Bits and Bobs – secondary causes of heart problems

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Not the heart…

Dextroposition

- Heart in the right chest with the apex to the left
- Often caused by left sided chest mass pushing heart over
- = pushed to the right
- Left CPAM

Levoposition

- Heart deviated further than usual into the left chest
- Apex pointing to the left
- Cardiac axis normal

- = Pushed to the left

- Right CPAM

Agenesis of the ductus venosus

- The duct controls blood flow from the UV to the fetus

- The UV connects directly to the fetal venous system –
  - To the ileac vein/IVC/other
  - Via the hepatic veins
  - To the heart
- Findings:
  - Large abnormal venous connection
  - Cardiomegaly
  - Hydrops
  - **High output cardiac failure**
- Treatment: delivery

Isomerism

- Abnormality of sidedness – right or left atrial isomerism
- Right – commonly have structural heart disease
- Left – heart may appear normal, but can have bradycardia (heart block)
- Non-cardiac issues:
  - Midline liver (biliary atresia – LAI)
  - Absent/multiple (sometimes non functional) spleens
  - Malrotation
  - Ciliary dyskinesia (respiratory issues)

Rhabdomyomas

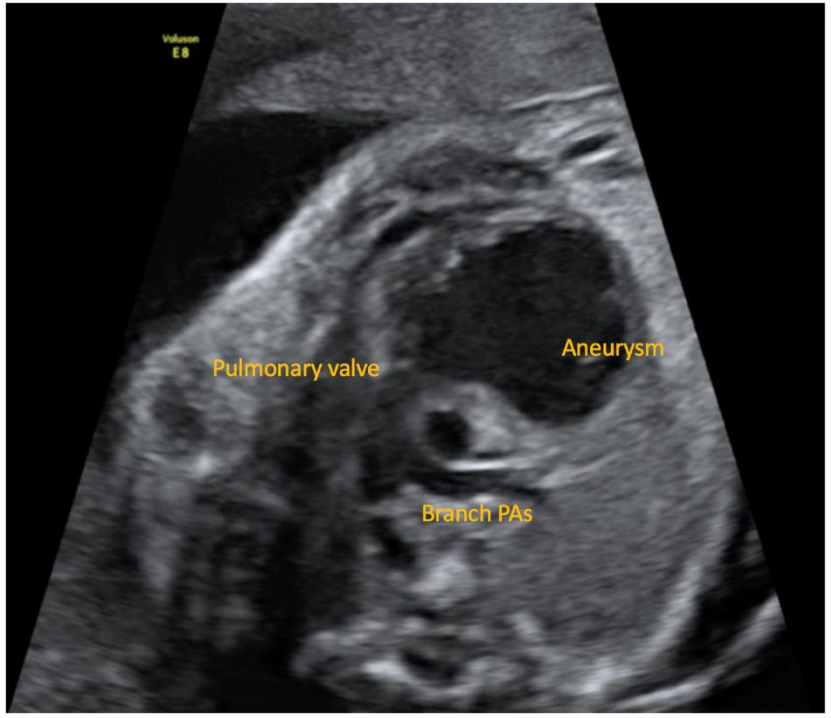
- **Multiple**
- Homogeneous
- Benign
- Get bigger until 32 weeks then slowly regress
- Can cause inflow/outflow obstruction
- Can have arrhythmias (SVT/VT/ectopy)
- Strong association with tuberous sclerosis
- Seizures, developmental delay, renal and cerebral tubers (wide spectrum)
- ? Parental history

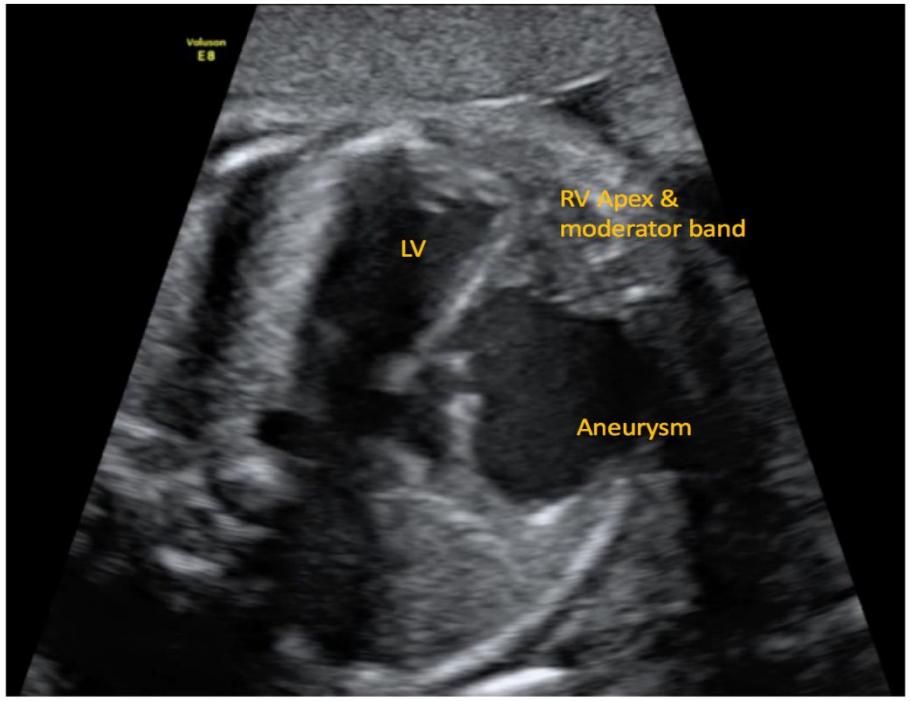
Aneurysms

- Thin walled outpouchings
- Broad based
- Poorly contracting

- Risk of arrhythmia, clot, hydrops, IUD

- ? Cause - ? Ischemia ? Infection





Ductal aneurysm

- Fetal echo findings:
- Long, tortuous, widened ductus arteriosus
- Demonstrated in the three vessel view and the sagittal plane
- The duct bulges out (may be saccular or fusiform) to the left of the aorta

Ductal aneurysm

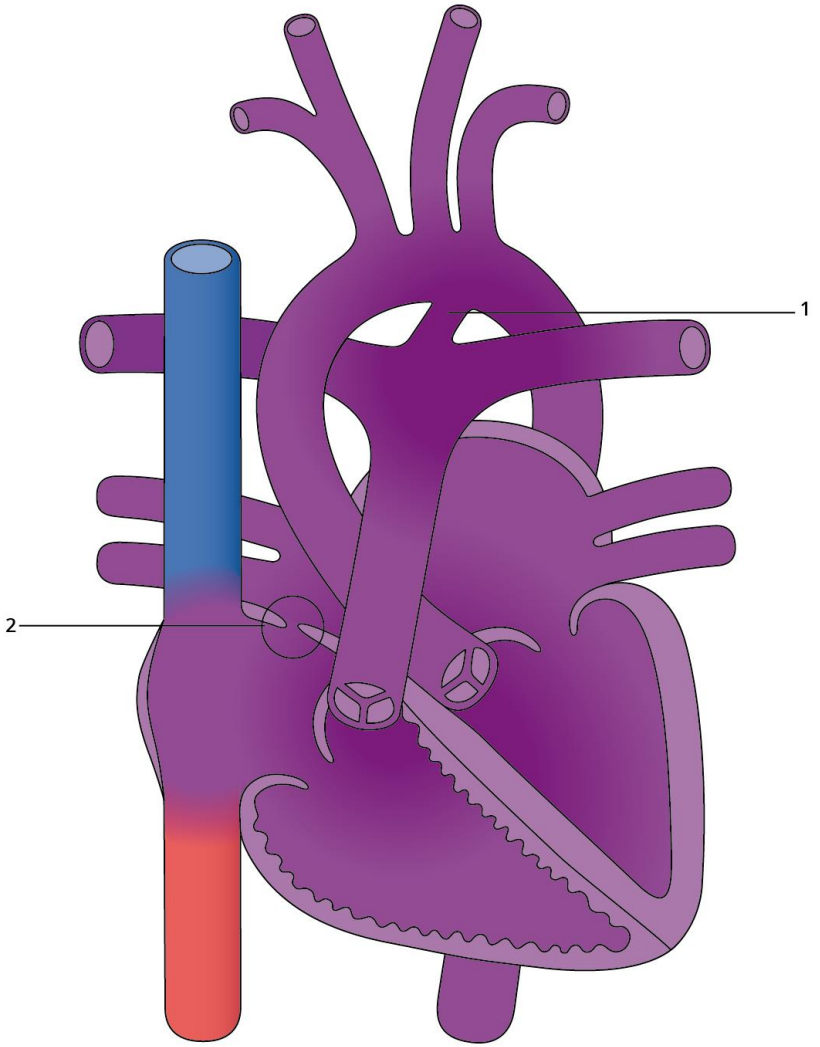
- The duct gets longer, wider and more tortuous as pregnancy progresses
- Ductal aneurysms happen increasingly in the third trimester
- By term, approx. 2% have an aneurysm and 8% have neonatal ductal aneurysms
- Mostly asymptomatic
- Some can rupture/fill with clot/embolise/compress surrounding structures
- Association with connective tissue disorders, mat GDM, SGA/LGA

Ductal constriction: fetal echo findings

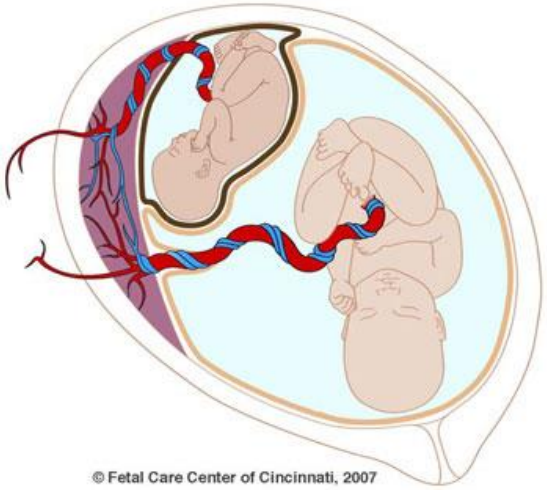
- Small appearance of the duct in 2D imaging
- Turbulent colour flow in the duct
- High velocity flow on pulsed Doppler with:
  - High systolic velocity
  - High diastolic velocity
  - Continuous flow throughout systole and diastole
  - Low pulsatility index :<1.8)

Ductal constriction

- The fetal lungs have high resistance →
- The majority of blood from the right ventricle goes through the duct to the lower body/placenta
- **Ductal constriction:**
- Results in high RV pressure →
  - In fetuses - PR, TR, dilated dysfunctional RV, hydrops, IUD
  - In neonates – pulmonary hypertension, cyanosis, death
- Is rare
- Usually happens in the third trimester
- Often idiopathic – but can be caused by NSAIDs and green tea, etc. (reversible causes)



Twin-twin transfusion syndrome



- Mono-chorionic diamniotic pregnancies
- “Donor” twin – small/absent bladder, oligohydramnios, abnormal Dopplers (UA)
- “Recipient” twin – hypertrophy and reduced fx (esp RV), tricuspid regurgitation, pulmonary stenosis, abnormal Dopplers (veins)

- Can be fatal
- Treatment – if bad enough – laser therapy → improvement in cardiomyopathy, pulmonary stenosis may improve

https://www.cincinnatichildrens.org/service/f/fetal-care/conditions/twin-twin-transfusion-syndrome

Ectopia cordis

- Very rare
- Heart either fully or partially outside the chest
- Can have chromosome probs (e.g. Trisomy 18)
- Outcomes tend to be poor
- May also have additional defects – omphaloceles, or pentalogy of Cantrell:
  - Diaphragmatic hernia
  - Defect of pericardium
  - Defect of anterior abdominal wall
  - **Intracardiac defects** (VSDs, tetralogy of Fallot, …)

Summary

- Heart position
- Agenesis of the ductus venosus
- Isomerism
- Tumors
- Aneurysms
- Ductal aneurysm
- Ductal constriction
- Twin-twin transfusion
- Ectopia cordis