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Pulmonary veins: anatomy & abnormalities

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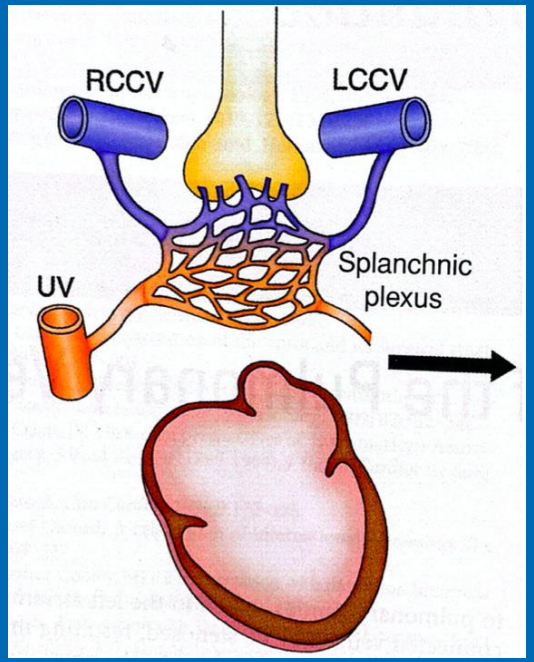
Here, it's different.™

No disclosures

Outline

- Embryology – yes, bear with me here!
- Normal and normal variants
- A framework for thinking about pulmonary venous abnormalities
- Can't-miss: TAPVR (may cause hemodynamic collapse), cor triatriatum
- Nice-to-know (causing right heart dilation, possible PH if untreated long-term): PAPVR, sinus venosus defect

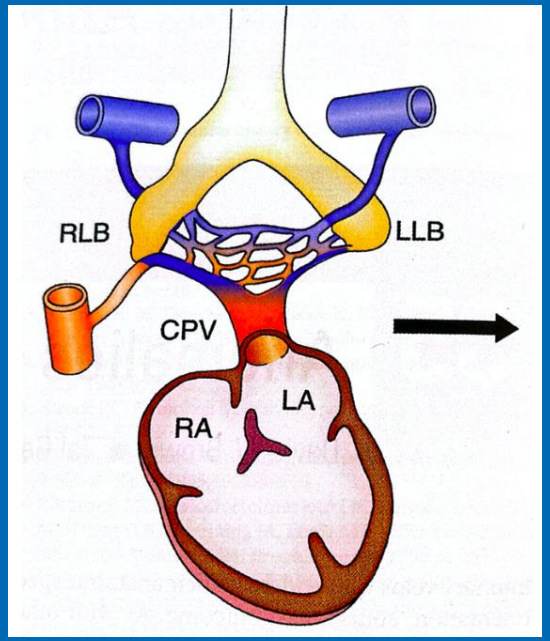
Embryology: ~4 weeks gestation



- Pulmonary blood supply grows from lungs, initially not connected to the heart (!)
- Connected to the systemic vein predecessors: cardinal veins = SVCs, umbilicovitelline system = IVC, portal veins

RCCV: right cardinal vein (becomes R-SVC); LCCV: left cardinal vein (becomes innominate vein +/- L-SVC); UV: umbilical vein

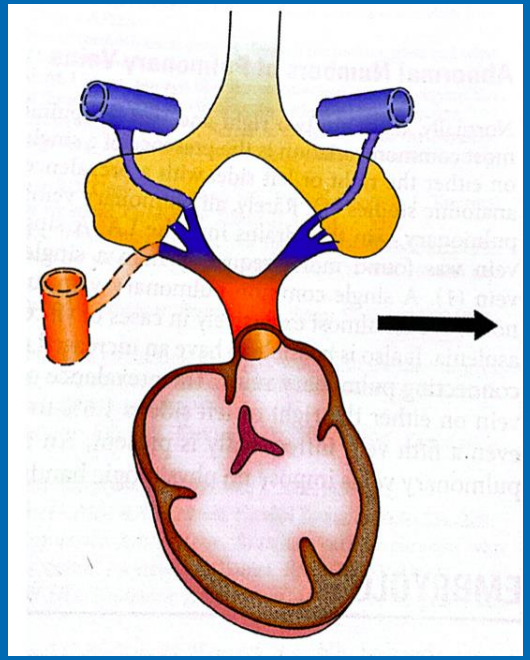
Embryology: 4-5 weeks



- Common pulmonary vein connects the pulmonary vascular bed to back of LA

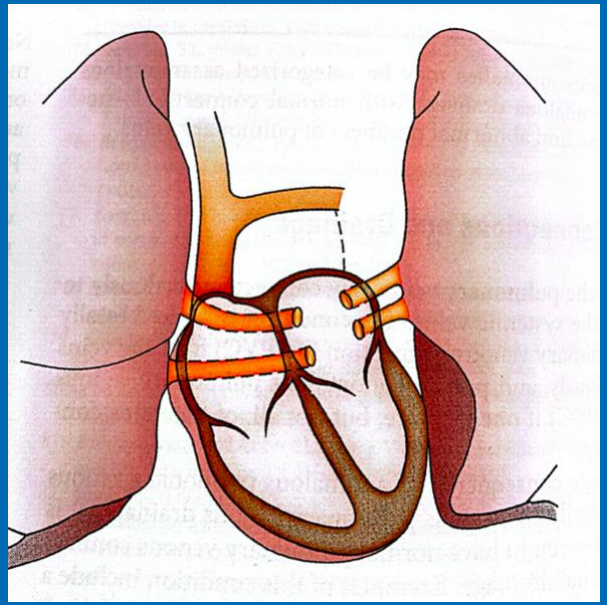
RLB: right lung bud; LLB: left lung bud; CPV: common pulmonary vein

Embryology: 5-6 weeks



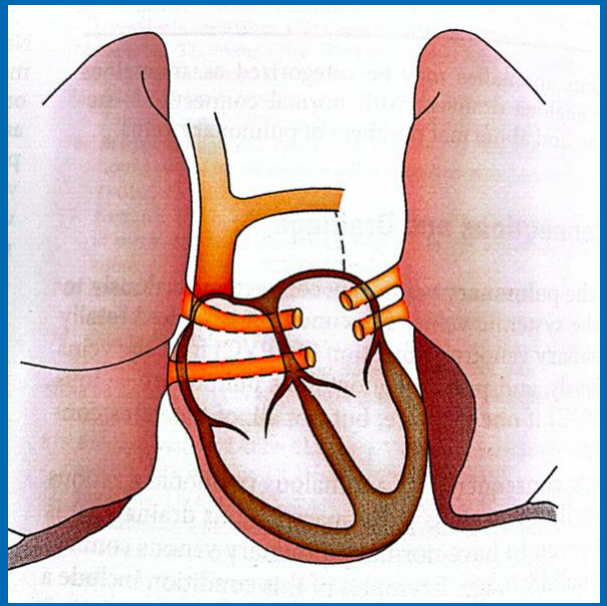
- Connections between pulmonary vascular bed and systemic veins involute

Embryology: term



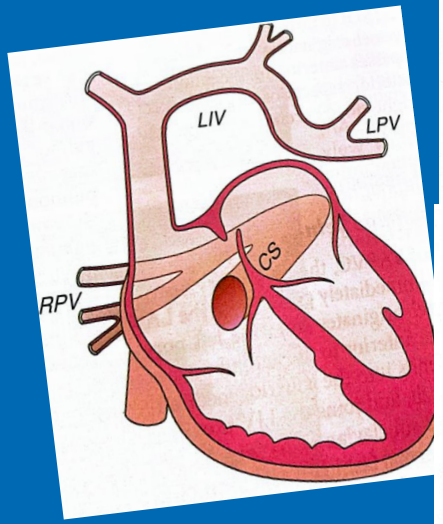
- Common pulmonary vein incorporates into back of LA
- Pulmonary veins drain individually into LA

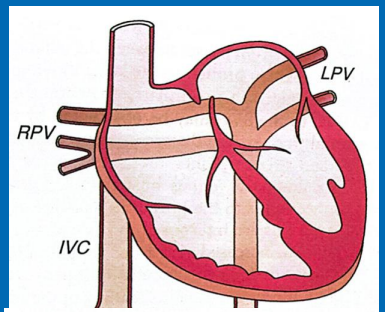
Normal and normal variants

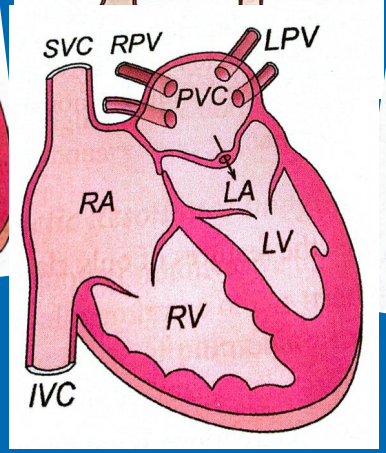


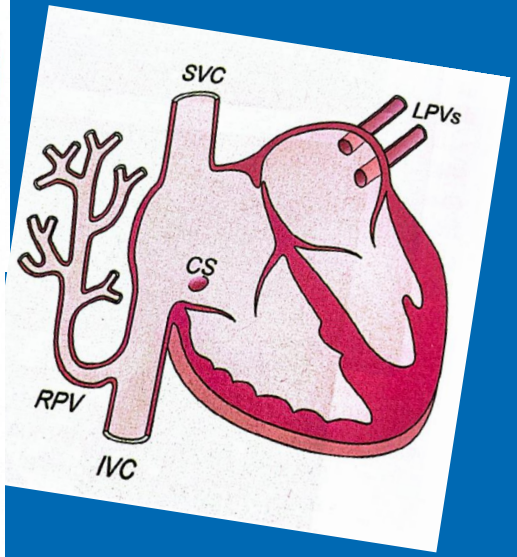
- Normal: two right and two left pulmonary veins
- Single pulmonary vein, left > right (~25%)
- An extra normally connecting pulmonary vein, left > right (1.5-2%)

What could possibly go wrong?

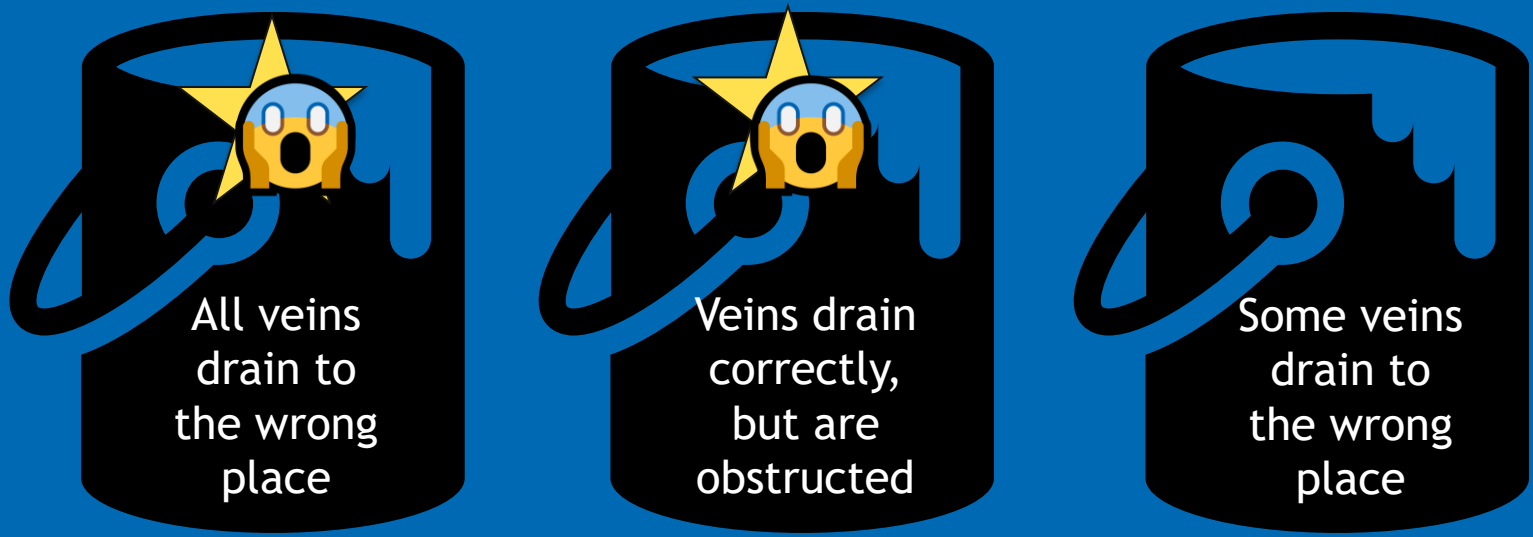








Three buckets of pulmonary vein problems



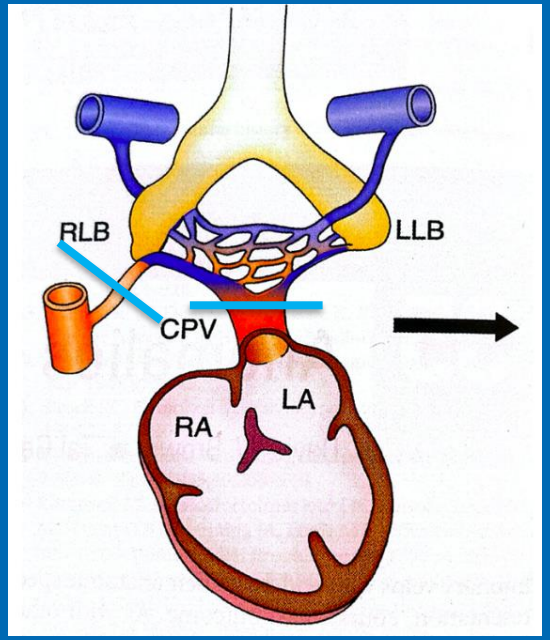
Totally anomalous pulmonary venous return

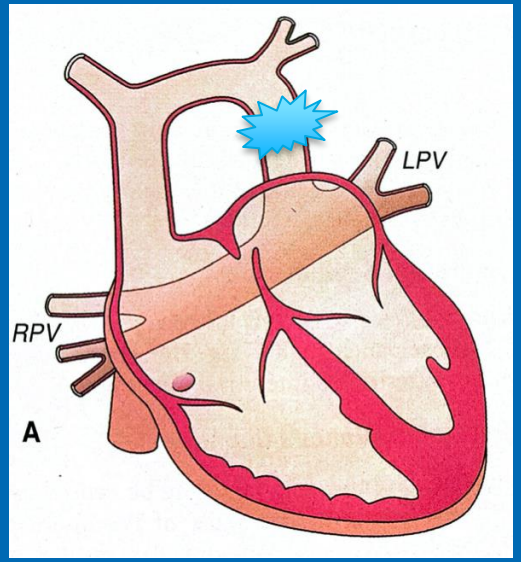
- Pulmonary veins drain to systemic venous return / right heart
- Supracardiac
- Cardiac
- Infracardiac
- Mixed

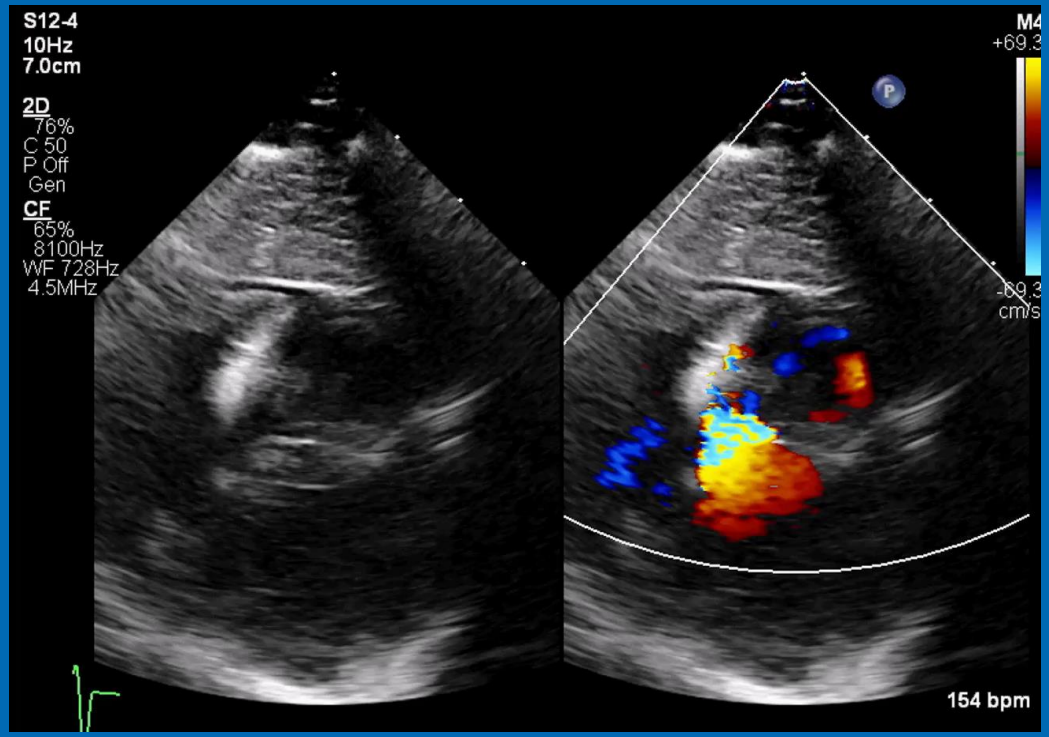
Totally anomalous pulmonary venous return

- Can't-miss: pulmonary venous drainage is often obstructed, especially in infra- and supracardiac variants
- Life-threatening if not surgically repaired emergently…not even PGE can save you! ☹

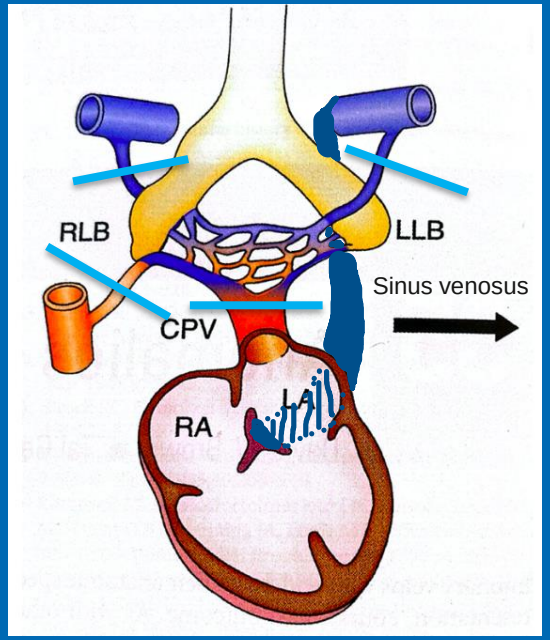
Supracardiac TAPVR

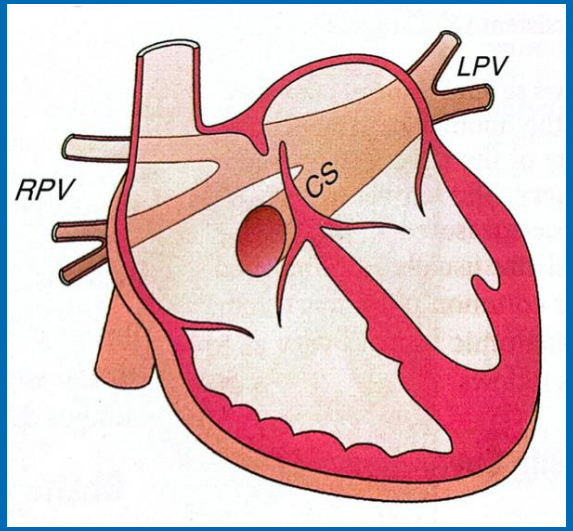


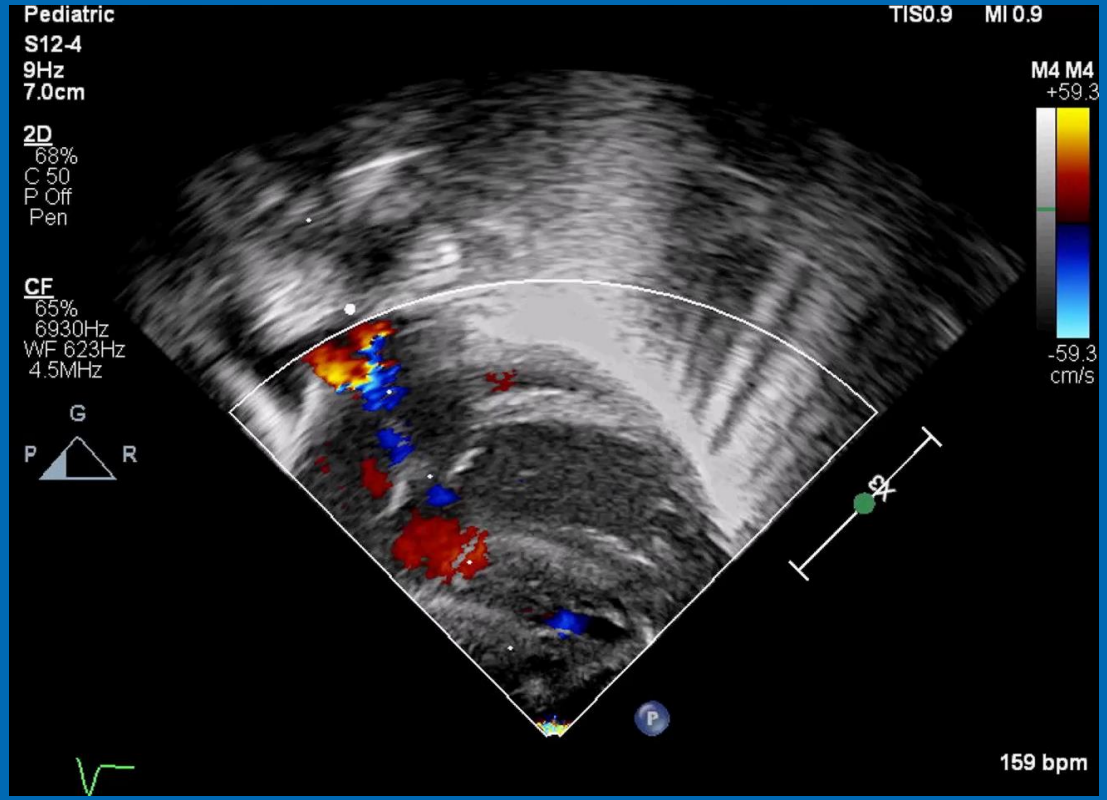


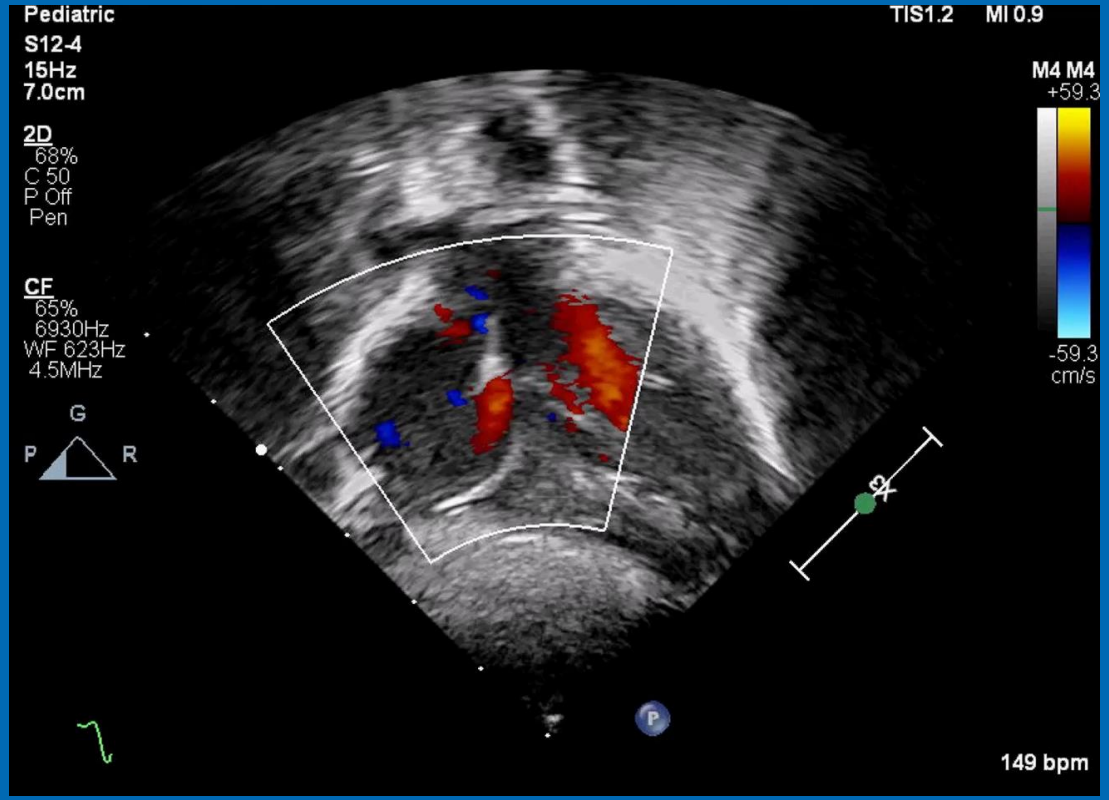

S12-4
10Hz
7.0cm
2D
76%
C 50
P Off
Gen
CF
65%
8100Hz
WF 728Hz
4.5MHz
M4
+69.3
-69.3
cm/s
154 bpm

Cardiac TAPVR (to CS)

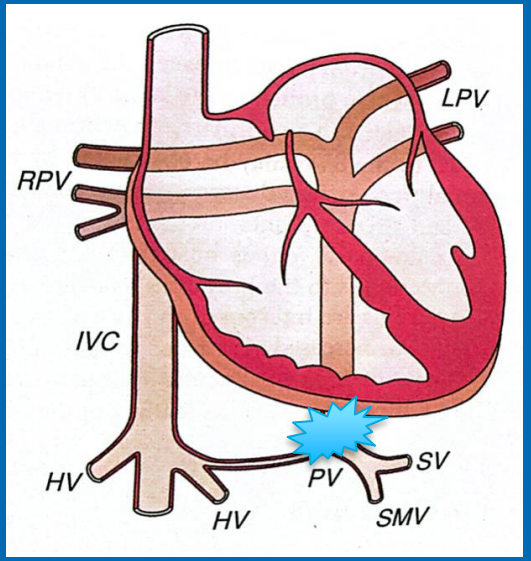


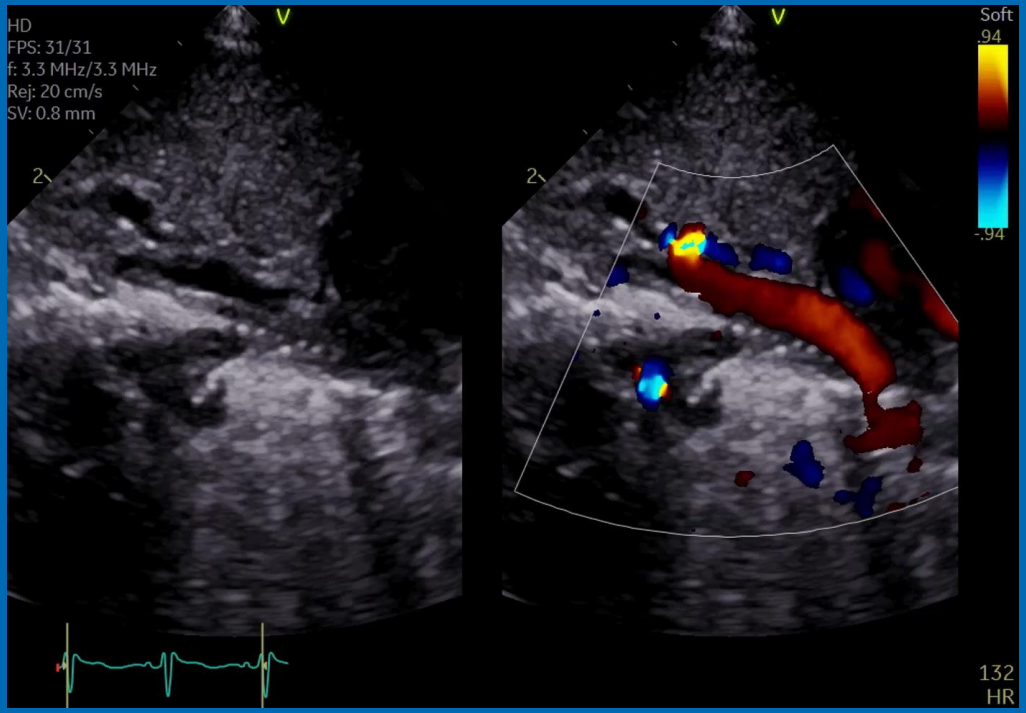


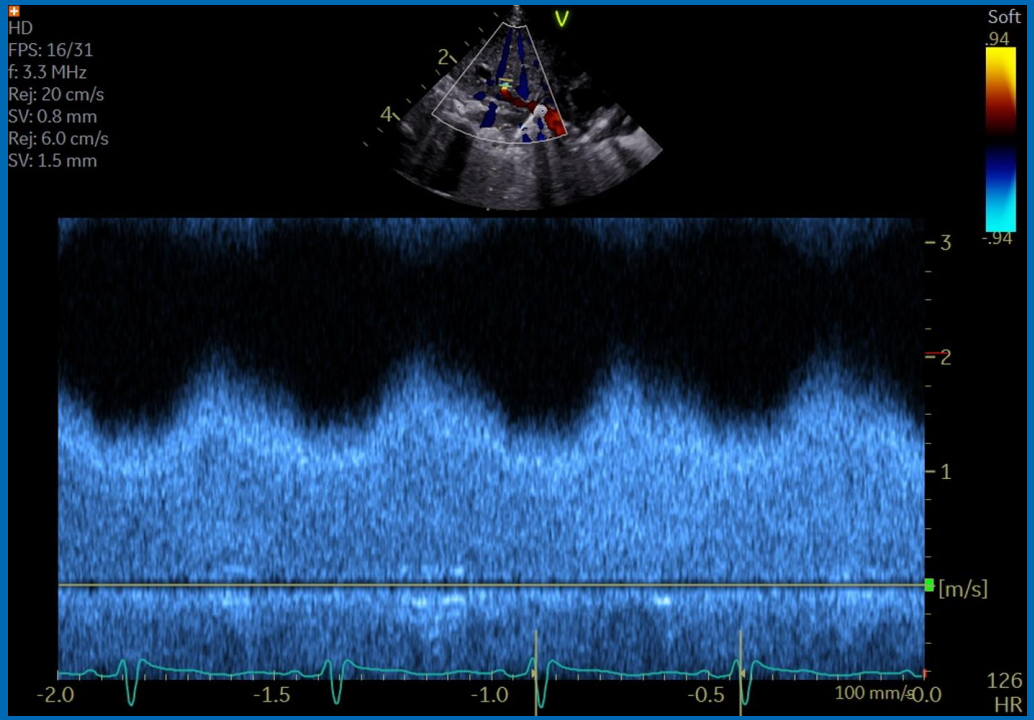

Pediatric
S12-4
9Hz
7.0cm
2D
68%
C 50
P Off
Pen
CF
65%
6930Hz
WF 623Hz
4.5MHz
G
P
R
TIS0.9
MI 0.9
M4 M4
+59.3
-59.3
cm/s
159 bpm


Pediatric
S12-4
15Hz
7.0cm
2D
68%
C 50
P Off
Pen
CF
65%
6930Hz
WF 623Hz
4.5MHz
G
P
R
TIS1.2
MI 0.9
M4 M4
+59.3
-59.3
cm/s
149 bpm

Infracardiac TAPVR



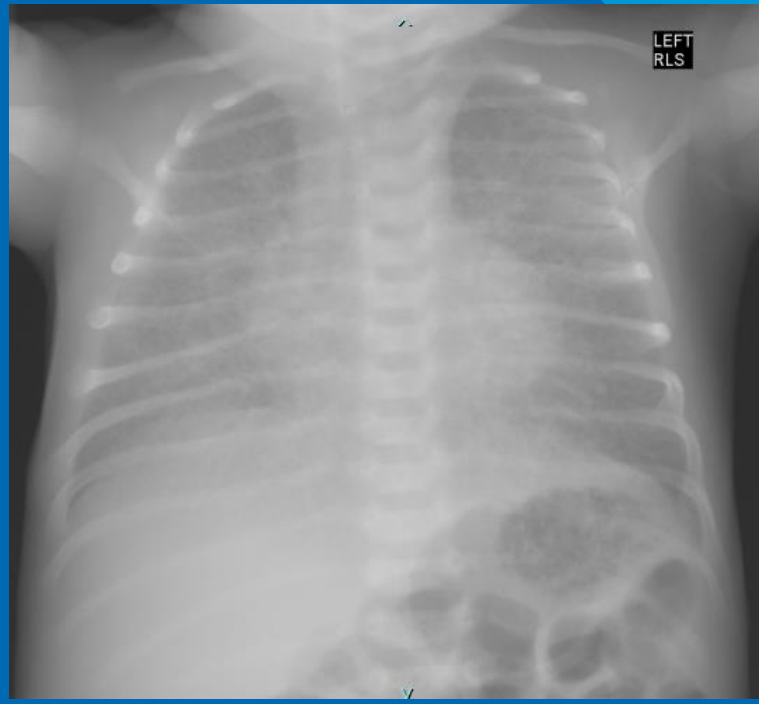

HD
FPS: 31/31
f: 3.3 MHz/3.3 MHz
Rej: 20 cm/s
SV: 0.8 mm
Soft
.94
-.94
132
HR



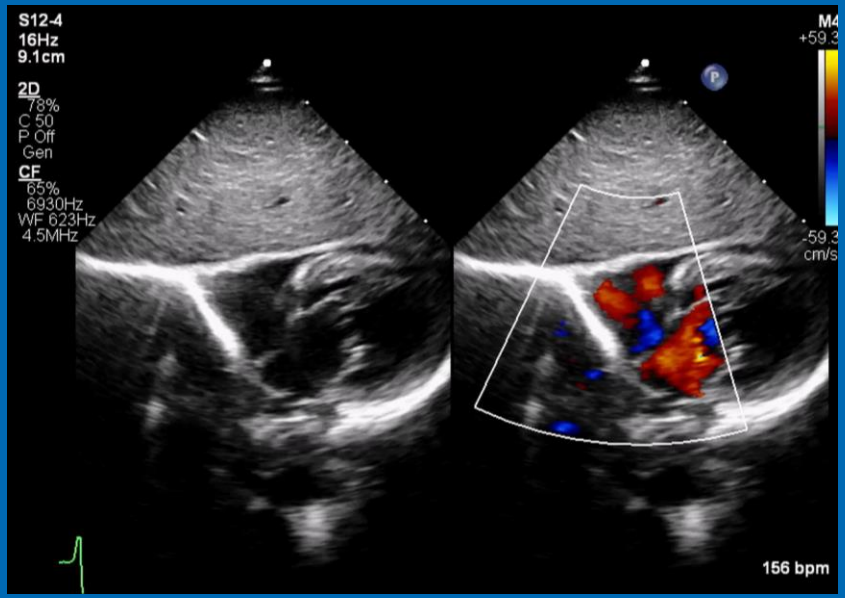
When in doubt, Doppler it out!

How to suspect TAPVR clinically?

- If unobstructed, often have mild hypoxemia / O2 requirement, can present like mild PPHN
- If obstructed, cardiorespiratory collapse: grey and blue baby
    - Not enough LV filling for adequate systemic perfusion
    - Lungs edematous, so SpO2 low / may not be responsive to FiO2
    - White-out lungs on CXR, apex elevated due to RV dilation
    - Intubated with bloody secretions from pulmonary hemorhage



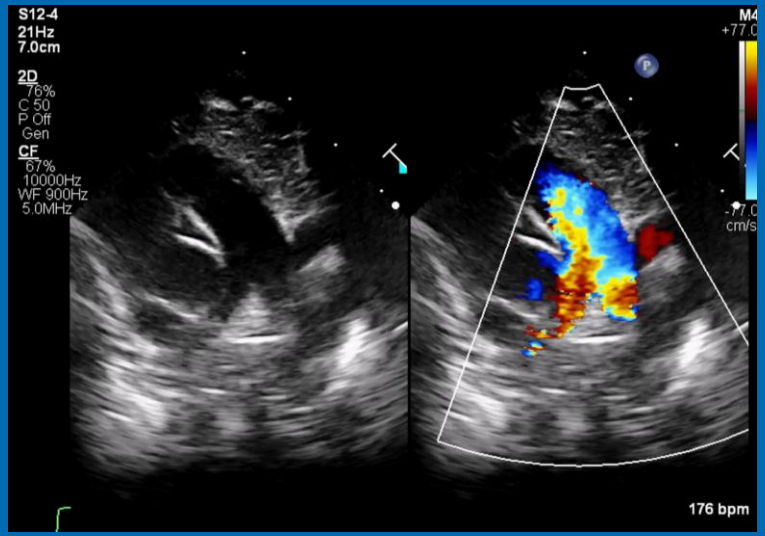
How to suspect TAPVR by echo?



Pure right-to-left shunt at atrial level

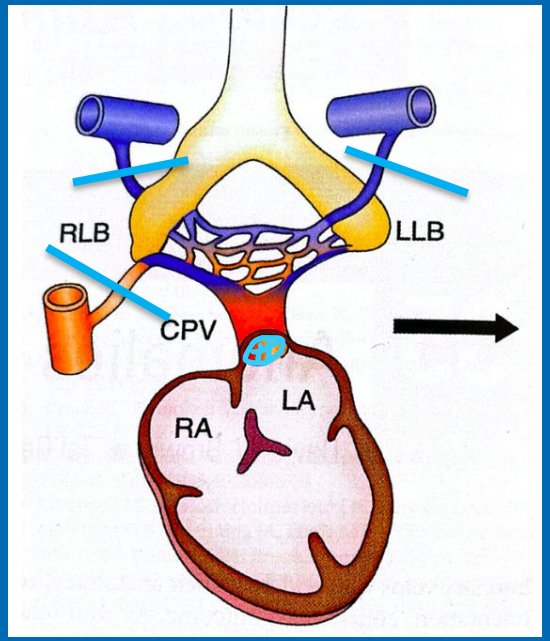
How to suspect TAPVR by echo?

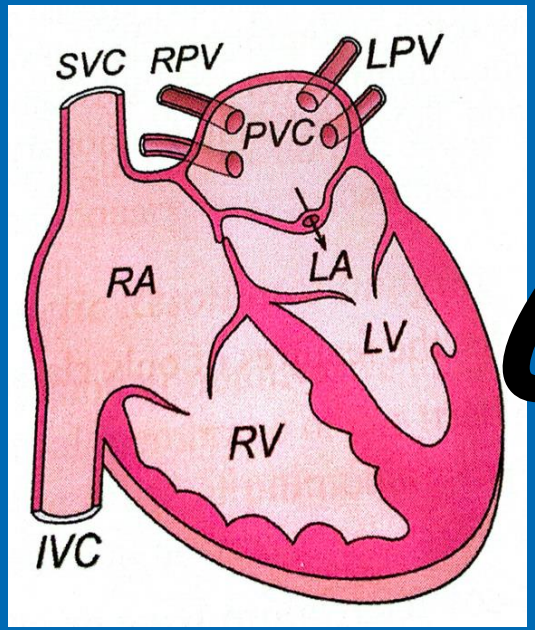
Weird venous flows in the parasternal/SSN



Weird venous flows in the liver

Cor triatriatum (“tri-atrial heart”)

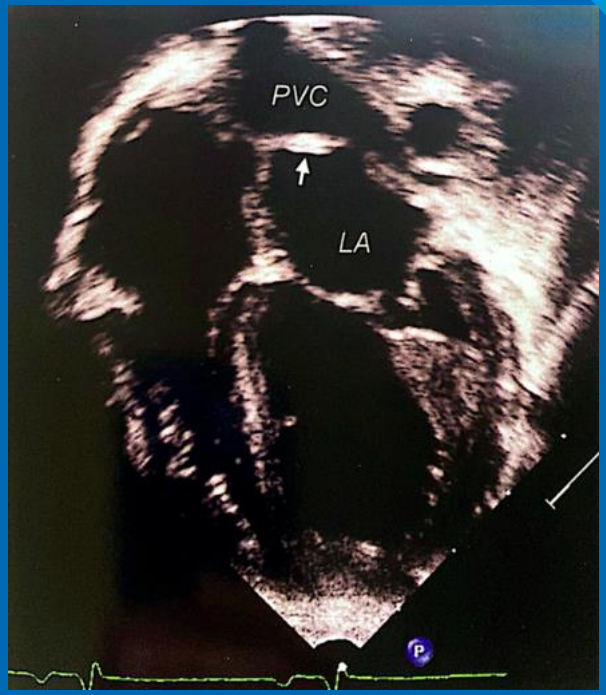


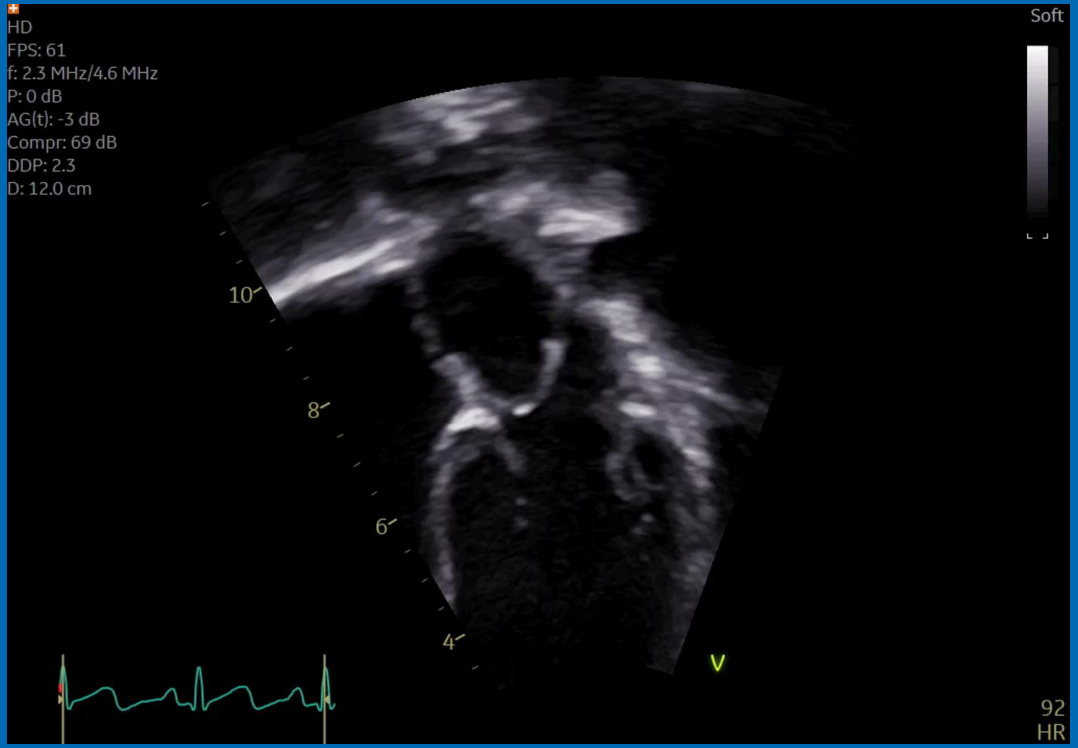


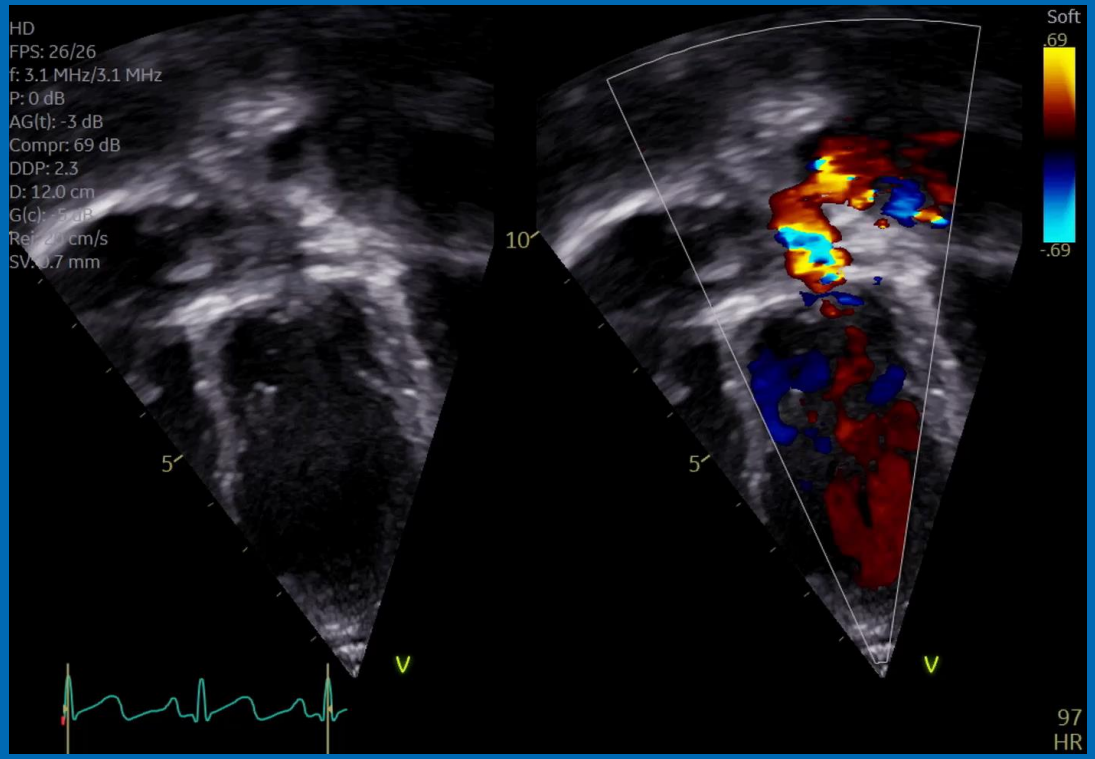
Veins drain correctly, but are obstructed

How to suspect cor triatriatum?

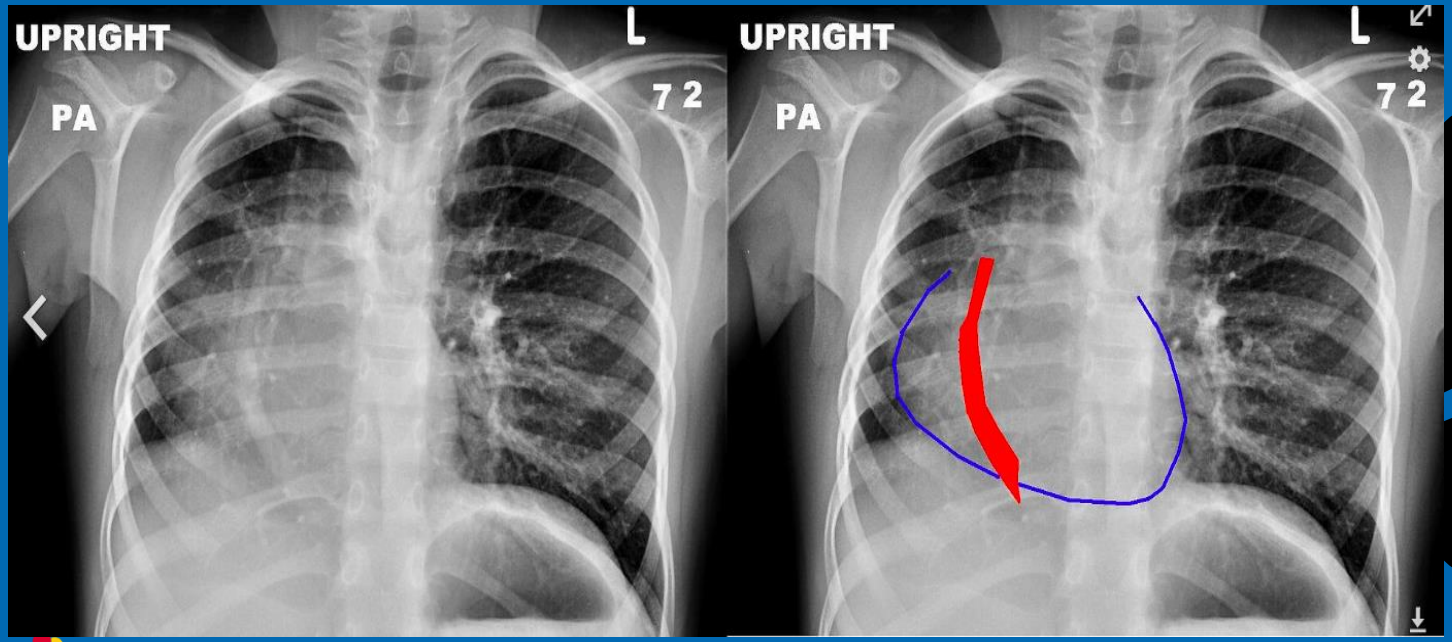
- Clinically:
  - Exercise intolerance
  - Exertional syncope
  - Murmur
- Echocardiographically:
  - Membrane above LA appendage
  - Flow disturbance in LA cavity
  - If chronic: evidence of elevated PA pressures (TR jet, septal flattening)




HD
FPS: 61
f: 2.3 MHz/4.6 MHz
P: 0 dB
AG(t): -3 dB
Compr: 69 dB
DDP: 2.3
D: 12.0 cm
Soft
10
8
6
4
V
92
HR

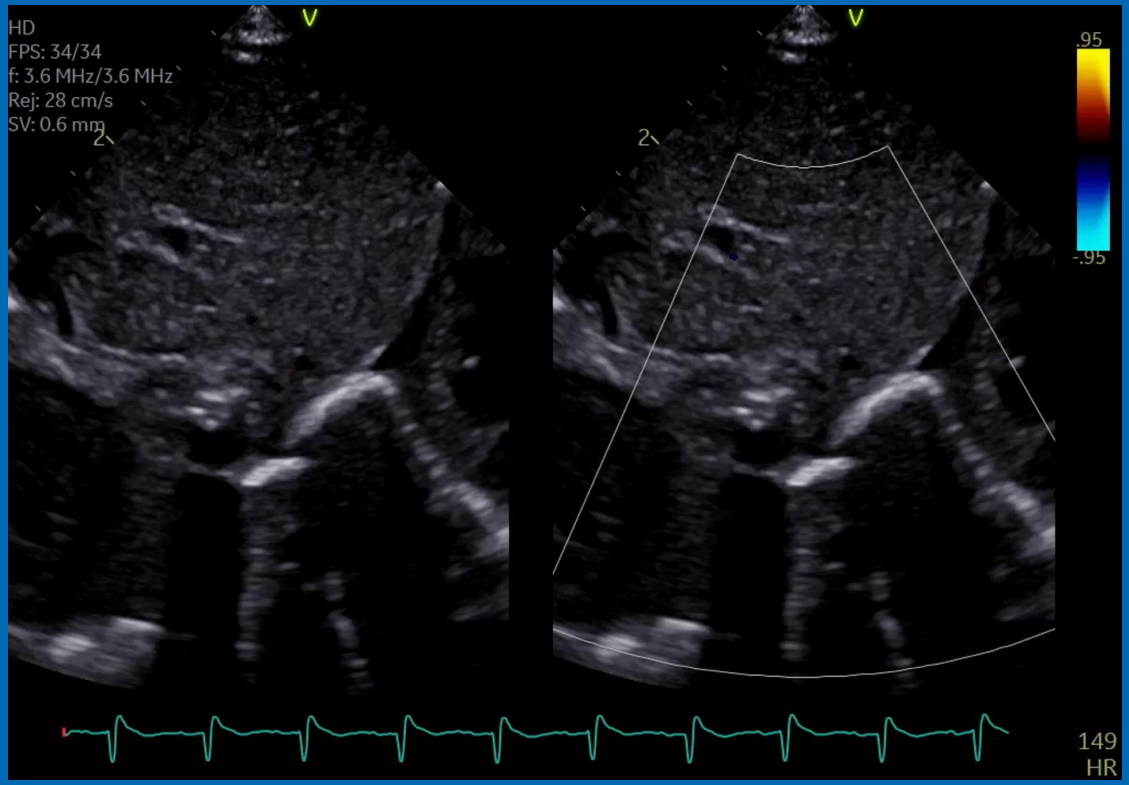

HD
FPS: 26/26
f: 3.1 MHz/3.1 MHz
P: 0 dB
AG(t): -3 dB
Compr: 69 dB
DDP: 2.3
D: 12.0 cm
Soft
.69
-.69
10
5
5
97
HR

Nice-to-know: PAPVR (Scimitar Syndrome)

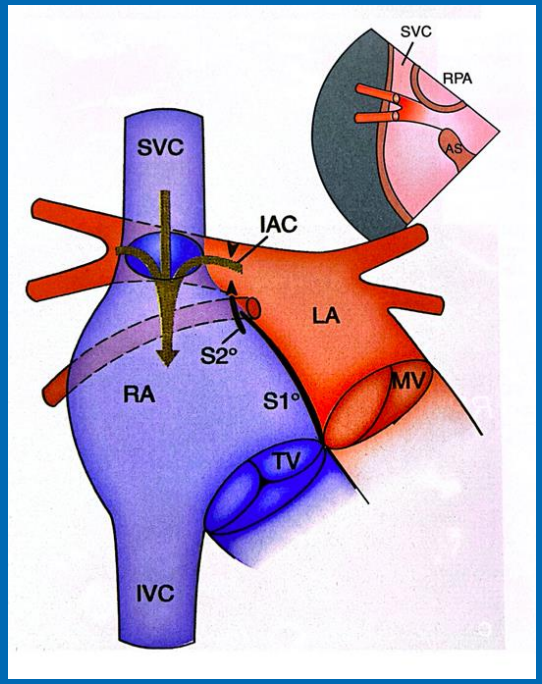


Some veins drain to the wrong place

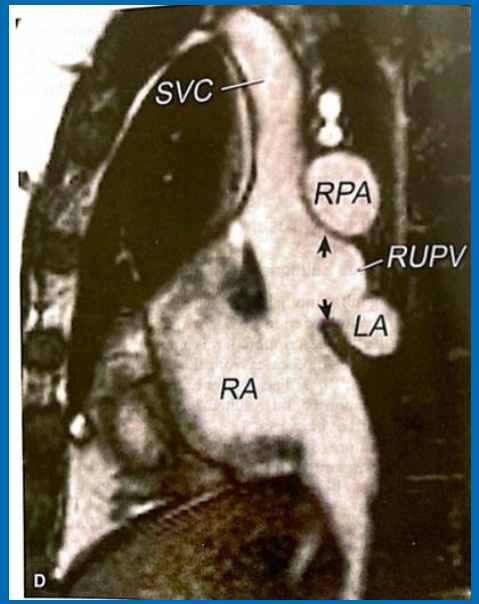
wikipedia.org

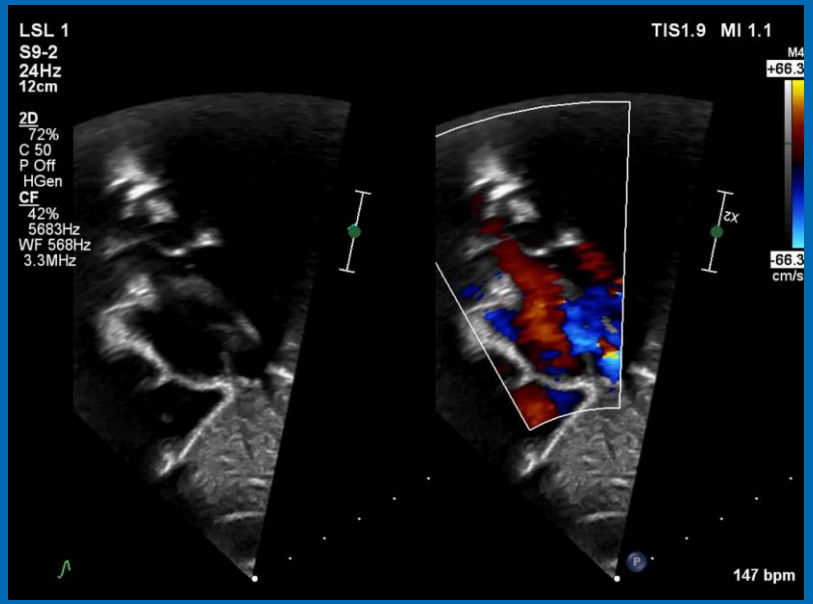

HD
FPS: 34/34
f: 3.6 MHz/3.6 MHz
Rej: 28 cm/s
SV: 0.6 mm
.95
-.95
149
HR

Nice-to-know: superior sinus venosus defect (RUPV PAPVR)



Superior sinus venosus defect





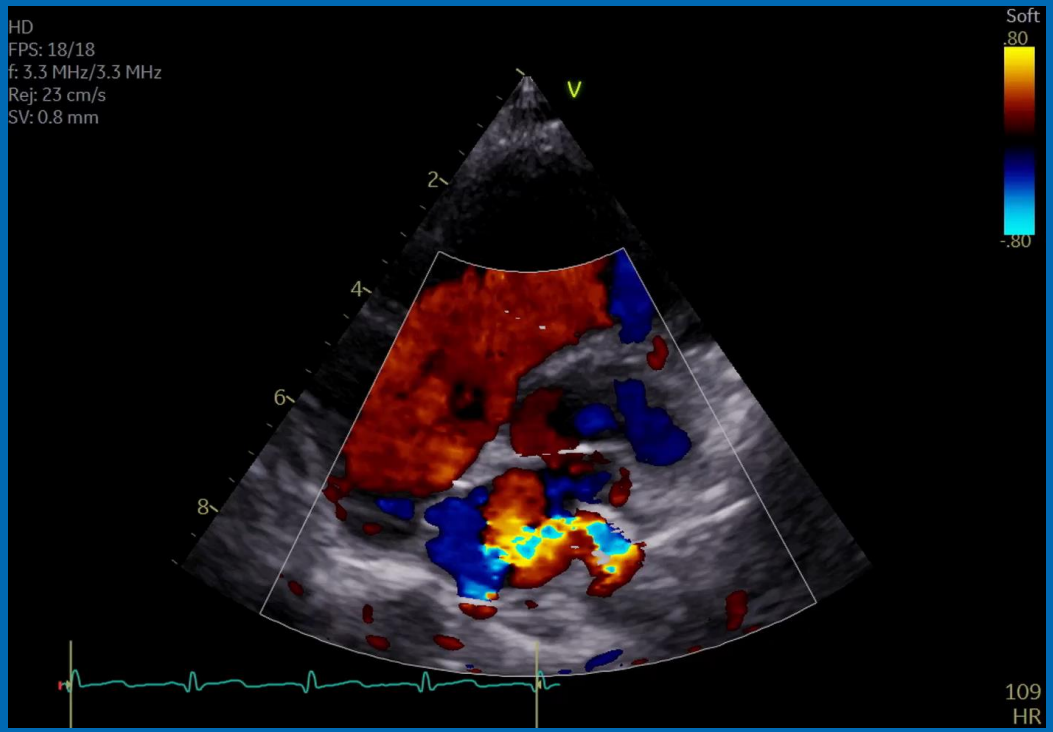
How to suspect PAPVR clinically

- Depends on how many veins drain anomalously
- Clinically silent → murmur, exercise intolerance
- Late: arrhythmias due to right heart dilation, PH due to chronic volume overload

How to suspect PAPVR by echo

- Right atrial and ventricular dilation without evidence of atrial level communication
- Bigger adolescents and adults, especially those at genetic risk for PAPVR may need CT or MRI to rule out if difficult windows (e.g. Turner Syndrome – up to 25%!)

A word about patients whose pulmonary veins have been intervened on:



- High rate of re-stenosis: 15-20%
- 40% three-year mortality if re-intervention needed ☹
- Look in medical history, they may be a cath lab frequent flyer with recurrent stenosis

Take-home points

- Pure R to L atrial septal shunt
- Find the veins!

- Obstructive membrane in LA
- E/o PH

- Right heart dilation with intact atrial septum

References

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Thank you!