

Pulmonary venous abnormalities

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- The **anomalous pulmonary venous connections** are so commonly associated with isomerism of the atrial appendages

Totally anomalous pulmonary venous connection

- Incidence and aetiology
 - Accounting for 1.5% of all patients with a cardiovascular malformation
 - Being seen once in 14,700 live births
 - Two-thirds with supracardiac and cardiac connections were males, while it was the infradiaphragmatic variant that showed an equal mix of genders
 - One report suggesting autosomal dominant inheritance
 - Identified a link between totally anomalous connection and paternal exposure to lead prior to conception

Totally anomalous pulmonary venous connection

- Anatomy

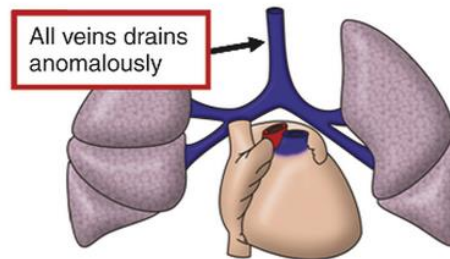
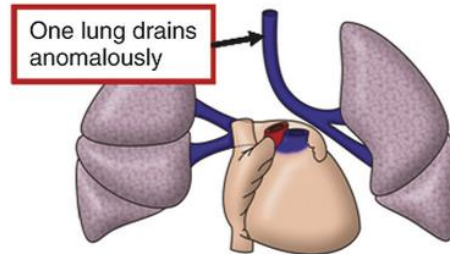
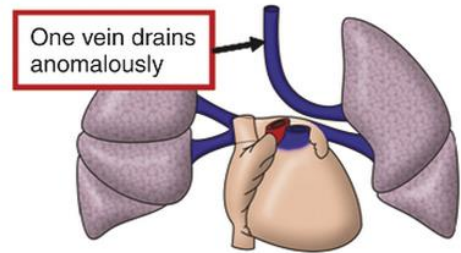


Figure 24-2

The cartoon shows the possibilities for anomalous connection of the pulmonary veins.

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Totally anomalous pulmonary venous connection

- Anatomy
 - Necessary to seek **stenotic areas or regions** along the route of anomalous drainage
 - To establish whether the anomalous pulmonary venous connection is **an isolated malformation**, or **part of a more complex anomaly**
 - Whether there are associated **structural malformation of the pulmonary vasculature**

Totally anomalous pulmonary venous connection

- Anatomy

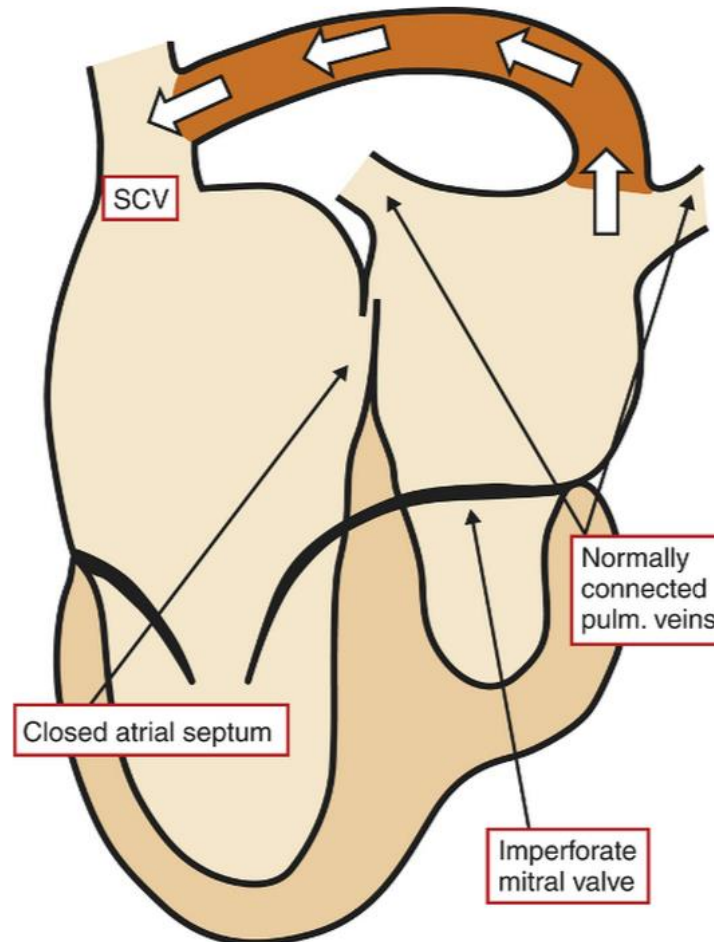


Figure 24-3

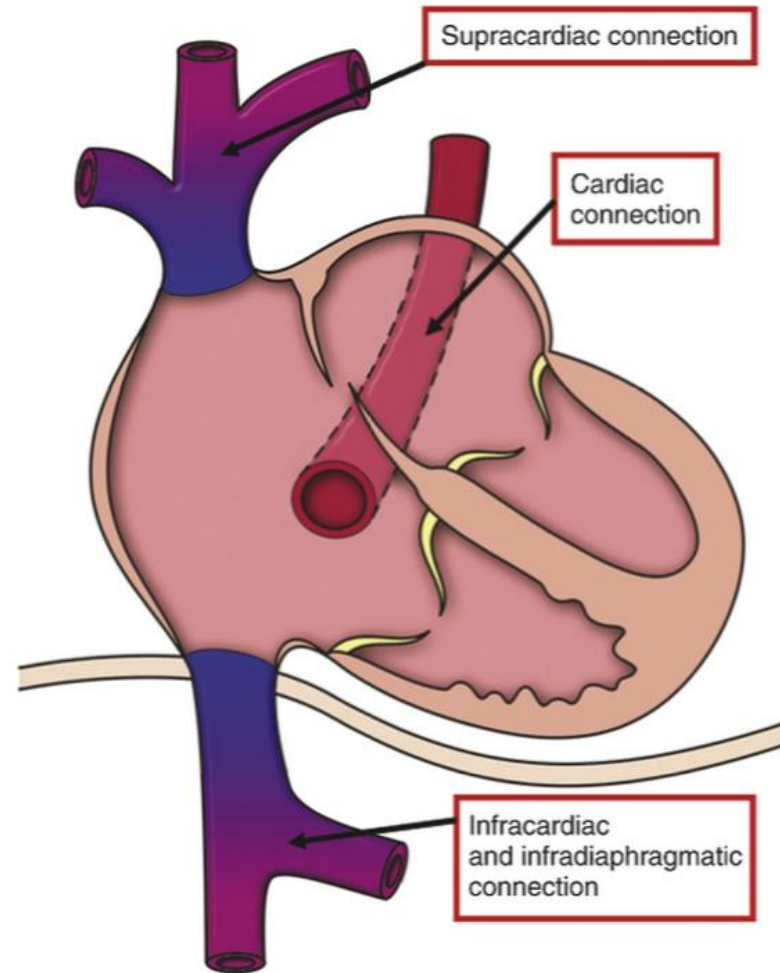
The cartoon shows an example of totally anomalous pulmonary venous drainage in the setting of normally connected pulmonary veins, in this instance because a so-called laevoatrial cardinal vein joins the left atrium to the superior caval vein (SCV) with mitral atresia and an intact atrial septum.

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Totally anomalous pulmonary venous connection

- Anatomy
 - The sites of anomalous connection are divided into **supracardiac, cardiac, and infracardiac** groups
 - The first two, taken together, constitute **supradiaphragmatic** drainage, while infracardiac drainage is at the same time **infradiaphragmatic**



Totally anomalous pulmonary venous connection

- Anatomy
 - The commonest site for anomalous connection is **supracardiac**, accounting for nearly half of cases (**supracardiac, 45%**)
 - cardiac and infracardiac anomalous connections account for approximately one-quarter each of the total group (**cardiac (intracardiac), 25%, infracardiac, 25%, mixed, 5%**)

Totally anomalous pulmonary venous connection

- Anatomy
 - Supracardiac connection can be to the **left brachiocephalic vein**, directly to the **right superior caval vein**, to the **azygos system** of veins, or to the **left superior caval vein**, albeit that when the left vein drains to the coronary sinus this is considered cardiac drainage
 - In the most common pattern, the four pulmonary veins usually join in turn to a venous channel behind the left atrium

Totally anomalous pulmonary venous connection

- Anatomy

- If the vein passes anterior to the left pulmonary artery, then this course is not associated with obstruction
- Should the vein pass between the left pulmonary artery and the left bronchus, these two structures clasp the channel in the so-called bronchopulmonary vice

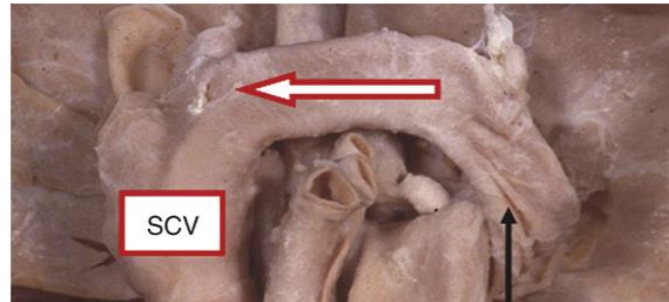


Figure 24-5

The picture shows the typical snowman pattern of supracardiac totally anomalous pulmonary venous connection to the superior caval vein (SCV). The anomalous venous pathway forms the head of the snowman, whilst the ventricular mass accounts for the body. The arrow shows the direction of flow of the anomalous pulmonary venous return through the brachiocephalic vein.

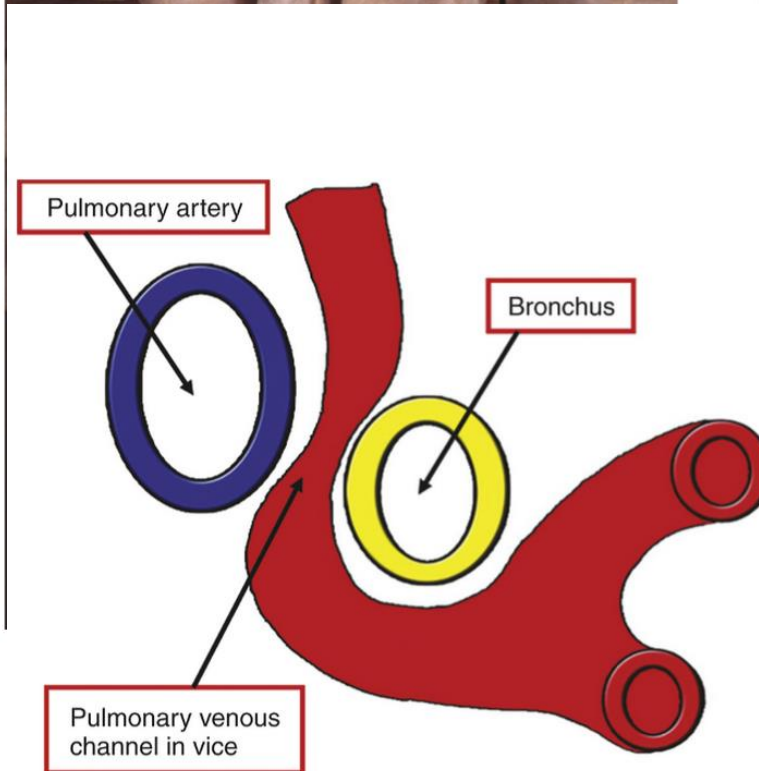


Figure 24-6

The cartoon shows the nature of the bronchopulmonary vice, which produces obstruction with supracardiac connections.

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Totally anomalous pulmonary venous connection

- Anatomy
 - **Obstruction** with this snowman pattern of anomalous connection can also occur, albeit rarely, **at the opening of the brachiocephalic vein** into the superior caval vein
 - Supracardiac connection can also be found when the vertical vein **joins directly with the right superior caval vein**
 - **Obstruction** may then occur **between the right pulmonary artery and the carina**

Totally anomalous pulmonary venous connection

- Anatomy

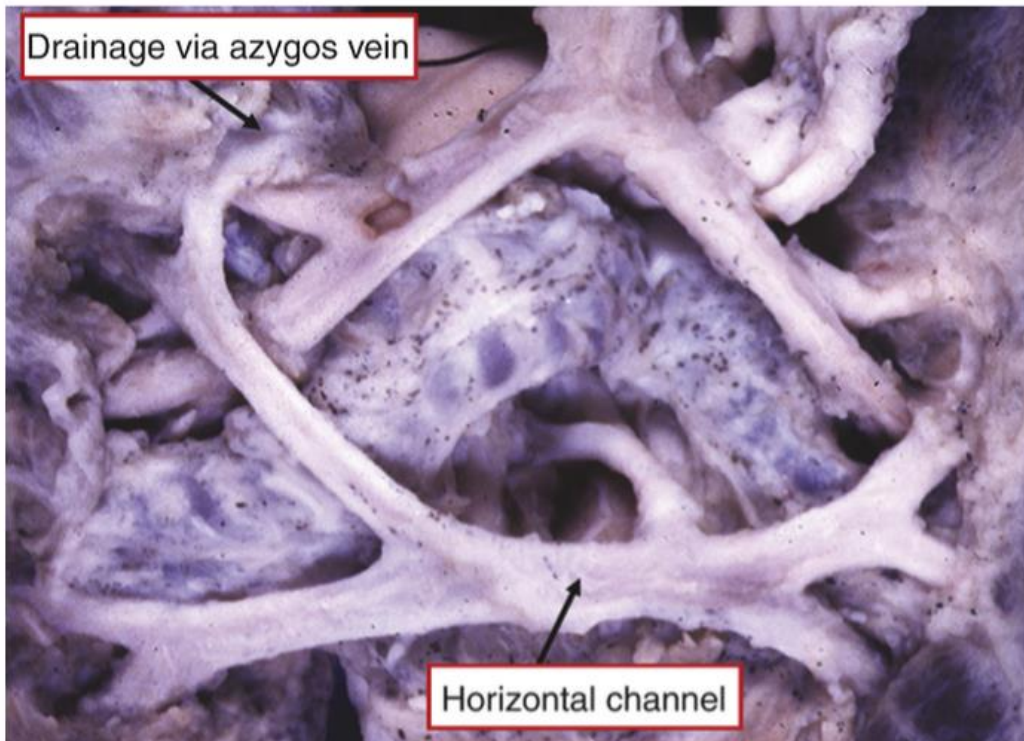


Figure 24-7

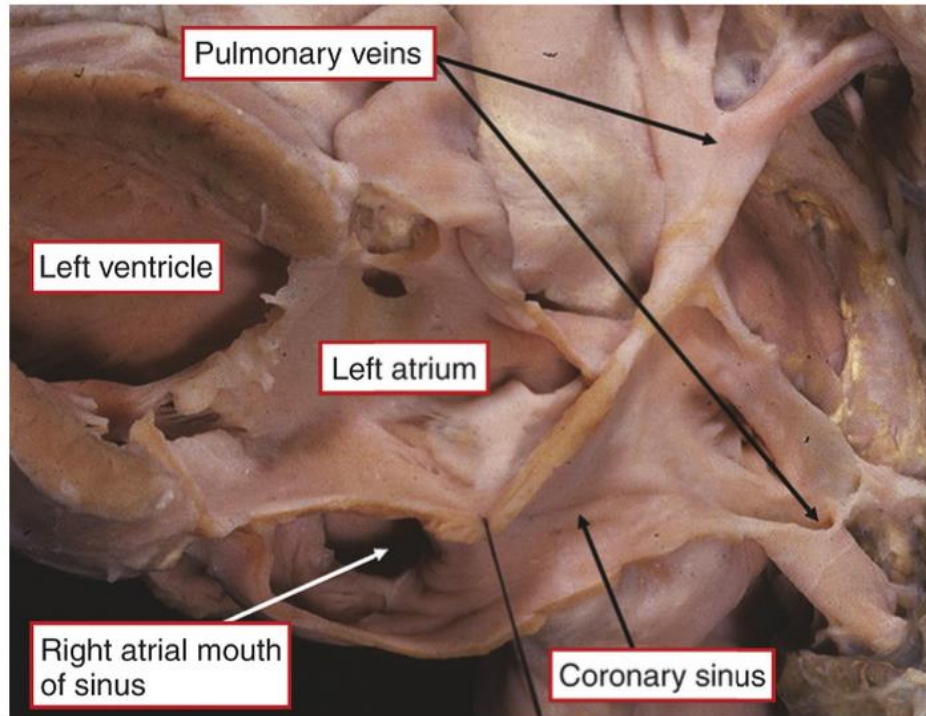
In this specimen, from a patient with isomeric right atrial appendages, the horizontal vein passes from left to right and ascends in the right paravertebral gutter, draining in supracardiac fashion through the azygos vein.

0 Notes

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Totally anomalous pulmonary venous connection

- Anatomy
 - When there is usual atrial arrangement, then almost always the **cardiac form of anomalous connection** is found when the pulmonary veins join the right atrium **via the coronary sinus**
 - **Obstruction is rare** when the pulmonary veins drain **through the coronary sinus**, but can be produced by **persistence of the Thebesian valve**, or **within the sinus** when the individual veins connect in unusual fashion



Totally anomalous pulmonary venous connection

- Anatomy
 - Direct connection of the pulmonary veins to the morphologically right atrium is exceedingly rare other than in the setting of isomerism of the right atrial appendages

Totally anomalous pulmonary venous connection

- Anatomy
 - The final site of anomalous connection is both **infracardiac** and **infradiaphragmatic**
 - The pulmonary veins join together, entering a **descending vertical vein** that passes into the abdomen through the **oesophageal orifice of the diaphragm**
 - It then usually **drains to the portal vein**, or to one of its tributaries, or **hepatic vein**
 - Drainage to the inferior caval vein is very rare

Totally anomalous pulmonary venous connection

- Anatomy
 - When the inferior connection is to the portal venous system, obstruction is almost always present subsequent to closure of the venous duct
 - Additional discrete stenosis can be found as the vertical vein passes through the diaphragm

Totally anomalous pulmonary venous connection

- Anatomy
 - As discussed, it is also possible for different veins to terminate in different anomalous sites ([mixed anomalous connection](#))

Totally anomalous pulmonary venous connection

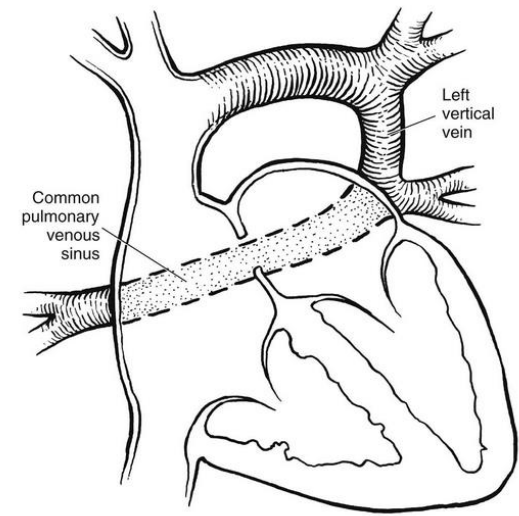
- Anatomy
 - A large proportion of the patients have significant associated malformations
 - Even when the anomalous connection is isolated, there is almost always an interatrial communication present, so that venous blood is able to reach the left side of the heart

Totally anomalous pulmonary venous connection

- Anatomy
 - At first sight, the left atrium and left ventricle seem small to the morphologist because of the disparate hyperplasia of the right atrium and right ventricle
 - Measurements, however, show that the left-sided structures are usually of adequate dimensions

Totally anomalous pulmonary venous connect

- Anatomy
- **Supracardiac type**



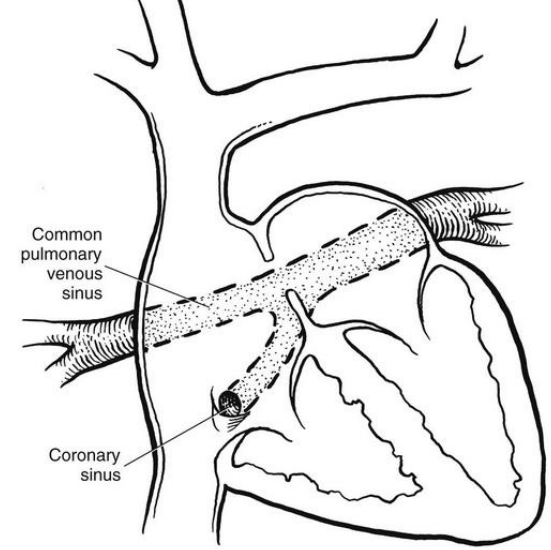
A

Supracardiac type (45%)



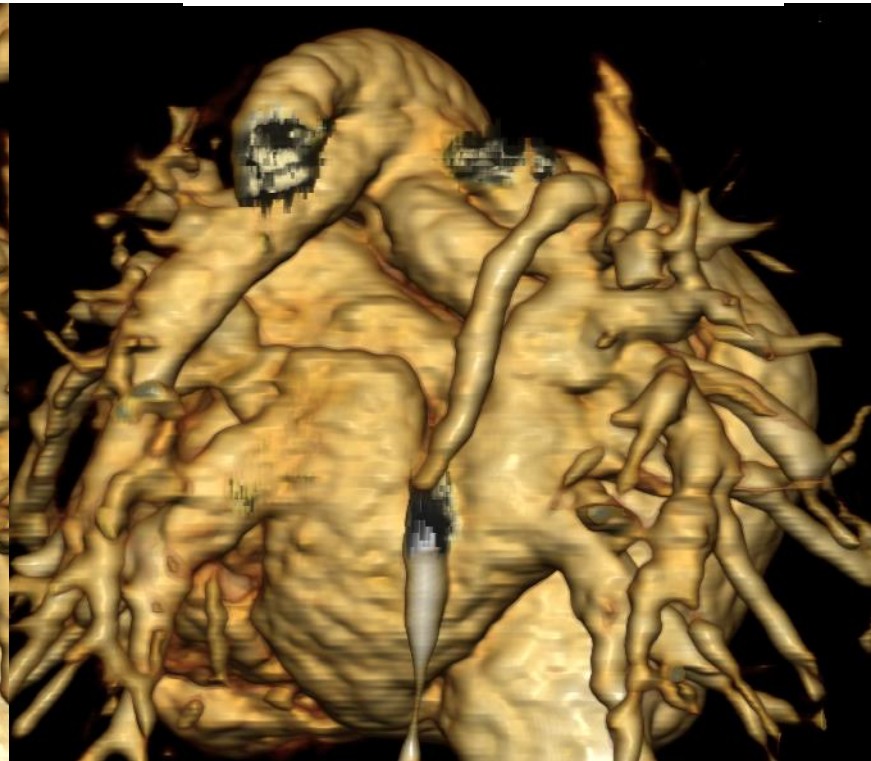
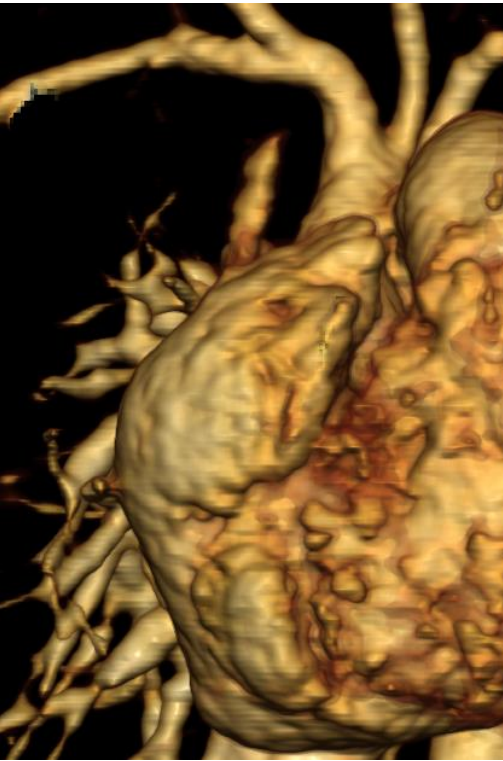
Totally anomalous pulmonary venous connection

- Anatomy
- **Cardiac type**



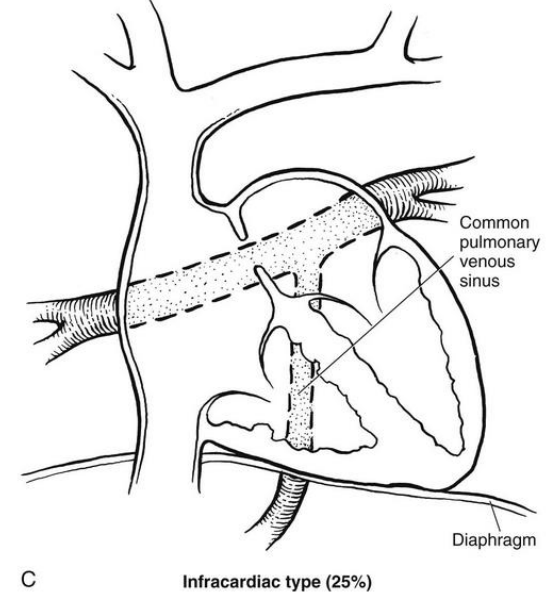
B

Cardiac type (25%)

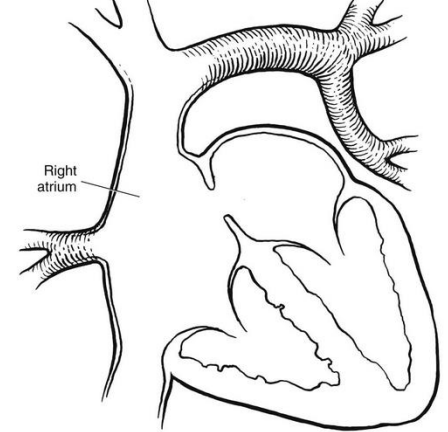


Totally anomalous pu venous connect

- Anatomy
- **Infracardiac type**



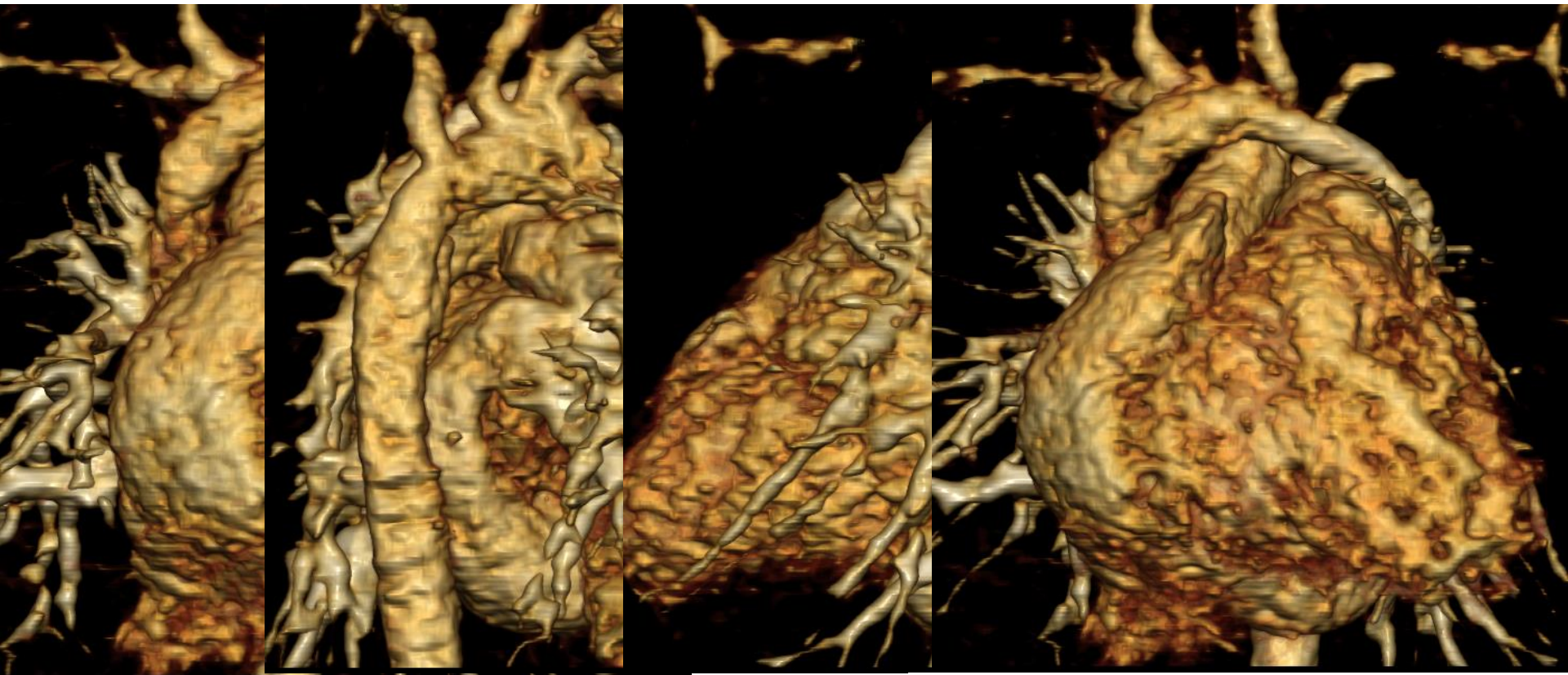
Totally anomalous pulm venous connection



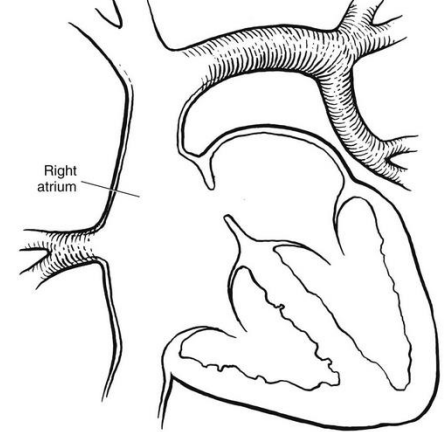
- Anatomy
- **Mixed type (cardiac + supracardiac, LUPV)**

D

Mixed type (5%-10%)



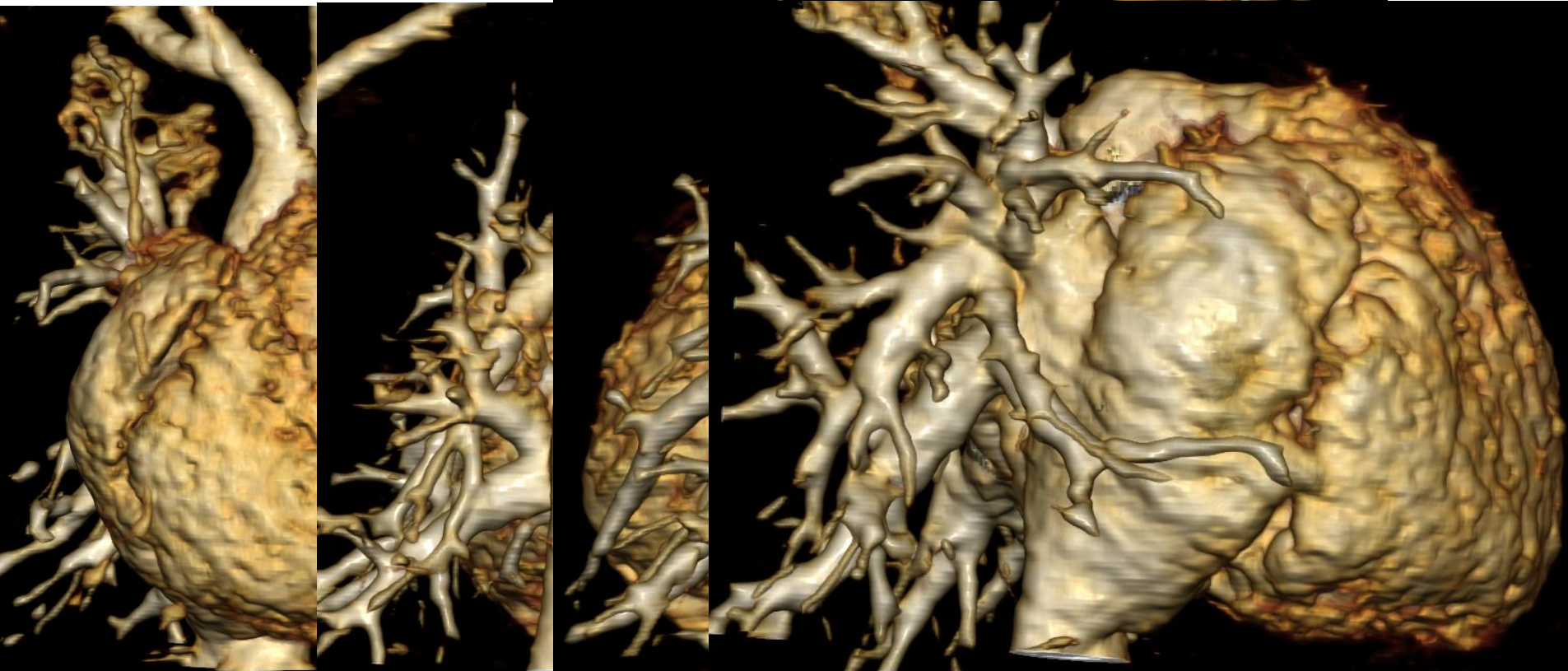
Totally anomalous pulm venous connector



- Anatomy
- **Mixed type (cardiac + supracardiac, RUPV)**

D

Mixed type (5%-10%)



Totally anomalous pulmonary venous connection

- Morphogenesis
 - Totally anomalous pulmonary venous connection is the consequence of failure of canalisation of the pulmonary venous channel in the mediastinum
 - Initially, since the lung buds themselves are derived from the foregut, the intrapulmonary veins also have connections to the systemic venous system
 - Should the pulmonary venous channel fail to develop, these anastomoses between pulmonary and systemic venous system persist and enlarge

Totally anomalous pulmonary venous connection

- Pathophysiology
- Shunt
 - There is an obligatory **left-to-right shunt**, since pulmonary venous return is to the systemic veins or right atrium
 - A **systemic output** can only be maintained if there is a **right-to-left shunt**, which is almost always at **atrial level**
 - Exceptional cases have been described in which the atrial septum was intact
 - In these patients, the right-to-left shunt occurred either at **ventricular, or ductal level**

Totally anomalous pulmonary venous connection

- Pathophysiology
- Obstruction to pulmonary venous return
 - Obstruction to pulmonary venous return can occur at any of the anatomical sites documented above
 - When there is definable obstruction, the right ventricular pressure is usually suprasystemic
 - Almost all patients with pulmonary vascular obstruction also had pulmonary venous obstruction

Totally anomalous pulmonary venous connection

- Pathophysiology
- Consequences of pulmonary venous obstruction
 - When **pulmonary venous return is unobstructed**, right ventricular diastolic pressure is low and right ventricular compliance relatively high
 - Since mixing of pulmonary and systemic venous blood is complete, apart from the minor degrees of streaming, right atrial and, therefore, systemic arterial blood is well oxygenated, with saturations of oxygen found in excess of 90%

Totally anomalous pulmonary venous connection

- Pathophysiology
- Consequences of pulmonary venous obstruction
 - In the presence of pulmonary venous obstruction, in contrast, pulmonary venous pressure is raised
 - The right ventricle becomes pressure rather than volume overloaded
 - Systemic arterial oxygen saturation may then fall to values of 20% to 30%
 - Results in tissue hypoxemia and metabolic acidosis

Totally anomalous pulmonary venous connection

- Presentation and symptoms
 - The main determinant of the clinical picture was the presence of pulmonary venous obstruction
 - Patients will be divided into those with and without pulmonary venous obstruction

Totally anomalous pulmonary venous connection

- Presentation and symptoms
 - Patients with severe pulmonary venous obstruction present in the first week or two of life with obvious cyanosis and difficulties with feeding and respiration
 - Patients without severe pulmonary venous obstruction tend to present in heart failure at 2 to 3 months of age
 - They have a history of difficulties with feeding and, sometimes, chest infections
 - Cyanosis is generally not a symptom

Totally anomalous pulmonary venous connection

- Presentation and symptoms
 - Those with severe pulmonary venous obstruction are sick neonates with obvious or severe cyanosis
 - Skin mottling is frequent, reflecting poor peripheral perfusion and metabolic acidosis
 - Tachypnea is usually marked, though respiration is quiet
 - Hepatomegaly is occasionally considerable, particularly when drainage is to the portal vein
 - The peripheral pulses are often somewhat weak

Totally anomalous pulmonary venous connection

- Investigations
- Chest radiography
 - Newborn with severe pulmonary venous obstruction have an extremely characteristic chest radiograph, with a **small or normally sized heart framed by ground-glass lung fields**



Totally anomalous pulmonary venous connection

- Investigations
- Chest radiography
 - Patients without severe pulmonary venous obstruction have enlarged hearts because of the right ventricular volume overload, together with engorged lung fields
 - The pulmonary trunk becomes prominent in older patients, as does the left vertical vein when this is the site of the anomalous venous connection -> the **snowman appearance**

Totally anomalous pulmonary venous connection

- Investigations
- Electrocardiography
 - The electrocardiogram shows **right-axis deviation** with a clock wise frontal plane loop and **right ventricular hypertrophy**
 - **Disturbances of conduction are rare**
 - Patients with pulmonary venous obstruction, who present younger, are much less likely to have right atrial hypertrophy than those without pulmonary venous obstruction

Totally anomalous pulmonary venous connection

- Investigations
- **Echocardiography**
 - Echocardiography is the **definitive non-invasive method of diagnosis**
 - Exclusive right-to-left shunting at the atrial level through an atrial septal defect or patent oval foramen -> it should be assumed that any patient with this finding has totally anomalous pulmonary venous connection until proven otherwise

Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - Other clues -> small left atrium, dilated superior caval vein, non-pulsatile caudally-directed flow seen below the level of the heart in subcostal imaging, or non-pulsatile cranially-directed flow seen above the level of the heart

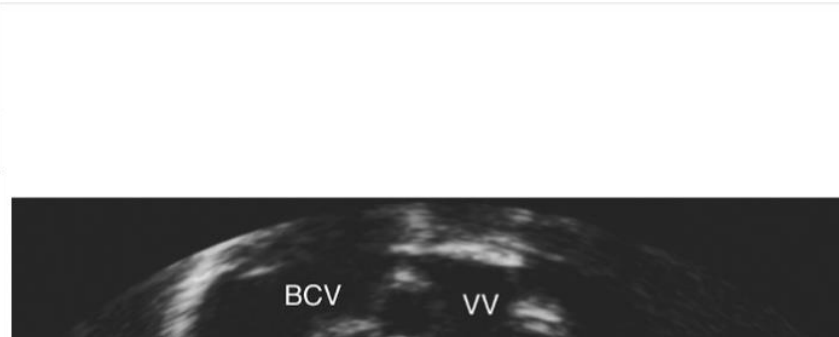


Figure 24-13

In this image, taken from the subcostal long-axis view angled posteriorly, note the severely dilated superior caval vein (SCV) emptying into the right atrium (RA), suggesting the presence of anomalous pulmonary venous connection. Further investigation revealed a confluence (C) behind the left atrium, leading to a vertical vein (VV), which connected to the left brachiocephalic vein (BCV). L, left; S, superior.

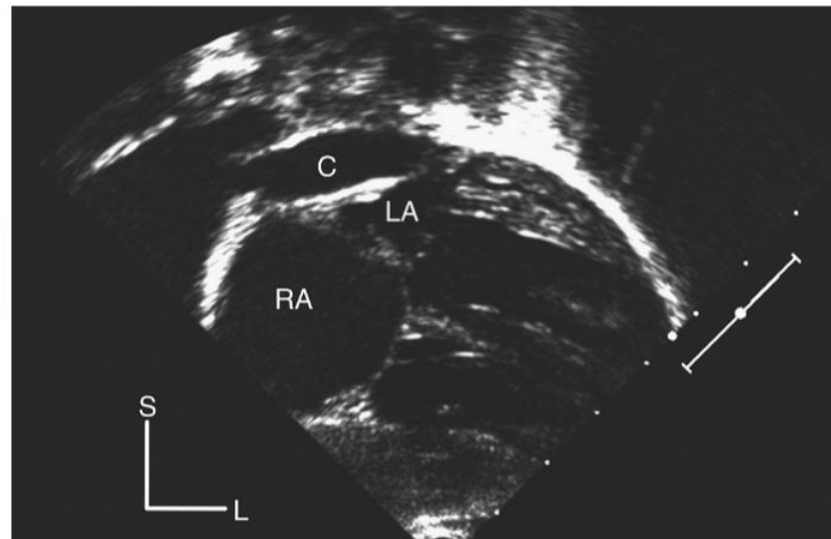


Figure 24-14

In most cases of totally anomalous pulmonary venous connection, echocardiography reveals an echo-free non-pulsatile region beyond and clearly separate from the left atrium. In this image, taken from the subcostal long-axis view, the pulmonary venous confluence (C) can be seen posterior and superior to the left atrium (LA). Colour Doppler can be applied to this view, clearly distinguishing this vessel from a pulmonary artery. L, left; RA, right atrium; S, superior.

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Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - When the pulmonary veins are connected anomalously to the coronary sinus, the collecting venous channel is the coronary sinus itself
 - Great care must be taken to distinguish between enlargement of the coronary sinus owing to persistence of the left superior caval vein and the pattern in which the pulmonary veins drain into it

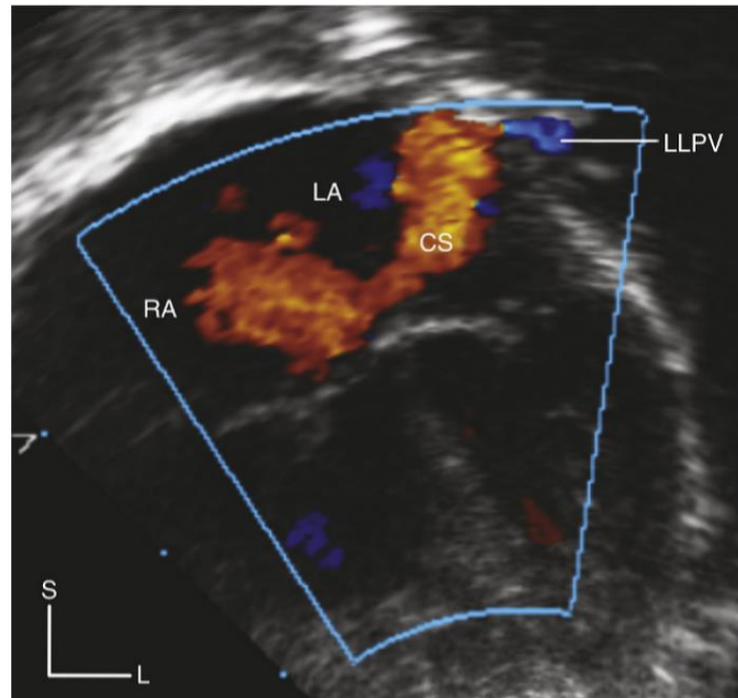


Figure 24-20

This apical four-chamber view reveals drainage of the left lower pulmonary vein (LLPV) into the coronary sinus (CS). L, left; LA, left atrium; RA, right atrium; S, superior.

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Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - A descending vein, as found in anomalous infradiaphragmatic connection, can sometimes be seen descending from the confluence from the suprasternal approach
 - Pulsed Doppler ultrasound or colour Doppler can be used to show that the descending pulmonary vein contains non-pulsatile blood moving inferiorly, while the inferior caval vein contains blood moving superiorly

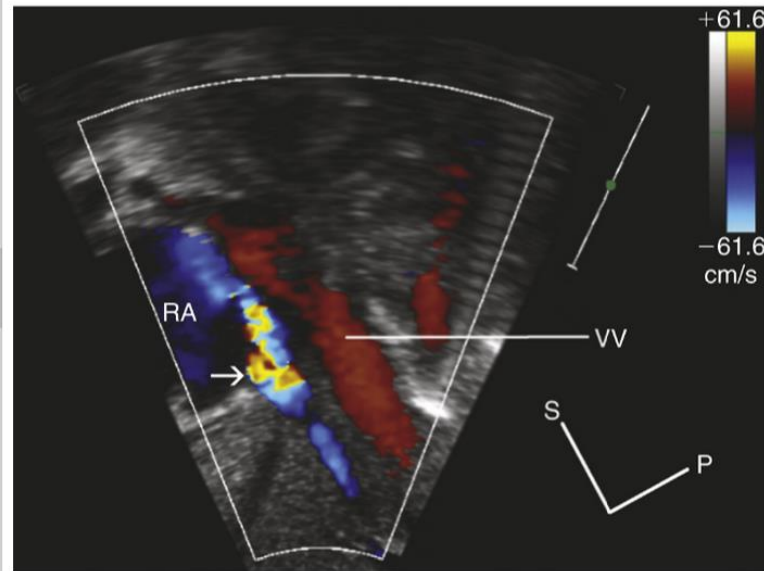


Figure 24-17

In this subcostal short-axis image, a large, posteriorly-directed vertical vein (VV) is seen. This vessel connects with the portal system inferior to the screen. Turbulence is seen in the region of the venous duct (arrow), representing obstruction to pulmonary venous return (see Fig. 24-22). P, posterior; RA, right atrium; S, superior.

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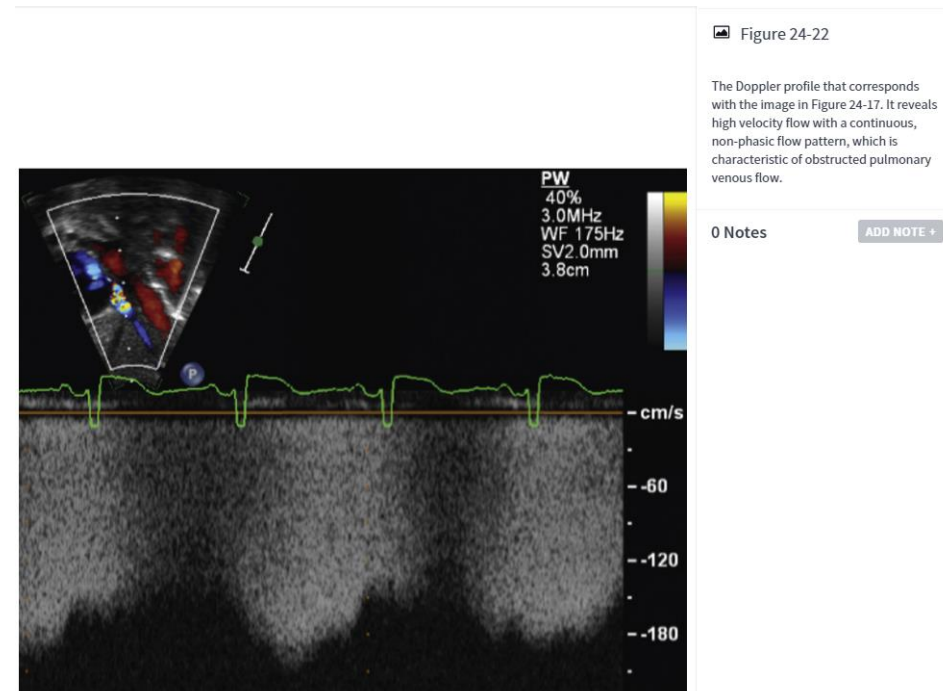
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Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - Anomalous connection to the right atrium can be diagnosed if there is no ascending or descending vein, the coronary sinus is of normal size and the pulmonary veins can be followed to their site of entry to the right atrium

Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - The echocardiographer should consider measuring the diameter of all four pulmonary veins between hilum and confluence, as the sum of these diameters is a strong and independent predictor of surgical survival
 - Sites of obstruction along the pulmonary venous pathway can be demonstrated as points of turbulence, or even absent flow, both pre- and post-operatively
 - In areas where colour Doppler suggests obstruction, pulsed wave Doppler offers an objective measure
 - The presence of a focal increase in flow velocity with a continuous, non-phasic flow pattern distally is a characteristic finding



Totally anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - If the clinical and cross sectional echocardiographic findings do not fit the clinical situation, additional imaging should be performed without hesitation
 - Fetal echocardiography

Totally anomalous pulmonary venous connection

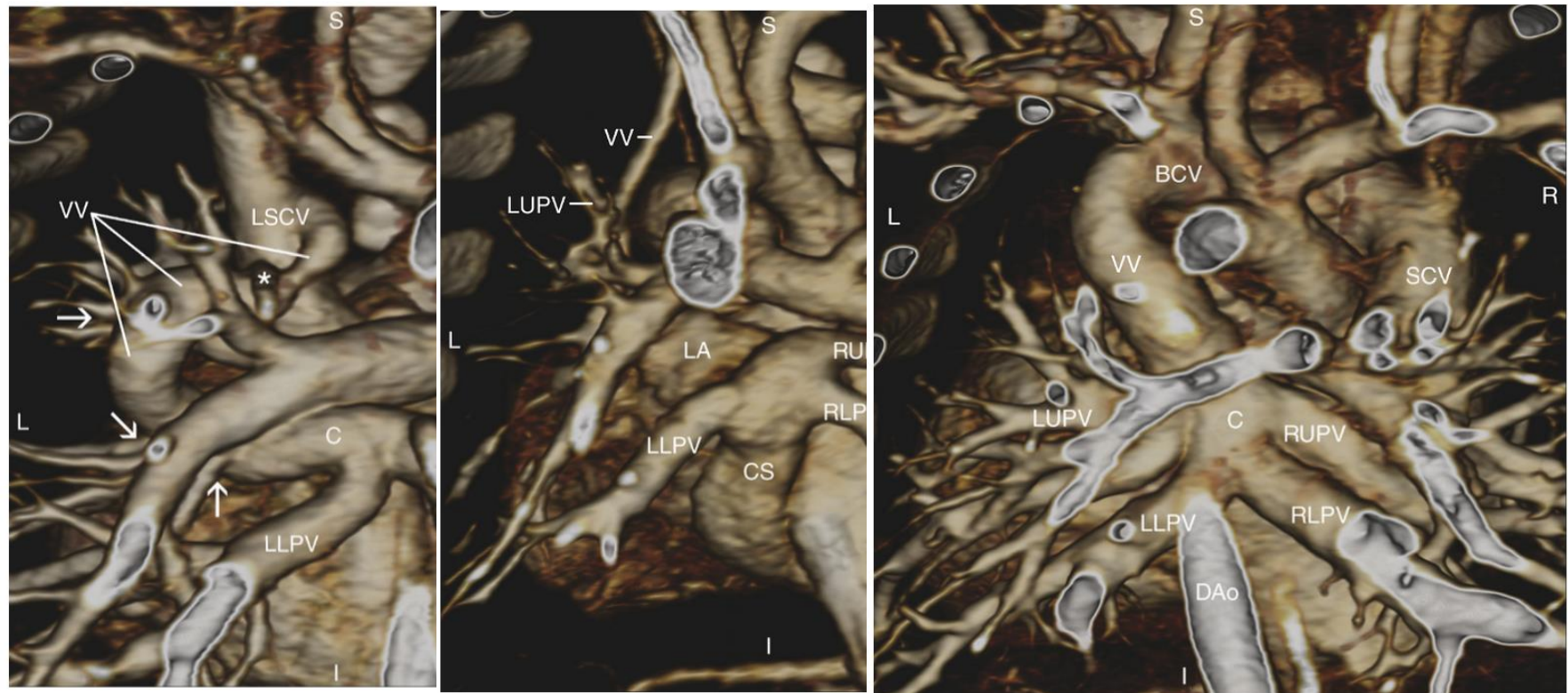
- Investigations
- Cardiac catheterisation
 - The pulmonary venous anatomy can almost always be delineated non-invasively, and the clinical scenario of pulmonary venous obstruction can almost always be determined without invasive testing
 - Pulmonary venous connections with a particularly tortuous course, as is often seen with infracardiac connection or drainage via the azygos system, can be difficult to follow by echocardiography

Totally anomalous pulmonary venous connection

- Investigations
- **Computed tomographic angiography**
 - Technological advances in medical imaging have increased the utilisation of computed tomographic angiography in the evaluation of patients with anomalous pulmonary venous connection

Totally anomalous pulmonary venous connection

- Investigations
- Computed tomographic angiography



Totally anomalous pulmonary venous connection

- Differential diagnosis
 - Unobstructed connection has to be distinguished from other conditions producing heart failure, mild cyanosis, and cardiomegaly with pulmonary plethora and right ventricular hypertrophy

Totally anomalous pulmonary venous connection

- Course and prognosis
 - With medical treatment alone, three-quarters of all children with totally anomalous pulmonary connection uncomplicated by isomerism were dead or had undergone **surgery** by their first birthday
 - **The only place for medical treatment, therefore, is in resuscitation of the critically ill neonate**

Totally anomalous pulmonary venous connection

- Course and prognosis
 - Atrial septostomy – Sano and colleagues found that in no case did septostomy result in sufficient clinical improvement in critically ill patients to permit deferral of the operation
 - In patients with severely obstructed pulmonary venous return, **stent placement** in the area of obstruction can be considered as a temporising measure **if surgery cannot be performed in a timely manner**

Totally anomalous pulmonary venous connection

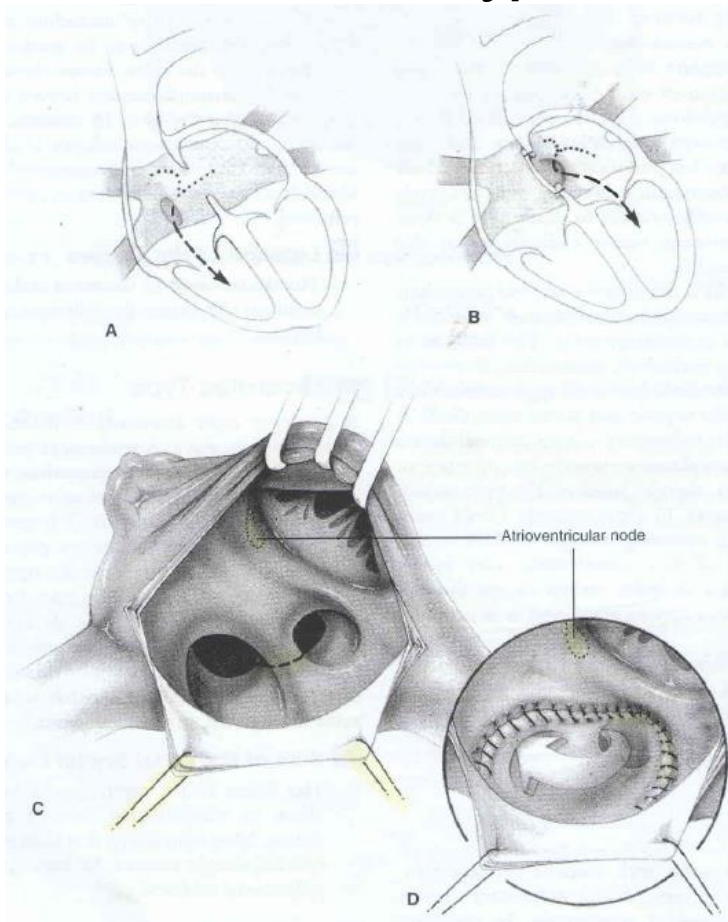
- Management
- Medical treatment
 - In the current era, medical management consists solely of **supportive measures** in preparation for surgical management

Totally anomalous pulmonary venous connection

- Management
- Surgery
 - Operations at this time included those in which **an anastomosis between the pulmonary venous confluence and the left atrium** was created but (the interatrial communication was not closed; the common pulmonary vein, if obstructed, was not ligated)
 - Because the left heart may have difficulty tolerating an acute increase in pulmonary venous return after surgery, resulting in low cardiac output, many surgeons leave the vertical vein intact after surgery

Totally anomalous pulmonary venous connection

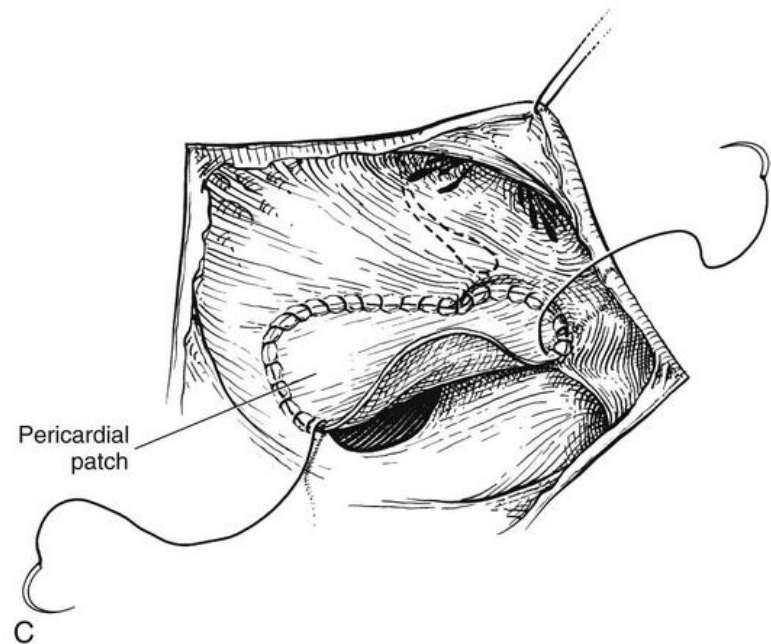
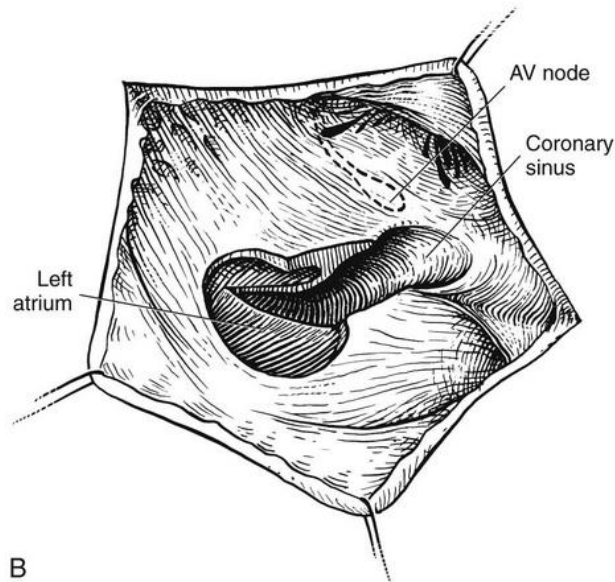
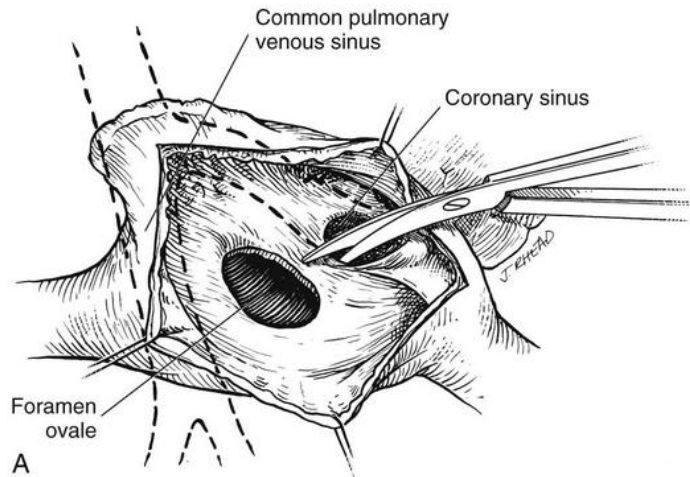
- Management
- Surgery
 - **Intracardiac type**



- . Whenever the common pulmonary vein returns to the coronary sinus, its orifice is extended superiorly to reach the atrial septal defect
- . This incision must be well away from the anterior margin of the coronary sinus to prevent damage to the atrioventricular node and the conduction system

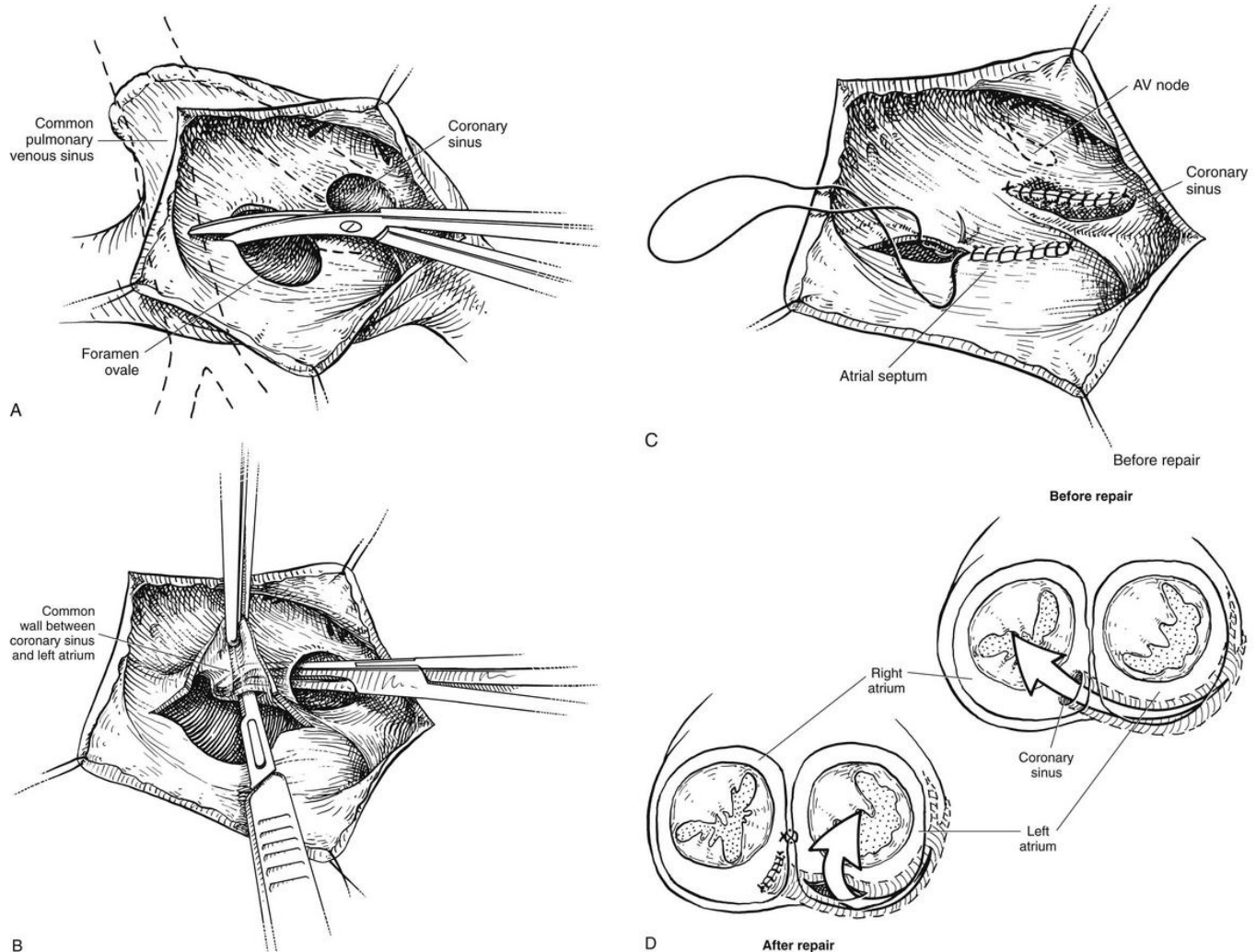
Totally anomalous pulmonary venous connection

- Management
- Surgery
 - **Intracardiac type**



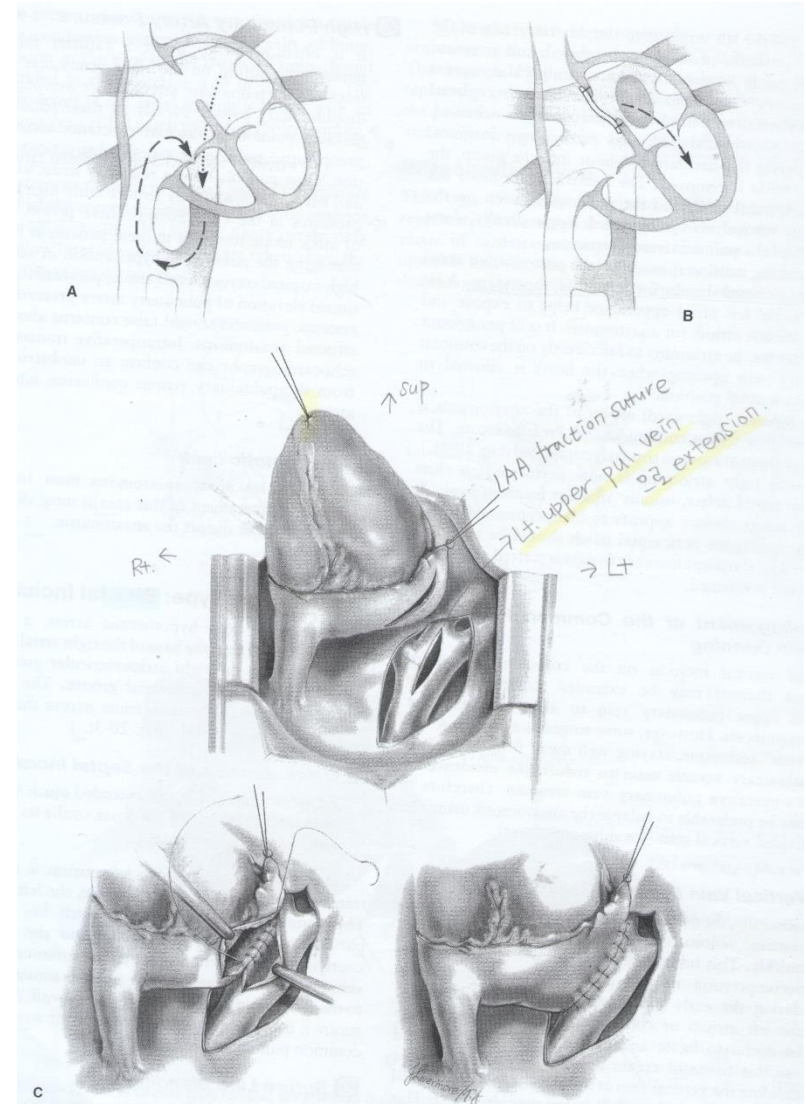
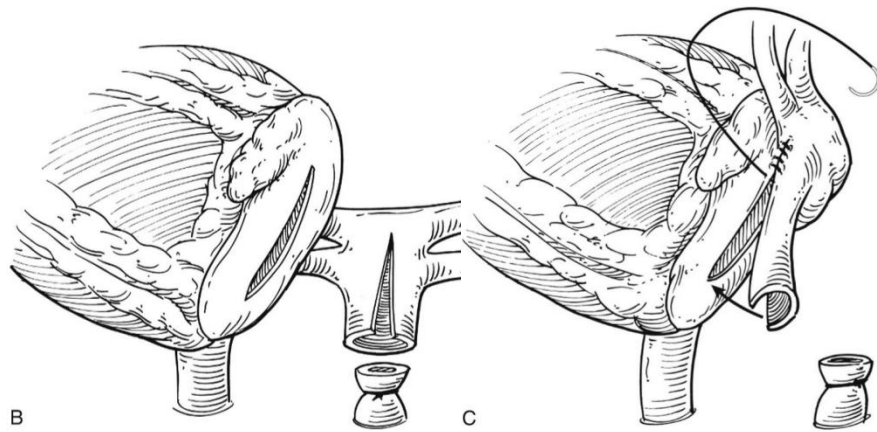
Totally anomalous pulmonary venous connection

- Management
- Surgery
- Intracardiac type, **Van Praagh method**



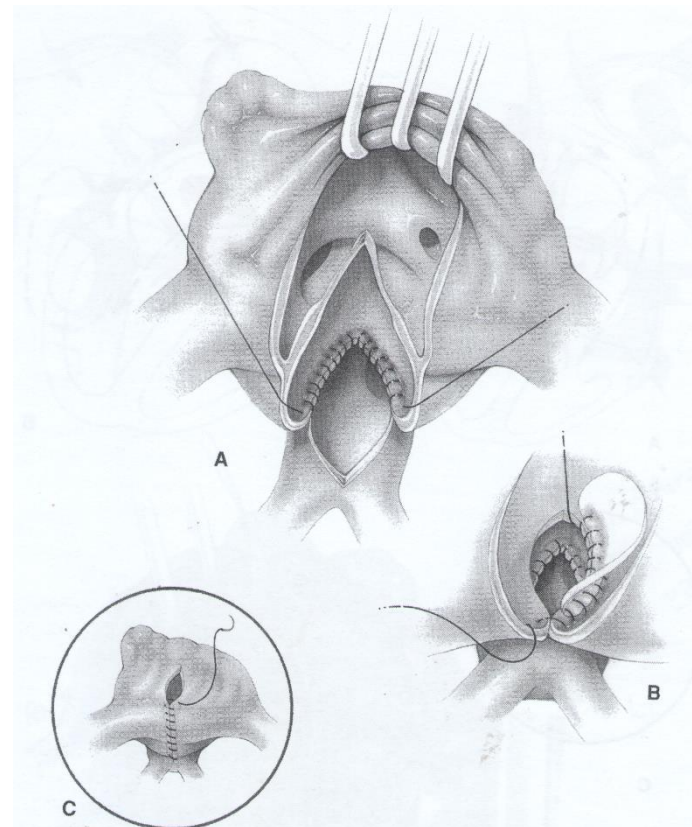
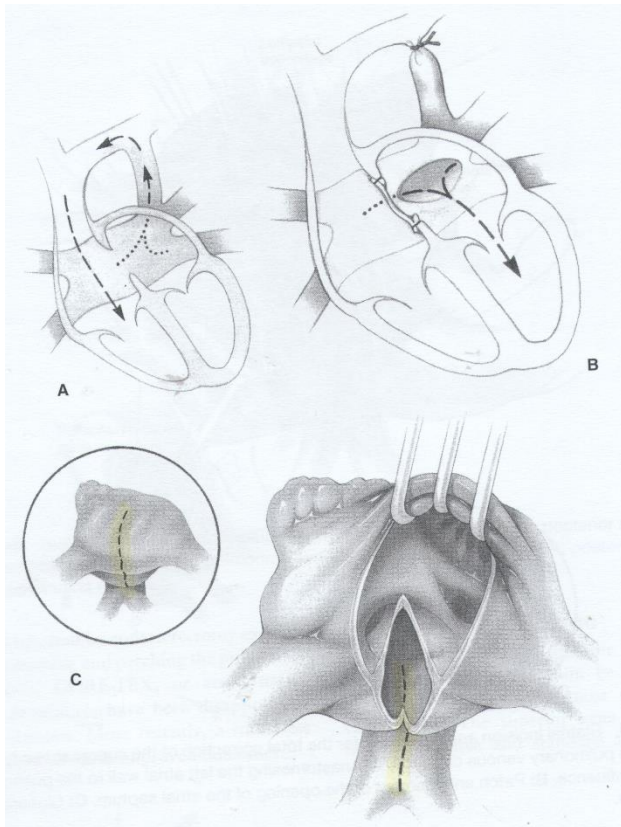
Totally anomalous pulmonary venous connection

- Management
- Surgery
 - Infracardiac type



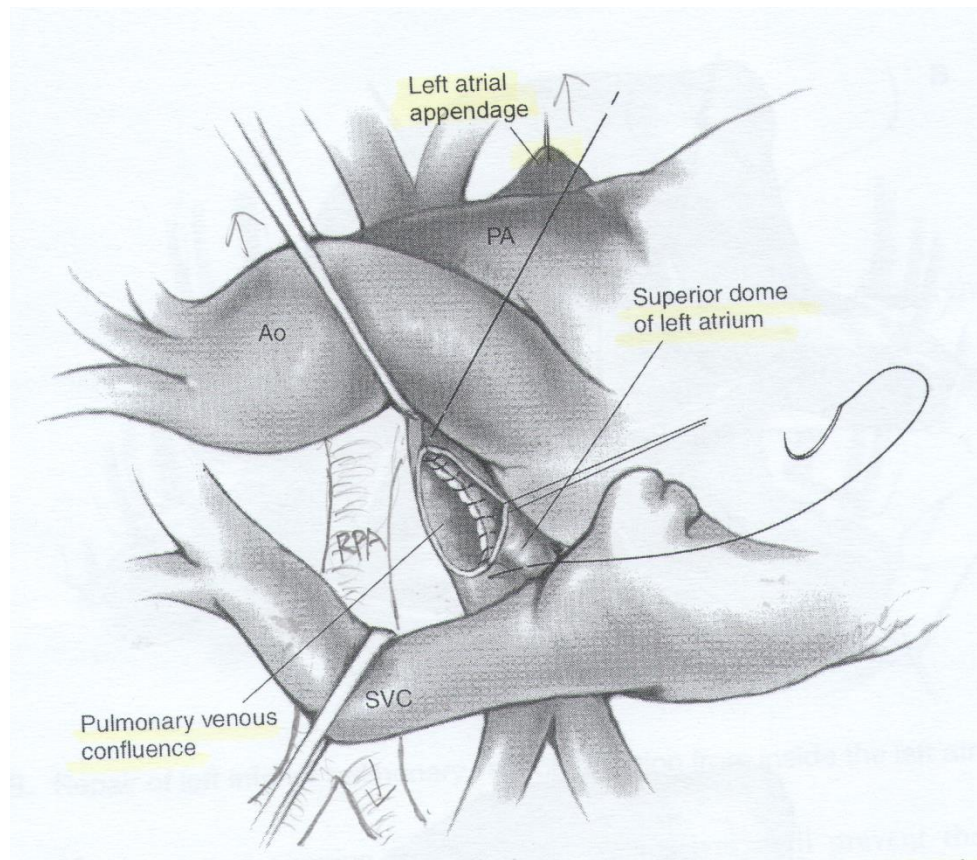
Totally anomalous pulmonary venous connection

- Management
- Surgery
 - **Supracardiac type (biatrial incision)**



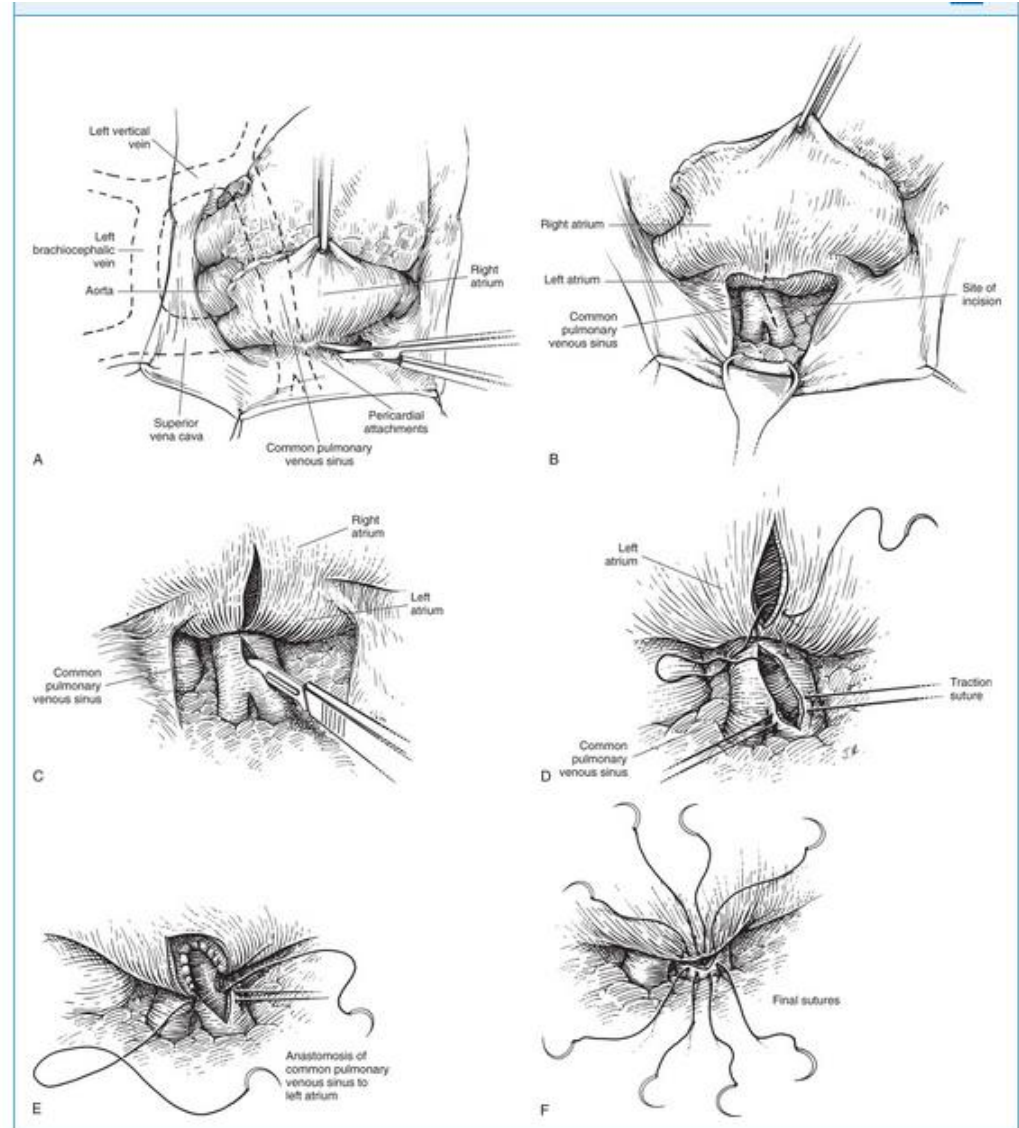
Totally anomalous pulmonary venous connection

- Management
- Surgery
 - **Supracardiac type (superior approach)**



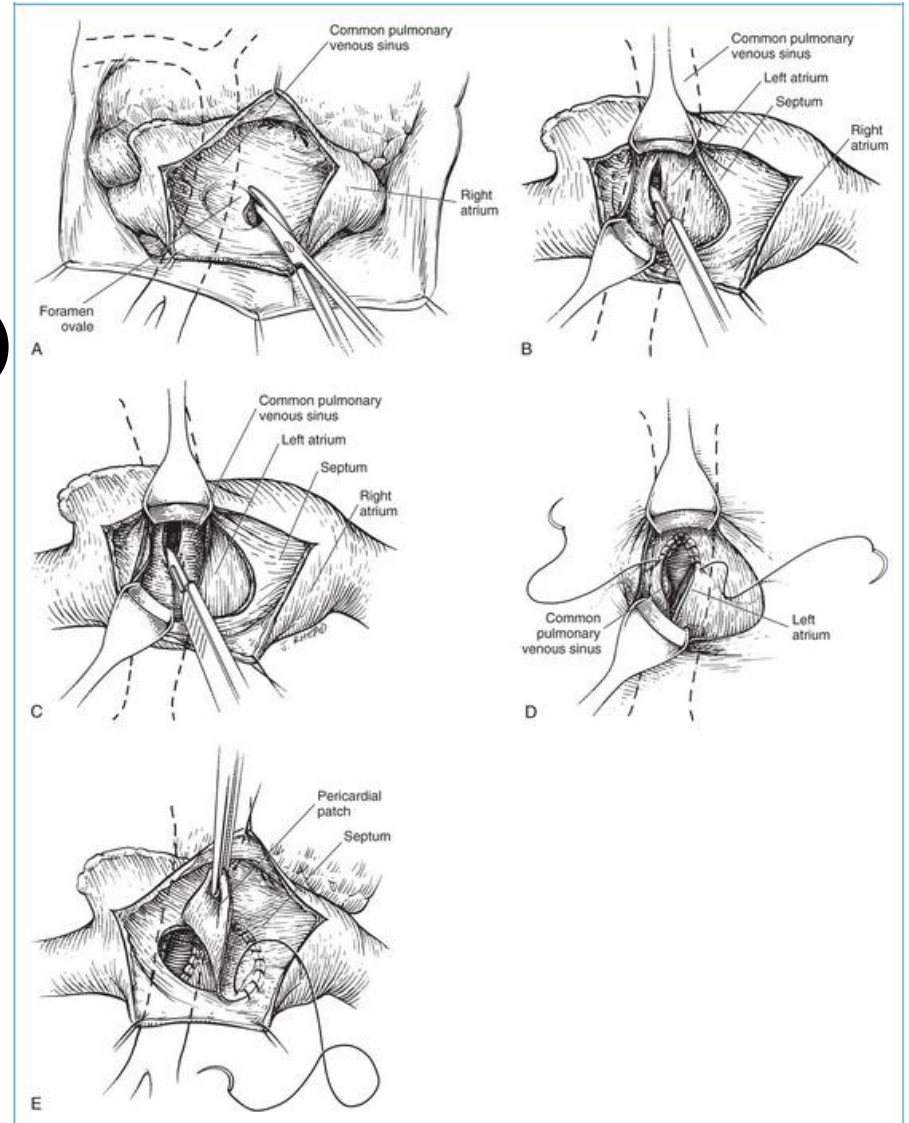
Totally anomalous pulmonary venous connection

- Management
- Surgery
 - **Supracardiac type (Rt. Lateral approach)**



Totally anomalous pulmonary venous connection

- Management
- Surgery
- **Supracardiac type (right atrial approach)**



Totally anomalous pulmonary venous connection

- Management
- Surgery

TABLE 24-1 Results of Repair of Totally Anomalous Pulmonary Venous Connection in Infancy

Report	Date of Operation	Hospital Mortality (Number [%])
Katz et al ¹⁰⁰	1974–1977	4/19 (21%)
Whight et al ¹²⁶	1969–1976	3/23 (13%)
Hammon et al ³¹	1969–1979	5/25 (20%)
Bove et al ¹³⁸	1971–1979	26/73 (36%)
Yee et al ²²¹	1975–1986	8/75 (11%)
Lamb et al ²⁹	1968–1985	14/80 (18%)
Lincoln et al ²²²	1973–1986	12/83 (14%)
Sano et al ⁸¹	1979–1987	1/44 (2%)
Raisher et al ¹⁰	1983–1990	1/20 (5%)
Korbmacher et al ²²³	1958–1992	18/52 (35%)
Lupinetti et al ⁹⁵	1985–1993	2/41 (5%)
Sinzobahamvya et al ⁹¹	1977–1994	6/71 (8%)
Bando et al ⁸⁷	1966–1995	10/105 (10%)
Calderone et al ⁸⁹	1982–1996	19/126 (15%)
Bogers et al ⁸⁸	1973–1998	6/44 (14%)
Michielon et al ⁹⁴	1983–2001	11/89 (12%)
Hyde et al ⁹²	1988–1998	6/85 (7%)
Hancock Friesen et al ⁹⁰	1989–2000	10/84 (12%)

Totally anomalous pulmonary venous connection

- Management
- Surgery
 - The post-operative course is frequently marked by pulmonary hypertensive crises
 - Post-operative pulmonary hypertension in this population was traditionally managed with 100% oxygen and epoprostenol (prostacyclin), but nitric oxide has shown promising results over the last 10 years

Totally anomalous pulmonary venous connection

- Results of surgery
 - The late results of repair are, in general, excellent
 - Nonetheless, late pulmonary venous obstruction is not uncommon
 - It occurs in about 10% of all large surgical series
 - Some authors have found it to be more common in patients with infracardiac or mixed drainage

Totally anomalous pulmonary venous connection

- Results of surgery
 - In earlier studies, reoperation carried a high mortality and a strong chance of recurrence, whatever the nature of the obstruction
 - In 1996, Lacour-Gayet and colleagues, introduced a **sutureless technique** for reoperation using in situ pericardium with promising results

Totally anomalous pulmonary venous connection

- Results of surgery
 - The diagnosis of restenosis, this will demonstrate a small anastomosis and/or a continuous, non-phasic relatively high-velocity Doppler flow signal and an enlarged right ventricle
 - Computed tomographic angiography as the imaging modality used to clarify questionable echocardiography findings

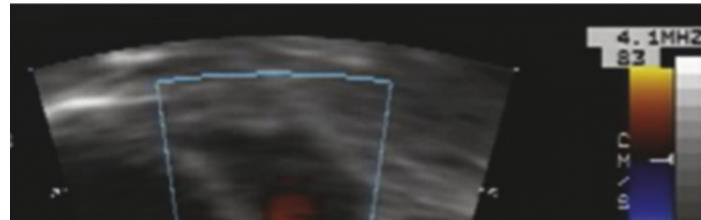


Figure 24-28

In this zoomed up apical image, angled posteriorly, turbulent flow (star) can be seen at the surgical anastomosis between the confluence (C) and the left atrium (LA). Note that the Nyquist limit is 83 cm/sec. Pulmonary venous flow should not alias in this setting, unless there is obstruction. L, left; P, posterior.

0 Notes

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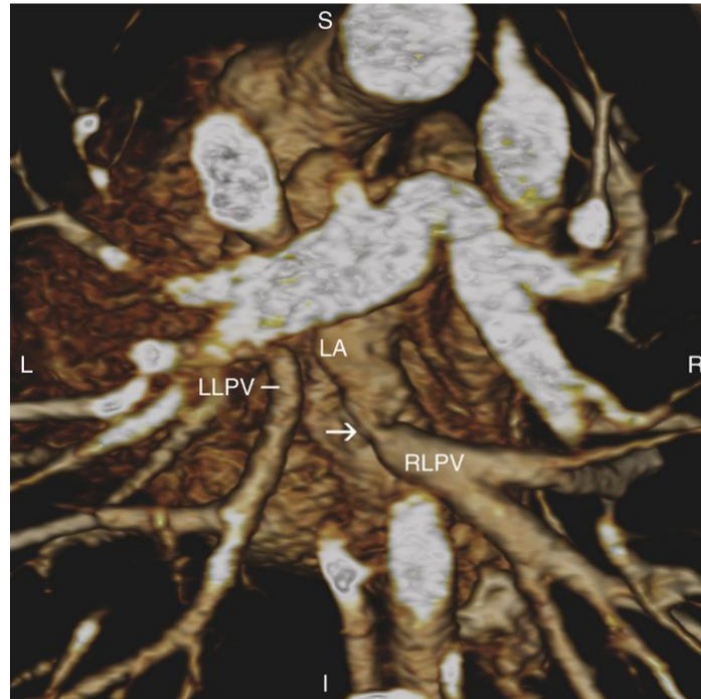


Figure 24-29

In this patient with repaired totally anomalous pulmonary venous connection, computed tomographic angiography reveals the substrate for significant obstruction (arrow). The right lower pulmonary vein (RLPV) is stenotic at its junction with the left atrium (LA), and the left lower pulmonary vein (LLPV) appears to no longer be connected to the left atrium. The left pulmonary veins drain via a vertical vein (not imaged here), which was left patent at surgery. I, inferior; L, left; R, right; S, superior.

0 Notes

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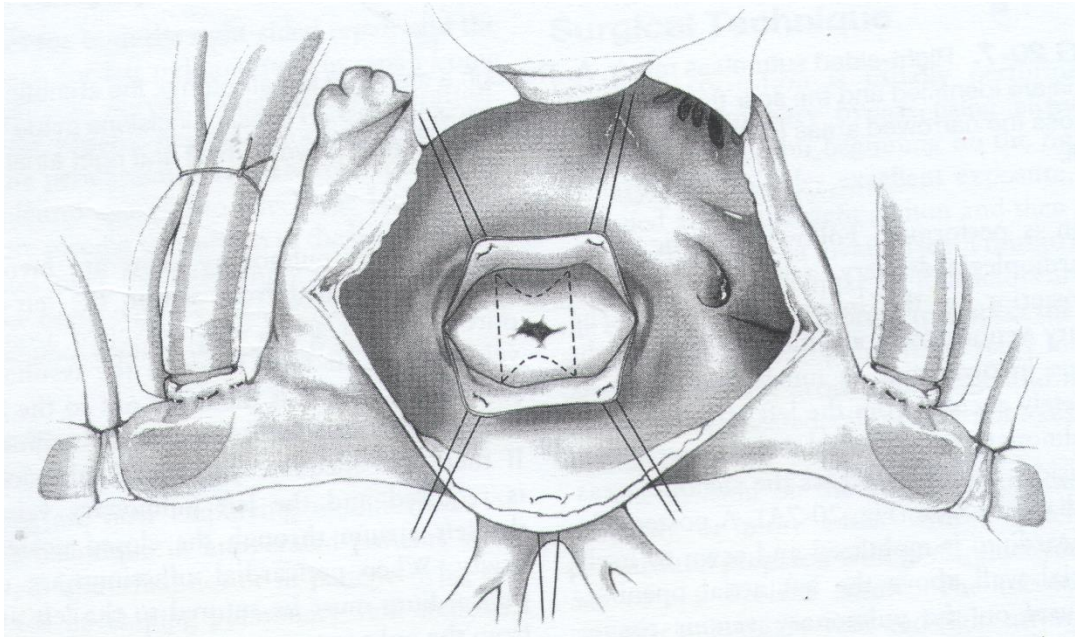
Totally anomalous pulmonary venous connection

- Results of surgery
- **Postoperative PV stenosis**



Totally anomalous pulmonary venous connection

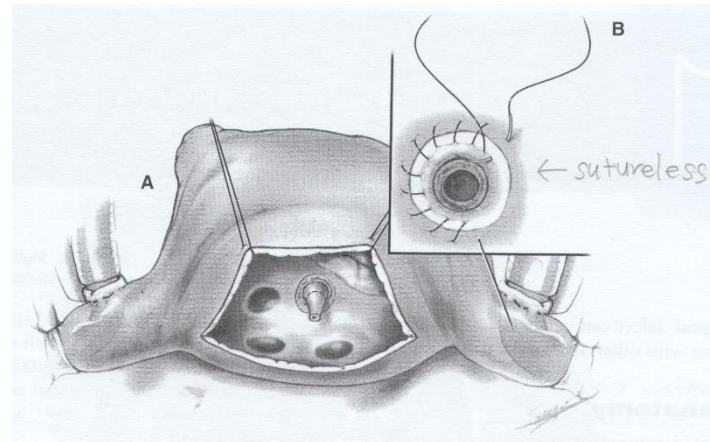
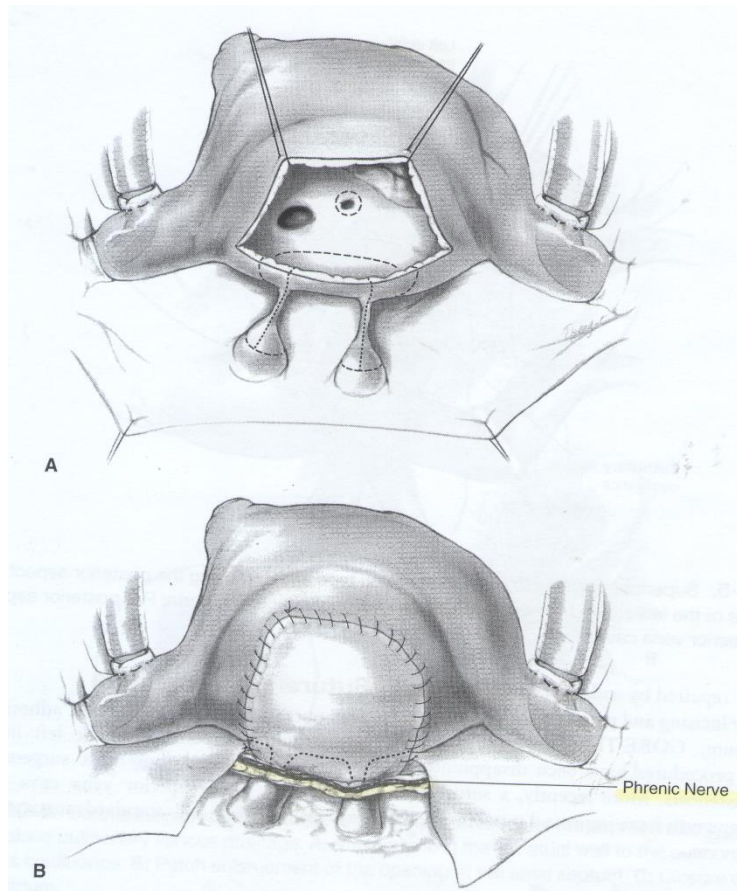
- Results of surgery
- Pulmonary venous stenosis after surgery
 - **Conventional technique**



- . An isolated anastomotic stenosis is approached through a right atriotomy and vertical incision on the atrial septum
- . The narrowed anastomosis is enlarged by removing as much of the tissue as possible between the posterior left atrium and the pulmonary veins

Totally anomalous pulmonary venous connection

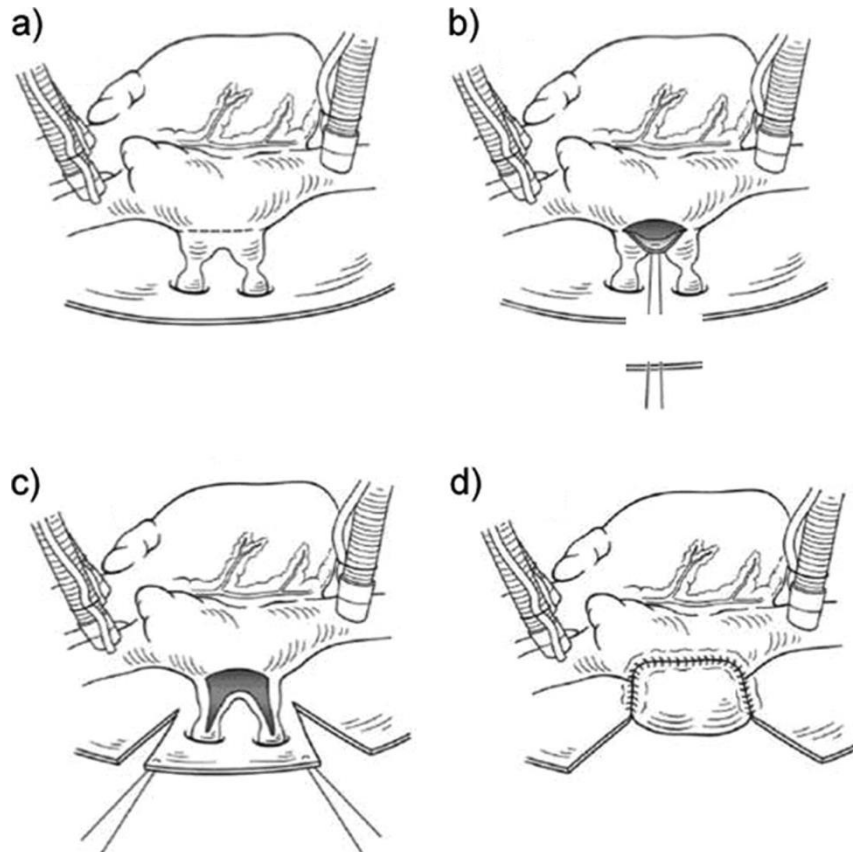
- Results of surgery
- Pulmonary venous stenosis after surgery
 - **Sutureless technique**



- . Often in a reoperation, **the course of the phrenic nerve** cannot be appreciated from within the pericardial space
- . Therefore, it is best to **open the pleural spaces to check the location of the nerve** before placing the sutures in the pericardium

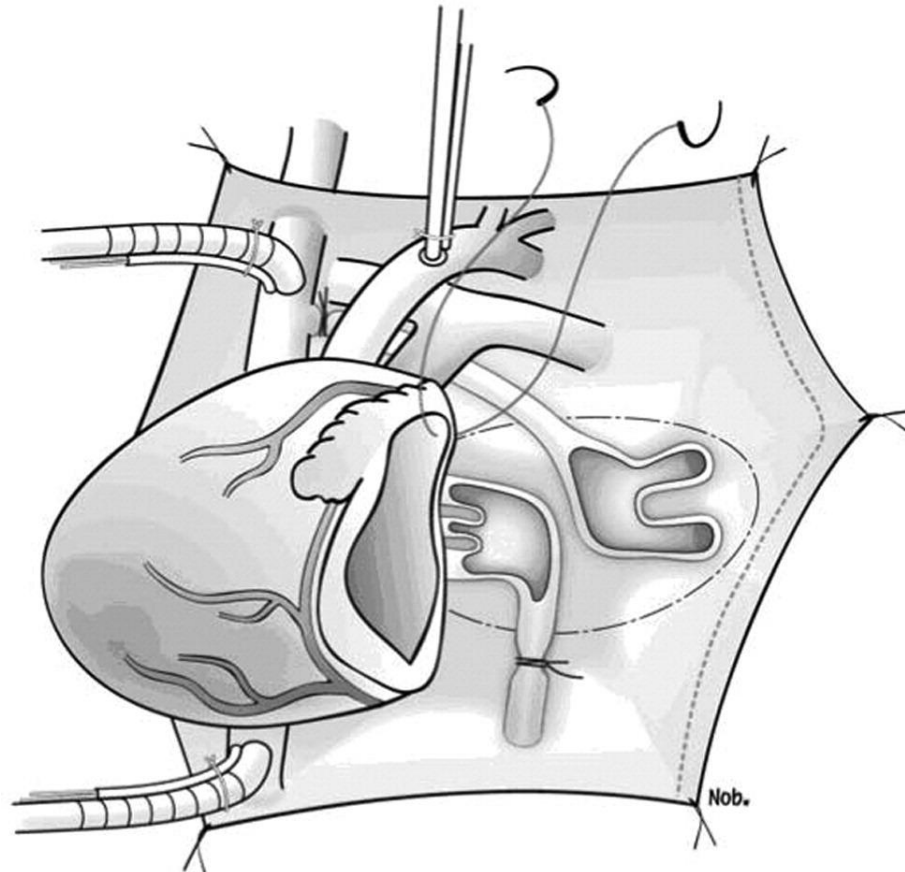
Totally anomalous pulmonary venous connection

- Results of surgery
- Pulmonary venous stenosis after surgery
 - **Sutureless technique**



Totally anomalous pulmonary venous connection

- Results of surgery
- Pulmonary venous stenosis after surgery
 - **Sutureless technique (primary application)**



Totally anomalous pulmonary venous connection

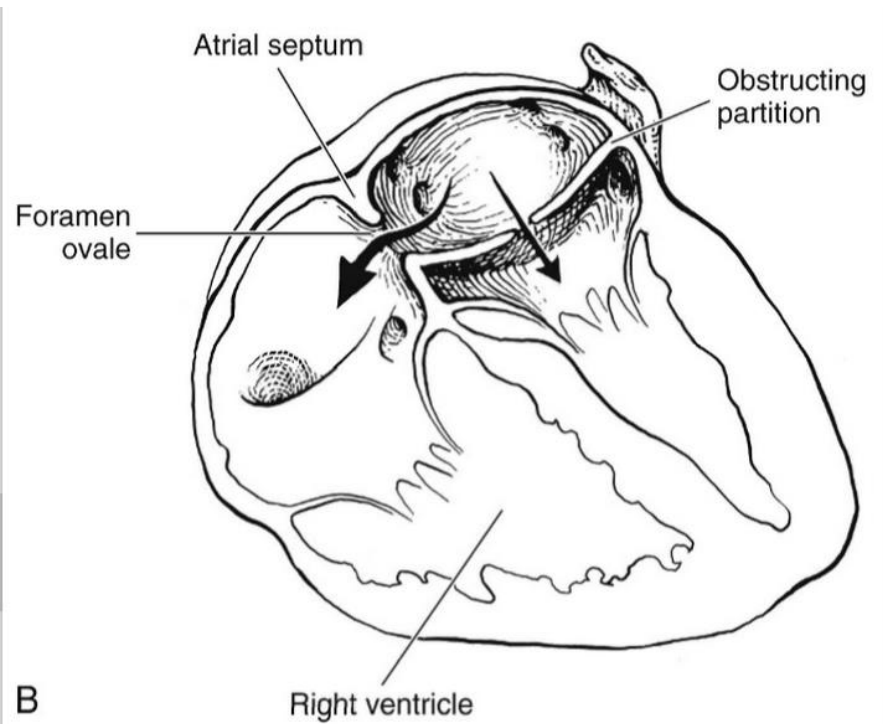
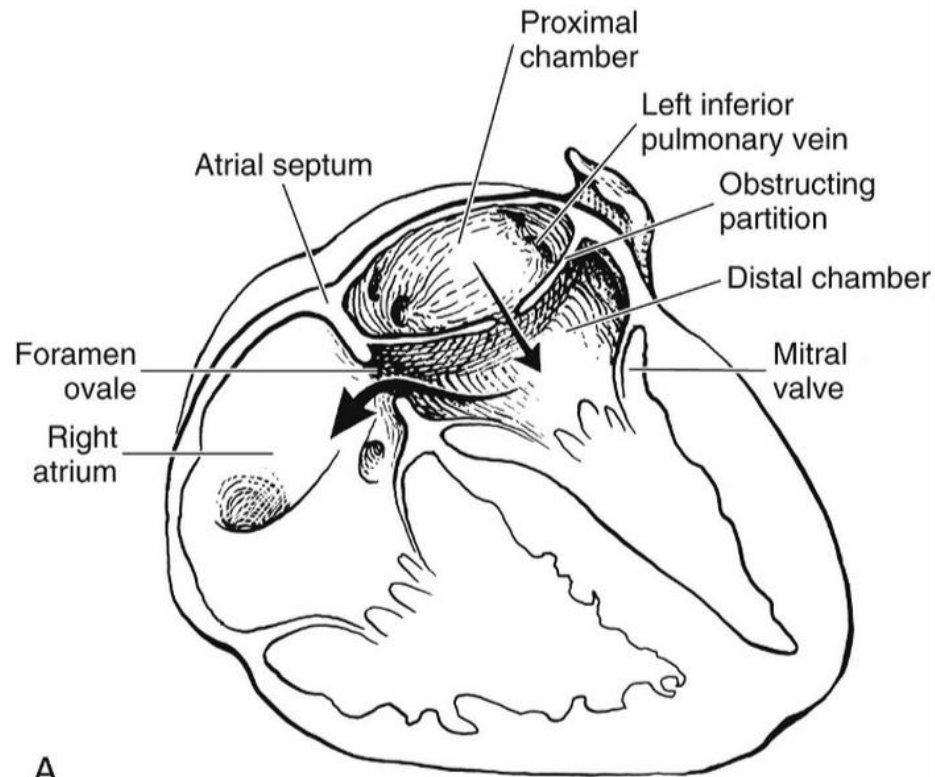
- Results of surgery
 - The poor results of surgical relief of post-operative pulmonary venous obstruction have led to placement of stents across the stenoses either percutaneously or intraoperatively, but the results have either been disappointing, or else satisfactory over a very short term

Totally anomalous pulmonary venous connection

- Results of surgery
 - A recent study by Kirshbom and colleagues evaluated long-term results of patients with totally anomalous pulmonary venous connection repaired between 1983 and 2005
 - They reported an 84% 17-year survival rate, with most deaths occurring within a few months after surgery
 - Over 90% of their patients reported excellent or good overall health, and school performance was average or better in 69% of subjects

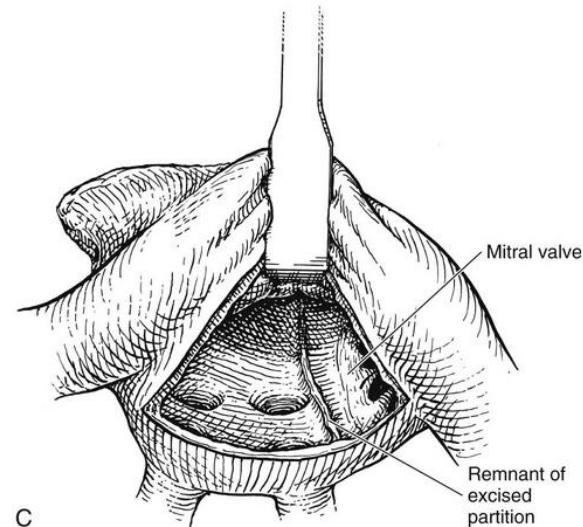
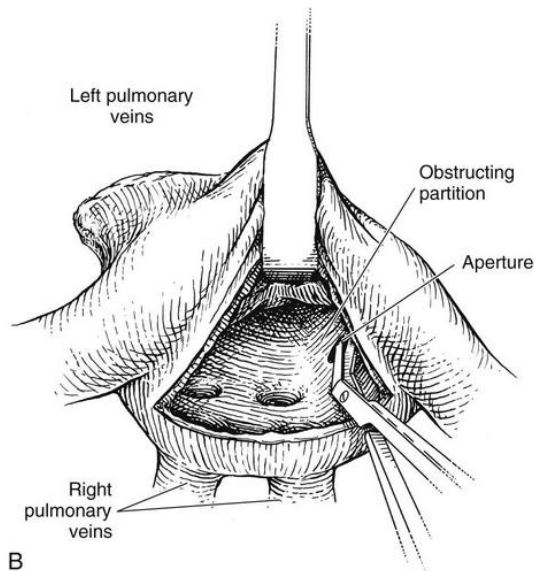
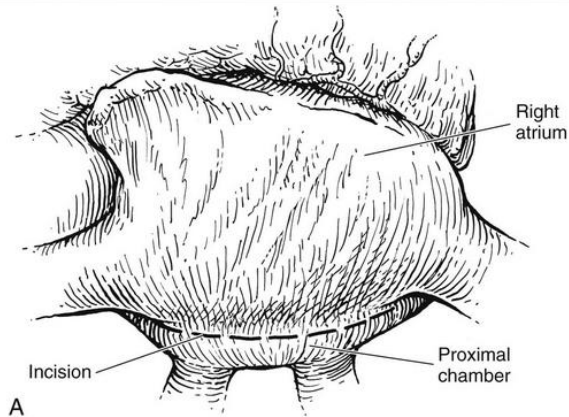
Cor Triatriatum

- Morphology



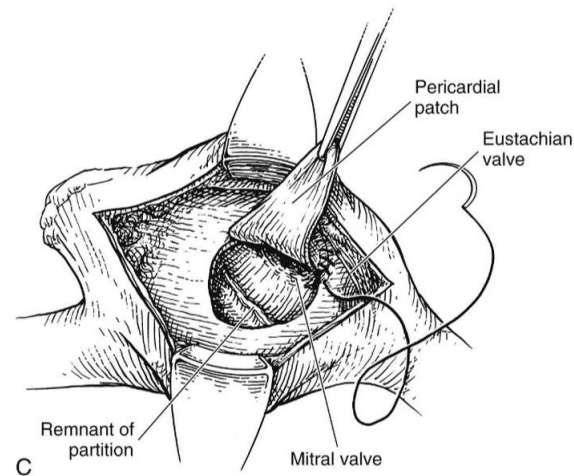
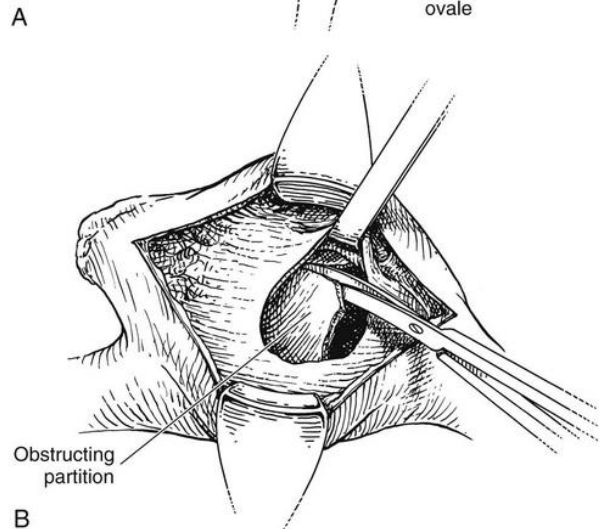
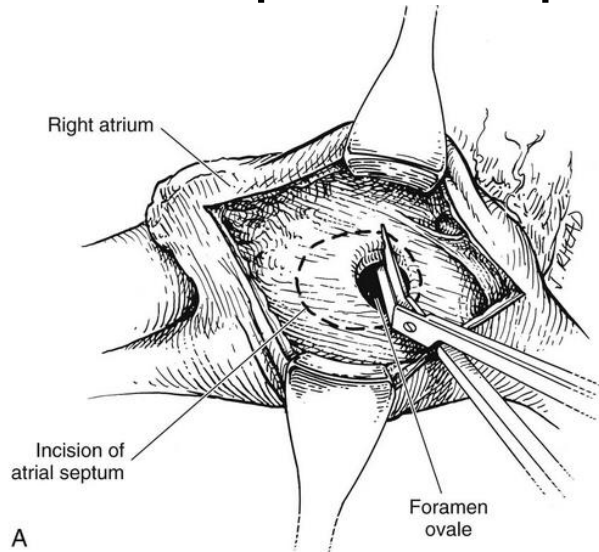
Cor Triatriatum

- Technique of operation (left-side approach)



Cor Triatriatum

- Technique of operation (right-side approach)



Atresia of the common pulmonary vein

- Anatomy
 - The essence of atresia of the common pulmonary vein is absence of functional connection between the pulmonary veins and the morphologically left atrium

Atresia of the common pulmonary vein

- Pathophysiology
 - There is **no direct route for blood to enter either the left atrium or any systemic vein** and yet patients have lived for up to a month with this condition
 - One suggested route is via bronchopulmonary venous anastomoses to the pleurohilar bronchial veins, which drain into the azygos, hemiazygos and brachiocephalic veins

Atresia of the common pulmonary vein

- Clinical presentation and investigation
 - The age at presentation, clinical findings, electrocardiogram and chest radiograph are as for totally anomalous connection with severe pulmonary venous obstruction
 - The diagnosis is difficult

Atresia of the common pulmonary vein

- Imaging
 - Given the often small pulmonary venous confluence and severely obstructed flow through the pulmonary veins, this diagnosis can be very challenging to make using echocardiography
 - Computed tomographic angiography can be attempted to define the anatomy

Atresia of the common pulmonary vein

- Management
 - There is **no medical treatment**
 - A few centers have reported success using aggressive haemodynamic stabilisation techniques, often including **extracorporeal membrane oxygenation**, and **early surgical repair**

Unilateral pulmonary venous atresia

- Unilateral pulmonary venous atresia is a very rare condition in which there is complete obliteration of luminal continuity between the pulmonary venous pathway of one lung and the left atrium
- Symptoms – within the first 3 years of life, displaying recurrent pulmonary infections, exercise intolerance, and/or haemoptysis
- Both reparative surgery and unilateral pneumonectomy have been successfully performed, although survival appears to be superior with pneumonectomy

Partially anomalous pulmonary venous connection

- Anatomy
 - Partially anomalous connection exists where one or more of the pulmonary veins is connected to the morphologically left atrium, while the rest are connected to a systemic vein or the right atrium
 - The association between [the sinus venosus interatrial communication](#) and anomalous drainage of the right pulmonary veins
 - The [Scimitar syndrome](#) – the lower lobe of the right lung is hypoplastic, and is supplied with arterial blood from the descending aorta, and its [pulmonary venous return is connected to the inferior caval vein](#)

Partially anomalous pulmonary venous connection

- Morphogenesis
 - Presumably the canalising pulmonary vein in patients with partially anomalous connection will have made connection only with part of the intraparenchymal pulmonary venous plexus

Partially anomalous pulmonary venous connection

- Pathophysiology
 - Because pulmonary venous obstruction is rarely present with partially anomalous venous connections, the haemodynamic effects are almost always the result of an obligatory left-to-right shunt through the anomalously connected segments of lung
 - Pulmonary hypertension is rare, except in some infants with scimitar syndrome

Partially anomalous pulmonary venous connection

- Presentation and symptoms
 - Patients with scimitar syndrome can present with recurrent pneumonia, wheezing, or haemoptysis
 - The physical signs in patients with an associated atrial septal defect are as for the atrial septal defect

Partially anomalous pulmonary venous connection

- Investigations
- Chest radiography

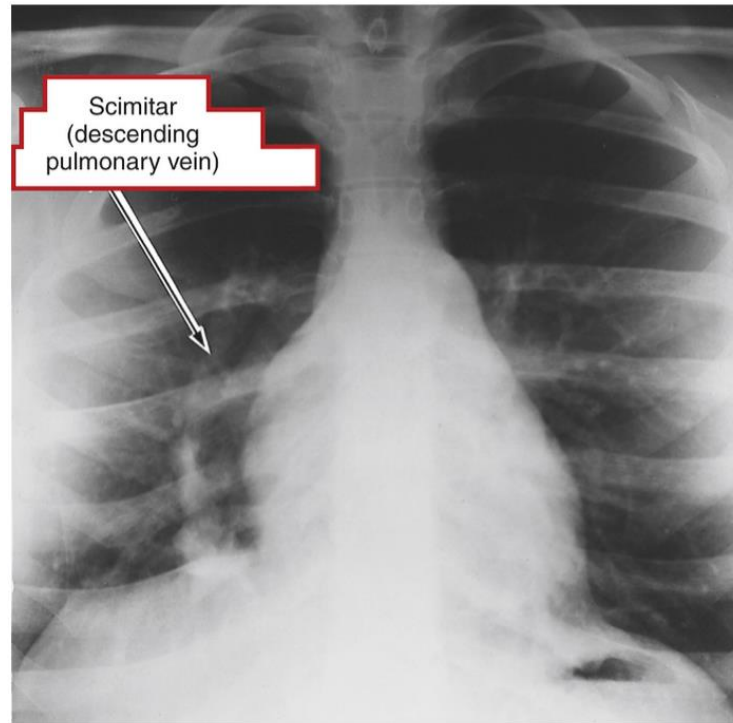
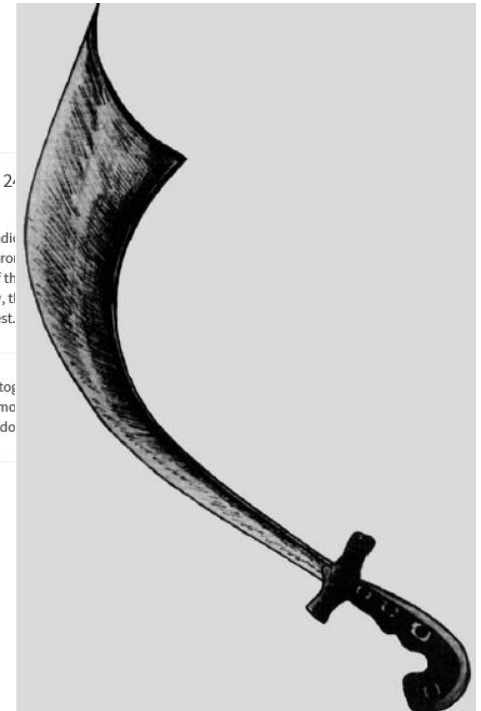


Figure 2.

Plain chest radiograph showing scimitar syndrome, hypoplasia of the left lung, and consequently, the scimitar in the left chest.

(Original photograph by the late Dr Simon, St. George's Hospital, London)

0 Notes



Partially anomalous pulmonary venous connection

- Investigations
- Echocardiography
 - Increased or atypical flow in the superior or inferior caval veins should alert one to the possibility of partially anomalous pulmonary venous connection

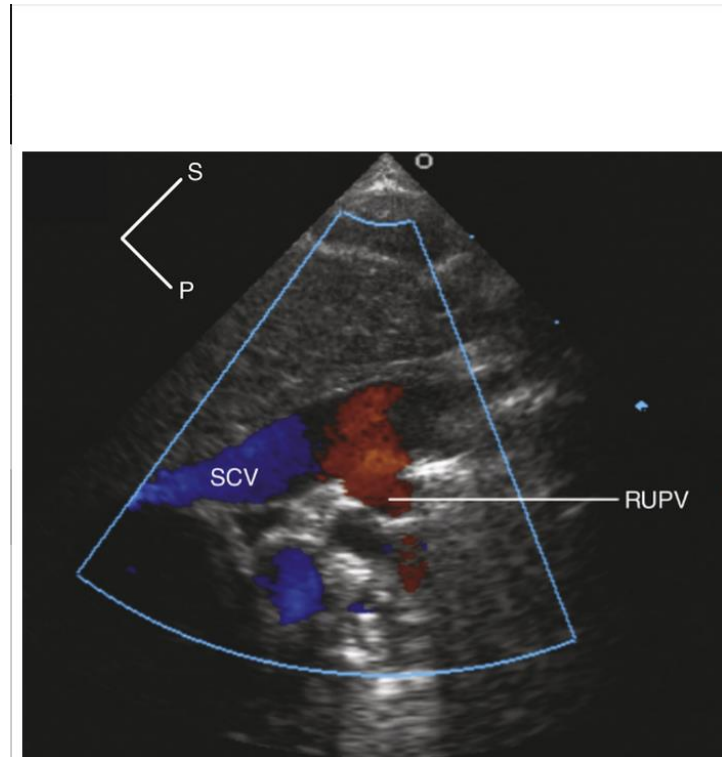


Figure 24-32

In this high right parasternal view, the right upper pulmonary vein (RUPV) can be seen entering the superior caval vein (SCV). P, posterior; S, superior.

0 Notes

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Partially anomalous pulmonary venous connection

- Investigations
- Computed tomographic angiography

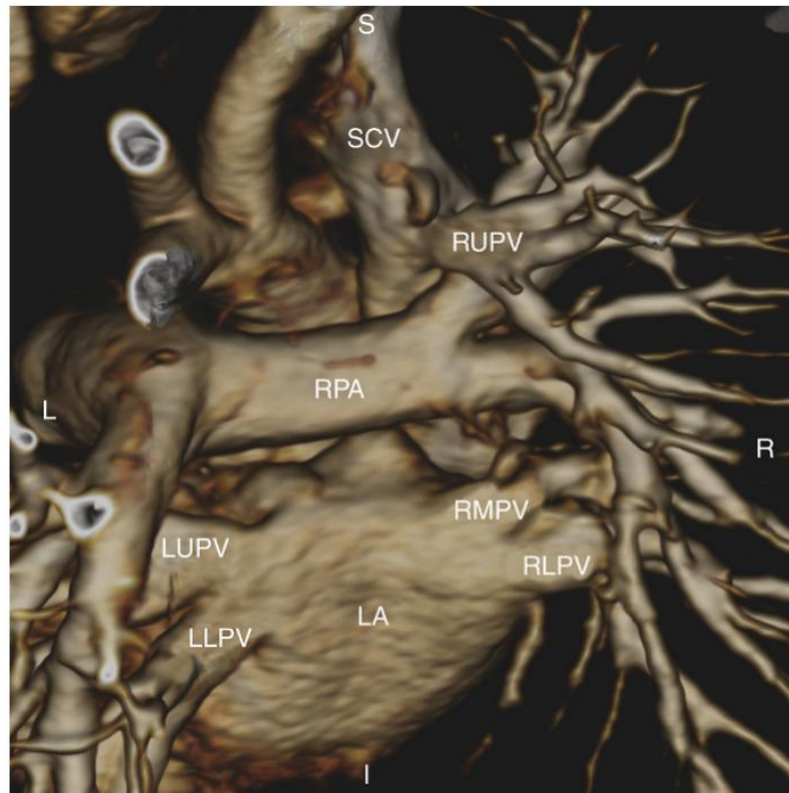


Figure 24-33

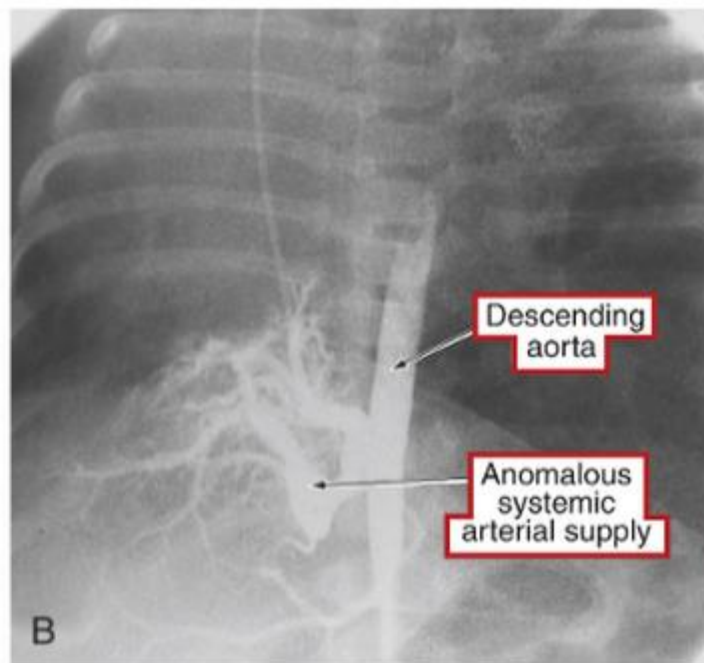
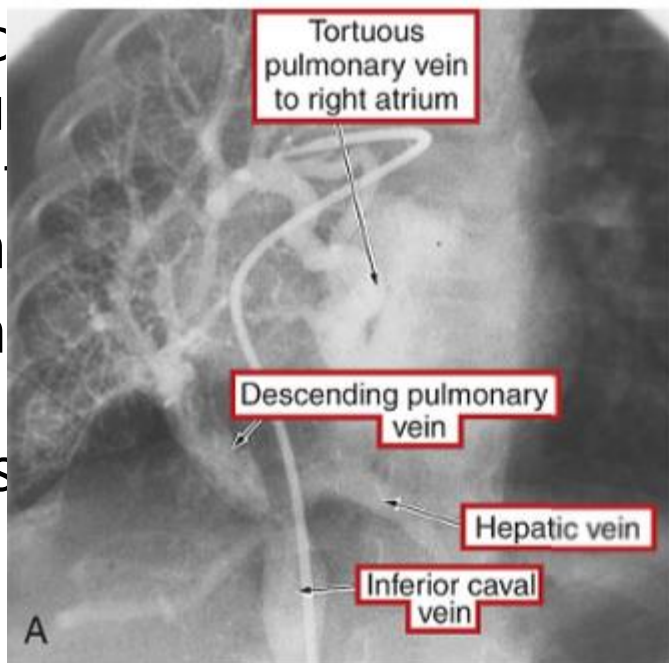
This computed tomographic angiogram shows the anomalous connection of the right upper pulmonary vein (RUPV) to the superior caval vein (SCV). Note that there are four pulmonary veins (LUPV, LLPV, RMPV, RLPV) entering the left atrium (LA). This highlights the challenge in detecting this anomaly by echocardiography. I, inferior; L, left; R, right; RPA, right pulmonary artery; S, superior.

0 Notes

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Partially anomalous pulmonary venous connection

- Investigations
- Cardiac catheterisation and angiocardiography
 - Cardiac catheterisation is rarely indicated for these patients
 - In diagnosing partially anomalous pulmonary venous connection, contrast injection in the superior or the inferior vena cava or the inferior pulmonary vena
 - In the case of a partially anomalous pulmonary venous connection, contrast injection in the superior vena cava is also essential

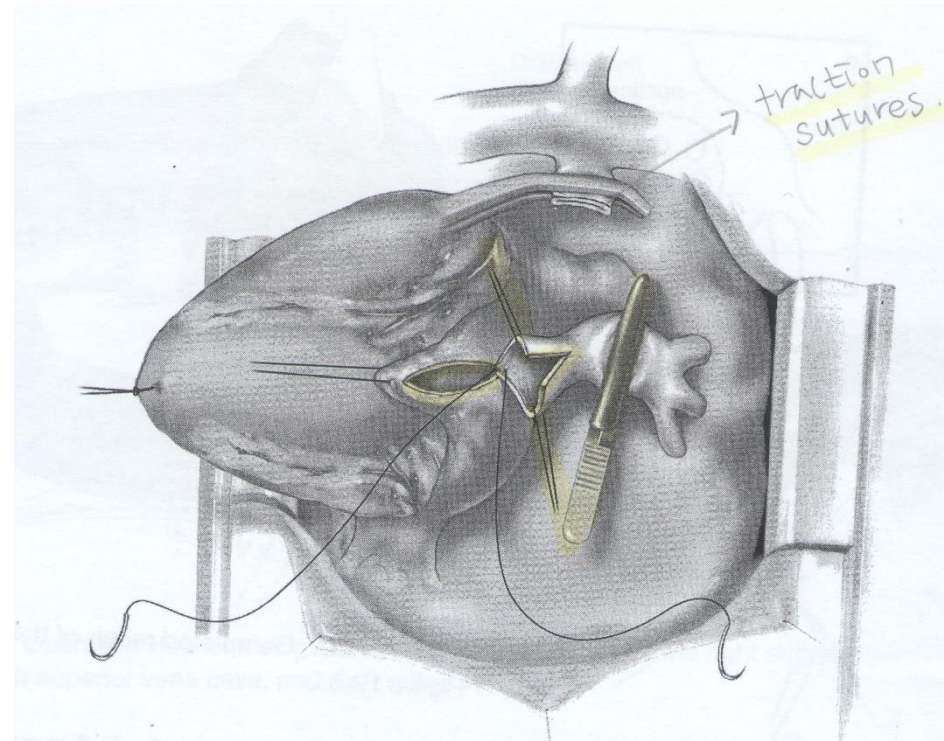


Partially anomalous pulmonary venous connection

- Course and prognosis
 - The course and prognosis for partially anomalous pulmonary venous connection are probably similar to those of an isolated atrial septal defect with a comparable left-to-right shunt
 - Prognosis for patients with scimitar syndrome is worse than other types of partially anomalous pulmonary venous drainage, particular among those who develop symptoms within the first year of life

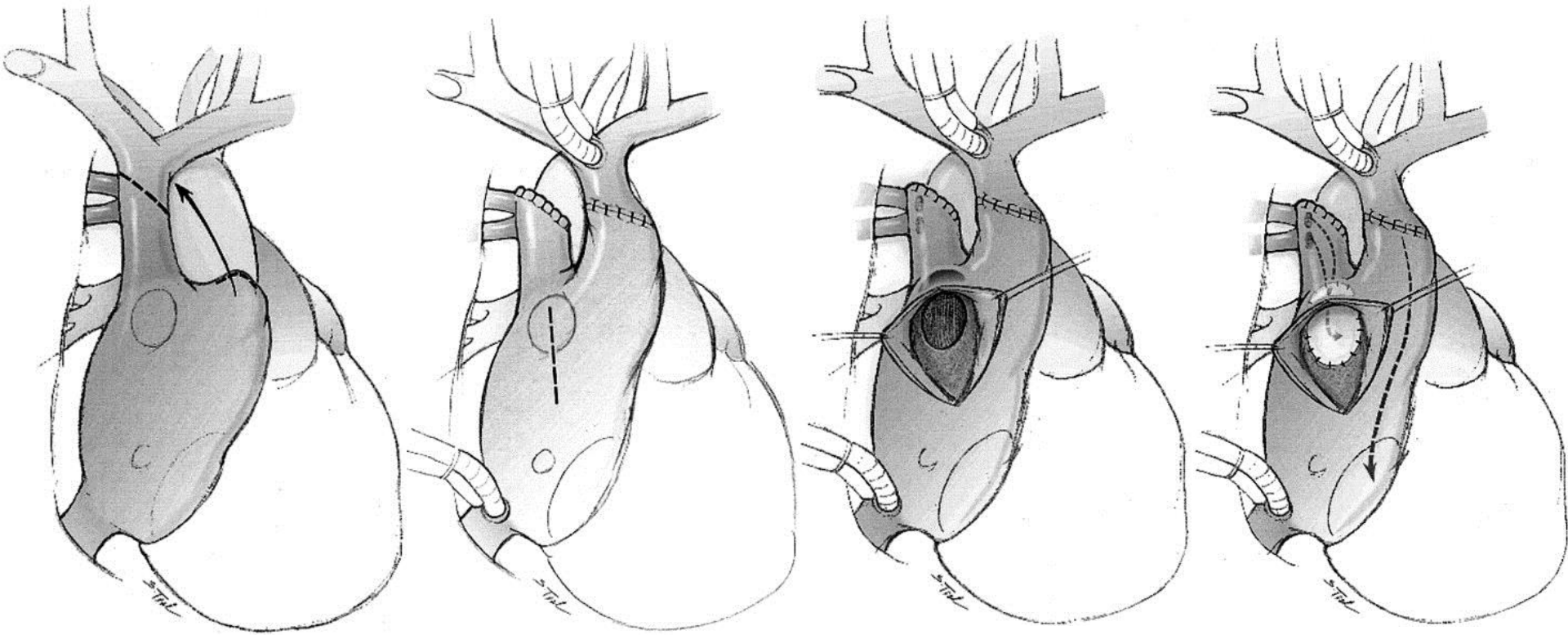
Partially anomalous pulmonary venous connection

- Management
- Surgery
 - Some patients with scimitar syndrome benefit from a concomitant resection of the pulmonary sequestration, if present
 - The usual method of repair of unilateral connection of all the veins from the left lung to a left vertical vein is to connect the common left pulmonary vein to the amputated stump of the left atrial appendage



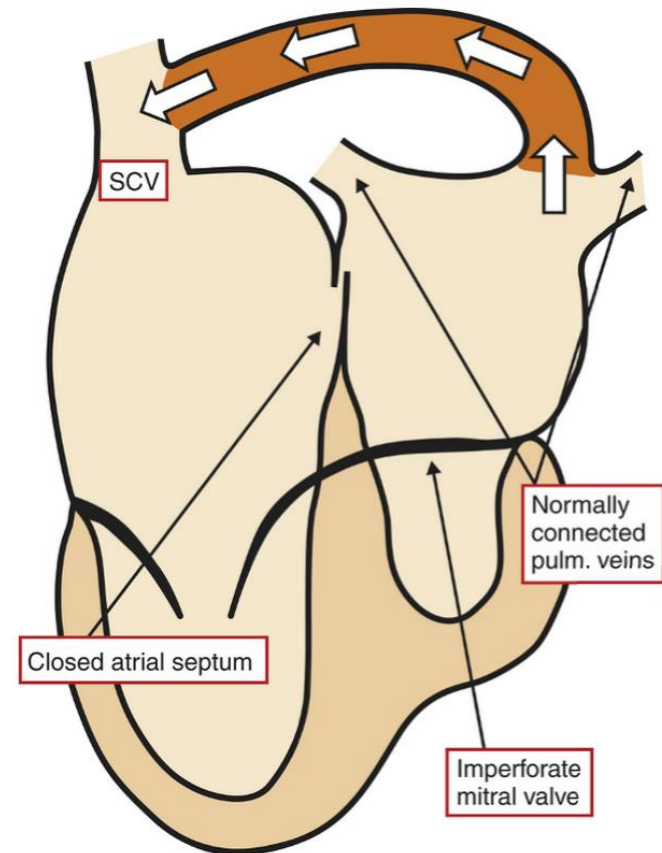
Patially anomalous pulmonary venous connection

- Management
- Surgery
 - Warden operation for sinus venosus atrial septal defect with PAPVC (RUPV to SVC)



Anomalous pulmonary-to-systemic collateral veins and the laevoatrial cardinal vein

- In these circumstances, the clinical presentation will be generally dominated by the lesion causing the pulmonary venous obstruction
- Along those lines, Vance reported placement of a stent in the laevoatrial cardinal vein as a bridge to surgery in a patient with severe pulmonary venous obstruction due to an intact atrial septum in the setting of mitral atresia



Stenosis or a pulmo

- Anatomy
 - Individual pulmonary veins can be stenosed at the junction with the atrium, or have tubular hypoplasia or significant intra- or extrapulmonary distal stenosis.
 - Pulmonary venous stenosis also results from acquired causes, such as constrictive pericarditis, mediastinal lymphadenopathy, pulmonary tuberculosis, and invasion by tumour.

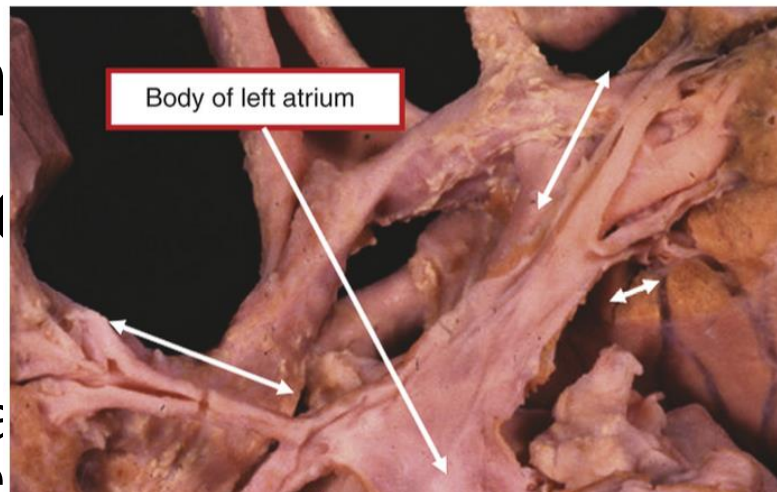


Figure 24-37

The heart from this patient, who died due to pulmonary venous stenosis, has been dissected to show the tubular hypoplasia of each of the four individual pulmonary veins (arrows).

0 Notes

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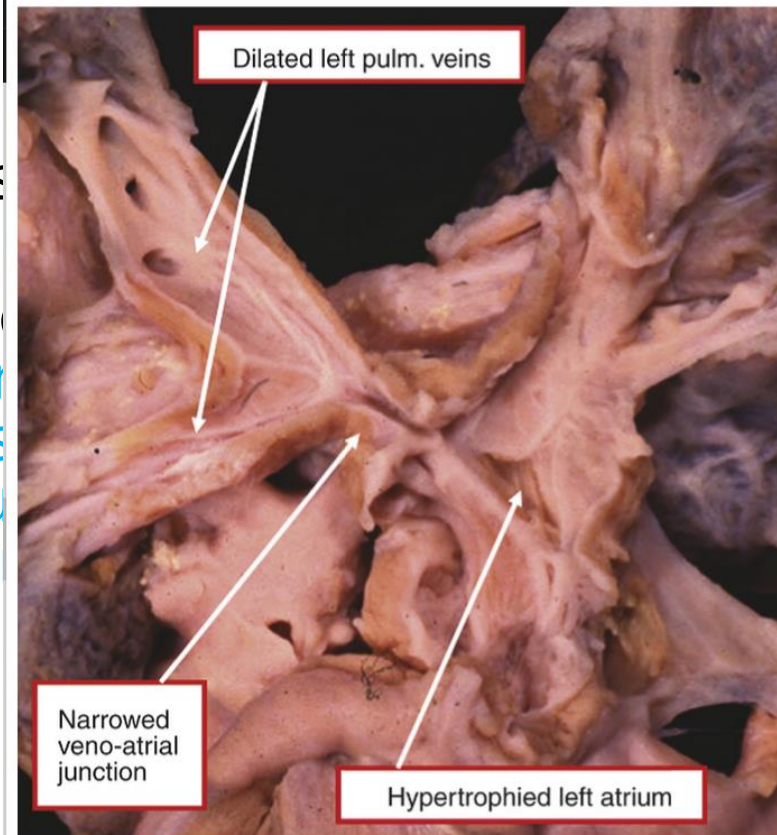


Figure 24-38

In this patient, who also suffered severe pulmonary stenosis in life, the stenotic sites are at the junctions of the individual pulmonary veins with the body of the left atrium. Note the dilated pulmonary veins, and the hypertrophied walls of the left atrium.

0 Notes

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Stenosis or atresia of individual pulmonary veins

- Pathophysiology
 - Just under half the patients described have had other congenital heart defects, varying from the simple to the complex
 - The greater the number of pulmonary veins obstructed, and the more severe the obstruction, the more severe will be the pulmonary hypertension

Stenosis or atresia of individual pulmonary veins

- Presentation and symptoms
 - Dyspnea and repeated pulmonary infections with failure to thrive are the rule
 - Many patients have haemoptysis and, occasionally, cyanosis

Stenosis or atresia of individual pulmonary veins

- Investigations
- Chest radiography
 - The heart is usually normal in size or slightly enlarged, with prominence of the pulmonary trunk
 - The lung show a reticular appearance or **ground-glass opacification**
 - There may be **hypoplasia of the lung on the affected side**

Stenosis or atresia of individual pulmonary veins

- Investigations
- Echocardiography
 - Turbulence or a focal increase in flow velocity with a continuous, non-phasic flow pattern distally in a pulmonary vein suggests stenosis

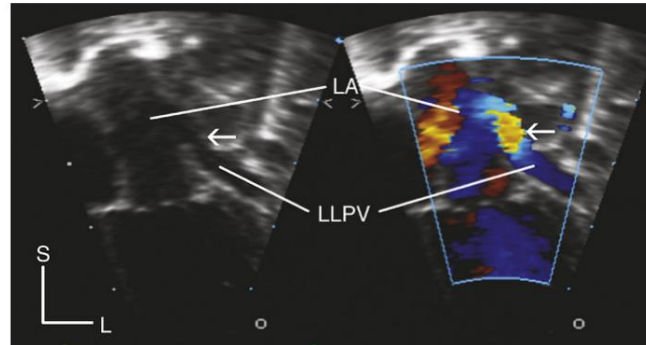


Figure 24-39

In this zoomed apical four-chamber view, shown with and without colour Doppler, an area of turbulence (arrow) can be seen as the left lower pulmonary vein (LLPV) enters the left atrium (LA). L, left; S, superior.

0 Notes

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Stenosis or atresia of individual pulmonary veins

- Investigations
- Computed tomographic angiography

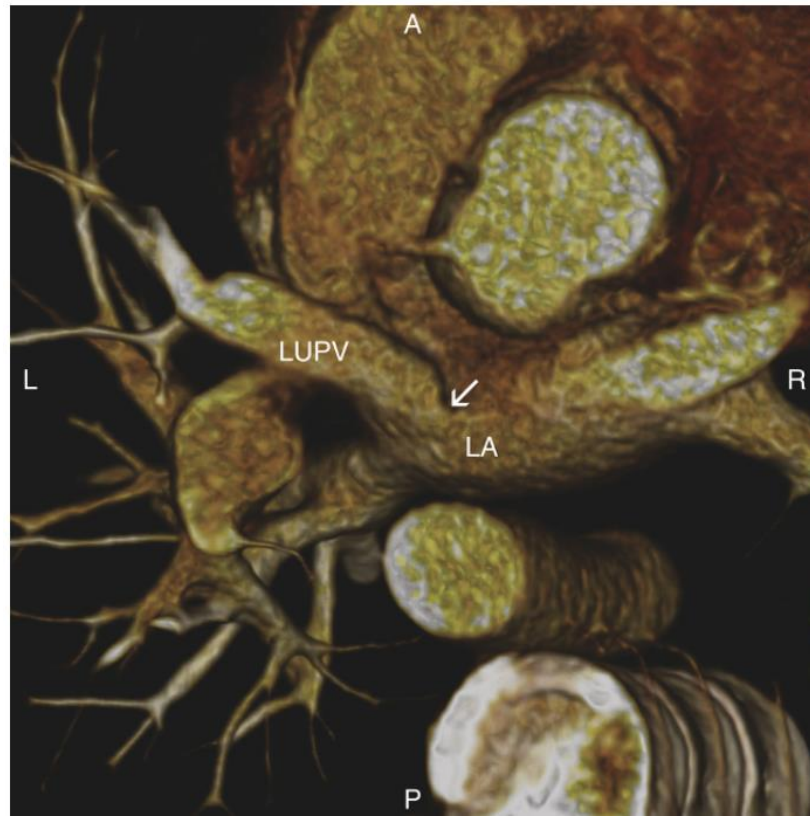


Figure 24-42

This computed tomographic angiogram shows stenosis (arrow) of the left upper pulmonary vein (LUPV) as it enters the left atrium (LA) in a patient after radio-frequency ablation for atrial fibrillation. A, anterior; L, left; P, posterior; R, right.

0 Notes

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Stenosis or atresia of individual pulmonary veins

- Differential diagnosis
 - Stenosis or atresia of one or two pulmonary veins must be distinguished from pulmonary infection
 - Stenoses or atresia of most or all pulmonary veins must be distinguished from other causes of generalized pulmonary venous hypertension

Stenosis or atresia of individual pulmonary veins

- Course and prognosis
 - Of these, 3/5 either died or underwent lung transplantation
 - 4/5 of those with a mean pulmonary arterial pressure higher than 33 mmHg experienced death or lung transplantation

Stenosis or atresia of individual pulmonary veins

- Management
- Surgery
 - Localised atresia or stenosis has been successfully treated by patch grafting, side-to-end anastomosis of the vein to the left atrium, excision of an obstructing membrane at the junction of the pulmonary vein with the left atrium, or by the so-called sutureless technique described for treatment of post-operative stenosis after repair of anomalous pulmonary venous connection
 - The sutureless technique is currently the most promising

Stenosis or atresia of individual pulmonary veins

- Management
 - While catheter-based interventions using **stents, and cutting balloons**, have been successful in adults with pulmonary venous stenosis occurring after catheter ablation for atrial fibrillation, the long-term results for other causes of pulmonary venous stenosis have been disappointing

Stenosis or atresia of individual pulmonary veins

- Management
 - The only effective treatment for long-segment atresia or severe pulmonary venous hypoplasia is **pneumonectomy**, when the disease is **unilateral** and the objective is to **cure massive haemoptysis**

Pulmonary varix

- This lesion is considered benign, and treatment is generally reserved for those who experience complications such as haemoptysis or cerebral embolic events, or for those in whom growth of the defect results in compression of surrounding structures