lung and right pulmonary artery, and cardiac dextro-position (Fig. 12). The right lung may have abnormal lobation with only two lobes (Konen et al. 2003). The atrial septum is usually intact (Geva and Van Praagh 2008b). An anomalous arterial supply to the right lower lung from the aorta or its branches is also frequent and often supplies pulmonary sequestration. This late finding is more commonly seen in infancy.

The presentation of the syndrome can be incomplete or complete. The scimitar syndrome is usually divided into three main forms: infantile form with symptoms and pulmonary hypertension, an “older” adult form, which is typically asymptomatic in infancy, and a form with associated CHD (Vida et al. 2010).

Goodman and colleagues described an entity closely related to scimitar syndrome: meandering pulmonary vein or pseudo-scimitar





**Fig. 11** Left superior anomalous pulmonary venous return at CT angiography. (a) Axial, (b) coronal thin maximum intensity projection, and (c) anterior volume ren-

dering images demonstrate an anomalous drainage of left superior pulmonary vein into the left brachiocephalic vein through an aberrant vertical vessel (white arrows)

syndrome (Goodman et al. 1972). It consists of an anomalous right pulmonary vein that courses through the right lung presenting as a scimitar-shaped structure on the chest radiograph, but ultimately drains into the left atrium (Goodman et al. 1972; Lee 2007). Most reported cases of pseudo-scimitar syndrome or scimitar variant connect to the left atrium and IVC simultaneously (Lee 2007; Mohiuddin et al. 1966; Morgan and Forker 1971; Gazzaniga et al. 1969; Pearl 1987; Tumbarello et al. 1991) (Fig. 13). Either surgery or transcatheter endovascular treat-

ments can be performed to close this dual venous drainage anomaly depending on individual anatomy.

#### (d) Other

Anomalous dual pulmonary venous drainage can exist as a PAPVR equivalent described with levoatrial cardinal vein (Türkvtan et al. 2017) (Fig. 14). The levoatrial cardinal vein connects the left atrium or one of the pulmonary veins to a systemic vein, usually the left innominate vein. In this situation, a portion of the lung drains into the left atrium and a systemic vein. The direction of the flow is predominantly

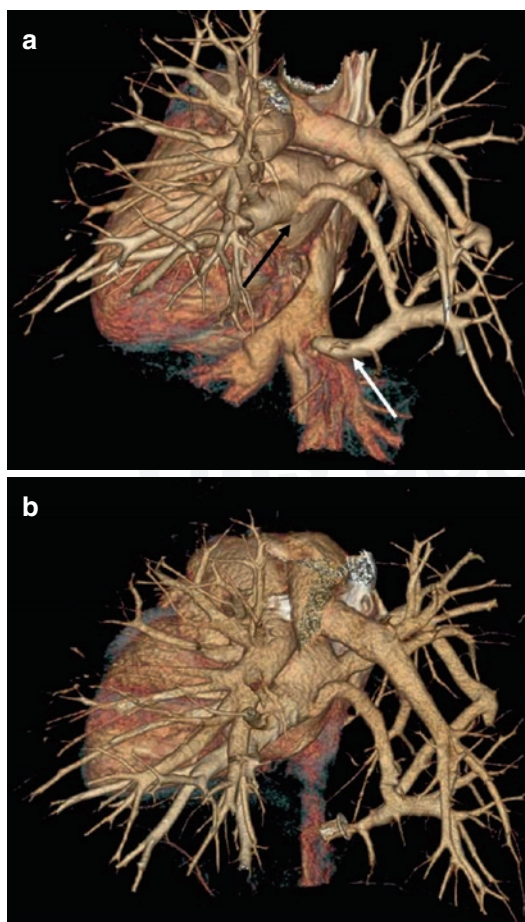


**Fig. 12** Scimitar syndrome. (a) Frontal chest radiograph shows hypoplasia of right lung, cardiac dextroposition, and a crescent-shaped abnormal vein (*black arrow*). (b) Coronal oblique view image obtained from cine FLASH

MRI sequence demonstrates a right inferior pulmonary vein draining into the inferior vena cava. (c) Coronal volume-rendered CT image helps confirm abnormal bronchial lobation with only two right pulmonary lobes

cephalad away from the left atrium, resulting in a left-to-right shunt. The flow can also be bidirectional, so patients are at risk of right-to-left shunting complications. Transcatheter endovascular treatments have been described (Peynircioglu et al. 2005).

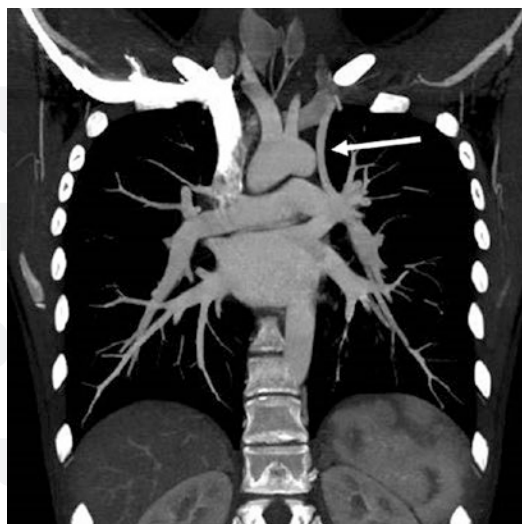
Also, this entity can be associated with obstructive left heart lesions. In this situation, this abnormal draining vein permits the decompression of pulmonary venous blood flow into the systemic venous circulation.



**Fig. 13** Pseudo-scimitar syndrome at CT angiography. (a) Posterior oblique volume rendering image demonstrates a right inferior pulmonary vein draining abnormally into the inferior vena cava (white arrow) and also into the left atrium (black arrow). (b) Posterior oblique volume rendering image from CT angiography realized after percutaneous intervention shows a device occluding the connection between the right inferior pulmonary vein and inferior vena cava

### 3.2.2 Total Anomalous Pulmonary Venous Return (TAPVR)

TAPVR is a rare form of CHD in which all pulmonary veins have no connection to the left atrium. Other cardiac lesions are found in 15% of patients (Seale et al. 2010). TAPVR can occur in patients with heterotaxy syndrome with asplenia. It is classified in four major subtypes based on the level of drainage as follows: type I, anoma-

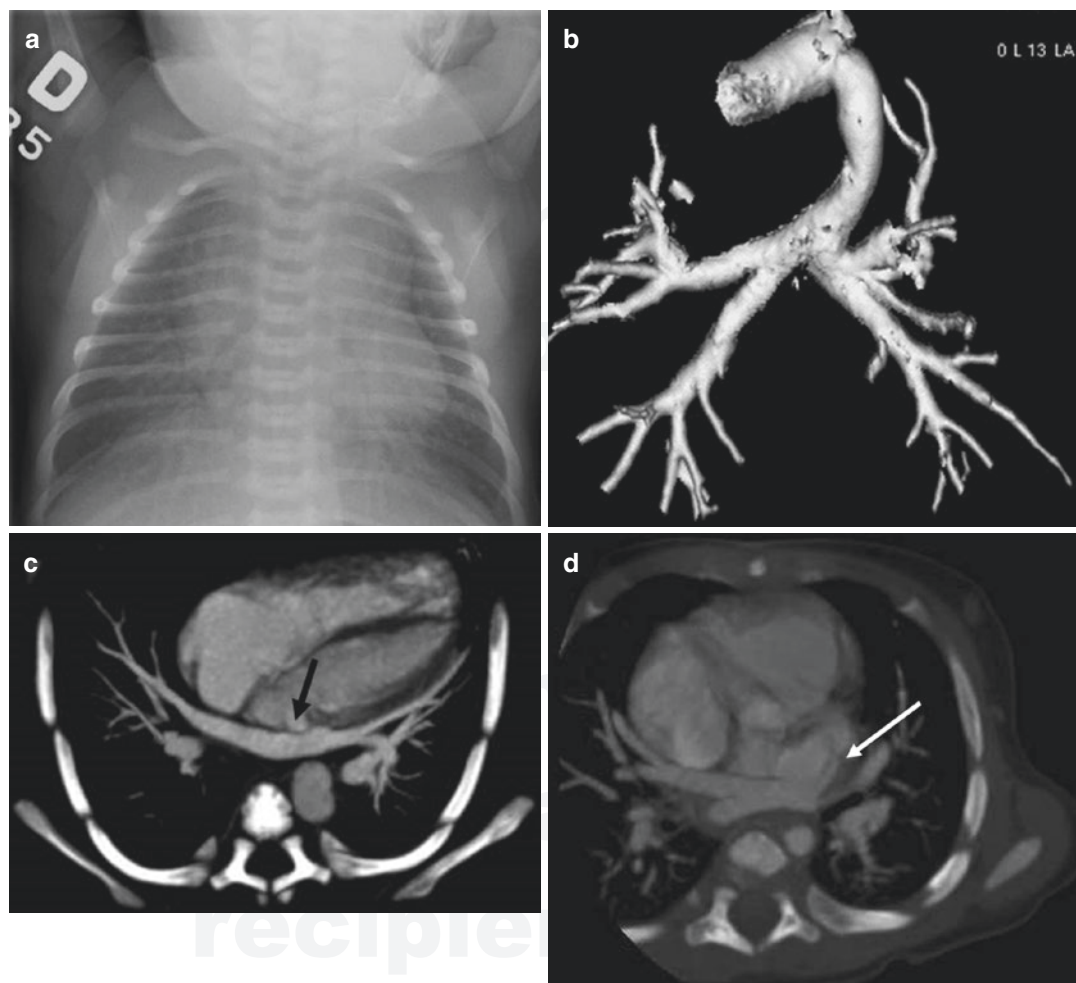


**Fig. 14** Levoatrial cardinal vein at CT angiography. Coronal thin maximum intensity projection image demonstrates a levoatrial cardinal vein (white arrow) connecting the left superior pulmonary vein to the left brachiocephalic vein. The left superior pulmonary vein drains also normally into left atrium

alous connection at the supracardiac level; type II, anomalous connection at the cardiac level; type III, anomalous connection at the infracardiac level; and type IV, anomalous connection at two or more above levels (Geva and Van Praagh 2008b).

The supracardiac type represents the most frequent subtype, 40–55% of cases (Katre et al. 2012). In this type, connection to the left innominate vein is found frequently (Karamlou et al. 2007), giving the classical snowman appearance on chest X-ray (Fig. 15). Other less common supracardiac venous connections include RSVC and azygos veins. In type II, the second most common type, the pulmonary veins connect to coronary sinus or the right atrium (Fig. 16). In type III, the pulmonary veins connect in 80–90% of cases to the portal vein through a common vertical descending vein which courses anterior to the esophagus and crosses the diaphragm at the esophagus hiatus. Rarely, the vertical vein can connect to the ductus venosus, IVC or hepatic veins (Katre et al. 2012). With this type, the





**Fig. 15** Supracardiac type of total anomalous pulmonary venous return (type 1). **(a)** Frontal chest radiograph shows an enlargement of superior mediastinum snowman shape and increased pulmonary arterial vascularization. **(b)** Anterior volume rendering image from CT angiography shows all pulmonary veins draining into common pulmonary vein that connects to left brachiocephalic vein through a vertical vein. **(c)** Four-chamber view thin

maximum intensity projection image demonstrates a posterior position to the left atrium of pulmonary vein confluence (*black arrow*). **(d)** Axial oblique thin maximum intensity projection image from CT angiography realized 10 days after surgical correction shows a large anastomosis between the left atrium and the pulmonary vein confluence (*white arrow*)

anomalous connection is virtually always accompanied by some degree of venous obstruction.

This entity induces cyanosis and an atrial septal defect or patent foramen ovale is necessary to sustain life. Anatomically, for most types of TAPVR, the pulmonary veins from each lung join to form a confluence posterior to the left atrium and then connect abnormally. The patients with such a condition should be corrected surgically as soon as possible.

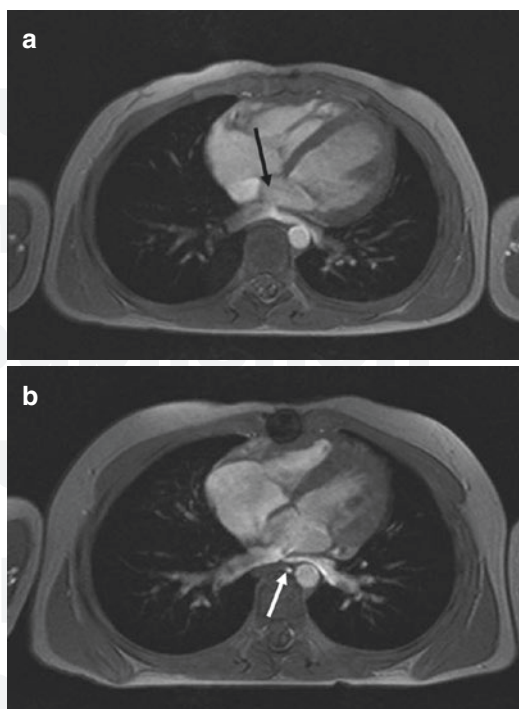
The basis of the correction is the creation of a large anastomosis between the left atrium and the pulmonary vein confluence (Geva and Van Praagh 2008b). The long-term prognostic appears to depend mainly on the state of the pulmonary vascular bed at the time of operation and the adequacy of the pulmonary venous-left atrial anastomosis. Despite an initial satisfactory course, pulmonary venous obstruction (PVO) develops in 5–18%, often within 3 to 6





**Fig. 16** Total anomalous pulmonary venous return into coronary sinus at CT angiography. Posterior volume rendering image shows all pulmonary veins draining into common pulmonary vein (*black arrow*) that connects to coronary sinus (*white arrow*)

months and can be progressive (Seale et al. 2010; Karamlou et al. 2007; Lacour-Gayet et al. 1999; Caldarone et al. 1998) (Fig. 17). Identified risk factors for development of postoperative pulmonary venous obstruction are preoperative hypoplastic/stenotic pulmonary veins and absence of a common confluence (Seale et al. 2010; Caldarone et al. 1998). Estimates of 30-day, 1-year, and 3-year survival for all patients with postoperative PVO were 95.8%, 62.0%, and 58.7%, respectively, which are worse than that of patients without postoperative PVO, in whom 30-day, 1-year, and 3-year survivals were 94.0%, 91.2%, and 91.2%, respectively (Seale et al. 2010, 2013). Risk factors for death included earlier presentation after TAPVC repair, diffusely small pulmonary veins at presentation of postoperative PVO, and an increased number of lung segments affected by obstruction (Seale et al. 2013). According to the study of Seale et al., patients presenting with pulmonary venous obstruction six months after surgical correction had less severe disease (Seale et al. 2013). Postoperative PVO may be a consequence of an inadequate anastomosis between the left atrium and the pulmonary venous confluence, inadequate postoperative growth of the surgical anastomosis, a reaction of

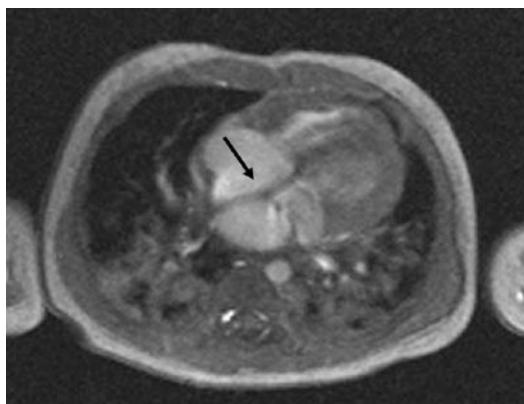


**Fig. 17** Postoperative pulmonary venous obstruction after surgical repair of total anomalous pulmonary venous return at MRI. (a) MRI realized one month after repair. Four-chamber view image obtained from cine FLASH MRI sequence demonstrates a large anastomosis between the left atrium and the pulmonary vein confluence (*black arrow*) and good size of pulmonary veins. (b) MRI realized four months after repair. Four-chamber image obtained from cine FLASH MRI sequence demonstrates a progressive stenosis on the left pulmonary vein near the venoatrial anastomosis (*white arrow*)

tissues to prosthetic material used, or pre/postoperative pulmonary vein hypoplasia/stenosis (Seale et al. 2010; Sano et al. 1989; Whight et al. 1978).

### 3.2.3 Cor Triatriatum

Cor triatriatum is a rare congenital anomaly defined by the presence of a fibromuscular membrane dividing the left atrium into proximal and distal chambers. The proximal (superior and posterior) accessory chamber receives inflow from all four pulmonary veins and the distal (inferior and anterior) main left atrial chamber (including the left atrial appendage and mitral orifice) delivers blood flow to the mitral valve (Figs. 18 and 19). The proximal chamber has been described as

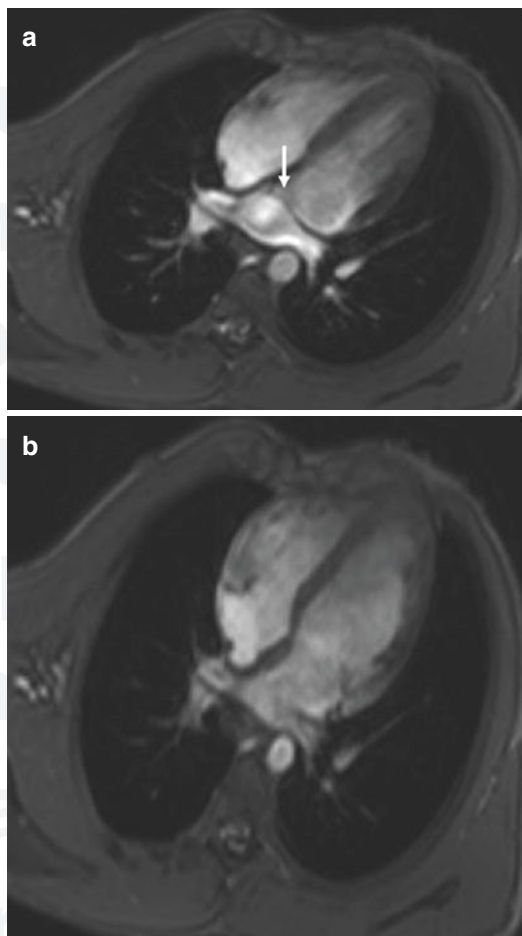


**Fig. 18** Obstructive cor triatriatum at MRI. Four-chamber view image obtained from cine FLASH MRI sequence shows an obstructive membrane (*black arrow*) dividing the left atrium into proximal and distal chambers. Pulmonary parenchymal edema was also noted

common pulmonary vein or third atrium. The reported incidence is 0.1–0.4% (Niwayama 1960).

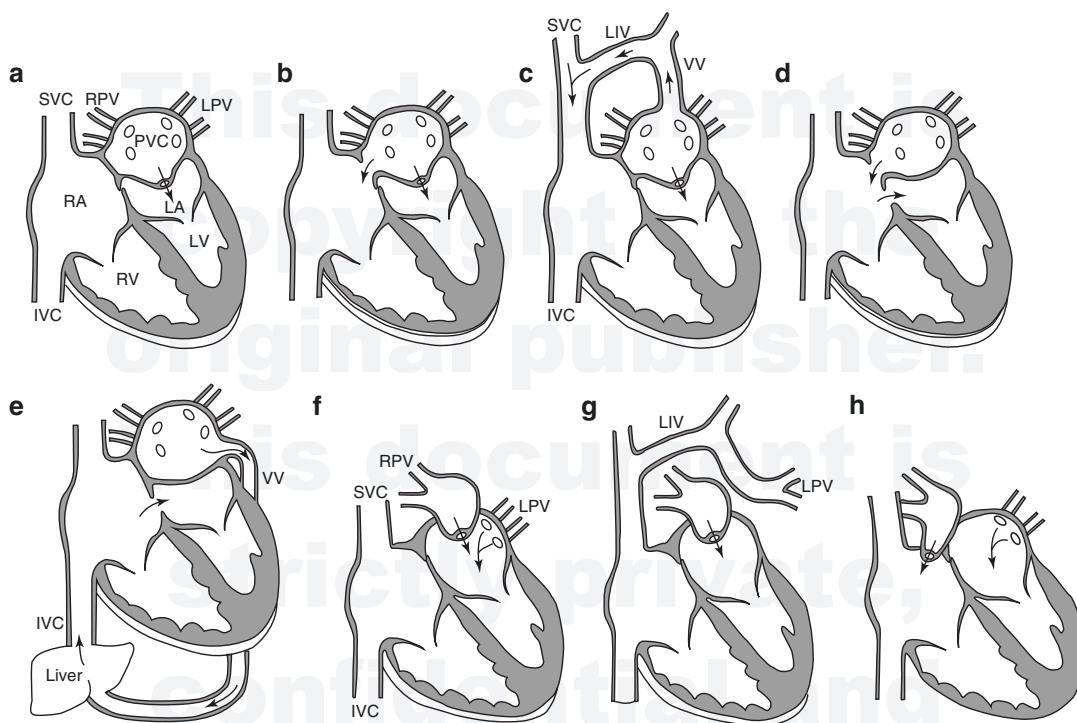
Many classifications exist and depend on the presence of other anomalies (Capdeville et al. 2014) (Fig. 20). The most common congenital defect is atrial septal defect or patent foramen ovale. A secundum atrial septal defect can communicate with either or both chambers, and the foramen ovale almost always communicates with the true left atrium. The other variation coexisting with this entity is total or partial anomalous pulmonary venous return (Kouchoukos et al. 2003). In right-sided partial anomalous return, the right-sided veins join with each other to drain into the anomalous proximal chamber, while left-sided pulmonary veins drain into the left atrium. In left-sided partial anomalous venous return, left-sided veins join to form a left vertical vein which connects to the left innominate vein and the right-sided pulmonary veins connect to the anomalous proximal chamber (Kouchoukos et al. 2003) (Fig. 21).

The natural history depends on the size of the membrane fenestration and the presence or absence of an atrial septal defect, as well as its location (Capdeville et al. 2014). When



**Fig. 19** Non-obstructive cor triatriatum at MRI. (a), (b) Four-chamber view images obtained from cine TruFISP MRI sequence show a membrane (*white arrow*) dividing the left atrium into proximal and distal chambers. During the cardiac cycle, a large opening was demonstrated through the membrane

fenestration is small and no ASD is present, the presentation is typically in infancy. Conversely, if an atrial septal defect communicates with the pulmonary venous chamber, partial decompression can occur, thereby potentially delaying the presentation. Symptoms usually associated with that pathology can be similar to the symptoms of mitral valve stenosis such as dyspnea, orthopnea, and hemoptysis, but some are asymptomatic (incidental finding) (Eichholz et al. 2013). It is



**Fig. 20** Schematic representation of various types of cor triatriatum. The difference between the various types is based mainly on the pulmonary venous return

and the location of the atrial septal defect. *Reprinted with permission from Allen et al. (Geva and Van Praagh 2008b)*

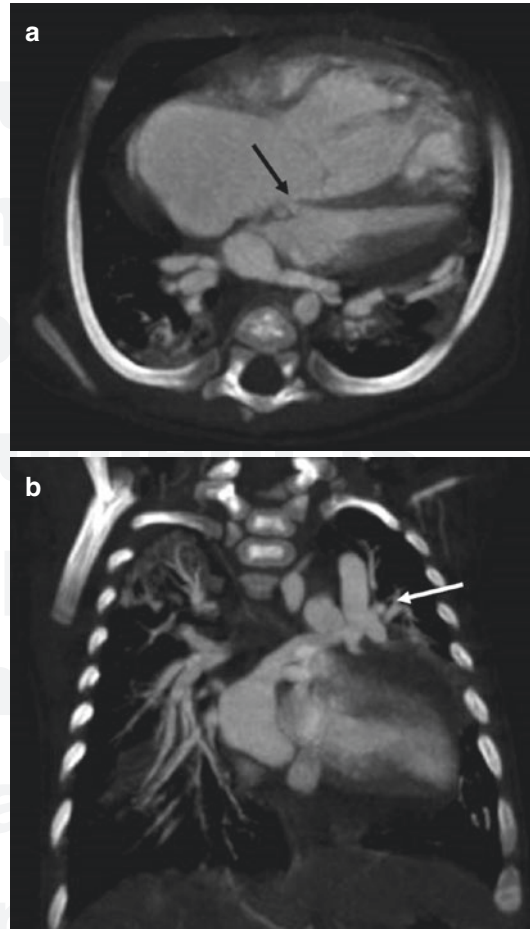
not clear why patients with large fenestration become symptomatic in adulthood. Potential factors include fibrosis or calcification of the membrane's orifice and the development of mitral regurgitation or atrial fibrillation (Eichholz et al. 2013).

### 3.2.4 Congenital Pulmonary Vein Stenosis

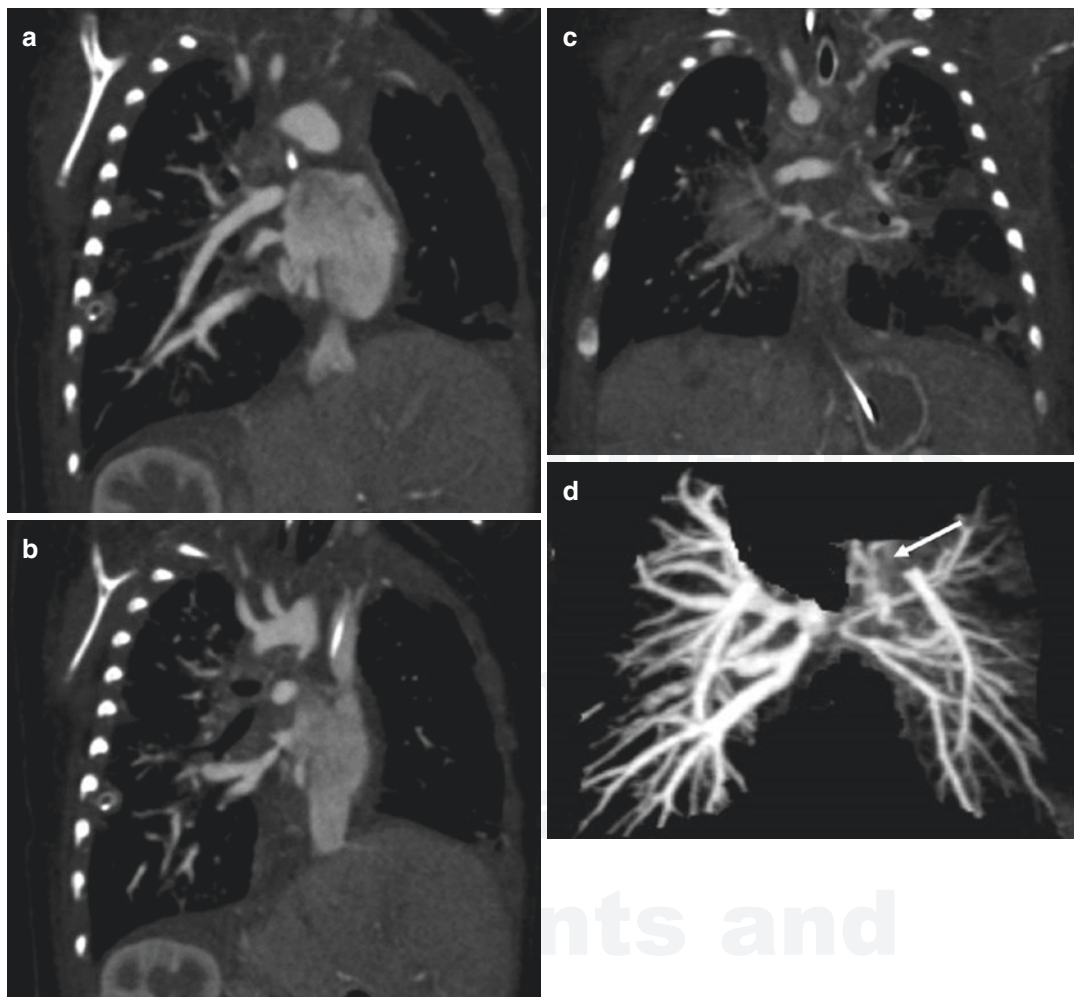
Congenital pulmonary vein stenosis is a rare and severe form of CHD. The congenital pulmonary vein stenosis may represent a hypoplasia of the entire vein or a focal lesion along the course of pulmonary vein (Türkvan et al. 2017) (Figs. 22, 23, and 24). This anomaly can be unilateral or bilateral. It is usually found during the newborn period and often leads to progressive pulmonary hypertension and premature death.

Few cases have been reported in the adult population (Kim et al. 2011; Kapoor et al. 2011; Omasa et al. 2004). No consensus exists regarding the optimal treatment strategy for this anomaly (Devaney et al. 2006; Gordon and Moore 2010; Seale et al. 2006; Peng et al. 2010). Several interventions have been proposed: balloon angioplasty, surgical dilatation, and surgical marsupialization. No statistically significant difference in mortality or re-intervention rate was present among these different therapeutic modalities according to Charlagorla et al. (Charlagorla et al. 2016). Patients with bilateral disease have increased mortality and decreased 5-year survival. According to this same study (the longest follow-up of this pathology to date), the overall survival rate at 58 months is 78% (Charlagorla et al. 2016).

**Fig. 21** Obstructive cor triatriatum and left partial anomalous pulmonary venous return at CT angiography. (a) Axial oblique and (b) coronal oblique thin maximum intensity projection images demonstrate membrane dividing the left atrium and a partial anomalous pulmonary venous return of the left superior pulmonary vein into a vertical vein (*white arrow*). The right pulmonary veins and the left inferior pulmonary veins drain into the anomalous proximal chamber. A small secundum atrial septal defect (*black arrow*) is also present between the right and the left atrium (distal chamber)



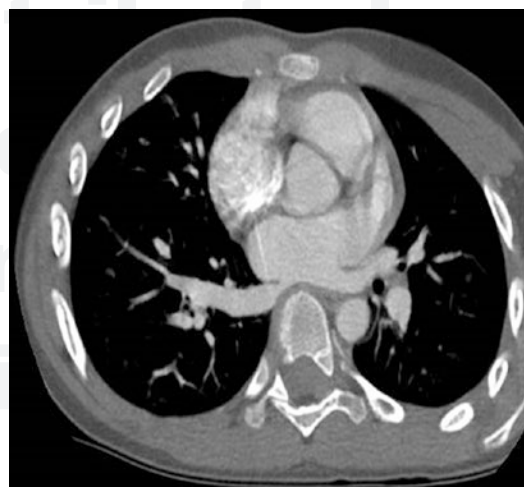


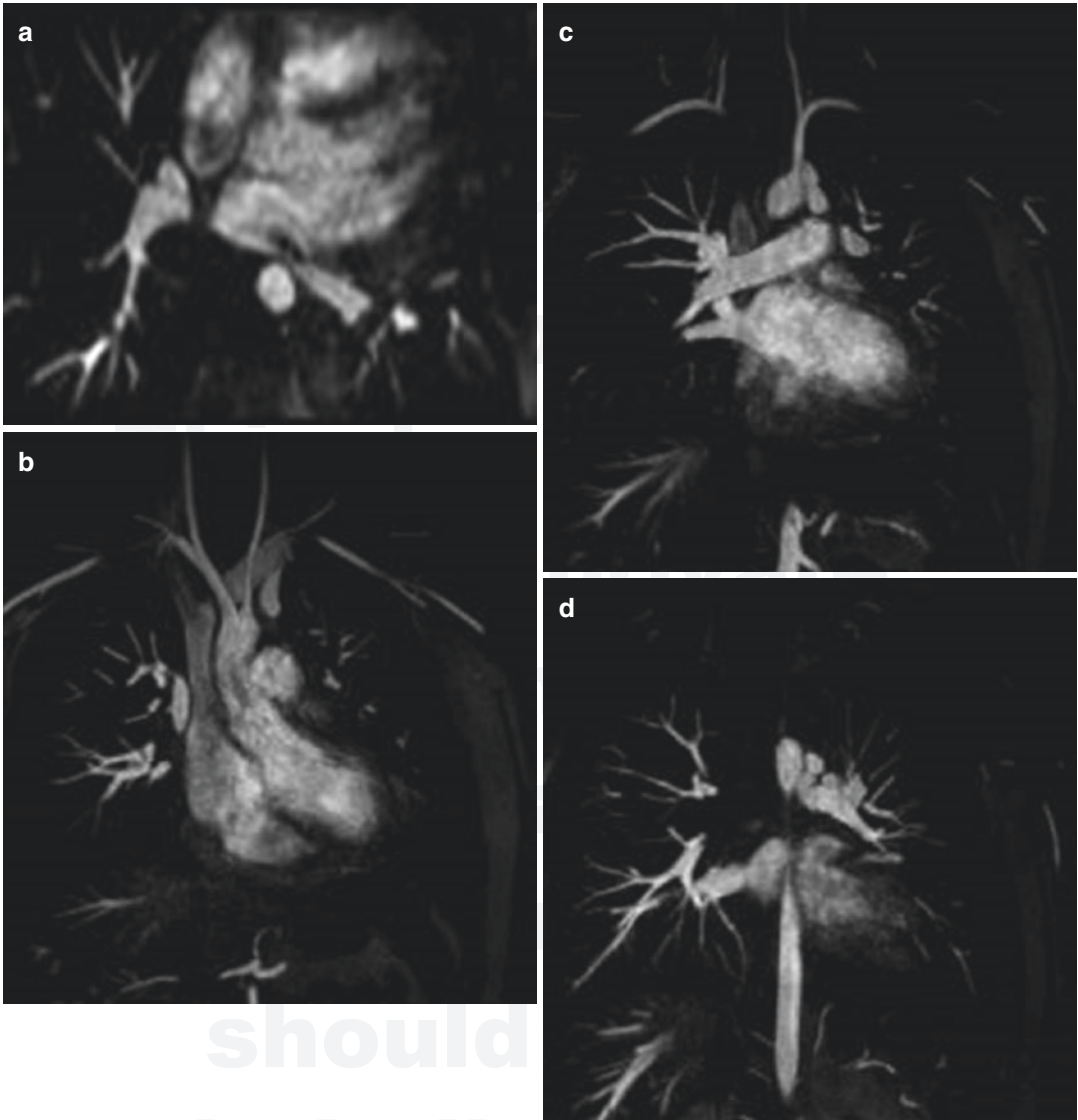


**Fig. 22** Pulmonary vein stenosis at CT angiography. (a)–(c) Coronal oblique thin maximum intensity projection (MIP) images and (d) volume rendering demonstrate hypo-

plasia of the left pulmonary veins and focal stenosis involving all the pulmonary veins at the venoatrial junction. On the left side, venous collaterals are visualized (*white arrow*)

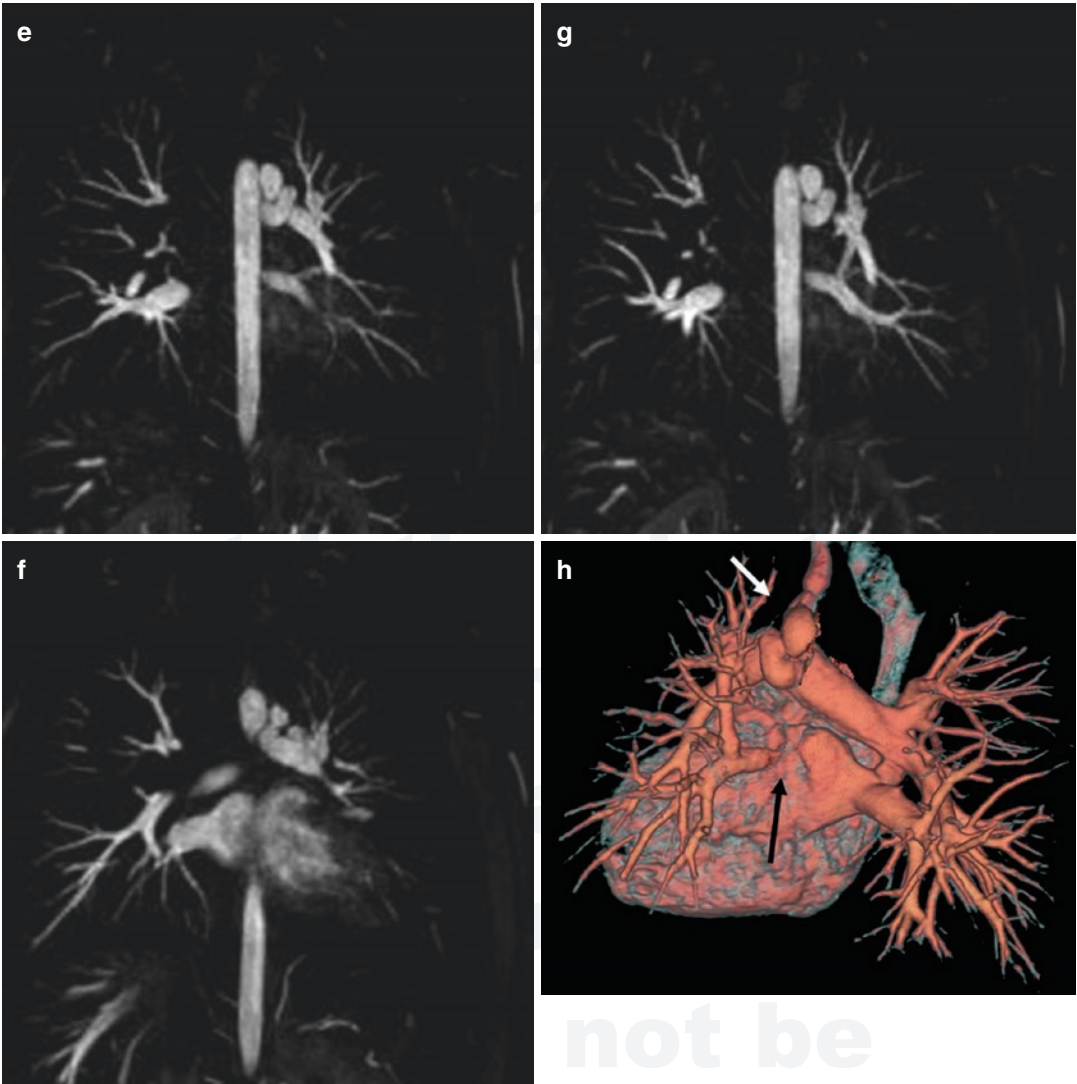
**Fig. 23** Right inferior pulmonary vein stenosis at CT angiography. Axial oblique thin maximum intensity projection images demonstrate a focal stenosis of right inferior pulmonary vein near the venoatrial junction





**Fig. 24** Left pulmonary vein stenosis at cardiac MRI. (a) Axial oblique view image obtained from cine TruFISP MRI sequence shows a severe stenosis involving the left common pulmonary vein. (b)–(g) Coronal thin maximum intensity projection and (h) posterior volume rendering

images from MR angiography demonstrate severe focal stenosis of left pulmonary vein (*black arrow*) with presence of venous collaterals draining into the left brachiocephalic through the left superior intercostal vein (*white arrow*)



**Fig. 24** (continued)

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## Conclusion

Congenital pulmonary and systemic venous anomalies have a broad spectrum and their clinical presentations vary. CT or MR imaging is usually necessary in order to characterize accurately these entities.

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