

# Pulmonary venous abnormalities

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# Pulmonary venous abnormalities

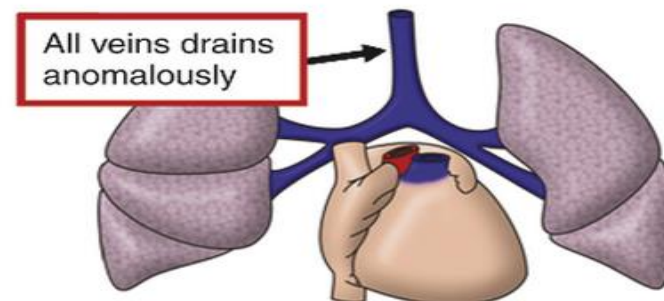
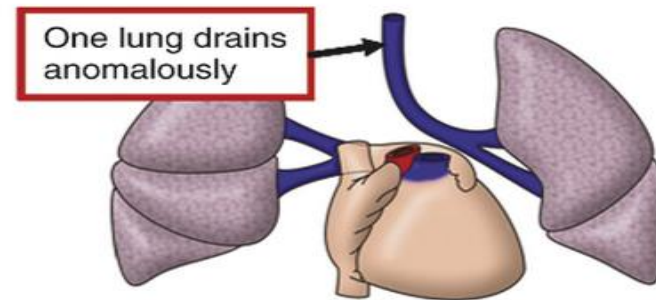
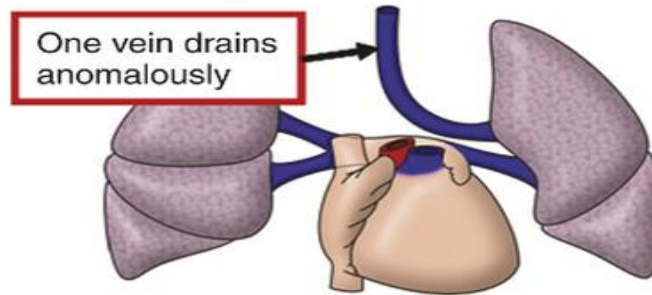
- TAPVC
- Cor Triatriatum
- PAPVC
- Stenosis or atresia of individual pulmonary veins

# Totally anomalous pulmonary venous connection

- Incidence and aetiology
  - 1.5% of all patients with a cardiovascular malformation
  - once in 14,700 live births
    - Two-thirds with **supracardiac** and **cardiac** connections
      - males
    - **infradiaphragmatic** variant - 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

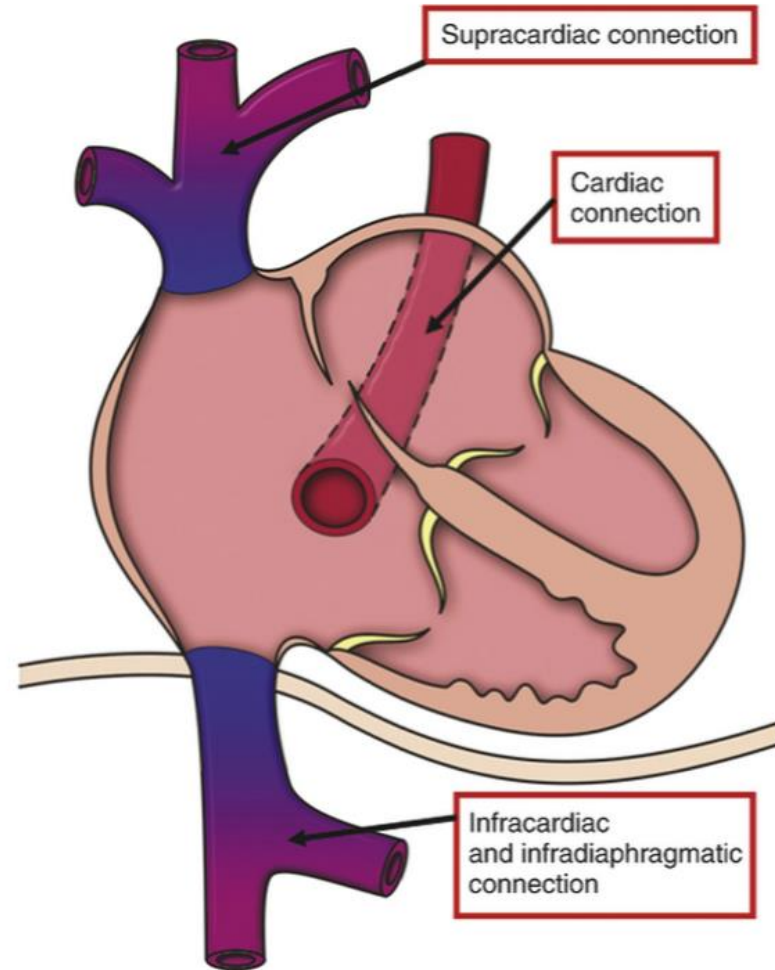


# Totally anomalous pulmonary venous connection

- 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
  - **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
  - supracardiac, 45%
  - cardiac (intracardiac), 25%
  - infracardiac, 25%
  - mixed, 5%

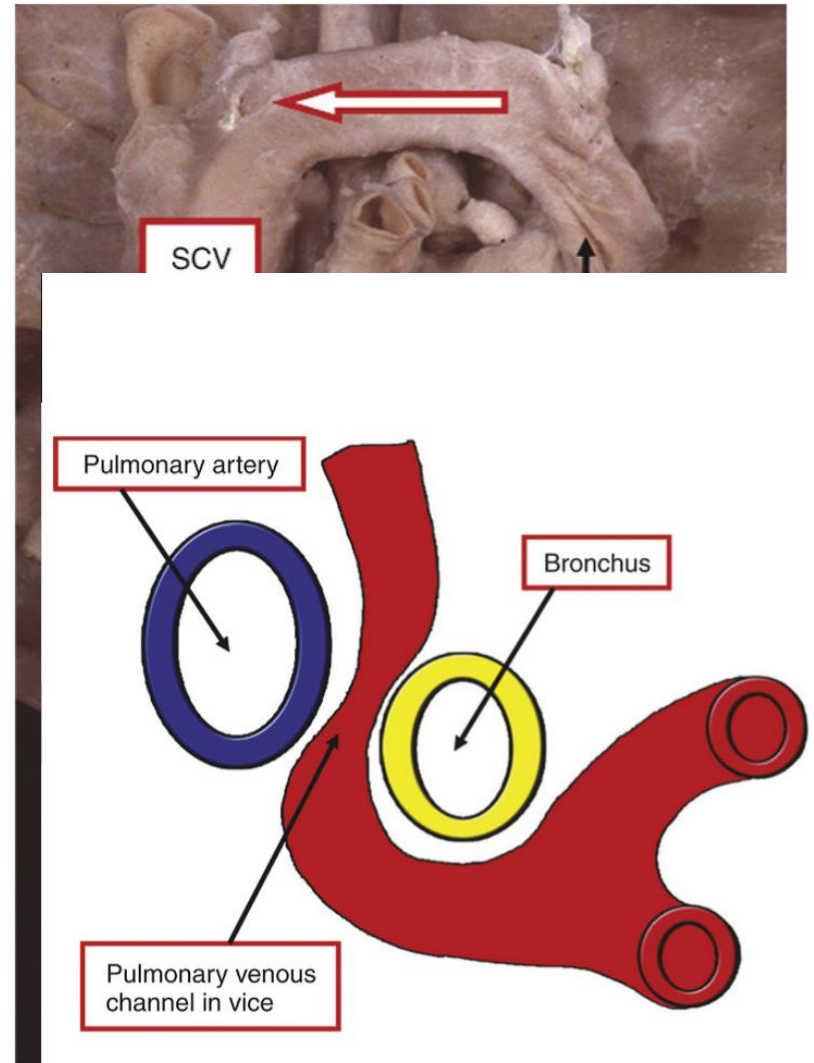
# Totally anomalous pulmonary venous connection (supracardiac type)

- Anatomy
  - Supracardiac connection
    - left brachiocephalic vein
    - right SVC
    - azygos vein
    - left SVC



# Totally anomalous pulmonary venous connection (supracardiac type)

- 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

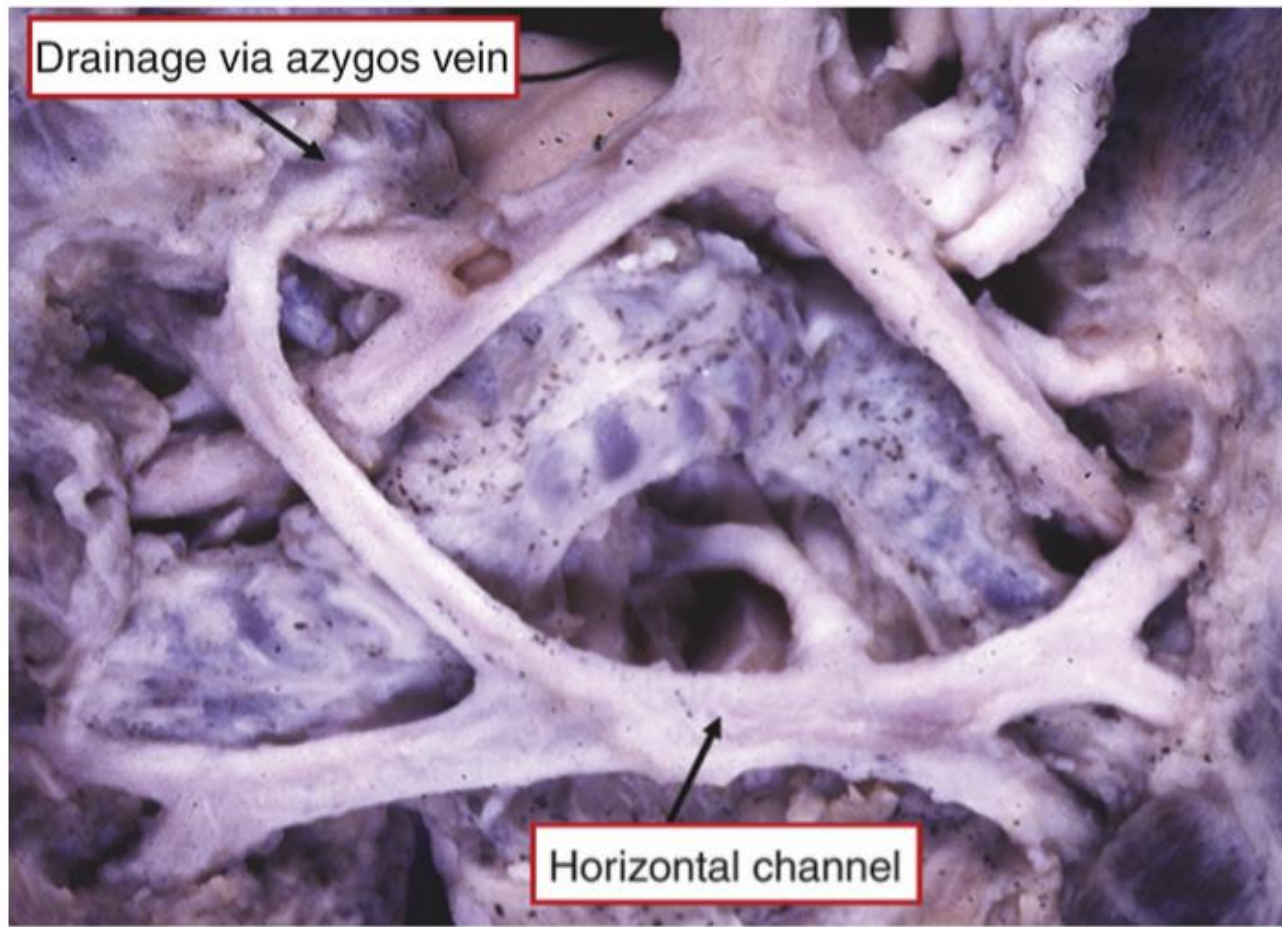


# Totally anomalous pulmonary venous connection (supracardiac type)

- 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**

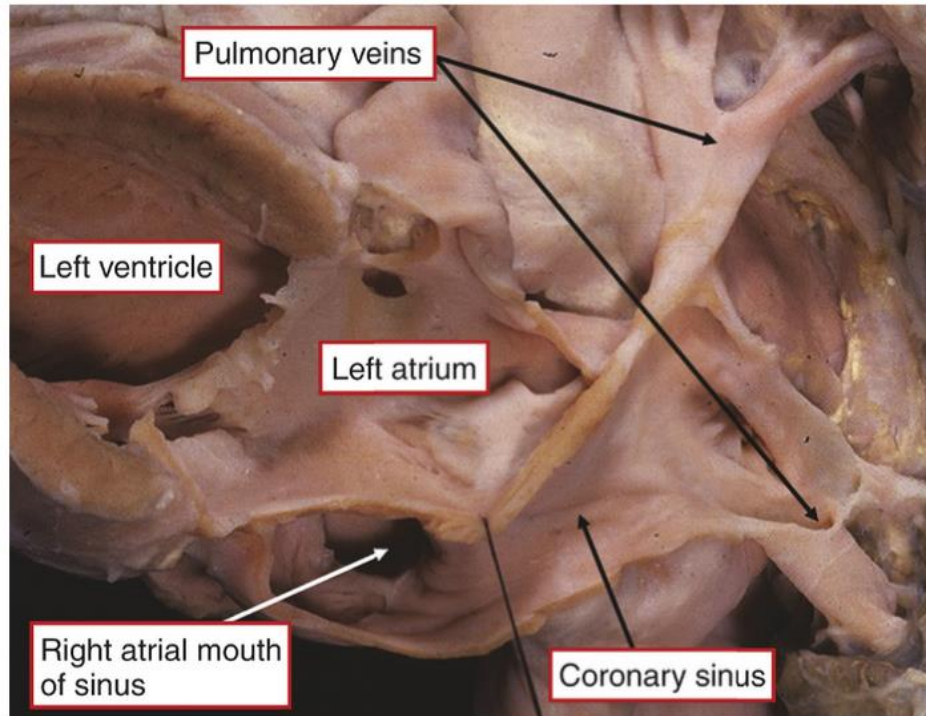
# Totally anomalous pulmonary venous connection (supracardiac type)

- Anatomy



# Totally anomalous pulmonary venous connection (cardiac type)

- Anatomy
  - cardiac form of anomalous connection - 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



# Totally anomalous pulmonary venous connection (cardiac type)

- Anatomy
  - Direct connection of the pulmonary veins to the morphologically right atrium is exceedingly rare

# Totally anomalous pulmonary venous connection (infracardiac type)

- 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 (infracardiac type)

- 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 (mixed type)

- Anatomy
  - possible for different veins to terminate in different anomalous sites (**mixed anomalous connection**)



# Totally anomalous pulmonary venous connection

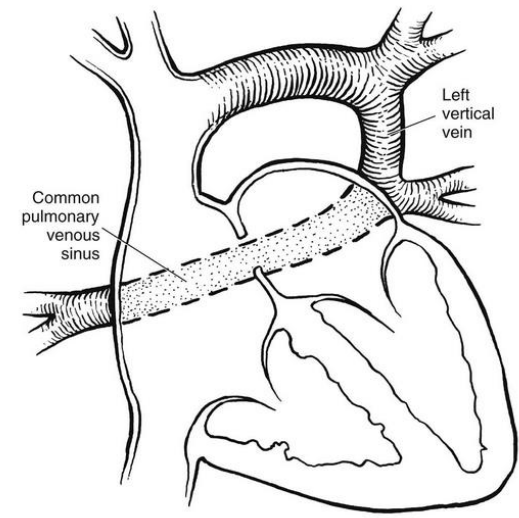
- Anatomy
  - A large proportion of the patients - **associated malformations**
  - an **interatrial communication** present, so that venous blood is **able to reach the left side of the heart**

# 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 connection

- Anatomy
- Supracardiac type



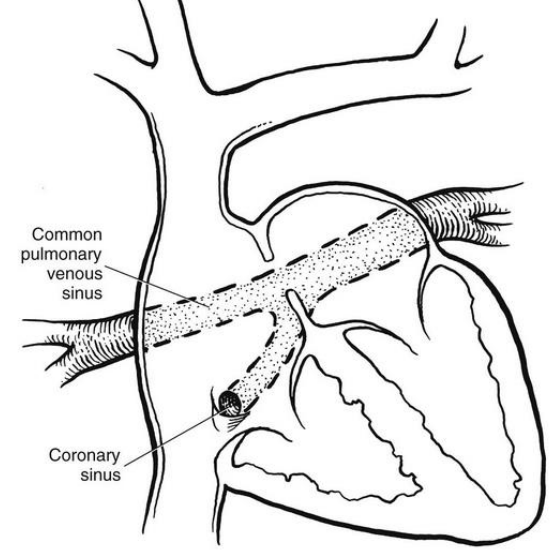
A

Supracardiac type (45%)



# Totally anomalous p venous connec

- Anatomy
- Cardiac type

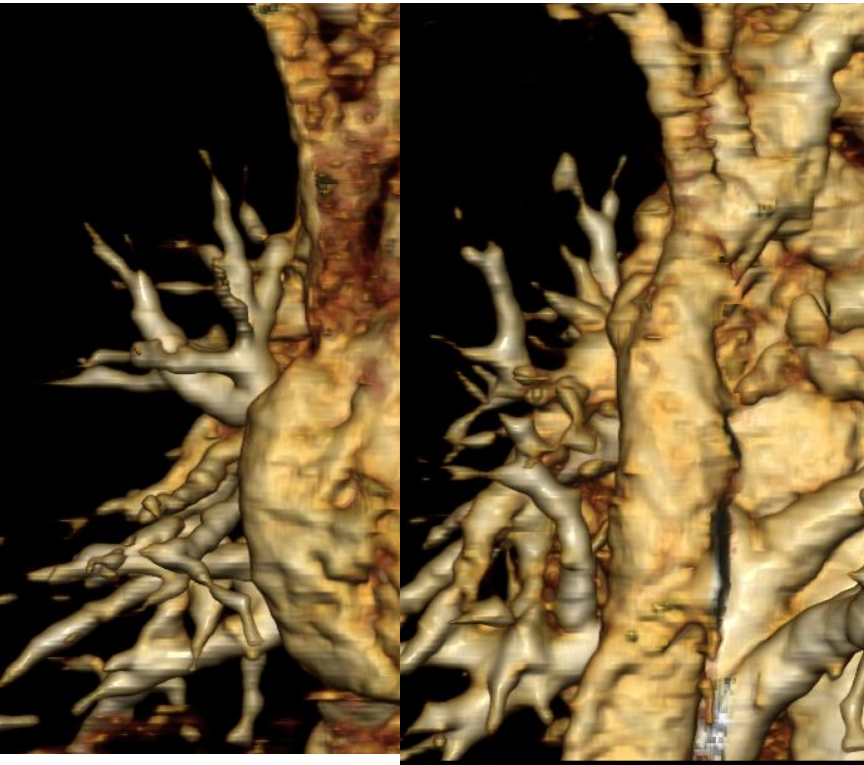
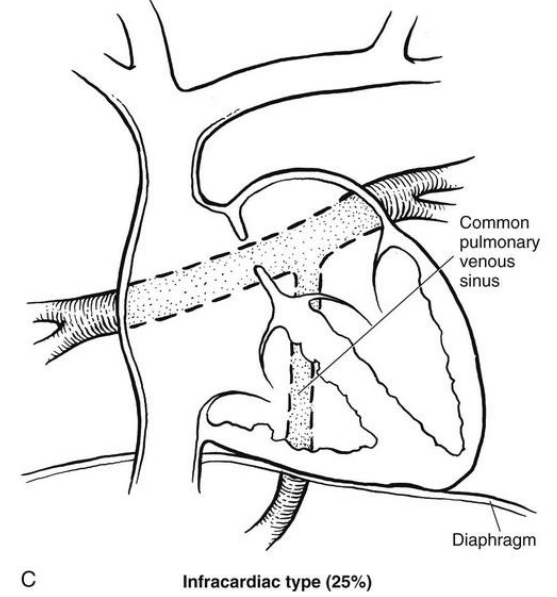


B Cardiac type (25%)

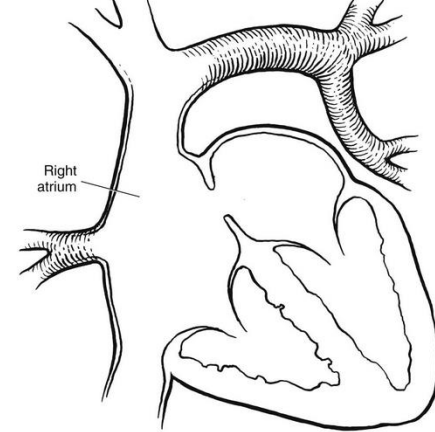


# Totally anomalous pulmonary venous connect

- Anatomy
- Infracardiac type



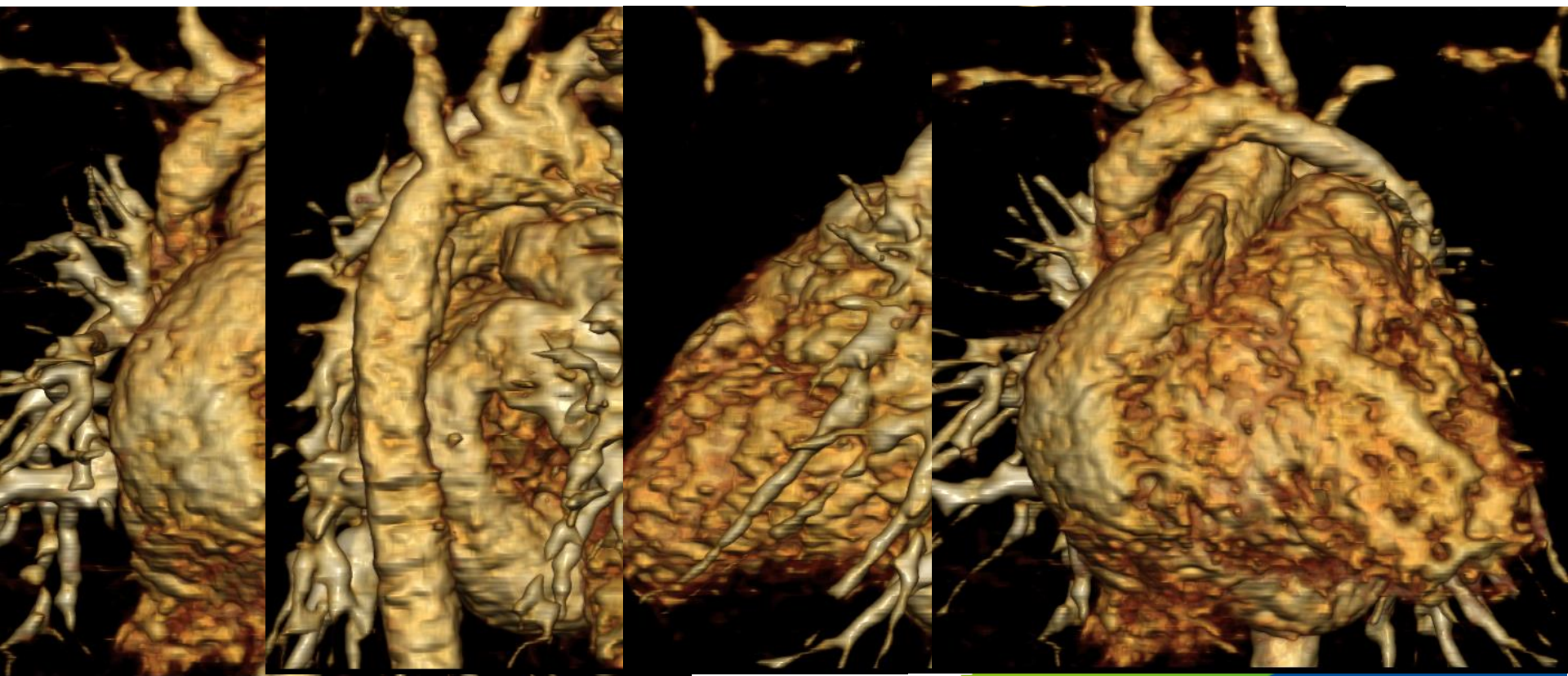
# Totally anomalous pulr venous connection



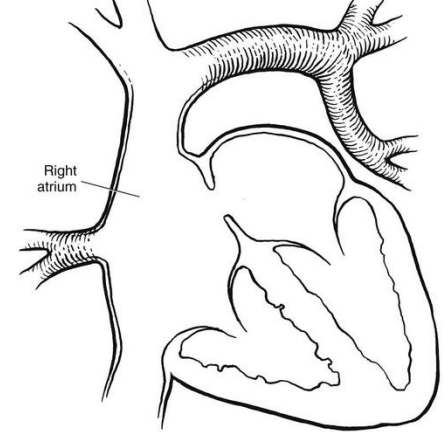
- Anatomy
- Mixed type (cardiac + supracardiac, LUPV)

D

Mixed type (5%-10%)



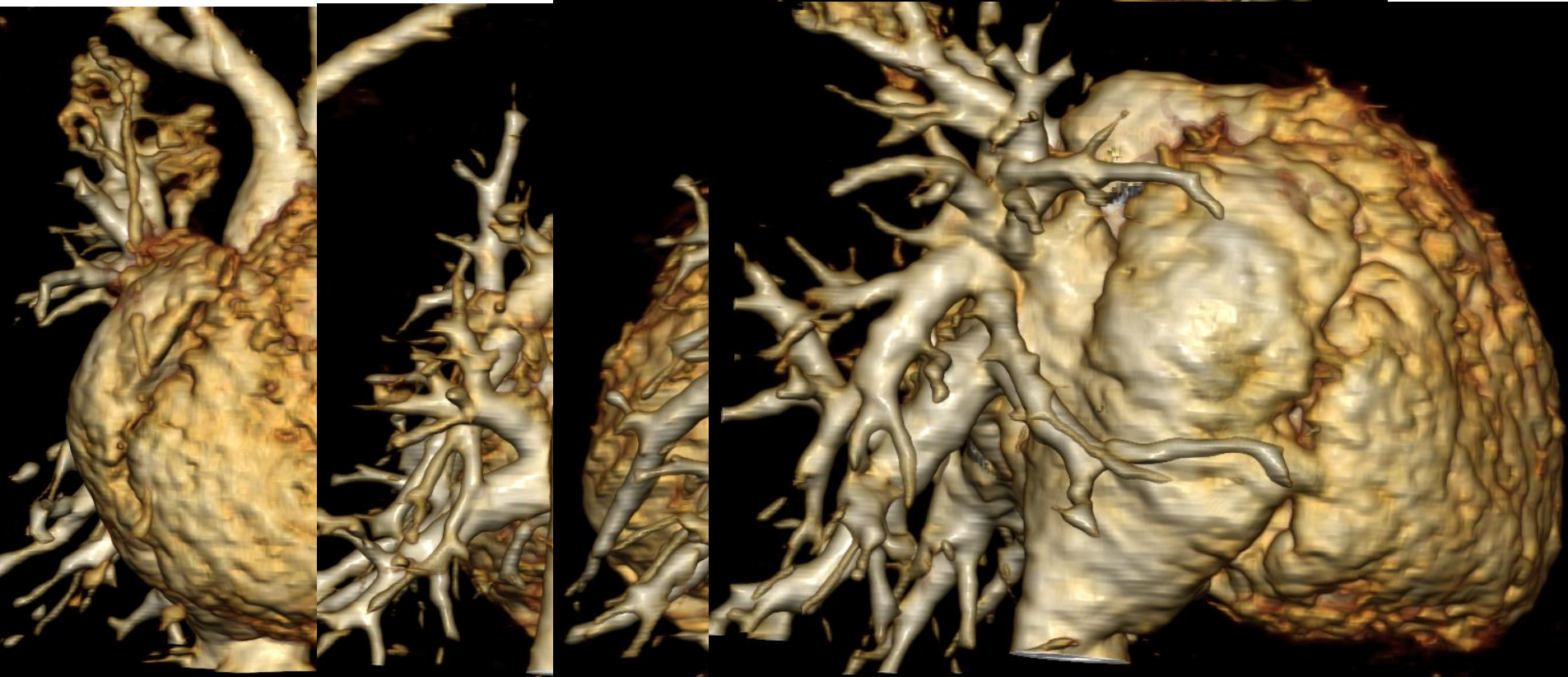
# Totally anomalous pulr venous connection



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

D

Mixed type (5%-10%)



# Totally anomalous pulmonary venous connection

- Morphogenesis
  - 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 (ASD)**
  - **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
    - **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 (supracardiac type)**

# Totally anomalous pulmonary venous connection

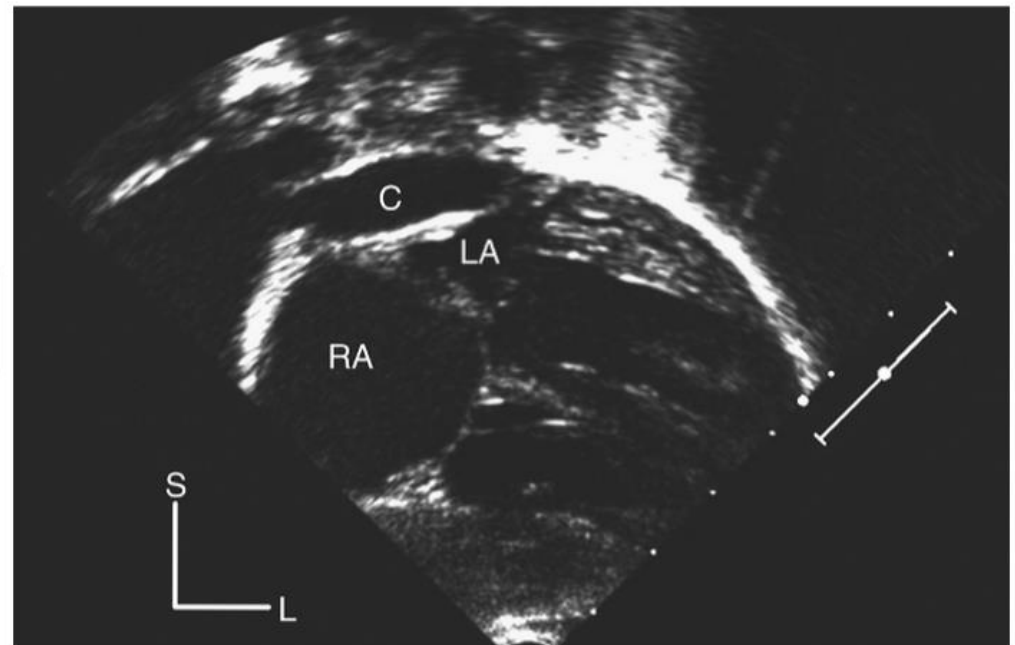
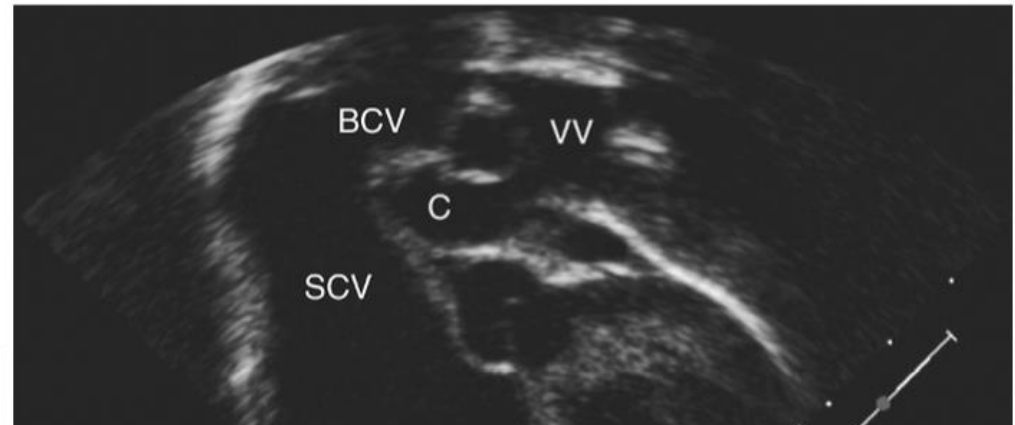
- Investigations
  - Electrocardiography
    - **right-axis deviation** with a clock wise frontal plane loop and **right ventricular hypertrophy**
    - Disturbances of conduction, rare
    - **Patients with pulmonary venous obstruction, who present younger, much less likely to have right atrial hypertrophy**

# Totally anomalous pulmonary venous connection

- Investigations
  - Echocardiography
    - definitive non-invasive method of diagnosis
    - Exclusive right-to-left shunting at the atrial level through ASD -> should be assumed that any patient with this finding has **totally anomalous pulmonary venous connection** until proven otherwise

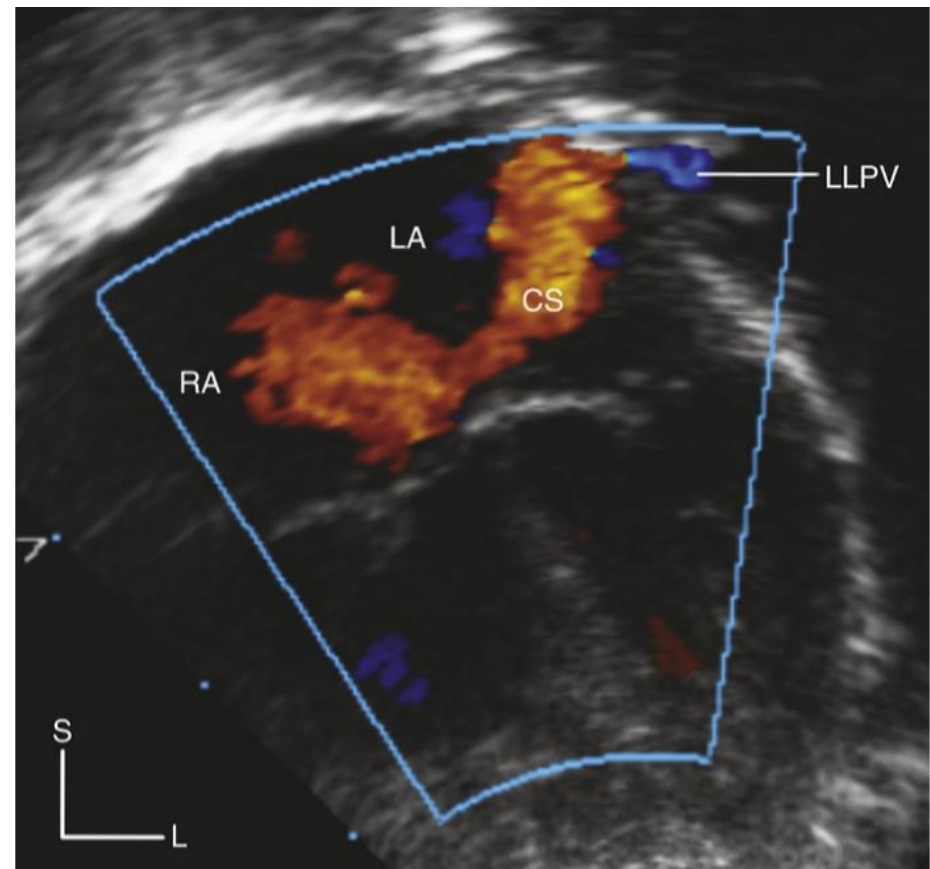
# Totally anomalous pulmonary venous connection

- Investigations
  - Echocardiography
    - Other clues
      - small left atrium
      - dilated SVC
      - non-pulsatile caudally-directed flow seen below the level of the heart in subcostal imaging
      - non-pulsatile cranially-directed flow seen above the level of the heart



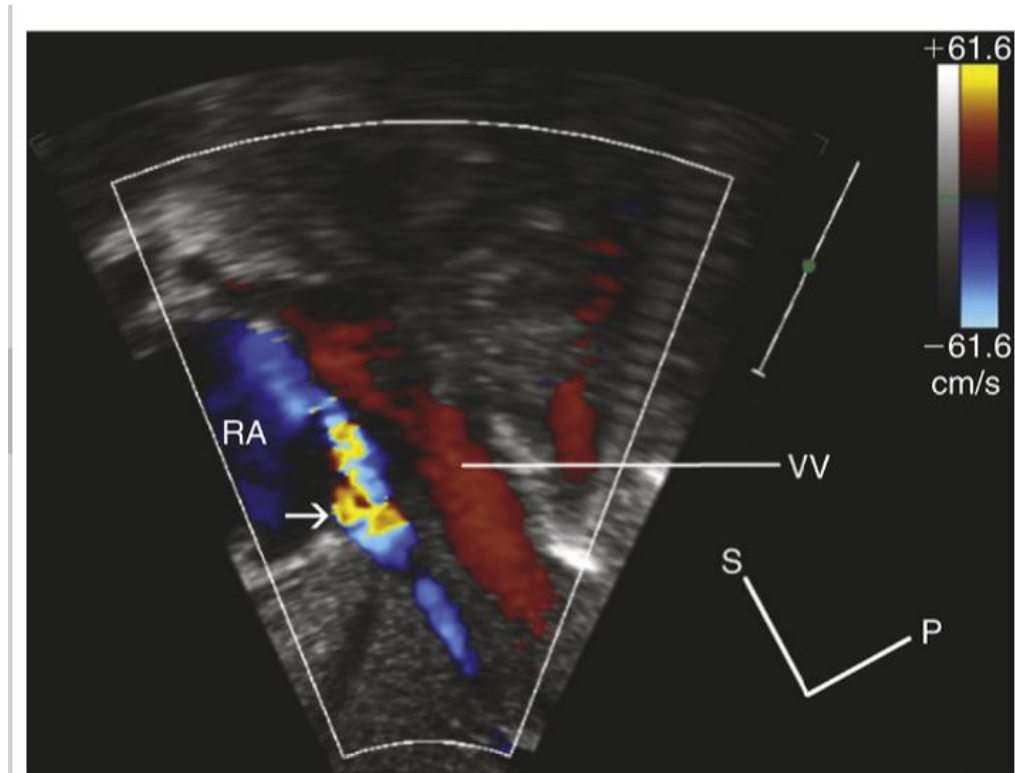
# Totally anomalous pulmonary venous connection

- 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**
    - **the pattern in which the pulmonary veins drain into it**



# 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



# Totally anomalous pulmonary venous connection

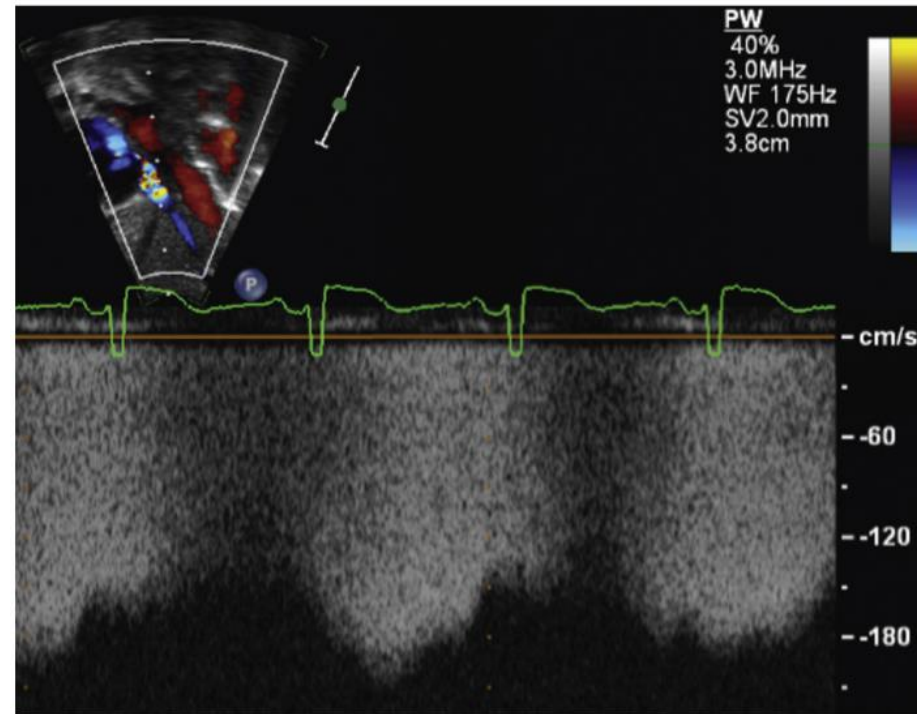
- Investigations
  - Echocardiography
    - **Anomalous connection to the right atrium** can be diagnosed if there is no ascending or descending vein, the coronary sinus is of normal size and the pulmonary veins can be followed to their site of entry to the right atrium

# 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

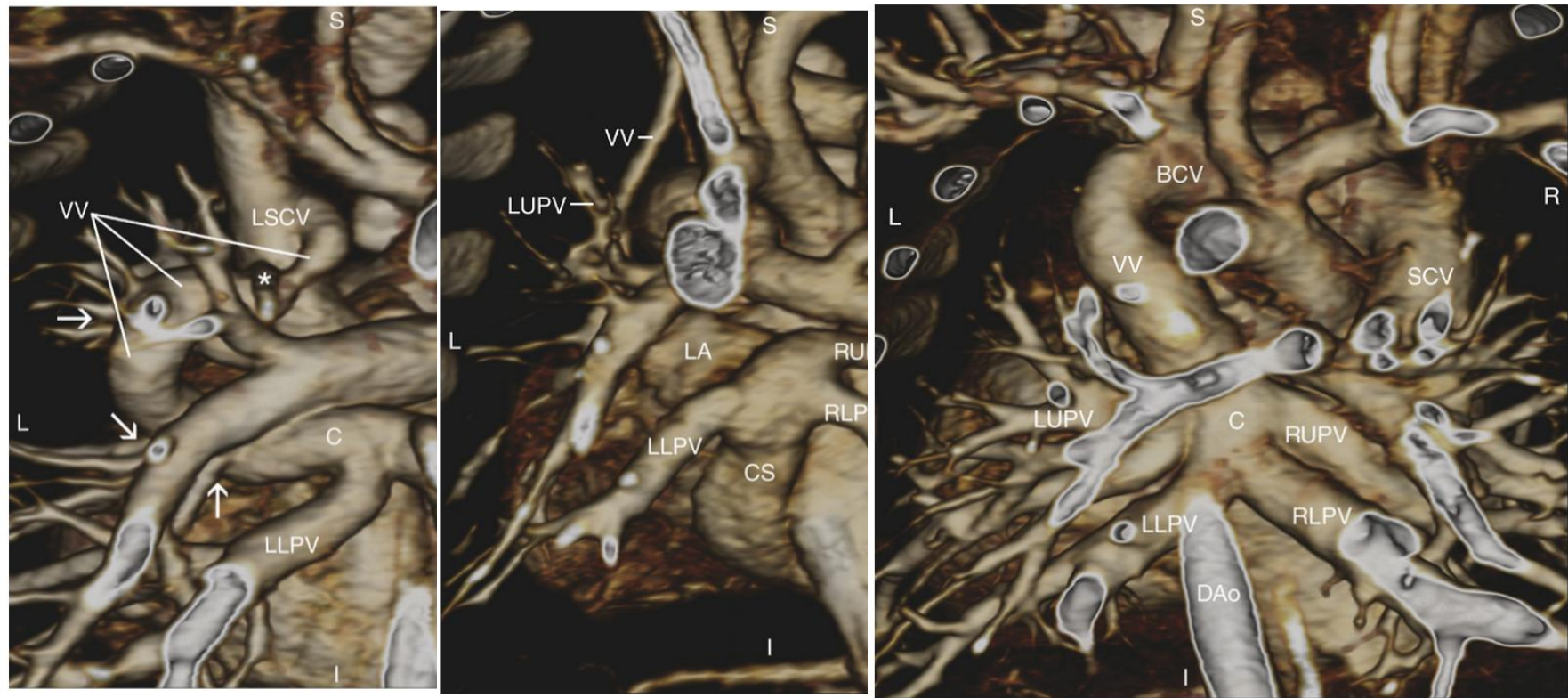
- Investigations
- **Cardiac catheterisation**
  - The pulmonary venous anatomy can almost always be delineated non-invasively, and **the clinical scenario of pulmonary venous obstruction can almost always be determined without invasive testing**
  - Pulmonary venous connections with a particularly tortuous 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

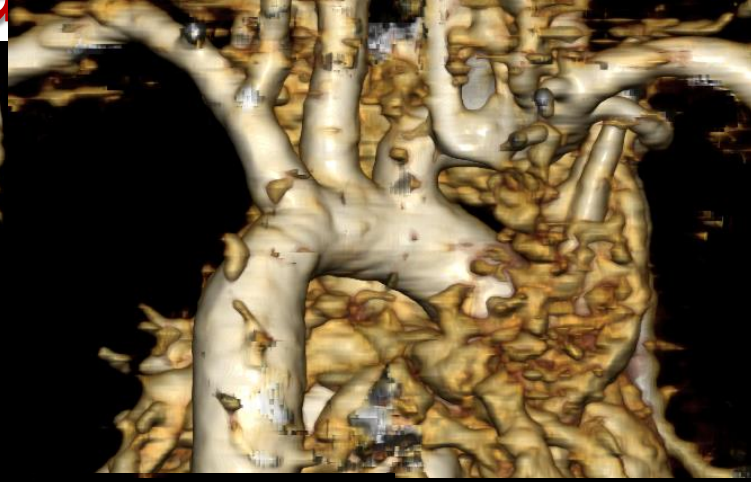
- Investigations
  - **Computed tomographic angiography**
    - Technological advances in medical imaging have **increased the utilization** of computed tomographic angiography in the evaluation of patients with anomalous pulmonary venous connection

# Totally anomalous pulmonary venous connection

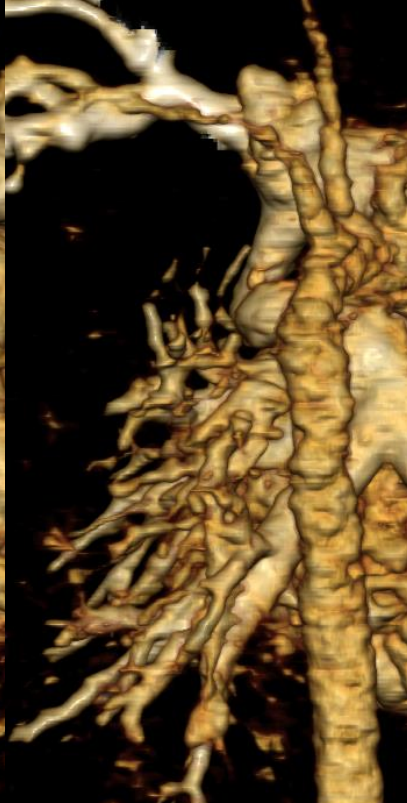
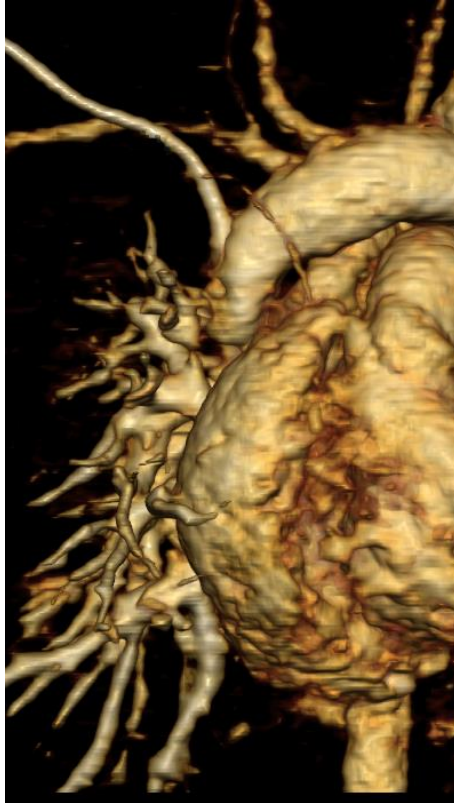
- Investigations
  - Computed tomographic angiography



# Totally anomalous pulmonary



return



# 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

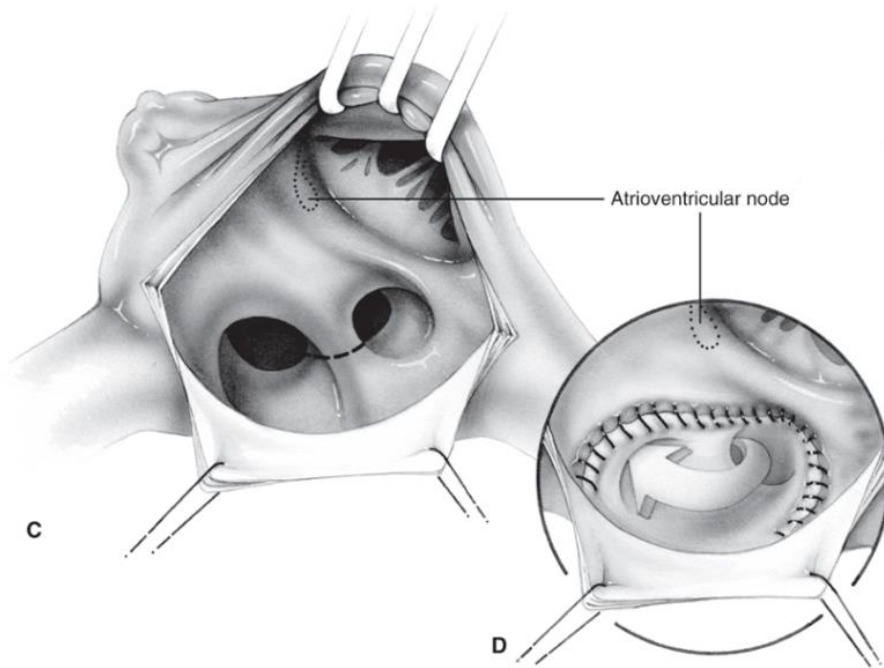
- Management
  - **Medical treatment**
    - In the current era, medical management consists solely of **supportive measures** in preparation for surgical management

# 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** (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**

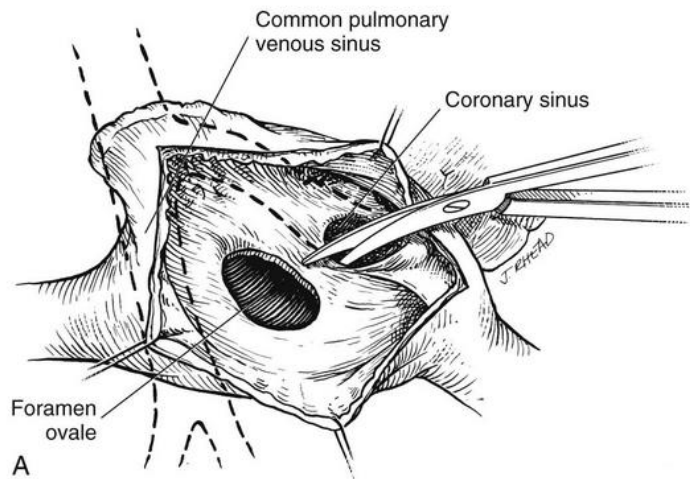
# Totally anomalous pulmonary venous connection

- Management
  - Surgery
    - Intracardiac type

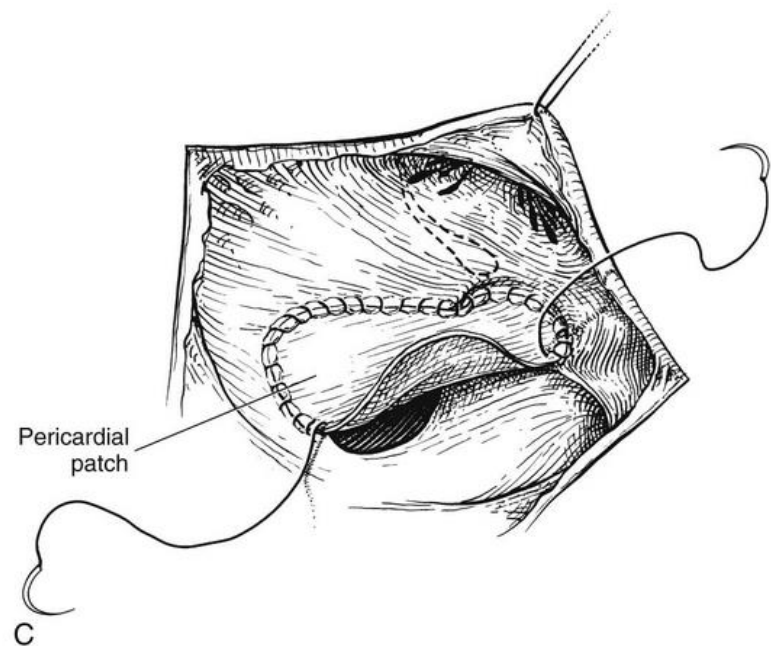
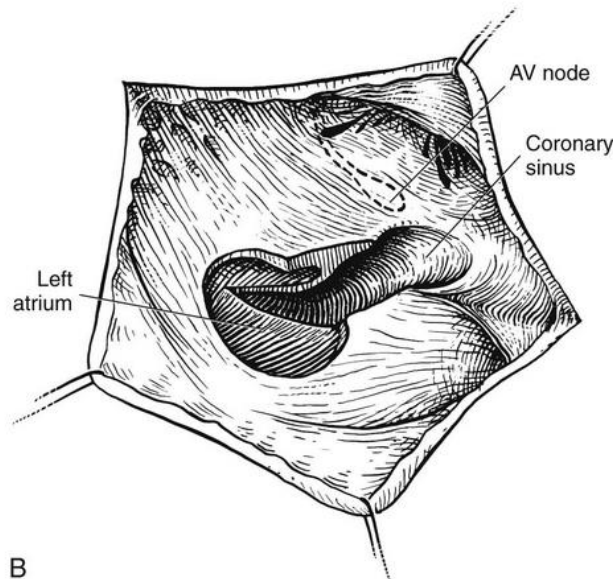


- . Whenever the **common pulmonary vein returns to the coronary sinus**, its orifice is **extended superiorly** to reach the atrial septal defect
- . This incision must be **well away from the anterior margin of the coronary sinus** to prevent damage to the **atrioventricular node and the conduction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

# Totally anomalous pulmonary venous connection

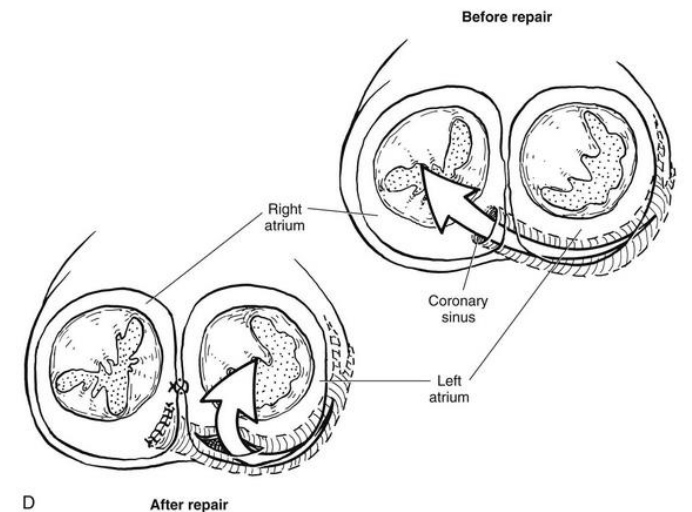
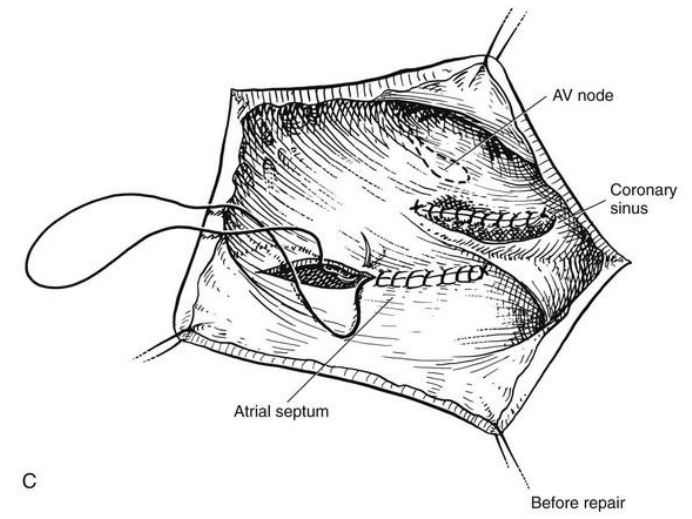
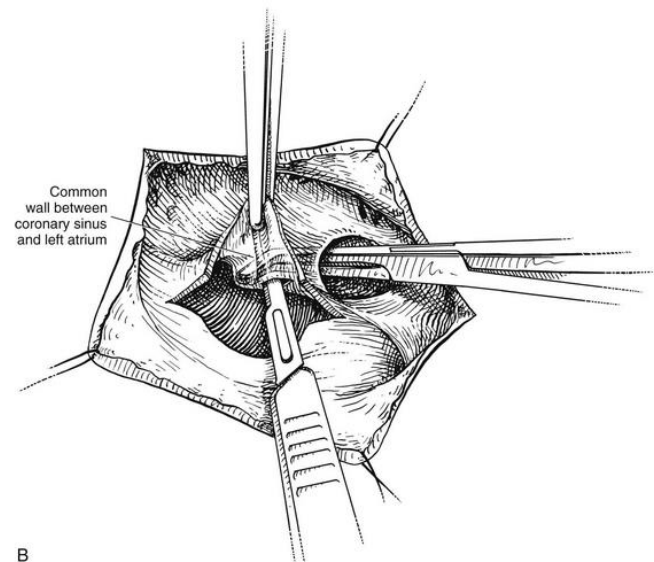
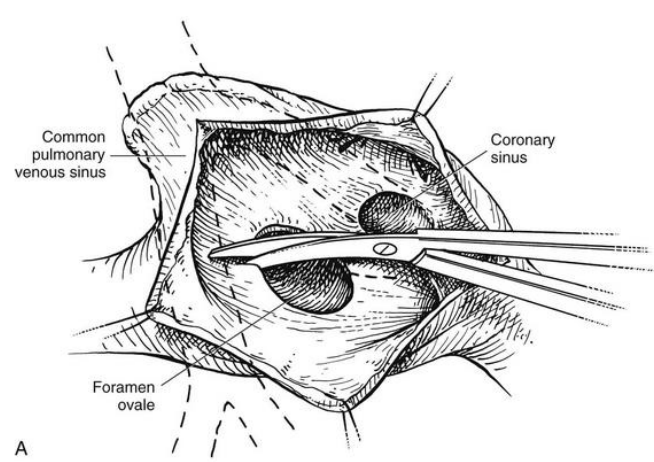


- Management
  - Surgery
    - Intracardiac type



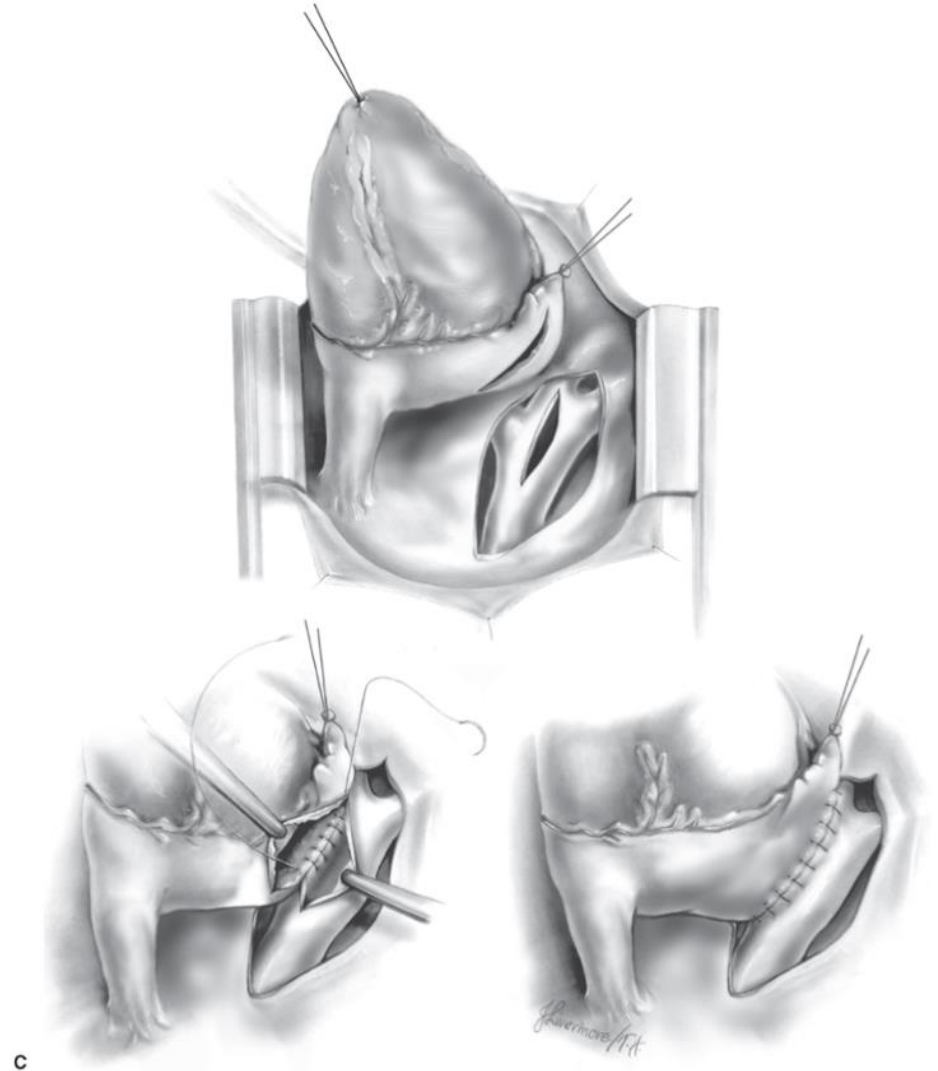
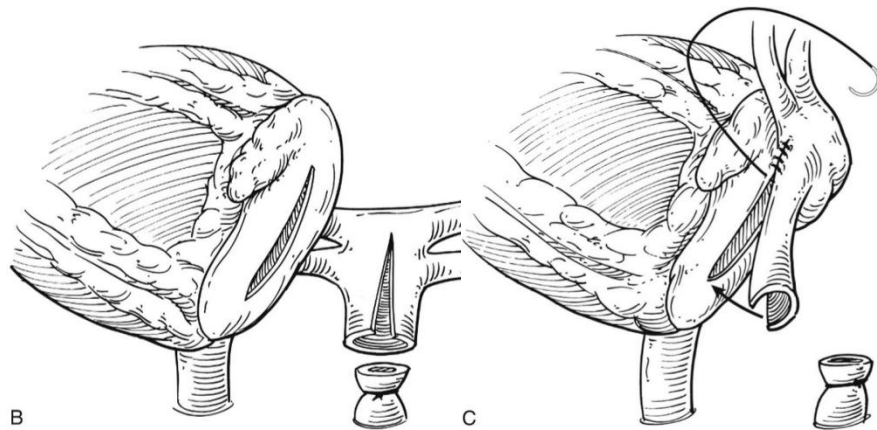
# Totally anomalous pulmonary venous connection

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



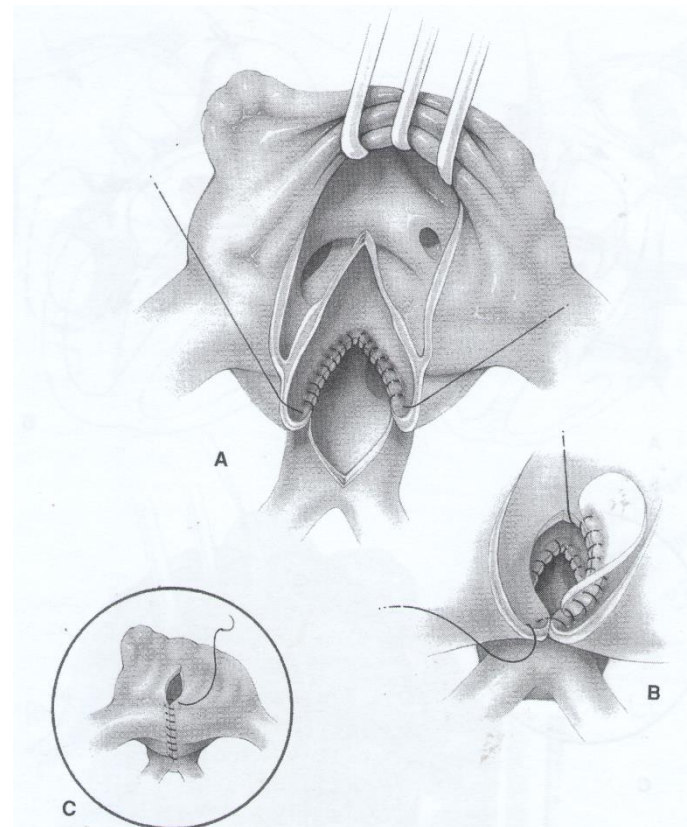
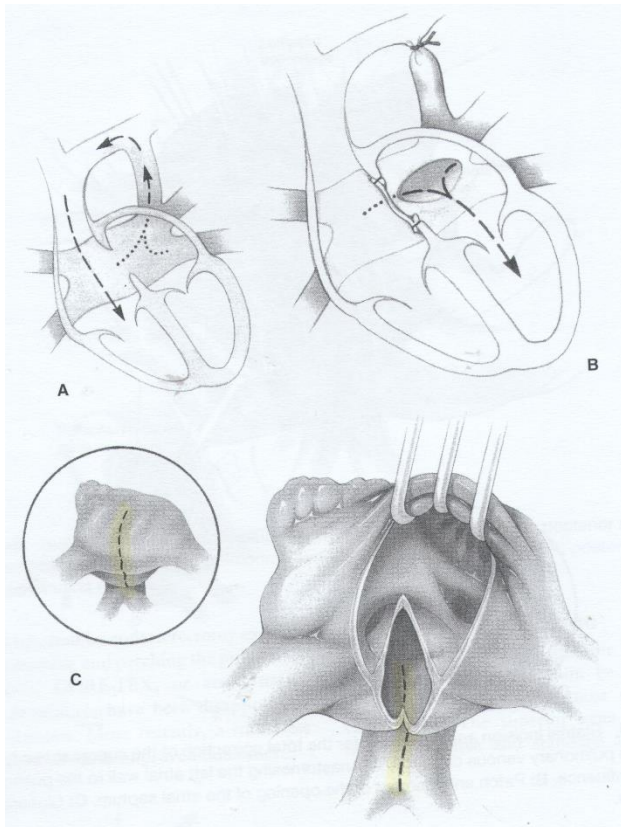
# Totally anomalous pulmonary venous connection

- Management
  - Surgery
    - Infracardiac type



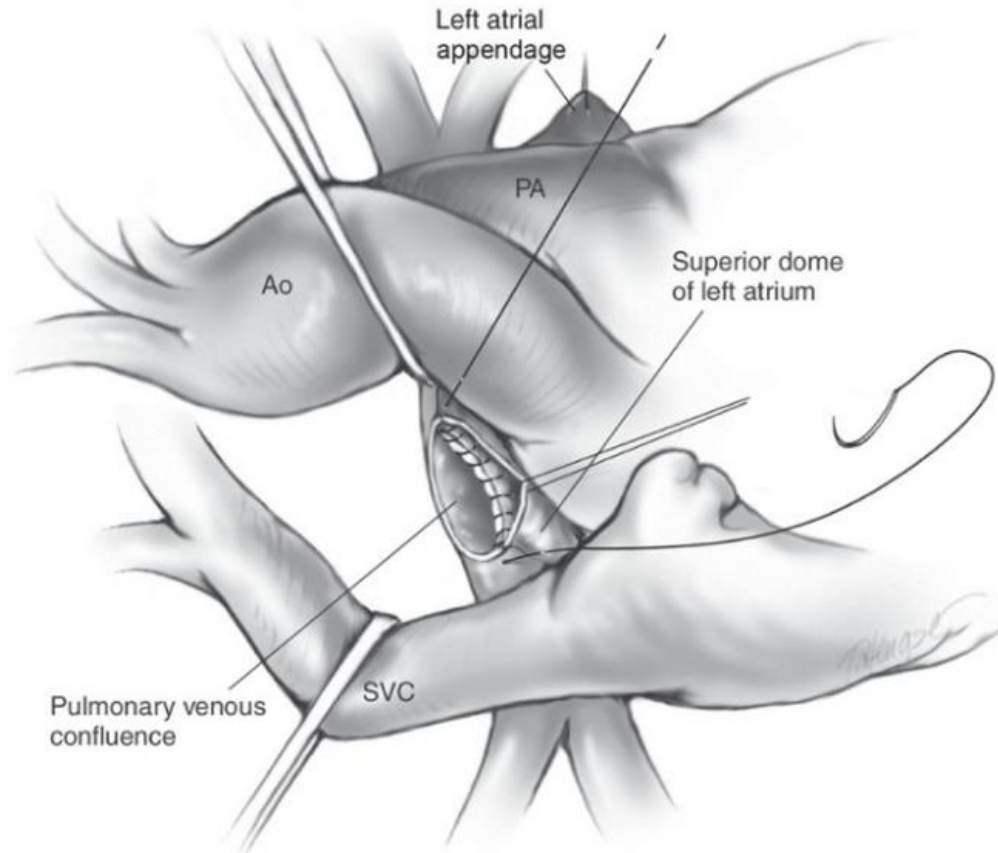
# Totally anomalous pulmonary venous connection

- Management
  - Surgery
    - Supracardiac type (biatrial incision)



# Totally anomalous pulmonary venous connection

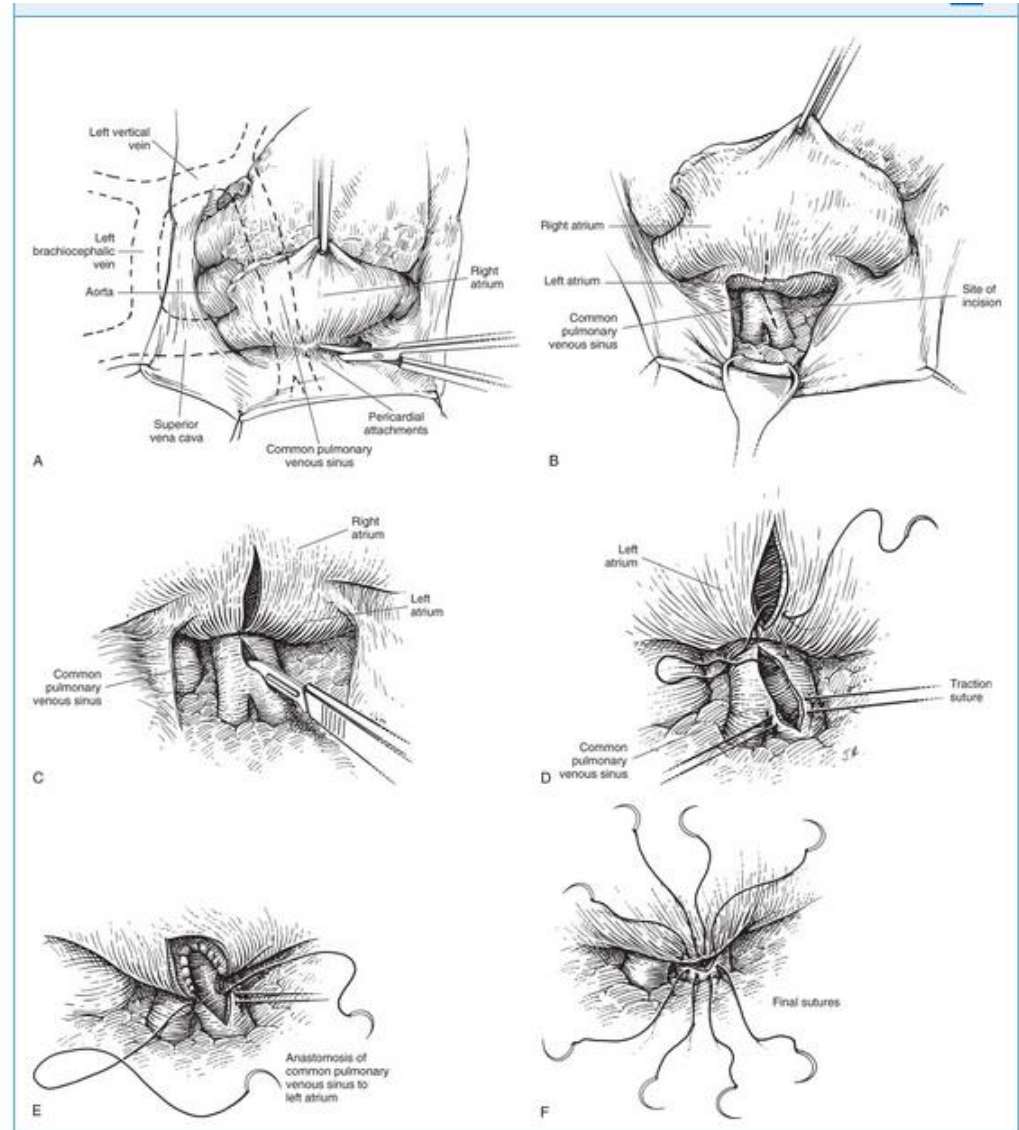
- Management
  - Surgery
    - Supracardiac type (superior approach)





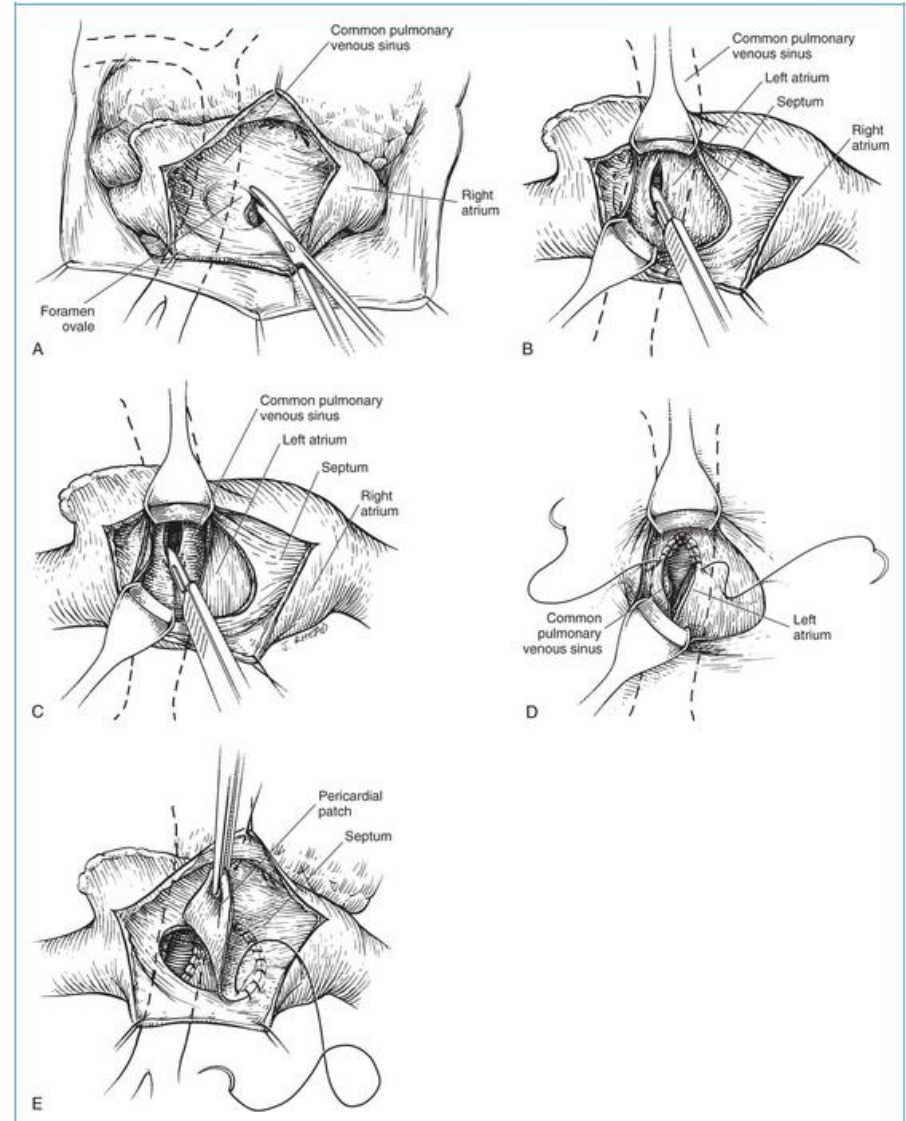
# Totally anomalous pulmonary venous connection

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

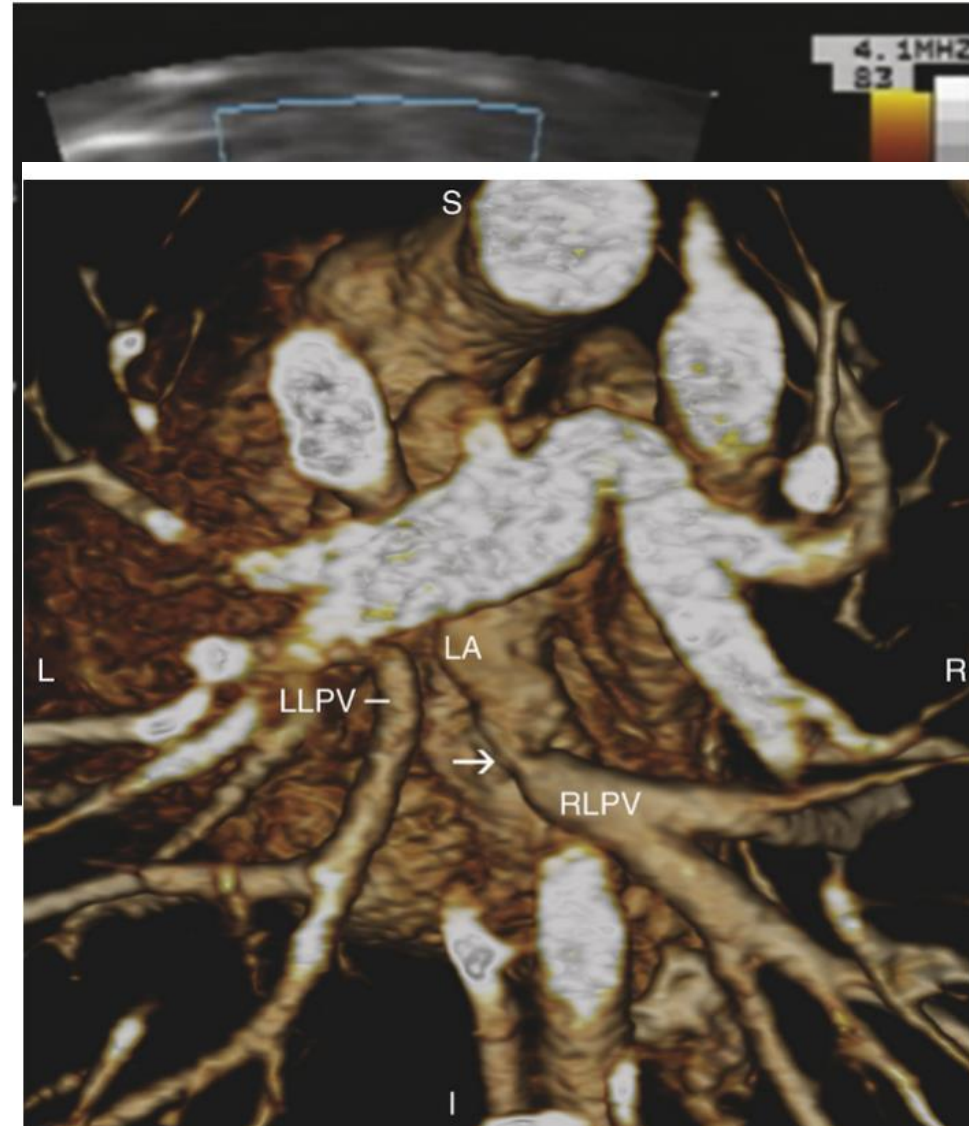
- Results of surgery
  - The **late results of repair** are, in general, **excellent**
  - Nonetheless, **late pulmonary venous obstruction** is not uncommon
  - It occurs in **about 10%** of all large surgical series
  - Some authors have found it to be **more common in patients with infracardiac or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



# Totally anomalous pulmonary venous connection

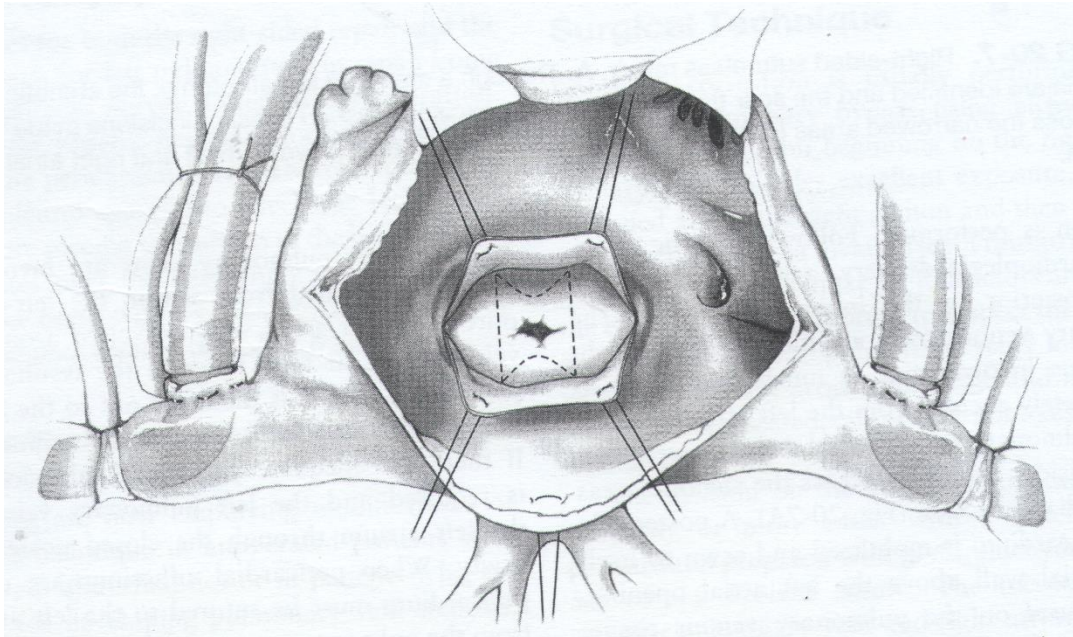
- Results of surgery
  - Postoperative PV stenosis





# Totally anomalous pulmonary venous connection

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

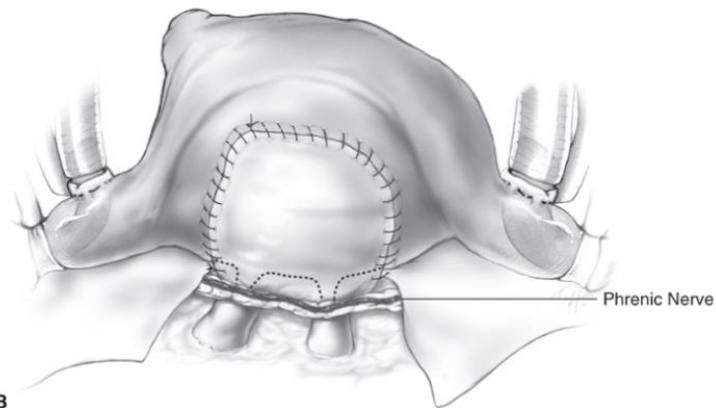
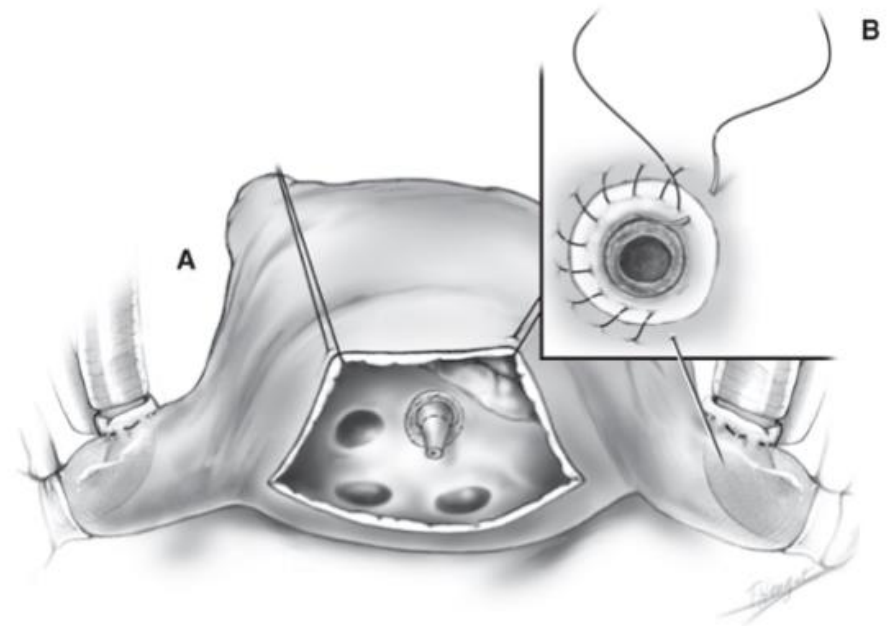
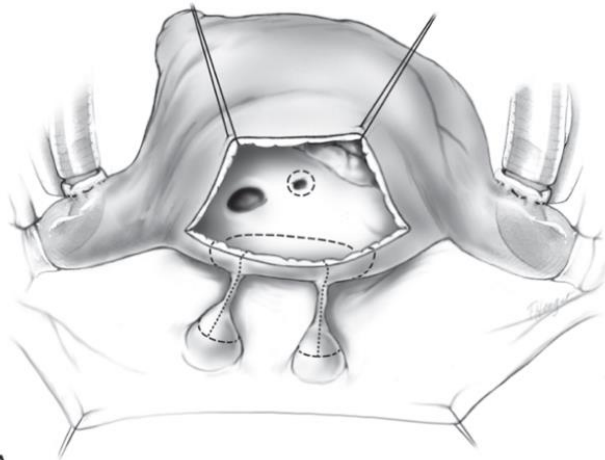


. An isolated anastomotic stenosis is approached through a right atriotomy and vertical incision on the atrial septum

. 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

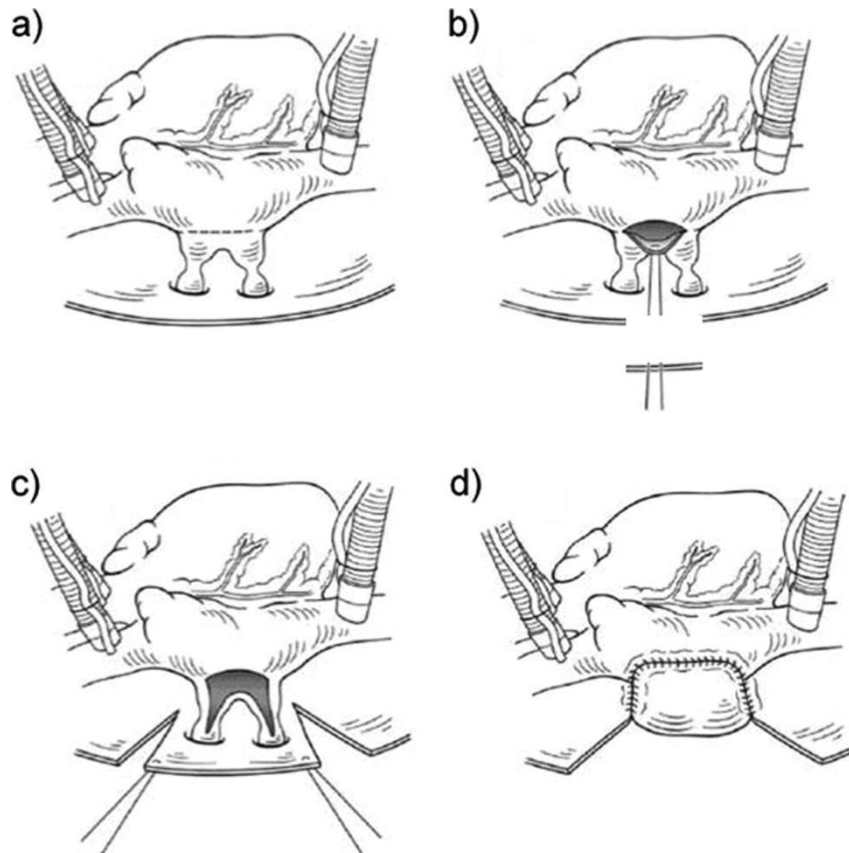
- Results of surgery
- Pulmonary venous stenosis after surgery
  - **Sutureless technique**



- . 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

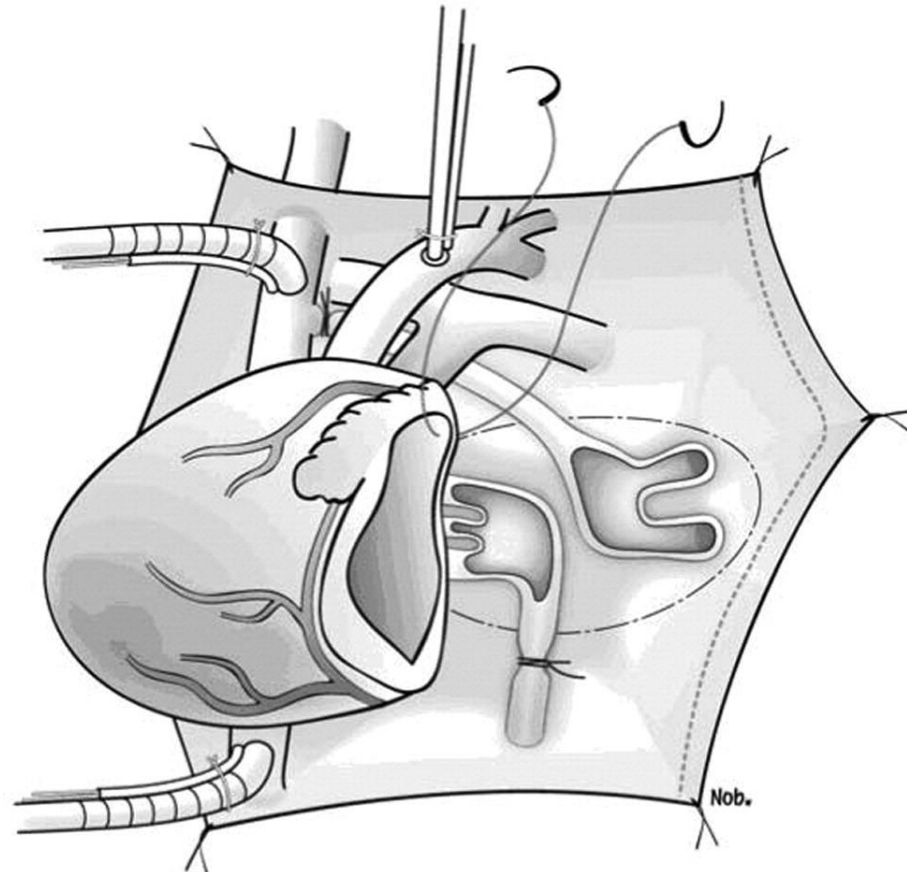
# Totally anomalous pulmonary venous connection

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

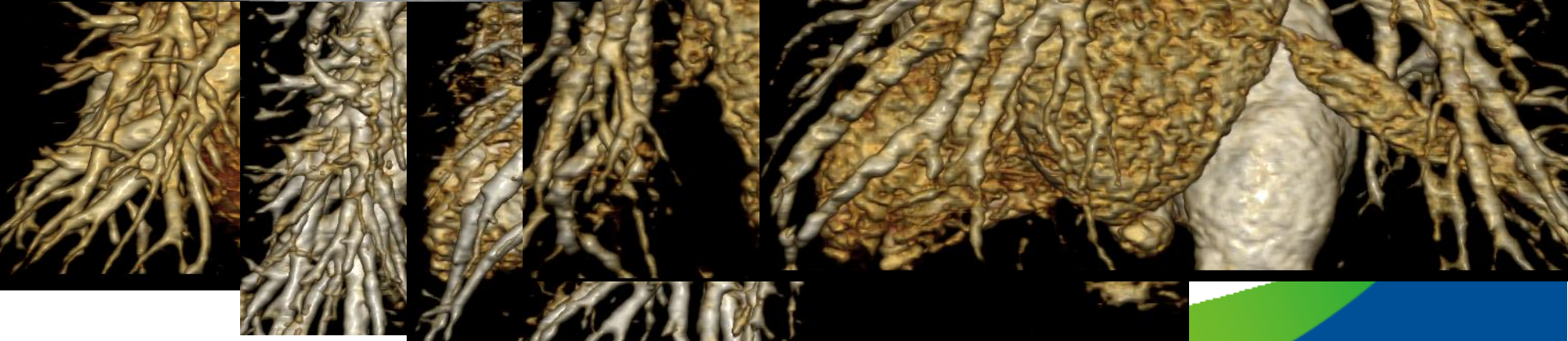
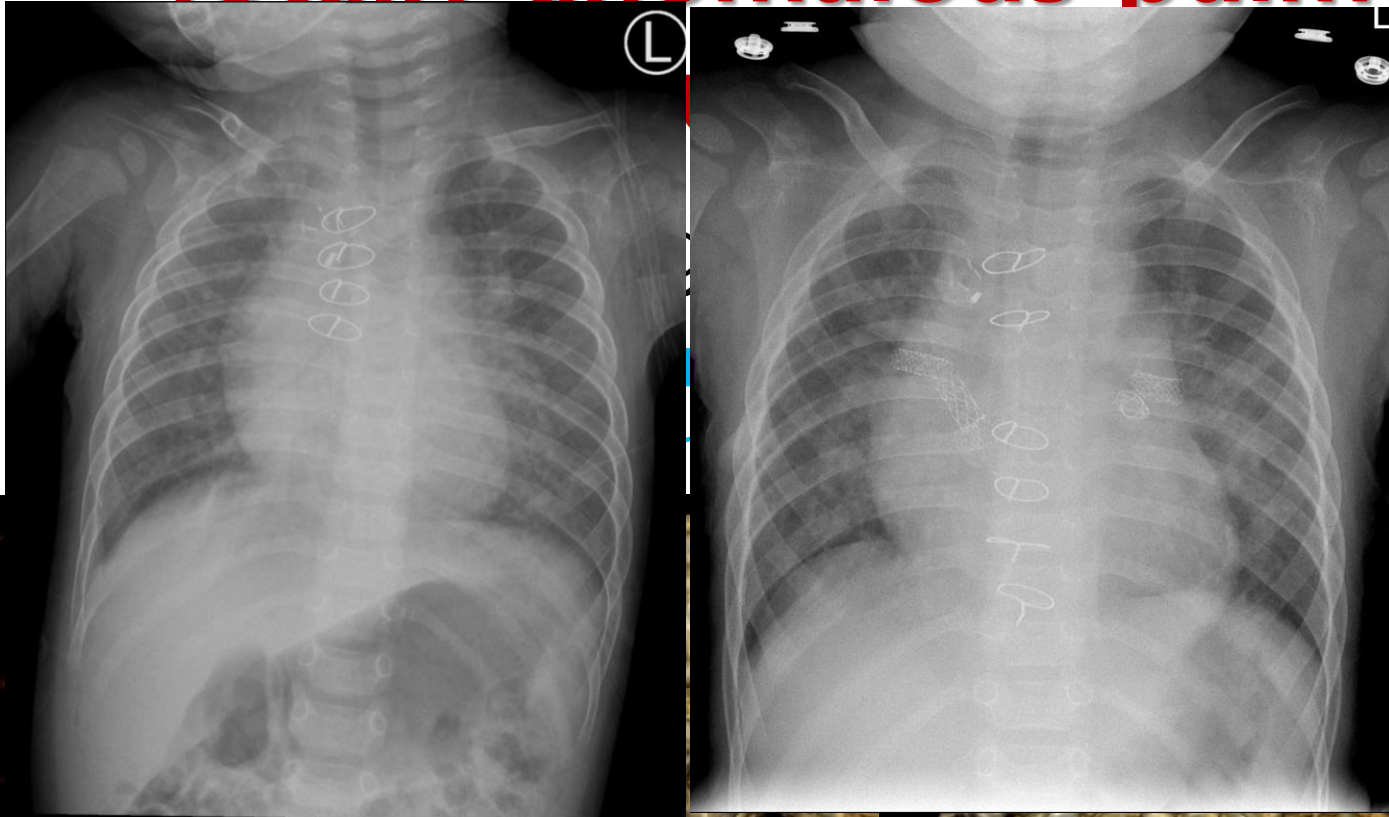


# Totally anomalous pulmonary venous connection

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



# Totally anomalous pulmonary

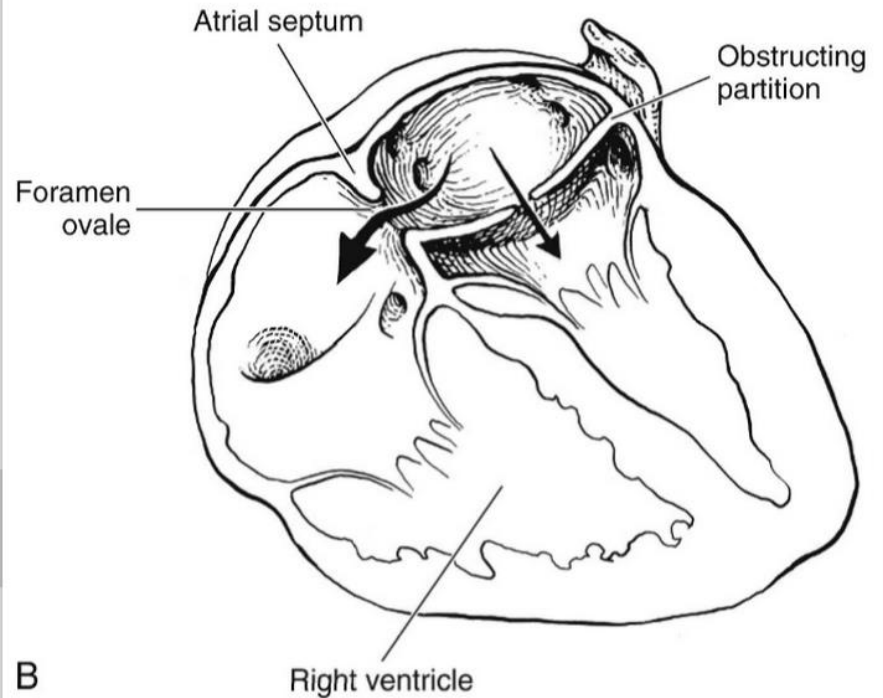
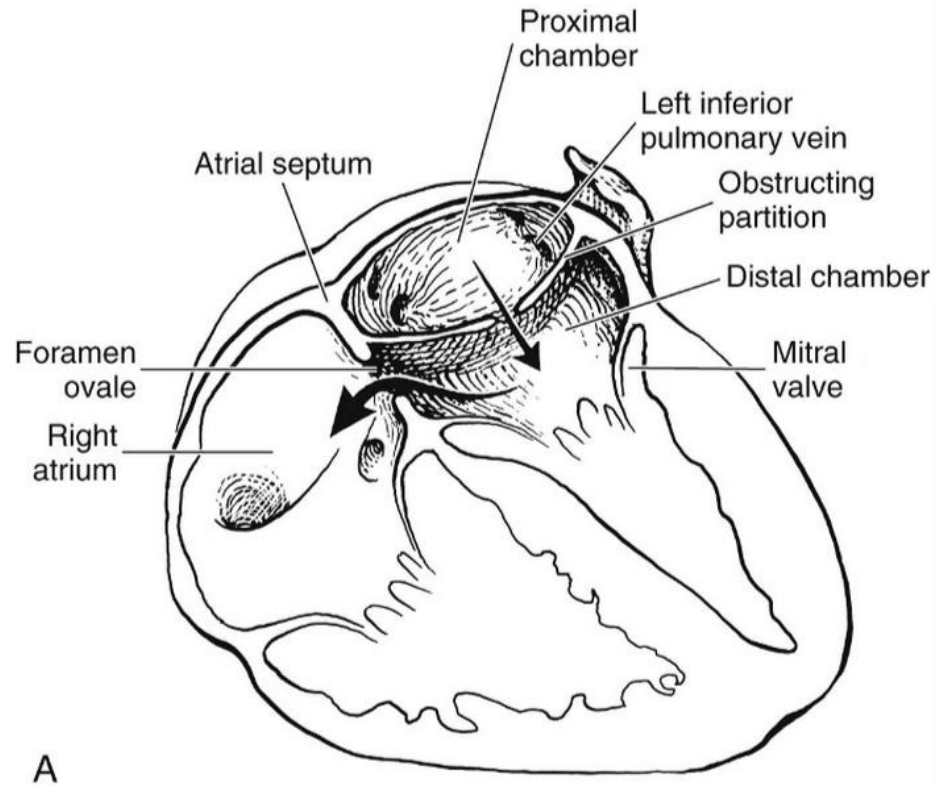


# Totally anomalous pulmonary venous connection

- 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



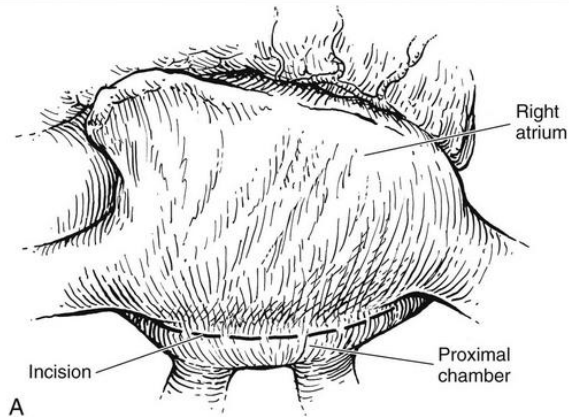
# Cor Triatriatum

- Morphology
  - Typically, the **proximal (common pulmonary venous) chamber** is somewhat **larger** than the distal (left atrial) chamber
  - **Proximal chamber** contains **all pulmonary vein** connections and is usually **thick-walled**, whereas **distal chamber** always contains **left atrial appendage**, leads into the **mitral valve**, and is **thin-walled**

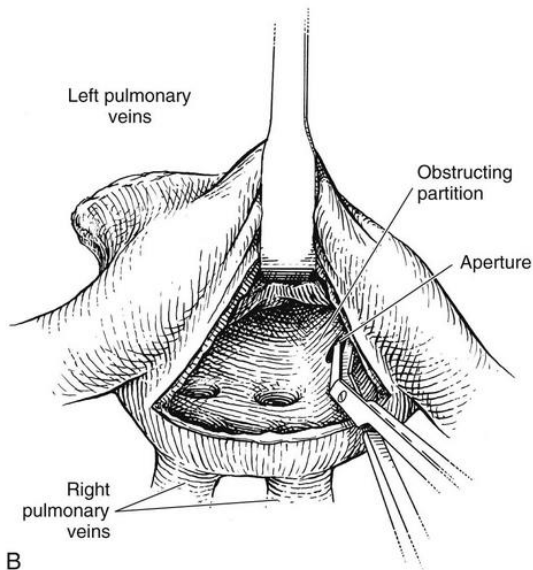


# Cor Triatriatum

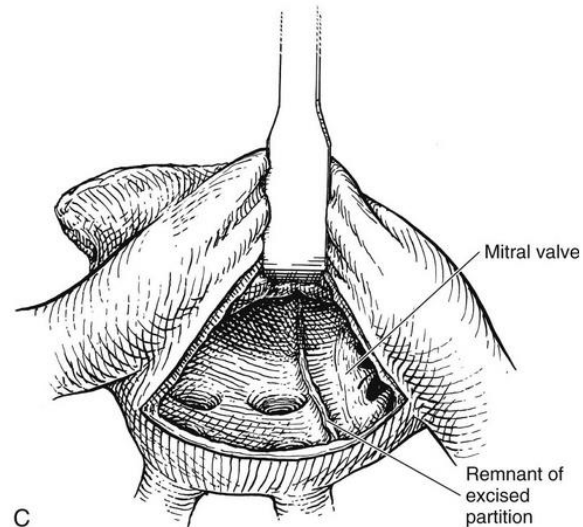
- Technique of operation (left-side approach)



A



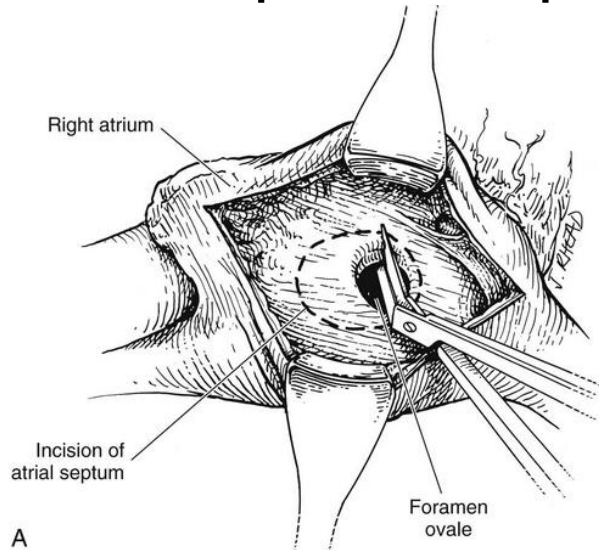
B



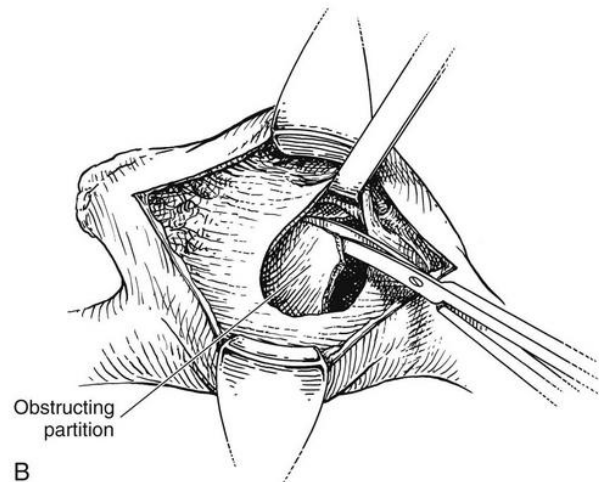
C

# Cor Triatriatum

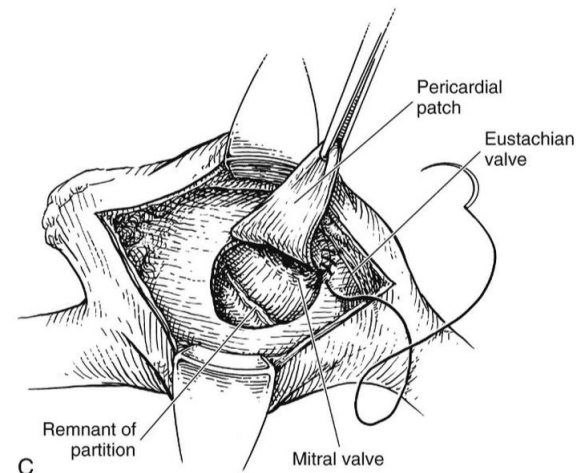
- Technique of operation (right-side approach)



A



B



C

# Partially anomalous pulmonary venous connection

- Anatomy
  - 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

# Patially anomalous pulmonary venous connection

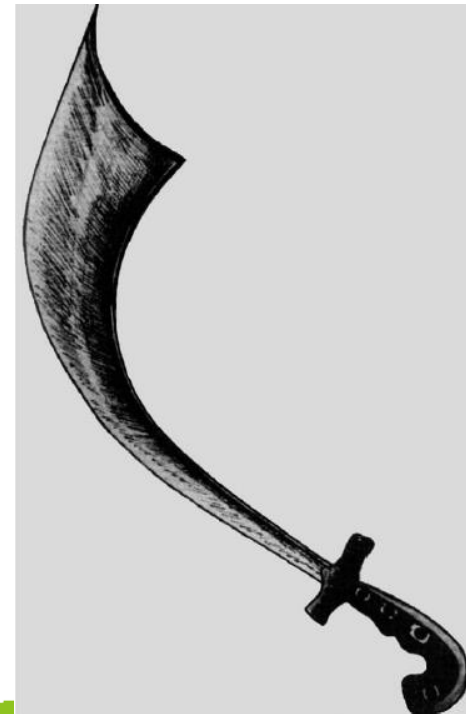
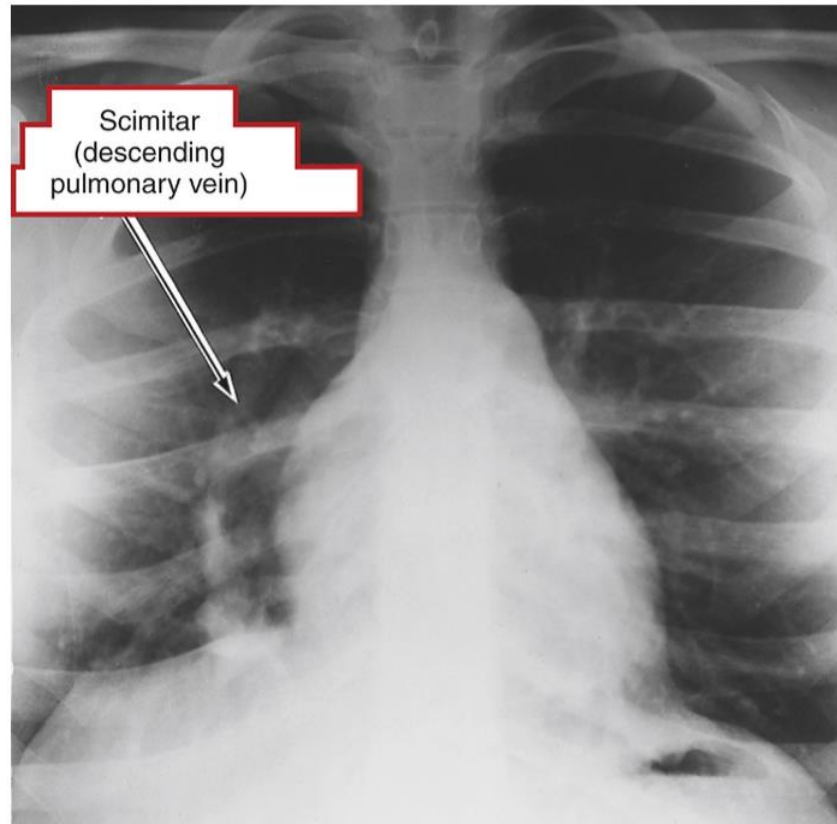
- Pathophysiology
  - Because **pulmonary venous obstruction is rarely present** with partially anomalous venous connections, the haemodynamic effects are almost always the result of an obligatory left-to-right shunt through the anomalously connected segments of lung
  - **Pulmonary hypertension is rare,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

# Patially 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

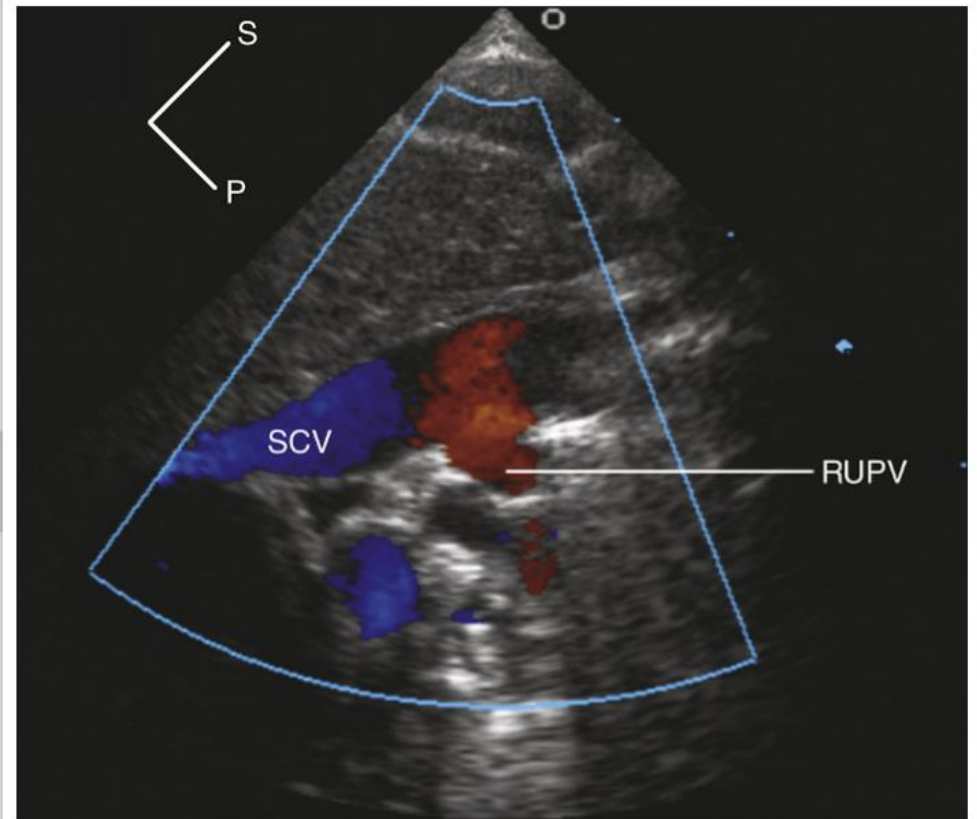
# Partially anomalous pulmonary venous connection

- Investigations
  - Chest radiography



# Partially anomalous pulmonary venous connection

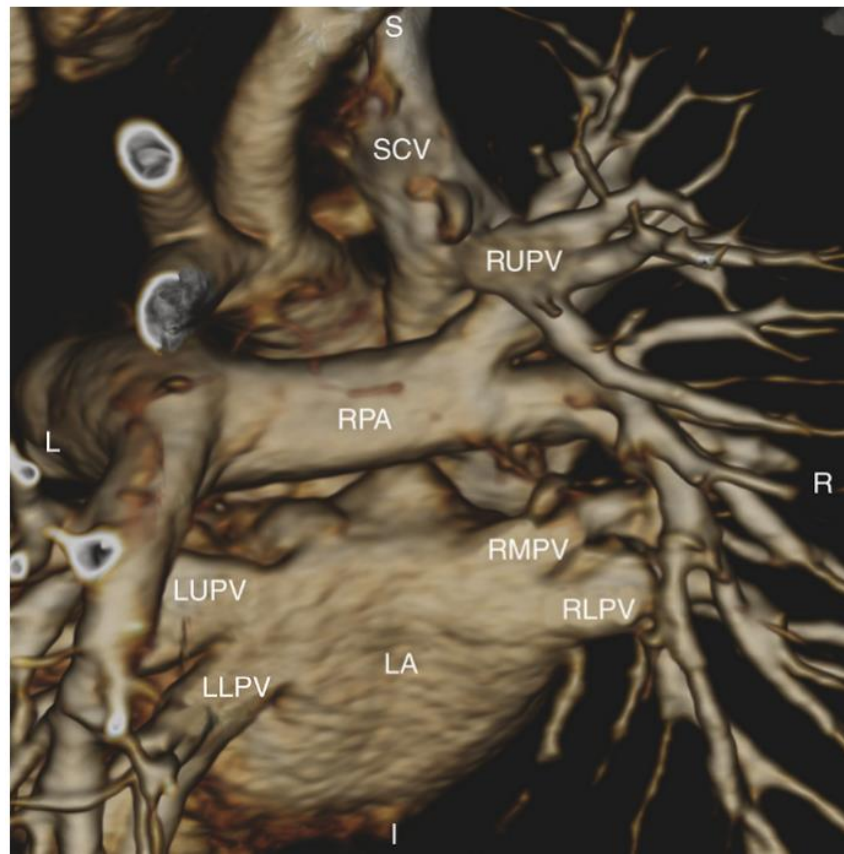
- Investigations
  - Echocardiography
    - **Increased or atypical flow in the superior or inferior caval veins** should alert one to the possibility of partially anomalous pulmonary venous connection





# Partially anomalous pulmonary venous connection

- Investigations
  - Computed tomographic angiography



# Partially anomalous pulmonary venous connection

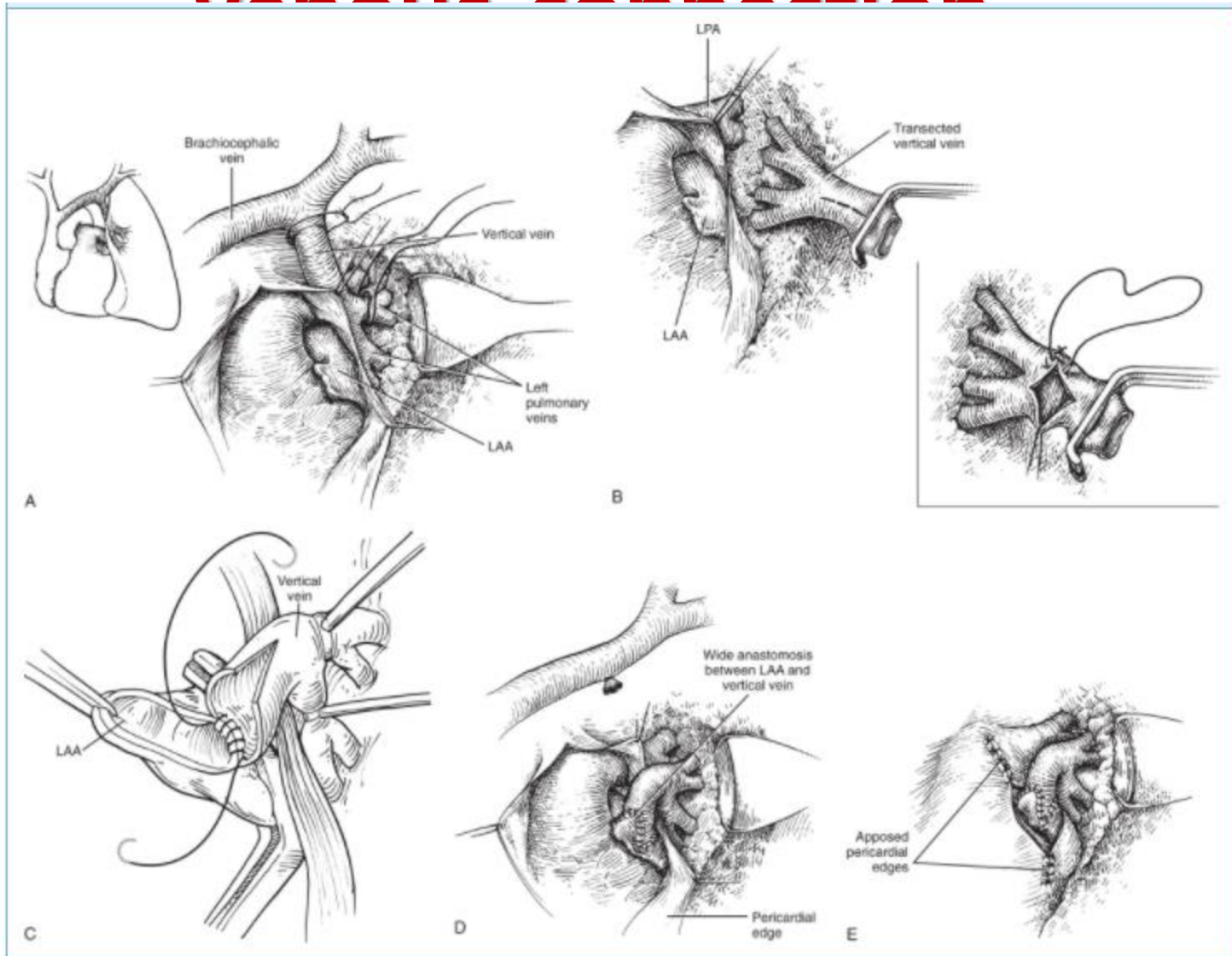
- Investigations
  - Cardiac catheterisation and angiocardiology
    - **Cardiac catheterisation is rarely indicated**
    - **a step-up in oxygen saturation** in the superior caval vein, the brachiocephalic vein, or the inferior caval vein is **suggestive of anomalous pulmonary veins draining into the respective site**
    - In the case of the **scimitar syndrome**, **selective injection of the systemic arterial supply to the lungs** 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

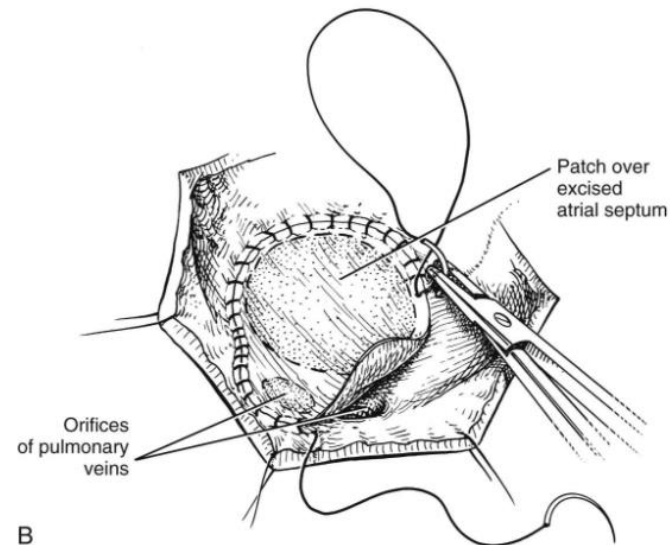
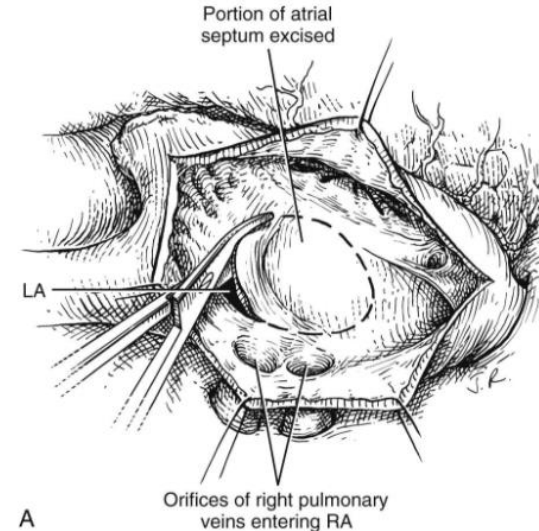
# Partially anomalous pulmonary venous connection

- Mar - S



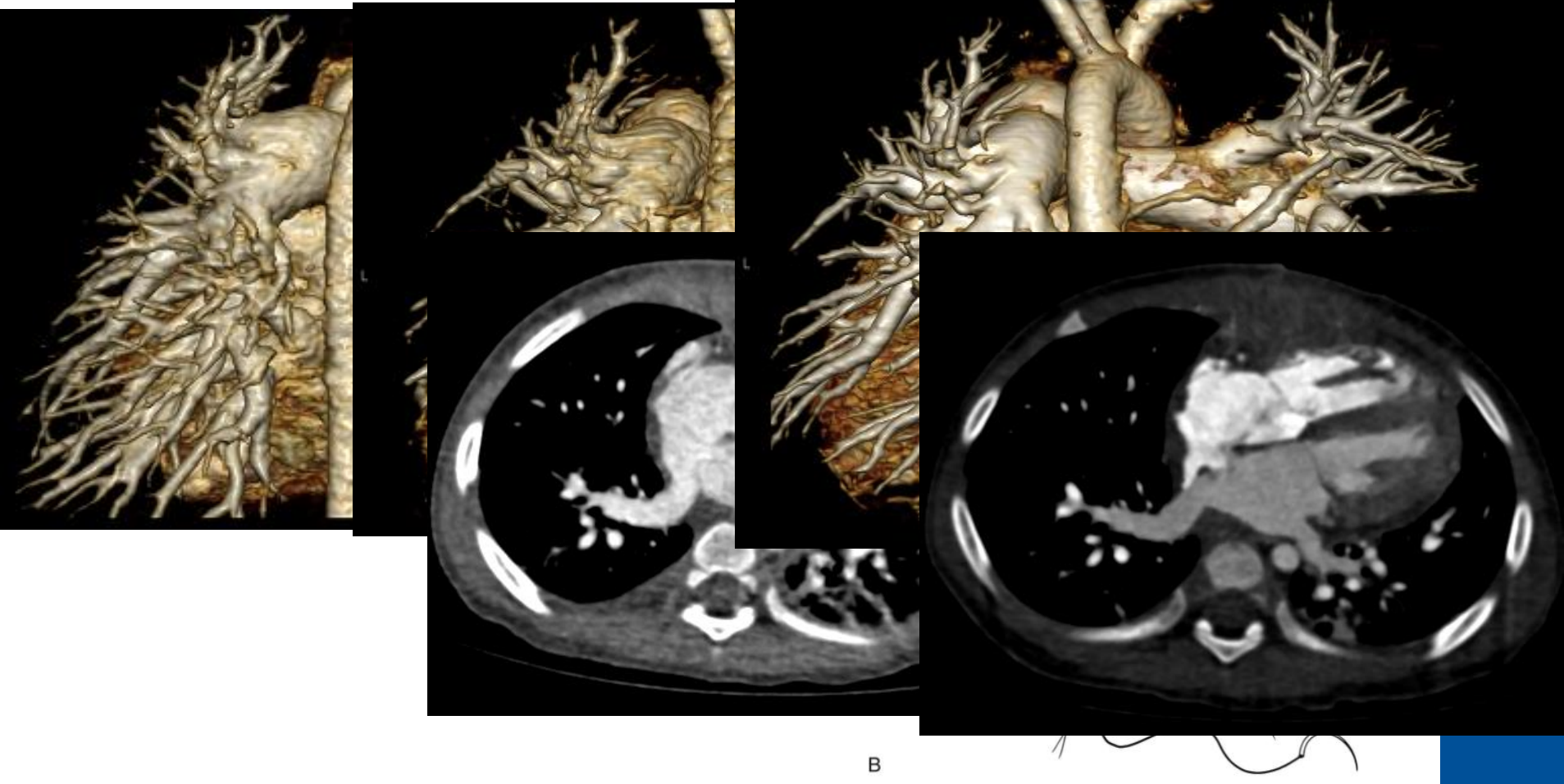
# Partially anomalous pulmonary venous connection

- Management
  - surgery

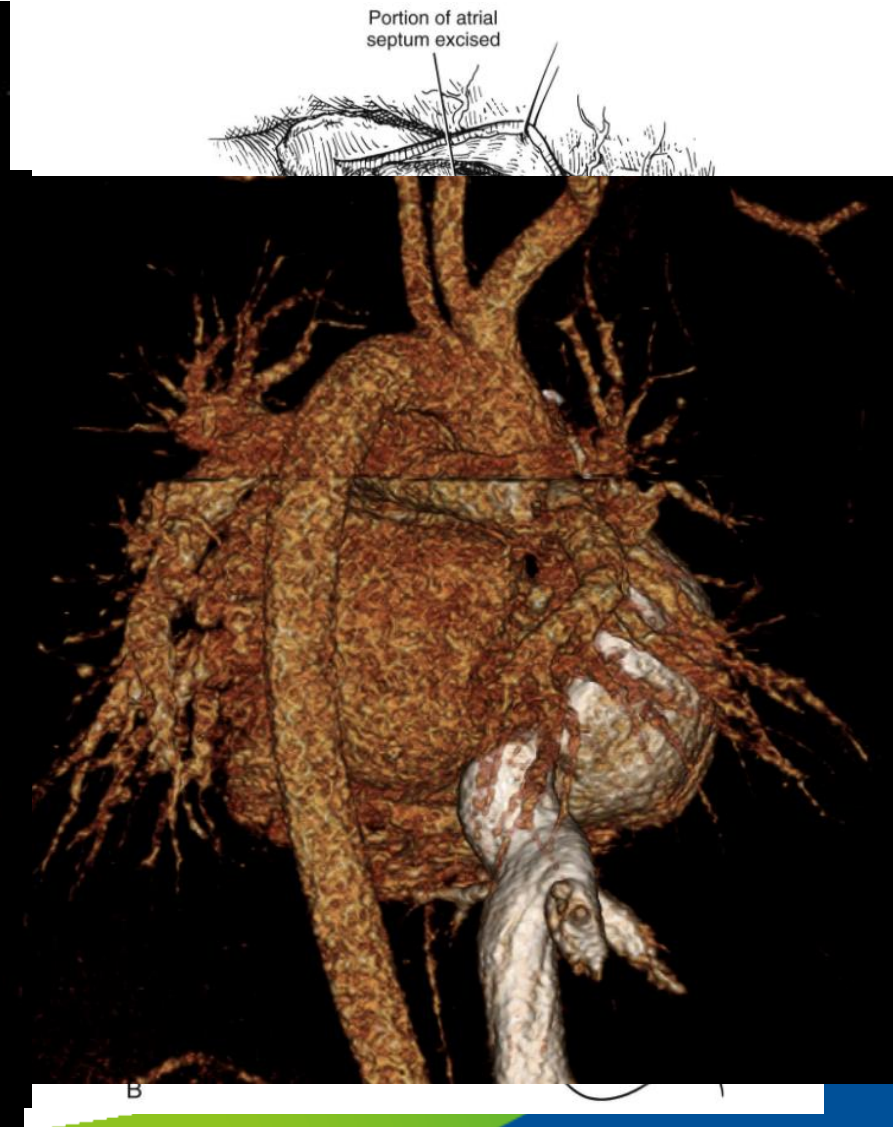
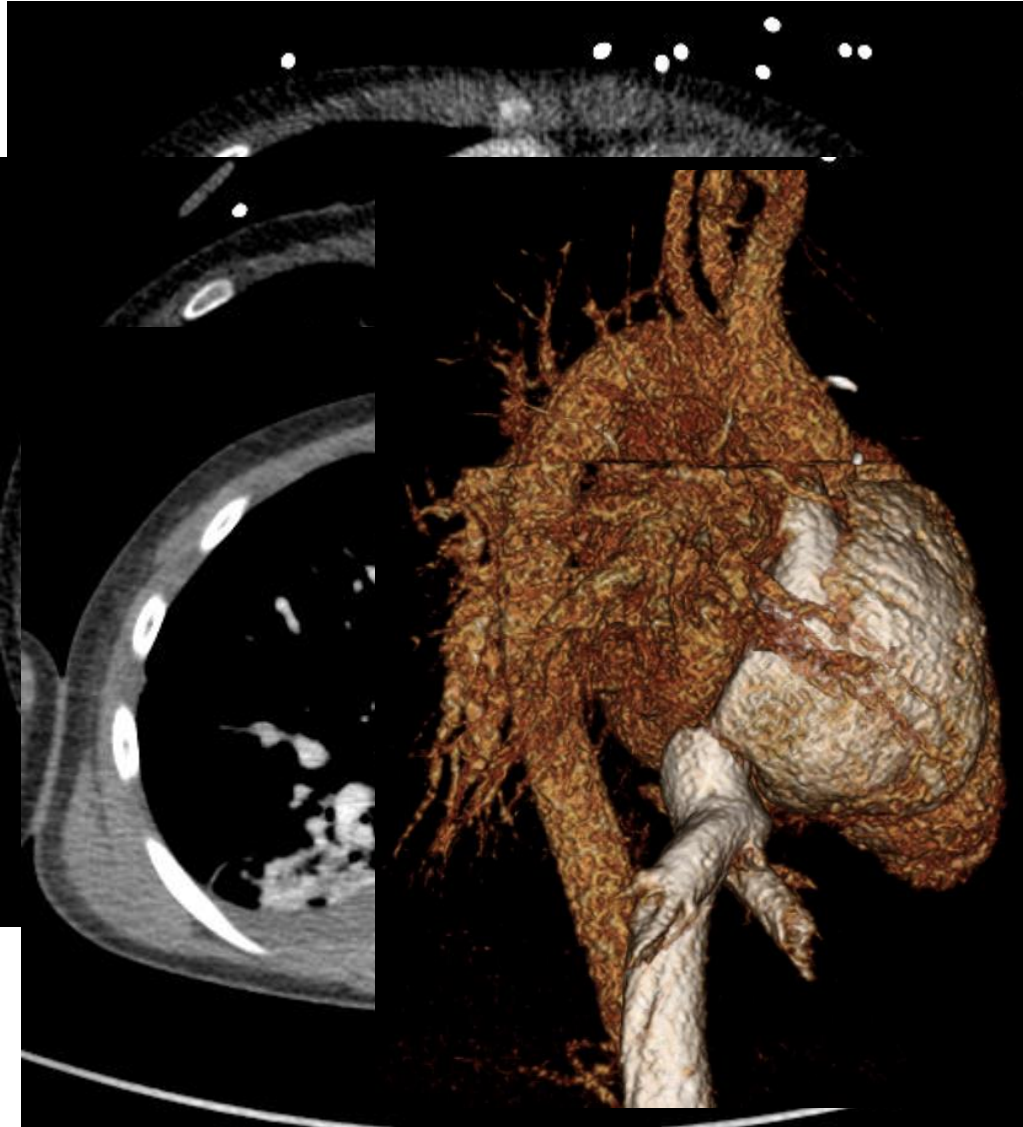


# Partially anomalous pulmonary venous connection

Portion of atrial septum excised

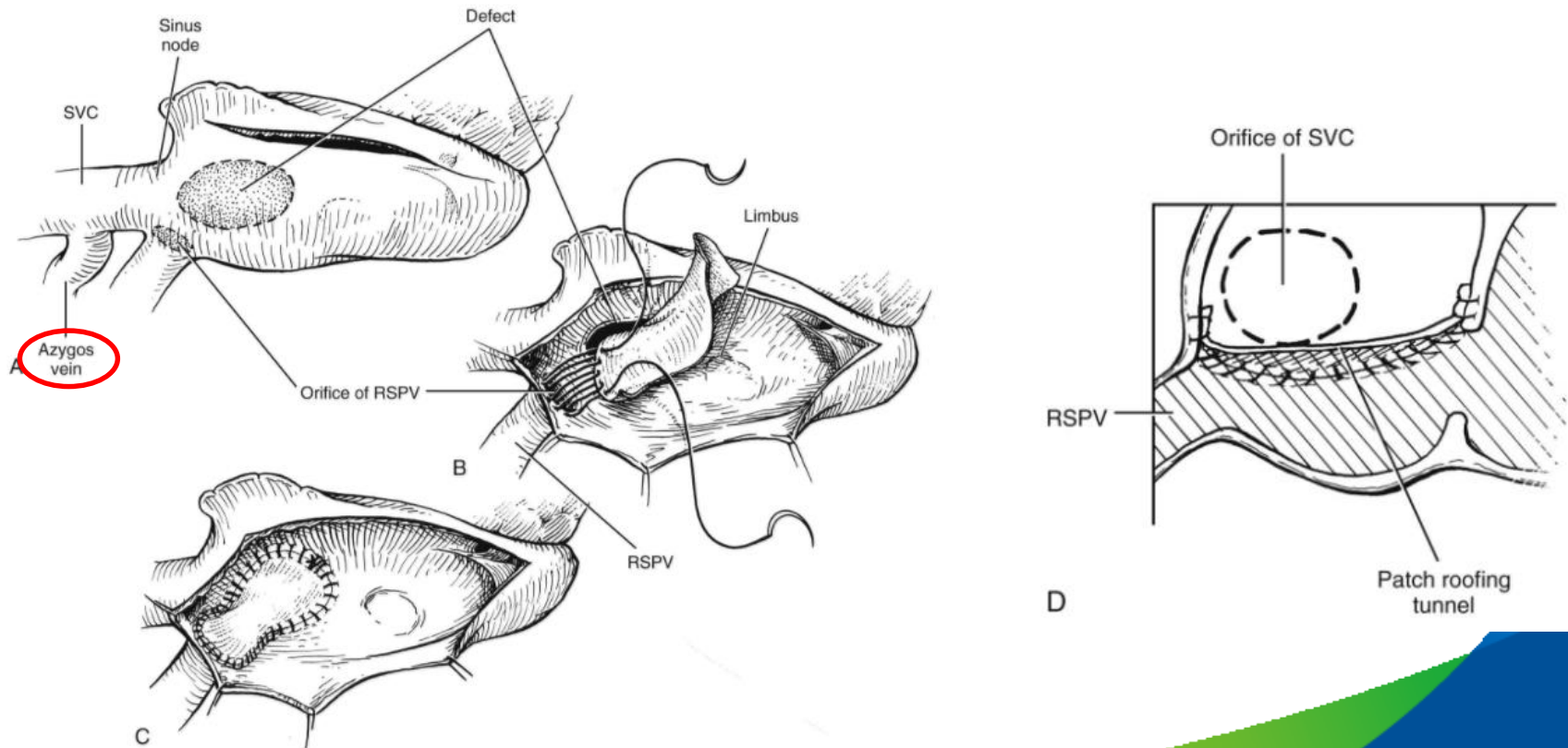


# Partially anomalous pulmonary venous connection



# Partially anomalous pulmonary venous connection

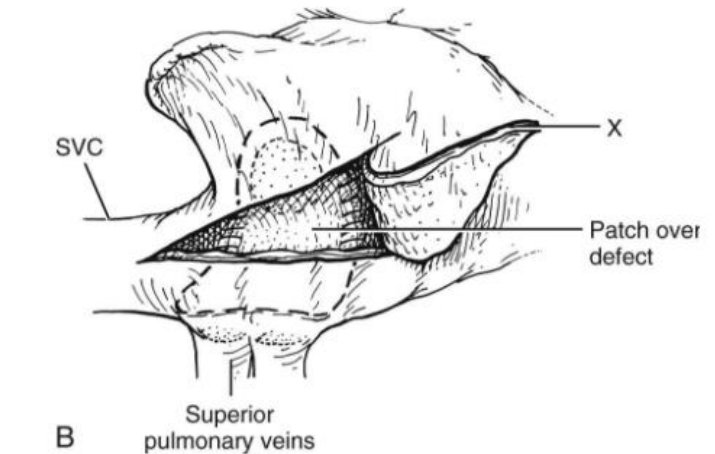
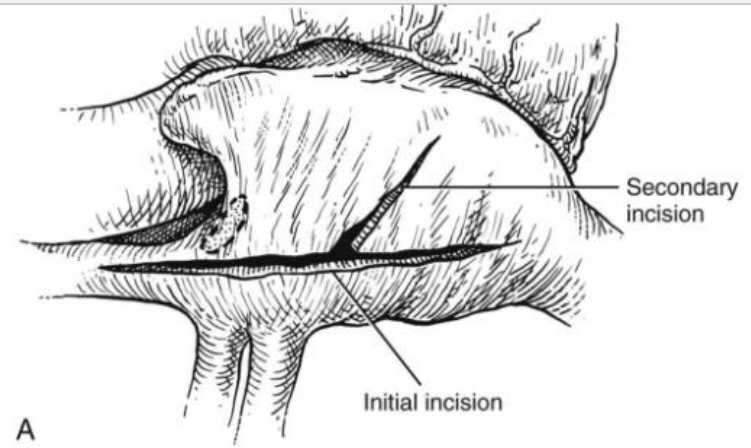
- Management
  - Surgery





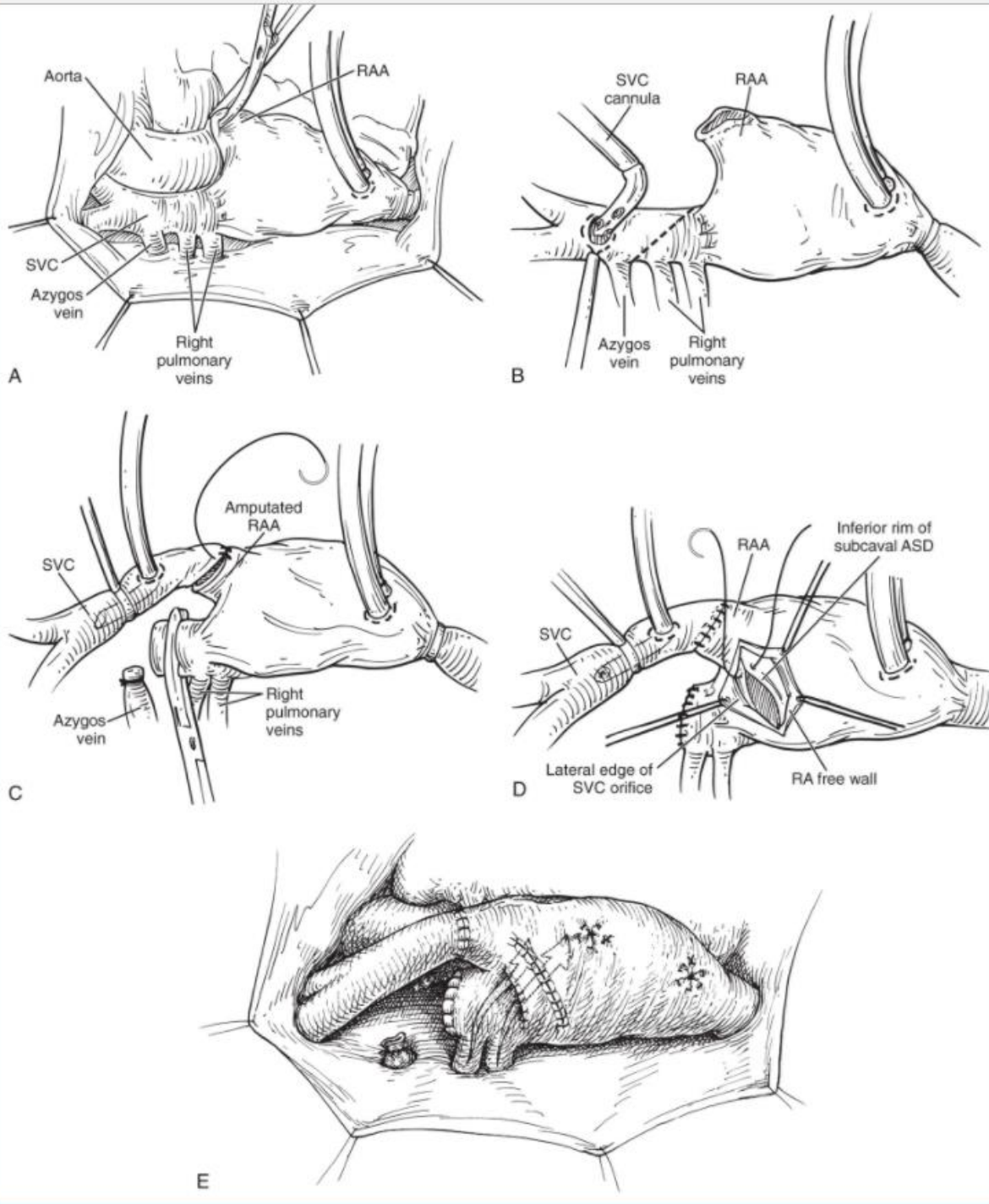
# Partially anomalous pulmonary venous connection

- Management
  - Surgery (V-Y atrioplasty technique for enlarging SVC)



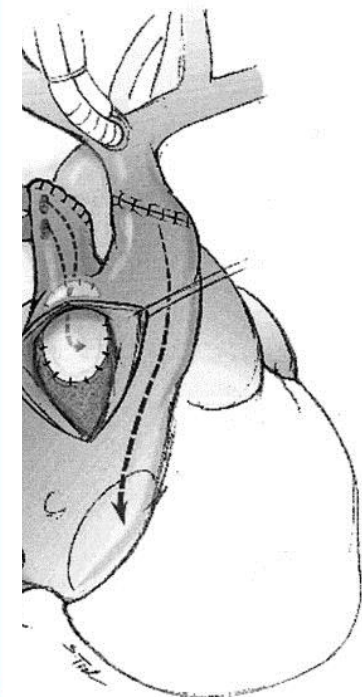
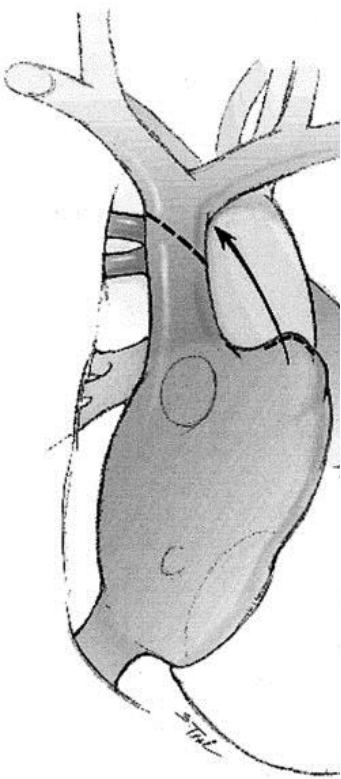
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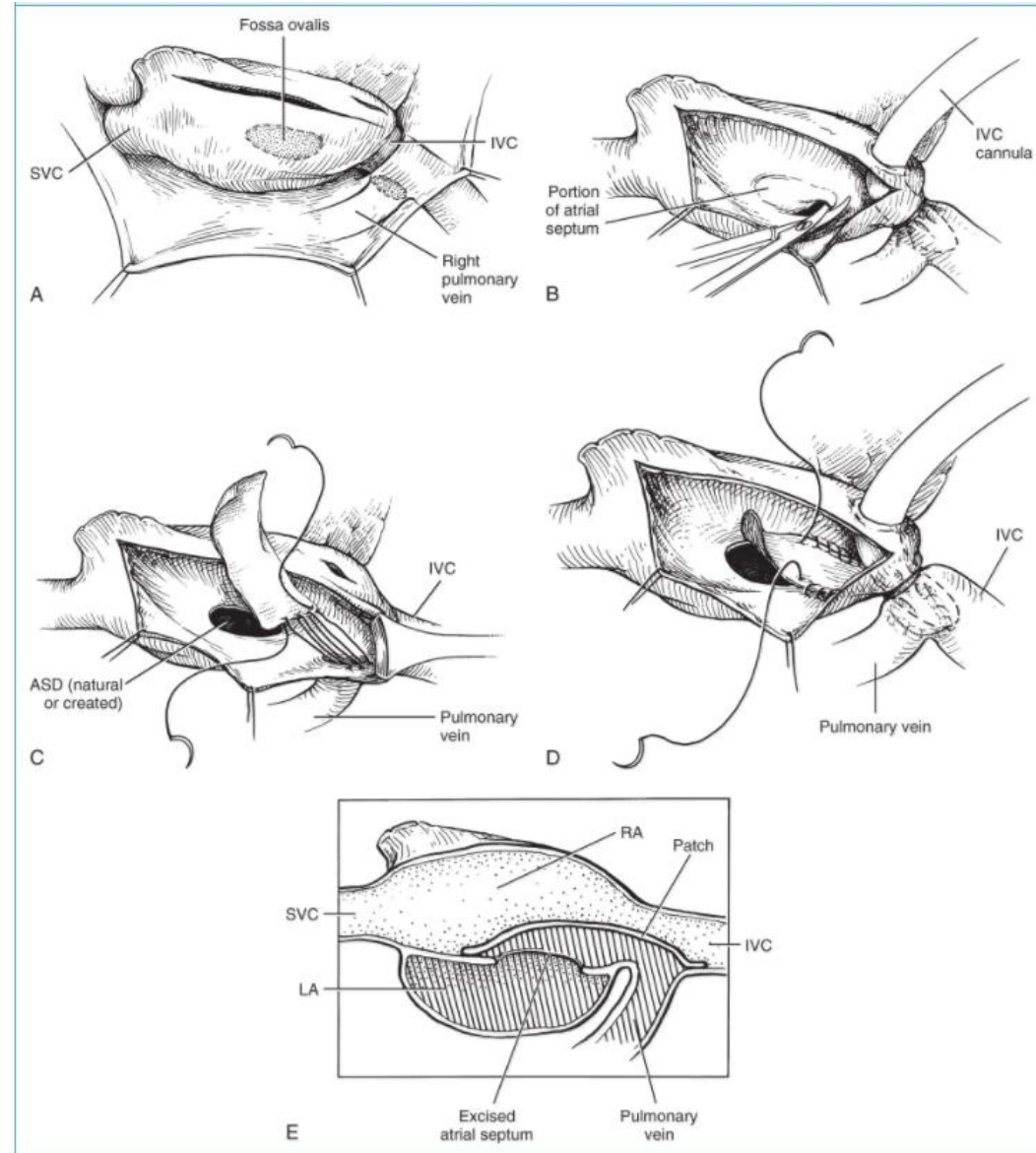
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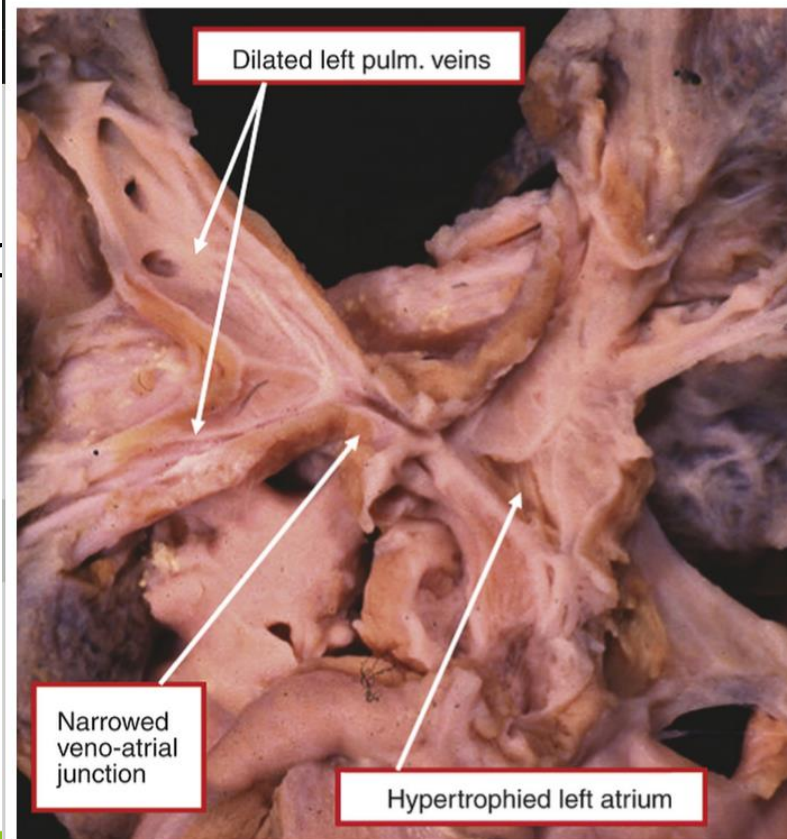
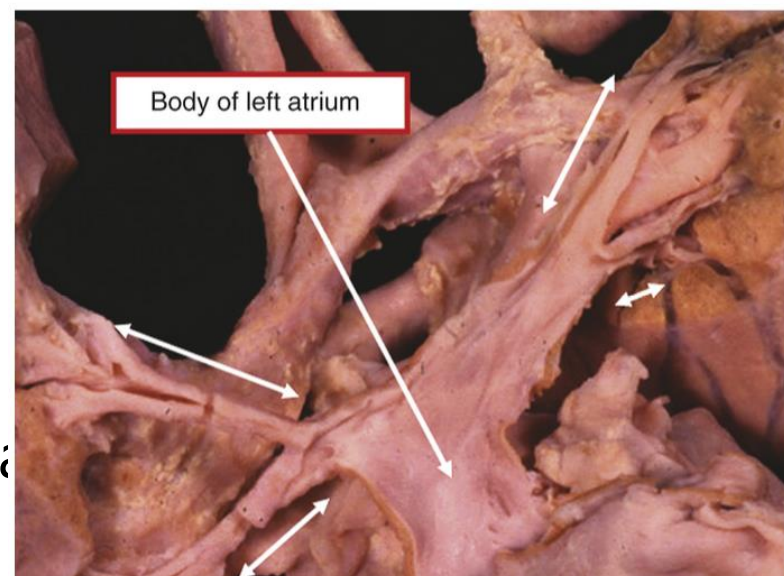
# Partially anomalous pulmonary venous connection

- Management
  - Surgery (scimitar syndrome)



# Stenosis or atresia pulmonary

- Anatomy
  - Individual pulmonary veins can be stenosed at their junction with the atrium, or exhibit tubular hypoplasia over a significant intra- and extrapulmonary distance
  - Pulmonary venous stenosis can also result from acquired causes, such as **constrictive pericarditis, mediastinitis, pulmonary tuberculosis or invasion by tumour**



# 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

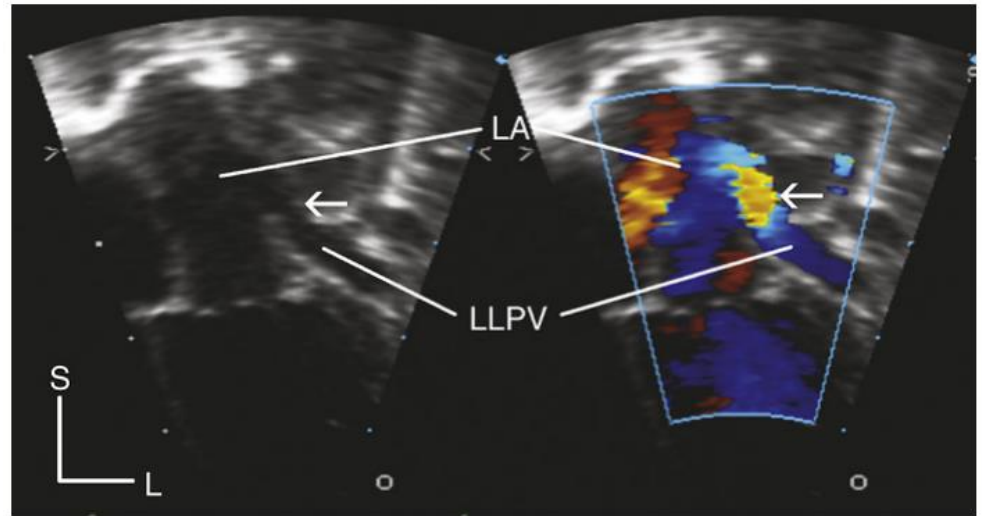
- Presentation and symptoms
  - Dyspnea and repeated pulmonary infections with failure to thrive are the rule
  - Many patients have haemoptysis and, occasionally, cyanosis

# 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

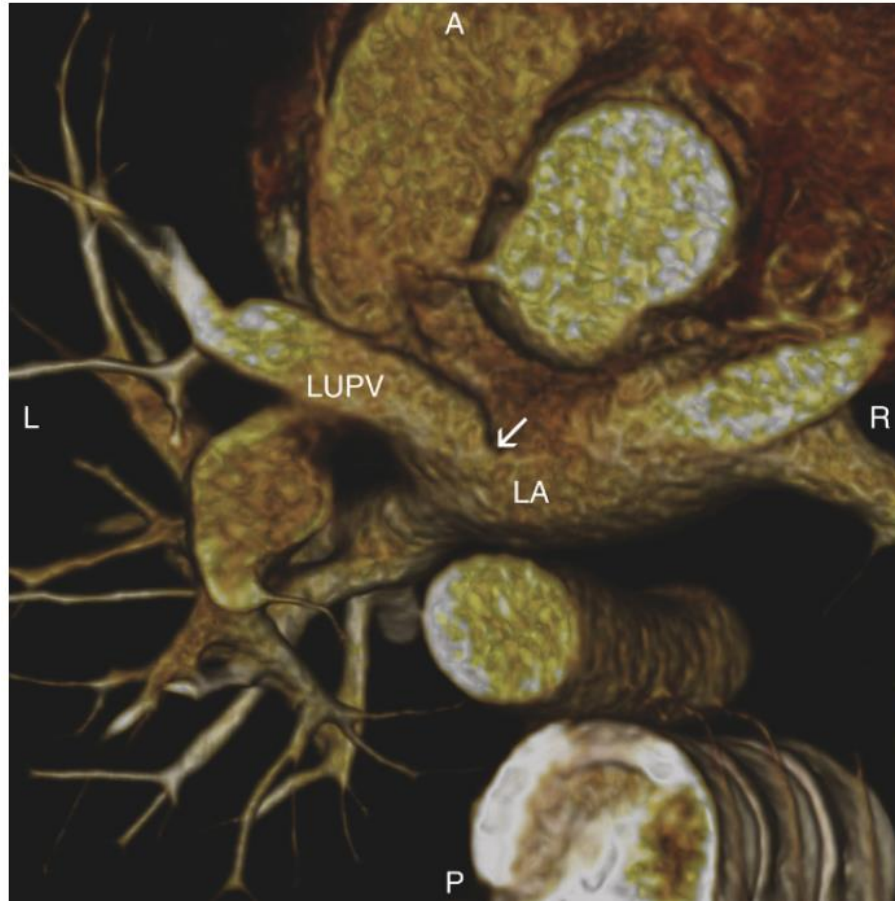
- Investigations
  - Echocardiography
    - Turbulence or a focal increase in flow velocity with a continuous, non-phasic flow pattern distally in a pulmonary vein





# Stenosis or atresia of individual pulmonary veins

- Investigations
  - Computed tomographic angiography



# 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**