

# Pulmonary veins

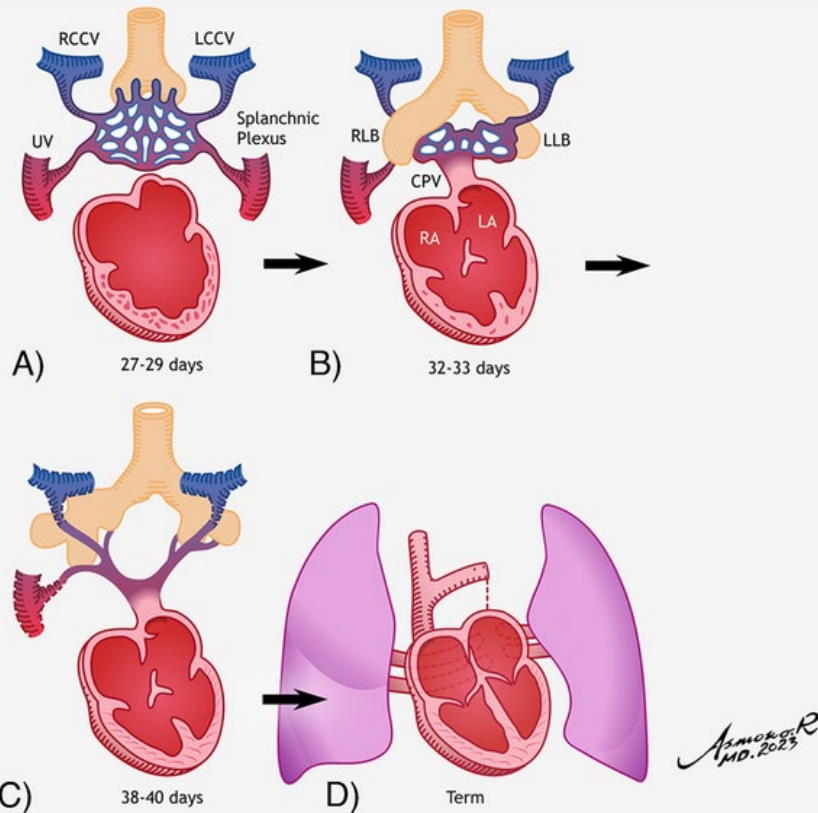
A “how to”

# DISCLOSURES

None

# Embryology

Simplified-ish



A: Lung buds connected to the splanchnic plexus. No connection to heart exists at this point. LCCV, left common cardinal vein; RCCV, right common cardinal vein; UV, umbilicovitelline vein.

B: Later in development, the common pulmonary vein (CPV) has evaginated from the left atrium (LA) and connected to the pulmonary venous plexus. The pulmonary veins are connected to the splanchnic plexus and the heart at this point in development. LLB, left lung bud; RLB, right lung bud.

C: The connections to the splanchnic plexus involute.

D: By term, the common pulmonary vein has become completely incorporated into the LA and the individual veins connect to the LA.

# Embryology

Very simplified

- At 32-33 days of gestation, the pulmonary veins subsequently establish a communication with the common pulmonary vein, which becomes incorporated into the posterior aspect of the developing left atrium
- If common pulmonary vein isn't absorbed by left atrium and a connection is created...
  - Blood needs to go somewhere-the cardinal vein or the umbilicovitelline vein remains

# Genetic associations

Most cases of pulmonary venous anomalies are sporadic

PAPVC

- Turner and Noonan syndromes

TAPVC

- Cat-eye, Holt-Oram and the asplenia(Heterotaxy)syndromes.

# Pulmonary venous abnormalities

- Cor triatriatum
- Abnormal number of pulmonary veins
- Sinus venosus defect
- Stenotic connections
- Atresia of the common pulmonary vein
- Partial anomalous pulmonary venous connection (PAPVC)
- Total anomalous pulmonary venous connection (TAPVC)

# PAPVC

- The most common form is the left upper pulmonary vein draining into the left innominate vein
- Other forms of PAPVC include
  - Pulmonary veins draining into: the SVC, coronary sinus, IVC, or azygos veins

# TAPVC

- Characterized by the abnormal connection of all four pulmonary veins, which eventually drain into the right atrium instead of the left atrium
- Entire oxygenated blood from the lungs returns to the right atrium
  - Atrial septal defect is crucial for blood mixing
  - Cyanosis occurs
- Complicated by pulmonary venous obstruction
  - Usually requires emergency surgery and is associated with severe neonatal morbidity and mortality
- More difficult to diagnose in a fetus vs a neonate or infant

# TAPVC without obstruction

- Supracardiac and cardiac type TAPVC are usually non obstructed\*
- At birth, the pulmonary and systemic vascular resistances are similar. In early weeks of life, there is a significant drop in pulmonary vascular resistance, leading to increased recirculation of flow in the lungs causing pulmonary over circulation
- Patients can be asymptomatic at birth but will become tachypneic and have feeding difficulties-if the atrial communication is adequate
- Leads to pulmonary hypertension and right heart failure

# TAPVC with obstruction

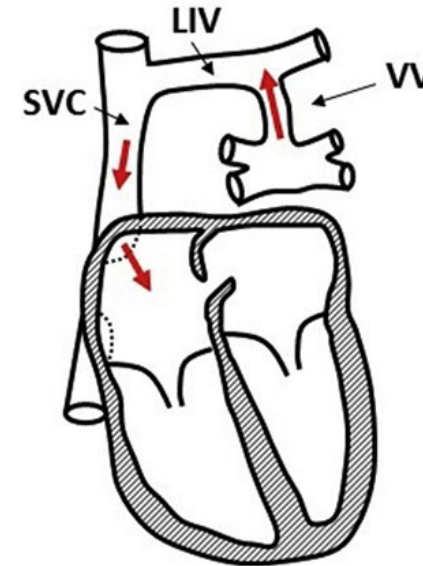
- Infradiaphragmatic is the most common type of obstructive TAPVC
- Pulmonary venous obstruction can occur at the level of the diaphragm, vein entering the IVC, or in the hepatics. Elevated pressures in the pulmonary venous system are transmitted to the lung bed leading to pulmonary edema
- Leads to pulmonary vascular resistance, pulmonary hypertension, right ventricular hypertrophy, and right-sided heart failure
- Presents in the neonatal period as severe respiratory distress

# Supracardiac

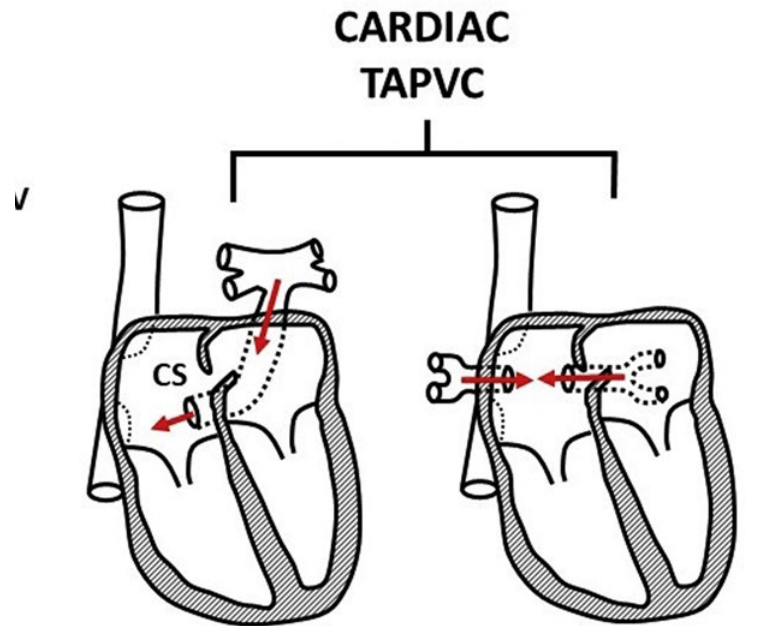
Pulmonary veins return to a confluence behind left atrium which goes to a vertical vein and drains to:

- Innominate\*
- Azygous
- Superior vena cava

## SUPRACARDIAC TAPVC



# Cardiac

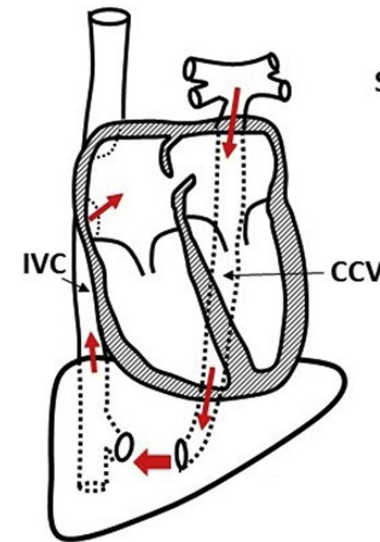


Pulmonary veins drain into:  
Coronary sinus\*  
Right atrium

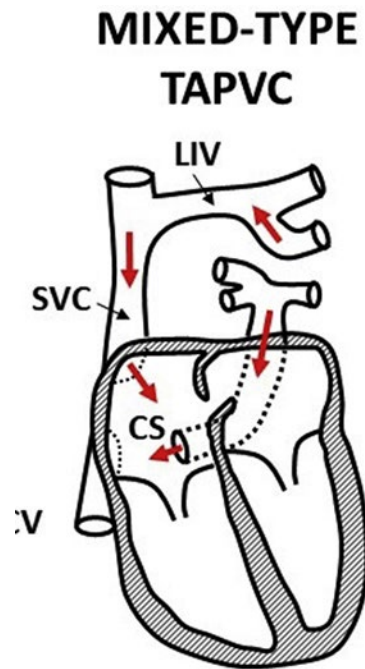
# Infradiaphragmatic

Pulmonary veins drain to a confluence into a vertical vein which goes below diaphragm and drains to inferior vena cava

INFRACARDIAC  
TAPVC

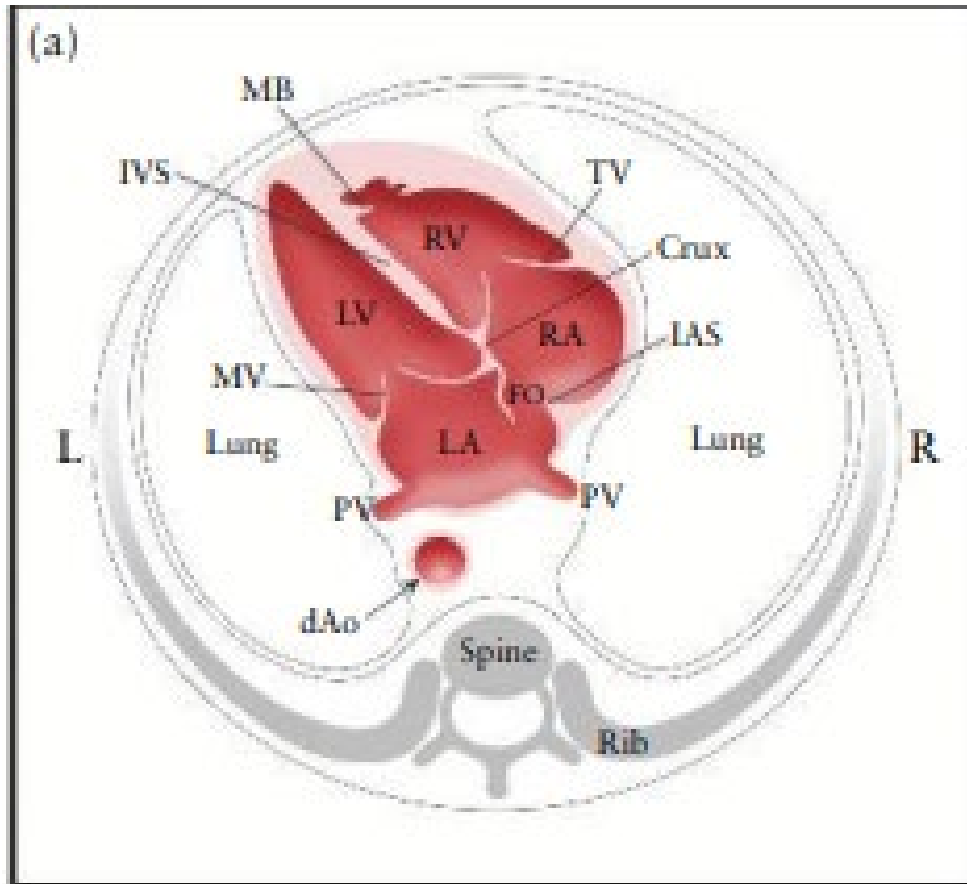


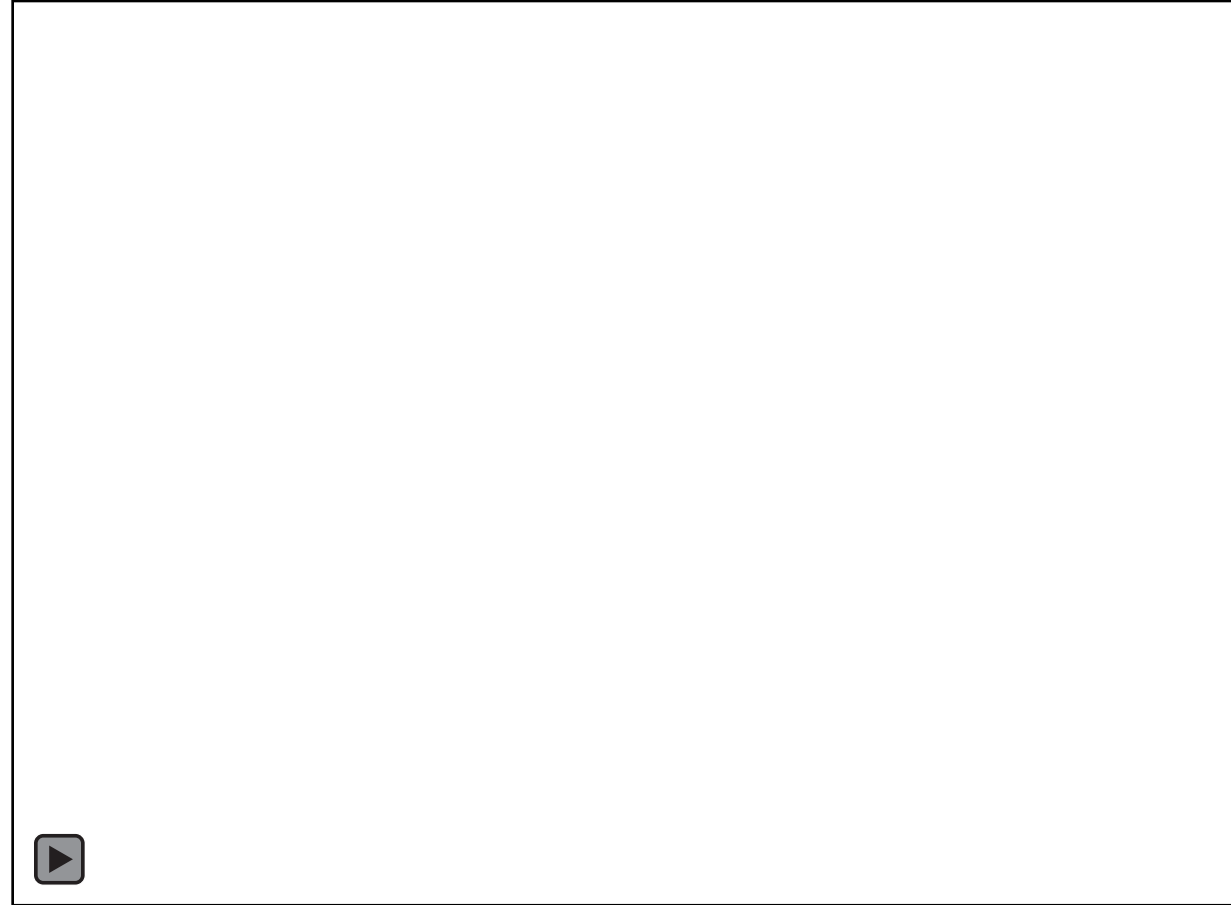
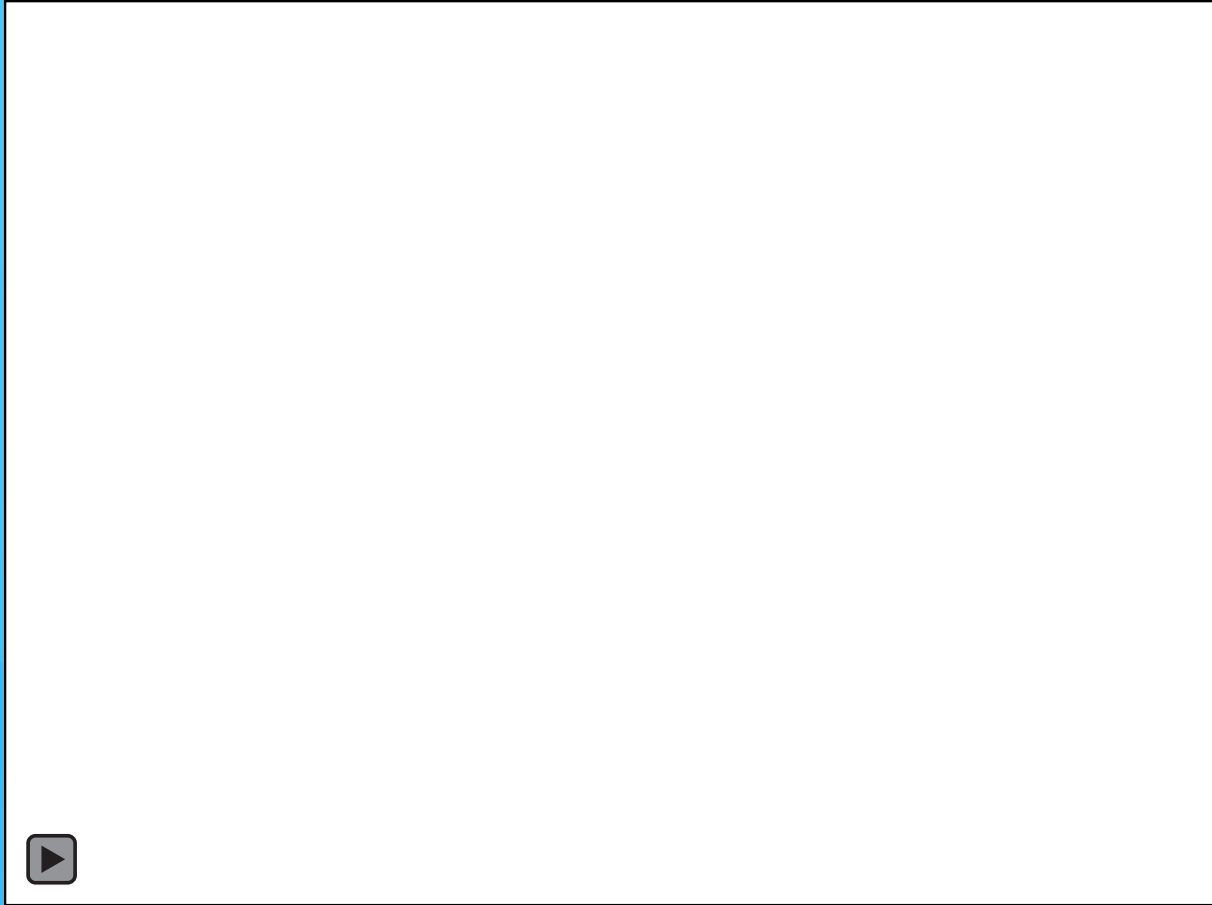
# Mixed



Any combo

# Know your normal

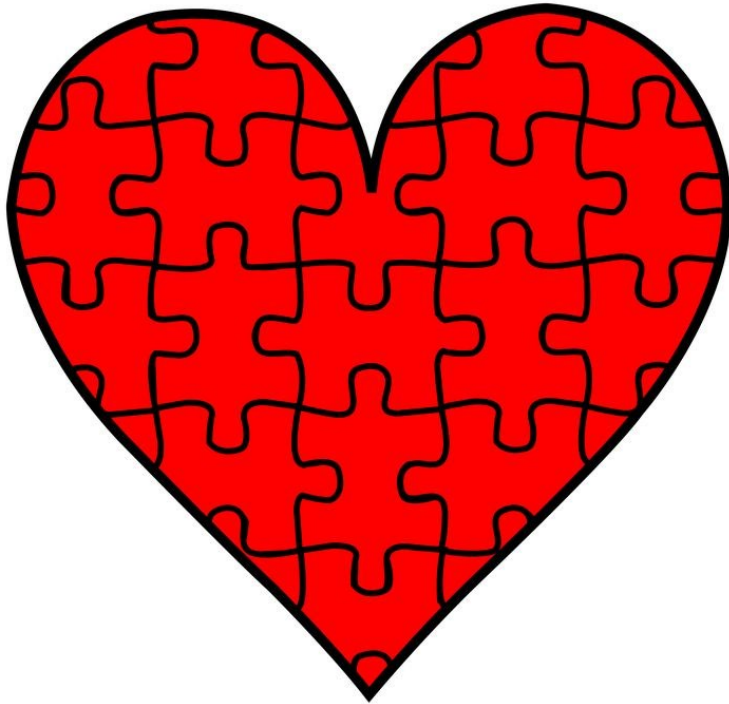




# Tips

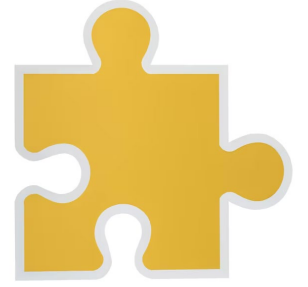
- Zoom-with landmarks still visible
  - Color scale down
  - Gain up
  - Smooth
  - Dual screen
- Check for landmarks
    - IAS
    - CS(dilated?)
    - MV
  - One vein from each side
  - Find the PAs

# The puzzle pieces

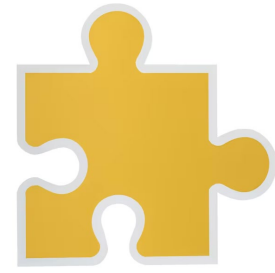


- IVC/ SVC dilation
- Right sided dilation
- Small left atrium
- Abnormal distance between aorta and left atrium
- Extra circles
- A funny flow...

# Four chamber view

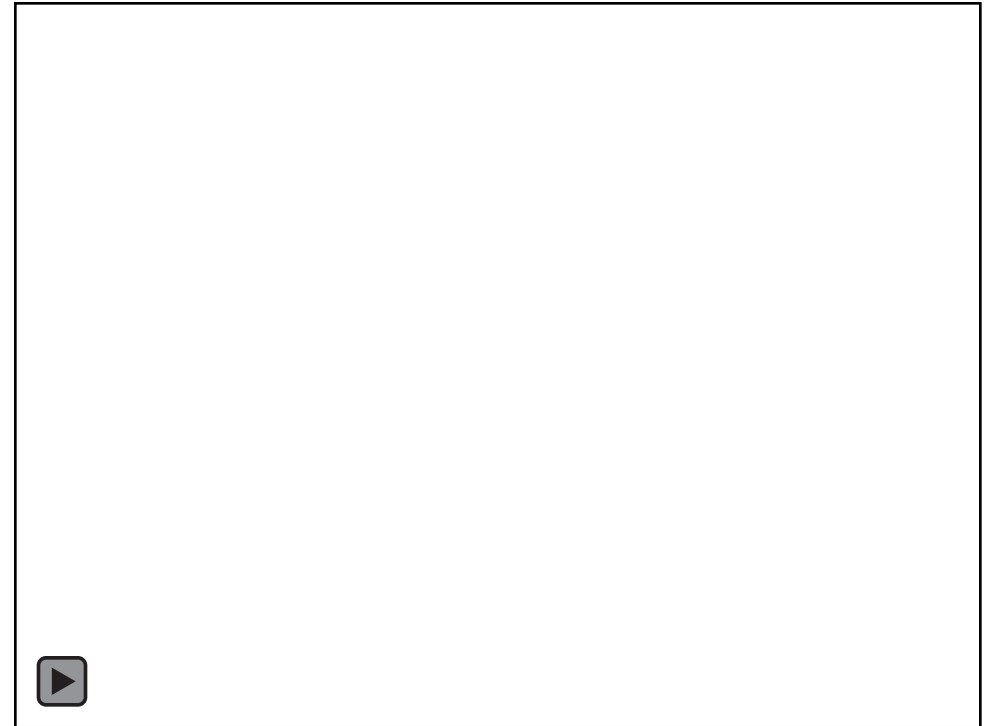


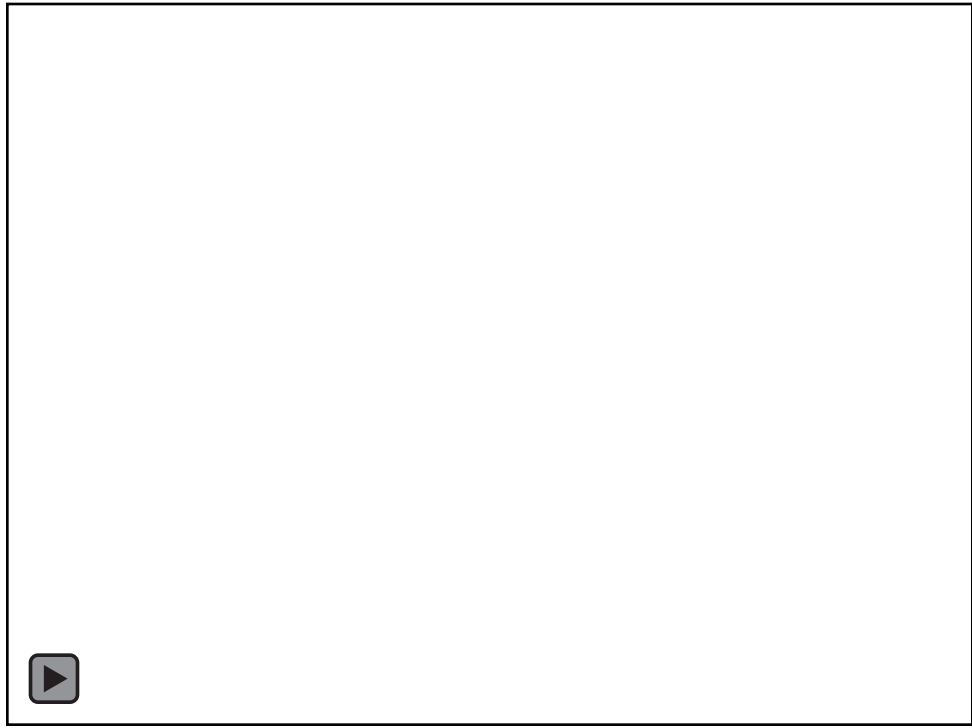
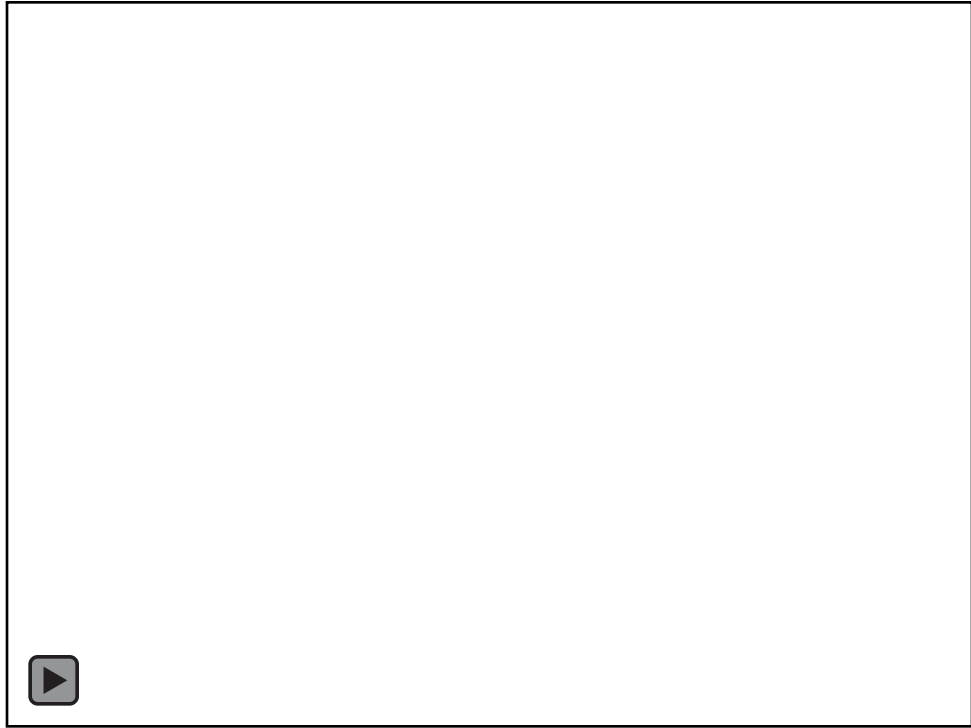
- Clues!
  - Extra circles
  - Distance between the aorta and left atrium
  - Right/ left discrepancy
  - Small left atrium
  - Smooth walled atrium
    - Coumadin ridge?
  - Extra:
    - 2d of left vein

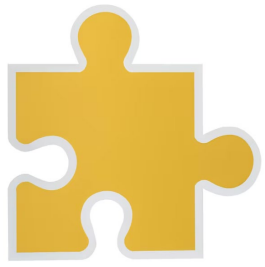


# Coumadin ridge??

- Part of the left atrium that lies between the left atrial appendage and left upper pulmonary vein
  - Described as tissue/ pericardial folding
  - The presence usually indicates that the pulmonary venous drainage is normal

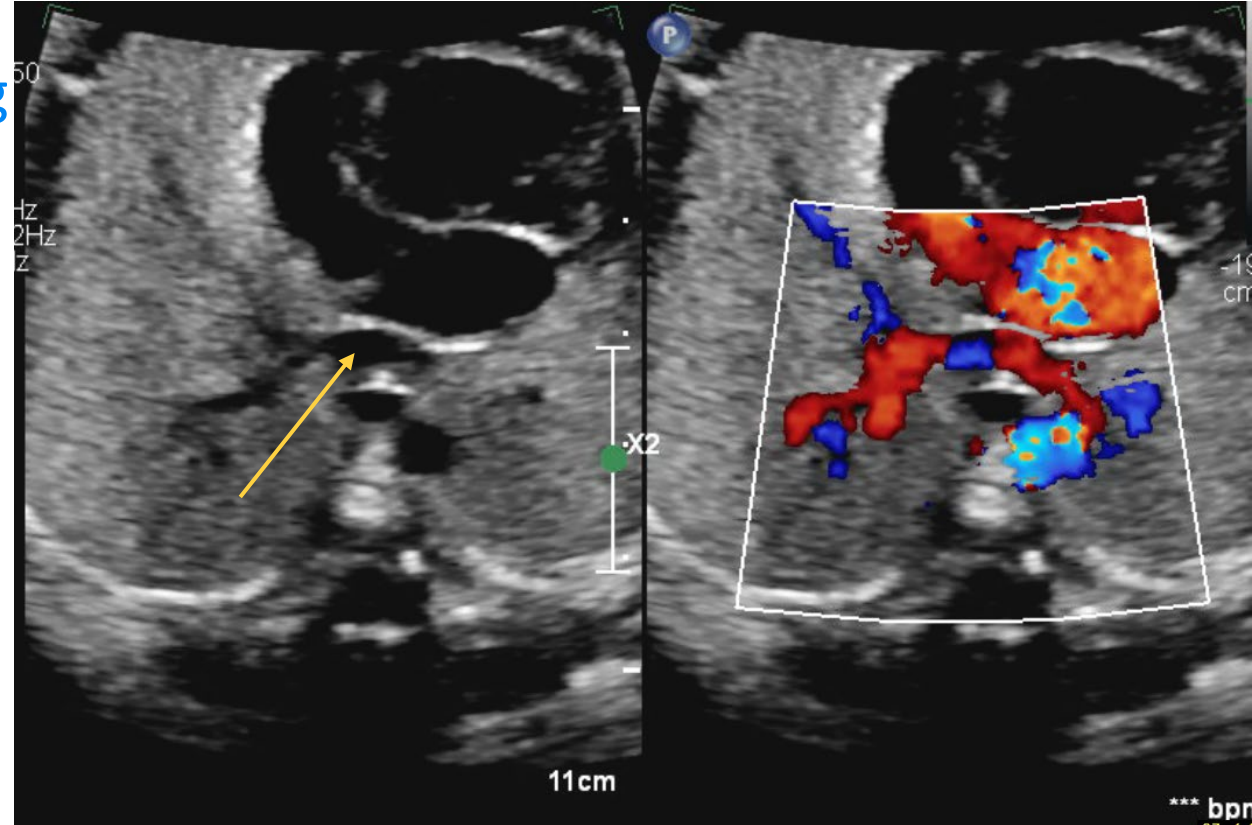
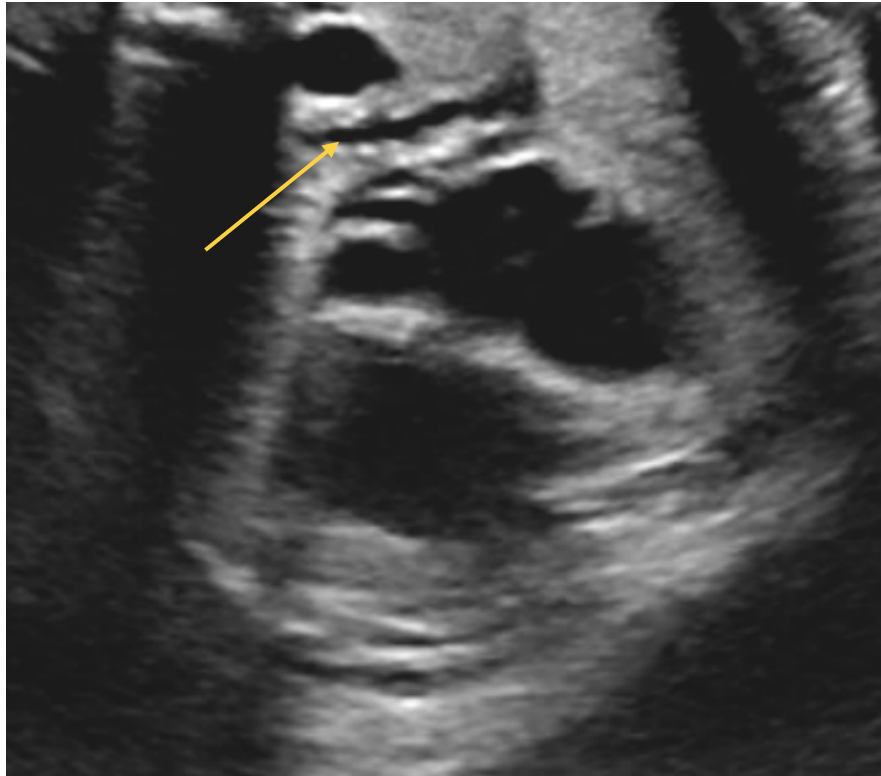


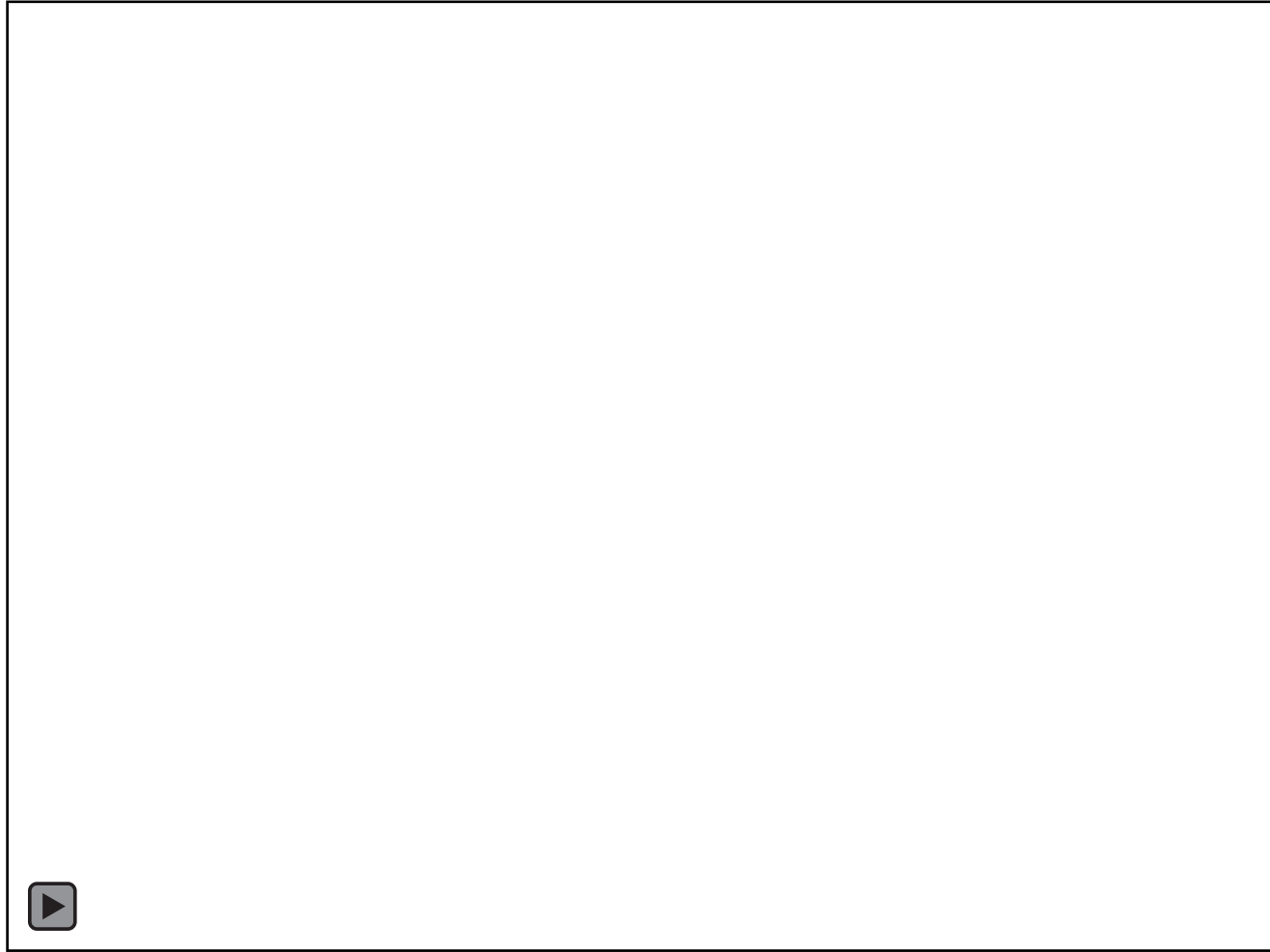




# The “twig sign”

- Bonus four chamber puzzle piece
- The posterior chamber resembling a twig





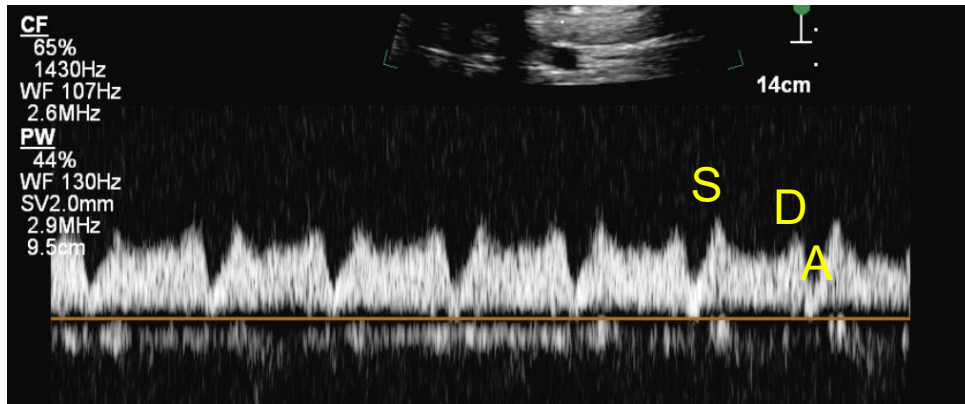
# Finding the veins



- Color from veins not “streaming” into left atrium
- Left vein color seems to “stick”



# Pulmonary vein doppler



In normal pulmonary vein doppler:

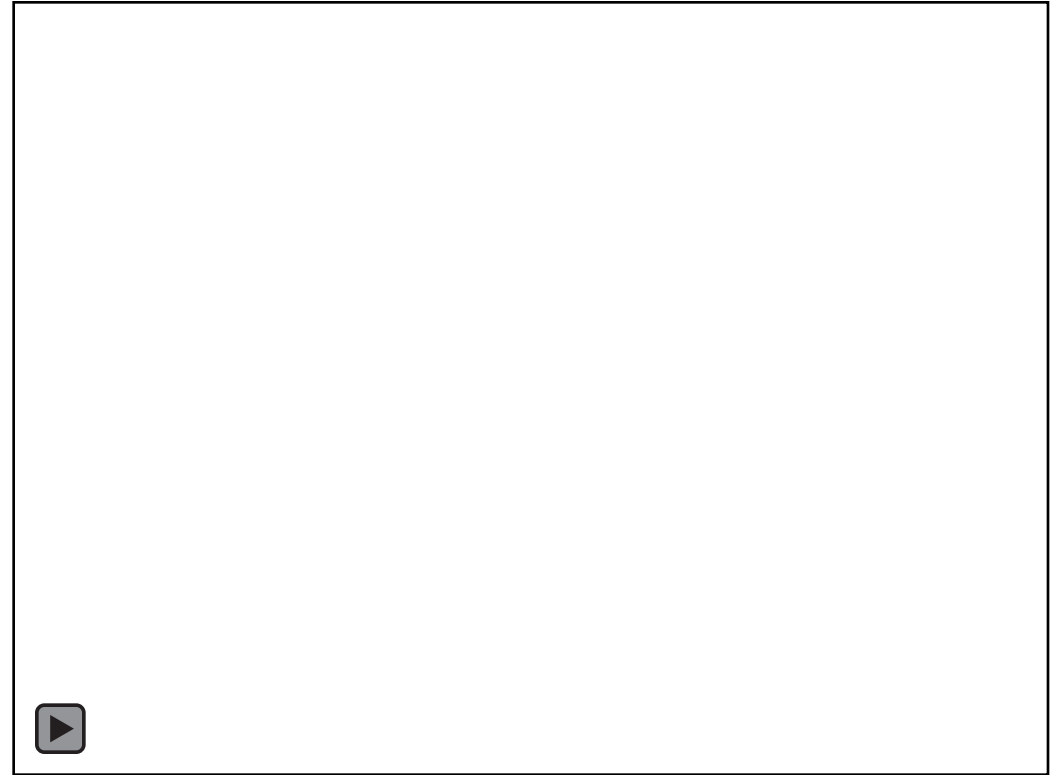
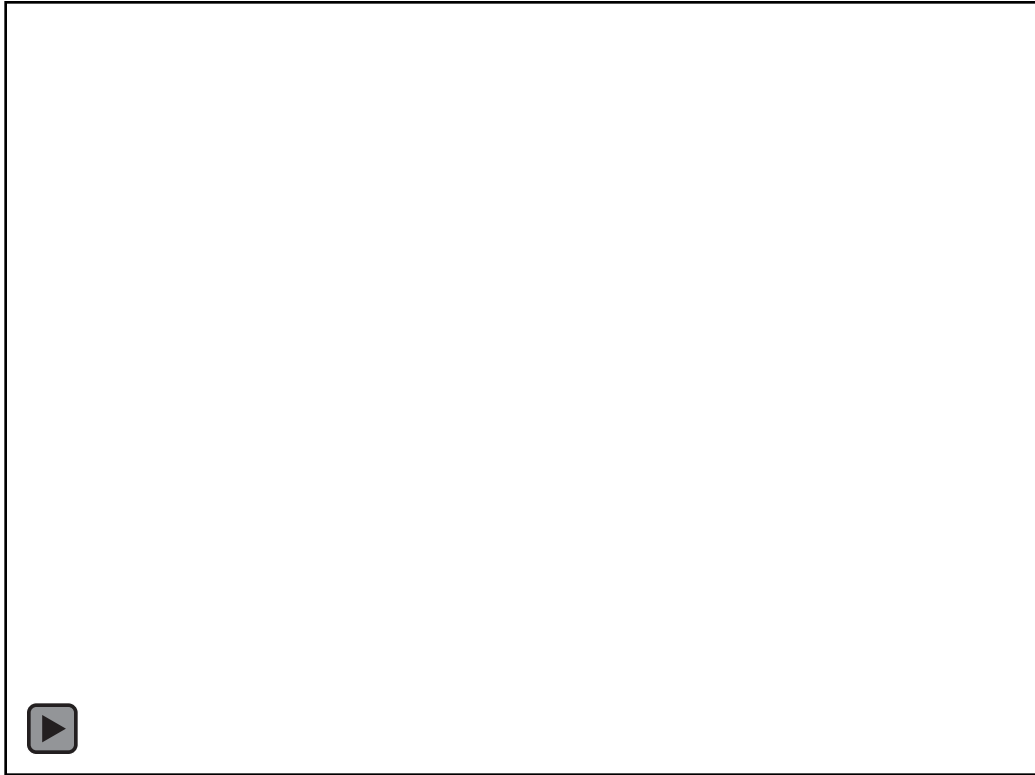
- Biphasic-can be triphasic

TAPVC it can be:

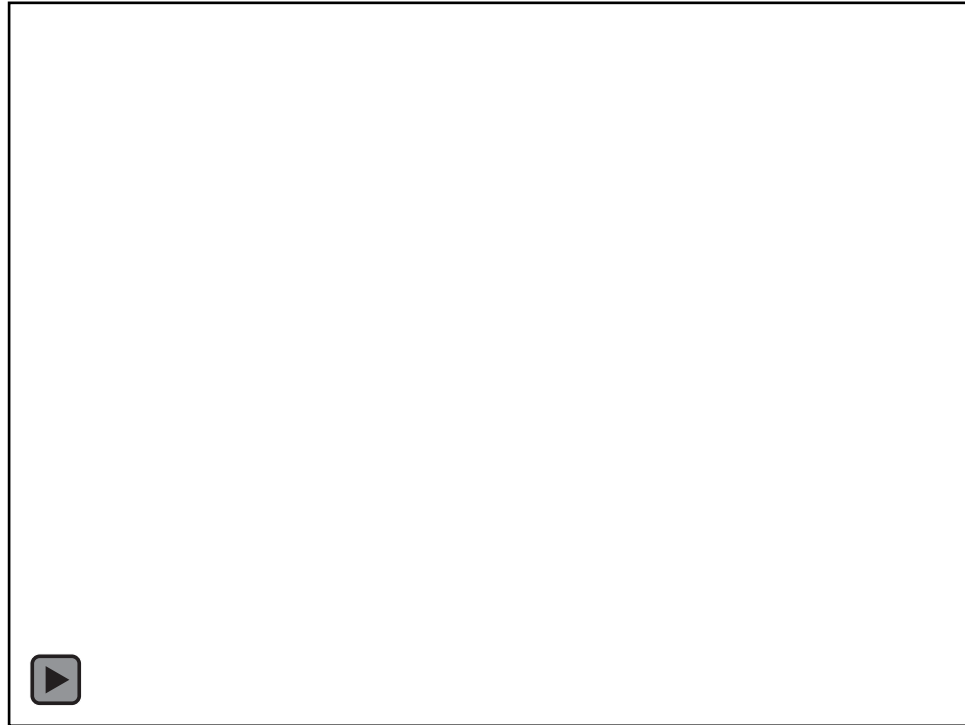
- Pseudonormal with slightly abnormal S, D
- monophasic
- low velocity monophasic/continuous



# Three vessel trachea view ...that's suspicious



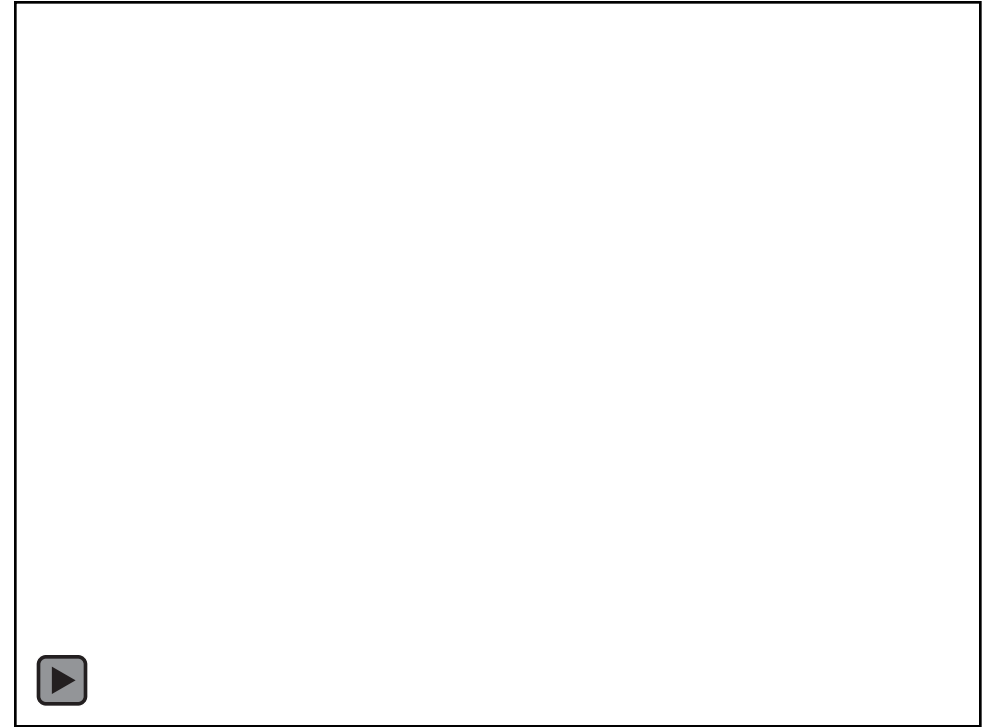
# Abdominal view



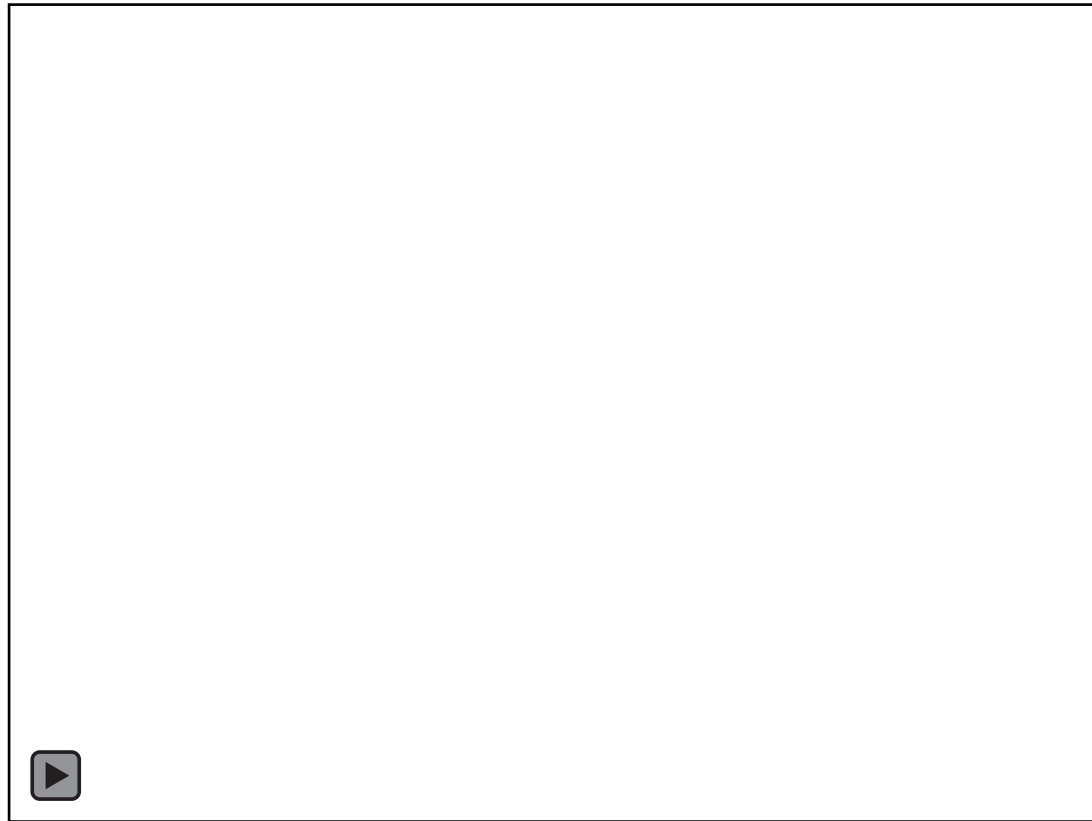
- Two separate turbulent flows
  - Ductus venosus
- What is the funny flow?

# The confluence

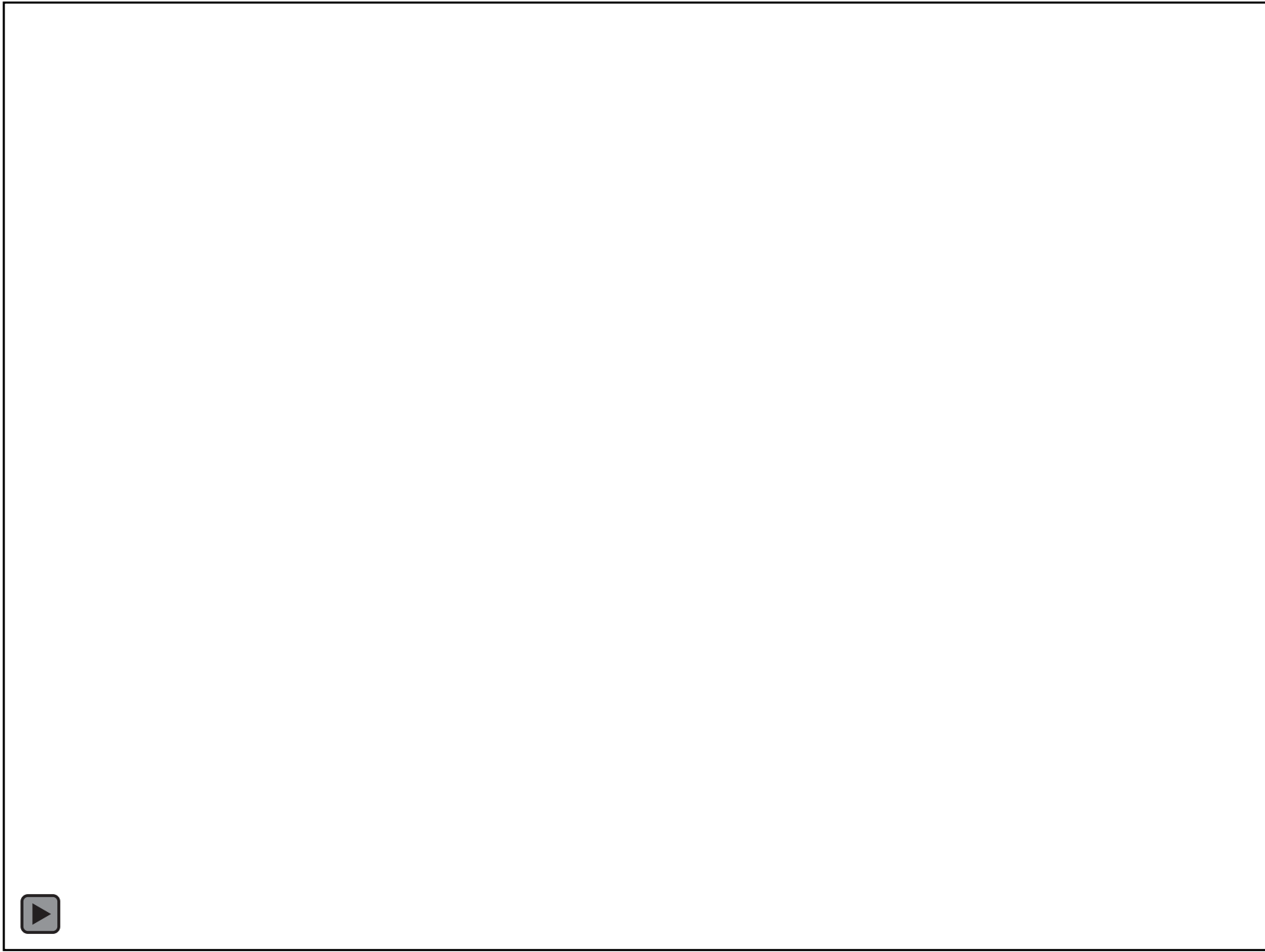
- NL is at 19
- Focus on the extra circle and in turn, on the pulmonary veins
- C9 probe being used..higher frequency can sometimes help define

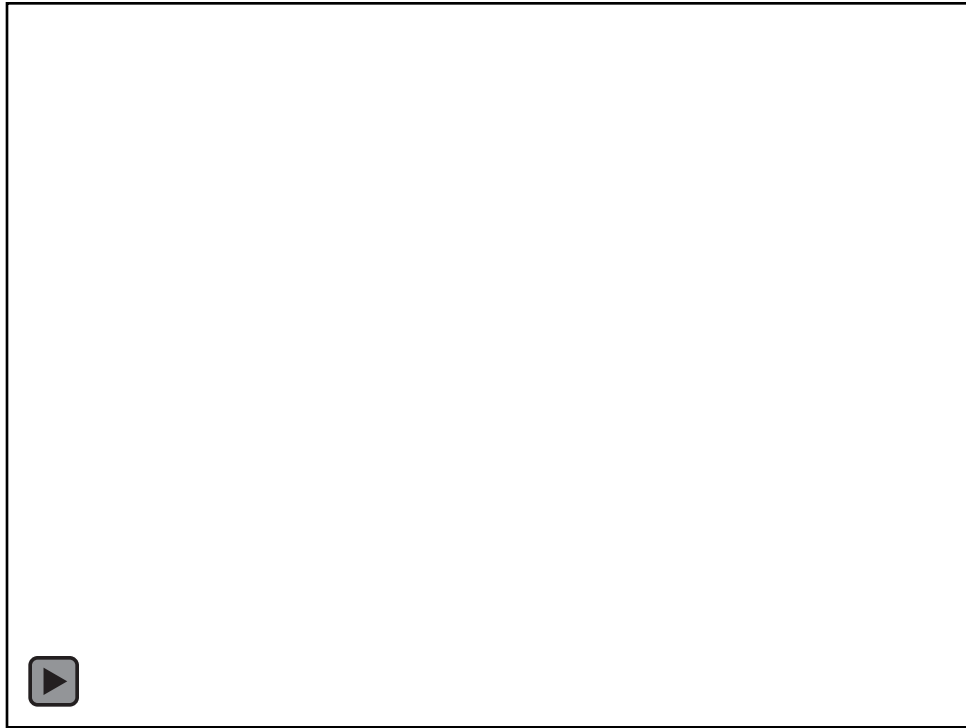


# The confluence (continued)



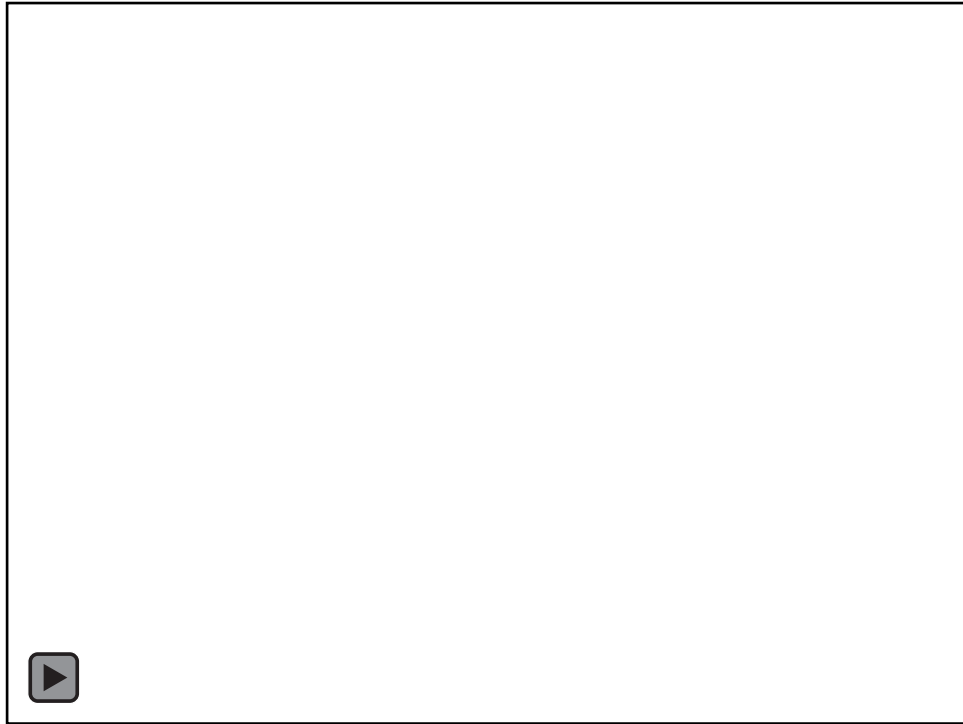
Color scale turned very low  
Follow extra circle  
Slow, careful sweeps  
Supracardiac TAPVC





- Small left atrium
- Extra circle
- No space between AO and LA?
- Right > left

# Importance of landmarks



- Small left atrium with septum shifted leftward
- Dilated coronary sinus
  - LSV or veins entering ?

# Cardiac TAPVC



- Move color box to show left veins\*
- This view combined with the 3vt view which showed an LSVC
- Explains the dilated coronary sinus and the dilated azygous next to aorta

# Case study

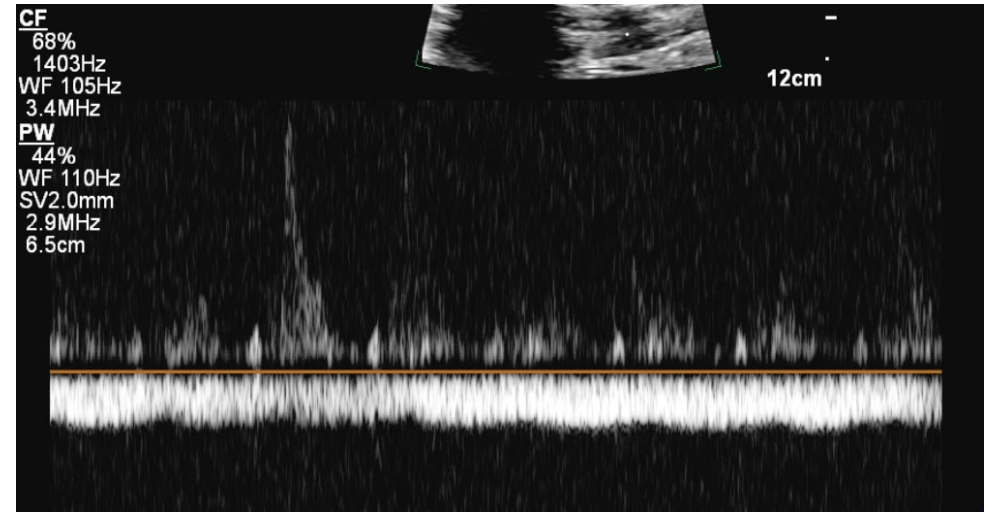
28 and 4/7 weeks

Referred for  
Tetralogy of  
Fallot

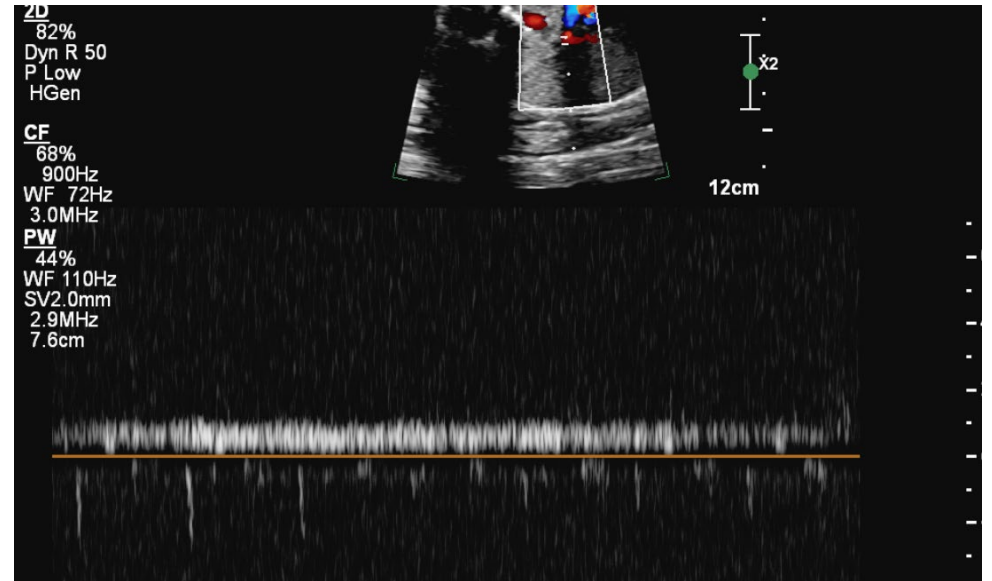
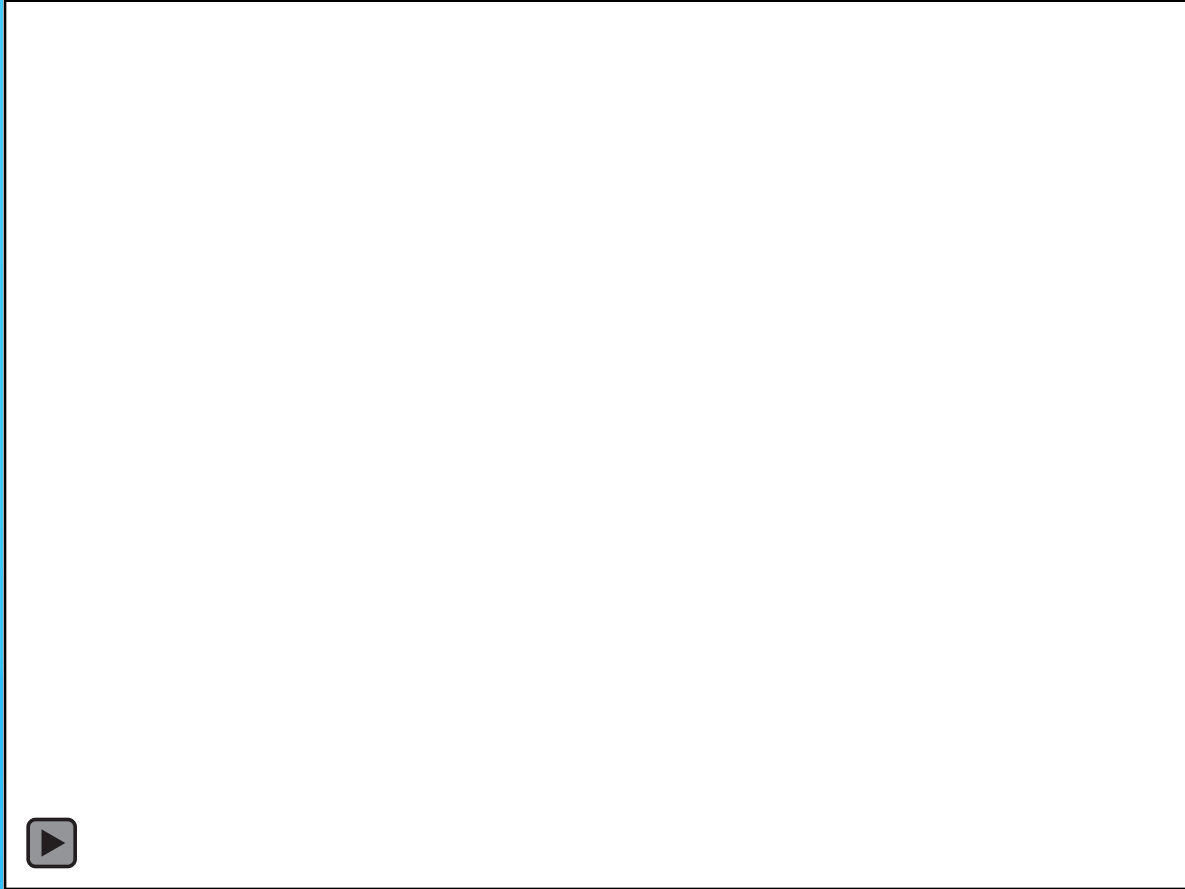
Low risk  
NIPT

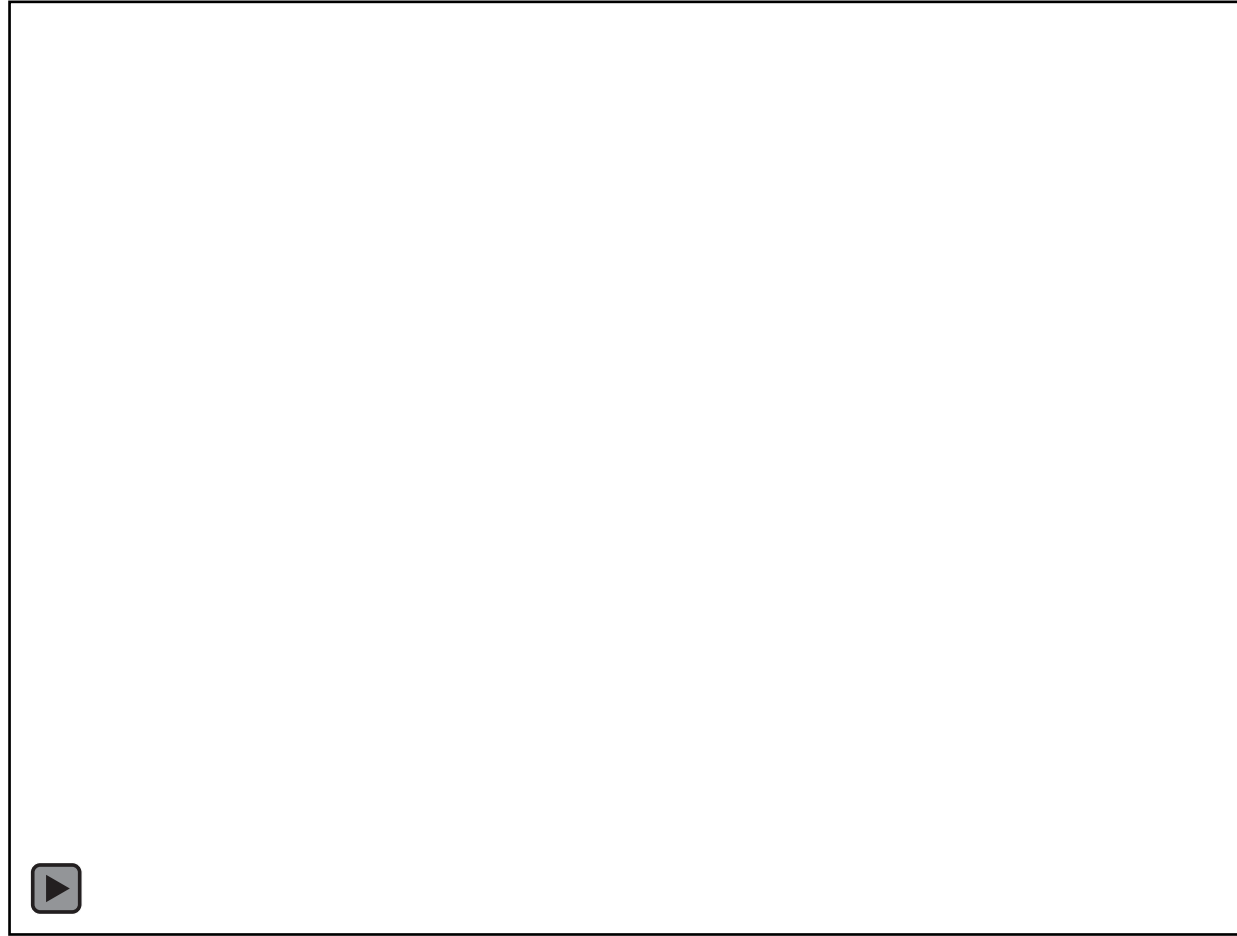


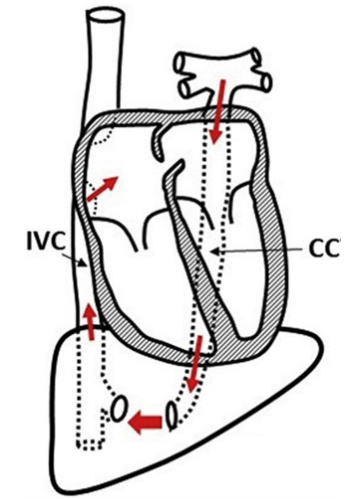
- Aorta distance from left atrium
- Extra circle
- Complete AV canal...difficult to determine left atrium size
  - Smooth atrial wall



Low-velocity monophasic and continuous flow







- Slow sweep following confluence or “extra circle”
- Color scale low
- Obstructed infradiaphragmatic TAPVC

# What's the plan?

Class 5 delivery

Planned C-section

In CVOR

Immediate intubation

Brief echo in OR

Delineate pulmonary venous drainage

If interventions aren't allowing time for echo-straight to OR with no additional imaging

After echo confirmation-TAPVC repair

## What is a class 5 delivery?

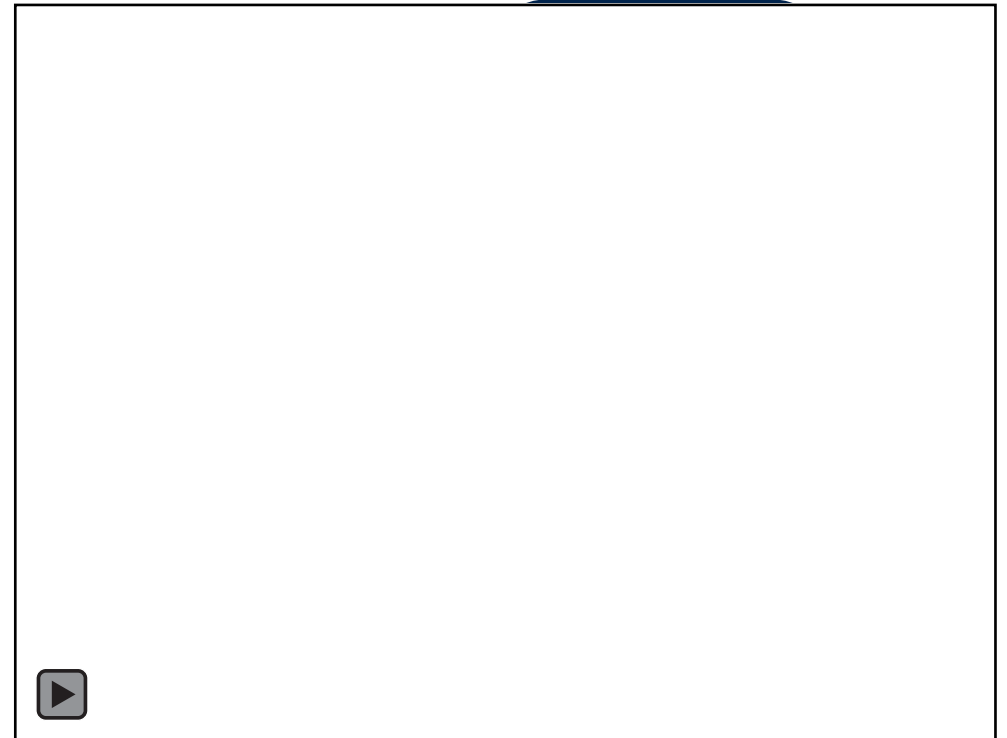
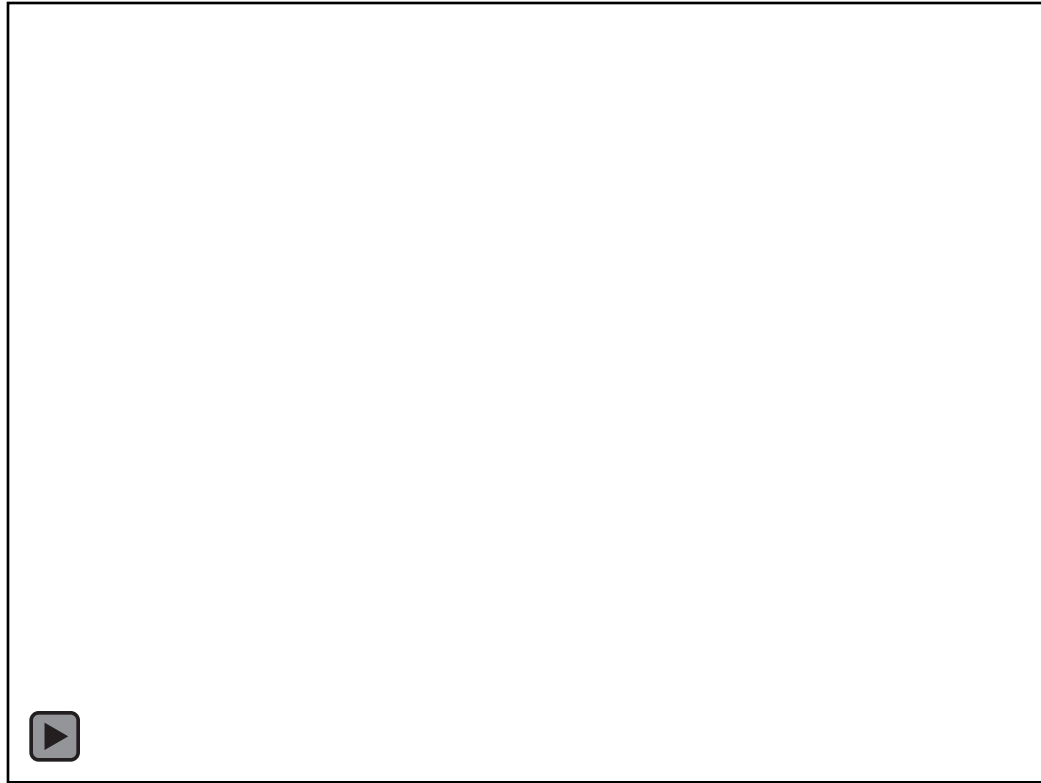
Neonatal instability is expected requiring immediate intervention. Delivery in the main OR with either CVOR or CVL on standby

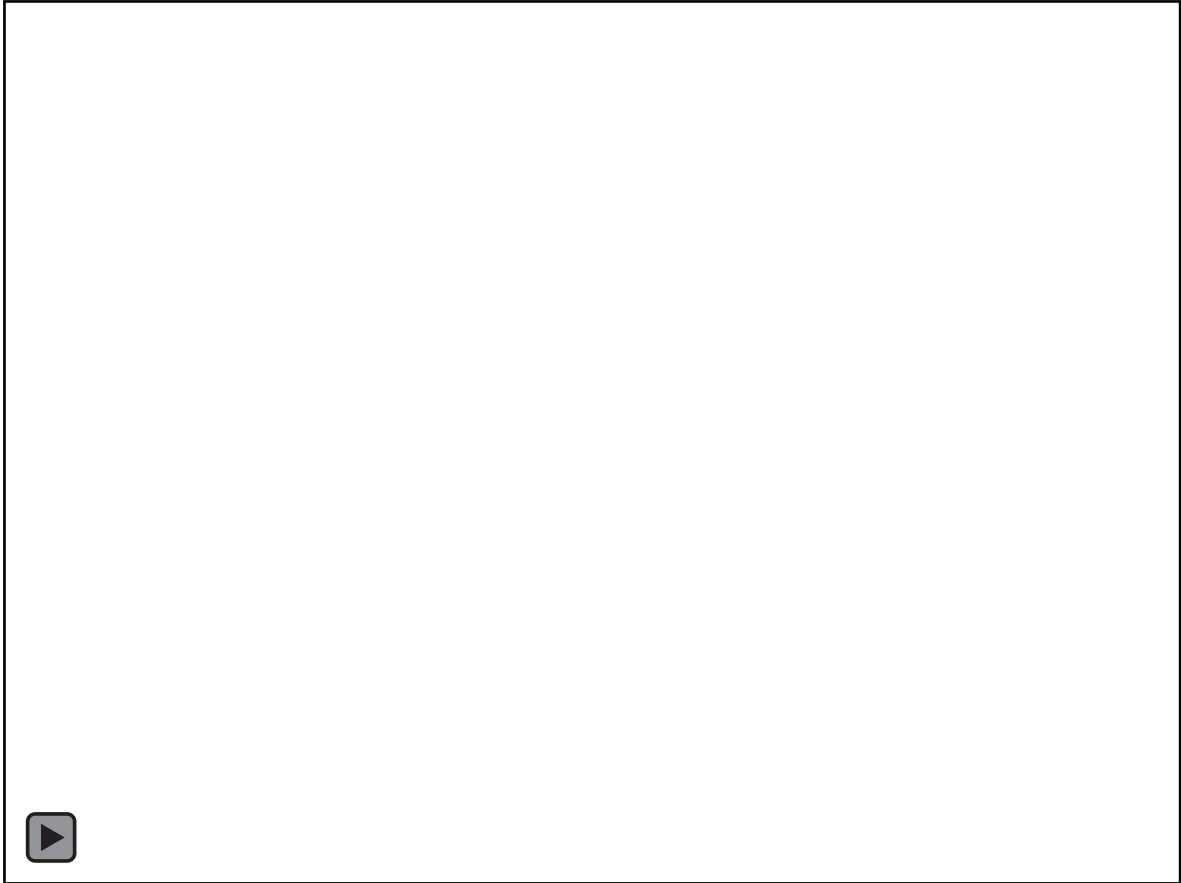
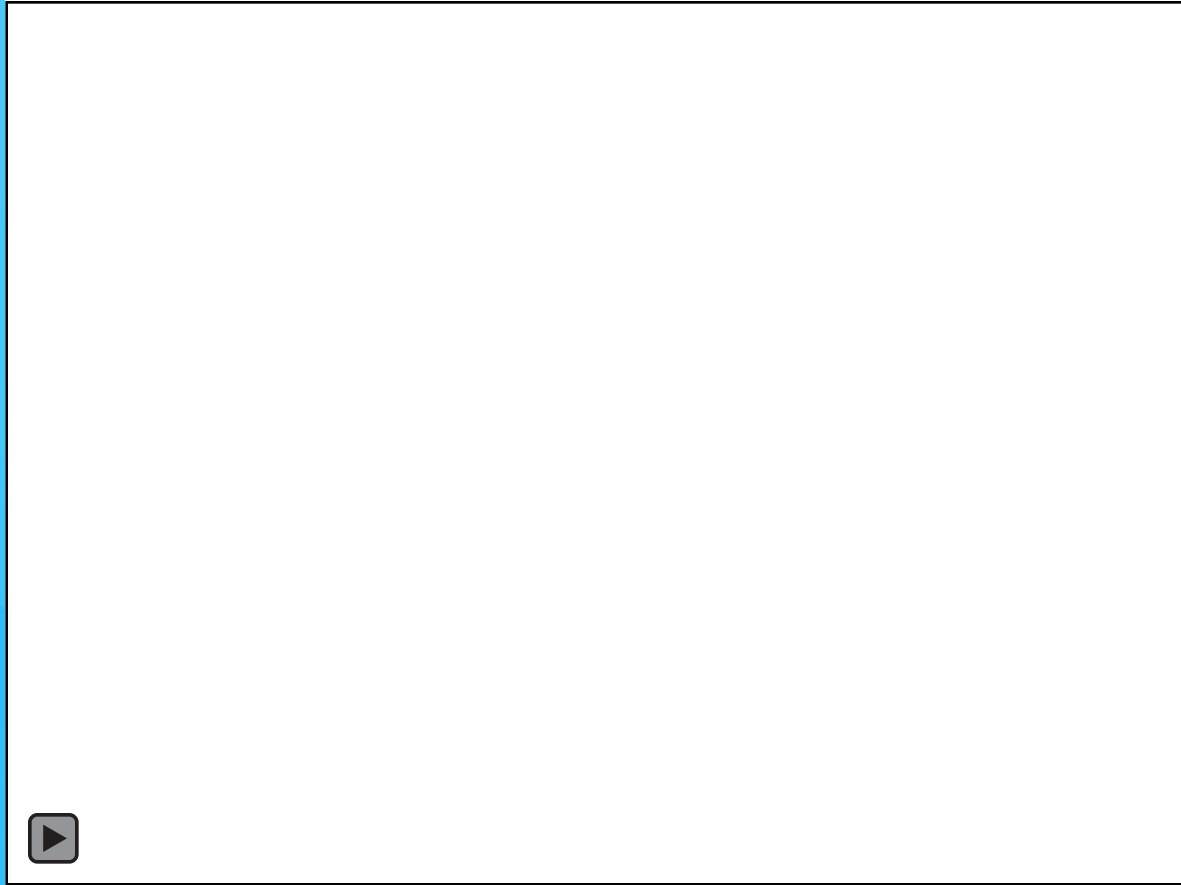
Involves multiple groups within the hospital working together

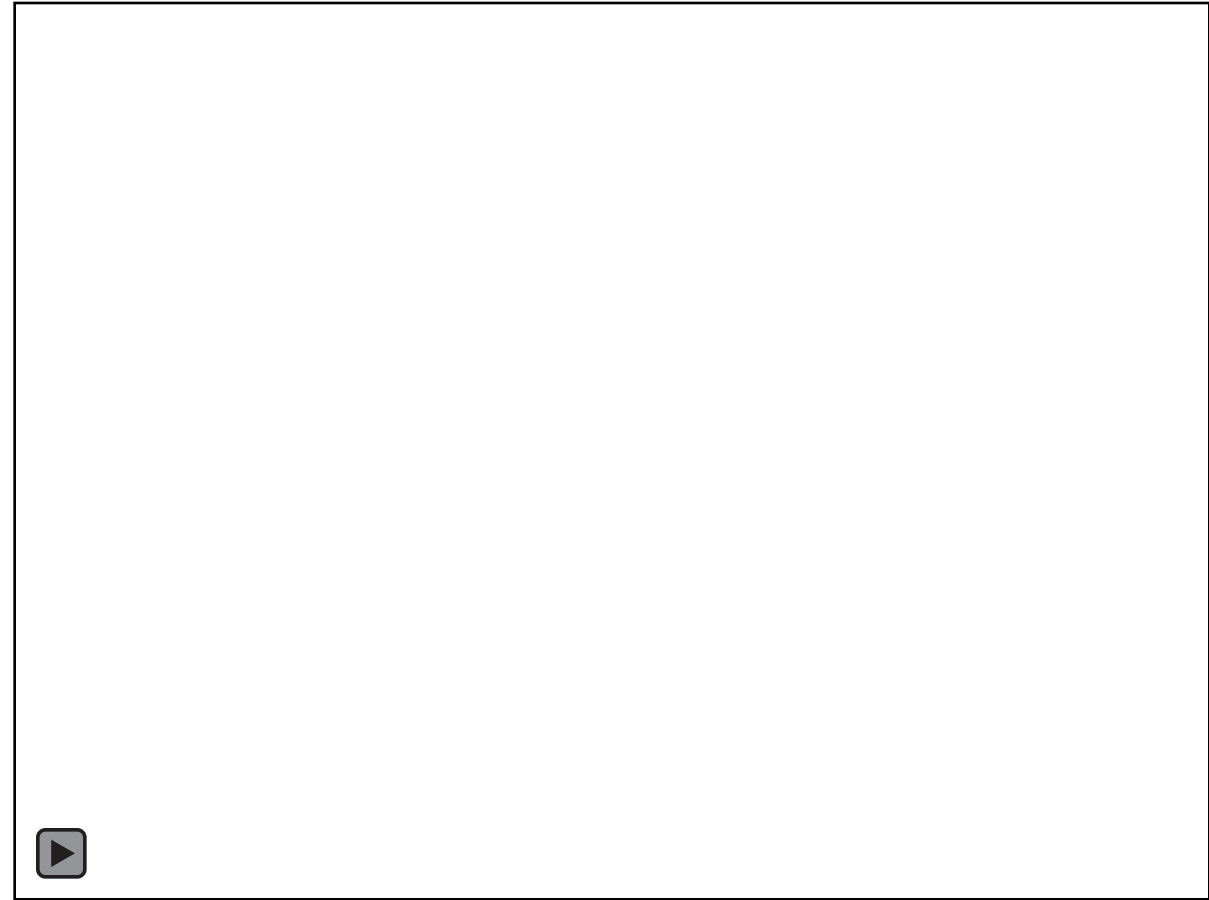
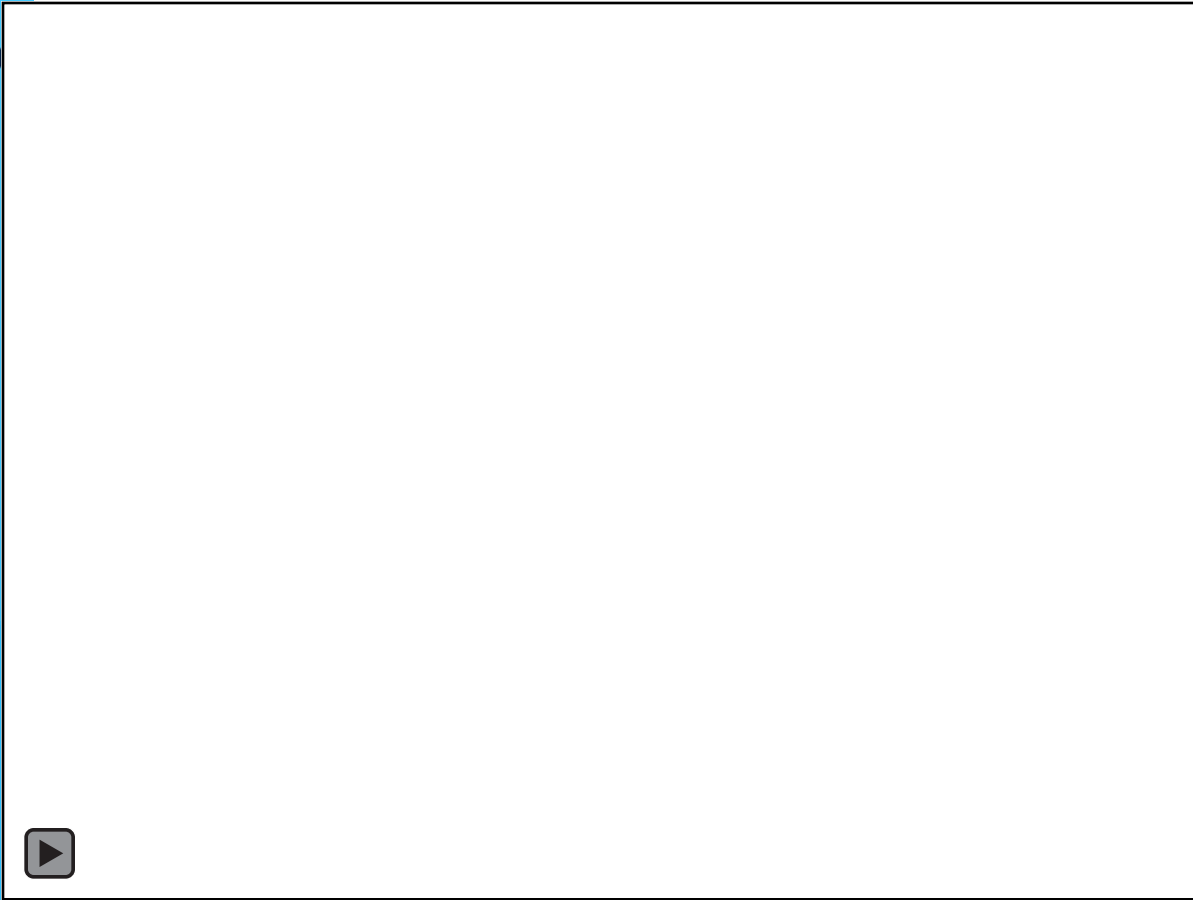
# What happened?

- Delivered via C-section
- Sats in 40s prior to intubation
- After intubation sats in 60s-70s
- Obtained limited echo images
- Sent to OR where underwent successful sutureless vein repair for obstructed TAPVC

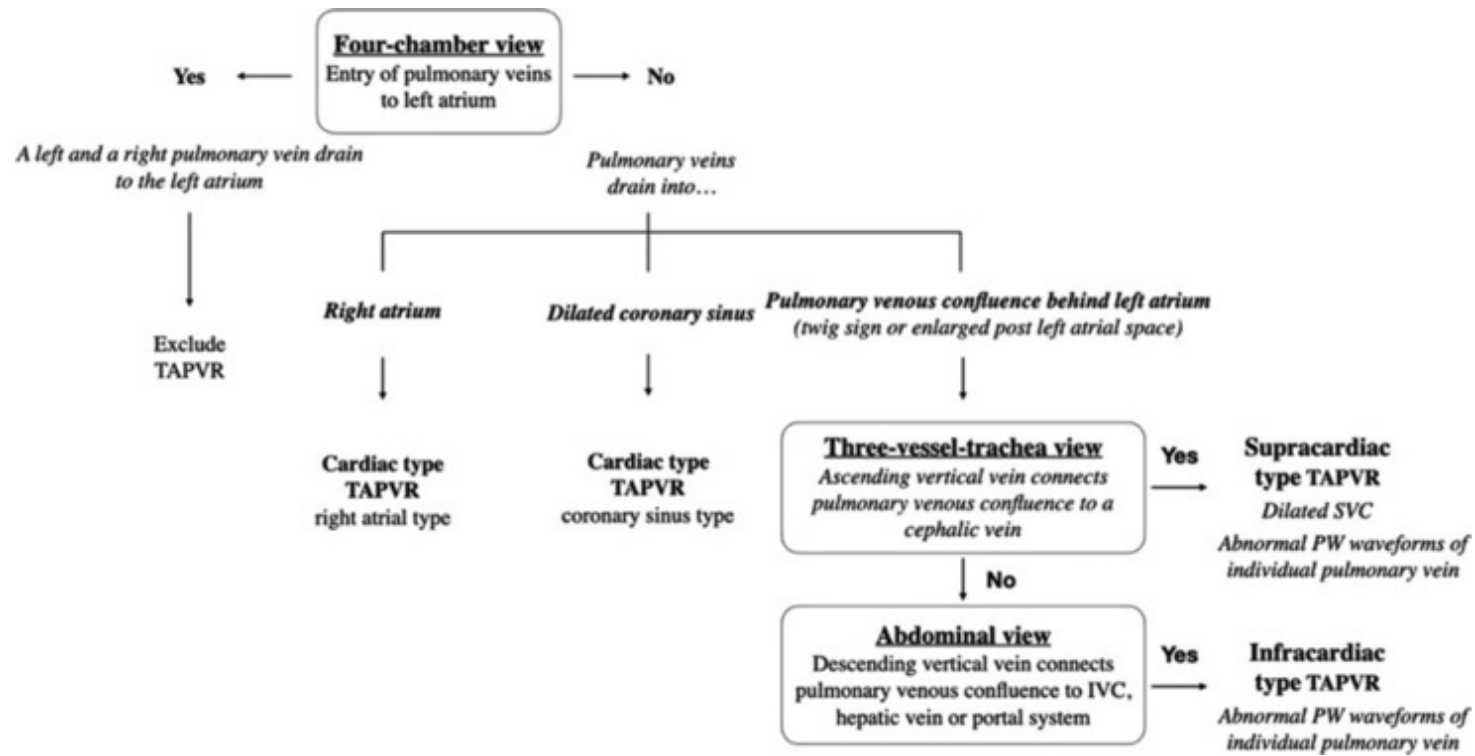
# Post natal images







# What could it be?



# \*Remember\*



- 12-20% of cardiac output is going to the lungs-low flow back to the heart
- Pulmonary veins are **SMALL**
- Prenatal detection is **HARD**
  - Indirect signs are your friend

# The views

Key takeaways

## Four chamber view

- Pulmonary vein flow is not entering left atrium
- Abnormal distance between left atrium and aorta
- “Twig sign”
- Small left atrium
- Dilated coronary sinus
- Size discrepancy R>L
- Extra circles
- No coumadin ridge

## 3 vessel view

- Extra circles
- Dilated SVC
- “Funny flow”

## Abdominal

- Presence of extra vessel
- Again with the “funny flow”

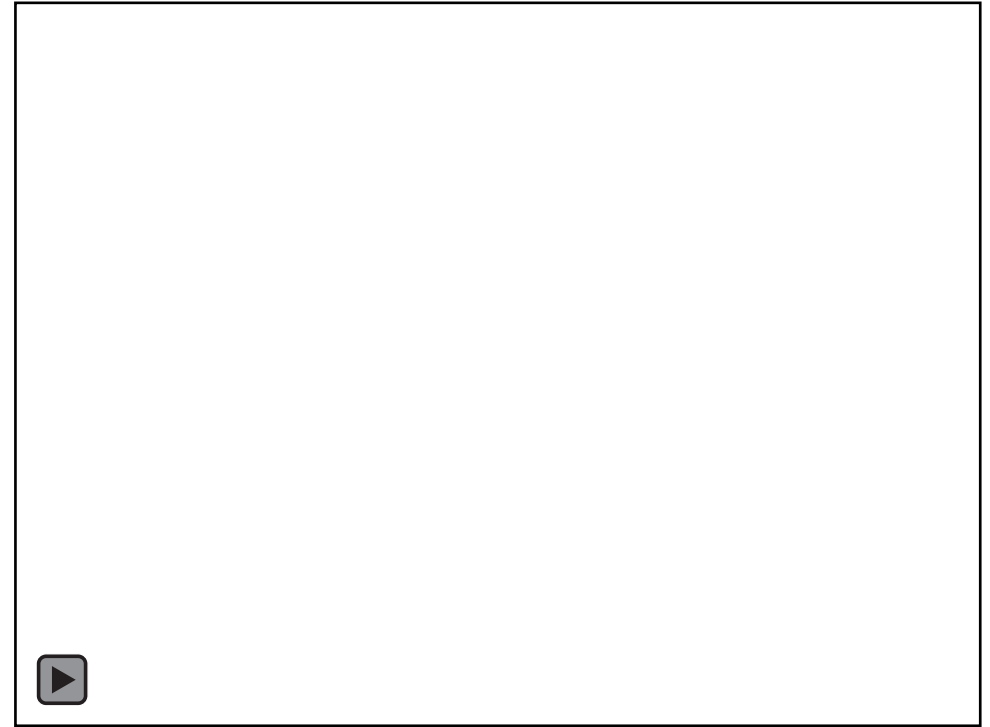
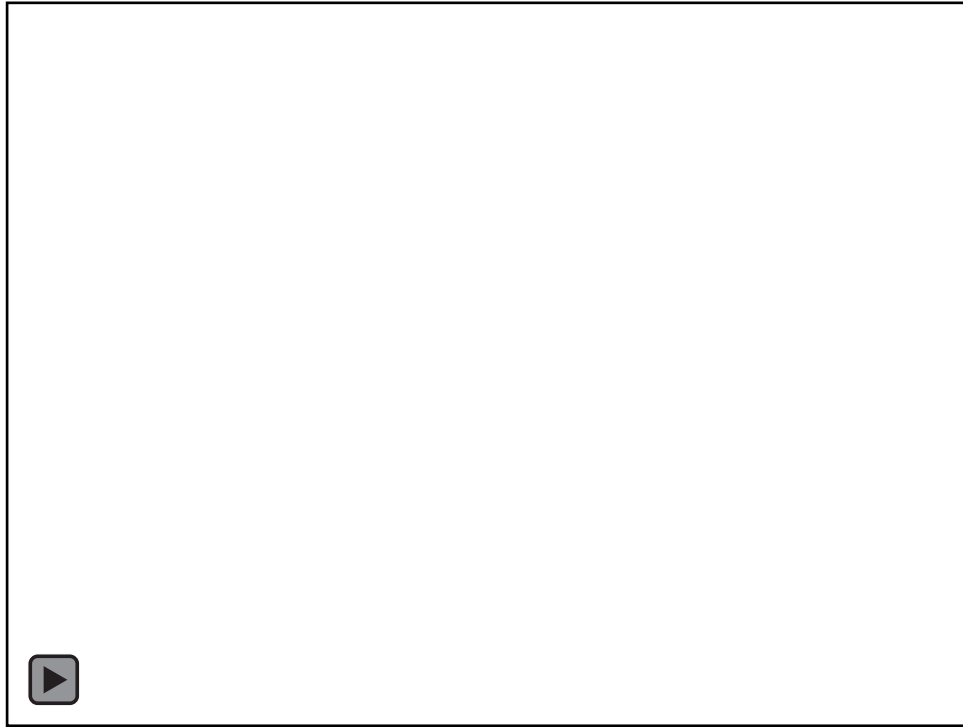


Just for fun... Name that vein in one sweep

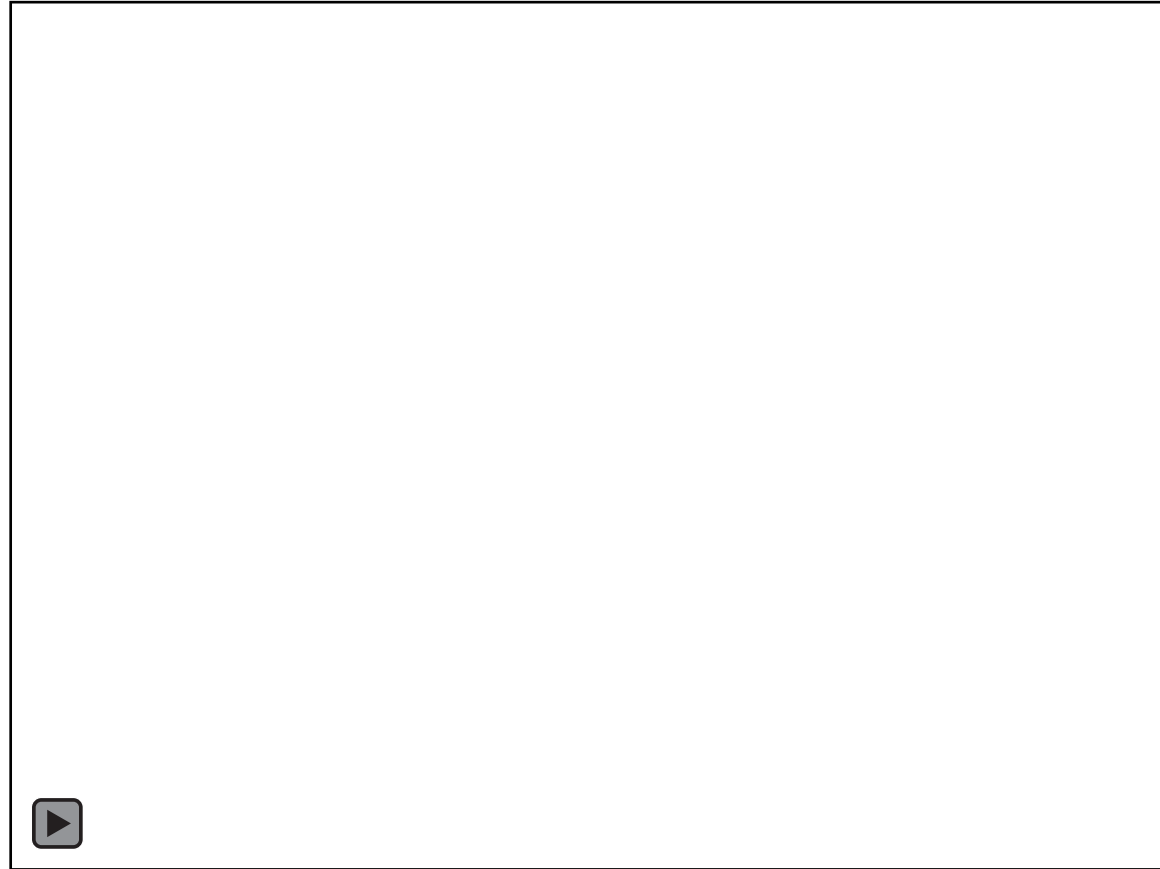


Supracardiac!

# Cardiac!



# Normal!!



# Thank you!!

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# References

Partial and Total Anomalous Pulmonary Venous Connection  
Anusha Konduri; Sanjeev Aggarwal.

Geva T, Van Praagh S. Anomalies of the pulmonary veins. In: Allen HD, Gutgesell HP, Clark EB, Driscoll DJ, eds. Moss and Adams' Heart Disease in Infants, Children and Adolescents. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2001:736–772. Originally adapted from work by R. C. Anderson

Chih WL, Ko H, Chang TY. Prenatal Ultrasound Markers of Isolated Total Anomalous Pulmonary Venous Return and a Sequential Approach to Reach Diagnosis. J Med Ultrasound. 2024 Apr 26;32(2):104-109. doi: 10.4103/jmu.jmu\_4\_24. PMID: 38882613; PMCID: PMC11175370.

Singh, U P1; Singh, Hargunbir2; Kaur, Ladbans3. A Journey from Adult to Fetal Echocardiography. Journal of The Indian Academy of Echocardiography & Cardiovascular Imaging 2(3):p 155-160, Sep–Dec 2018. | DOI: 10.4103/jiae.jiae\_26\_18

Prenatal Findings in Total Anomalous Pulmonary Venous Return

A Diagnostic Road Map Starts With Obstetric Screening Views

Suguna Ganesan, MD, Michael M. Brook, MD, Norman H. Silverman, MD, Anita J. Moon-Grady, MD