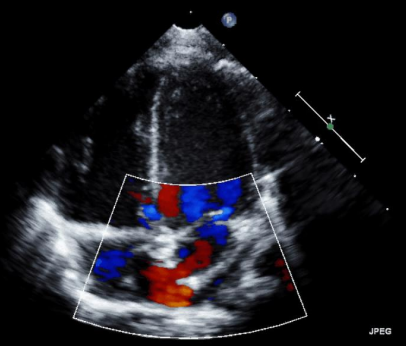
KSC 2014

# Pulmonary Venous Anomaly - Embryology and Anatomy




Lucy Youngmin Eun, MD, PhD
Pediatric Cardiology
Yonsei University Severance Hospital

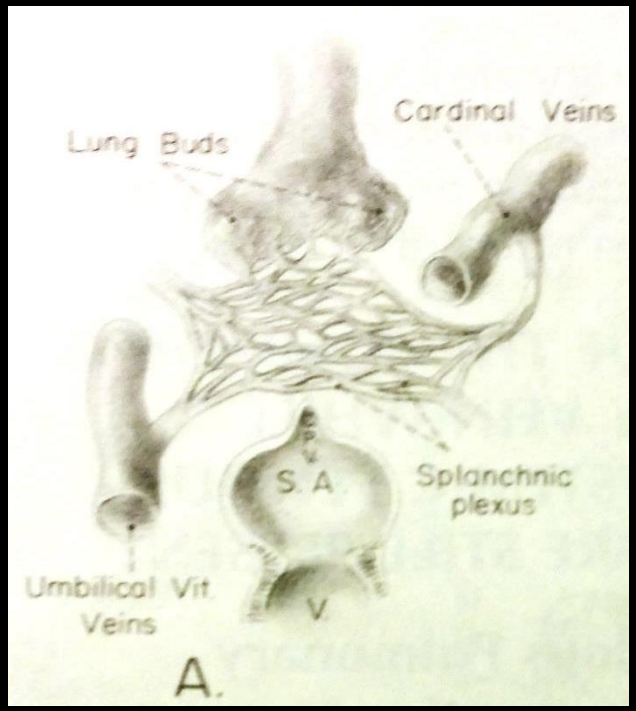
# Pulmonary Vein Anomaly

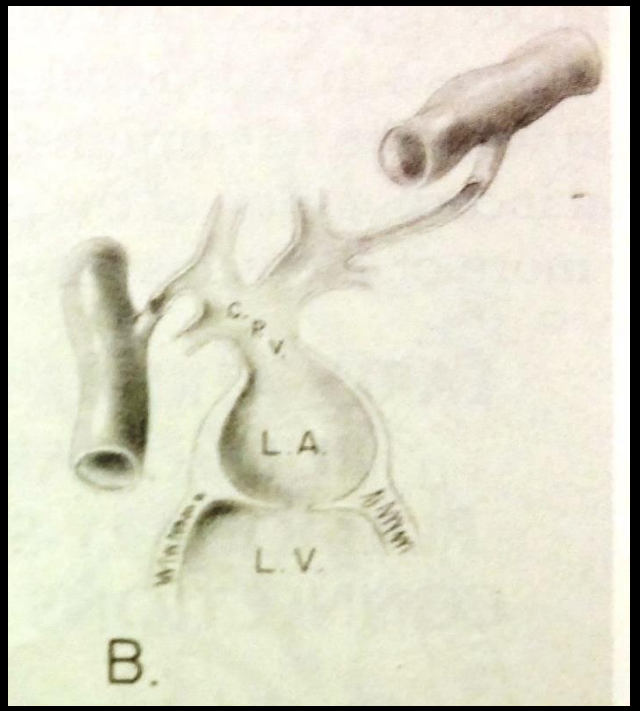
- Total Anomalous Pulmonary Venous Connection
- Partial Anomalous Pulmonary Venous Connection
- Common Pulmonary Vein Stenosis or Atresia
- Unilateral Pulmonary Vein Stenosis or Atresia

# Embryology

- The development of pulmonary vein begins at 27~29 days of gestation.
- The vein arise from the lung buds that are part of the vascular plexus of the forgut, the splanchnic plexus.
- There are multiple connections to the umbilicovitelline and cardinal venous system.
- *Coalesce to form four vessels that join with common pulmonary vein that emerges from the back wall of atrium.*

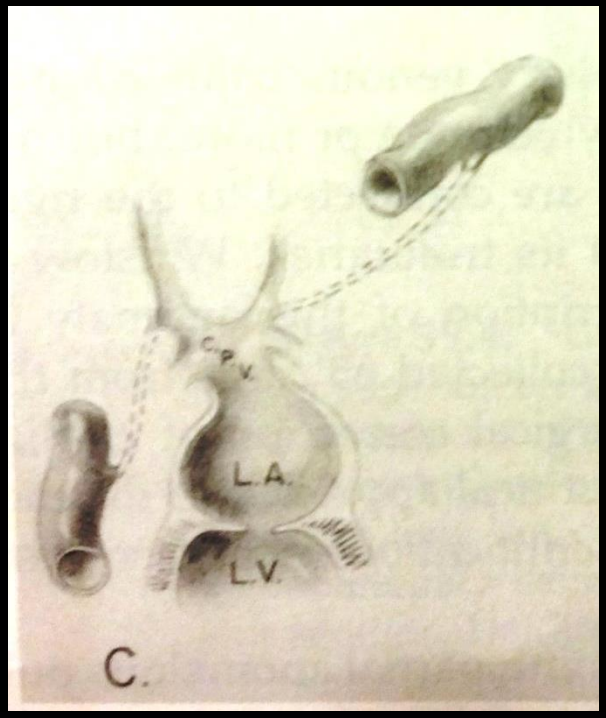
# Normal Development

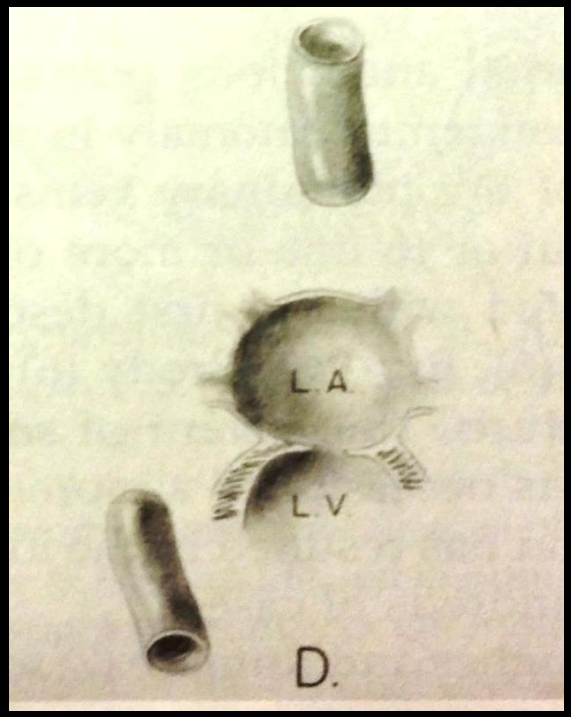



A. Splanchnic plexus drains the lung buds. It shares the root of drainage of splanchnic plexus, cardinal venous system, and the umbilicovitelline systems.
B. The common pulmonic vein originates as an invagination from the left atrial side of the common atrium and establishes communication with the splanchnic plexus.

# Normal Development




C. No longer necessary, the primitive pulmonary venous connections disappear.

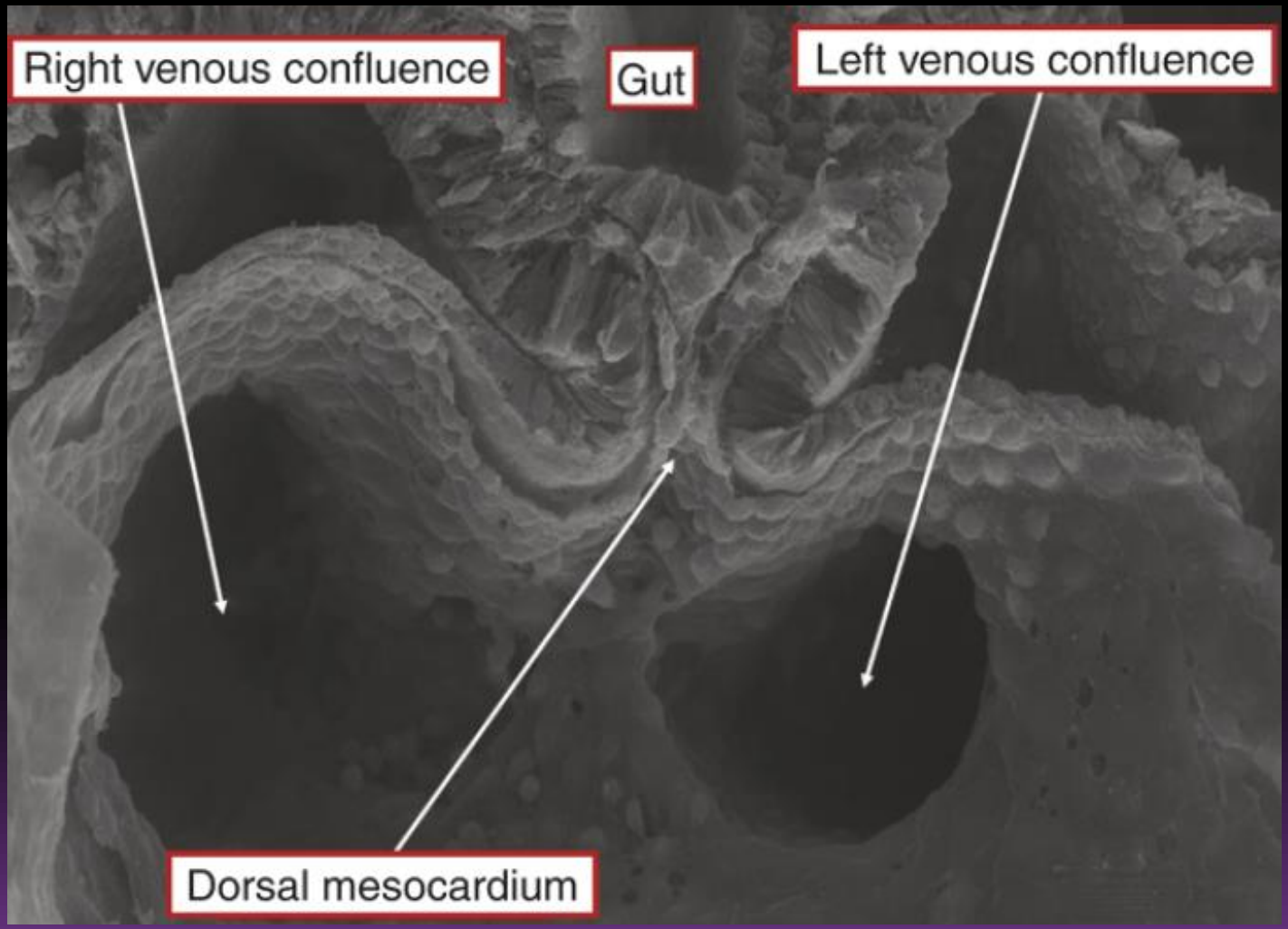
D. Finally, the individual pulmonary veins are incorporated into the left atrium, and the common pulmonary vein no longer exists.

# Embryology

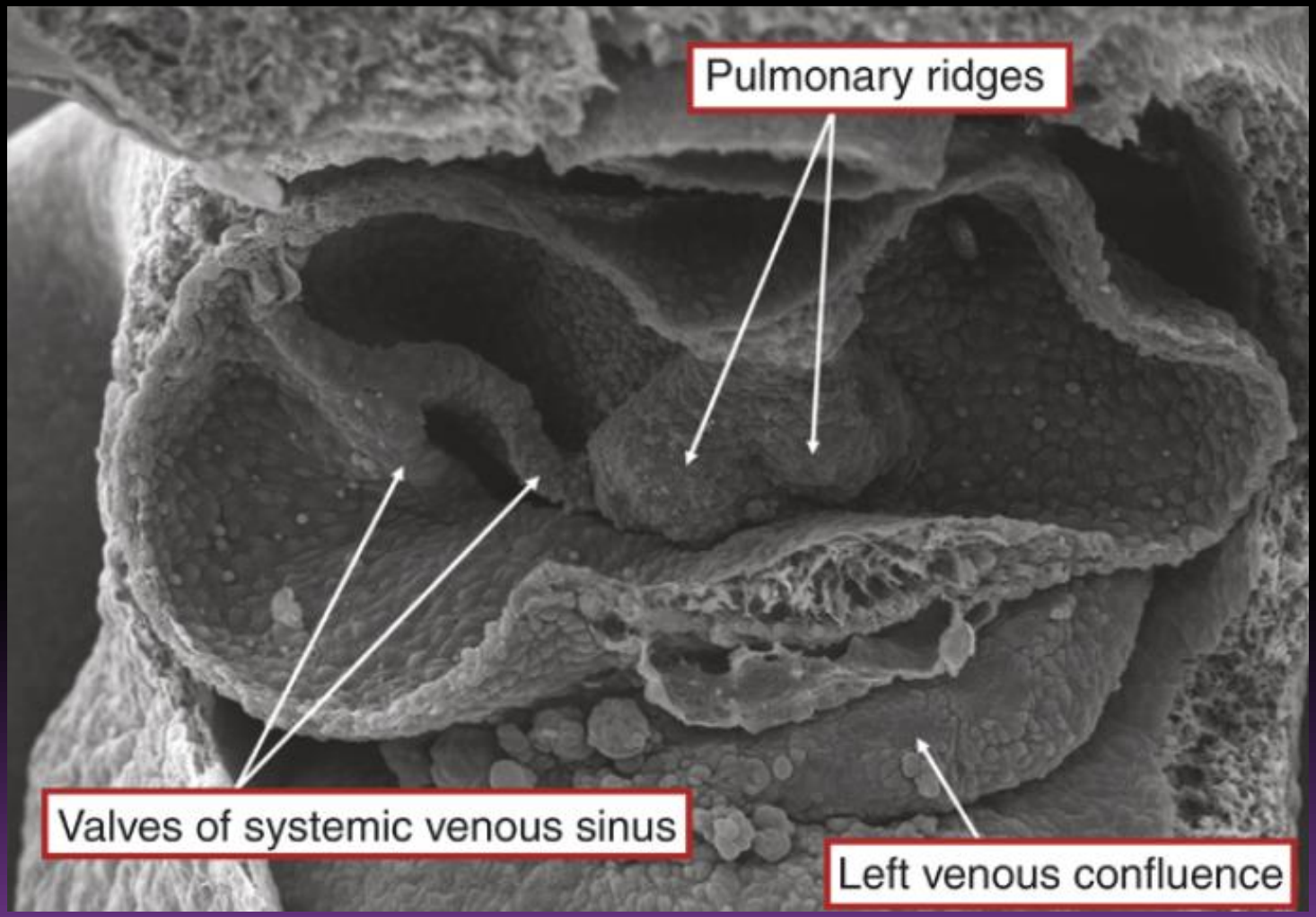
The Systemic venous tributaries identified in molecular terms expression of the transcription *factor Tbx18*.

The Pulmonary veins
→ do not contain this protein.

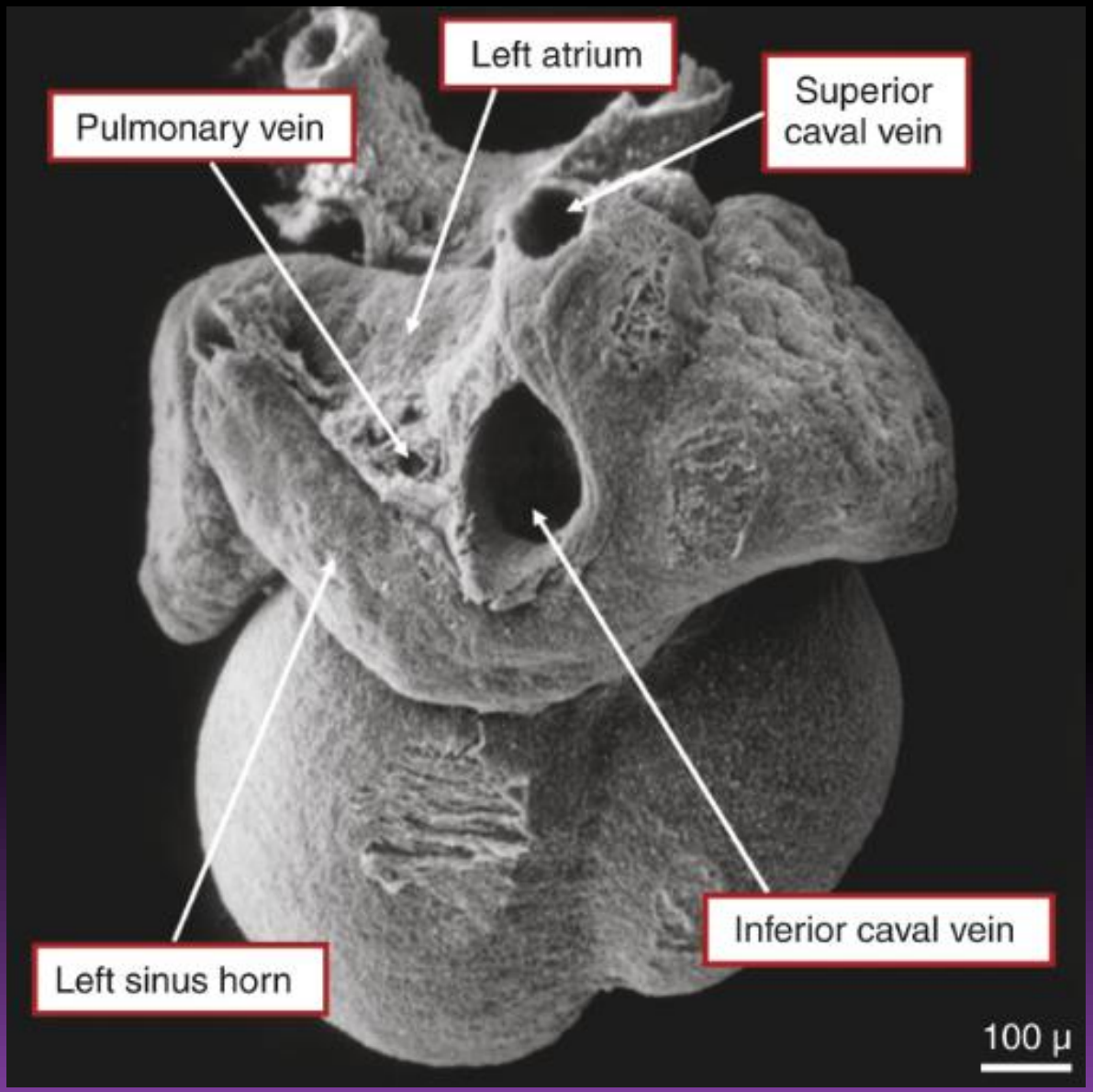
# Embryology - 4 wks



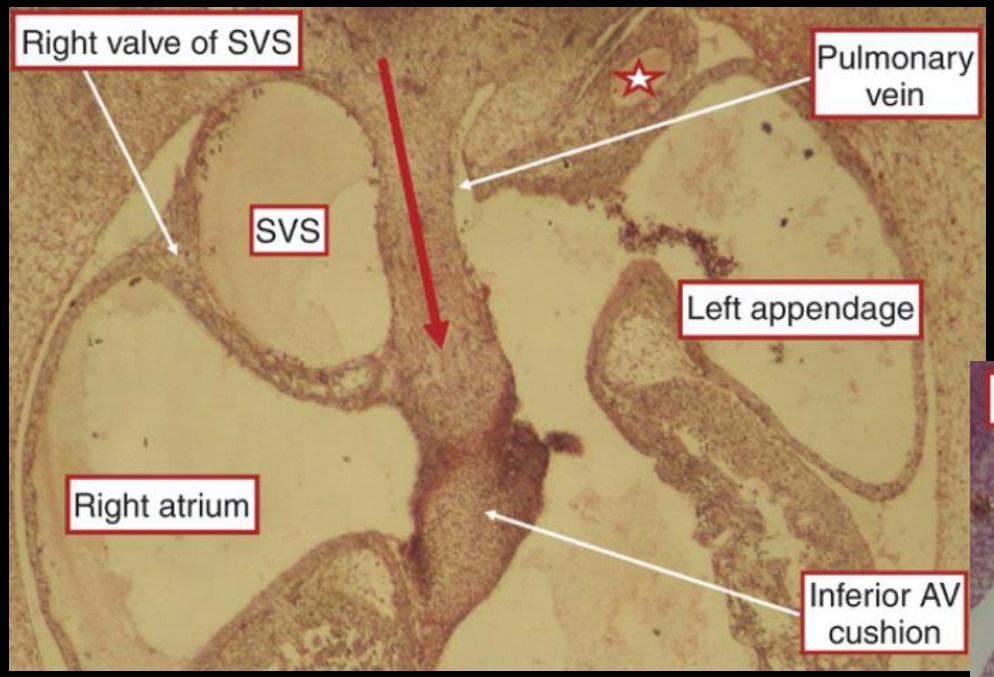
# Embryology – 5 wks

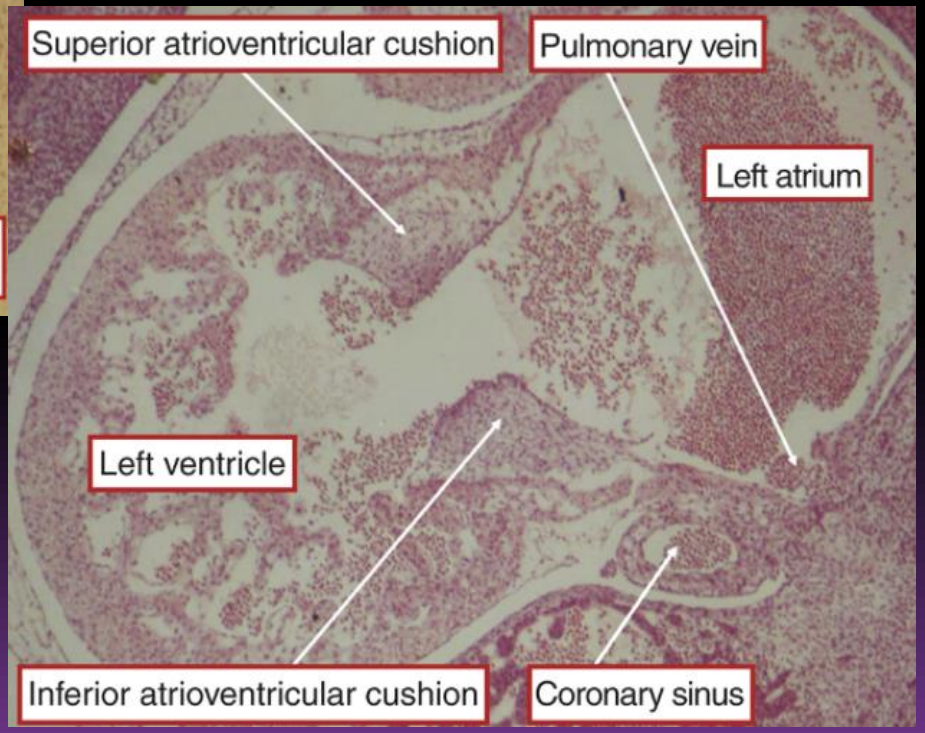


# Embryology – 6 wks

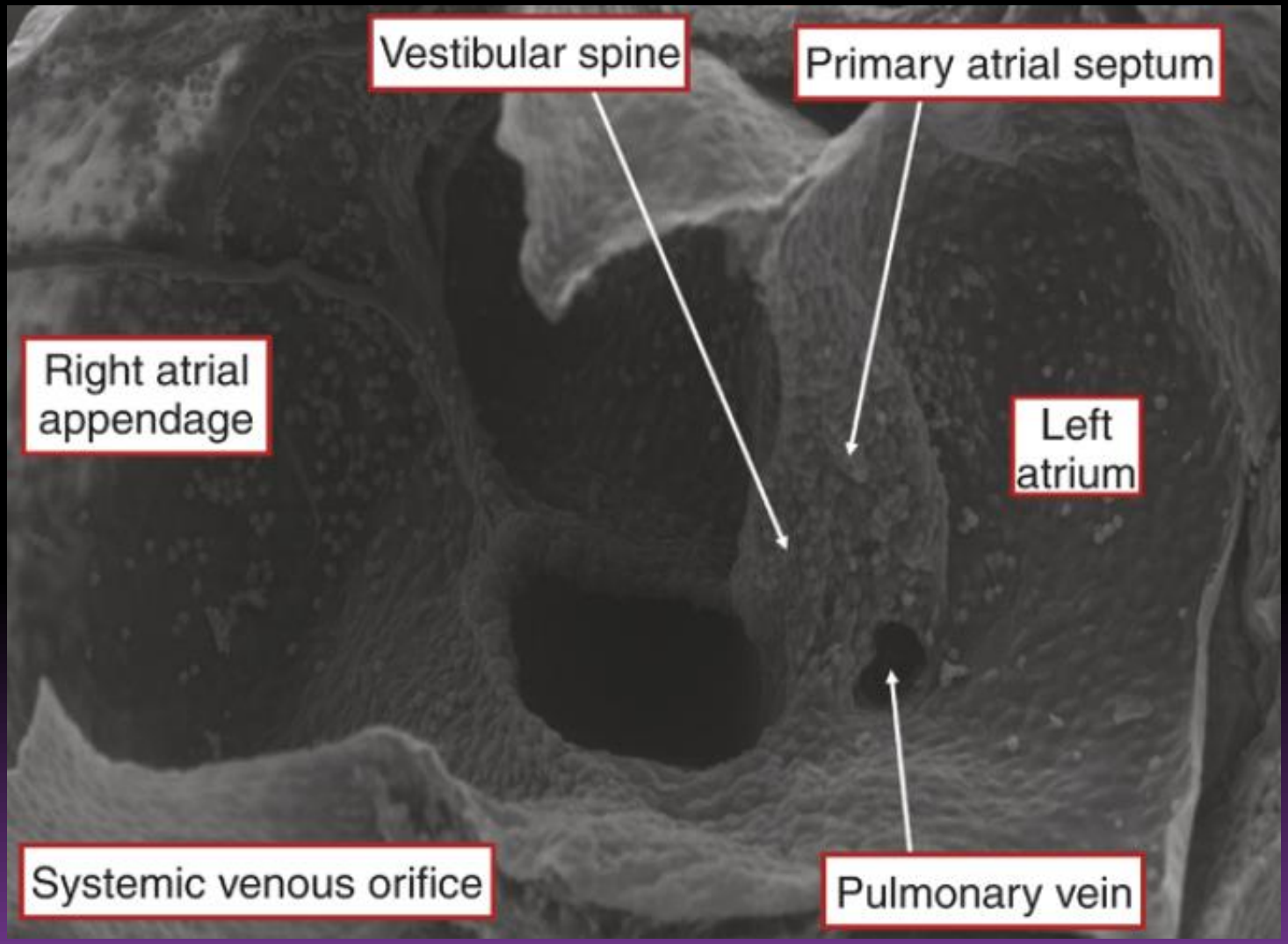


# Embryology – 51 wks

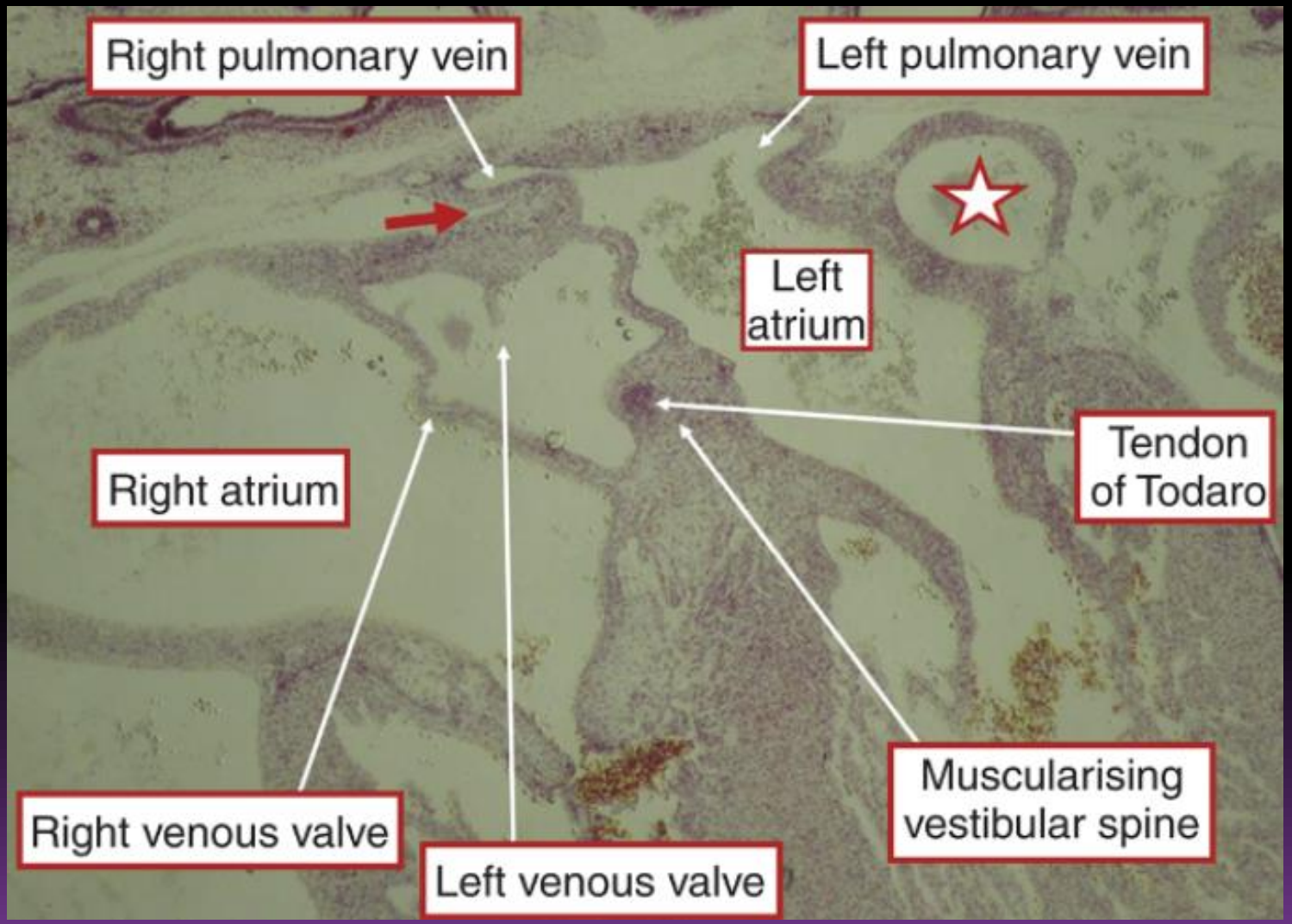


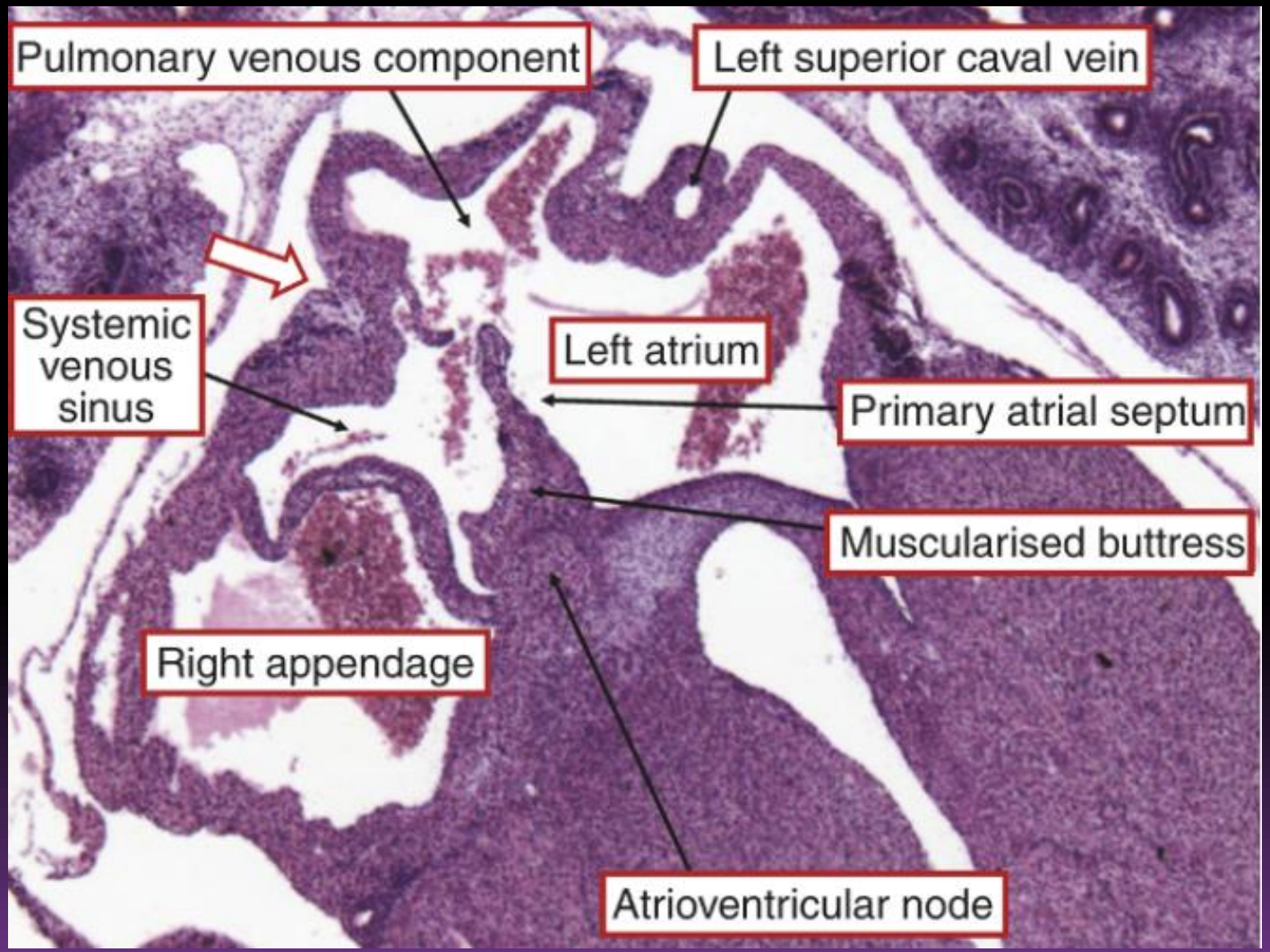
# Embryology


Vestibular spine
Primary atrial septum
Right atrial appendage
Left atrium
Systemic venous orifice
Pulmonary vein

# Embryology


Right pulmonary vein
Left pulmonary vein
Left atrium
Tendon of Todaro
Right atrium
Muscularising vestibular spine
Right venous valve
Left venous valve

# Embryology


Pulmonary venous component
Left superior caval vein
Systemic venous sinus
Left atrium
Primary atrial septum
Muscularised buttress
Right appendage
Atrioventricular node

# Embryologic Classification

I. Atresia of common pulmonary vein while pulmonary systemic venous connections are still present

A. Partial anomalous pulmonary venous connection

B. Total anomalous pulmonary venous connection

II. Atresia of the common pulmonary vein after pulmonary systemic venous connections are obliterated

- Atresia of the common pulmonary vein

III. Stenosis of the common pulmonary vein

- Cor triatriatum

IV. Abnormal absorption of the common pulmonary vein into the left atrium

A. Stenosis of the individual pulmonary vein

B. Abnormal number of pulmonary vein

# Anomalous Connection

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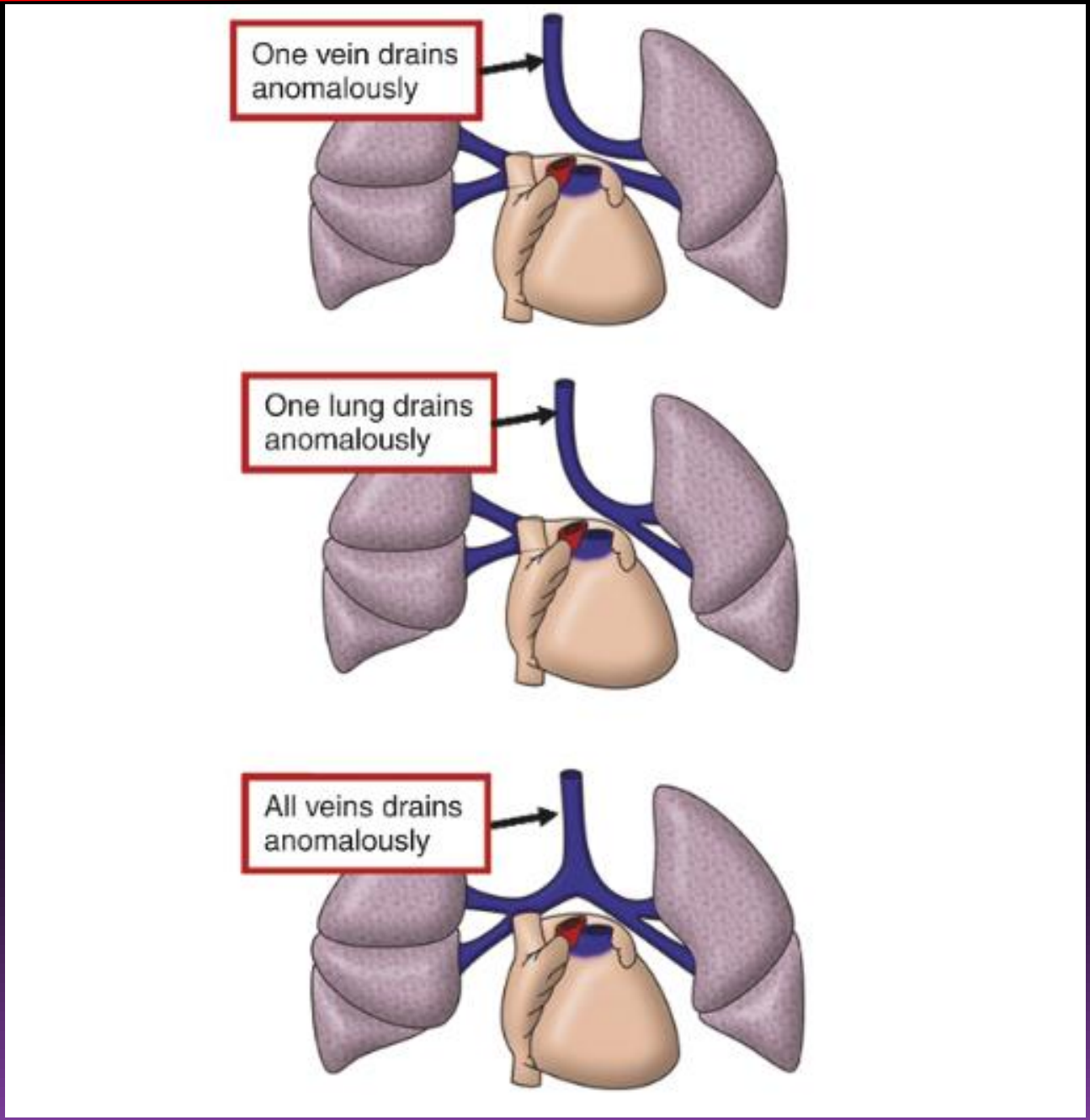
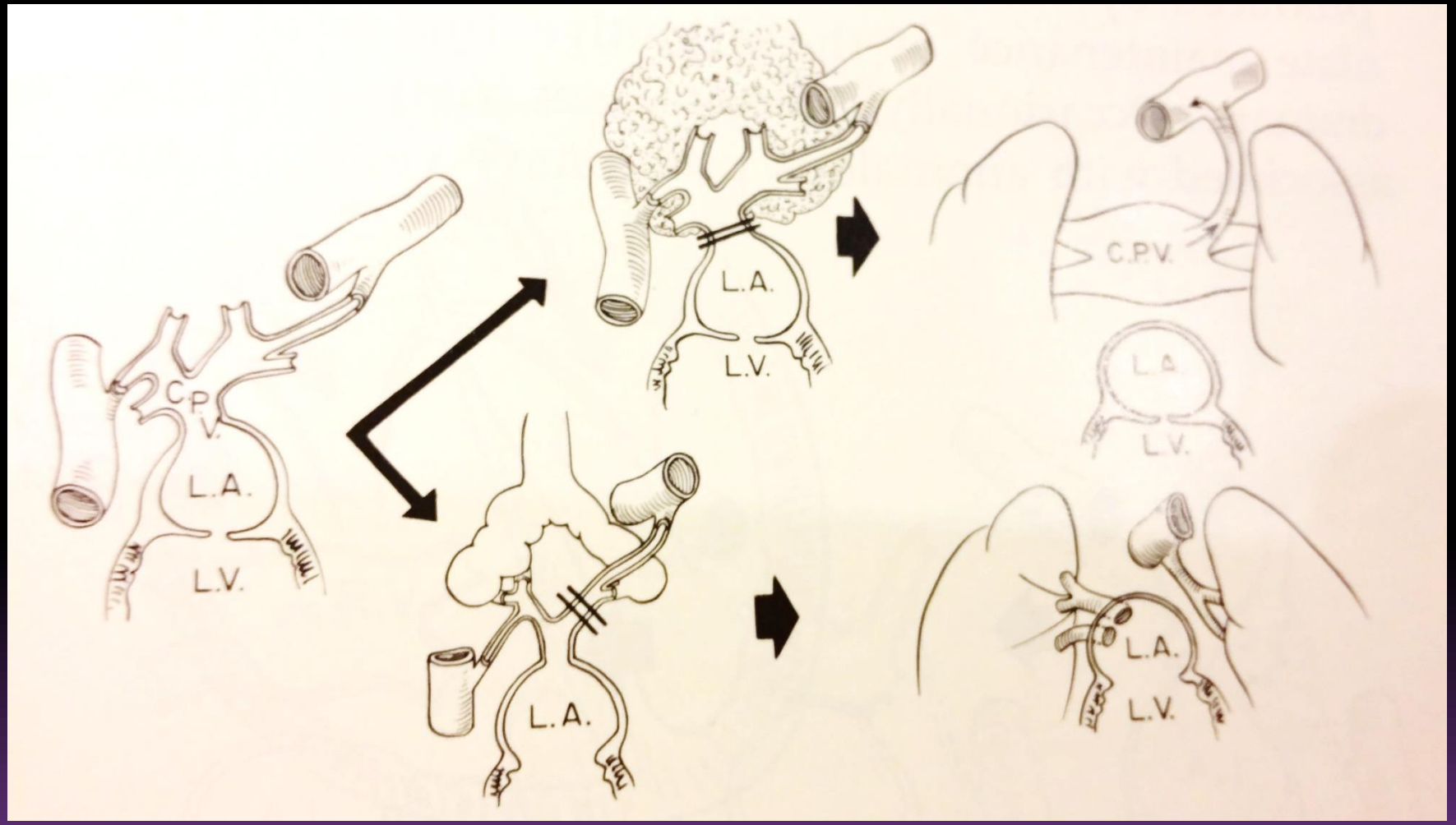
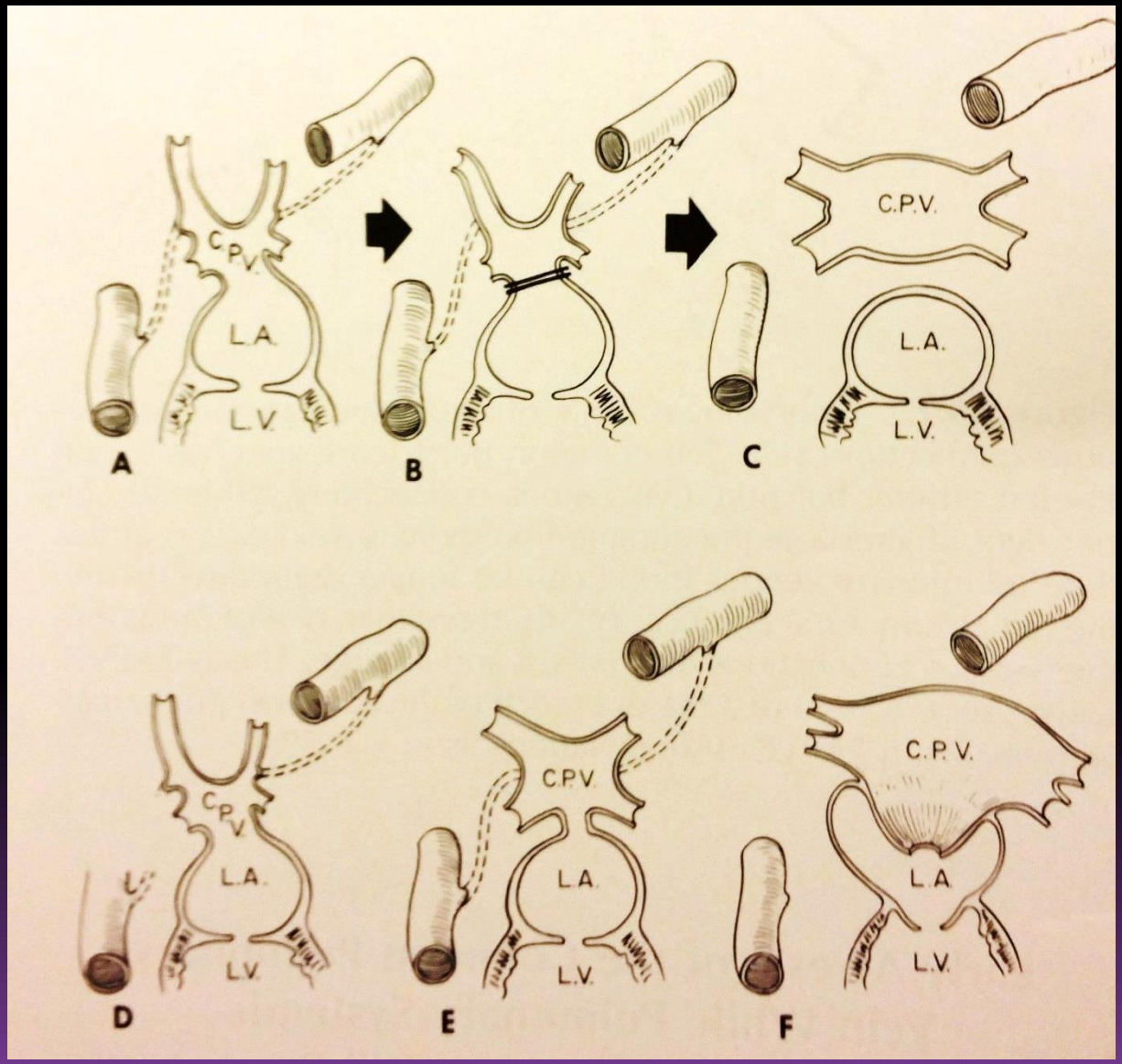


# Why Anomalous Connection?



# Why Anomalous Connection?



# Pulmonary Vein Anomaly

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- Common Pulmonary Vein Stenosis or Atresia
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# Total Anomalous Pulmonary Venous Connection

***A pulmonary vein is connected anomalously only when it is attached to a site other than the morphologically left atrium***

- 1 % of all congenital heart disease
- Boy : Girl = 4 : 1

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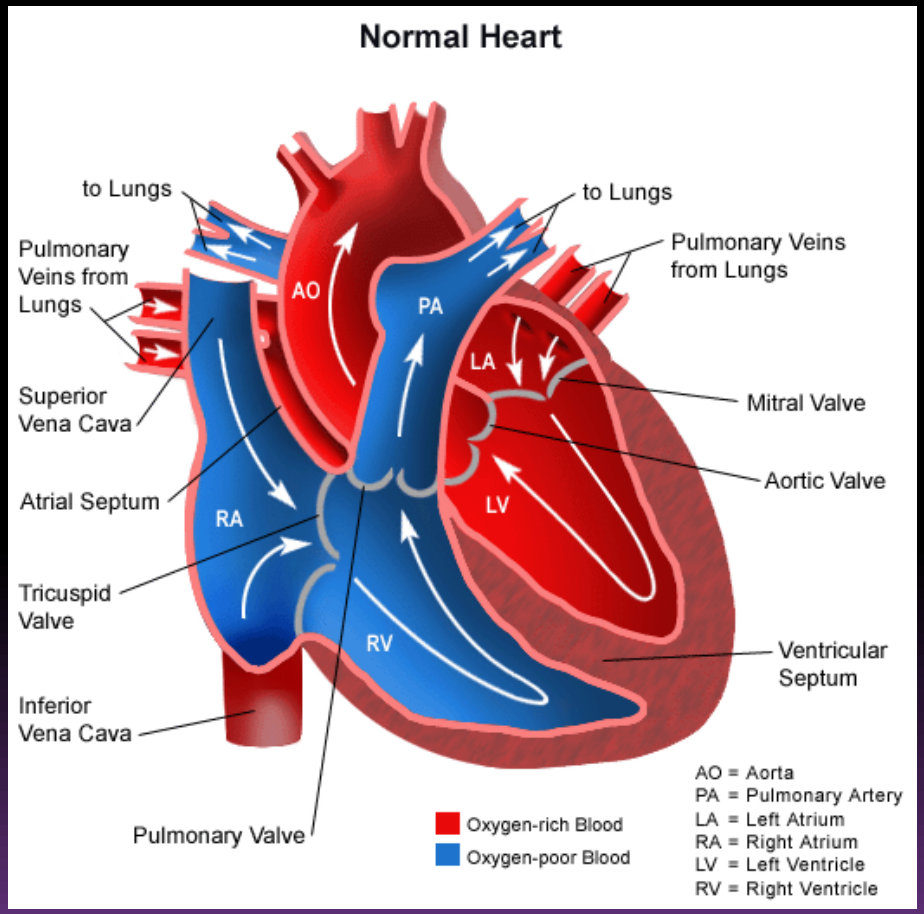
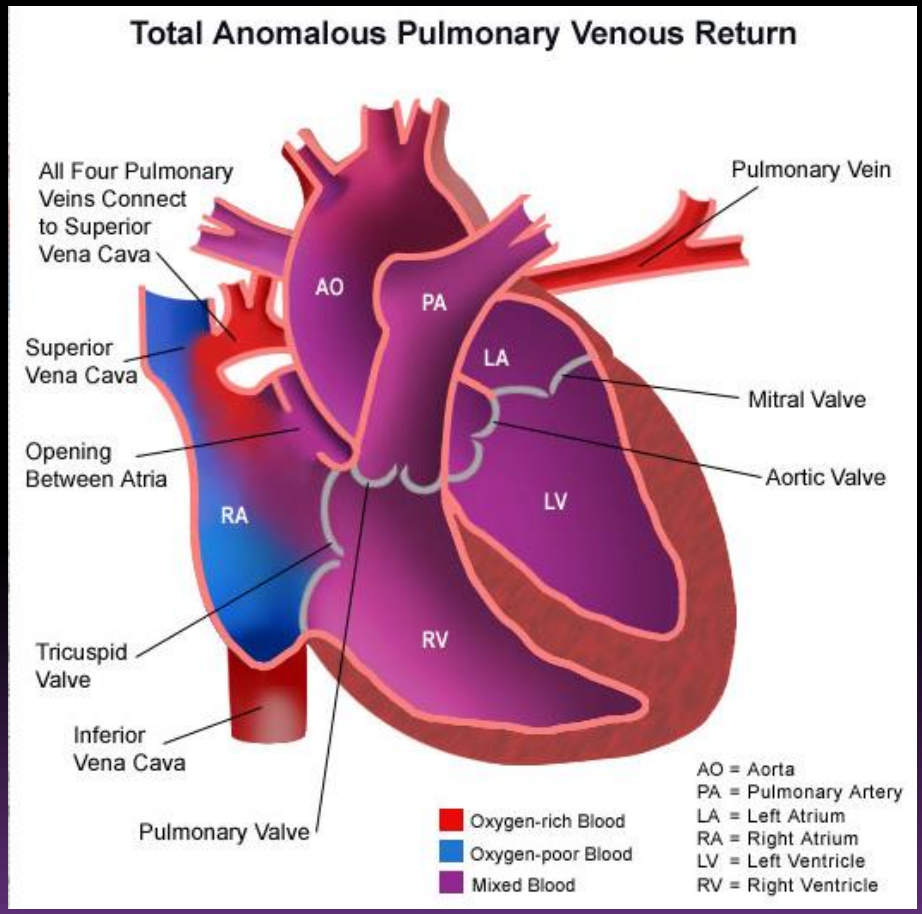
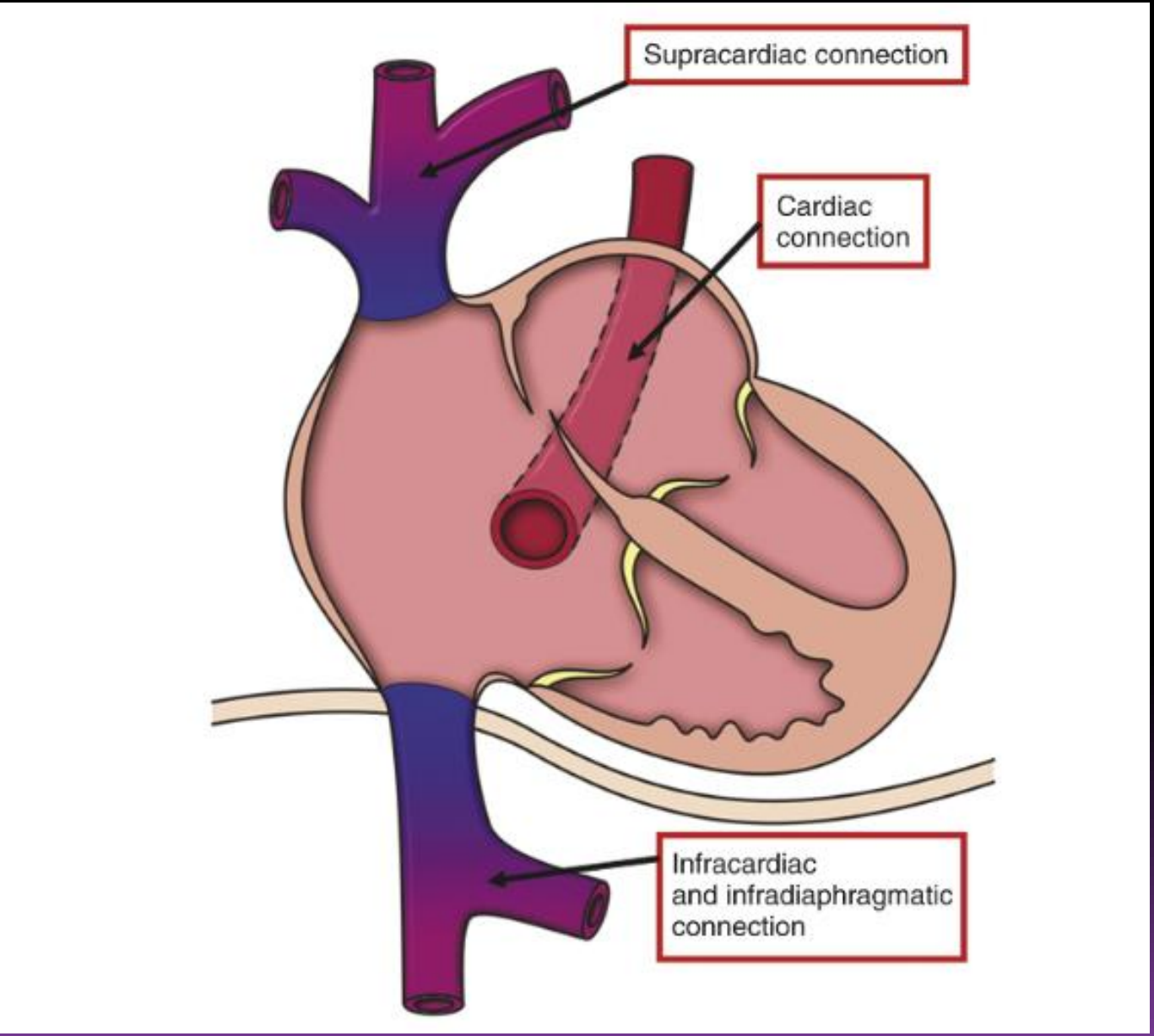
# Total Anomalous Pulmonary Venous Connection

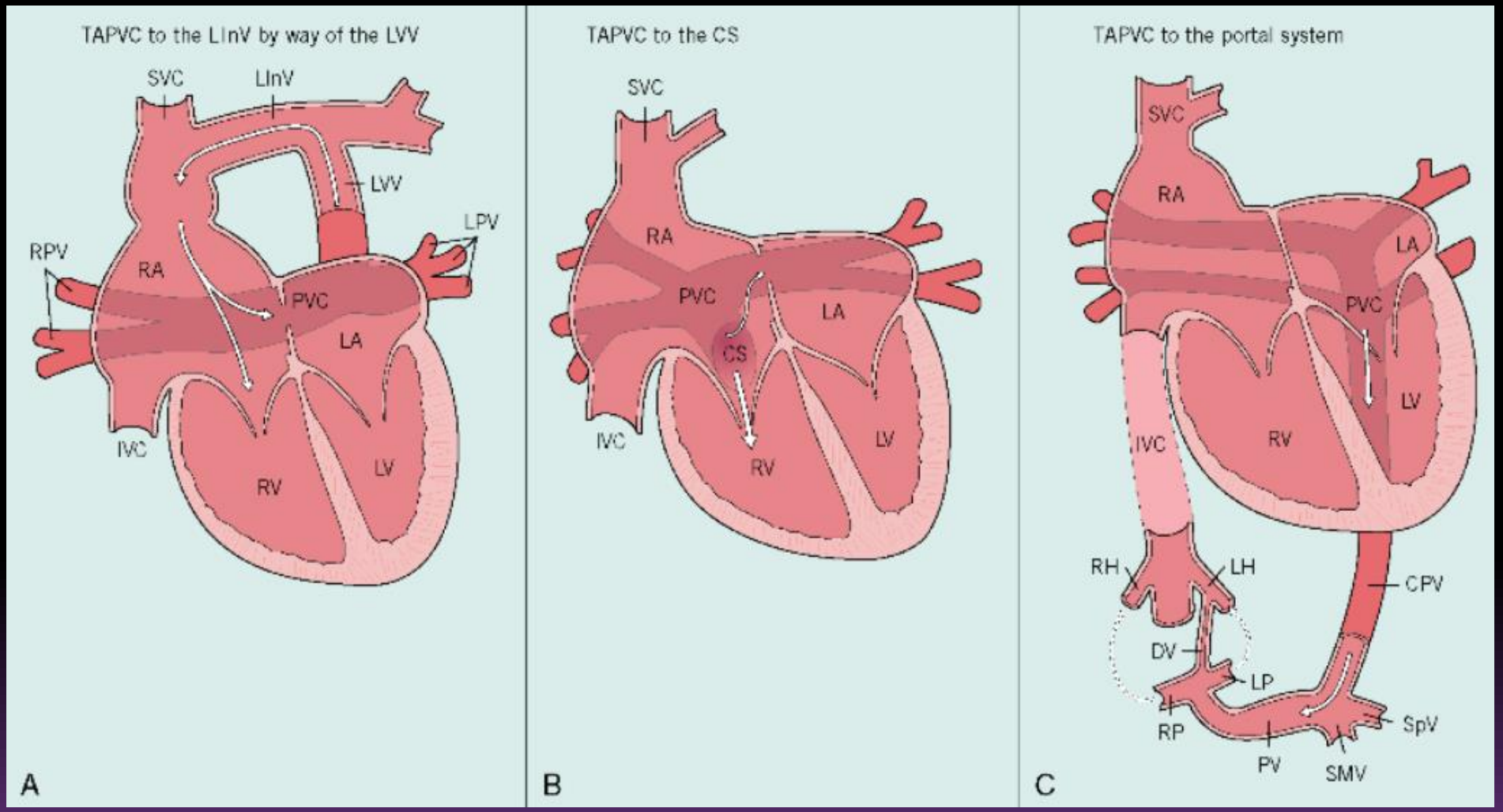


# Different Sites of Anomalous Connection



# Different Sites of Anomalous Connection



# Anatomy of TAPVC

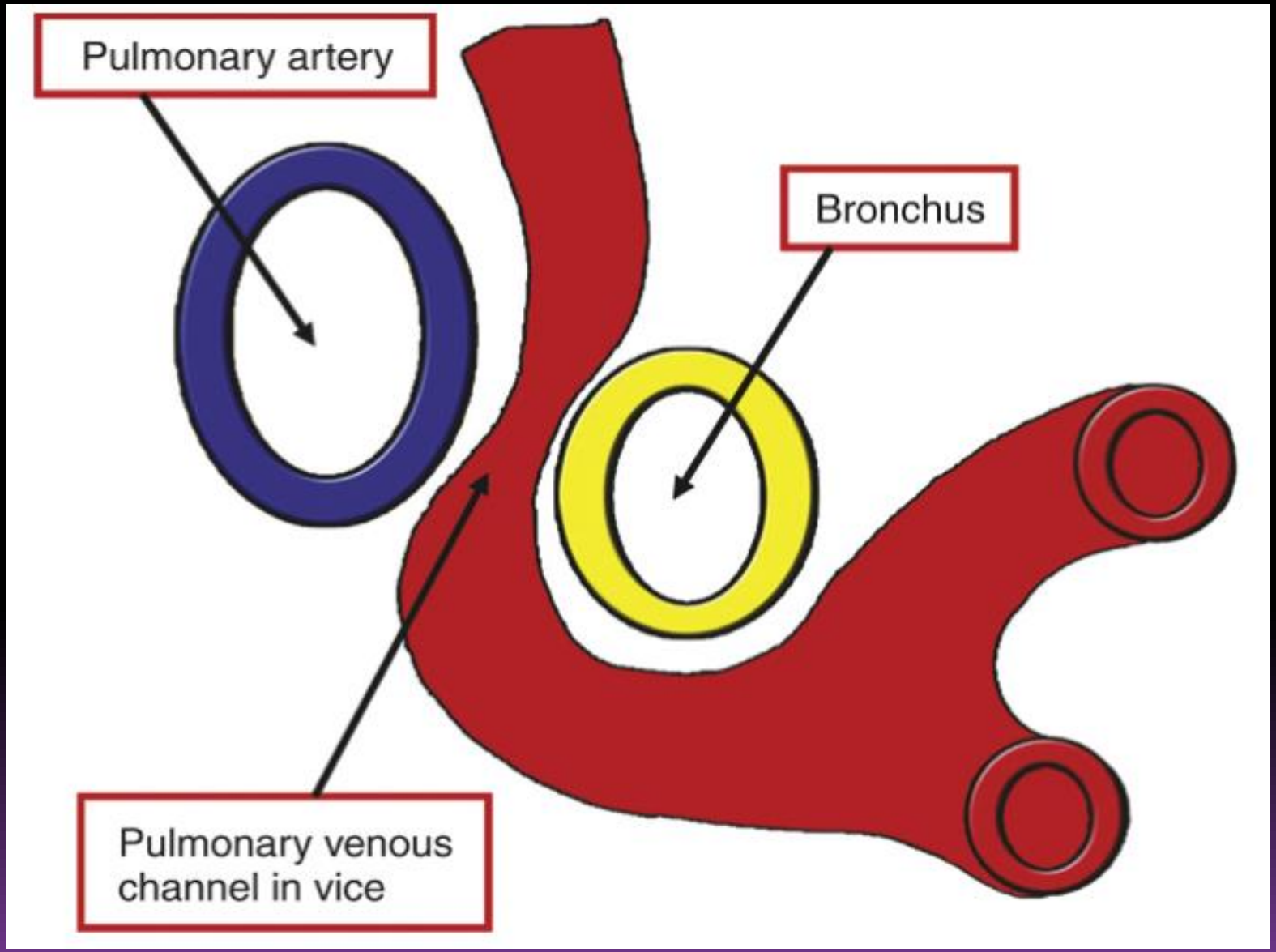
- Anomalous connections
- Stenotic connections
- Abnormal numbers of pulmonary veins

# Anatomy of TAPVC

KEY to the diagnosis of TAPVC

- the anatomic connections of all four pulmonary veins
- the size and location of each vein
- how and where the four veins enter the heart, to seek stenosis

# Nature of the Structure



# Anatomy of TAPVC

- to establish whether the anomalous pulmonary venous connection is:
  - an isolated malformation?
  - part of a more complex anomaly?
  - whether there are associated structural malformations of the pulmonary vasculature?

# Total Anomalous Pulmonary Venous Connection

2/3 of patients :

TAPVC is an isolated anomaly, associated only with the required interatrial communication.

1/3 of patients :

significant other cardiac defects occur, truncus arteriosus, pulmonary atresia, AVSD, TGA, single ventricle physiology, or heterotaxy syndrome (asplenia or polysplenia).

# TAPVC /c Pulmonary Venous Obstruction

Without severe pulmonary venous obstruction:
- Present in heart failure at 2~3 months of age.
- History of difficulty of feeding, pneumonia.
- Cyanosis is rare.

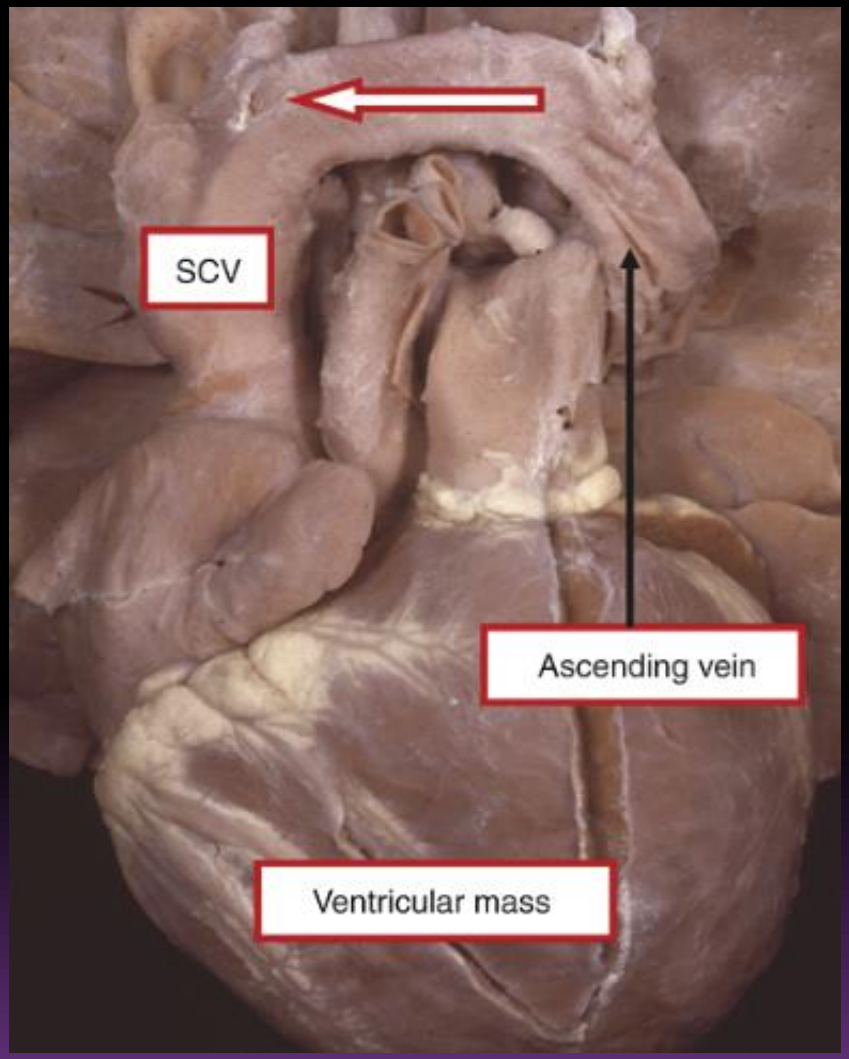
With severe pulmonary venous obstruction:
- Obvious severe Cyanosis
- Skin mottling reflecting poor peripheral perfusion and metabolic acidosis
- Tachypnea

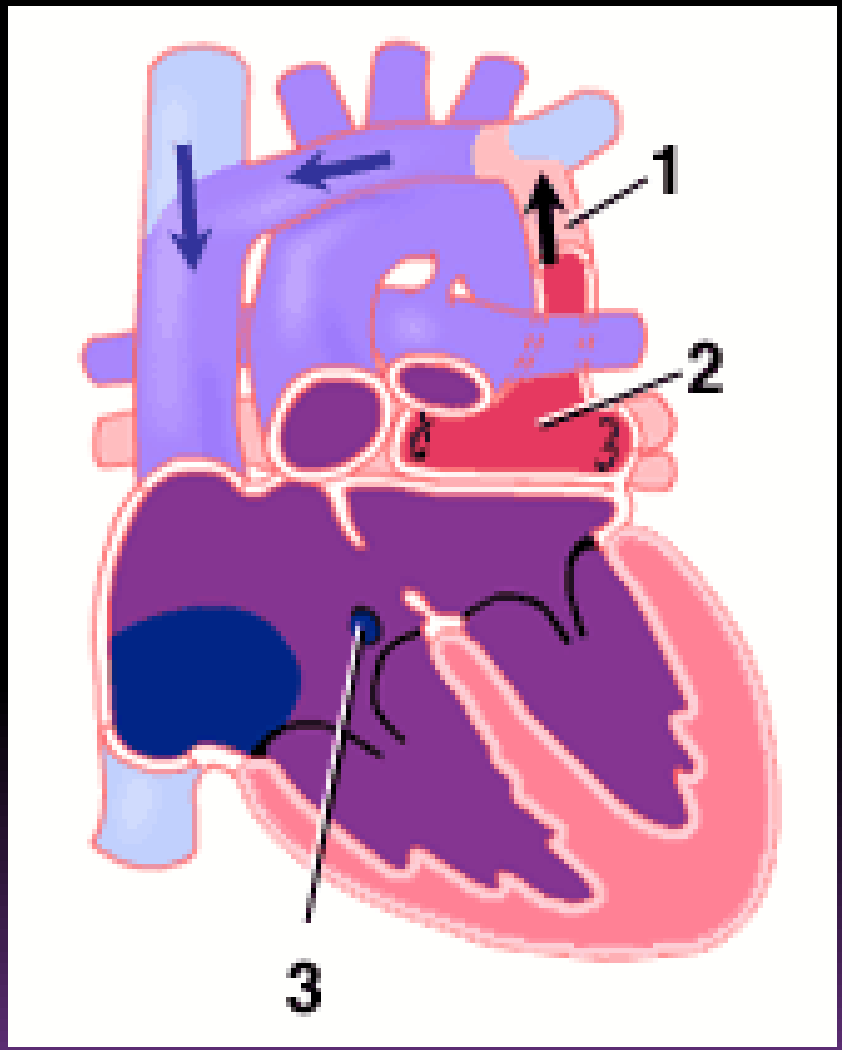
# Inheritance of TAPVC

- Cat eye syndrome –

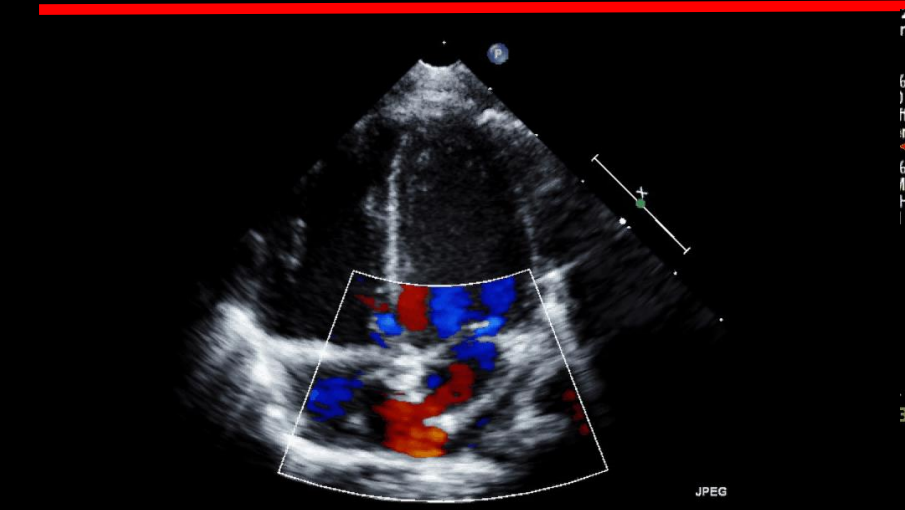
  trisomy of the centromeric portion of chromosome 22q
- Association with a deletion of chromosome 2q31-q33
- Holt-Oram syndrome
- Asplenia syndrome

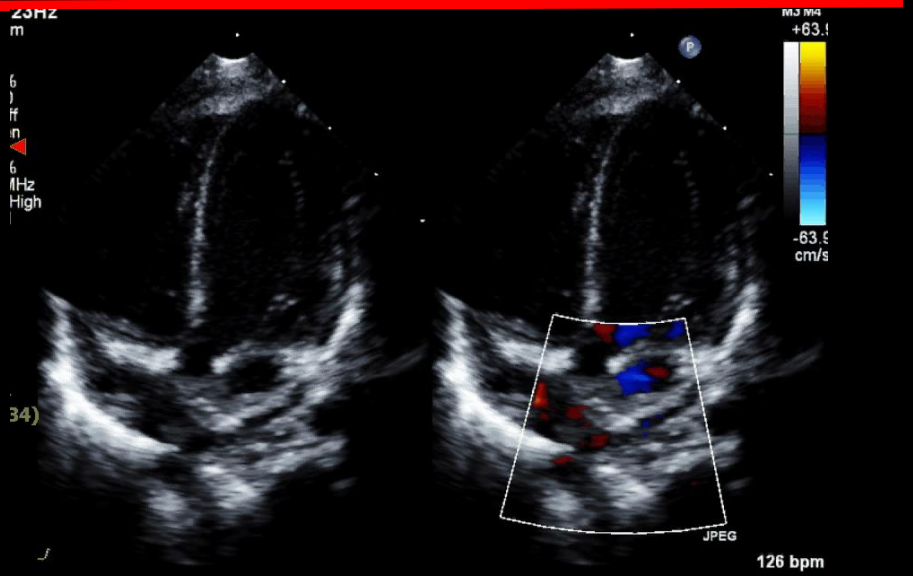
# TAPVC- Supracardiac

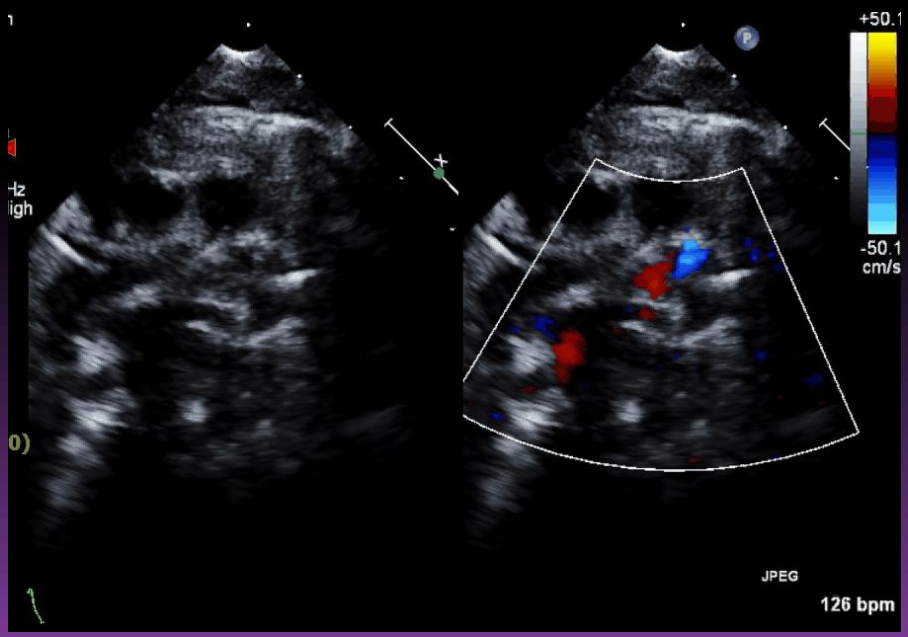


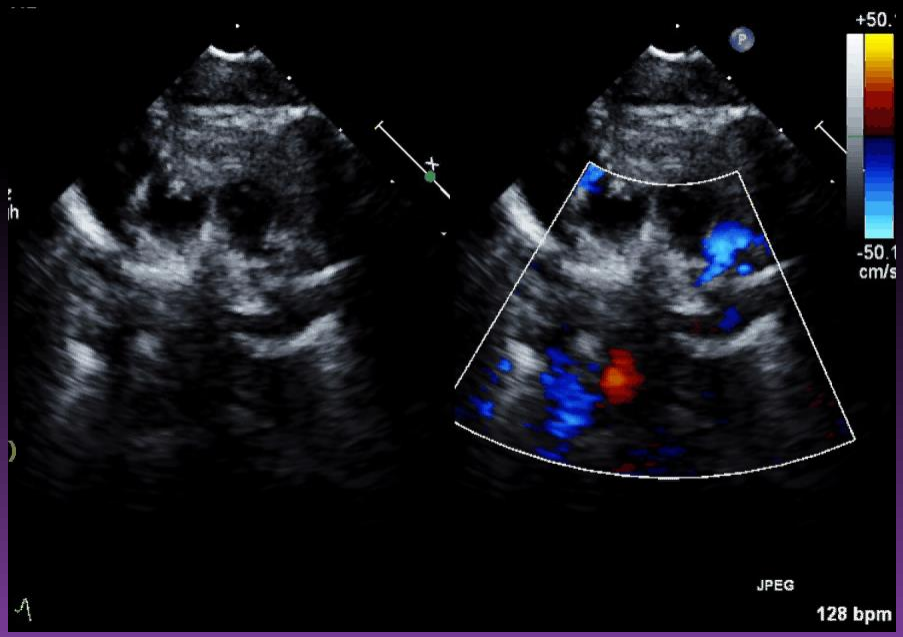



# TAPVC CASE

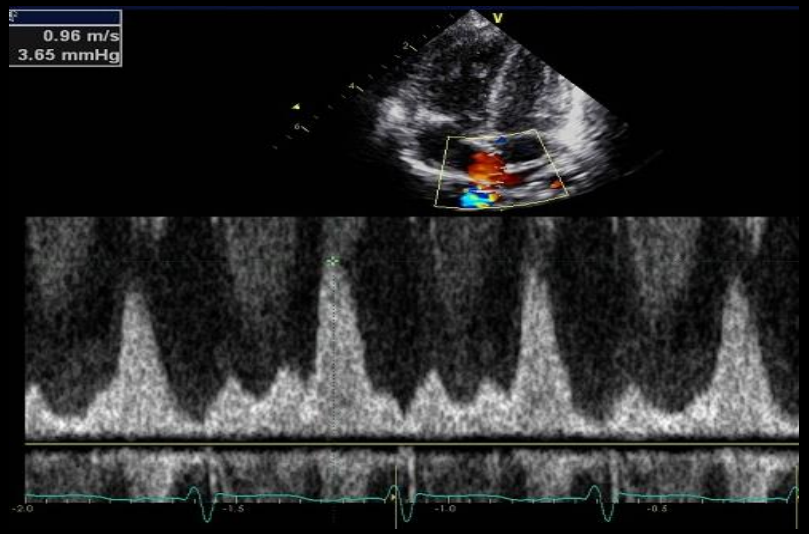


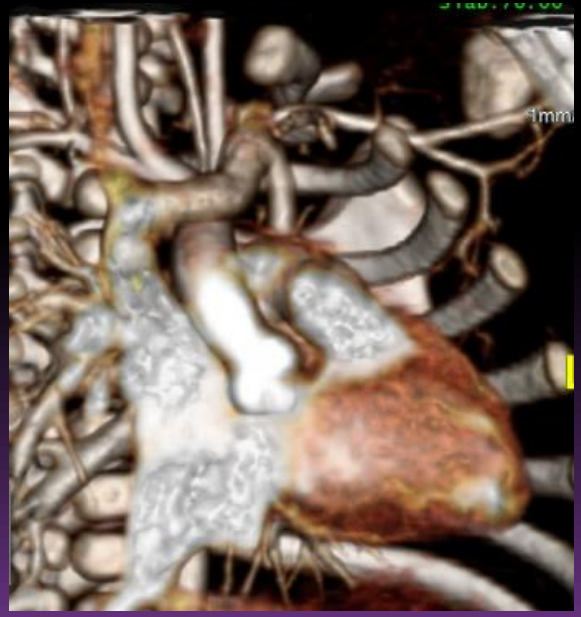



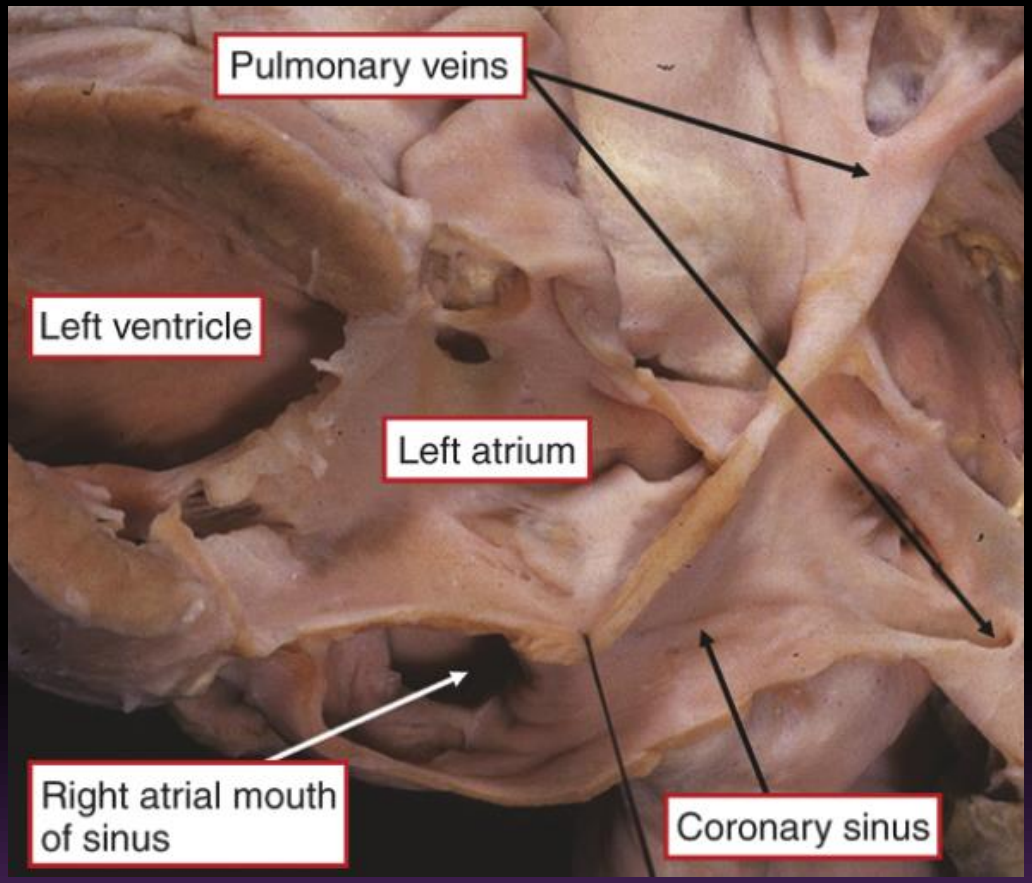
# TAPVC- Supracardiac and Cardiac

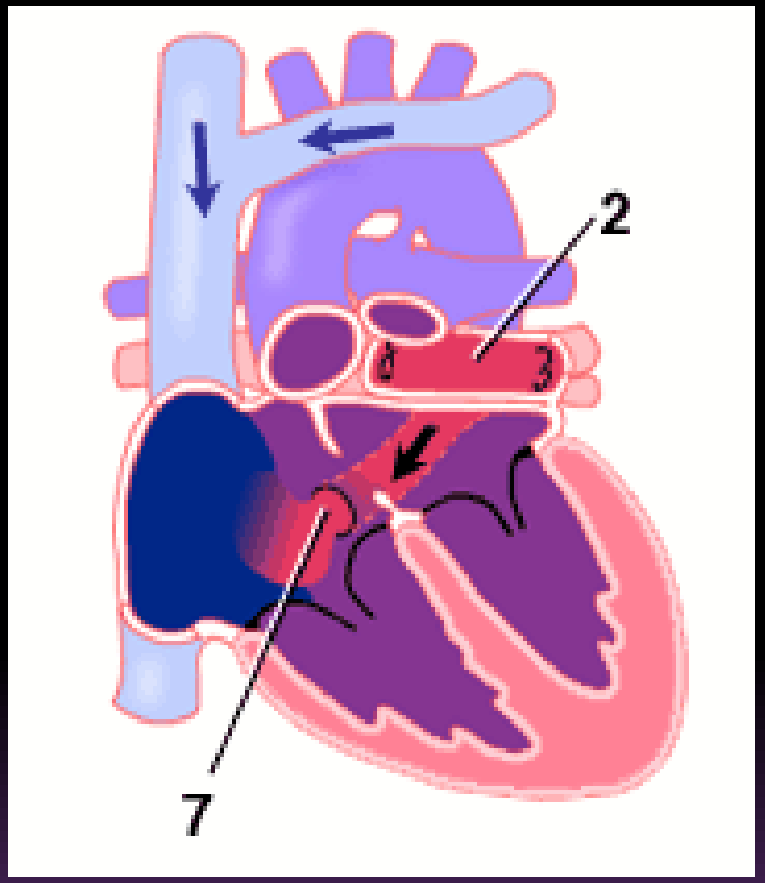




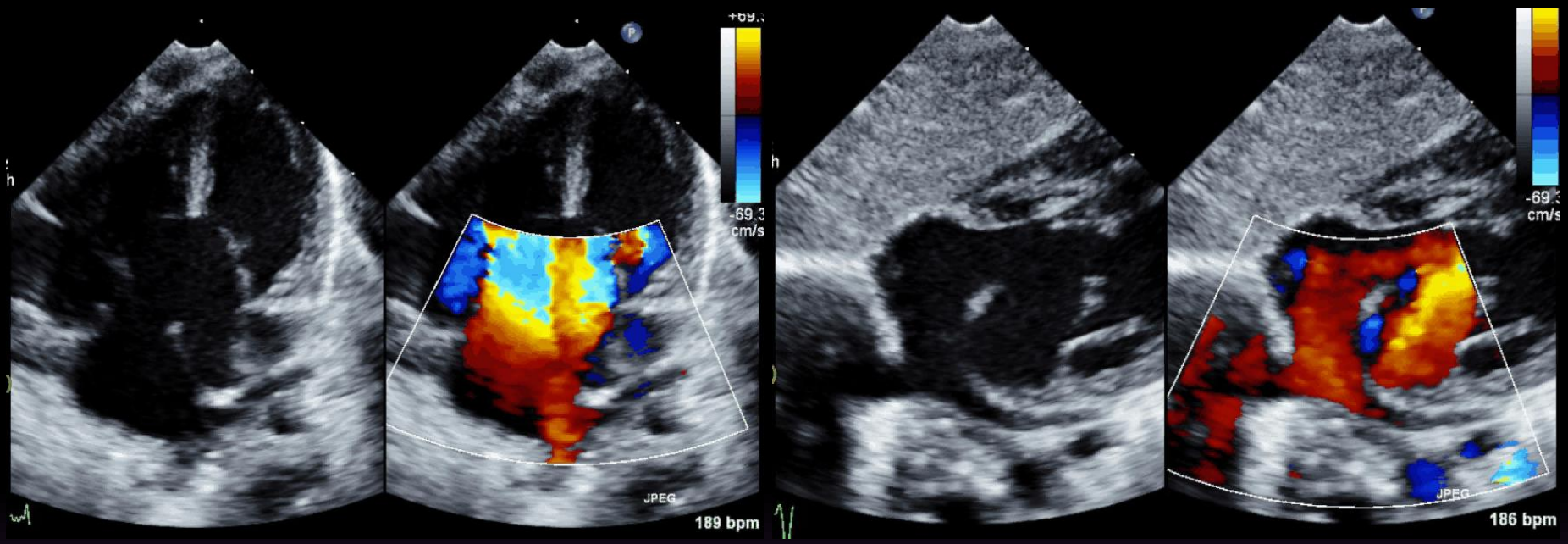


# TAPVC- Cardiac

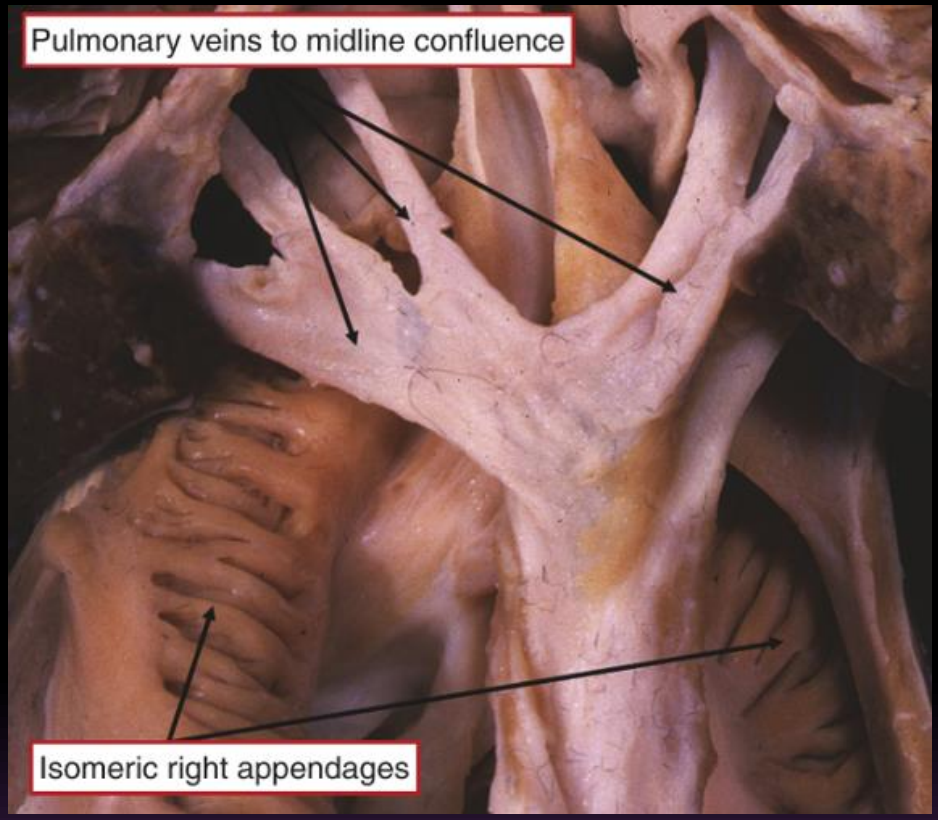


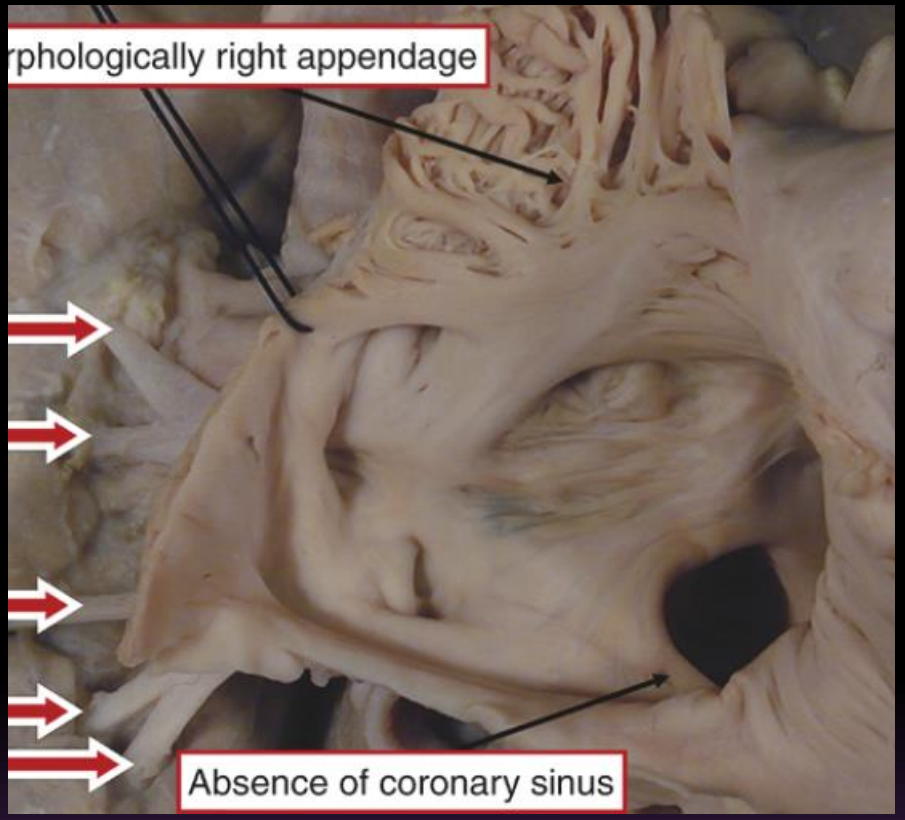



# TAPVC- Cardiac

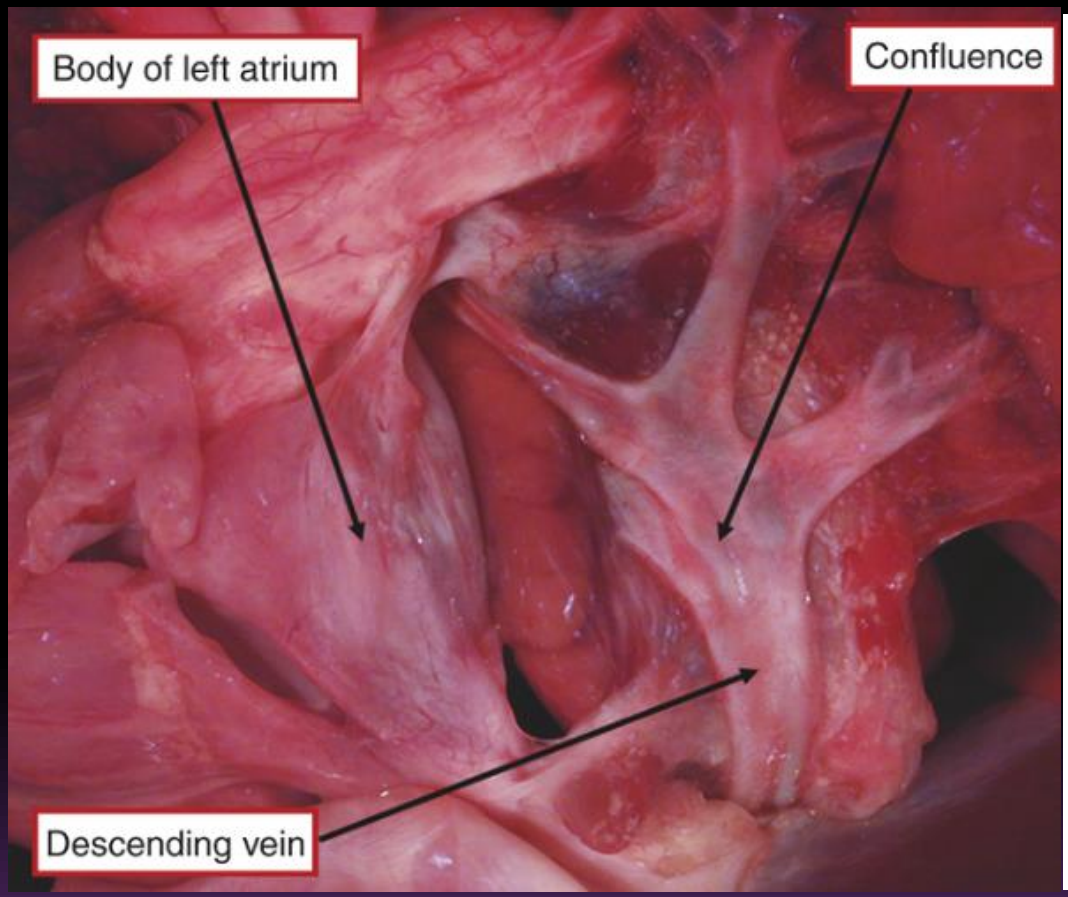


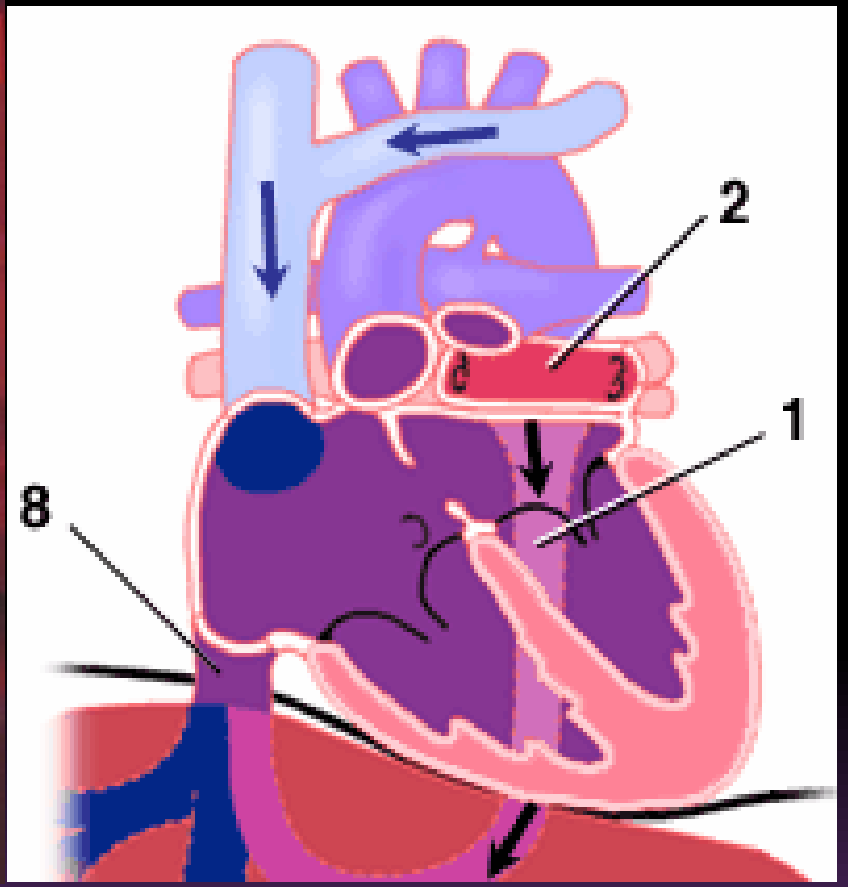
# TAPVC- Right Isomerism



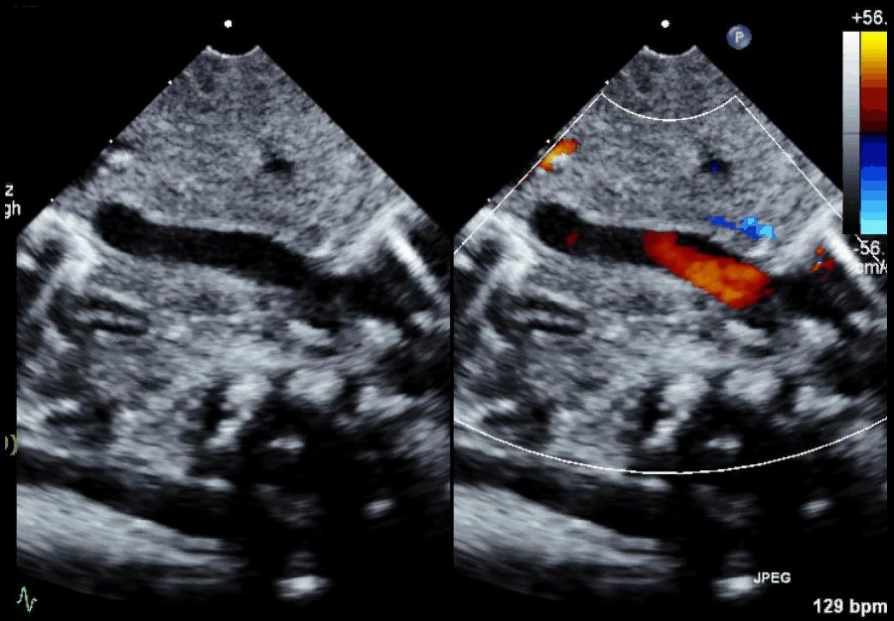



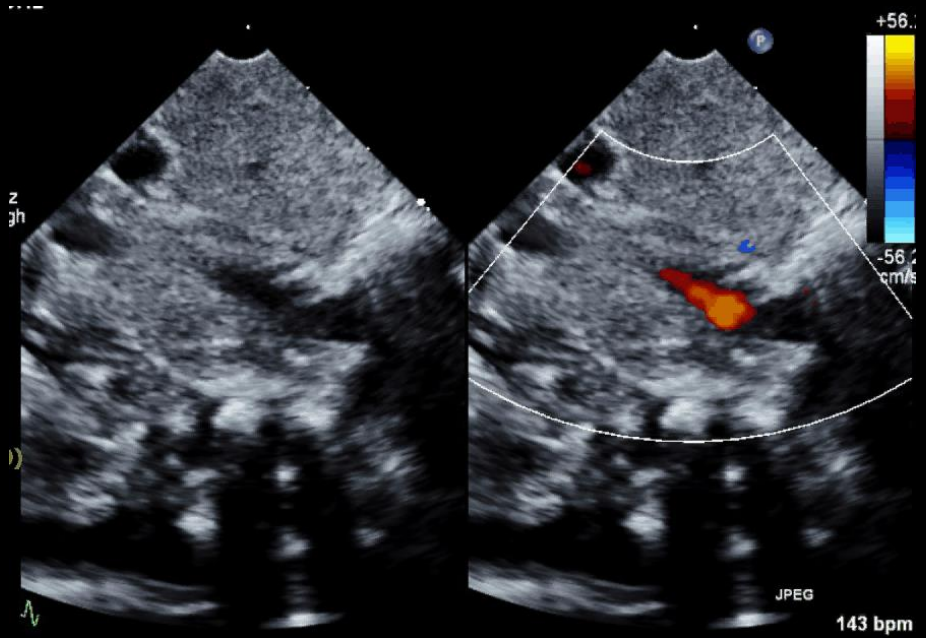
# TAPVC- Infracardiac

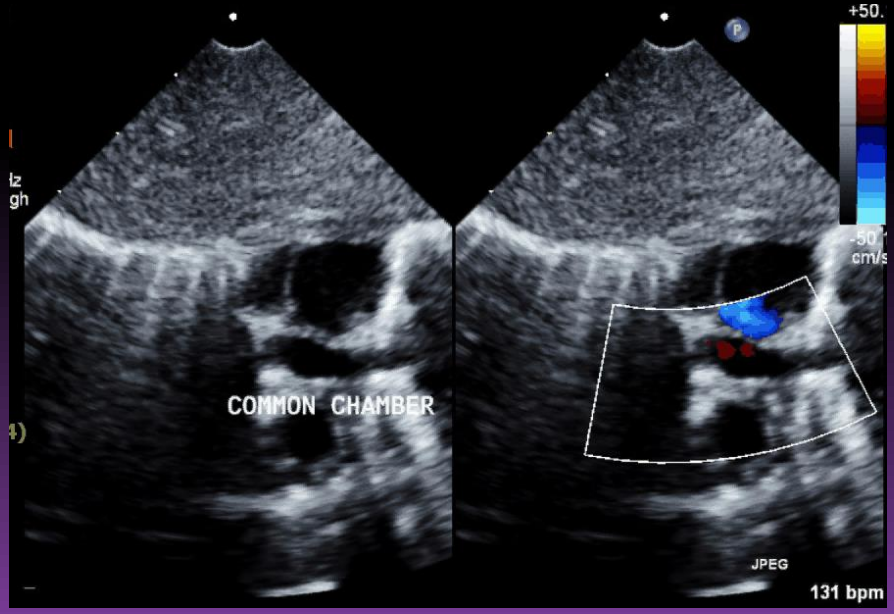


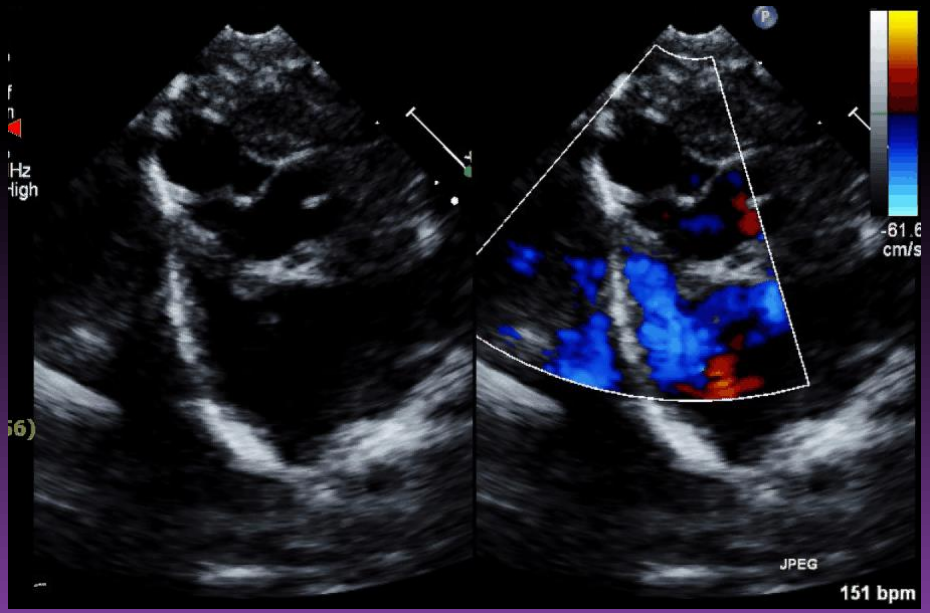



# TAPVC- Infracardiac



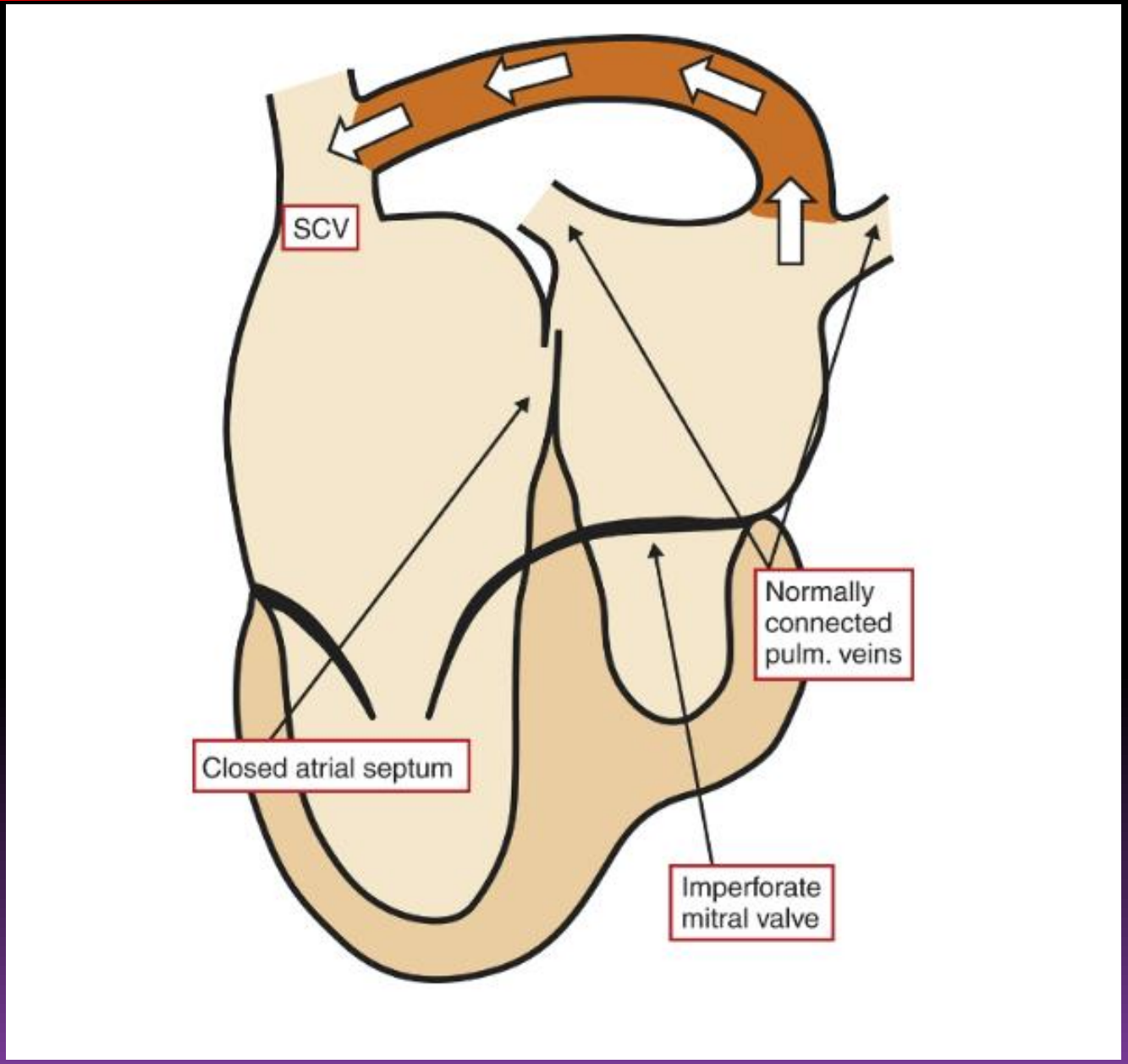



# TAPVC or not ?

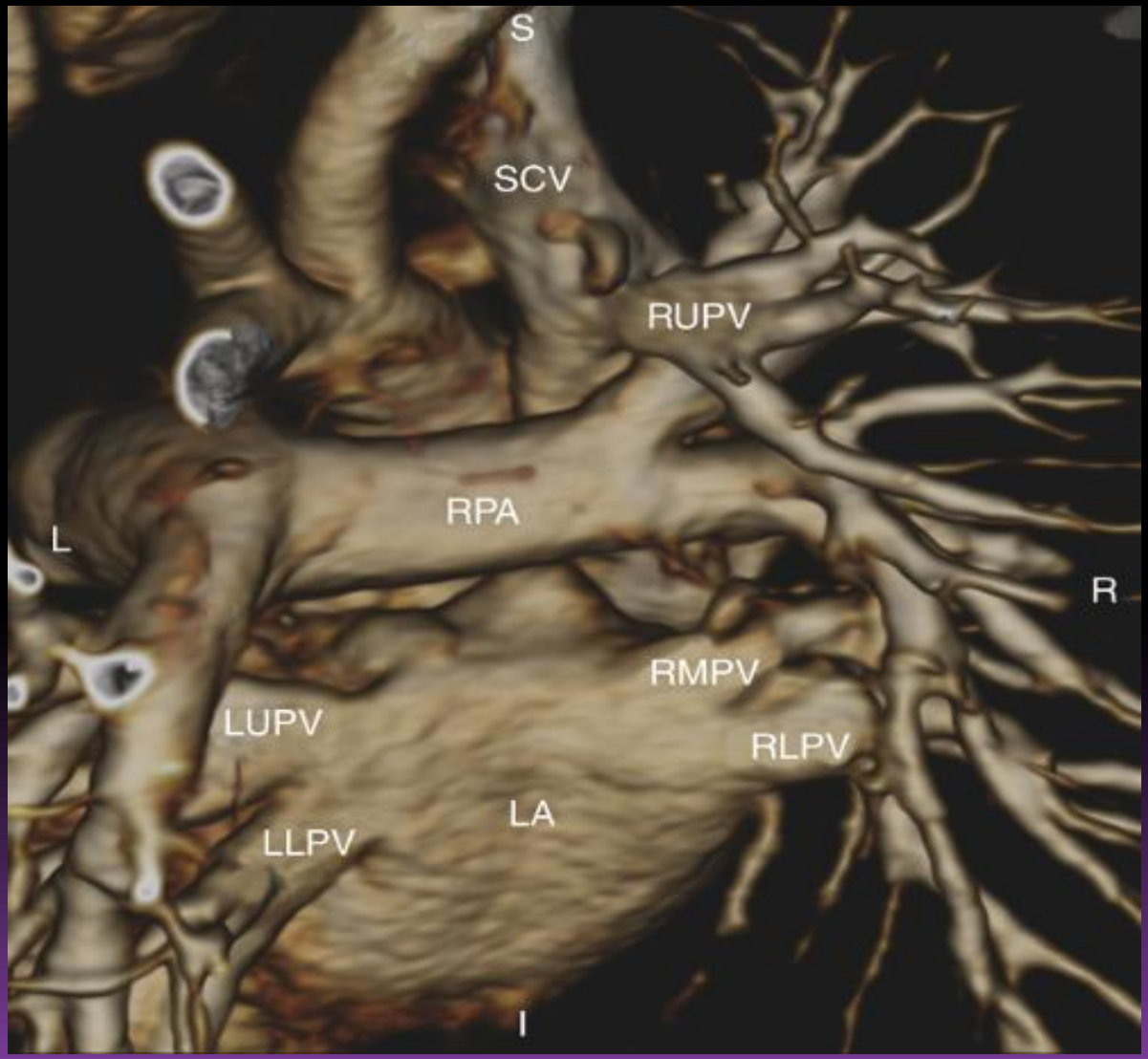
Differential Diagnosis

- Complete TGA with large VSD
- AVSD with common atrium
- Levoatrial cardinal vein associated with mitral atresia and intact atrial septum

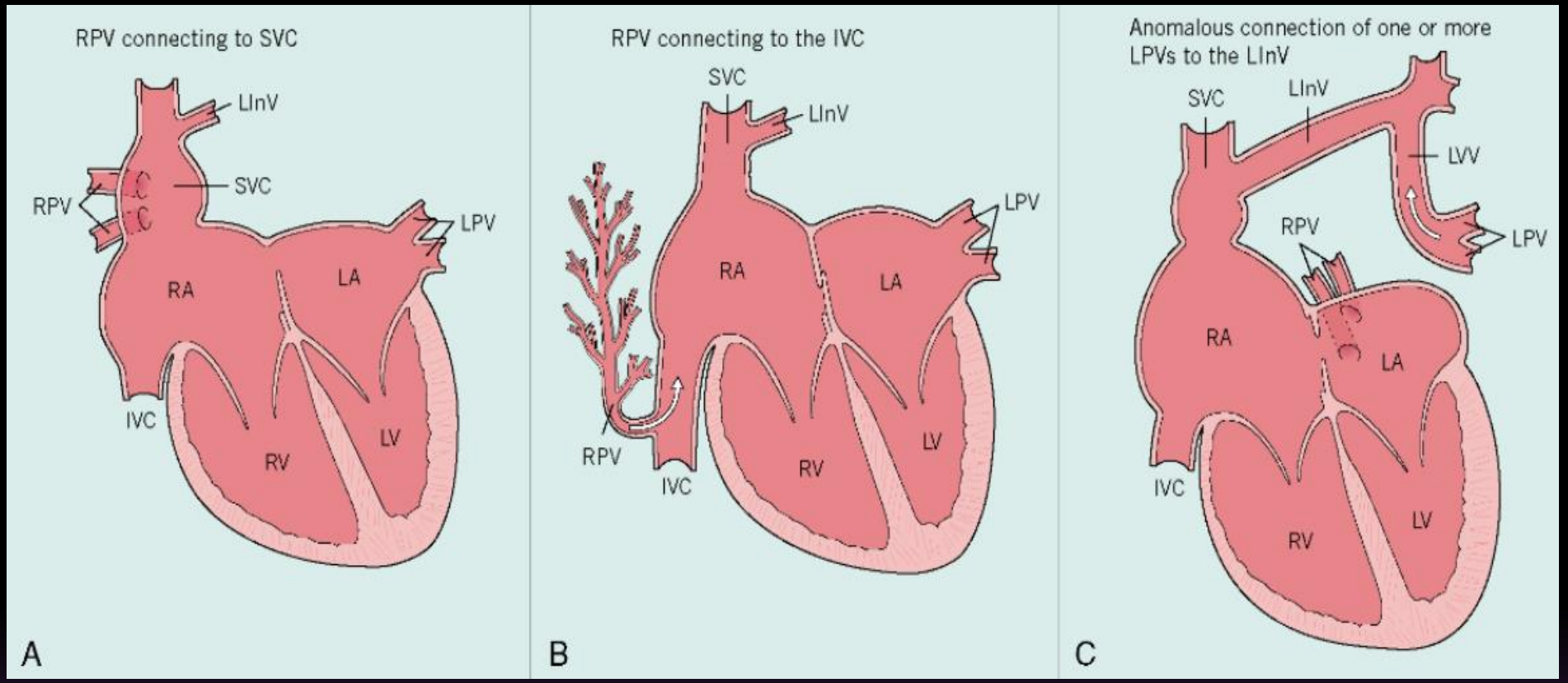
# Normal, but Abnormal Connection



# Partial Anomalous Pulmonary Venous Connection



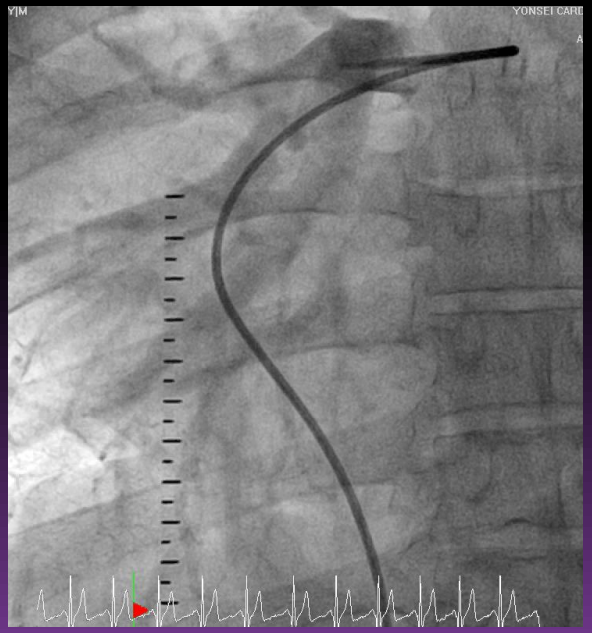
# Different Sites of Anomalous Connection

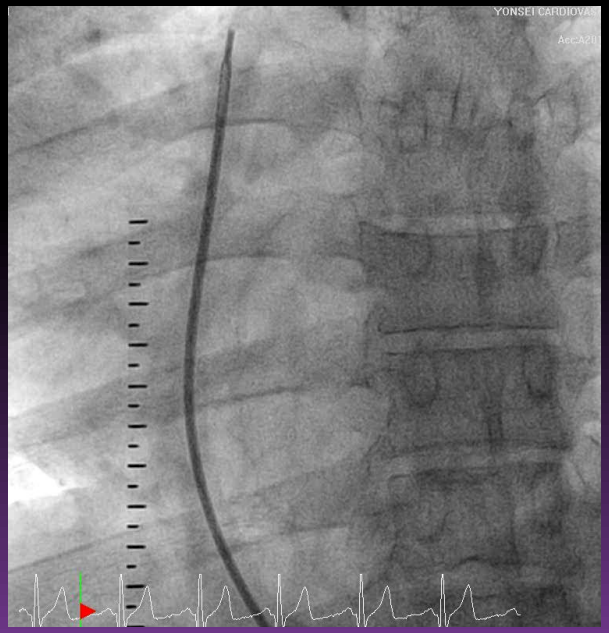


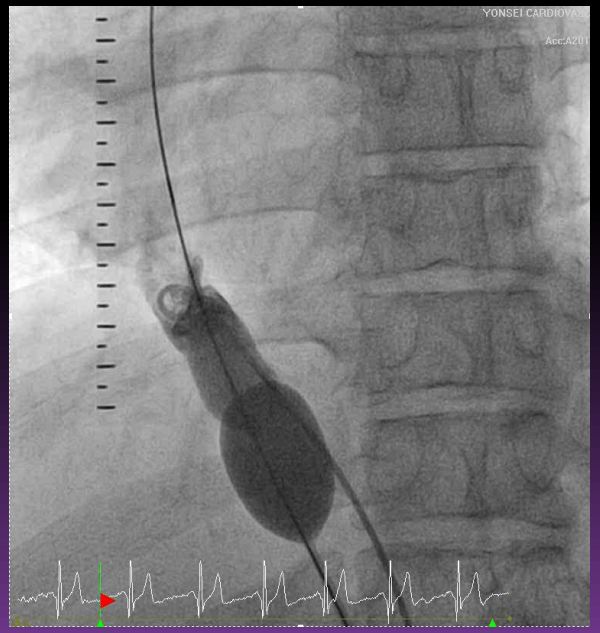

- Right pulmonary veins to right SVC or azygos vein
- Right pulmonary veins to right atrium
- Right pulmonary veins to IVC (Scimitar syndrome)
- Left pulmonary veins to the left innominate vein.

# CASE

- M/ 19 yr,  Palpitation, DOE
- RLL, RML, part of RUL drained by SCMV
  connected to the intrahepatic IVC
- Scimitar variant with non-restrictive connection
  of meandering vein to LA







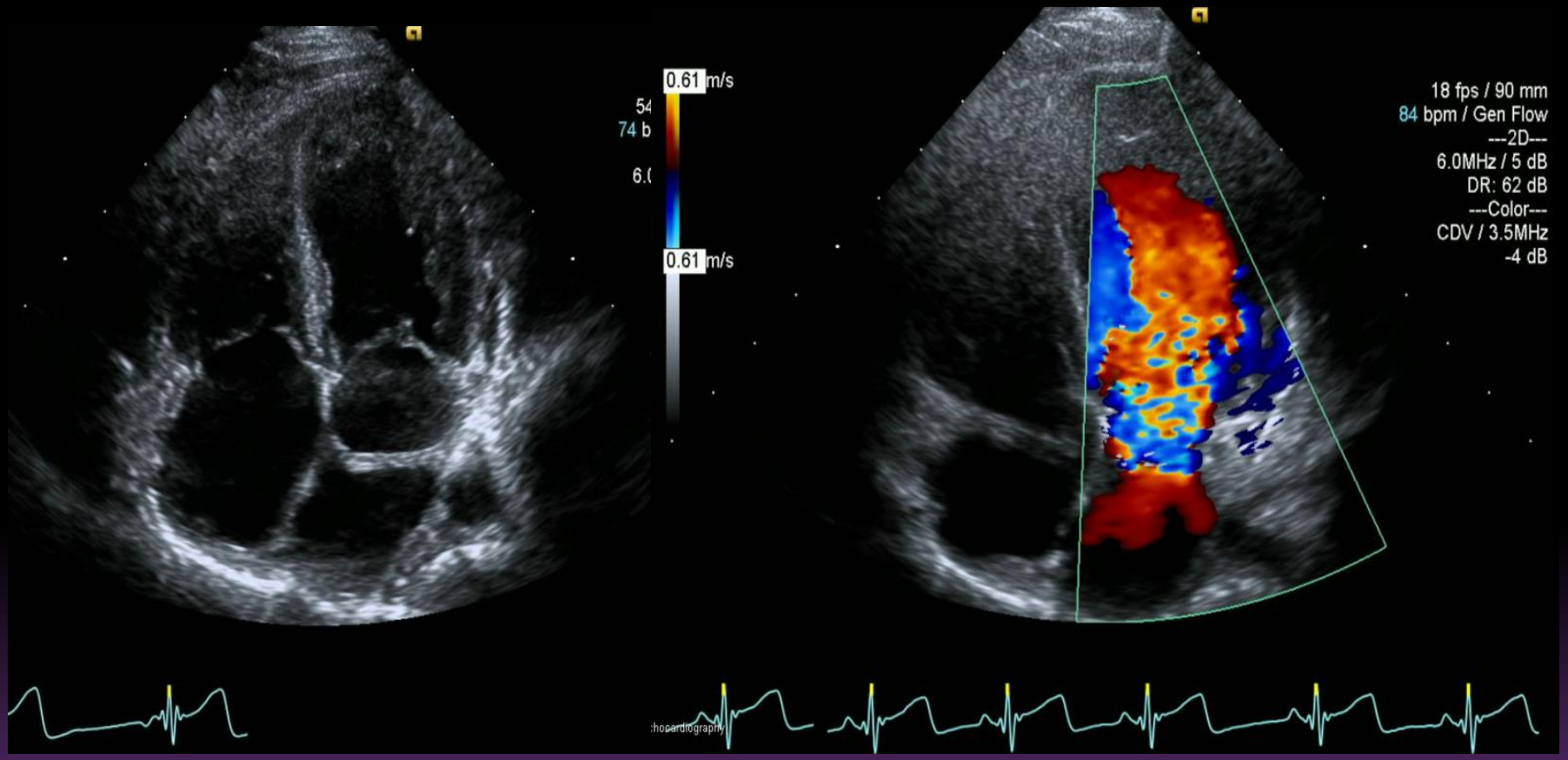
# Common Pulmonary Vein Stenosis - Cor Triatriatum

I. Accessory atrial chamber receives all pulmonary veins and communicates with left atrium
   - A. no other connection – classic cor triatriatum
   - B. other anomalous connection – RA or TAPVR

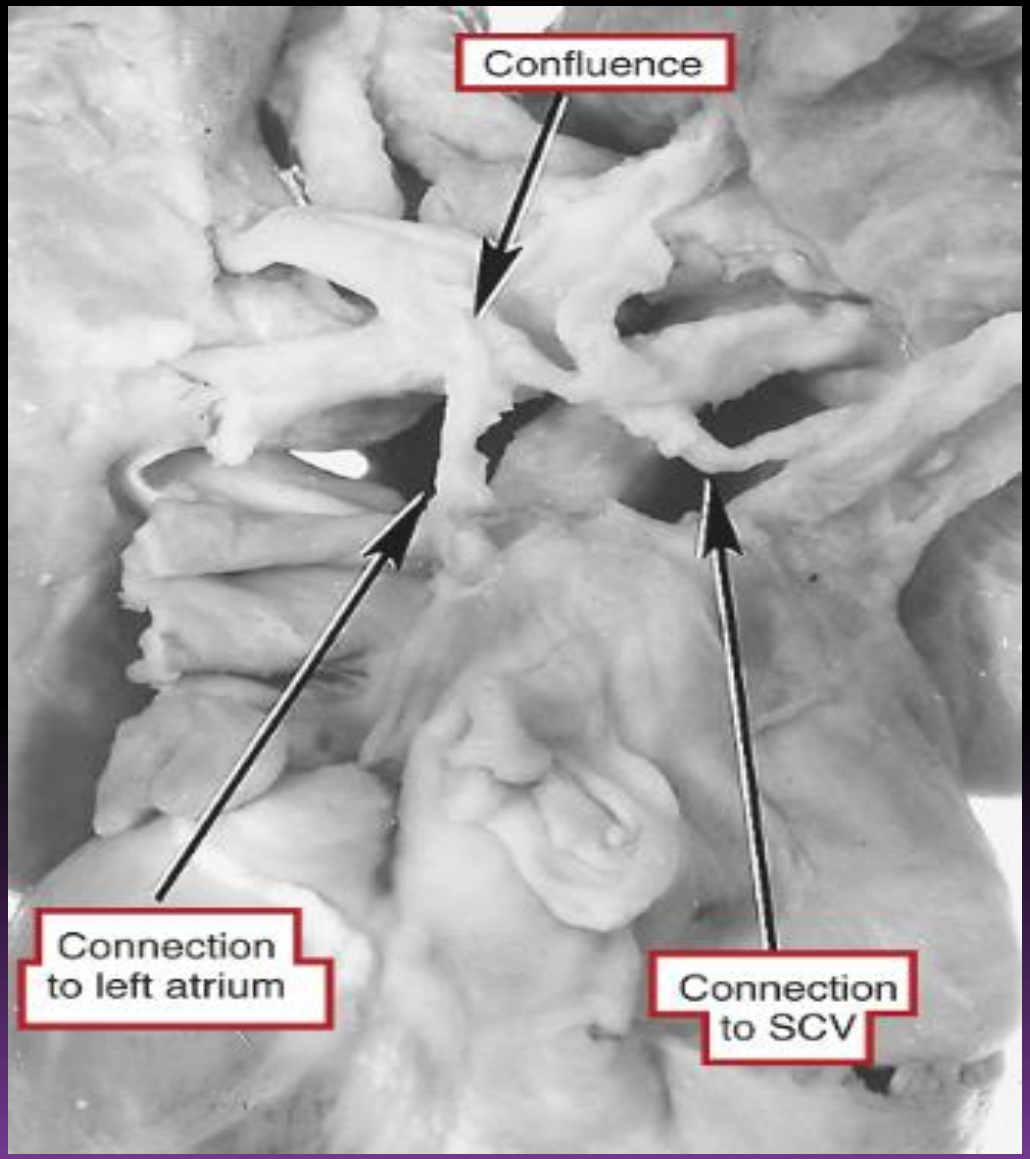
II. Accessory atrial chamber receives all pulmonary veins and does not communicate with left atrium
   - A. Anomalous connection to right atrium directly
   - B. With total anomalous pulmonary connection

III. Subtotal Cor triatriatum
   - A. Accessory atrial chamber receives part of pulmonary veins and connect to left atrium
   - B. Accessory atrial chamber receives part of the pulmonary veins and connects to right atrium

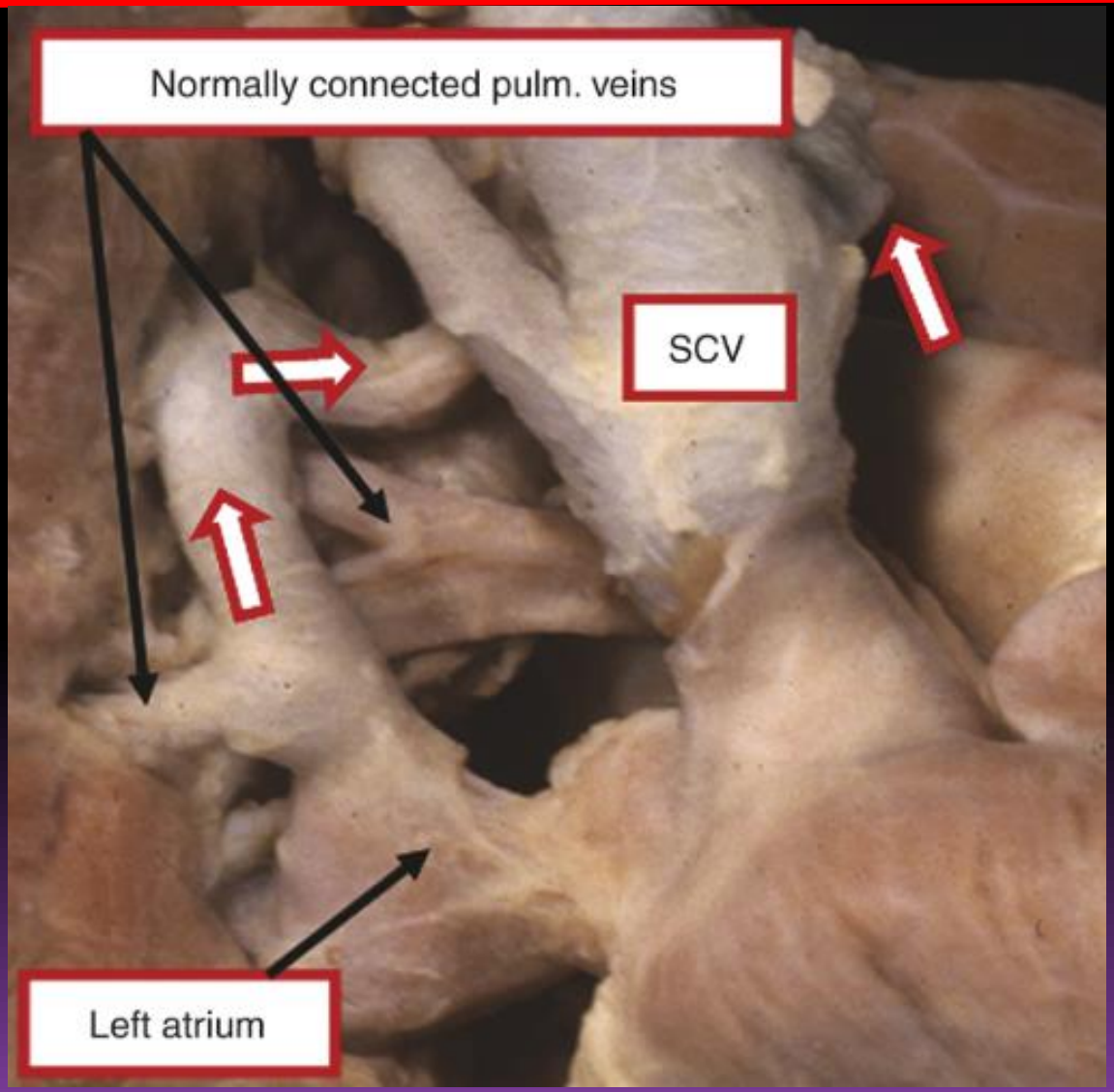
# Common Pulmonary Vein Stenosis - Cor Triatriatum



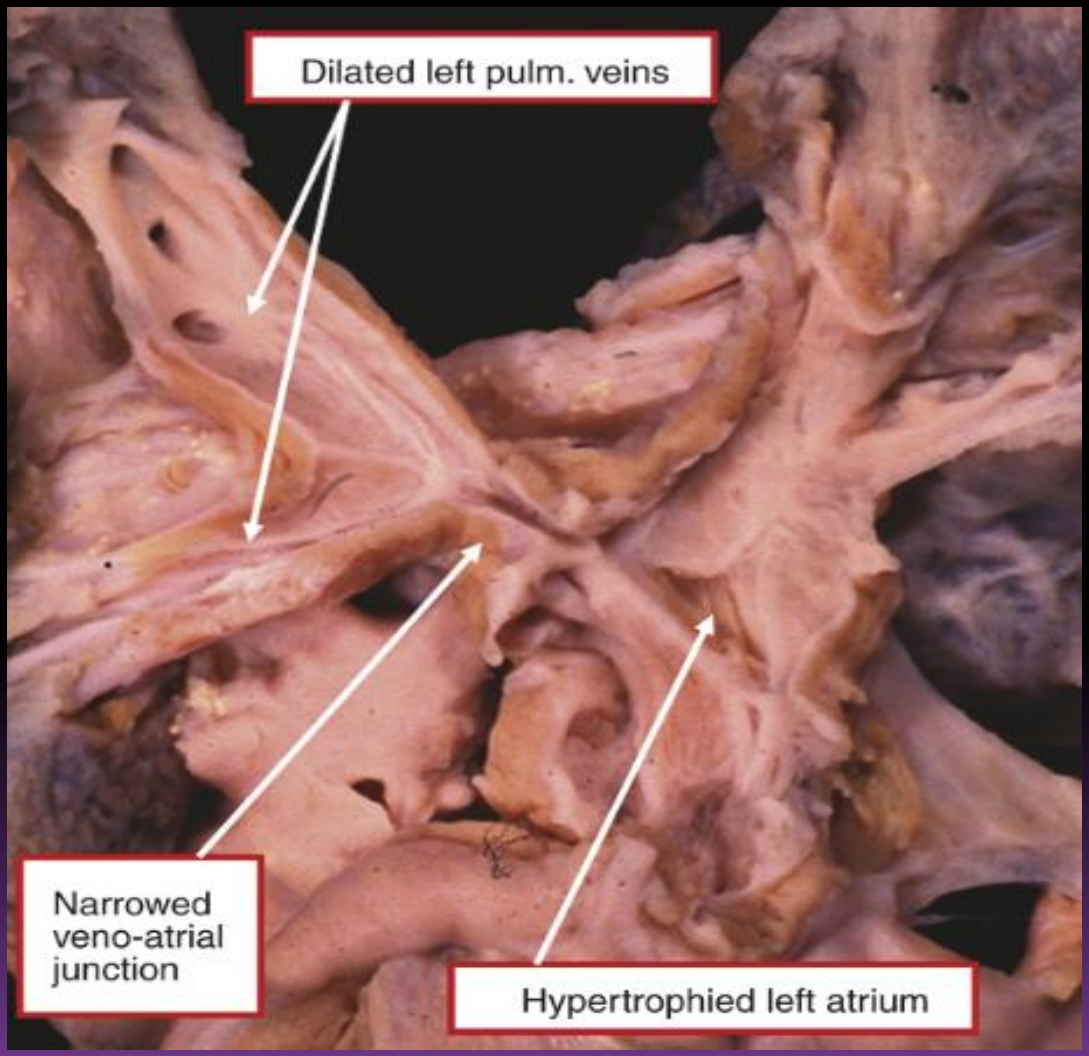
# Common Pulmonary Vein Atresia



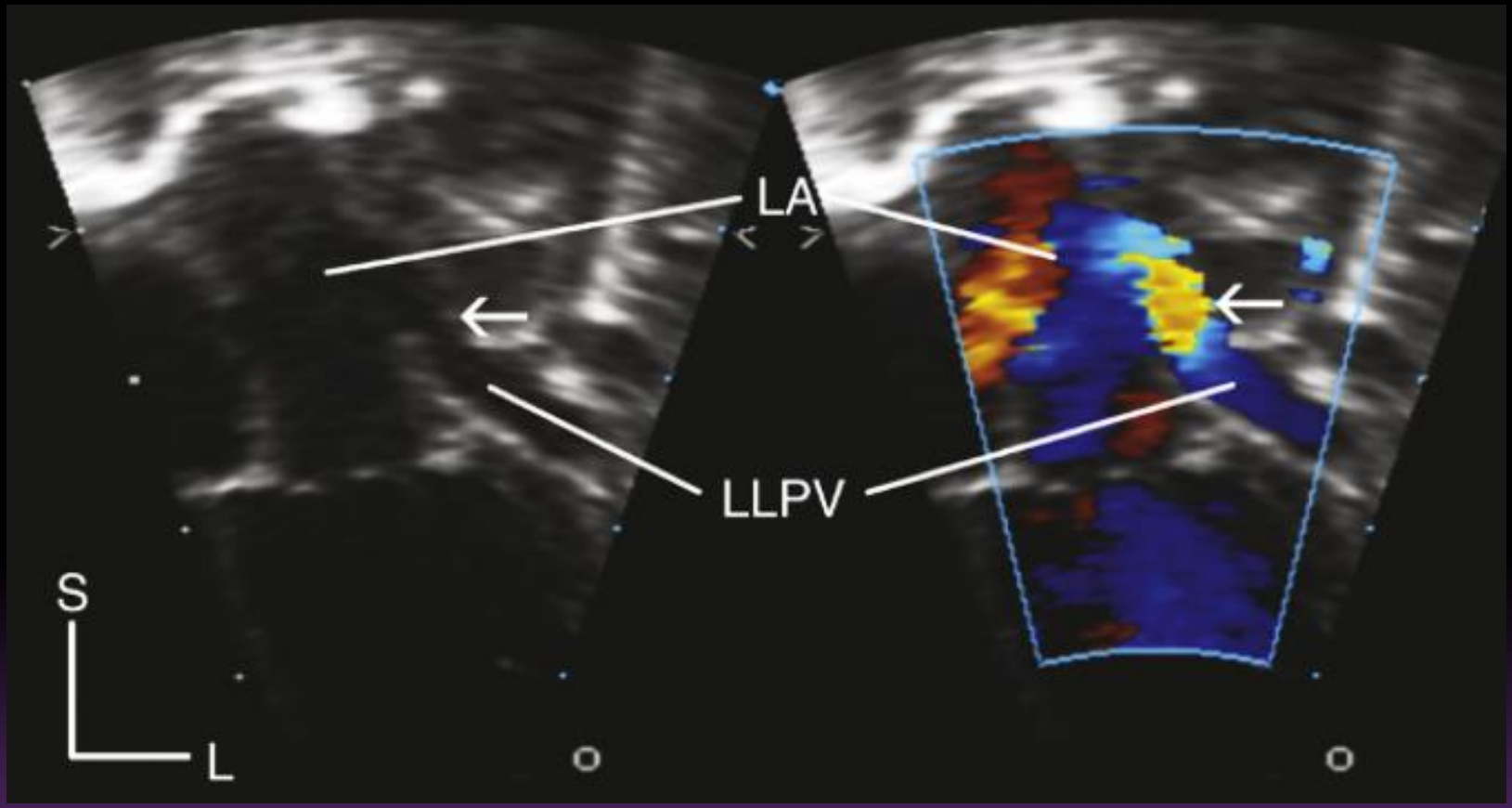
# Anomalous Pulmonary to Systemic Collateral Vein & Levoatrial Cardinal Vein



# Stenosis or Atresia of Individual Pulmonary Vein

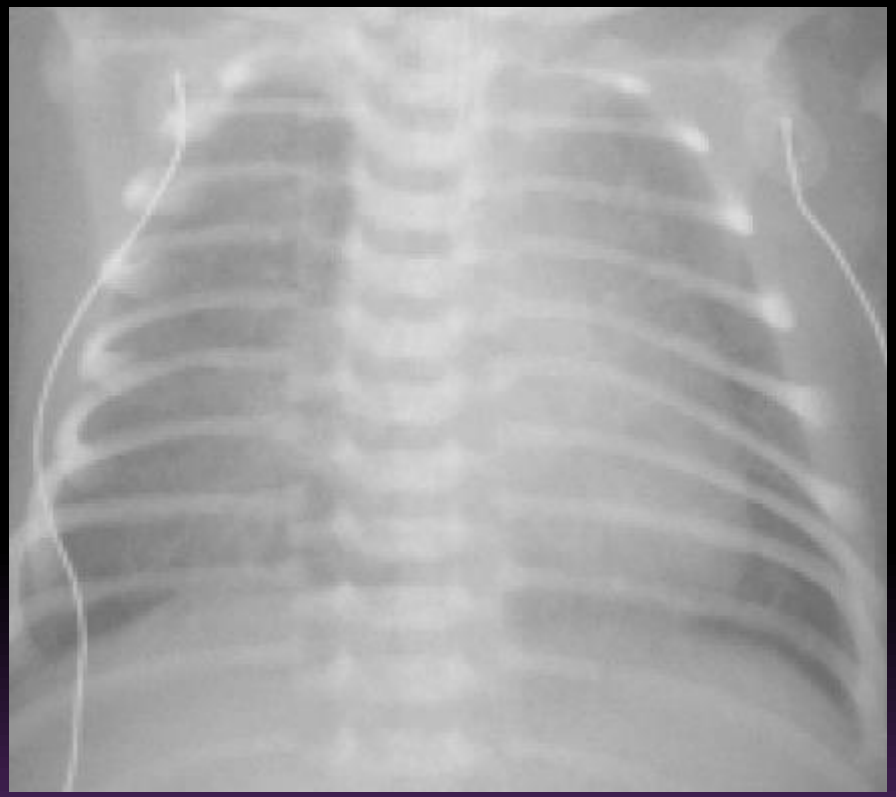


# Stenosis or Atresia of Individual Pulmonary Vein

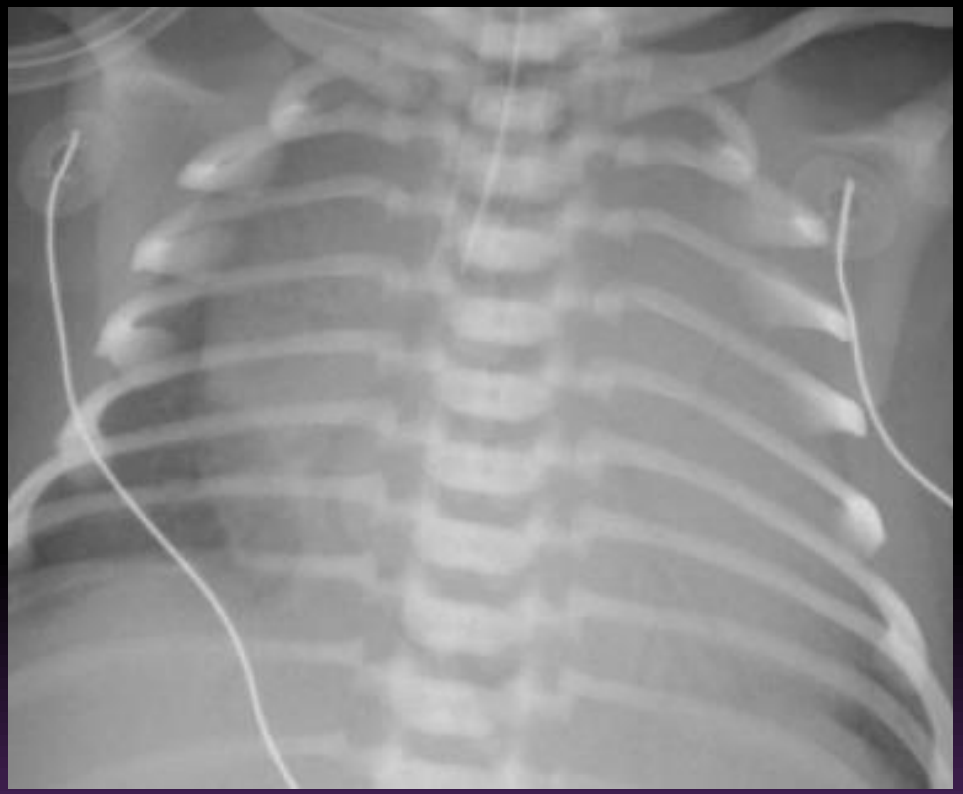


# CASE

- Baby at 2 hours after birth, tachypnea, cyanosis



Baby 1

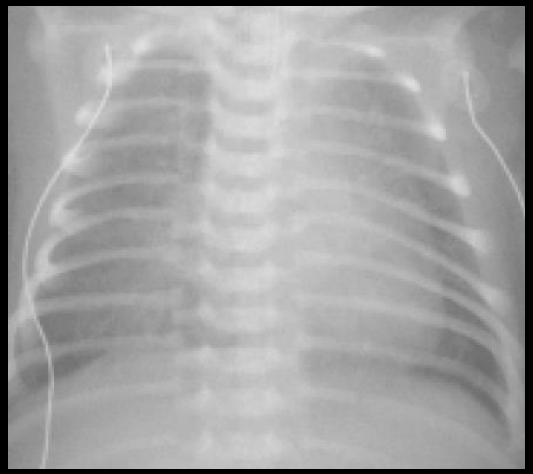


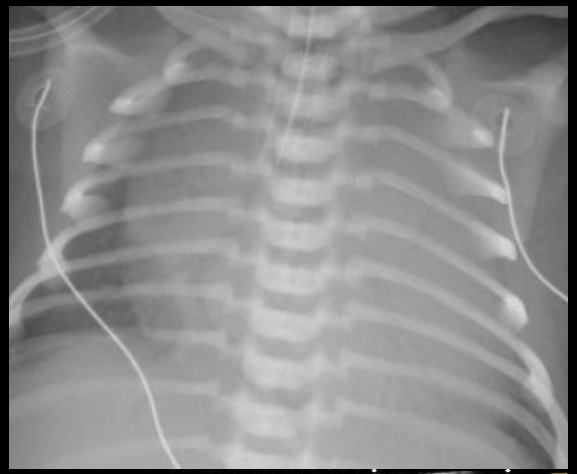
Baby 2

# CASE

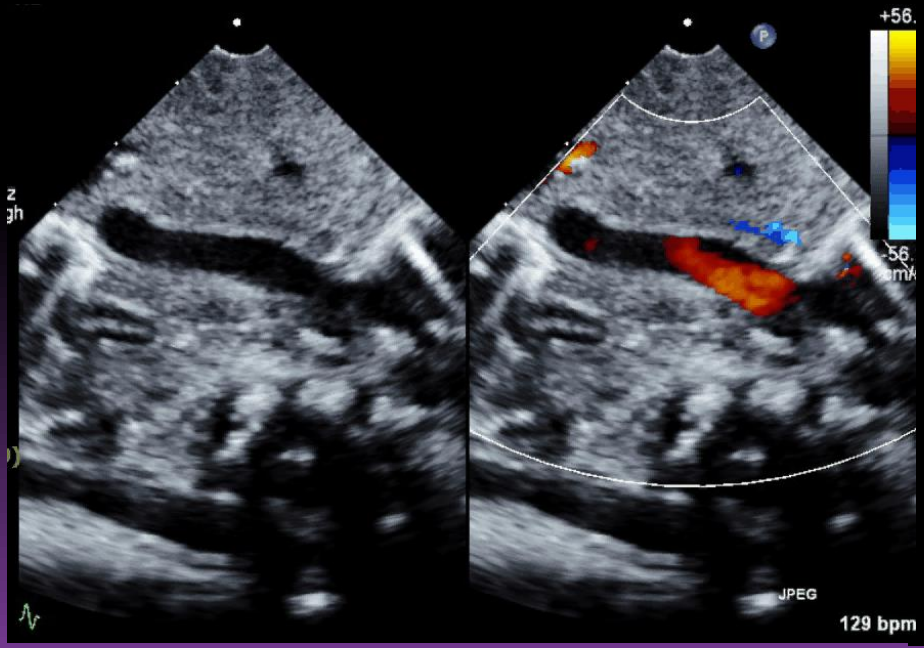
- Baby at 2 hours after birth, tachypnea, cyanosis

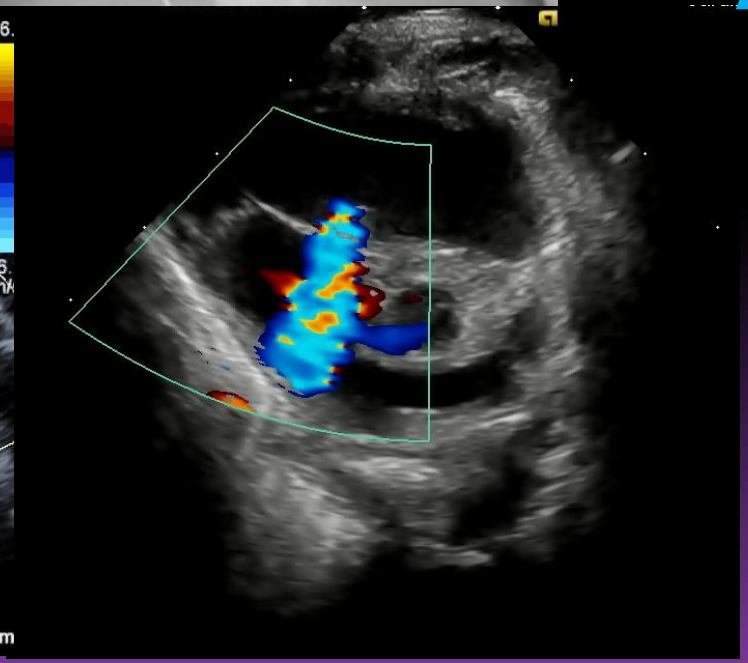
**Baby 1**





**Baby 2**



# Thank You !

# 감사합니다.