

Jonathan R. Dillman¹ Sai G. Yarram^{1,2} Ramiro J. Hernandez¹

Keywords: anomalous pulmonary venous connection, congenital anomaly, CT, MRI, pulmonary veins

DOI:10.2214/AJR.08.1526

Received July 12, 2008; accepted after revision October 1, 2008.

¹Department of Radiology, Section of Pediatric Radiology, University of Michigan Health System, C. S. Mott Children's Hospital, 1500 E Medical Center Dr., Ann Arbor, MI 48109. Address correspondence to J. R. Dillman (jonadill@umich.edu).

²Present address: FW Radiology, Fort Wayne, IN

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AJR 2009; 192:1272-1285

0361-803X/09/1925-1272

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Imaging of Pulmonary Venous Developmental Anomalies

OBJECTIVE. The purpose of this article is to review pulmonary venous embryology and to present the imaging findings of a variety of pulmonary venous developmental anomalies, including total anomalous pulmonary venous connection, partial anomalous pulmonary venous connection, partial anomalous pulmonary venous connection, pulmonary venous connection, pulmonary venous connection, and cor triatriatum.

CONCLUSION. There are numerous developmental pulmonary venous anomalies. Although these conditions have traditionally been evaluated with echocardiography and angiography, they can be accurately diagnosed using both MRI and MDCT.

bnormal embryonic pulmonary vein development may result in a wide spectrum of congenital anomalies. Although pulmonary venous developmental anomalies have been evaluated traditionally with echocardiography and angiography, MRI and MDCT are playing increasing roles in their characterization. This article will review pulmonary venous embryology and present the imaging findings of numerous pulmonary venous developmental anomalies, including total anomalous pulmonary venous connection (TAPVC), partial anomalous pulmonary venous connection (PAPVC), pulmonary vein stenosis and hypoplasia/atresia, and cor triatriatum.

Embryology of Pulmonary Veins

The development of the pulmonary veins is a complicated process that occurs early in embryonic life. Several theories regarding pulmonary vein formation have been proposed based on human and animal embryonic research [1]. One commonly accepted theory suggests that blood returning from the lung buds initially drains into the splanchnic plexus, which communicates with paired cardinal veins as well as umbilicovitelline veins. The right cardinal venous system eventually develops into the right superior vena cava (SVC), whereas the left cardinal venous system mostly disappears and may potentially develop into a left SVC (seen in < 1% of individuals). The umbilicovitelline veins develop into the inferior vena cava (IVC), portal venous system, and ductus venosus. A primitive common pulmonary vein arises as an outpouching from the dorsal wall of the left atrium. With time, the common pulmonary vein communicates with the portion of the splanchnic plexus that drains blood flow from the lungs. Pulmonary venous connections to the cardinal and umbilicovitelline veins normally involute, and the common pulmonary vein becomes incorporated into the dorsal wall of the left atrium, ultimately typically giving rise to four separate pulmonary veins [2].

Pulmonary venous developmental anomalies happen if any of these processes fails to occur properly. If the common pulmonary vein fails to connect to the splanchnic plexus and a splanchnic plexus communication with a cardinal or umbilicovitelline vein persists, some type of TAPVC or PAPVC will occur. If the common pulmonary vein fails to properly incorporate into the dorsal left atrial wall, pulmonary vein stenosis/atresia or cor triatriatum will occur [2].

Imaging Techniques

Echocardiography is the initial imaging technique of choice in the evaluation of congenital heart disease, including pulmonary venous anomalies [2, 3]. Although echocardiography has a variety of strengths, including portability and lack of ionizing radiation, evaluation of pulmonary veins may be suboptimal in some patients. This is because echocardiography has a limited field of view ("echo window") that may prevent assessment of pulmonary venous structures peripheral to the expected location of the left atrial ostia. In

addition, this imaging technique may not be able to completely characterize or even detect more distant associated vascular anomalies.

CT excellently depicts vascular structures peripheral to the heart in the thorax [2-5]. Additional benefits of this imaging technique when using newer MDCT scanners include very rapid imaging that may obviate patient sedation and multiplanar reformatting capabilities. Axial and 3D reconstructed images both excellently depict anomalous pulmonary venous structures with statistically similar detection rates that approach 100% [4]. The primary disadvantage of CT is that it requires the use of ionizing radiation. CT also requires the use of IV iodinated contrast material, which may adversely affect renal function or, rarely, result in acute allergic-like reactions. In addition, timing of the IV contrast material bolus is critical because a suboptimally timed bolus may limit opacification of pulmonary venous structures. CT may be useful for imaging pulmonary venous structures in patients who are incompletely evaluated by echocardiography and who cannot, for whatever reason, undergo an MRI examination.

Regarding CT technique, as thin as possible detector collimation (or detector configuration) should be used to produce isotropic reformatted and 3D volume-rendered images. Nonionic low-osmolality (or iso-osmolality) iodinated contrast material (1.5-2.0 mL/kg, 120-150 mL maximum volume) is typically administered IV using a power injector at injection rates ranging from 1 to 4 mL/s, depending on patient weight and quality of IV access. Hand injection of contrast material may be required in small children with tenuous venous access. In the evaluation of PAPVC, dual anomalous pulmonary venous drainage, and cor triatriatum in older patients, timing of imaging after the injection of contrast material may be precisely determined using either a timing bolus or realtime contrast material tracking. For patients with suspected TAPVC as well as for small children, we typically image immediately after the injection of contrast material at our institution. Cardiac gating is not required for the evaluation of pulmonary venous structures, although it may prove useful if the patient is being specifically evaluated for cor triatriatum or central pulmonary vein hypoplasia or stenosis.

MRI is the preferred imaging technique for the evaluation of pulmonary venous structures after echocardiography. MRI can be used to depict complex thoracic cardiovascular anomalies, including pulmonary vein abnormalities [3, 6–9]. Advantages of this imaging technique include a lack of ionizing radiation, multiplanar capability, and the ability to acquire multiple imaging phases using a single IV bolus of gadolinium-containing contrast material. Disadvantages of MRI include the amount of time required for image acquisition, the frequent need for patient sedation, and its susceptibility to metal-related artifact.

The pulmonary venous system can be evaluated using a variety of MRI techniques [6, 8–10]. Images are ideally acquired using a dedicated phased-array cardiac coil [3]. Both black blood and bright blood images may be obtained without IV contrast material [6]. High-resolution double-inversion-recovery fast (or turbo) spin-echo (or black blood) images are typically acquired to assess anatomy [6]. Gradient-recalled echo (GRE) and 2D balanced steady-state free precession (SSFP) pulse sequences provide bright blood cine images that are useful for the evaluation of the cardiac chamber and valvular function [6, 8]. These sequences can also be used to evaluate the central pulmonary veins and the left atrium. Respiratorytriggered ECG-gated free breathing 3D balanced SSFP pulse sequences allow the acquisition of a near-isotropic volumetric data set that can be reformatted and reviewed in any plane.

Gadolinium-enhanced MR angiography (MRA) is an extremely valuable tool in the evaluation of pulmonary venous structures [8-10]. This non-ECG-gated 3D spoiled GRE imaging technique allows rapid dynamic imaging of thoracic vascular structures, including during arterial and venous (pulmonary and systemic) vascular phases [8-10]. Images can be acquired using multiple breath-holds or during quiet breathing [10]. Gadolinium-containing contrast material may be either injected by hand or power-injected at < 1-2 mL/s using a double dose (0.20 mmol/kg) [3, 8-10]. These images have a large field of view, may be acquired in either the sagittal or coronal plane, and possess excellent spatial resolution [3, 9, 10]. Although reviewing the individual source images is important, these images are also typically reformatted in multiple planes as well as three-dimensionally reconstructed as either maximum-intensity-projection (MIP) or volume-rendered images [3, 8-10].

Total Anomalous Pulmonary Venous Connection

TAPVC or total anomalous pulmonary venous return (TAPVR) accounts for approximately 1–5% of cardiovascular congenital anomalies [7, 11, 12]. This condition is a cause of neonatal cyanosis and may rapidly result in death when blood is not shunted from the right heart (or pulmonary circulation) to the left heart (or systemic circulation). This shunting typically occurs through either an atrial septal defect (ASD)/patent foramen ovale (PFO) or, less commonly, a patent ductus arteriosus (PDA) [11]. An increased frequency of TAPVC is seen in patients with heterotaxy syndromes, particularly asplenia [7, 12] (Figs. 1 and 2).

TAPVC occurs in four types that are classified on the basis of the location of pulmonary venous drainage, including supracardiac, cardiac, infracardiac, and mixed forms. Supracardiac drainage (type I TAPVC) is most common, accounting for approximately 44% of cases according to a large study of 377 children with TAPVC by Karamlou et al. [12]. In this type, drainage most commonly occurs via a vertical vein to the left brachiocephalic vein. Rarely, supracardiac TAPVC may drain directly to a right SVC, left SVC, or azygous system [7] (Figs. 1 and 2). In the cardiac type (type II TAPVC), which represents approximately 21% of TAPVC cases, pulmonary veins drain either to the coronary sinus or directly into the right atrium [12] (Fig. 3).

The infracardiac type (type III TAPVC) represents approximately 26% of cases of TAPVC and drains below the diaphragm to either a systemic vein-the IVC, a hepatic vein, or the azygos system-or the portal venous system [12] (Fig. 4). The draining pulmonary veins in this type of TAPVC may be obstructed, frequently at the level of the diaphragm, because of extrinsic narrowing, resulting in neonatal pulmonary edema with a normal-size cardiac silhouette on chest radiography. Although pulmonary venous obstruction may occur in any type of TAPVC (Figs. 1 and 2), it is most common in the infracardiac form, being present in up to 78% of patients [7, 12].

The final type of TAPVC is diagnosed when the location of pulmonary vein drainage is mixed (type IV TAPVC). In this form, pulmonary veins drain to at least two different locations, including a brachiocephalic vein, SVC, azygos vein, coronary sinus, right atrium, or below the diaphragm. This type of pulmonary venous drainage accounts for approximately 9% of TAPVC cases [12] (Figs. 5–7).

Accurate diagnosis of TAPVC and knowledge of the exact pattern of pulmonary venous drainage are important to allow appropriate preoperative planning. Contrast-enhanced MRA and CT are particularly valuable in the characterization of mixed forms of TAPVC; echocardiography may be markedly limited in such cases. Advances in diagnostic imaging and refined surgical techniques have allowed markedly decreased postoperative mortality in TAPVC patients over the last several decades. Karamlou et al. [12] have documented 5-year survival rates of 97% for TAPVC patients repaired since 2000.

Partial Anomalous Pulmonary Venous Connection

PAPVC or partial anomalous pulmonary venus return (PAPVR) has a prevalence of 0.4-0.7% and may be incidentally detected on either CT or MRI [5, 7, 11]. In PAPVC, at least one pulmonary vein drains to a location other than the left atrium. Patients with PAPVC are typically acyanotic and most commonly only mildly symptomatic or asymptomatic. PAPVC is thought to more frequently affect the right lung than the left lung, although PAPVC affecting the left lung may be more often detected at CT [5, 13]. This partial anomalous drainage results in a left-to-right shunt, similar to an ASD, ventricular septal defect (VSD), or PDA. Some authors have suggested that PAPVC becomes clinically significant when 50% or more of the pulmonary blood flow returns anomalously [5]. The ratio of pulmonary to systemic blood flow can be accurately quantified using velocity-encoded phase contrast MRI.

PAPVC most commonly involves the anomalous drainage of the right superior pulmonary vein to the right atrium or SVC (Fig. 8). This pattern of pulmonary venous drainage is frequently associated with a sinus venosus ASD [7]. Left anomalous pulmonary venous structures commonly drain to the left brachiocephalic vein via a vertical vein (Fig. 9) or to the coronary sinus [7]. Other patterns of PAPVC may also be observed (Fig. 10).

Scimitar syndrome—hypogenetic lung or pulmonary venolobar syndrome—is a rare form of PAPVC that almost always involves the right lung [14]. In this condition, a portion or all of right lung pulmonary venous blood flow drains to the IVC (above or below the diaphragm), the azygos system, right atrium, portal venous system, or a hepatic vein [7, 13]. Other findings observed in patients with scimitar syndrome include a small ipsilateral hemithorax, cardiac dextroposition, and pulmonary artery hypoplasia or aplasia. Systemic arterial collateral blood supply to the ipsilateral lung may also be seen arising from the aorta or its branch vessels such as the celiac axis [14] (Fig. 11).

Anomalous dual pulmonary venous drainage is present when a portion of lung drains to both the left atrium and a systemic venous structure. Patients with anomalous dual drainage may be asymptomatic or may present later in life with congestive heart failure [15, 16]. Reports in the literature have described transcatheter endovascular management of this condition by either embolizing (or occluding) the anomalous pulmonary vessel or covering the vessel's anomalous insertion with an endograft [15, 16] (Fig. 12).

Although it is not traditionally considered a form of PAPVC, anomalous pulmonary venous drainage in the setting of extralobar pulmonary sequestration also occurs [7]. Extralobar pulmonary sequestration is a congenital malformation in which a portion of lung fails to communicate with the central tracheobronchial tree. These structures are invested in pleura that is separate from that investing the remaining normal lung [14, 17]. In addition, there is typically systemic arterial supply from the aorta or a branch vessel as well as lack of normal pulmonary venous drainage to the left atrium. Venous drainage may be either systemic-to the IVC or azygos/hemiazygos system-or to the portal venous system. Extralobar pulmonary sequestrations can be identified by ultrasound, CT, or MRI. Contrast-enhanced CT or MRI reformatted images can be used to elucidate feeding arterial and draining venous structures [14] (Fig. 13). These structures are typically left-sided [14, 17] and may occur above, within, or below the diaphragm. Additional anomalies, such as congenital diaphragmatic hernia and congenital cystic adenomatoid malformation, may also coexist [14, 17].

Congenital Pulmonary Vein Stenosis and Hypoplasia/Atresia

Congenital pulmonary vein stenosis and hypoplasia/atresia reflect a spectrum of the same abnormality and may affect individual or multiple pulmonary veins. The abnormality may be focal or may involve a long segment of pulmonary vein [2, 11]. Congenital pulmonary vein stenosis and hypoplasia/atresia are rare causes of neonatal pulmonary edema that may be unilateral or bilateral (Figs. 14 and 15). This condition may also present later in life with pulmonary artery hypertension or hemoptysis [2]. A recent study by Ou et al. [18] determined that 64-MDCT without gating more completely characterized pulmonary vein stenosis than did echocardiography in children. Congenital pulmonary vein stenosis is associated with congenital heart disease in approximately 50% of affected patients [2].

Cor Triatriatum

Cor triatriatum is due to faulty incorporation of pulmonary venous structures into the left atrium. In this condition, the left atrium is divided into proximal (or posterior) and distal (or anterior) chambers by a diaphragm or membrane (Figs. 16 and 17). The posterior chamber receives blood flow from the pulmonary veins, whereas the anterior chamber delivers blood to the mitral valve. Blood may flow from the posterior chamber to the anterior chamber through a widely patent or restrictive fenestration in the membrane. Alternatively, blood may flow from the posterior chamber to the right atrium via an ASD or anomalous venous structure. Rarely, a patient may present with subtotal cor triatriatum. This form is diagnosed when an accessory left atrial chamber receives a portion of pulmonary venous blood flow, whereas the remaining portion of pulmonary venous blood flow drains to the proper left atrium [11].

Summary

A wide spectrum of pulmonary venous developmental anomalies exist that can be detected by imaging. Accurate characterization of these abnormalities is important because they all can be associated with significant patient morbidity and mortality. Numerous recent studies have confirmed that both MDCT and MRI are noninvasive imaging techniques that should play increasingly important roles in the evaluation of these anomalies. CT and MRI are most useful in the evaluation of patients with mixed TAPVC and PAPVC as well as pulmonary vein stenosis and hypoplasia/ atresia because these specific lesions may be difficult to characterize at echocardiography.

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Fig. 1—3-month-old boy with supracardiac total anomalous pulmonary venous connection (type I), asplenia, dextrocardia, right aortic arch with mirror-image branching, and bilateral superior venae cavae.

A and B, Coronal contrast-enhanced subvolume maximum-intensity-projection MR images show bilateral pulmonary venous drainage to common pulmonary vein (*white asterisks*, A). Common pulmonary vein then drains into diminutive right superior vena cava (*black asterisks*, B). Anomalous vessel (*black arrow*, B), possibly patent ductus arteriosus, is seen communicating between proximal left subclavian and left pulmonary artery. LPA = left pulmonary artery, RPA = right pulmonary artery, LSPV = left superior vena cava, AA = aortic arch. Arrowheads indicate right pulmonary veins.

(Fig. 1 continues on next page)

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Fig. 1 (continued)—3-month-old boy with supracardiac total anomalous pulmonary venous connection (type I), asplenia, dextrocardia, right aortic arch with mirror-image branching, and bilateral superior venae cavae.
C, Contrast-enhanced volume-rendered MR image, posterior view, confirms drainage of common pulmonary vein (*white asterisks*) into small right superior vena cava (*black asterisks*). Note pulmonary vein substruction at level of white arrow. Black arrow indicates anomalous vessel, possibly patent ductus arteriosus.





Fig. 2—1-month-old girl with total anomalous pulmonary venous connection, asplenia, complete atrioventricular septal defect, double-outlet right ventricle, and bilateral superior venae cavae. AA = aortic arch.

A and B, Sagittal oblique (A) and coronal (B) contrast-enhanced subvolume maximum-intensity-projection images show drainage of left inferior pulmonary vein (LIPV) into common pulmonary vein (*asterisks*, A). Vertical vein drains into markedly dilated left superior vena cava (LSVC). Both LSVC and hepatic veins (*asterisks*, B) drain into left atrium (LA). Note moderate obstruction of LSVC as it enters LA (*black arrow*).

(Fig. 2 continues on next page)



Fig. 2 (continued)—1-month-old girl with total anomalous pulmonary venous connection, asplenia, complete atrioventricular septal defect, double-outlet right ventricle, and bilateral superior venae cavae. AA = aortic arch.

C and **D**, Contrast-enhanced volume-rendered MR images in posterior (**C**) and anterior (**D**) views confirm drainage of all pulmonary veins into common pulmonary vein (*white asterisks*, **C**). Small right superior vena cava (RSVC) and inferior vena cava (IVC) drain into right atrium. Note portions of distal aortic arch and descending thoracic aorta have been removed on posterior image. Black asterisks show hepatic veins. LSVC = left superior vena cava, LSPV = left superior pulmonary vein, RSPV = right superior pulmonary vein, RIPV = right inferior pulmonary vein, LIPV = left inferior pulmonary vein, RPA = right pulmonary artery, LPA = left pulmonary artery.



Fig. 3—3-day-old girl with cardiac total anomalous pulmonary venous connection (type II). Side-by-side gray-scale and color Doppler echocardiographic images show common pulmonary vein confluence (PVC) posterior to heart that empties into right atrium (RA) via coronary sinus. LA = left atrium; LV = left ventricle. (Courtesy of Gregory J. Ensing, M.D., University of Michigan Health System, Department of Pediatrics and Communicable Diseases, Ann Arbor, MI)



Fig. 4—5-day-old boy with infracardiac total anomalous pulmonary venous connection (type III). Gray-scale echocardiographic image shows common pulmonary vein confluence (PVC) posterior to heart. Common pulmonary vein (CPV) empties into infradiaphragmatic inferior vena cava. LA = left atrium, RA = right atrium, RV = right ventricle, HV = hepatic vein). (Courtesy of Gregory J. Ensing, M.D., University of Michigan Health System, Department of Pediatrics and Communicable Diseases, Ann Arbor, MI)

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Fig. 5—1-month-old boy with mixed total anomalous pulmonary venous connection (type IV). AA = aortic arch, SVC = superior vena cava, RPA = right pulmonary artery, LPA = left pulmonary artery, RIPV = right inferior pulmonary vein, LIPV = left inferior pulmonary vein, RSPV = right superior pulmonary vein. A, Contrast-enhanced maximum-intensity-projection MR image shows drainage of left superior pulmonary vein (*arrows*) to left brachiocephalic vein via vertical vein (*arrowheads*).

B, Contrast-enhanced volume-rendered MR image, posterior view, shows that left inferior and right pulmonary veins (*asterisks*) drain to right atrium via dilated coronary sinus (CS). Arrowheads = vertical vein.



Fig. 6—6-day-old boy with mixed total anomalous pulmonary venous connection (type IV). Coronal oblique contrast-enhanced maximum-intensity-projection MR image shows that right pulmonary veins drain to superior vena cava (SVC) just above superior cavoatrial junction. Left pulmonary veins drain to left brachiocephalic vein (LBCV) via vertical vein (*arrowheads*). Pulmonary arteries (*asterisks*) are seen as separate structures. RSPV = right superior pulmonary vein, RIPV = right inferior pulmonary vein, LIPV = left inferior pulmonary vein.



Fig. 7—11-day-old girl with cyanosis at birth and mixed total anomalous pulmonary venous connection (type IV). IVC = inferior vena cava, SVC = superior vena cava, LBCV = left brachiocephalic vein.

A, Posterior oblique contrast-enhanced volume-rendered MR image shows drainage of right pulmonary veins (*black arrows*) to superior vena cava (SVC). Asterisks indicate pulmonary arteries.

B, Contralateral posterior oblique contrast-enhanced volume-rendered image shows drainage of left pulmonary veins (LPV) to left hepatic vein. Draining vein is obstructed at level of diaphragm (*black arrow*). AA = ascending thoracic aorta.



Fig. 8—51-year-old man with pulmonary hypertension, right ventricular dilatation, and increase in oxygen saturation between inferior vena cava (IVC) and main pulmonary artery. Contrast-enhanced volume-rendered MR image, posterior oblique view with pulmonary arteries and portion of descending thoracic aorta removed, shows drainage of right pulmonary veins to lower superior vena cava (SVC) (*arrowhead*) and upper right atrium (RA) (*black arrow*). Left pulmonary veins drain normally to left atrium (LA). CAJ = superior cavoatrial junction.

Fig. 9—14-year-old boy with newly diagnosed atrial septal defect and partial anomalous pulmonary venous connection. Contrast-enhanced volume-rendered MR image shows that left superior pulmonary vein drains to left brachiocephalic vein (*black arrowhead*) via vertical vein (VV). Remaining pulmonary veins drain normally to left atrium. Ao = ascending thoracic aorta, MPA = main pulmonary artery, RBCV = right brachiocephalic vein, SVC = superior vena cava.

Fig. 10—7-day-old boy with partial anomalous pulmonary venous connection.





A, Coronal contrast-enhanced subvolume maximum-intensity-projection (MIP) MR image shows anomalous drainage of both right and left inferior pulmonary veins (RIPV

B, Another coronal subvolume MIP image shows drainage of right and left superior pulmonary veins (RSPV and LSPV, respectively) to left atrium (LA).

and LIPV, respectively). Bilateral inferior pulmonary veins form confluence (black arrow) that drains into inferior vena cava.



Fig. 11—5-day-old boy with scimitar syndrome. Coronal contrast-enhanced maximum-intensity-projection MR image shows small right lung and hypoplastic right pulmonary artery (not shown). Note anomalous right lung pulmonary venous drainage (*arrowheads*) to inferior vena cava just below level of right atrium (RA). Artery arising from celiac axis (black arrow) provides systemic arterial supply to right lung. AA = aortic arch, LPA = left pulmonary artery, LIPV = left inferior pulmonary vein.



Α

Fig. 12—38-year-old woman with anomalous dual pulmonary venous drainage of left lung. A, Coronal contrast-enhanced subvolume maximum-intensity-projection MR image shows left superior pulmonary venous drainage to both left atrium (*arrow*) and left brachiocephalic vein (arrowheads). LA = left atrium.

B, Catheter venography confirms dual anomalous drainage from left superior pulmonary vein. Balloon catheter is seen in right atrium, superior vena cava, and left brachiocephalic vein (LBCV). Arrowheads indicate anomalous pulmonary vein; lower arrow denotes left superior pulmonary vein.

(Fig. 12 continues on next page)

Fig. 12 (continued)—38-year-old woman with anomalous dual pulmonary venous drainage of left lung.

C, Catheter venography shows no blood flow in anomalous vein that connects left superior pulmonary vein to left brachiocephalic vein after occlusion with vascular plug device (*arrowheads*).



Fig. 13—5-month-old boy with extralobar pulmonary sequestration. Coronal subvolume maximum-intensity-projection image from contrast-enhanced CT examination shows "mass" (*arrowheads*) just inferior to left lung lower lobe above left hemidiaphragm. This structure receives arterial supply from abdominal aorta (*black arrow*). Draining vein can be followed below diaphragm (*white arrow*).





Fig. 14—5-month-old girl with congenital pulmonary venous stenosis. A, Contrast-enhanced volume-rendered MR image, posterior view with pulmonary arteries and portion of descending thoracic aorta removed, shows severe stenosis of right superior and bilateral inferior pulmonary veins (white arrows). Left superior pulmonary vein is patent and drains into left atrium near its appendage (black arrow). L = left. B, Left atrial endoluminal images from contrast-enhanced MRI examination after surgical repair show now nonobstructed right superior pulmonary vein (RSPV). Note residual stenosis of bilateral inferior pulmonary veins. LSPV = left superior pulmonary vein, RIPV = right inferior pulmonary vein, LIPV = left inferior pulmonary vein, LAA = left atrial appendage.



Fig. 15—2-day-old boy with congenital pulmonary vein atresia, bilateral superior venae cavae, and aberrant right subclavian artery arising from left aortic arch. A, Neonatal chest radiograph shows left unilateral pulmonary edema and right upper lobe atelectasis. B, Coronal contrast-enhanced CT subvolume maximum-intensity-projection (MIP) image shows severe central atresia of left pulmonary veins (arrow). (Fig. 15 continues on next page)



Fig. 15 (continued)—2-day-old boy with congenital pulmonary vein atresia, bilateral superior venae cavae, and aberrant right subclavian artery arising from left aortic arch. C, Coronal subvolume MIP image shows presence of both right and left superior venae cavae (RSVC and LSVC, respectively) and no bridging vein. D, Another coronal subvolume MIP image confirms presence of aberrant right subclavian artery (*arrows*).



Fig. 16—24-year-old woman with cor triatriatum.

A and B, Coronal black blood MR images show presence of membrane (*black arrow*) in left atrium that divides chamber into proximal and distal portions. Pulmonary veins drain to proximal portion of chamber. Membrane contains restrictive perforation (*arrowheads*, B). RPA = right pulmonary artery, LPA = left pulmonary artery, RA = right atrium, LV = left ventricle.



Fig. 17—39-year-old woman with recurrent atrial flutter and unsuspected cor triatriatum. Axial oblique reformatted image from contrast-enhanced CT examination performed to evaluate pulmonary venous anatomy before radiofrequency catheter ablation procedure shows thin membrane (*arrows*) in left atrium.

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