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Anomalies of the Systemic

and Pulmonary Veins

Lars Grosse-Wortmann and Arno Roest

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Abstract

References.....

The range of pathologies involving the systemic or pulmonary veins is wide. Many of these abnormalities are either clinically insignificant or have been surgically "corrected" during childhood. This chapter focuses on two types of clinical scenarios relevant to the care of adults with congenital heart disease: (1) Venous lesions that may present during or remain untreated until adulthood, and (2) those that are sequelae of previous interventions.

Total Anomalous Pulmonary Venous

Connection.....

Cor Triatriatum.....

Pulmonary Vein Stenosis.....

Overview of Imaging Techniques for Systemic and Pulmonary Venous Anatomy

In many adults with known congenital heart disease the anatomy of the systemic and pulmonary veins has been established through previous imaging and/or direct inspection at the time of prior operations.

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More than a century after the rays that Wilhelm Conrad Röntgen discovered were first used to image the thorax, chest X-ray remains an important imaging modality in congenital heart disease. To the trained eye, chest radiographs reveal many clues to systemic and pulmonary venous anomalies: For example, an abnormal caval vein may produce a characteristic shadow while pulmonary edema can suggest pulmonary venous obstruction. Furthermore, a chest X-ray can reveal important associated cardiac and extracardiac pathologies such as situs anomalies.

Echocardiography is often diagnostic for venous anomalies. It is important to interrogate the entire venous anatomy. On the systemic side, this includes the bridging (so-called innominate) vein and coronary sinus. Bilateral superior caval veins must be searched for routinely. On the pulmonary side, each pulmonary vein and its connection to the left atrium must be demonstrated. Anomalous pulmonary venous connections and obstructions within the left atrium must be carefully ruled out. Color Doppler maps the direction of blood flow and identifies areas of stenosis which are further evaluated by pulsed-wave Doppler.

If echocardiographic images are insufficient, or if information on blood flow is desired, cardiac magnetic resonance (CMR) is often the next applicable imaging modality. All cardiovascular acquisitions, except for conventional contrastenhanced MR angiograms and some real-time applications, are electro- or vectorcardiogramgated. Most centers have abandoned turbo-spin echo ("black blood") techniques in favor of gradient echo ("white blood") sequences. The latter are fast, offer excellent blood tissue contrast and can be performed in multiple phases per cardiac cycle, which are later arranged as cine clips. With a view to shorten scan time and increase signalsteady-state-free-precession to-noise ratio (SSFP) is the preferred pulse sequence for white blood imaging. However, spoiled gradient-echo sequences are occasionally preferred for their robustness towards field inhomogeneities and to visualize flow turbulence. Frequently, even static gradient echo sequences in the three orthogonal planes, obtained as "scouts" or "localizers"

delineate the venous anatomy sufficiently. Information on ventricular volumes, which may be necessary to evaluate lesions that preload an atrial or ventricular chamber, is derived from multiphase ("cine") multislice sequences in the axial or short axis orientation. Three-dimensional (3D) contrast-enhanced magnetic resonance angiography provides a more detailed delineation of the luminal vascular anatomy. As mentioned, the conventional angiogram is a non-gated, either static or time-resolved, acquisition during breathholding. The use of blood-pool contrast agent allows for a slower acquisition, using cardiac gating and diaphragm navigation to reduce blurring from pulsatility and respiration. The latter approach is also feasible with a non-enhanced 3D-SSFP-technique which can be employed when contrast is to be avoided. Phase-contrast flow velocity mapping is part of most protocols for patients with congenital heart disease, including those with venous pathologies. It clarifies the direction of blood flow, quantifies flow volume and maximum flow velocities, and depicts alterations of the expected flow velocity pattern. The use of 3D flow velocity mapping ("4D flow") in venous anomalies is hampered by the low signalto-noise ratio and experimental at present. Computed tomography (CT) is used frequently in adults with congenital heart disease, particularly when CMR is contraindicated, poor CMR image quality is expected, or when pulmonary or mediastinal pathology is to be visualized. Electrocardiographic gating is employed when intracardiac or small vascular structures are the focus of the examination. Typically, CT is performed contrast-enhanced. For the systemic veins in particular, a CT angiogram must be carefully planned: First-pass imaging after injection into an upper-extremity vein often causes beamhardening artifacts in the superior caval vein while delayed imaging may lead to subdiagnostic signal intensity in the veins (Choi et al. 2004). Careful selection of the injection site as well as adjustments to the concentration and timing of the contrast bolus help in minimizing artifacts. Cardiac catheterization is rarely necessary for the diagnosis of isolated venous anomalies, but the systemic and pulmonary veins are shown on

nearly all fluoroscopic angiographies. Changes in sampled blood oxygen saturations or pressure as the catheter passes through the heart are clues to shunts, including anomalous pulmonary or systemic venous connections. Of the nuclear imaging applications only lung perfusion scans for the quantification of pulmonary blood flow are employed with any frequency in congenital or postoperative venous pathologies. With the advent of phase contrast CMR lung perfusion scans have become an infrequently used tool at most centers.

2 Systemic Venous Abnormalities

When abnormalities of the systemic venous system occur in isolation they often cause minimal symptoms and are diagnosed coincidentally in later child- or even adulthood. Many patients do not require an intervention. Many complex types of congenital heart disease, on the other hand, are associated with systemic venous abnormalities. In these situations systemic vein anomalies can be a major determinant of surgical options and outcomes, as is the case in patients with heterotaxy syndrome and single ventricle circulations (Van Praagh and Van Praagh 1990). The development of the systemic veins is complex. As a result, the spectrum of possible deviations from the normal morphogenesis is wide and can not be covered in detail here. This chapter focuses on isolated systemic venous pathologies. Those that are part of complex congenital heart disease such as Fontan circulations are discussed in other chapters of this book.

2.1 Normal and Pathological Development of the Systemic Venous System

An understanding of the development aids in the understanding of pathologies of the systemic veins and, in many cases in the choice, acquisition, and interpretation of diagnostic studies. The precursors of the systemic veins are the cardinal veins, the umbilical veins, and the omphalomesenteric or vitelline veins. These develop as paired structures which drain into the horn of the sinus venosus on either side of the primitive heart. The cardinal veins eventually develop into the superior caval veins while the vitelline veins are the precursors of the hepatic veins and of the suprahepatic inferior caval vein (Smallhorn and Anderson 2010). Septation of the sinus venosus leads to a separation into right and left atrium and, in normal development, isolates the left atrium from the drainage of the cardinal, umbilical, and vitelline veins. The coronary sinus forms from the left horn of the sinus venosus and the left common cardinal vein. It drains the coronary venous blood and, if present, that from a persistent left superior caval vein, to the right atrium.

2.2 Persistent Left Superior Caval Vein and Anomalies of the Coronary Sinus

Of the initially paired superior caval veins the left one normally involutes around week seven of gestation. Persistence of the left superior caval vein occurs in 0.3% of the general population (Sanders 1946), but is much more frequent in individuals with congenital heart disease, particularly Tetralogy of Fallot, atrioventricular septal defect, and left-sided obstructive lesions (de Leval et al. 1975; Cochrane et al. 1994).

2.2.1 Anatomy

A persistent left superior caval vein always drains into the coronary sinus because both structures share a similar embryological origin: The left common cardinal vein. In approximately 10% of patients with a persistent left superior caval vein the coronary sinus is partially or completely unroofed, resulting in a right-to-left shunt (Meadows and Sharp 1965). Conversely, a completely or partially unroofed coronary sinus almost never occurs without a left superior caval vein. In rare instances the left may be the only superior caval vein present (Bartram et al. 1997). In patients with anomalies of the thoracic situs the "morphologically right" superior caval vein may be on the patient's left.

2.2.2 Presentation

In isolated bilateral superior caval veins the presentation depends on whether or not the coronary sinus is intact, delivering the desaturated blood from the left superior caval vein to the right atrium. In severe cases an aneurysmally enlarged coronary sinus can cause obstruction to mitral inflow, leading to a clinical picture similar to cor triatriatum or pulmonary vein stenosis (below) (Agnoleti et al. 1999; Tham et al. 2007). A coronary sinus aneurysm is associated with Wolff–Parkinson–White syndrome (Guiraudon et al. 1988).

If the coronary sinus is deficient a bidirectional shunt results: Desaturated blood from the persistent left superior caval vein and the coronary veins enters the left atrium while pulmonary venous blood reaches the right atrium via the coronary sinus ostium. The degrees of heart failure (from the left-to-right shunt) and cyanosis (from the right-to-left shunt) depend on the presence and size of a left superior caval vein, on the presence or absence of bridging innominate (or left brachiocephalic) vein, and on the size of the coronary sinus defect. Rarely, the right-to-left shunt is sufficient to manifest in cyanosis.

2.2.3 Diagnosis

The majority of individuals with bilateral superior caval veins (and most other anomalies of the superior caval veins and coronary sinus) are asymptomatic. A persistent left superior caval vein must be differentiated from other veins in the left mediastinum, including anomalously connecting left pulmonary veins and a levoatrial cardinal vein. In contrast to a left superior caval vein, flow in these two situations is directed cephalad. A levoatrial cardinal vein is typically associated with left-sided obstructive lesions with restrictive atrial septae, providing an egress for pulmonary venous blood (Bernstein et al. 1995; Geva 2013). Its course is typically posterior to the left pulmonary artery while a left superior caval vein travels anterior to it (Lucas Jr. et al. 1962). Cephalad flow in the left superior caval vein itself occurs if the coronary sinus ostium into the right atrium is partially or completely occluded. In this situation, the only egress of coronary venous blood is via the superior caval vein and into the (bridging) innominate vein.

The presence of a left superior caval vein can be suspected on chest radiographs via a shadow along the left mediastinum. Echocardiography is typically sufficient to secure the diagnosis (Huhta et al. 1982). A left superior caval vein and a bridging innominate or left brachiocephalic vein are best seen from a high left parasternal or a suprasternal sagittal probe position. A dilated coronary sinus seen from a slightly lower right parasternal probe position or, in the rare adult with sufficient transhepatic windows, from subcostal, signifies a persistent left superior caval vein until proven otherwise. The integrity of the coronary sinus can be challenging to visualize conclusively by echocardiography. Imaging during injection of a bolus of agitated saline (socalled "bubble study") into a left arm vein is helpful in demonstrating a left superior caval vein: The bubbles will appear in the coronary sinus before the right atrium. If the coronary sinus is partially or completely unroofed the left atrium fills with bubbles before the right atrium. If these measures do not result in a definitive delineation of the caval and bridging venous as well as coronary sinus anatomy CMR establishes it with certainty. Contrast-enhanced angiography is usually not needed. Computed tomographic angiography accomplishes the same anatomical delineation as CMR. Catheterization may be obtained for reasons other than the systemic veins or with a view to occlude an abnormal vein.

2.2.4 Treatment

No treatment is necessary for isolated persistent left superior vena cava with an intact coronary sinus. If the coronary sinus is completely or partially unroofed with a small to moderate size left superior caval vein and good size bridging innominate vein the left caval vein can be ligated or occluded in the catheter laboratory with minimal penalty from drainage of coronary venous blood into the left atrium. A larger left superior caval vein without adequate connecting vein needs to be baffled into the right atrium, a procedure that also directs coronary venous blood into the right atrium. Aneurysmal enlargement of the superior caval vein does not require surgery unless there is compression of neighboring structures, as the risk of rupture is exceedingly low (Geva 2013).

2.3 Connection of the Right Superior Caval Vein to the Left Atrium

In addition to an unroofed coronary sinus another, much more rare, route of systemic venous drainage into the left atrium is formed by an anomalously connected right superior caval vein (Van Praagh et al. 2003; Shapiro et al. 1981).

2.3.1 Anatomy

The right superior caval vein is partially or entirely committed to the left atrium, overriding an interatrial communication to varying degrees. This situation has been described as the systemic venous analogy of a sinus venosus atrial septal defect and anomalous pulmonary venous connection (Geva 2013).

2.3.2 Presentation

The cardinal sign, central cyanosis, depends on how much of the desaturated blood is directed towards the left atrium. Many patients are asymptomatic until late child- or well into adulthood. In addition to cyanosis patients are at risk for paradoxical emboli, resulting in cerebrovascular accidents and intracranial abscess formation.

2.3.3 Diagnosis

A high index of suspicion is key to making the diagnosis. While echocardiography can be sufficient to establish the anomalous venous connection the pathology is not infrequently missed (Chin 1994). This risk is greater in patients with absent subcostal windows as this is the best view to identify the roof of the atria. Parasternal and high parasternal views from both sides of the sternum are also helpful in this regard. Scanning during injection of agitated saline ("bubble

study") may be useful in understanding the systemic venous drainage. Cross-sectional imaging with CMR or CT delineate the cavo-atrial connection with greater fidelity. The assessment includes a description of the degree of "overriding" of the interatrial communication by the superior caval and the presence of ostial venous stenosis. In addition, CMR phase-contrast flow measurements in the ascending aorta and the pulmonary artery permit a quantification of the right-to-left shunt in bi- or left atrial caval connection. Cardiac catheterization is usually not necessary and nuclear medicine techniques are not helpful.

2.3.4 Treatment

Management is surgical and consists of redirecting the right superior caval venous blood to the right atrium, either by dividing and anastomosing the vein to the right atrial appendage or by baffling it through the atrial communication, which may need to be enlarged, into the right atrium.

Complications consist of stenosis of the anastomosed vein or the baffle as well as baffle leaks.

2.4 Stenosis of the Systemic Veins

While primary stenosis of a major systemic vein is exceedingly rare and most often caused by expansive tumors of the mediastinum, like lymphoma, adults with congenital heart disease are predisposed to this complication of previous interventions: The most frequent mechanism is occlusion with a venous thrombus (Fig. 1). Clot formation is promoted by vessel wall injury through (a) instrumentation, (b) hypercoagulable states which occur during and after every surgery, (c) as side effects of cardiovascular pharmacotherapy, or (d) as a result of venous stasis.

2.4.1 Anatomy

The venous lumen is narrowed. Early in the disease process an intraluminal clot may be visible by imaging, which later becomes partially dissolved, organized, or integrated into the vessel wall. Fig. 1 CMR, bilateral superior vena cava occlusion. The patient was born with intracardiac total anomalous pulmonary venous connection and persistent left superior vena cava (LSVC) to the roof of the left atrium. Following the repair which included baffling of the LSVC to the right atrium, the patient developed occlusion of the left lower and severe stenosis of the left upper pulmonary vein. She also had complete thrombotic obstruction of her right and left superior vena cava (asterisks), with both veins draining via networks of venovenous collaterals. RA right atrium

2.4.2 Presentation

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The venous drainage of the upper body is most often affected. The presence and extend of symptoms of superior vein stenosis depend on location and severity of the obstruction as well as the degree of collateralization. When severe narrowing develops rapidly before collaterals can form and there is only one superior caval vein, signs and symptoms are readily apparent: The most frequent presenting symptoms are dyspnea and headaches (Sharoni et al. 2001; Rice et al. 2006). Nasal congestion and nose bleeds are also common. Other symptoms include cough, arm swelling, hoarseness, and pleural effusions (Blalock et al. 1936).

2.4.3 Diagnosis

Echocardiography and Doppler ultrasound of the neck and mediastinum may show a narrowed lumen and flow acceleration by color and pulsedwave Doppler. Computed tomography and CMR

are the noninvasive methods of choice to visualize venous thrombi. The use of so-called blood pool contrast agents and electrocardiographicgated inversion-recovery CMR sequences offers excellent visualization of thrombi. Invasive venography confirms the diagnosis but is typically reserved for cases in which a percutaneous treatment is planned in the same setting.

2.4.4 Treatment

Patients who are diagnosed acutely with a recently formed thrombus are treated with thrombolysis and anticoagulation. Surgical thrombectomy or percutaneous recanalization are sometimes undertaken. Percutaneous stent placement is the treatment of choice in patients with superior vena cava syndrome. Venous bypass grafts are rarely used. Lifelong follow-up is required to screen for recurrent obstruction.

2.5 **Retroaortic Innominate Vein**

Retroaortic innominate veins are found in ~0.5% of patients with congenital heart disease, most frequently tetralogy of Fallot (Kulkarni et al. 2008).

2.5.1 Anatomy

Normally, the bridging or innominate vein courses from left to right high in the mediastinum and anterior to the proximal aortic arch. In individuals with a retroaortic innominate vein the bridging innominate vein crosses the midline posterior to the ascending aorta and aortic arch and inserts into the right superior caval vein caudal, rather than cranial, to the azygos vein (Fig. 2) (Gerlis and Ho 1989; Choi et al. 1990; Smallhorn et al. 1985).

Presentation 2.5.2

Individuals are asymptomatic. The lesion is discussed here for its surgical implications during the repair of associated heart disease.

2.5.3 Diagnosis

A retroaortic innominate vein is typically diagnosed coincidentally by echocardiography or cross-sectional imaging. By ultrasound, a





Fig. 2 CMR, retroaortic innominate vein. The coronal steady state free precession "localizer" image shows a left innominate vein (asterisk) beneath the aortic arch. *A* transverse arch, *RPA* right pulmonary artery

horizontal sweep from a high right parasternal probe position defines the relationship of the aortic arch to the innominate vein. The proximal portion of the innominate vein must not be mistaken for a persistent left superior caval vein.

2.5.4 Treatment

No treatment is indicated. However, the presence of a retroinnominate vein may necessitate alterations in surgical approach to associated lesions, for example during creation of a bidirectional cavopulmonary connection (Geva 2013). The superior caval vein cannula must be placed more caudally during cardiopulmonary bypass to avoid obstruction of the innominate vein (Chen et al. 2005). The anomalous innominate vein may cause technical difficulties during pacemaker insertion or central venous line placement through the left arm approach (Kulkarni et al. 2008).

2.6 Interrupted Inferior Caval Vein

Although interruption of the inferior caval vein can occur in isolation it should be viewed as a sign of anomalies of the visceral and thoracic situs until proven otherwise.

2.6.1 Anatomy

The inferior vena cava is interrupted along its abdominal course and an intrahepatic segment is absent. Venous blood from the extremities returns to the heart via a prominent azygos or hemiazygos vein (Fig. 3). In the abdomen, interruption of the inferior caval vein is associated with polysplenia, biliary atresia, and intestinal malrotation. In the thorax, it commonly indicates left isomerism of the atrial appendages, bronchi, and lungs. The left and right hepatic veins may drain via a common confluence to the same atrium or to different atria.

2.6.2 Presentation

Patients with isolated interruption of the inferior caval vein are asymptomatic because the azygos continuation ensures normal venous drainage despite an abnormal connection. The clinical importance of this anomaly is its frequent association with abdominal heterotaxy and left isom-



Fig. 3 CMR, absent infrahepatic inferior vena cava, large azygos vein. Volume rendered reformat of a magnetic resonance angiogram, seen from levo-posterior. This patient with "azygos continuation" (arrowheads) of the systemic venous return below the liver had a normal situs and tetralogy of Fallot with a right aortic arch. *DAO* descending aorta

erism of the atrial appendages. In this setting, congenital heart disease is common, most often in the form of left-sided obstructive lesions.

2.6.3 Diagnosis

In adults with significant congenital heart disease in the face of left isomerism of the atrial appendages the presence of an interrupted inferior caval vein with azygos continuation is known from previous diagnostic tests or surgery. Occasionally, it is diagnosed in adulthood. Chest plain films that show left isomerism of the main bronchi should prompt an echocardiogram to assess for interruption of the inferior caval vein and associated heart disease. Echocardiography shows a missing intrahepatic inferior vena cava segment and the dilated (hemi-)azygos vein as a prominent channel behind the liver in the paravertebral gutter (Huhta et al. 1984). In patients with sufficient acoustic windows the (hemi-)azygos vein can be shown in its entirety and connection with the superior caval vein. Cardiac magnetic resonance imaging and CT are definitive in the diagnosis. Cardiac catheterization is not needed except for in the diagnosis and treatment of associated heart disease. Imaging, preferably by abdominal ultrasound or CMR, delineates the abdominal situs, including the position of the stomach and liver; presence size, number, and position of the spleen(s), anatomy of the biliary system, and rotation of the intestine.

2.6.4 Treatment

When associated with complex congenital cardiac malformations which require a Fontan-type palliation the presence of an interrupted inferior caval vein changes the surgical strategy. At the time of the bidirectional cavopulmonary connection the blood from most of the lower body is diverted into the lungs via the azygos vein and its connection with the superior caval vein (Kawashima operation). At the time of the Fontan completion the hepatic venous flow is collected in a lateral tunnel or extracardiac conduit towards the pulmonary circulation.

Follow-up and life expectancy depend on the associated heart defects and their surgical palliation.

2.7 Anomalous Drainage of the Inferior Caval Vein

2.7.1 Anatomy

Whether a direct connection of the inferior caval vein to the morphological left atrium is embryologically possible is under debate (Smallhorn and Anderson 2010; Geva 2013). However, drainage of blood from the inferior caval vein into the left atrium via an atrial septal defect, particularly in a setting of a prominent persistent Eustachian valve, is a well-recognized occurrence (Lucas Jr. and Krabill 1995). Rarely, the inferior caval vein connects to the coronary sinus. Partial anomalous hepatic venous connection to the coronary sinus has also been described (Nabarro 1903). These anomalies are unlikely to occur in isolation and their discovery should prompt a search for other congenital heart disease.

2.7.2 Presentation

The anomalous drainage of desaturated blood to the left atrium results in cyanosis. The right-toleft shunt also carries the risk of paradoxical emboli.

2.7.3 Diagnosis

The diagnosis can usually established by transthoracic echocardiography. In patients with subdiagnostic acoustic windows cross-sectional imaging establishes the venous connections.

2.7.4 Treatment

Venous blood flow is surgically redirected to the right atrium.

2.8 Portosystemic Shunts

2.8.1 Anatomy

Congenital portosystemic shunts are divided into extra- and intrahepatic. In extrahepatic shunts the portal vein is either absent or part of the portal venous blood is diverted into the inferior caval vein. Intrahepatic portosystemic shunts are connections between branches of the portal vein and the hepatic veins.

2.8.2 Presentation

Ammonia and other toxins which are normally cleared by the liver result in hepatic encephalopathy, manifesting in tremors, seizures, and psychiatric symptoms. Other complications include failure to thrive, hepatopulmonary syndrome, and pulmonary hypertension. Patients may suffer from urate kidney stones because of increased amounts of uric acid in circulation which are excreted by the kidneys.

2.8.3 Diagnosis

Ultrasound with color Doppler imaging is the first-line imaging tool for the evaluation of portosystemic shunts (Franchi-Abella et al. 2010). Magnetic resonance imaging and CT angiography readily visualize the anatomy (Kakitsubata et al. 1996). Doppler ultrasound and magnetic resonance phase-contrast flow velocity mapping can be used to quantify the portovenous shunt ratio by dividing the total blood flow volume in the shunt by that in the portal vein (Kudo et al. 1993). Ratios <24–30% do not appear to result in hepatic encephalopathy while ratios >60% are an indication for intervention (Maeda et al. 1993). Nuclear medicine studies have also been used to quantify the magnitude of the shunt (Uchino et al. 1996).

2.8.4 Treatment

Asymptomatic patients do not require treatment. Percutaneous shunt occlusion or surgical closure of the shunt have been described. However, the portal venous system must be shown to be definitively intact before shunt occlusion to prevent mesenteric venous congestion and bowel ischemia.

2.9 Chiari Network

2.9.1 Anatomy

Chiari networks are the most frequent type of persistent embryonic valves within the right atrium. They consist of strands of tissue extending across the cavity of the right atrium from the terminal crest to the Thebesian or Eustachian valves (Smallhorn and Anderson 2010; Chiari 1897).

2.9.2 Presentation

Most patients are asymptomatic. Large Chiari networks can protrude through the tricuspid valves and cause obstruction to inflow. Endocarditis and thrombus formation within the network with embolization have also been described.

2.9.3 Diagnosis

Chiari networks are readily detected by echocardiography.

2.9.4 Treatment

Active management is usually not necessary. If there are complications resection may be considered.

3 Pulmonary Venous Abnormalities

3.1 Normal and Pathological Development of the Pulmonary Venous System

Around the fourth week in human development the blood from the primitive lung buds drains into the splanchnic plexus and from there into the paired common cardinal and umbilicovitelline veins (Lucas Jr. et al. 1963). A single primitive pulmonary vein develops shortly afterwards and fuses with the left atrium. For a short period of time, the pulmonary venous plexus is connected to both this early pulmonary vein and to the cardinal and umbilicovitelline veins. Soon afterwards, the connections to the latter two involute. These transient multiple connections of the pulmonary venous contributors serve to conceptualize all cases of anomalous pulmonary venous connection while an incomplete incorporation of the primitive pulmonary vein is the substrate for cor triatriatum: Anomalous connection to the systemic venous system are thought of as a result of the atresia of a pulmonary vein while pulmonaryto-systemic venous connections still exist. Cor triatriatum is the consequence of stenosis or incomplete fusion during the incorporation of the primitive single pulmonary vein into the left

atrium, after the pulmonary-to-systemic venous connections have been lost.

3.2 Partial Anomalous Pulmonary Venous Connection

Most people have two right- and two left-sided pulmonary veins, although a right middle pulmonary vein is not uncommon. In partial anomalous pulmonary venous connection (PAPVC) some, but not all, of the pulmonary veins connect to the systemic venous system instead of the left atrium. Usually, but not always, abnormal connection leads to abnormal drainage, and vice versa.

3.2.1 Anatomy

Anomalous pulmonary venous connection most often involves the right upper and middle pulmonary veins to the superior caval vein or right atrium (Fig. 4) as well as the left upper pulmonary vein to the left innominate vein (Fig. 5). Partial anomalous pulmonary venous connection of the right



Fig. 4 CMR, partial anomalous pulmonary venous connection. This patient with multiple anomalously connected veins draining the right upper and part of the right middle lobe (arrow) also had a sinus venosus type interatrial communication. Together, the two left-to-right shunts amounted to a pulmonary to systemic blood flow ratio of 3.2, causing severe right atrial and right ventricular enlargement. The main pulmonary artery is also dilated. *RA* right atrium, *RV* right ventricle, *SVC* superior vena cava



Fig. 5 CMR, repaired total anomalous pulmonary venous connection with residual left vertical vein. The volume rendered reformat of a magnetic resonance angiogram demonstrates a persistent left vertical vein in a patient who was born with total anomalous pulmonary venous connection. The connection of the left upper pulmonary vein to the left innominate vein was not corrected because the patient developed pulmonary hypertension during intra-operative test occlusion of the vertical vein. *LA* left atrium, *LLPV* left lower pulmonary vein, *RLPV* right lower pulmonary vein, *VV* vertical vein

upper plus/minus right middle pulmonary vein(s) is often associated with a superior sinus venosus defect, a complex malformation involving the pulmonary veins, the superior caval vein/right atrium junction, and the superior wall between the right and left atria. An inferior sinus venosus defect constitutes an interatrial communication and anomalous drainage of the right lower pulmonary vein.

3.2.2 Presentation

In many ways, the clinical picture of PAPVC is similar to that of an atrial septal defect. Both cause a left-to-right shunt which, if large enough, can lead to right heart dilatation, arrhythmias, and pulmonary arterial hypertension. Factors that influence the degree of left-to-right shunting include the number of anomalously connected pulmonary veins, the presence and size of any atrial septal defect (ASD), and the compliance of the right ventricle (Brown and Geva 2013; Seller et al. 2018).

3.2.3 Diagnosis

Patients with a significant shunt show increased pulmonary vascular markings and dilatation of the right ventricle and the pulmonary artery segment on chest X-ray. The X-ray may also provide clues to the type of PAPVC: In Scimitar syndrome, for example, the right lung is hypoplastic and the mediastinum shifted to the right, with elevation of the right hemidiaphragm (Yoo and Epelman 2010). There is retrosternal soft tissue density. The anomalously connected right-sided pulmonary veins are often recognized as a crescent-shaped shadow within the right mediastinum. With long-standing, unrepaired PAPVC pulmonary vascular disease can occur and manifest in "pruning" of the peripheral pulmonary arterial branches (Saalouke et al. 1977; Babb et al. 1981). Echocardiography is often, but not always, sufficient in the delineation of the pulmonary venous anatomy. The junction of the right upper pulmonary vein to the left atrium, in particular, can be difficult to ascertain. It is important that all pulmonary veins are detailed in their connection to either the left atrium or, anomalously, to a systemic venous structure, including the right atrium. Left and right parasternal, suprasternal, and apical acoustic windows are most useful to show the pulmonary veins. In slim individuals with favorable windows the subcostal approach may present another option. A complete echocardiography study also detects any associated lesions, most importantly an interatrial communication. The severity of left-to-right shunting can be gauged from the degree of right atrial and right ventricular enlargement. Flatting of the interventricular septum at end-diastole is an indicator of right ventricular volume overload. While transesophageal echocardiography can typically clarify the venous anatomy in patients with insufficient transthoracic windows this information can be obtained noninvasively and more comprehensively by CMR or CT. As mentioned in the imaging overview at the beginning of this chapter the CMR "localizer" or "scout" images are often sufficient to detail the pulmonary venous connections. However, contrastenhanced angiography is beneficial, especially when more than one anomalously connected vein

is suspected. When all of the right pulmonary veins are found to connect to the right atrium, inferior caval vein, or a hepatic veins the possibility of a Scimitar syndrome must be strongly considered (Fig. 6). Patients with Scimitar syndrome have other abnormalities, including, mentioned, hypoplasia of the right lung and the right pulmonary artery, lung segmentation and bronchial branching abnormalities, and often a prominent aortopulmonary collateral artery from the abdominal aorta to the base of the right lung. Adults with this condition are usually symptomatic and a substantial proportion have pulmonary arterial hypertension. In addition to the anatomy, the degree of right ventricular dilatation and the magnitude of the left-to-right shunt is an important factor in surgical decision-making. Today's gold standard for ventricular volumetry and shunt quantification is CMR. Phase-contrast CMR allows for flow assessments in each individual pulmonary vein as well as across the atrial septum (Beerbaum et al. 2001; Goo et al. 2009; Grosse-Wortmann et al. 2007). Separating the contributions of shunt across the atrial communication and via the anomalously draining pulmonary vein is of clinical relevance in patients with

RA S HV

Fig. 6 CT, Scimitar syndrome. The computed tomography contrast-enhanced angiogram shows complete anomalous connection of the entire right lung via a scimitar vein to the suprahepatic inferior vena cava. *HV* hepatic vein, *RA* right atrium, *S* scimitar vein

PAPVC of the left upper pulmonary vein with secundum type atrial septal defect: If the shunt via the vein is minor percutaneous device closure of the atrial septal defect may be the preferred approach as opposed to a "full repair" on cardiopulmonary bypass. Given the performance of echocardiography and cross-sectional imaging cardiac catheterization is not routinely necessary in PAPVC, unless pulmonary arterial hypertension is suspected. The ratio of pulmonary (Qp) to systemic blood flow (Qs) is calculated via oximetry and the Fick principle. The levophase of a fluoroscopy after right ventricular or pulmonary arterial injection of contrast delineates the pulmonary venous drainage and connection. The anomalously connected vein may be entered retrogradely and injected selectively with dye.

3.2.4 Treatment

The natural history of PAPVC is not favorable in all cases. Anecdotal case reports of patients developing pulmonary vascular disease in the affected lung lobe in the presence of only one anomalously draining pulmonary vein document the risk of pulmonary hypertension in these patients (Saalouke et al. 1977; Babb et al. 1981). The indication for repair of PAPVC with or without atrial septal defect depends on symptoms, shunt magnitude, and degree of right ventricular dilatation. A pulmonary (Qp) to systemic flow (Qs) ratio ≥ 2.0 is widely used as a criterion for the operative repair of simple left-to-right shunt lesions, including atrial level shunts and PAPVC. Using CMR, we recently found that asymptomatic patients with isolated anomalous connection of the left upper pulmonary vein without an atrial septal defect are unlikely to have a significant left-to-right shunt and typically do not require surgery (Seller et al. 2018). On the other hand, the combination of right-sided PAPVC with (sinus venosus) atrial communication is routinely associated with a significant left-to-right shunt. The most important postoperative sequelae are pulmonary vein stenosis (see below) and atrial arrhythmias. The scimitar vein is typically baffled to the right atrium, either via route of an atrial septal defect, that can be associated with the Scimitar Syndrome or surgically created. Surgical

indications and postoperative complications in patients with Scimitar syndrome are identical to other types of PAPVC, although Scimitar syndrome carries a worse prognosis, owing to the accompanying lung hypoplasia as well as bronchial branching and lung segmentation abnormalities, but also due to the higher prevalence of native pulmonary venous obstruction.

3.3 Total Anomalous Pulmonary Venous Connection

Anomalous connection of all pulmonary veins hardly ever presents in an adult and is only discussed superficially in this book (Cheng et al. 2015). However, the sequelae of surgical repair of total anomalous pulmonary venous connection (TAPVC), residual anomalous connection of one or more pulmonary veins or pulmonary vein stenosis, are not uncommon among adults.

3.3.1 Anatomy

The connection of the pulmonary veins to the systemic venous system may be at the supracardiac, (intra-)cardiac, or infracardiac level (Craig et al. 1957). Combinations of these three types are possible within the same patient.

3.3.2 Presentation

As mentioned, TAPVC typically manifests, and is repaired, in early childhood. The very few that survive to adulthood with this diagnosis have excellent atrial mixing and minimal obstruction of the pulmonary venous pathway. Given the propensity of obstruction in infracardiac TAPVC adult cases are invariably of the supracardiac, intracardiac, or mixed type. There is an obligatory left-to-right shunt and systemic cardiac output can only be maintained if there is also a right-to-left shunt, usually across the atrial septum. Consequently, patients present in heart failure with varying degrees of cyanosis.

3.3.3 Diagnosis

The chest X-ray shows cardiomegaly, increased pulmonary vascularity, and pulmonary edema. It is sometimes possible to detect dilated pulmonary-to-systemic venous connections, such as in supracardiac TAPVC and the wellknown "snowman sign" (Yoo and Epelman 2010). Echocardiography is often sufficient to define the pulmonary venous anatomy comprehensively in neonates. There are few lesions that produce an exclusive right-to-left shunt across the atrial septum. Total anomalous pulmonary venous connection is, one of them. The left ventricle in TAPVC is underfilled. With more advanced age at presentation cross-sectional imaging, i.e., CT or CMR, take on an increasingly important role in delineating the anatomy. It is imperative to define the anatomy of each pulmonary vein, the common pathway, if present, as well as any accompanying heart lesions. An important association exists between TAPVC and right isomerism of the atrial appendages so that the latter must be actively ruled out. Any obstruction within the pulmonary venous drainage or the obligatory right-to-left shunt at the atrial level must be assessed. Cardiac catheterization with assessment of pulmonary vascular resistance is advised prior to surgical correction in an adult patient.

3.3.4 Treatment

In the absence of prohibitive pulmonary vascular disease the pulmonary veins are anastomosed to the left atrium.

3.4 Cor Triatriatum

Cor triatriatum is the result of failure of incorporation of the primitive common pulmonary vein into the left atrium (Brown and Geva 2013).

3.4.1 Anatomy

The left atrium is divided into an upper and a lower chamber. The pulmonary veins enter the "upper chamber." Significant heterogeneity in presentation and management is introduced by the presence or absence of communications between either chamber and the right atrium and between the upper and lower left atrial compartments (Niwayama 1960). Cor triatriatum is commonly associated with PAPVC.

3.4.2 Presentation

The physiology and clinical presentation of Cor triatriatum varies widely according to atrial level shunts and associated PAPVC (47). If no communication between the upper chamber and the right atrium exists and the connection to the lower chamber is stenotic the clinical picture is that of mitral stenosis. If both left atrial chambers communicate with the right atrium and the connection between upper and lower chambers is either stenotic or atretic the physiology is that of TAPVC. (Brown 2009).

3.4.3 Diagnosis

It is important to delineate the membranous barrier between the upper and lower left atrial compartments. In contrast to supravalvar mitral stenosis the intraatrial membrane is situated above the left atrial appendage. All interatrial communications must be visualized in terms of size, estimated gradients, and flow directions. Given the common association with PAPVC all pulmonary veins must be interrogated. In many cases, transthoracic echocardiography visualizes the pertinent features with sufficient detail and certainty. The right ventricle is often dilated and hypertrophied. Depending on the physiology the chest X-ray shows evidence of pulmonary venous obstruction, with pulmonary edema and enlargement of the central pulmonary arteries. The assets of CMR and CT are the same as for TAPVC. The intraatrial membrane can be challenging to see by cross-sectional imaging, especially on an ungated CT angiogram. Invasive manometry during cardiac catheterization reveals pulmonary arterial hypertension unless the upper left atrial chamber has an unobstructed outlet, either into the right atrium or into the lower chamber. Selective injections into the pulmonary arteries opacify the pulmonary veins and the upper left atrial chamber. If there is a defect in the core membrane, the lower chamber and the left ventricle fill after a small delay and the membrane itself can be appreciated as a thin filling defect between the two chambers.

3.4.4 Treatment

The definite management is typically surgical and consists of resection of the intraatrial membrane. Any associated PAPVC is corrected at that time, unless deemed to be hemodynamically insignificant. Percutaneous relief of obstruction with or without closure of unnecessary communications with the right ventricle is an alternative approach in select cases (Kerkar et al. 1996; Li et al. 2015). The prognosis in "uncomplicated" and repaired Cor triatriatum is good. Any associated PAPVC predisposes to postoperative pulmonary vein stenosis.

3.5 Pulmonary Vein Stenosis

Pulmonary vein stenosis manifests as a primary lesion or after pulmonary vein surgery. Primary pulmonary vein stenosis occurs as an isolated defect or associated with other types of congenital heart disease.

3.5.1 Anatomy

The morphology of pulmonary vein stenosis is either a discrete narrowing (Fig. 7) at the ostium into the left atrium or a diffuse, long-segment hypoplasia of the entire vein and often also its peripheral branches. Combinations of these two features are common as ostial stenosis often entails hypoperfusion which leads to shrinking of the contributing channels.

3.5.2 Presentation

Patients present with signs of pulmonary arterial hypertension, including tachypnea, reduced exercise tolerance and, sometimes, cyanosis. Recurrent pneumonia is a frequent complication; pulmonary hemorrhage occurs in advanced disease.

3.5.3 Diagnosis

In advanced disease the chest X-ray shows increased interstitial markings and dilation of the pulmonary lymphatic vessels with ground-glass changes (Yoo and Geoffray 2010). Long-standing pulmonary vein stenosis may lead to decreased ipsilateral pulmonary artery size and lung hypoplasia. The heart size is typically normal. Echocardiography is routinely used in the postoperative surveillance for obstruction following



Fig. 7 CT, pulmonary vein stenosis. The coronal multiplanar reformat of the computed tomography angiogram demonstrates severe focal stenoses (asterisk) of all three contributors to the right lower pulmonary vein in a 3-year old. The right pulmonary artery is enlarged, secondary to pulmonary hypertension. The other three pulmonary veins were also obstructed. The stenoses recurred following surgical and catheter interventions and the patient eventually received a heart and lung transplant. *LA* left atrium, *RPA* right pulmonary artery

pulmonary vein repair. An unobstructed pulmonary vein has laminar flow by color Doppler and low velocity, phasic flow pattern with one or two systolic peaks, a diastolic peak and a brief nadir at the end of diastole (Grosse-Wortmann et al. 2007). A stenosed pulmonary vein lacks some or all of these features. An elevated mean gradient estimate by pulsed-wave Doppler confirms the stenosis. Relatively low velocity flow, however, does not rule out significant obstruction, especially if the classic phasic fluctuations across the cardiac cycle are attenuated, as flow redistribution away from the obstruction can mask a higher gradient. It has been shown repeatedly that echocardiography lacks sensitivity as well specificity in the detection of pulmonary vein stenosis, in comparison to CMR (Valsangiacomo et al. 2003a, b; Greenway et al. 2011). Magnetic resonance offers anatomical and functional delineation: In significant obstruction, the ipsilateral branch pulmonary artery is usually hypoplastic as a reflection of the decreased blood flow to the

lung with the affected vein (Roman et al. 2005). Diffusely small peripheral pulmonary venous contributors are a sign of severe obstruction and shunting of blood away from the stenosed vessel. This appearance may also reflect diffuse disease with so-called arterialization of long segments of the pulmonary veins. It is sometimes possible by CMR or CT to visualize venous collateral channels that deviate blood away from the obstructed egress (Grosse-Wortmann et al. 2007). Phasecontrast velocity CMR demonstrates the hemodynamic effects of the stenosis. This information aids in gauging the severity of obstruction and is particularly helpful in distinguishing compression of a vein (typically the left lower) between the descending aorta and the heart without obstruction to flow from significant stenosis. Flow mapping should be performed in the pulmonary arteries and in the pulmonary veins. Significant pulmonary vein obstruction often leads to redistribution of blood flow, away from the area drained by the obstructed lung. This redistribution can be seen at the arterial level as an imbalance of right vs. left pulmonary artery flow volume. It can also be observed at the venous level with a greater proportion of the ipsilateral pulmonary blood flow passing through the nonobstructed vein. The possibilities of multi-site and bilateral obstruction must be taken into consideration when interpreting flow data. The pulmonary arterial flow curve may show signs of pulmonary arterial hypertension, with an earlier systolic peak, diminished peak velocity, additional late systolic and diastolic peaks and a deep nadir in early diastole. The flow profile of the affected pulmonary vein is typically dampened with decreased amplitude. The average velocity of flow in the pulmonary vein is typically low when measured "upstream" of the stenosis and accelerated "downstream" from it (Grosse-Wortmann et al. 2007; Valsangiacomo et al. 2003b). Similar to phase-contrast CMR a lung perfusion scan with 99 m-technetium-labeled macroaggregated albumin shows reduced relative perfusion of the affected lung (Drubach et al. 2015). Cardiac catheterization is useful in the quantification of the severity of pulmonary arteanatomical rial hypertension and in the

delineation of the stenotic segment. When evaluating a fluoroscopic angiogram in the postero-anterior projection it should be remembered, however, that the cranio-caudal diameter in this view is often preserved despite significant narrowing in the antero-posterior dimension.

3.5.4 Treatment

In principle, two different surgical techniques are used to connect the pulmonary veins to the left atrium: With the "conventional repair," the pulmonary veins are directly sutured to the left atrium. In contrast, with the atriopericardial anastomosis, often referred to as "sutureless repair," a wide communication between the left atrium and the incised pulmonary veins is created using a pericardial patch. Both approaches are burdened with risks of restenosis (Greenway et al. 2011). Postoperative pulmonary venous obstruction manifests most commonly during the first year after surgery and can develop rapidly. Imaging surveillance consists out of a combination of echocardiography and cross-sectional imaging. Given the limitations of echocardiography in the diagnosis of pulmonary vein stenosis CT or CMR is recommended at least once or twice during the first postoperative year and in greater intervals thereafter. Percutaneous approaches to pulmonary vein obstruction with angioplasty and stenting have yielded disappointing results. Attempts at pharmacological interventions that target the fibrotic vascular remodeling give rise to optimism, but are experimental at the time of writing (Zhu et al. 2014).

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