Evaluation of the Atria, Atrioventricular Valves, and Veins

5

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One should begin cardiac CT evaluation by assessing the atria, the systemic and pulmonary veins draining to them, and the atrioventricular (AV) valves. Examine the following:

(a) Anatomy of the right atrium, left atrium, or a common atrium. Assess whether there is atrial inversion, and determine the size of each atrium (hypoplasia or dilatation).

(b) Atrial appendages. The right atrial appendage is triangular and directed anteriorly and superiorly, whereas the left atrial appendage is tongue shaped and directed laterally (leftward). In some complex heart defects, the atrial appendages may be difficult to identify. Look for an abnormal right-sided or a left-sided juxtaposition of the atrial appendages.

(c) Superior vena cava (SVC) and inferior vena cava (IVC). The normal SVC inserts into the right-sided morphologic right atrium, whereas the IVC inserts into the medial, posterior, and inferior aspect of the right atrium. The hepatic veins communicate with the IVC. A left SVC may be present with or without a crossing brachiocephalic vein that connects to a right SVC. It is important to determine whether the left SVC empties into a coronary sinus that then drains to the right atrium, or whether it empties directly into the left atrium.

(d) Pulmonary veins. Typically, there are four pulmonary veins connecting to the morphologic left atrium. However, sometimes two veins may fuse into one before joining the left atrium, or multiple pulmonary veins may drain to one side before they reach the left atrium. Pulmonary veins that drain anomalously should be identified—one, some, or all of the pulmonary veins may drain to the IVC, hepatic vein, SVC, or brachiocephalic vein. Total anomalous pulmonary venous connection occurs if all pulmonary veins drain anomalously either together through a common pulmonary venous chamber or through mixed or different locations. A common association of a right upper or lower partial anomalous pulmonary vein on the right vein is the presence of a sinus venosus type of atrial septal defect (ASD). A partial anomalous pulmonary venous connection of all or most of the right-sided pulmonary veins is seen in scimitar syndrome, in which the anomalous connection is into the IVC. This is associated with a hypoplastic right lung and right pulmonary artery as well as systemic arterial supply to the right lower lobe.

(e) Atrial septum. Defects in the atrial septum may be difficult to see and may require assessment of different phases of the scan to

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optimize visualization. Echocardiography provides optimal visualization of the type, size, extent, and number of ASDs.

(f) AV valves. There are two AV valves: the tricuspid valve, which drains into the right ventricle, and the mitral valve, which drains into the left ventricle. Anomalies include dilatation, hypoplasia, dysplasia, atresia, overriding annulus, straddling chordae, common AV valve, and the presence of Ebstein's malformation. Ebstein's anomaly, a spectrum of tricuspid anomaly that occurs when there is apical displacement of the septal leaflet and dysplasia of the posterior leaflet, and when the anterior leaflet becomes sail-like, with some degree of tethering to the right ventricular (RV) wall. This results in atrialization of a portion of the right ventricle and, rarely, obstruction of the RV outflow tract.

Normal Anatomy of the Atria and Veins

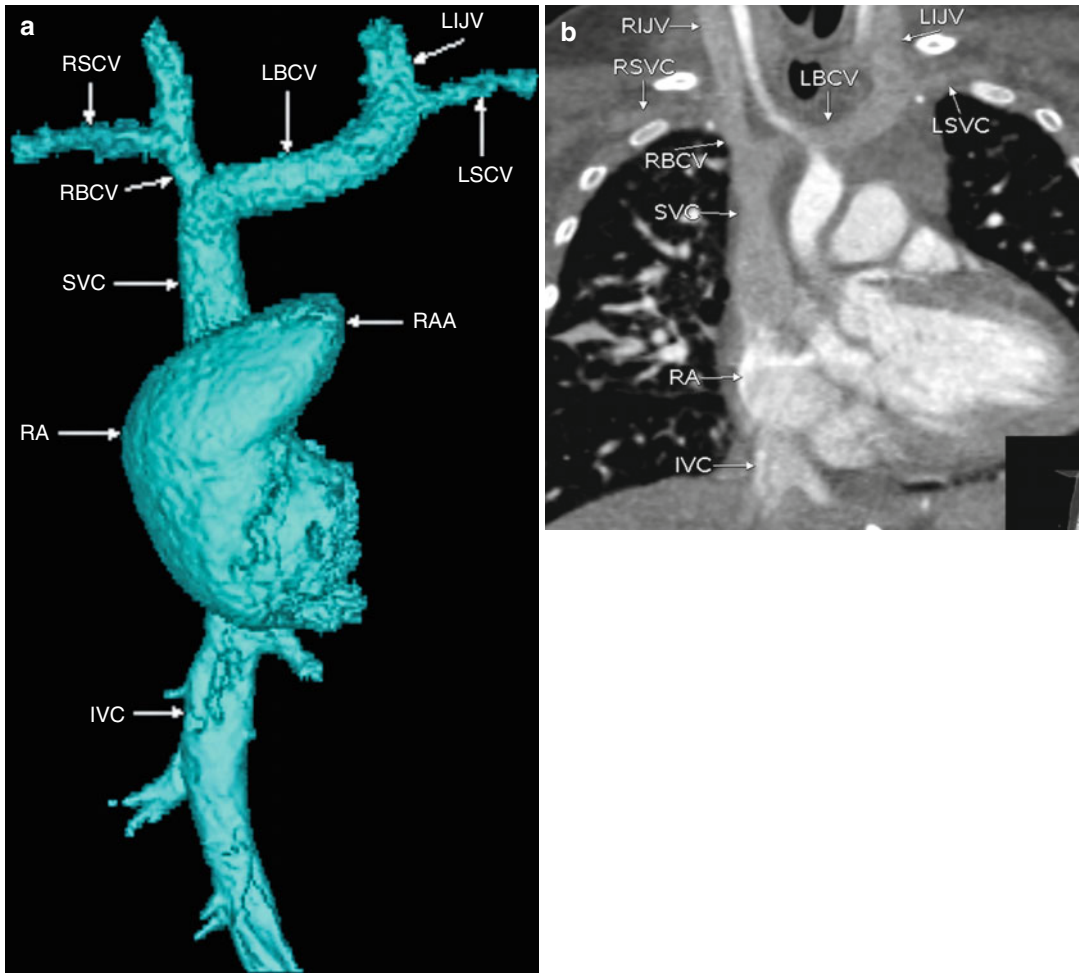

Fig. 5.1 Frontal projection three-dimensional (3D) model (**a**) and coronal maximum-intensity projection (MIP) image (**b**) from a cardiac CTA shows normal anatomy of the normal right-sided SVC (*RSVC*) draining the left brachiocephalic vein (*LBCV*) and right brachiocephalic vein (*RBCV*) into the right atrium (*RA*). Note the typical triangular shape of the right atrial appendage (*RAA*). *LIJV* left internal jugular vein, *LSCV* left subclavian vein, *LSVC* left SVC, *RIJV* right internal jugular vein, *RSCV* right subclavian vein, *IVC* inferior vena cava, *SVC* superior vena cava

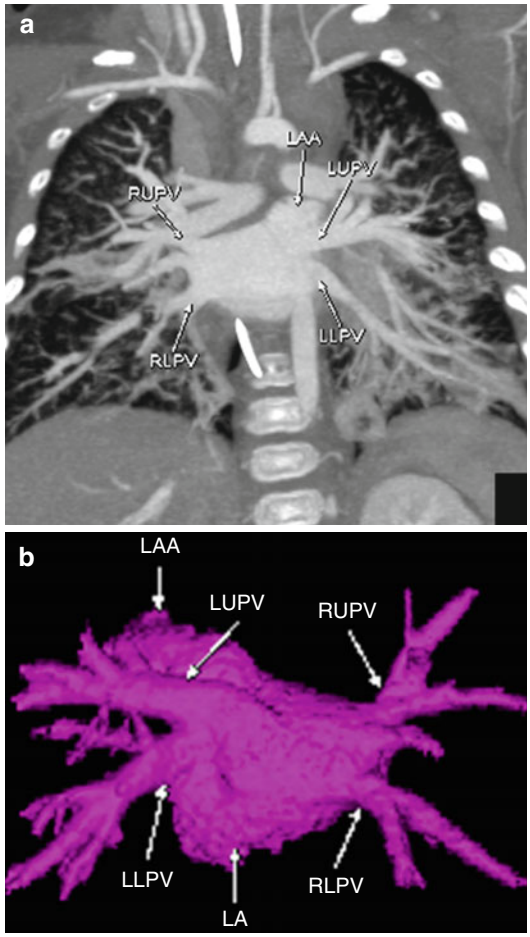

Fig. 5.2 Coronal MIP (**a**) and posterior projection 3D color-coded (**b**) images from a cardiac CTA scan demonstrating normal anatomy, with right and left upper and lower pulmonary veins (*RUPV* right upper pulmonary vein, *LUPV* left upper pulmonary vein, *RLPV* right lower pulmonary vein, *LLPV* left lower pulmonary vein, respectively) draining to the left atrium (*LA*) and a normal-appearing left atrial appendage (*LAA*). Note the elongated, tongue-like appearance of the left atrial appendage

Left Superior Vena Cava

Left SVC is a common variant, especially in patients with congenital heart disease. It is important to make this finding for presurgical planning in patients undergoing SVC shunts so that there is no collateral flow around the shunt. In most instances, there is duplicated SVC physiology, with the left SVC draining blood from the left

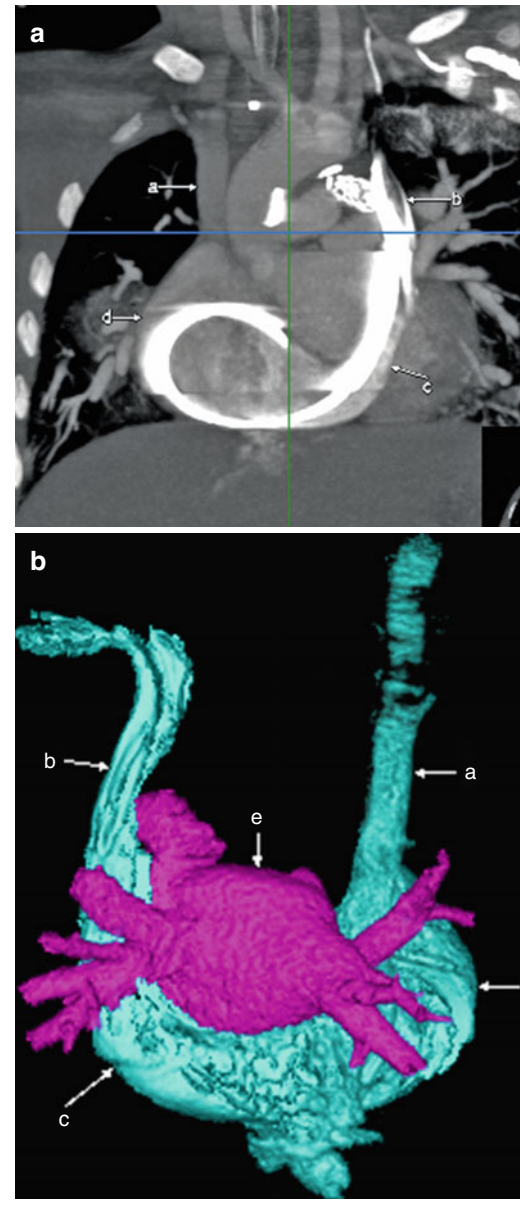

Fig. 5.3 Coronal MIP (**a**) and posterior projection color-coded 3D (**b**) images from a cardiac CT scan showing a left SVC (*b*) draining to the right atrium (*d*) via a large coronary sinus (*c*). Notice the distal right SVC (*a*) draining normally into the right atrium (*d*). Notice the elongated left atrial appendage adjacent to the left SVC (*b*)

internal jugular and left subclavian veins and the right-sided systemic veins draining to a right SVC. In patients with atrial inversion that may be associated with heterotaxy syndrome, the left SVC may be the morphologic right SVC.

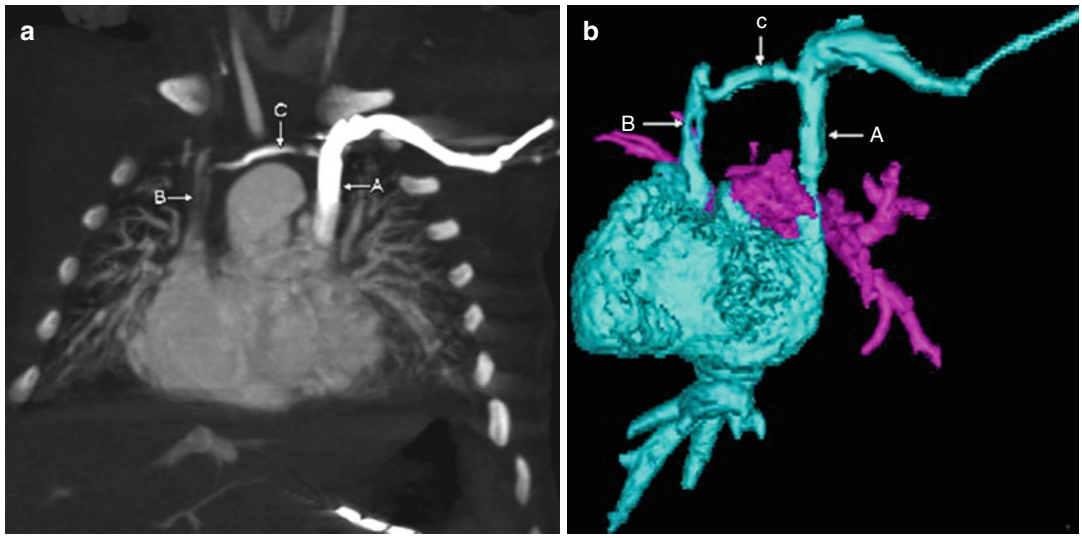

Fig. 5.4 Coronal MIP (**a**) and frontal projection color-coded 3D (**b**) images from a cardiac CT scan showing a right (*B*) and left (*A*) SVC with a connecting brachiocephalic vein (*C*).

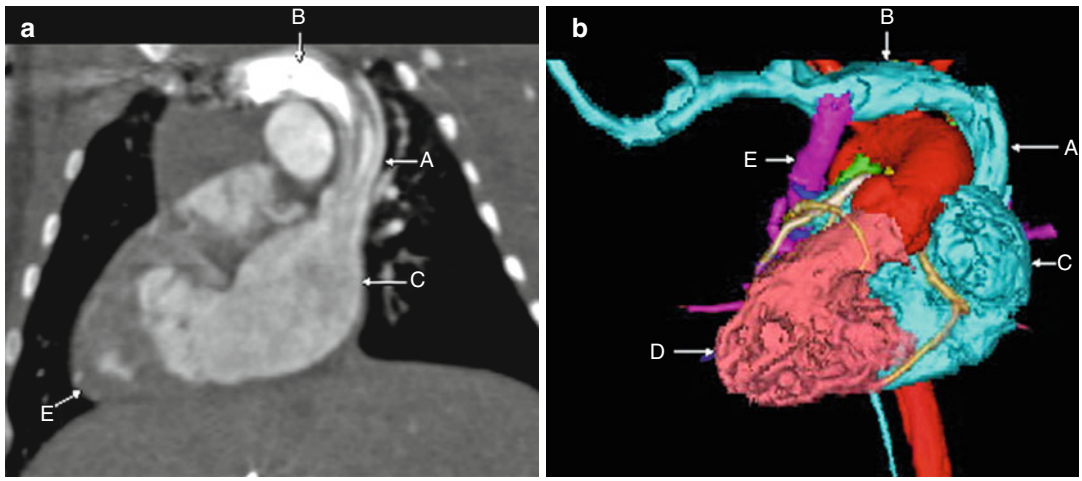

Fig. 5.5 Coronal MIP (**a**) and frontal projection color-coded 3D (**b**) images from a cardiac CT scan showing a right brachiocephalic vein (*B*) emptying into a left SVC (*A*) draining to the left atrium (*C*) in a patient with situs inversus. This may be associated with other congenital anomalies, such as total anomalous pulmonary venous return (TAPVR). In this case, a right-sided vertical vein (*E* in **b**) drains blood from the pulmonary veins (*pink*) to the brachiocephalic vein (*B*). Also notice that the apex of the heart is on the right, consistent with dextrocardia and known situs ambiguous in a patient with heterotaxy syndrome

Partial Anomalous Pulmonary Venous Return

Partial anomalous pulmonary venous return (PAPVR) has the following characteristics:

1. One or more, but not all, pulmonary veins flow to a systemic vein
2. One pulmonary lobe typically is involved, although the whole lung may be involved
3. It typically is asymptomatic
4. If symptoms occur, they usually are the result of right heart overload from increased volume
5. It is associated with sinus venosus ASD and scimitar syndrome.

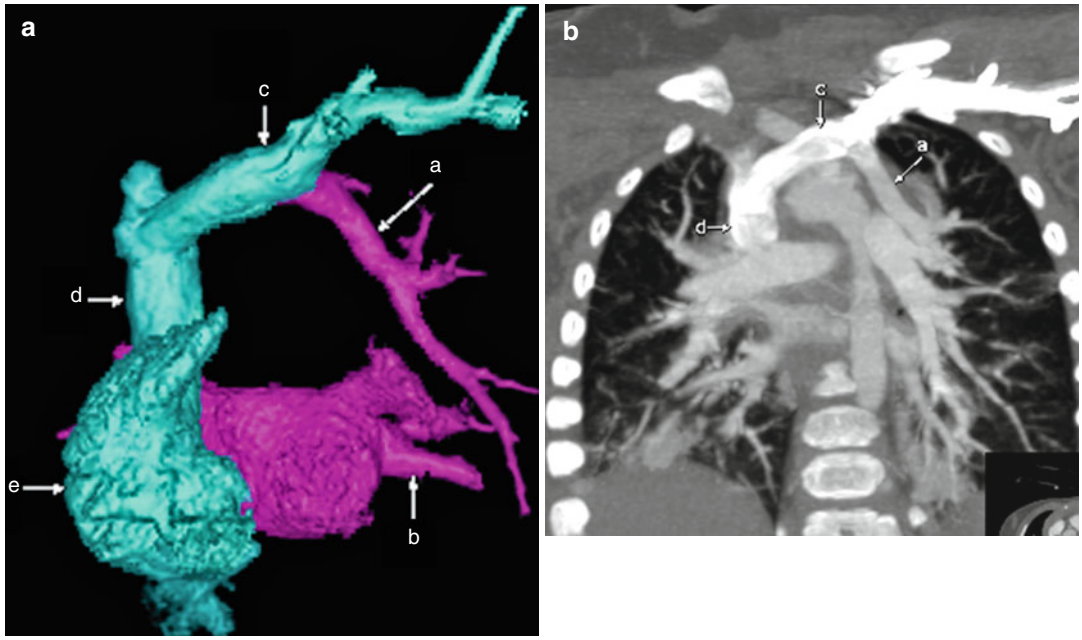

Fig. 5.6 Frontal projection 3D color-coded (**a**) and coronal MIP (**b**) images demonstrating a left upper lobe pulmonary vein (*a*) anomalously draining to the left brachiocephalic vein (*c*). The SVC (*d*) and right atrium (*e*) are mildly prominent. Notice that there is normal drainage of the left lower lobe pulmonary vein (*b*) to the left atrium

Supracardiac TAPVR Type 1

In the most common type of supracardiac TAPVR, the pulmonary veins form a common chamber connected to a vertical vein that in turn drains to the left brachiocephalic vein, which connects to the SVC. Occasionally, supracardiac TAPVR may connect to the right-sided SVC through a vessel that crosses the midline. Obstruction to the pulmonary venous blood flow may occur as the vertical vein crosses the bronchi or at its entry to the systemic vein. All types of TAPVR are left-to-right shunts that form obligatory right-to-left shunts through an ASD or patent ductus arteriosus after mixing with systemic venous blood. Thus, patients are cyanotic. This lesion may be isolated or associated with complex congenital heart disease and heterotaxy syndromes. All total anomalous pulmonary venous connections (TAPVCs) result in increased pulmonary blood flow.

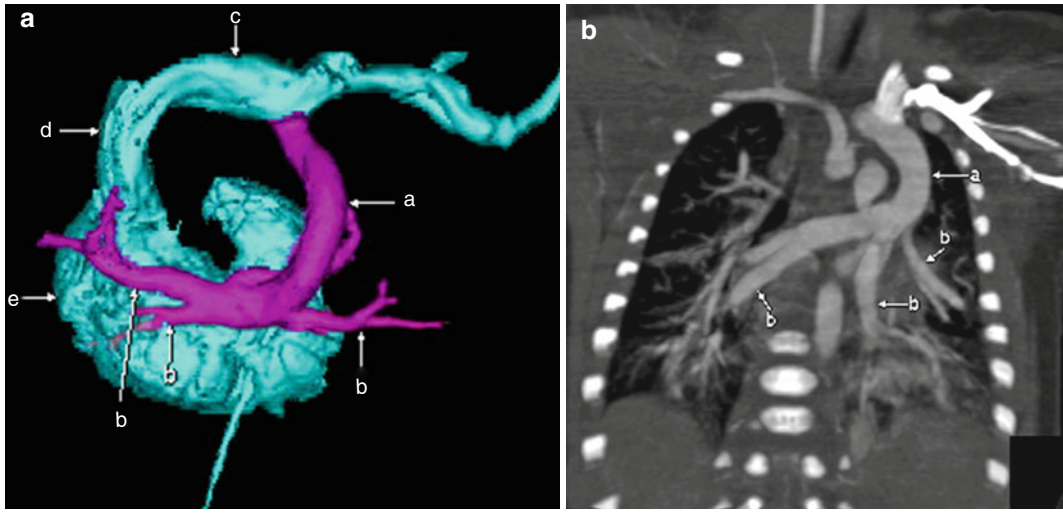

Fig. 5.7 Posterior projection 3D color model from CTA (**a**) and coronal MIP (**b**) images demonstrating multiple pulmonary veins (*b*) draining to a vertical vein (*a*), which connects to the brachiocephalic vein (*c*) and to the SVC (*d*) back to the systemic atrium (*e*)

Intracardiac TAPVR

Intracardiac TAPVR is also a left-to-right shunt that requires an obligatory right-to-left shunt through an ASD for adequate systemic cardiac output. Thus, patients may appear cyanotic. TAPVC draining into the coronary sinus is the second most common type of TAPVC; direct right atrial drainage is less common. Pulmonary venous obstruction is unusual.

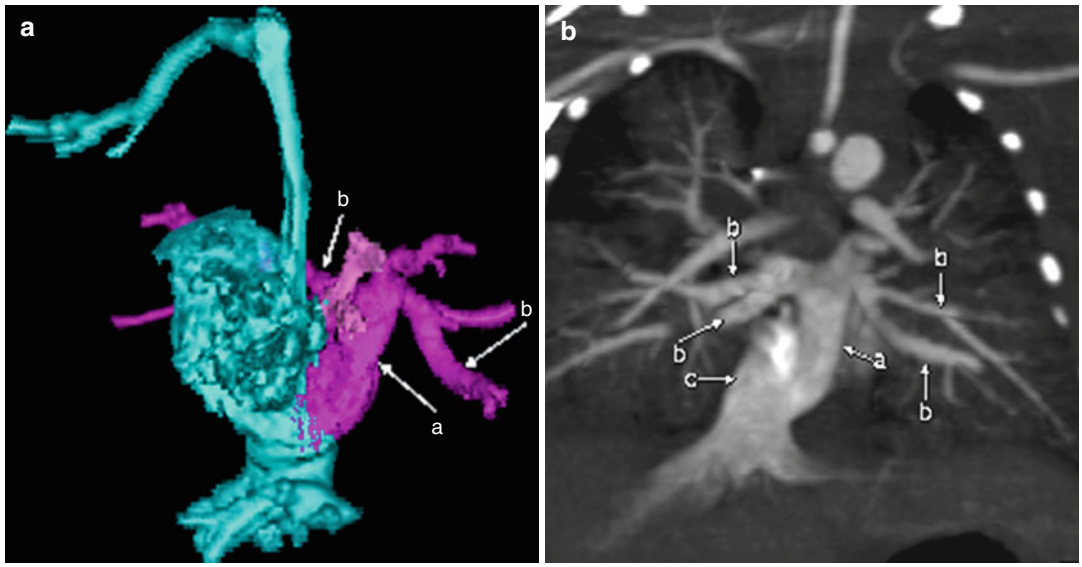

Fig. 5.8 Anterior projection 3D color model (**a**) and coronal MIP image (**b**) showing multiple pulmonary veins (*b*) draining directly to the coronary sinus (*a*) back to the right atrium (*c*)

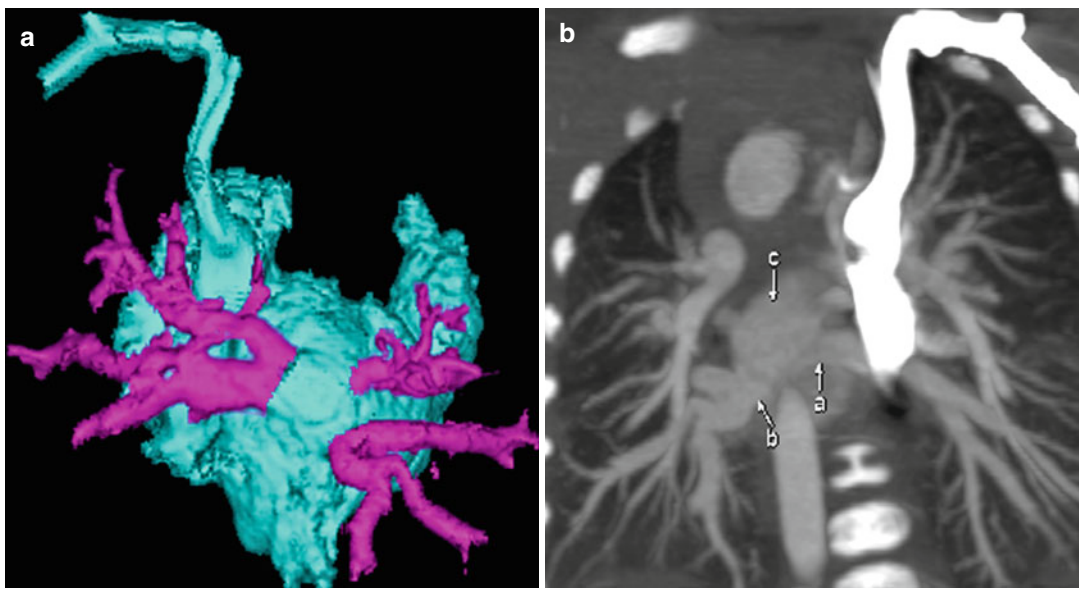

Fig. 5.9 Posterior projection 3D color model (**a**) and coronal MIP image (**b**) demonstrating multiple pulmonary veins (*pink*, *a* and *b*) draining directly to the right atrium (*light blue*, *c*) in a patient with TAPVR

Infracardiac TAPVR

In infracardiac TAPVR, the pulmonary veins drain to a vertical vein that empties into the IVC, portal vein, or hepatic veins. This type of TAPVR is associated with congestive heart failure due to obstruction of the pulmonary venous drainage. Pulmonary venous obstruction also may result from narrowing of the vertical vein as it passes through the diaphragm, narrowing at the insertion into the intrahepatic vessel, with blood having to pass through the hepatic capillary bed (if the insertion is to a portal vein) or a hypoplastic vertical vein. This may be associated with asplenia or polysplenia.

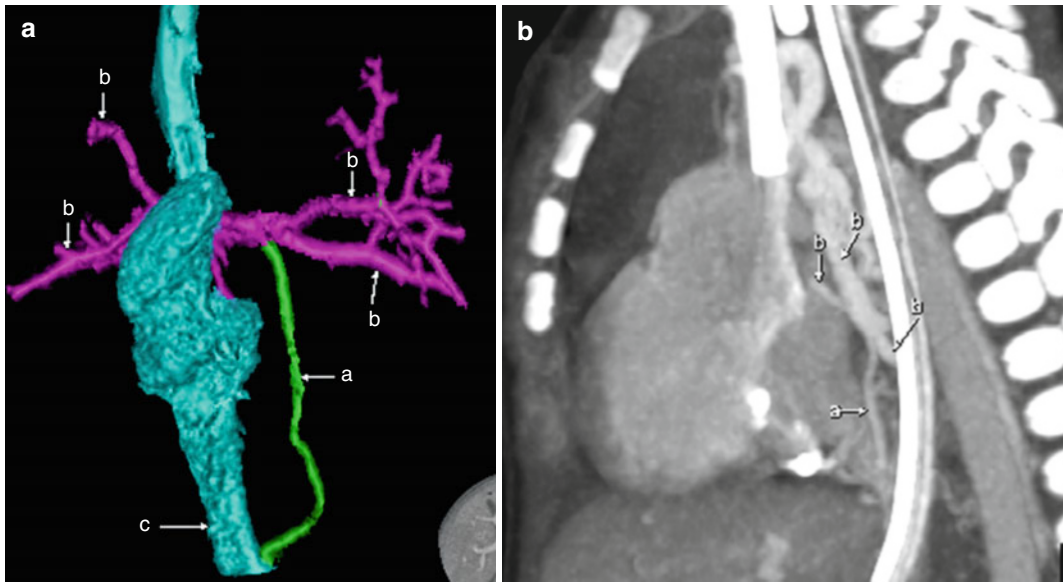

Fig. 5.10 Frontal oblique 3D color model (**a**) and sagittal MIP image (**b**) demonstrating multiple pulmonary veins (*b*) draining to a markedly hypoplastic vertical vein (*a*), which connects to the IVC (*c*)

Mixed-Type TAPVR

Mixed-type TAPVR, the least common form, is a combination of the previously described anomalous connections. In this condition, CTA is effective in delineating complex anatomy.

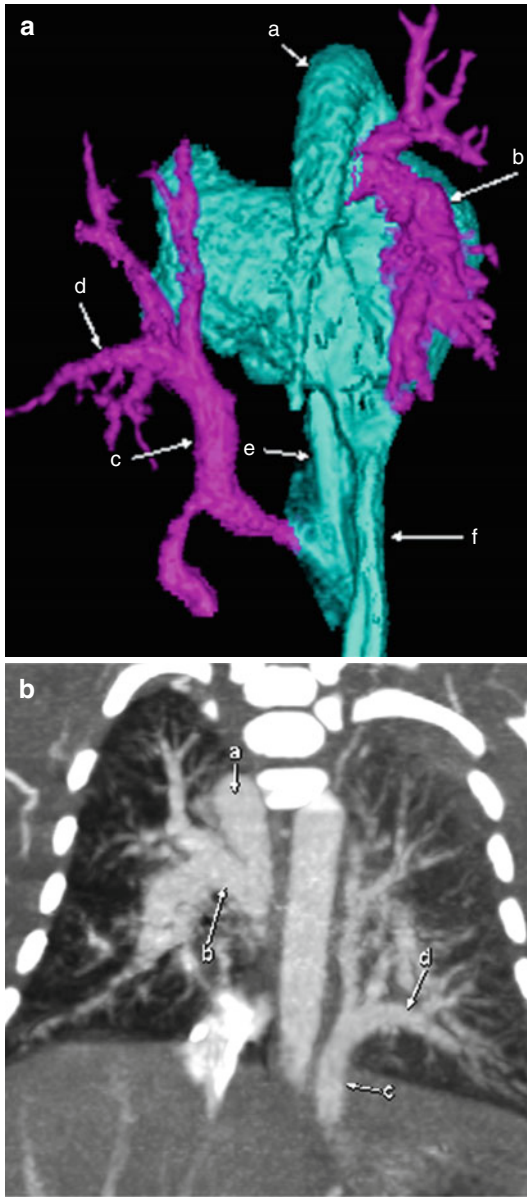

Fig. 5.11 Posterior projection of a 3D color model (**a**) and coronal MIP image (**b**) demonstrating mixed-type TAPVR. The left pulmonary veins (*d*) drain to the IVC (*e* and *f*) via a vertical vein (*c*). The right pulmonary veins (*b*) drain directly to the azygous vein (*a*)

Hypogenetic Lung (Scimitar) Syndrome

In hypogenetic lung syndrome, also known as scimitar syndrome, there is a triad of findings: (1) PAPVR of the right lung, typically to the IVC or right atrium, (2) a hypoplastic right lung and right pulmonary artery, and (3) a systemic arterial supply to the right lower lobe. Patients are acyanotic and typically asymptomatic. The name *scimitar* comes from the curved appearance of the right lower lobe pulmonary vein, which becomes larger as it approaches the diaphragm and may look like a scimitar sword, especially on chest radiography.

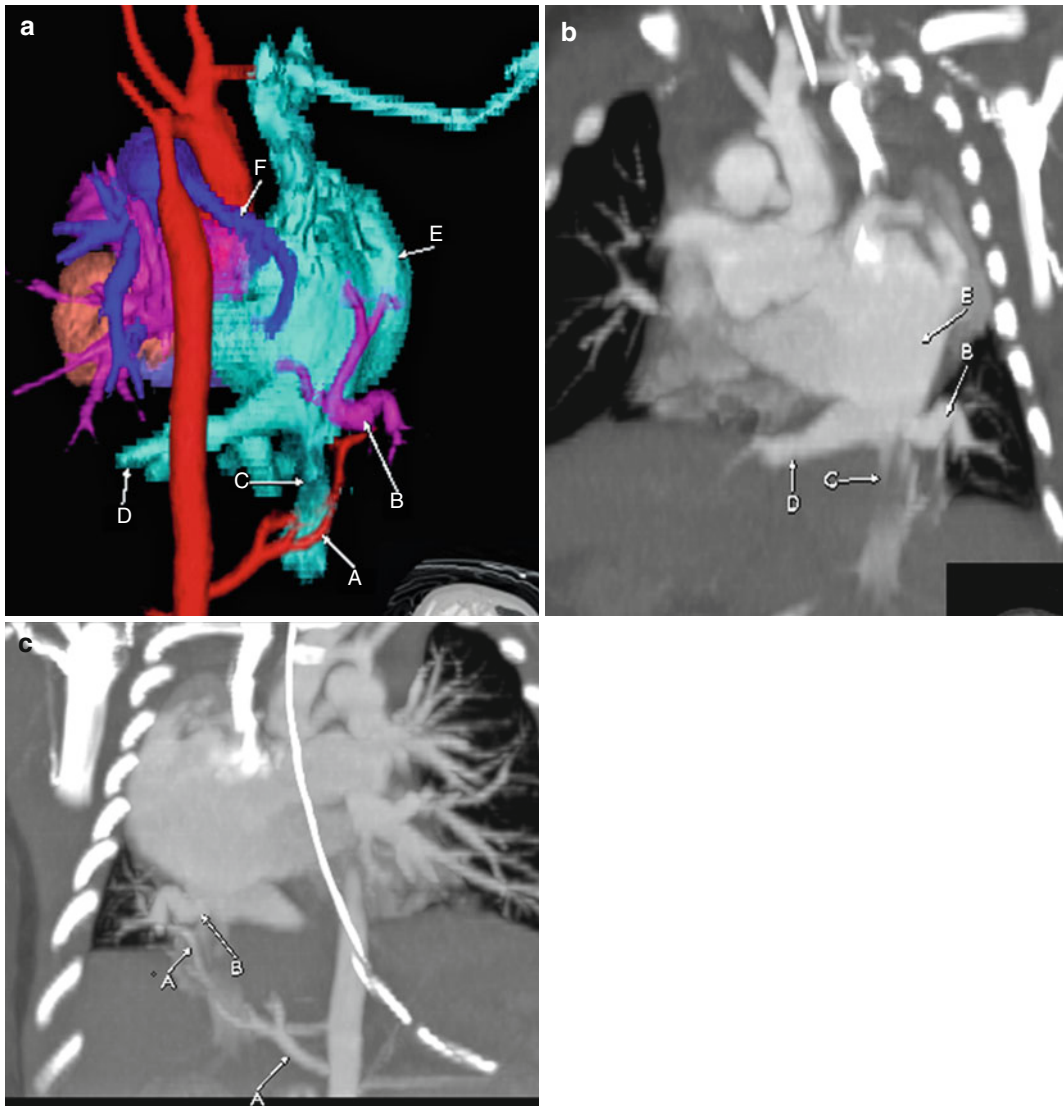

Fig. 5.12 Posterior projection color-coded 3D model (**a**) and coronal MIP images (**b** and **c**) from a chest CTA showing right lower lobe partial anomalous venous return (*B*) to the junction between the IVC (*C*) and right atrium (*E*). There also is a systemic arterial supply (*A*) to the right lower lobe from the descending aorta and a hypoplastic right pulmonary artery (*F*). Notice the drainage of the hepatic veins (*D*) to the right atrium

Azygous Continuation of the IVC

In azygous continuation, the prehepatic suprarenal IVC is interrupted, with most of the lower extremity venous return occurring through a dilated azygos (usually) or hemiazygos vein; the intrahepatic IVC is absent. Typically, the hepatic veins form a confluence of a posthepatic IVC that drains normally into the right atrium. Rarely, the hepatic veins drain directly into the right atrium. This condition is associated with ASD, ventricular septal defect (VSD).

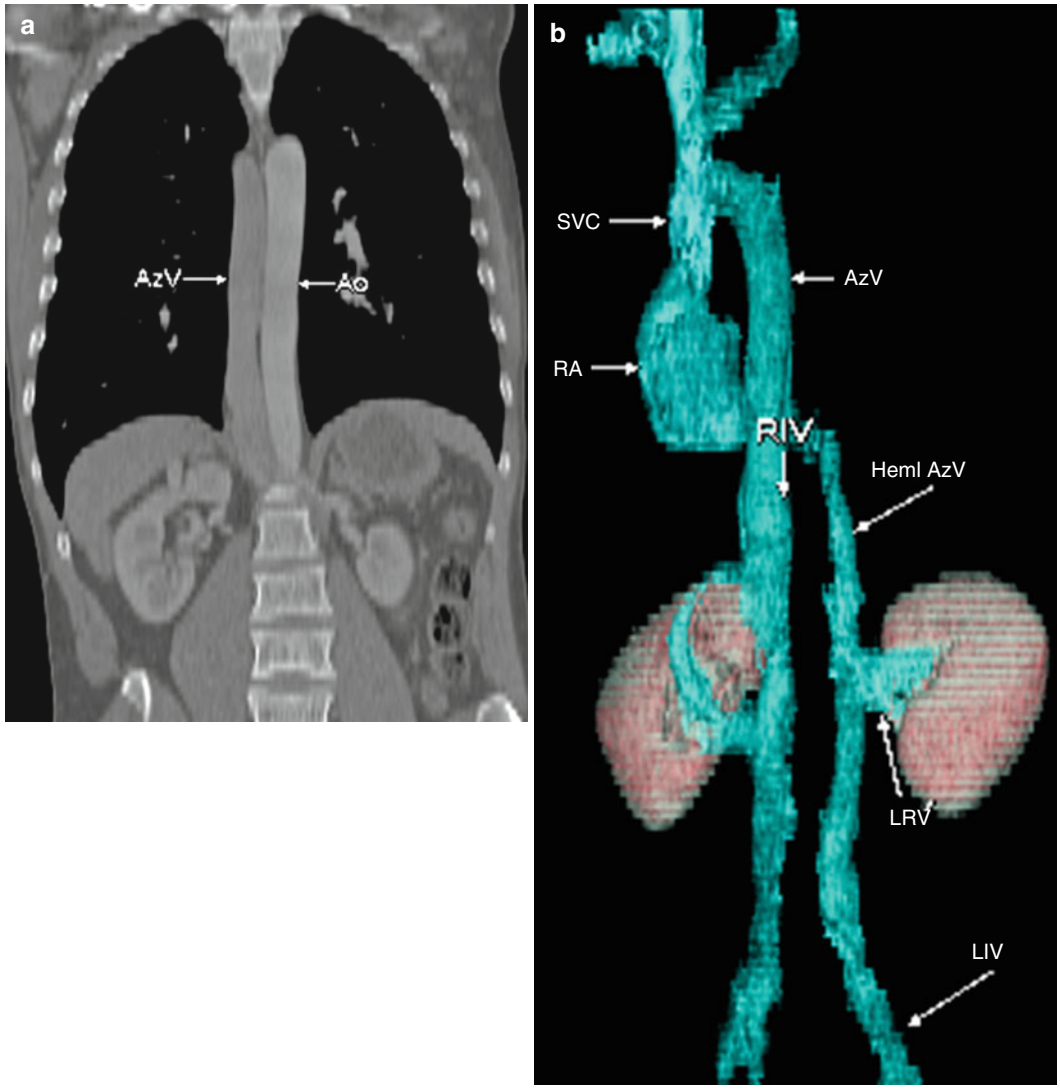

Fig. 5.13 Coronal MIP image (**a**) and anterior projection of a 3D color model (**b**) demonstrating a markedly dilated azygos vein (*AzV*) receiving inflow from an interrupted IVC (*RIV*) and the hemiazygos vein (*Hemi AzV*). Left-sided lower-extremity blood drains to the left common iliac vein (*LIV*), which coalesces with the left renal vein (*LRV*) into the Hemi AzV (variant duplicated caval system). No intrahepatic IVC was present on subsequent images. *Ao* aorta, *SVC* superior vena cava, *RA* right atrium

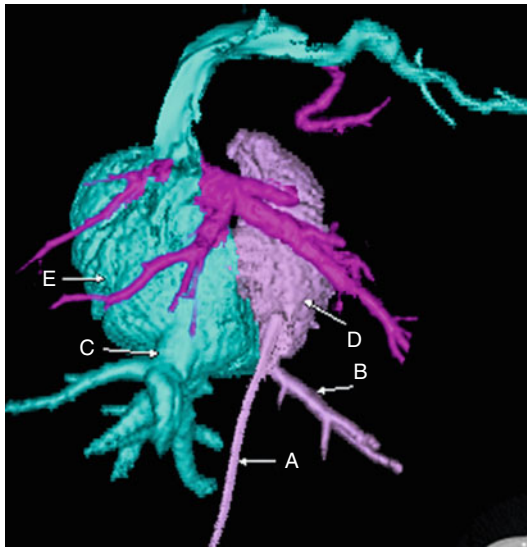

Fig. 5.14 Posterior projection of a 3D color CTA scan showing an umbilical venous catheter (*A*) coursing through one of the right-sided hepatic veins (*B*) to enter the right aspect of the right atrium (*D*). A left-sided IVC (*C*) enters the left aspect of the right atrium (*E*) as well. The presence of normal anatomic hepatic venous drainage in addition to a left IVC is evidence of a duplicated caval system

Duplicated (Bilateral) IVC

In duplicated, or bilateral, IVC, the left common iliac vein ascends to the left of the aorta rather than merging with the right common iliac vein. At the level of the left renal vein, it receives renal inflow and usually crosses anterior to the aorta to unite with the right-sided IVC. Duplicated IVC may be asymmetric, with the right side usually dominant and larger. Incomplete IVC duplication is common (see Fig. 5.13), with only separation of the hepatic venous drainage.

Patent Foramen Ovale

In patent foramen ovale (PFO), there is a normal interatrial channel in the posteroinferior atrial septum that allows blood flow to bypass the pulmonary circulation *in utero*. At birth, because the left atrial pressure exceeds the pressure in the right atrium, a preexisting left atrial tissue flap closes the channel across the fossa ovalis. This usually is permanent. It may remain open in conditions in which the right atrial pressure is elevated, such as tricuspid atresia or Ebstein's anomaly. An asymptomatic right-to-left shunt may exist into adulthood.

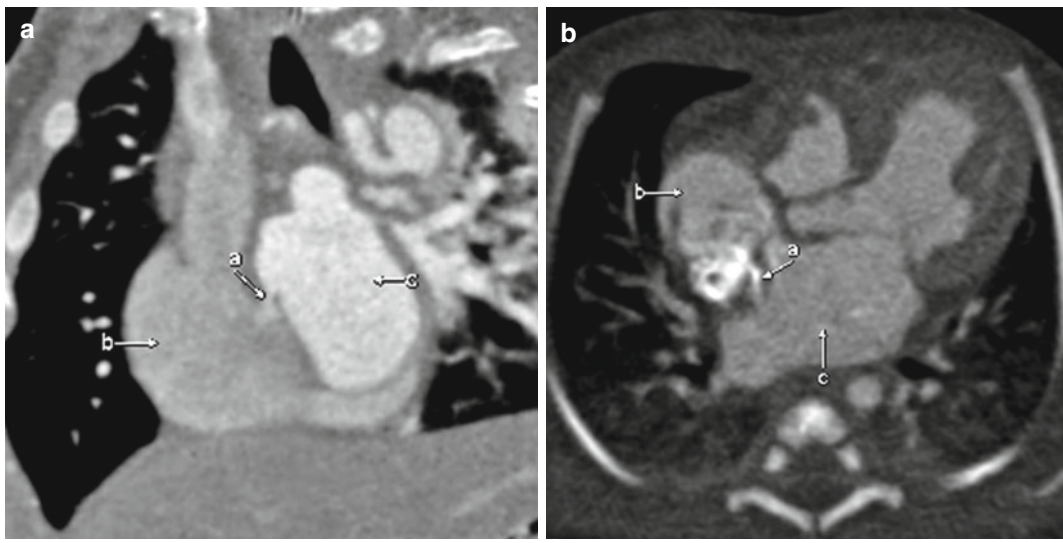

Fig. 5.15 Coronal (**a**) and axial MIP (**b**) images demonstrating a jet of contrast passing through a PFO (*a*) between the right (*b*) and left (*c*) atria

Ostium Secundum ASD

Ostium secundum ASD is the most common ASD type. It is located in the midportion of the atrial septum and typically is a result of the deficiency of the atrial septum primum to form a flap with the atrial septum secundum. It may be asymptomatic into adulthood or eventually result in shunt reversal and Eisenmenger's syndrome. It may close spontaneously or require percutaneous intervention and flap closure.

Ostium Primum ASD

In ostium primum ASD, the most basic of the endocardial cushion/AV septal defects, the most anteroinferior atrial septum is deficient. This condition may coexist with a defect in the anterior mitral valve leaflet and may or may not be associated with a VSD.

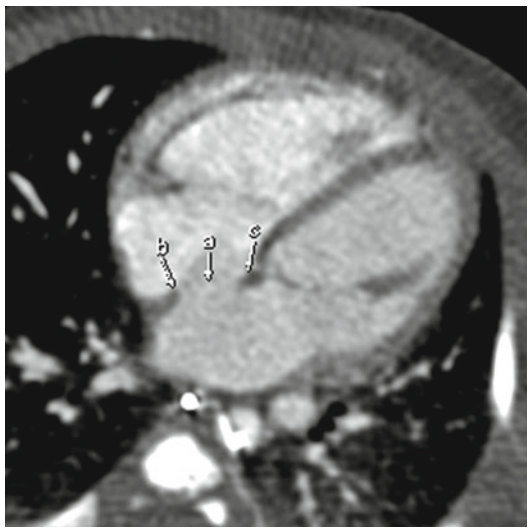

Fig. 5.16 Axial image showing a large secundum-type ASD (*a*) bordered by the posteromedial (*b*) and anterolateral (*c*) flaps of the atrial septum

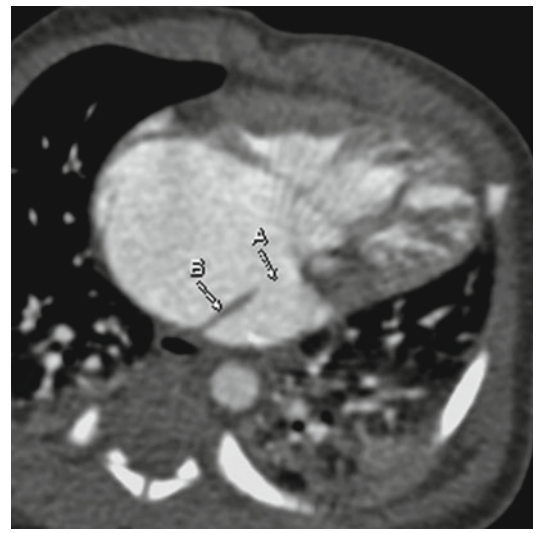

Fig. 5.17 Axial CTA image demonstrating an interatrial connection through a septum primum–type ASD (*A*). The posterior atrial septum (*B*) is intact

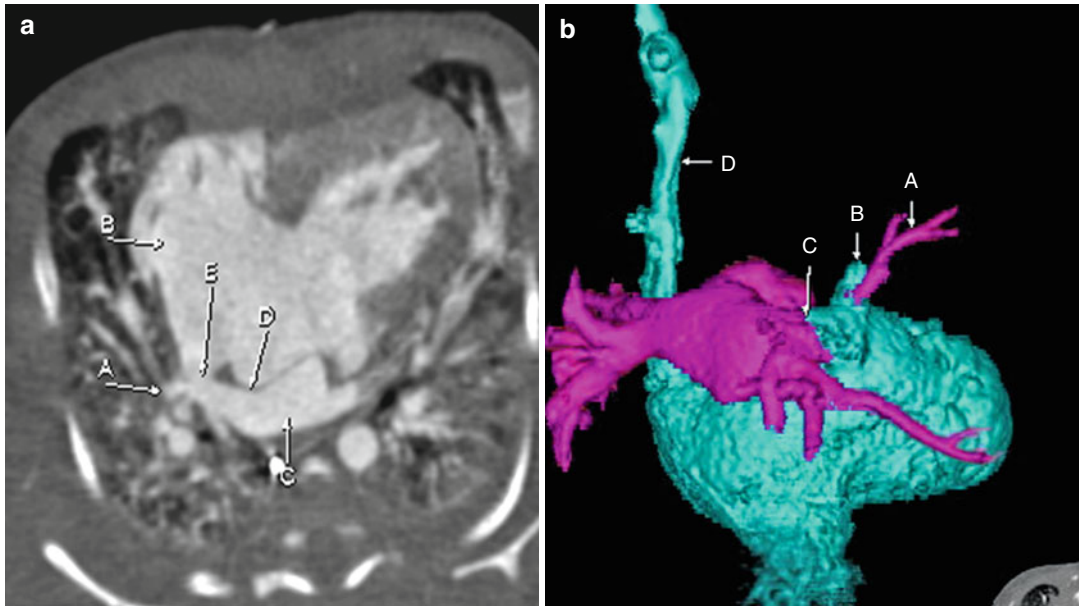

Fig. 5.18 Axial cardiac CTA (**a**) and posterior projection 3D model (**b**) images showing a posterosuperior defect (*E*) in the atrial septum (*D*) with communication between the left (*C*) and right atria (*B* in axial image). This nearly always is associated with PAPVR of the right upper lobe (*A*), which drains to the junction of the SVC (*B* in 3d image) and right atrium

Sinus Venosus–Type ASD

Sinus venosus–type ASD occurs in the posterosuperior or posteroinferior atrial septum and is contiguous with the SVC or IVC, respectively. It nearly always is associated with right upper lobe PAPVR because of the deficiency of the septum separating the right atrium from the left atrial–right pulmonary venous junction. Occasionally, a pulmonary vein is inserted high up the SVC, distant from the sinus venosus ASD.

Complete AV Septal Defect (Complete Common AV Canal Defect)

Complete AV septal defect is a defect in the development of the endocardial cushion to form the tricuspid and mitral valves, and to complete the septation of the atrial septum and ventricular septum. This results in a common AV valve with variable chordal attachments to the ventricle, ostium primum ASD, and AV canal–type (inlet)

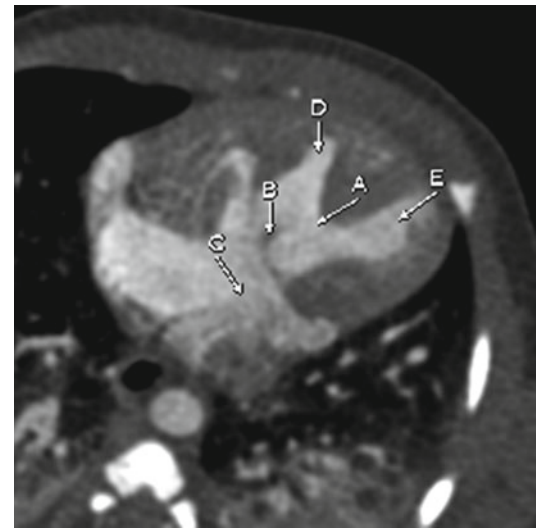

Fig. 5.19 Axial CTA image demonstrating a septum primum ASD (*C*) as well as a posterior-type VSD (*A*). Blood from the atria and from the left (*E*) and right (*D*) ventricles may mix freely. A common AV valve (*B*) is present.

VSD. This may be associated with trisomy 21 as well as heterotaxy syndromes. There also may be a concomitant presence of tetralogy of Fallot.

Tricuspid Atresia

Tricuspid atresia is a congenital agenesis of the right AV (tricuspid) valve, preventing forward flow of systemic venous blood from the right atrium to the ventricle. Newborns with tricuspid atresia must have a coexisting ASD to survive. This condition may be associated with a VSD allowing pulmonary perfusion, or transposition of the great arteries. The size of the heart depends on the size of the VSD. If there is no VSD and the great arteries have a normal relationship, there will be pulmonary atresia, and the patient requires ductal patency for survival.

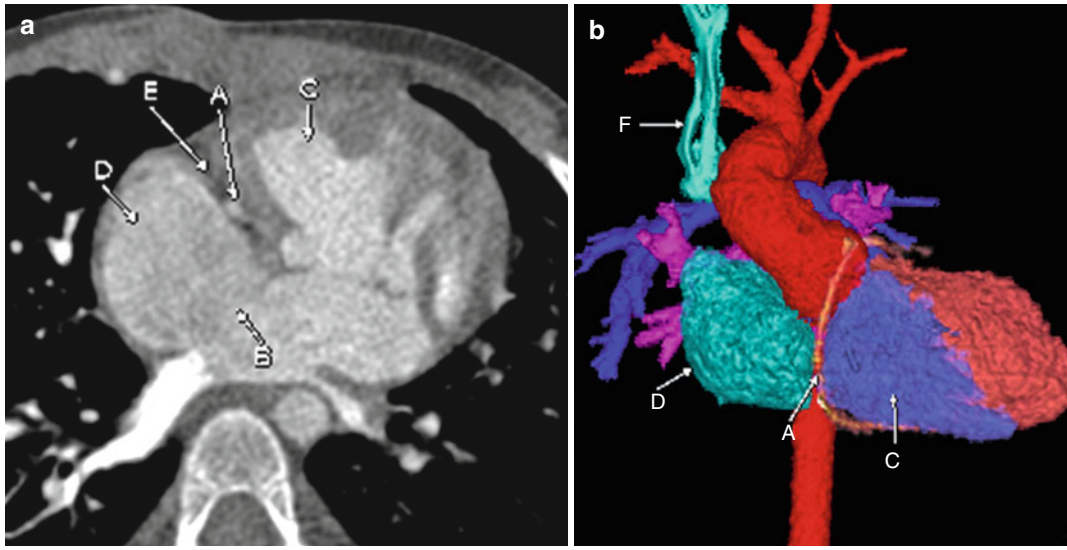

Fig. 5.20 (**a**) Axial CTA image showing fat (*E*) and fibrous tissue, with the right coronary artery (*A*) in the expected location of the tricuspid valve. Blood cannot pass directly from the right atrium (*D*) to the right ventricle (*C*), so it crosses through an ASD (*B*) into the left atrium. (**b**) Anterior projection 3D model demonstrating SVC (*F*) drainage to the pulmonary arteries from a post-surgical Glenn shunt

Ebstein's Anomaly

In Ebstein's anomaly, the septal leaflets of the tricuspid valve may be dysplastic, hypoplastic, and displaced. The anterior leaflet typically is large and sail-like; it may have tethering into the RV free wall and may extend up to the RV outflow tract apically into the right ventricle, leading to "atrialization" of the right ventricle. The posterior variable degree of non-coaptation of the leaflets results in tricuspid regurgitation, which often is severe and results in massive cardiomegaly.

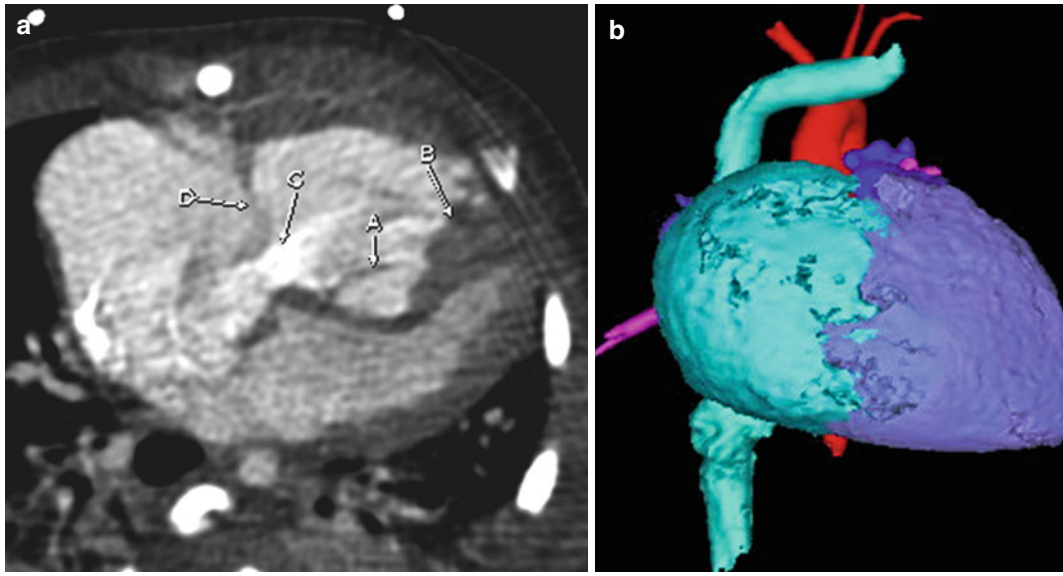

Fig. 5.21 (**a**) Axial cardiac CTA image demonstrating the sail-like, apically displaced septal leaflet (*A*) of the tricuspid valve, which is tented by dysplastic cordae tendineae (*B*) seen along the apicoseptal portion of the left ventricle. The anterior leaflet (*D*) of the tricuspid valve is visualized in a relatively normal position. This causes regurgitation along the septal portion of the valve (*C*). (**b**) Frontal projection color 3D image showing massive dilatation of the right atrium (*light blue*) and right ventricle (*purple*)

Mitral Stenosis

Mitral stenosis may result from congenital heart disease or acquired heart disease, classically from rheumatic fever. In this condition, an impediment to left ventricular inflow results in left atrial enlargement. Eventually, elevated left atrial and pulmonary pressures result in pulmonary edema and pulmonary hypertension.

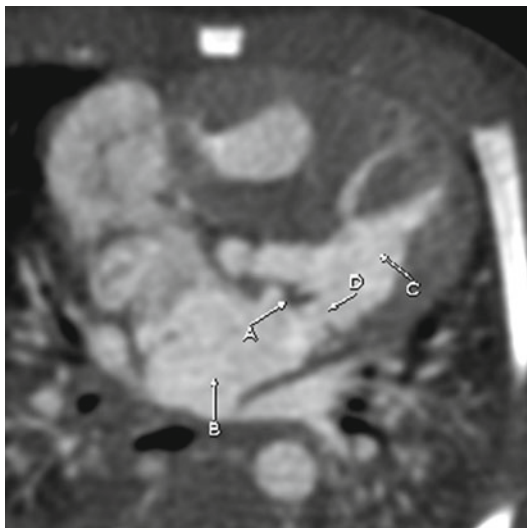

Fig. 5.22 Axial cardiac CTA image demonstrating a thickened anterior mitral leaflet (*A*), resulting in a stenotic mitral orifice (*D*) for oxygenated blood to flow from the left atrium (*B*) to the ventricle (*C*) during diastole. The left atrium is mildly enlarged

Single Atrium

Single atrium is the result of a development failure in the components of the atrial septum. This condition typically is part of a heterotaxy syndrome, usually with a complete AV septal leaflet. It may be associated with single ventricle, usually a single right ventricle.

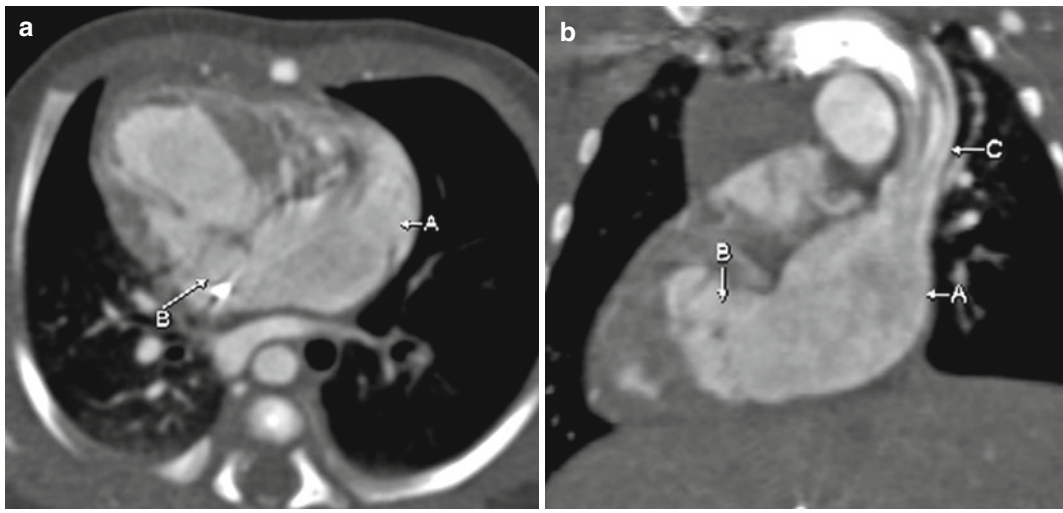

Fig. 5.23 Axial (**a**) and coronal (**b**) cardiac CTA images showing a right (*B*) and left (*A*) atria with no formed interatrial septum. Notice the left- sided SVC (*C*), which drains to the left atrium in this patient with asplenia-type heterotaxy