MMed and DCH Lectures

Weekly by Zoom

Prof Trevor Duke

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Congenital heart disease II

July 20, 2020

Prof Trevor Duke

Congenital heart disease: pathophysiology

Week 1. Acyanotic (75%)

- Left to right shunts (VSD, PDA, ASD, AVSD)
- Left ventricular outflow tract obstruction

Week 2. cyanotic

- Obstructed right ventricular outflow tract
- Mixing lesions

Pulmonary hypertension

Presentations of cyanotic CHD

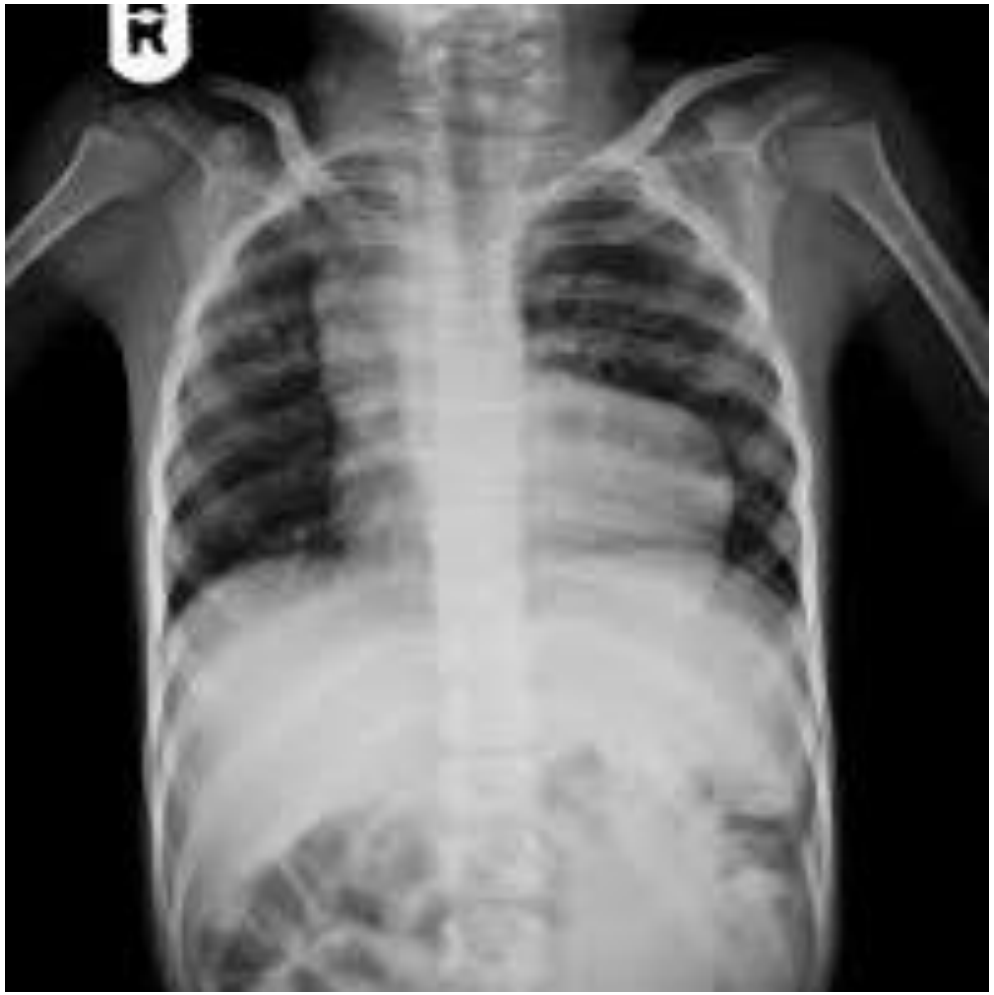
- The infant or child with cyanotic episodes
- The newborn who is cyanosed or soon after at birth

Case

- 4 month old boy presents with seizures and collapse occurring when upset and crying
- Normal looking infant, pulses normal
- Chest hyperinflation but auscultation clear, no hepatomegaly
- Mild cyanosis, SpO_2 88%
- Differential diagnosis…?

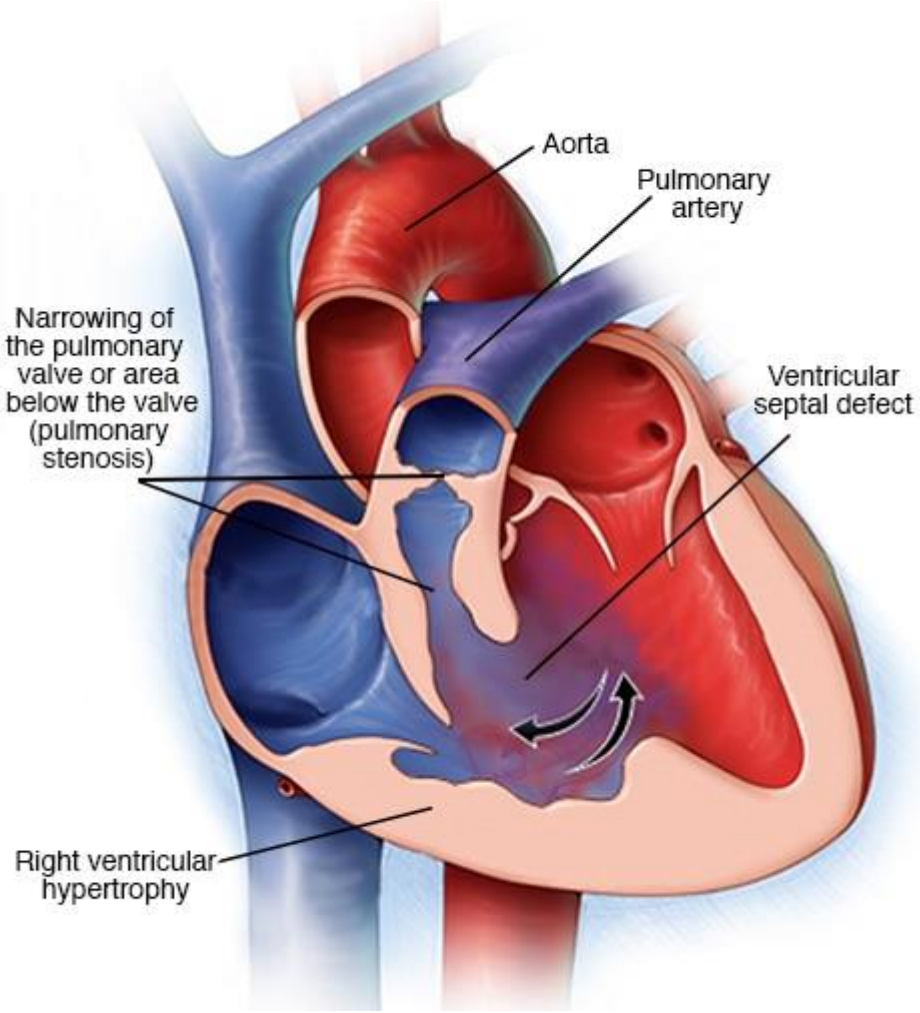
Differential diagnosis

- Respiratory event
  - breath holding
  - aspiration
- Neurological events?
  - Seizures, infantile spasms?
- Tetralogy of Fallot
  - Cyanosis and desaturation, especially when upset
  - Murmur of PS
  - Chest xray
- Arrhythmia?
  - ECG



Cyanotic heart disease

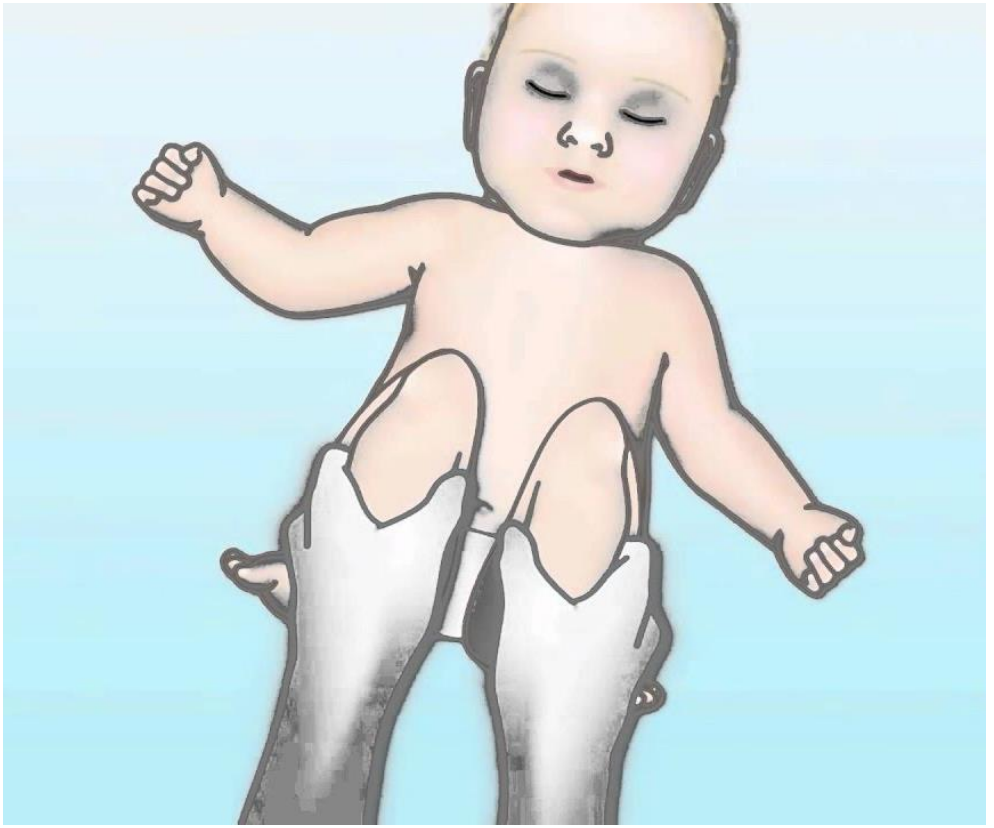
- Deoxygenated blood from the systemic veins (SVC / IVC) is being directed back into the systemic arterial circulation, bypassing the lungs (right-to-left shunt).
- In Tetralogy of Fallot
  - Pulmonary blood flow reduced by obstruction to RVOTO (PS and RV *infundibular* hypertrophy)
  - Septal defect (VSD) which means blood preferentially follows the "path of least resistance" through the "over-riding" aorta.

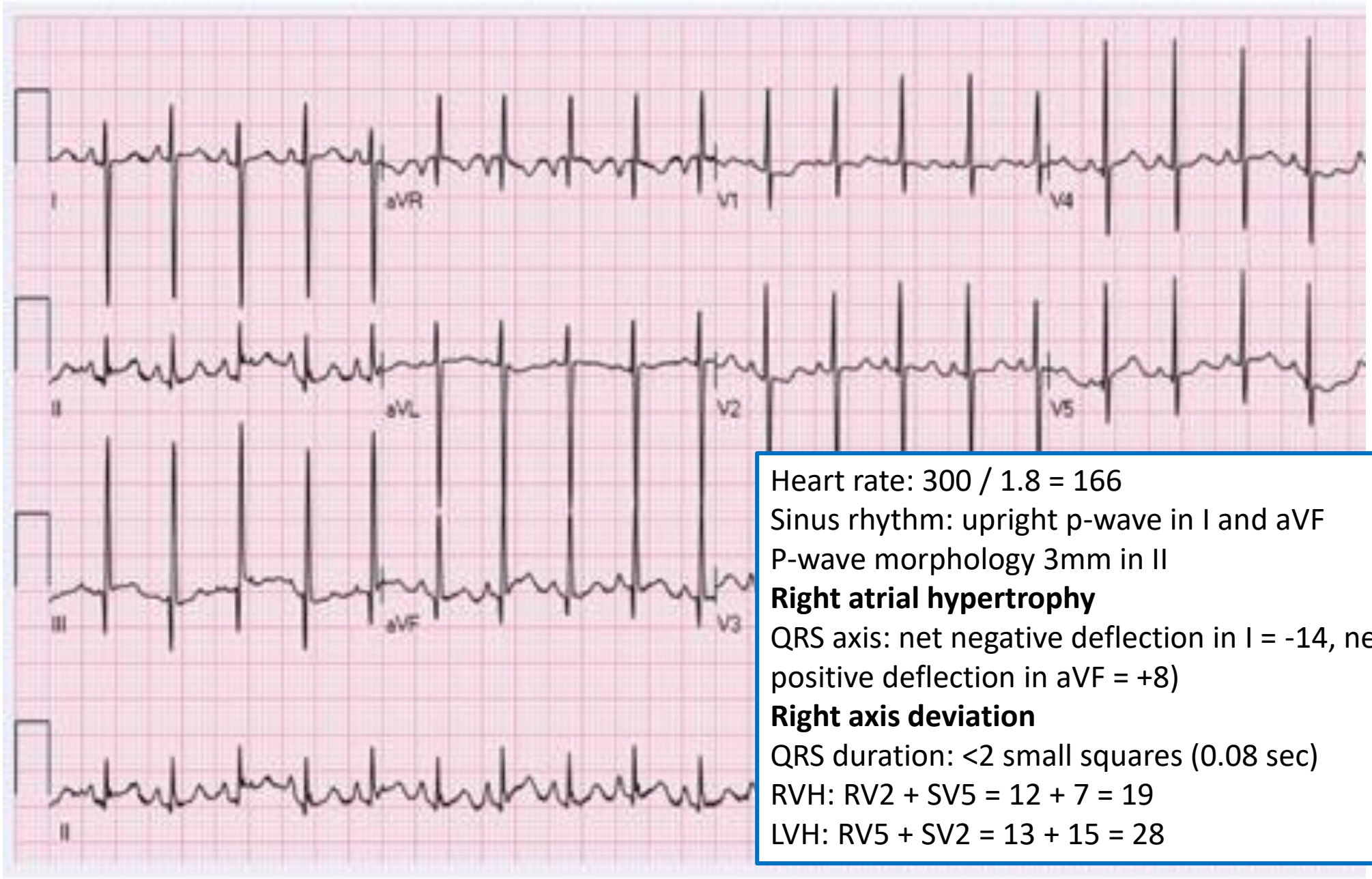


- Normally *not* cyanosed at birth
- SpO_2 may be low or normal in first week of life
- Development of cyanotic spells in first 6 months of life
- ESM upper left sternal edge (pulmonary area), radiates to back

Treatment of cyanotic spells

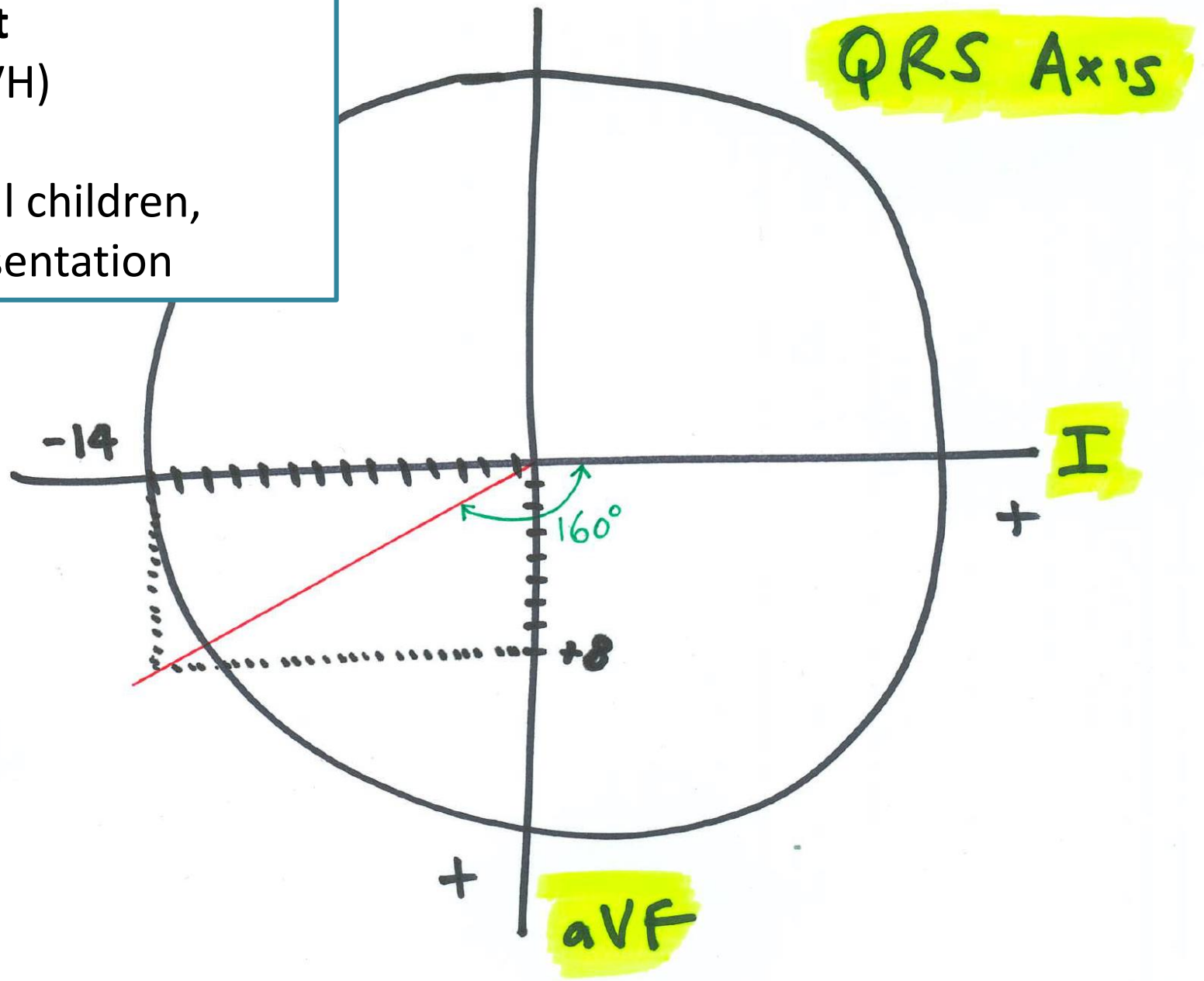
- Calm the child if distressed
- Oxygen
- IM morphine
- IV fluid bolus
- Propranolol
- ‘Squatting’ / knee-chest position increases SVR → less R-L shunt → increased pulmonary blood flow





Heart rate: 300 / 1.8 = 166
Sinus rhythm: upright p-wave in I and aVF
P-wave morphology 3mm in II
Right atrial hypertrophy
QRS axis: net negative deflection in I = -14, net positive deflection in aVF = +8)
Right axis deviation
QRS duration: <2 small squares (0.08 sec)
RVH: RV2 + SV5 = 12 + 7 = 19
LVH: RV5 + SV2 = 13 + 15 = 28

ECG: Tetralogy of Fallot
Right axis deviation (RVH)
Peaked p-waves (RAH)
Not all are present in all children, especially at initial presentation



QRS axis age changes

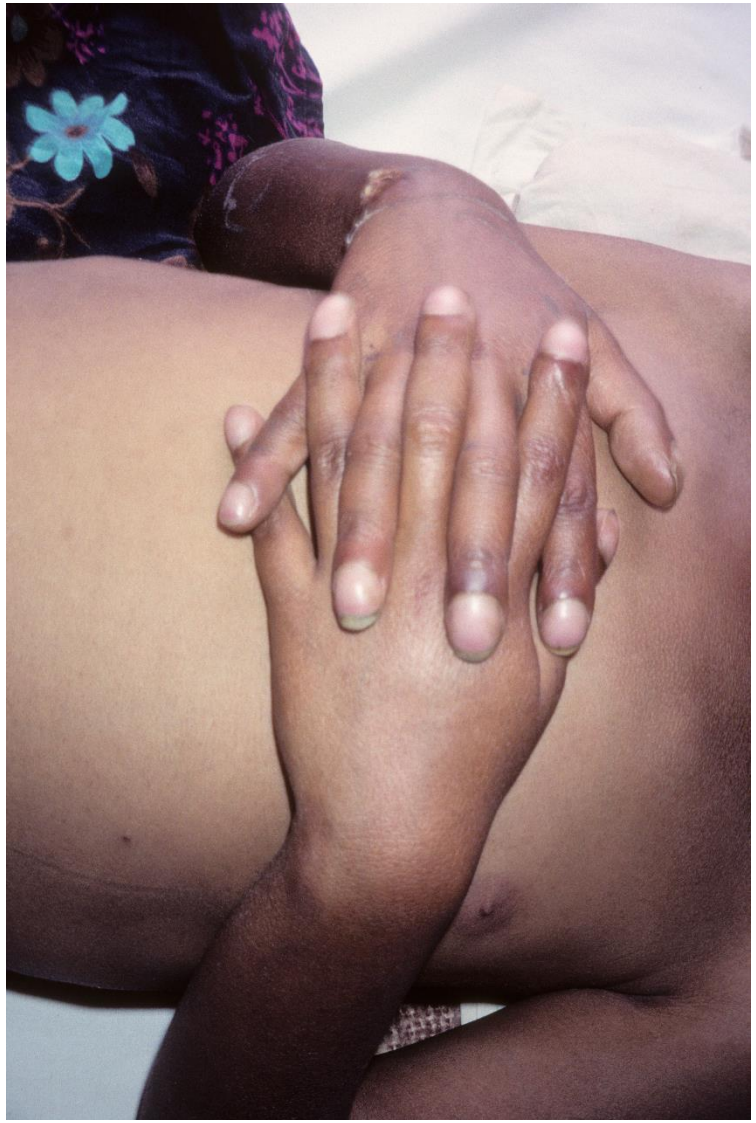
Age	Mean	Range
1 - 3 months	+70°	+10° to +125°
3 months - 3 years	+60°	+10° to +110°
3 years +	+60°	+20° to +120°
Adult	+50°	-30° to +105°

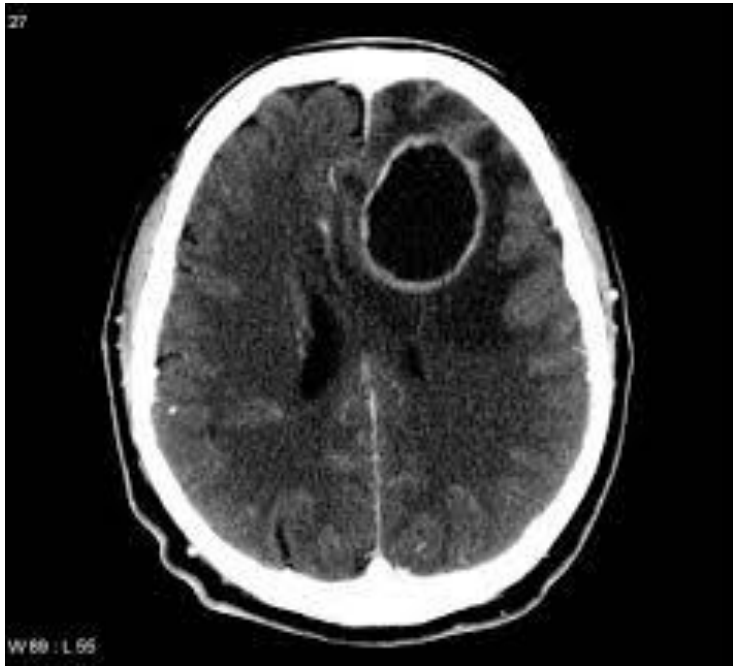
Complications of Tetralogy of Fallot

Stroke

Cerebral abscess

- Right to left shunt (no lung filter for bacterial micro-emboli)
- Hypoxia increases anaerobic bacteria
- Hyperviscosity leads to slow blood flow in brain





Management of Tetralogy of Fallot

- Advise parents on home care of cyanotic spells
- Propranolol
- Reduce hyperviscosity (if Hb >15-17 g/dL)
- Surgery:
  I. complete repair
  II. shunt (subclavian A to pulmonary A on same side) – prosthetic tube graft (Blalock-Taussig-Thomas* shunt)
- Long term complications (post correction):
  - Recurrent RVOTO
  - Pulmonary regurgitation – may need valve replacement
  - Arrhythmias (30%) in adults (atrial and ventricular)

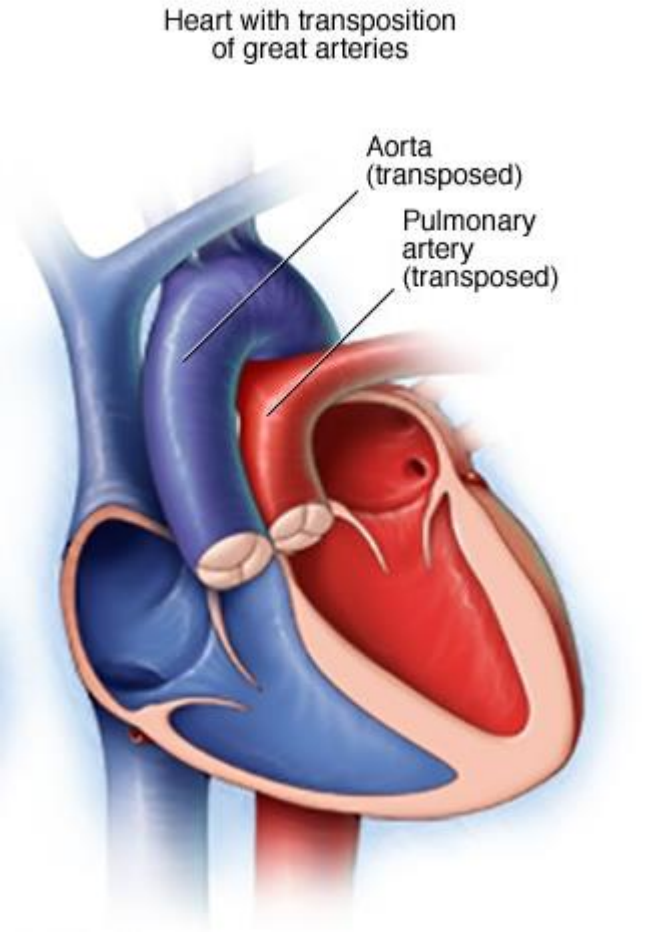
* Vivien Thomas. “Something the Lord Made” Alfred Blalock, Helen Taussig

The newborn with cyanosis

1. The neonate who is *well at birth* then collapses with *shock* in the first week
    I. Congenital heart disease (duct dependent lesion, such as coarctation, HLHS)
    II. Sepsis
    III. Metabolic – inborn error of metabolism hyperammonaemia
    IV. Asphyxia (SIDS)
2. The neonate who is cyanosed *from the first day of life*
    I. Birth asphyxia
    II. Pneumonia / sepsis
    III. Congenital heart disease (e.g. Transposition of the Great Arteries)
    IV. Other congenital airway / lung abnormalities (such as diaphragmatic hernia)

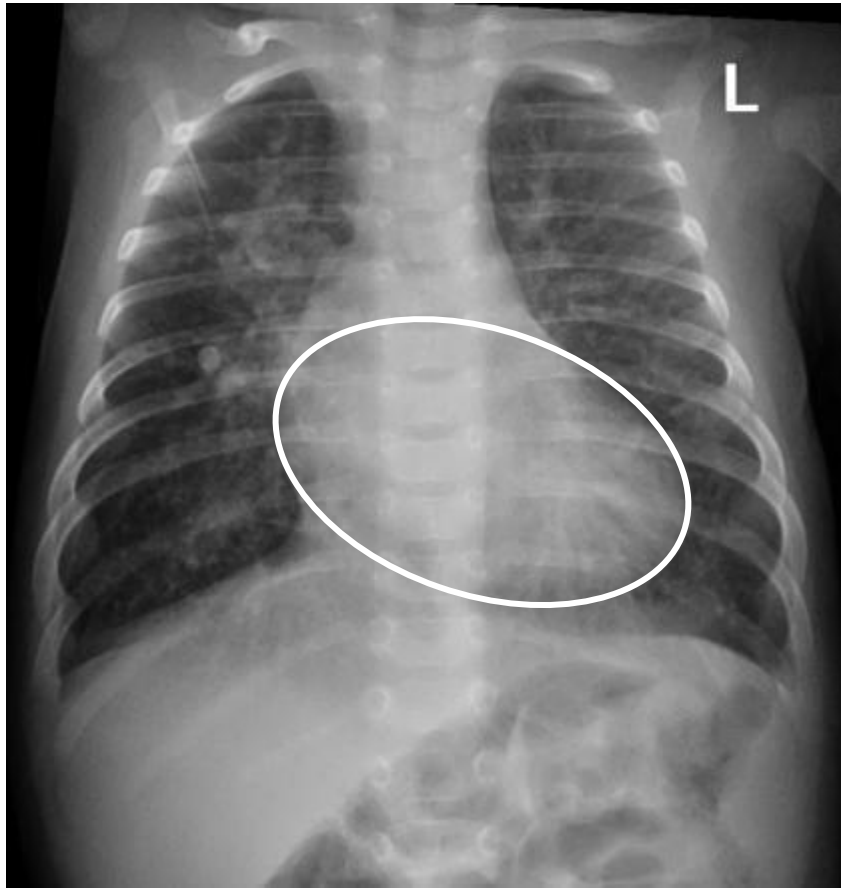
Transposition of the Great Arteries

- Aorta arises from RV
- Pulmonary artery arises from LV
- *Parallel* circulations – incompatible with life
- Need mixing to survive even a few minutes of extra-uterine life: PFO, duct, VSD



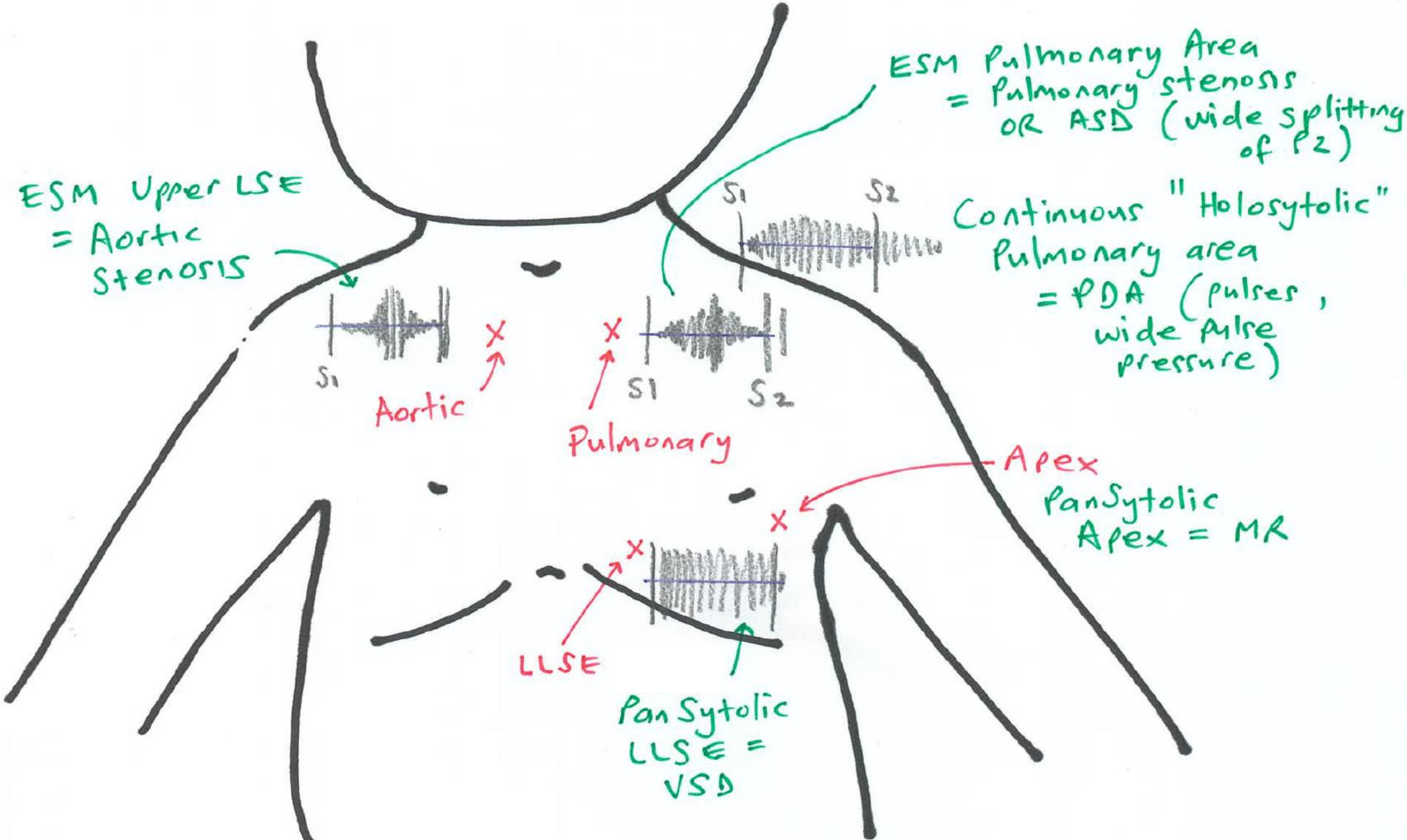
Presentation of TGA

- Cyanosis in early hours of life, progresses over first days
- Metabolic (lactic) acidosis
- Baby is blue, tachycardia, right ventricular impulse ++
- Often no murmur
- Cardiac surgery required but baby can be normal (usually no chromosome or other problems)

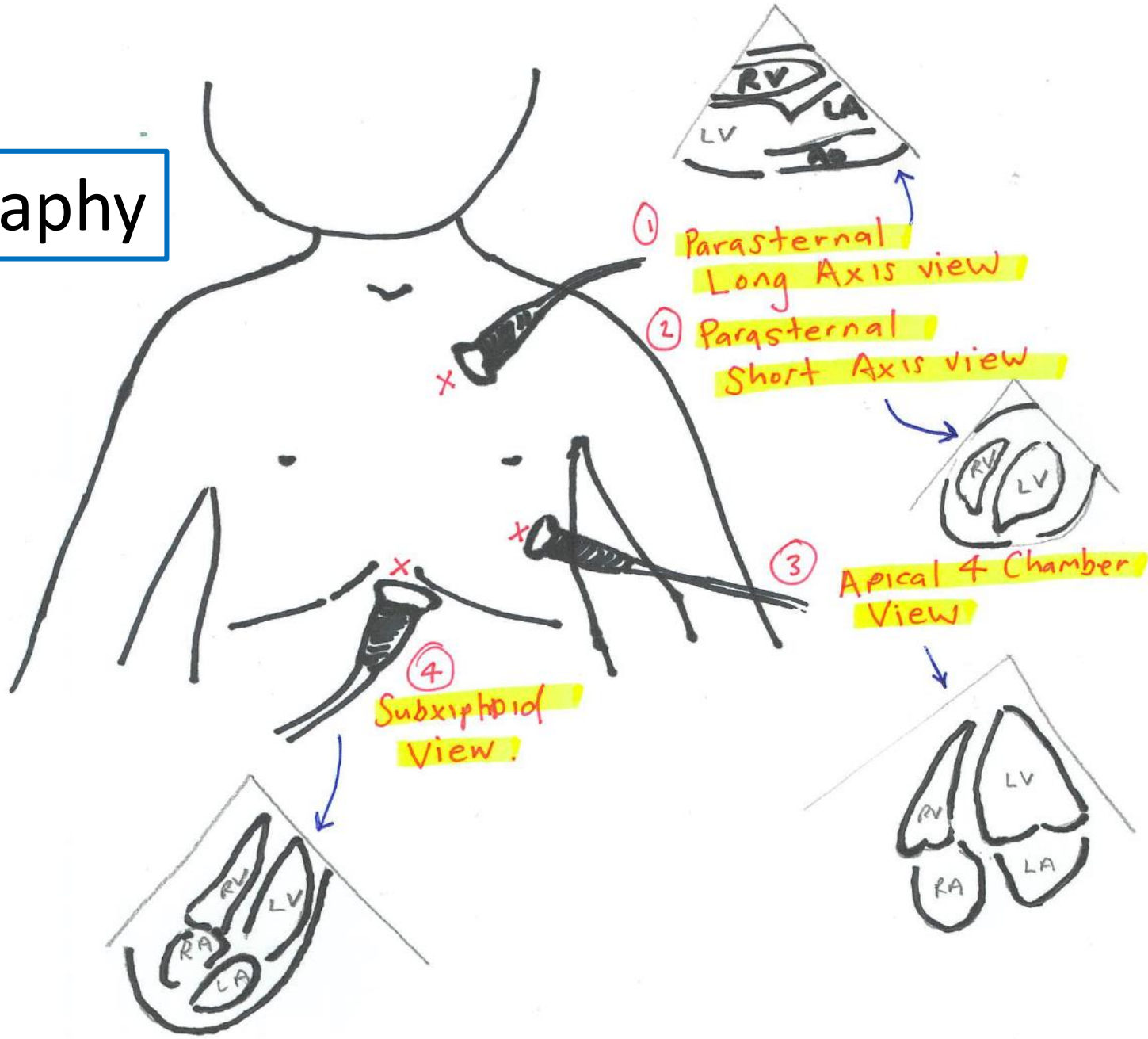


The newborn with cyanosis - CHD

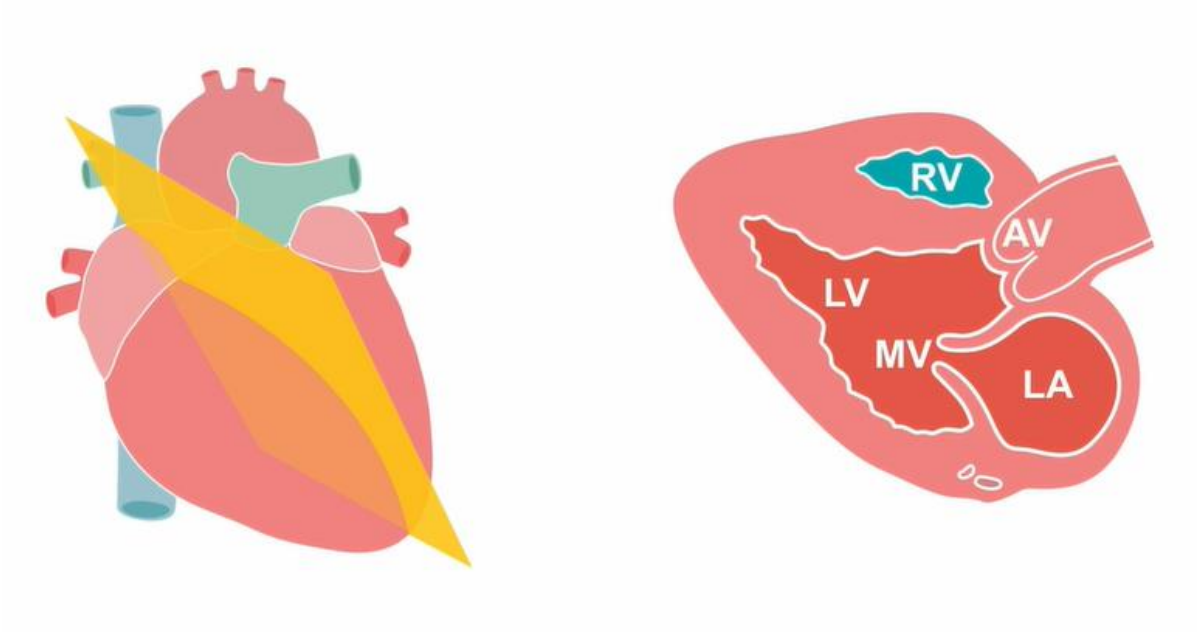
- DDx: Duct dependent pulmonary circulations
  - pulmonary atresia
  - tricuspid atresia
- Truncus arteriosus

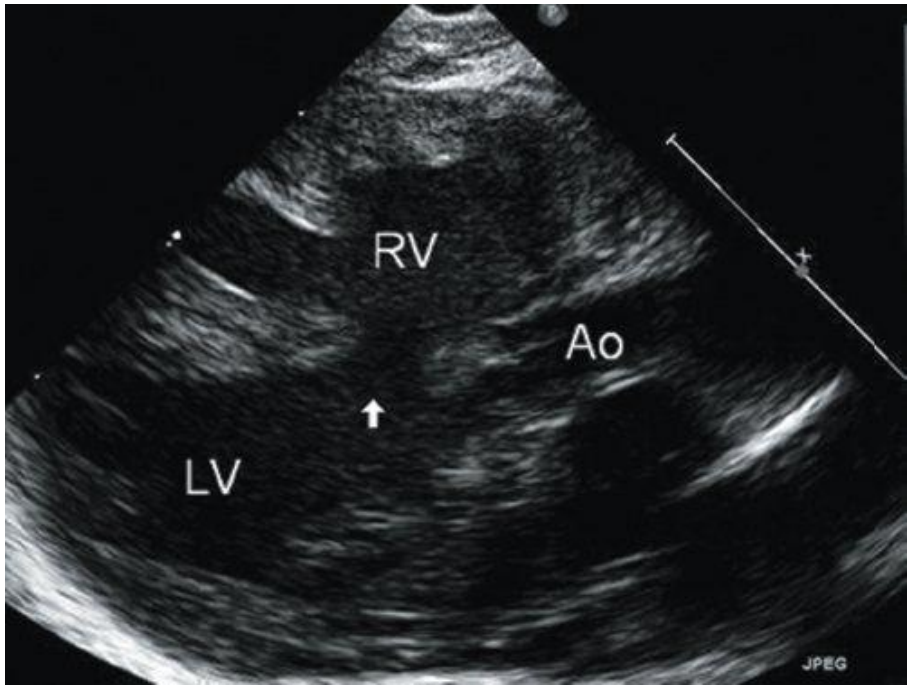

ESM Pulmonary Area
= Pulmonary stenosis
OR ASD (wide splitting
of P2)
ESM Upper LSE
= Aortic
Stenosis
S1
S2
Continuous "Holosytolic"
Pulmonary area
= PDA (pulses,
wide pulse
pressure)
S1
Aortic
Pulmonary
S1
S2
Apex
PanSytolic
Apex = MR
LLSE
PanSytolic
LLSE =
VSD

Echocardiography



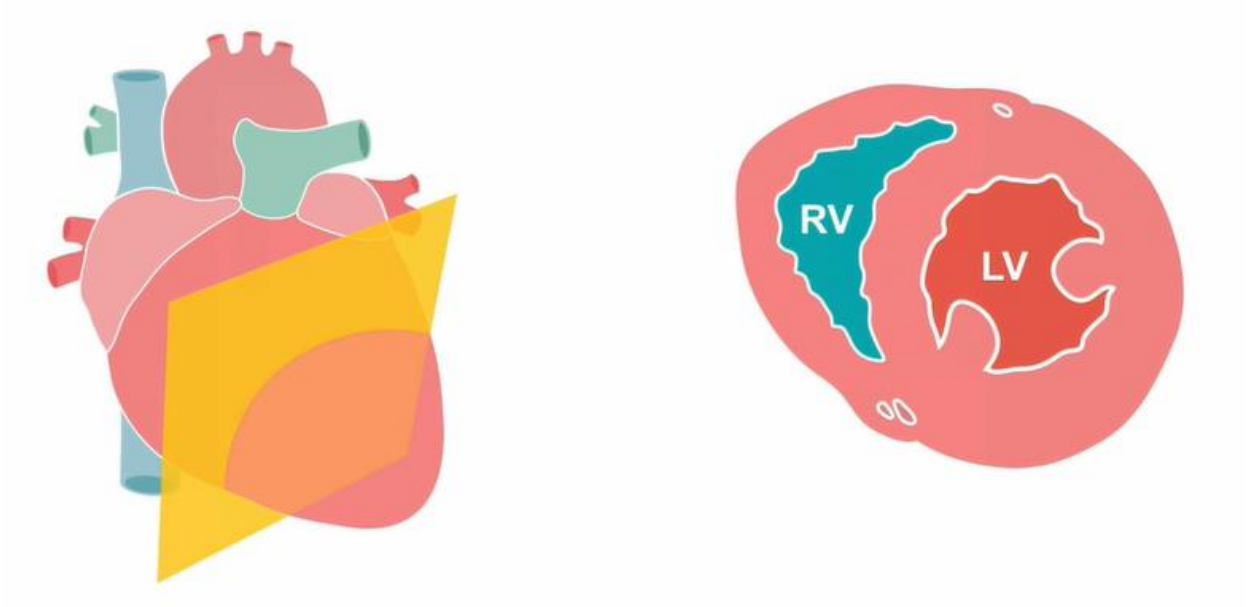
1. Parasternal long axis view

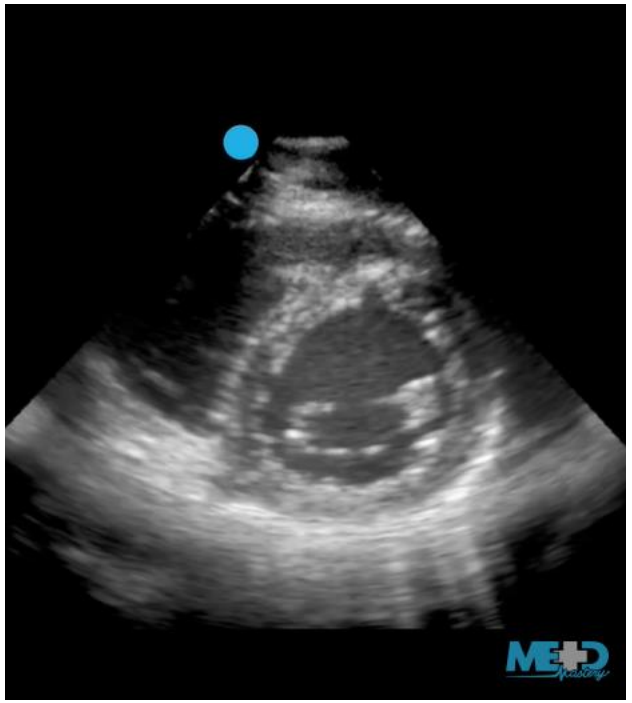




Probe placed to left of the lower third of the sternum 3rd / 4th ICS.
Probe marker pointing towards the right shoulder.

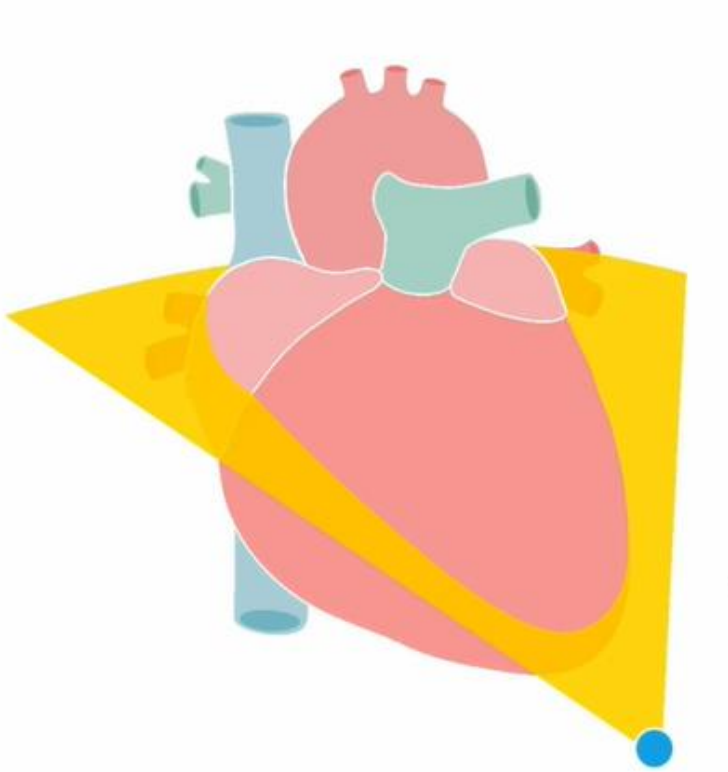
2. Parasternal short axis view

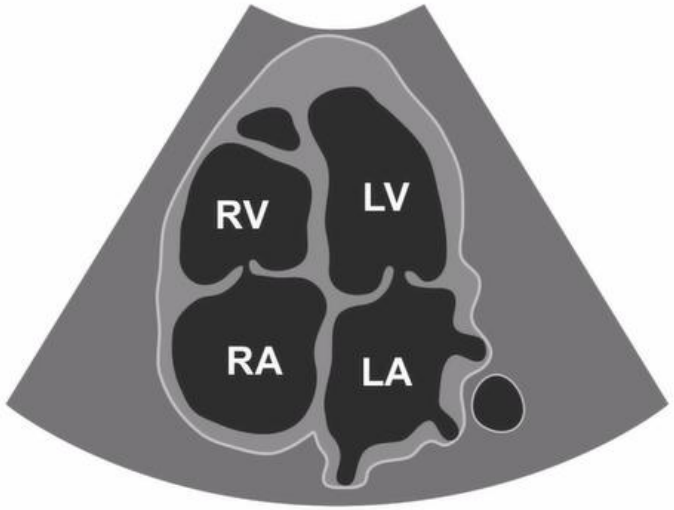


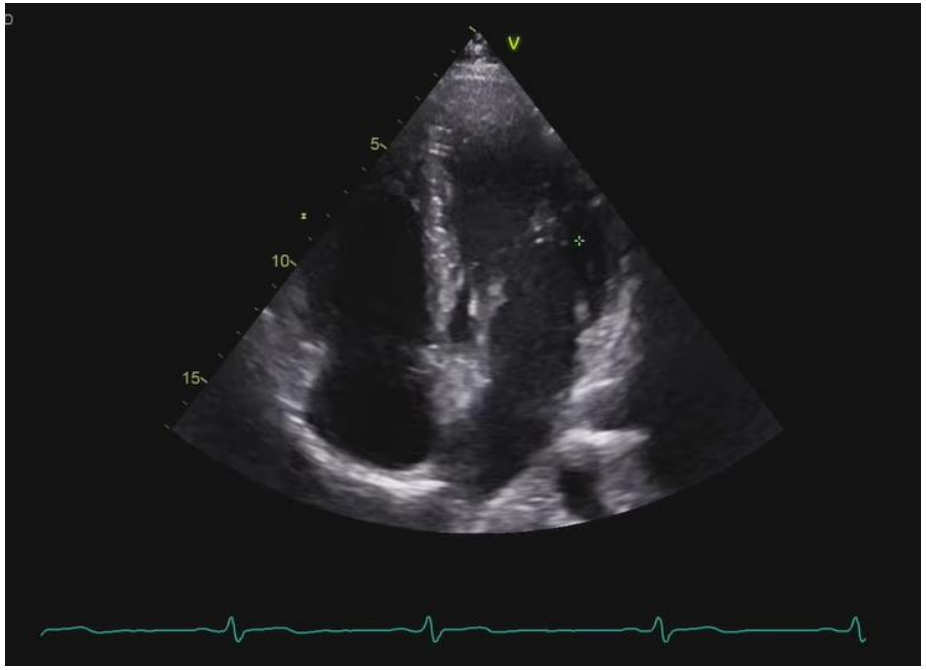


Probe placed to left of the lower third of the sternum 3rd / 4th ICS.
Probe marker pointing towards the left shoulder.

3. Apical 4-chamber view

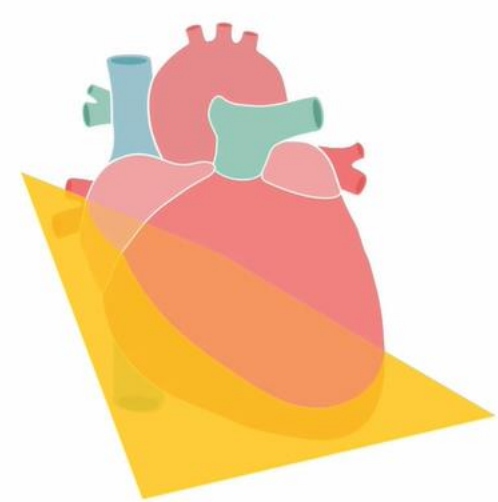


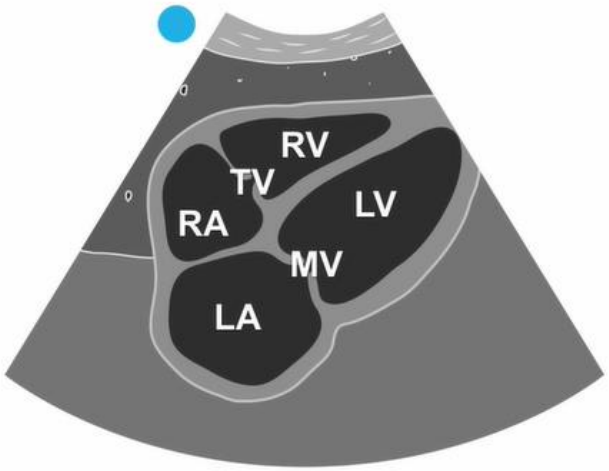


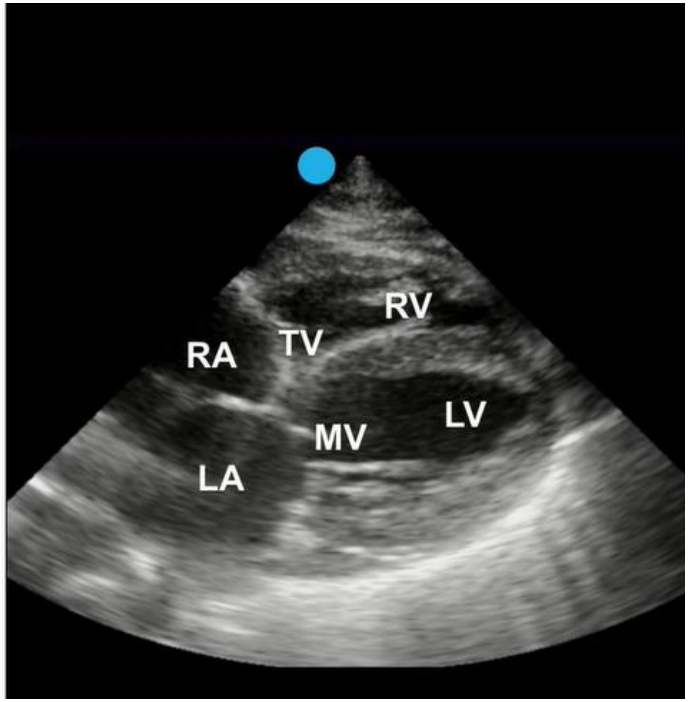


Place probe at apex, at an angle pointing towards the right shoulder.
The positioning marker on the probe faces the left shoulder

4. Sub-xiphoid view







Place probe under xiphoid process
Positioning indicator to right.

Pulmonary hypertension: key concepts

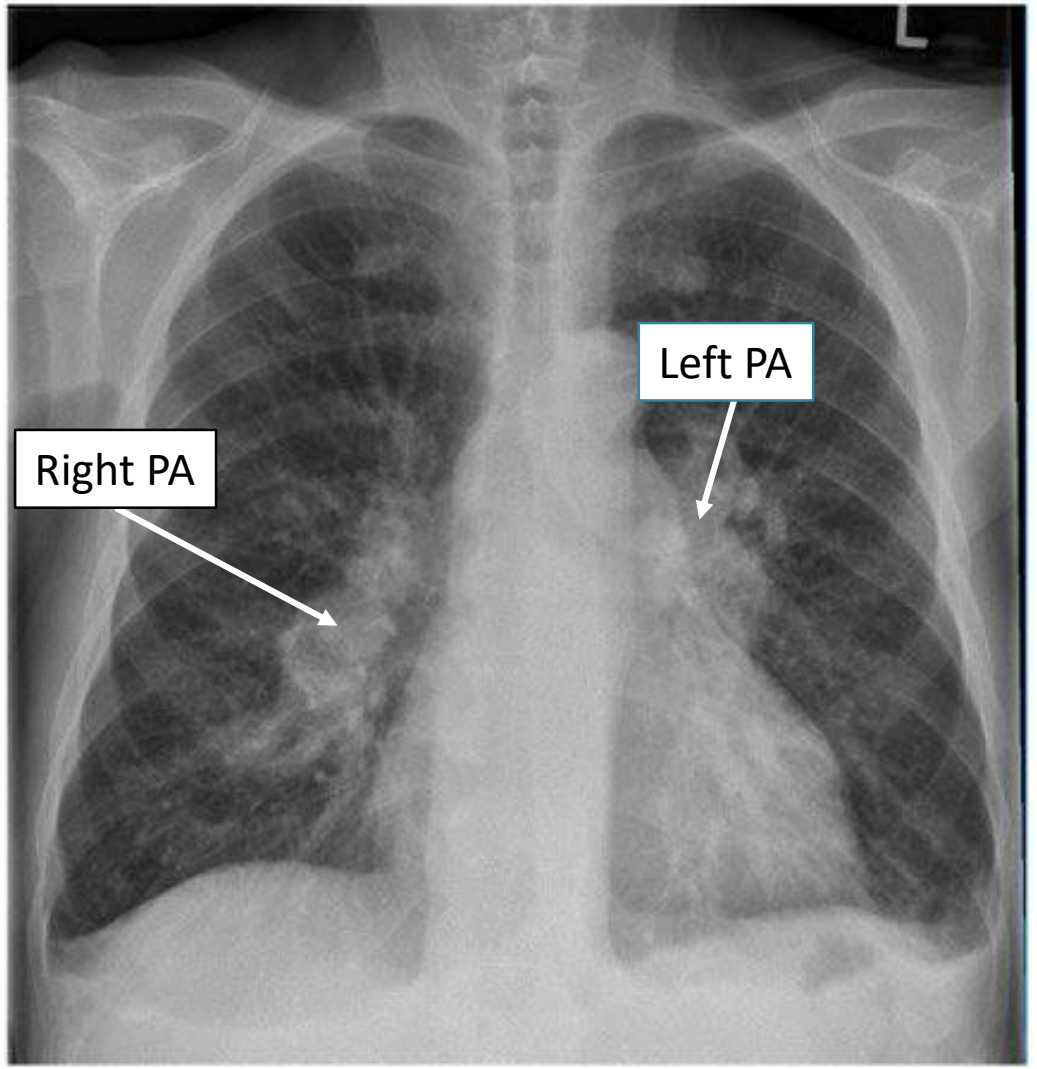
- Definition: PAP mean >20mmHg
- Pulmonary vascular resistance (PVR) ↓↓ rapidly after birth, but neonates remains susceptible to stimuli that increase PVR, e.g. hypoxia
- Primary or secondary
- Pulmonary *arterial* hypertension or pulmonary *venous* hypertension
- PHT affects the function of the right ventricle (a weaker pump than the LV)
- Conditions at the time of assessment: distress, agitation, hypoxia ↑↑PVR

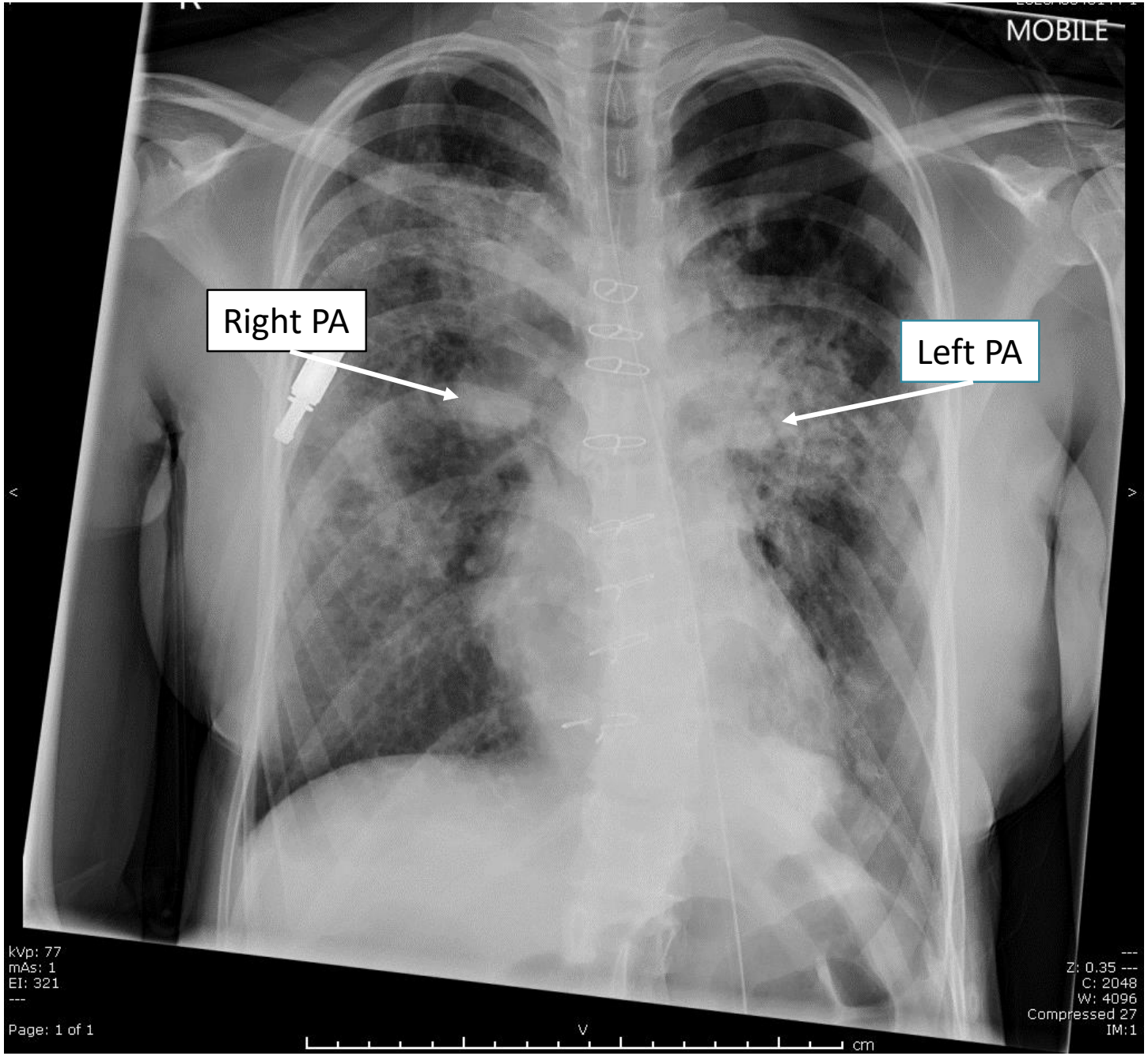
Pulmonary hypertension

- PPHN
- High pulmonary blood flow (VSD, ASD, PDA, AVSD)
- Pulmonary venous hypertension (mitral stenosis, TAPVD)
- Pneumonia in the highlands
  - Altitude associated hypoxic pulmonary vasoconstriction
- Chronic lung disease
  - Bronchopulmonary dysplasia, Pulmonary fibrosis (TB / HIV)
  - Pulmonary hypoplasia / CDH
- Pulmonary vascular problems
  - Pulmonary emboli, Sickle cell disease, COVID-19
- Other – scurvy

Pulmonary hypertension: x-ray changes

- Large pulmonary arteries
- Cardiomegaly (if right heart is failing)
- Pruning of pulmonary vascular marking (think proximal and disappear distally)

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Left PA
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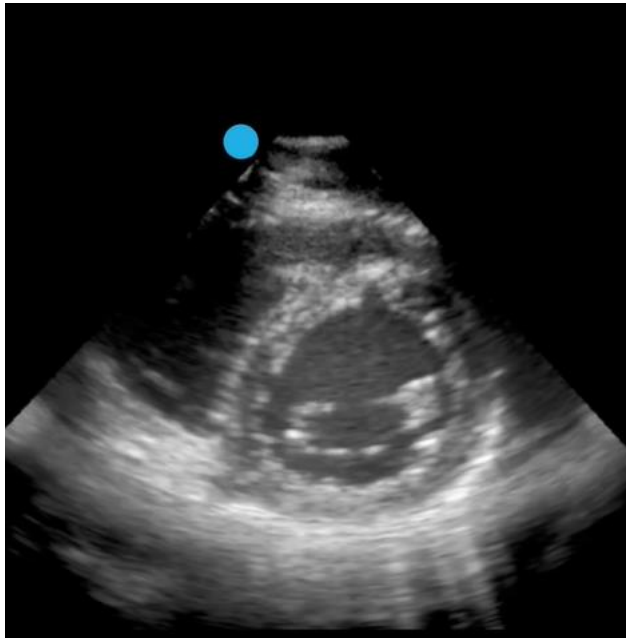
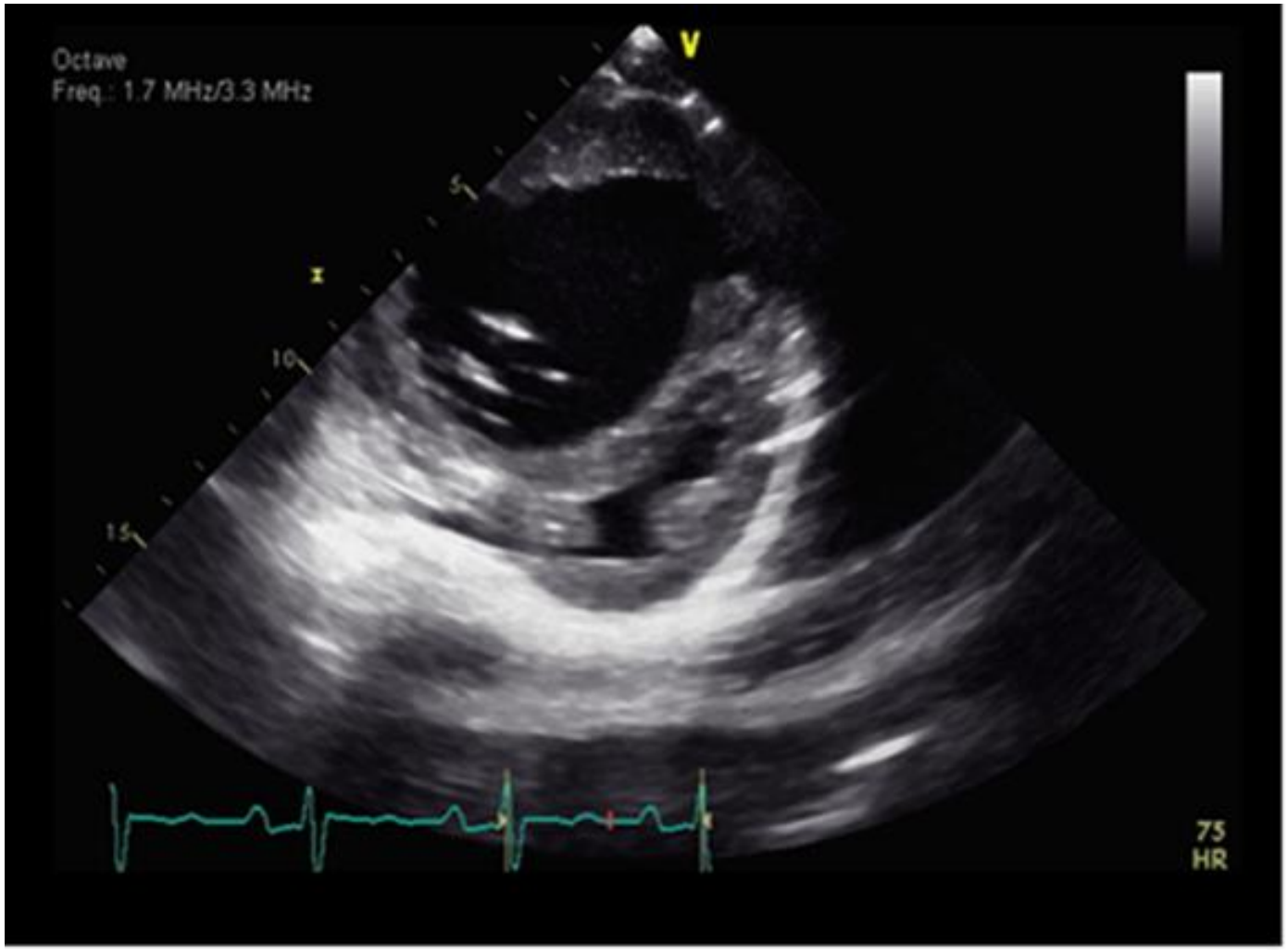
IM:1

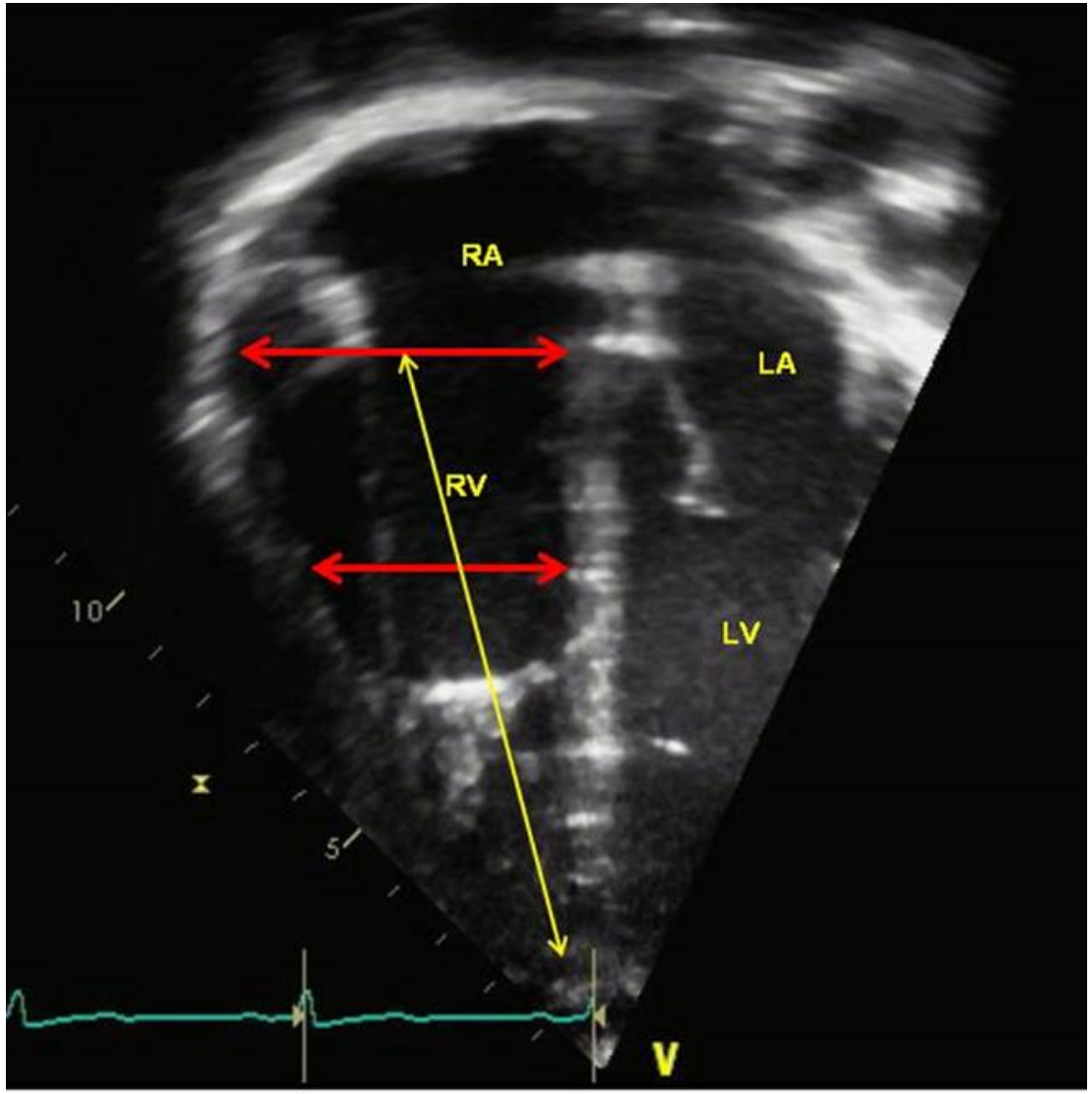
Pulmonary hypertension (parasternal short axis)

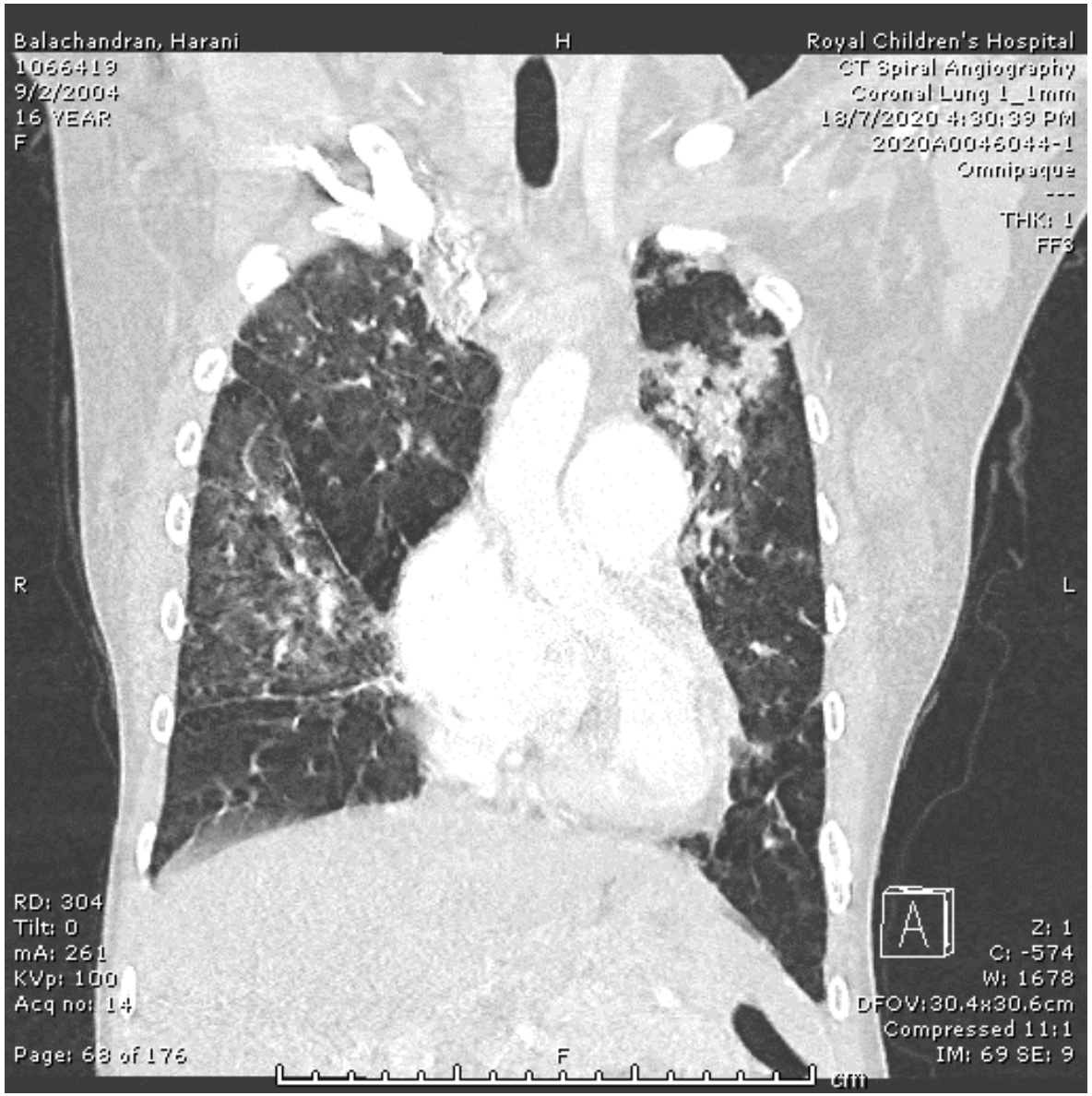




Pulmonary hypertension (apical 4 chamber)

- Right atrial enlargement
- RV enlargement
- Left shift of septum in systole
- TR

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Royal Children's Hospital
CT Spiral Angiography
Coronal Lung 1_1mm
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mA: 261
KVp: 100
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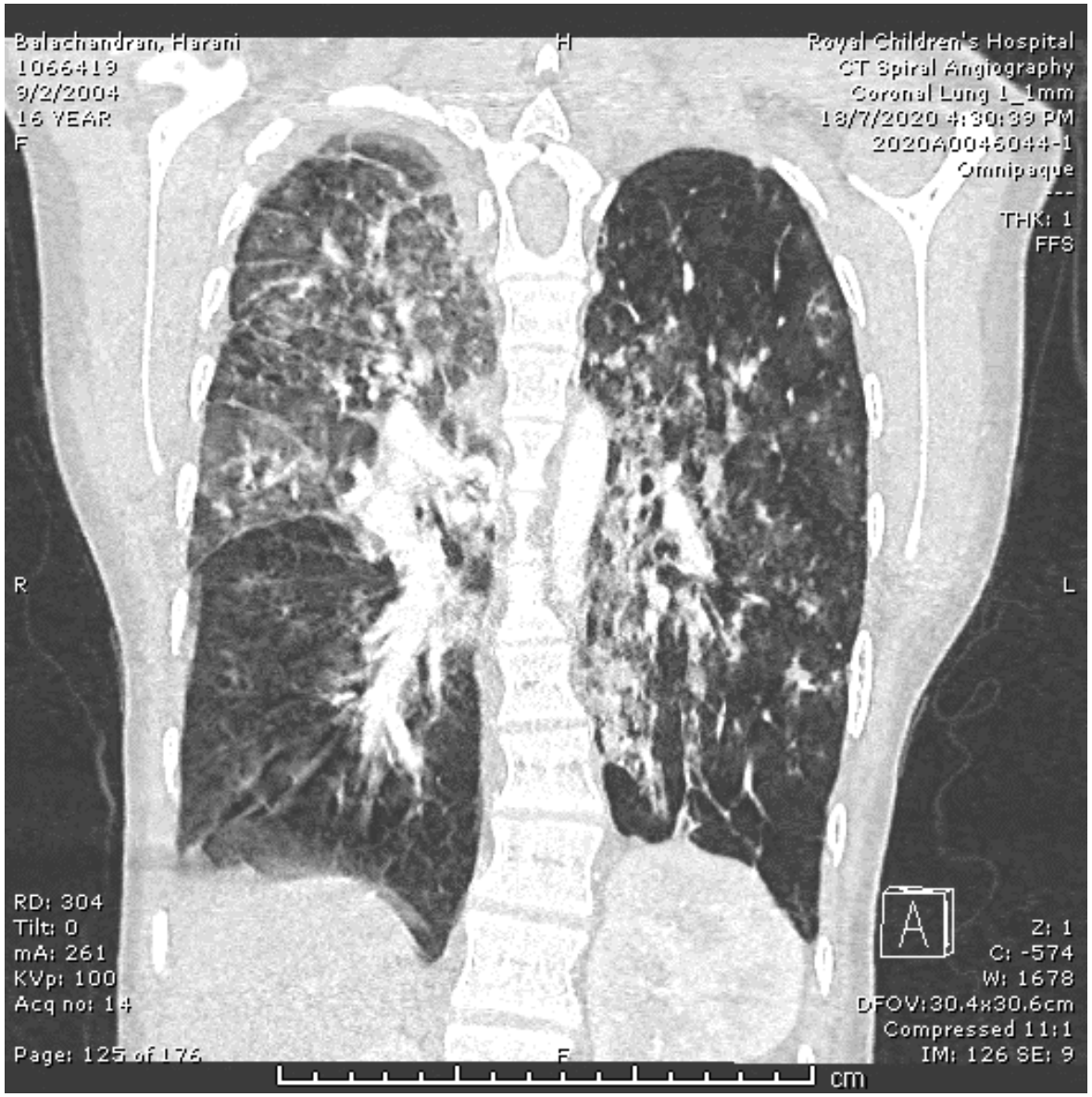
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Page: 125 of 176
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cm

Investigation and treatment of pulmonary hypertension

- Find the cause(s)
- Is it cardiac or respiratory?
- Is it acute (e.g. pneumonia related hypoxia) or chronic?
- What is reversible?
  - Hypoxia – oxygen is the best pulmonary vasodilator
  - Nutrition (vitamin C)
  - Treat cardiac failure
  - Sildenafil (phosphodiesterase inhibitor), pulmonary vasodilator.
    - *Do not* use if L→R shunt (e.g. VSD, ASD)