MMed and DCH Lectures

Weekly by Zoom

Prof Trevor Duke

MMed and DCH Lectures

Congenital heart disease I

July 20, 2020

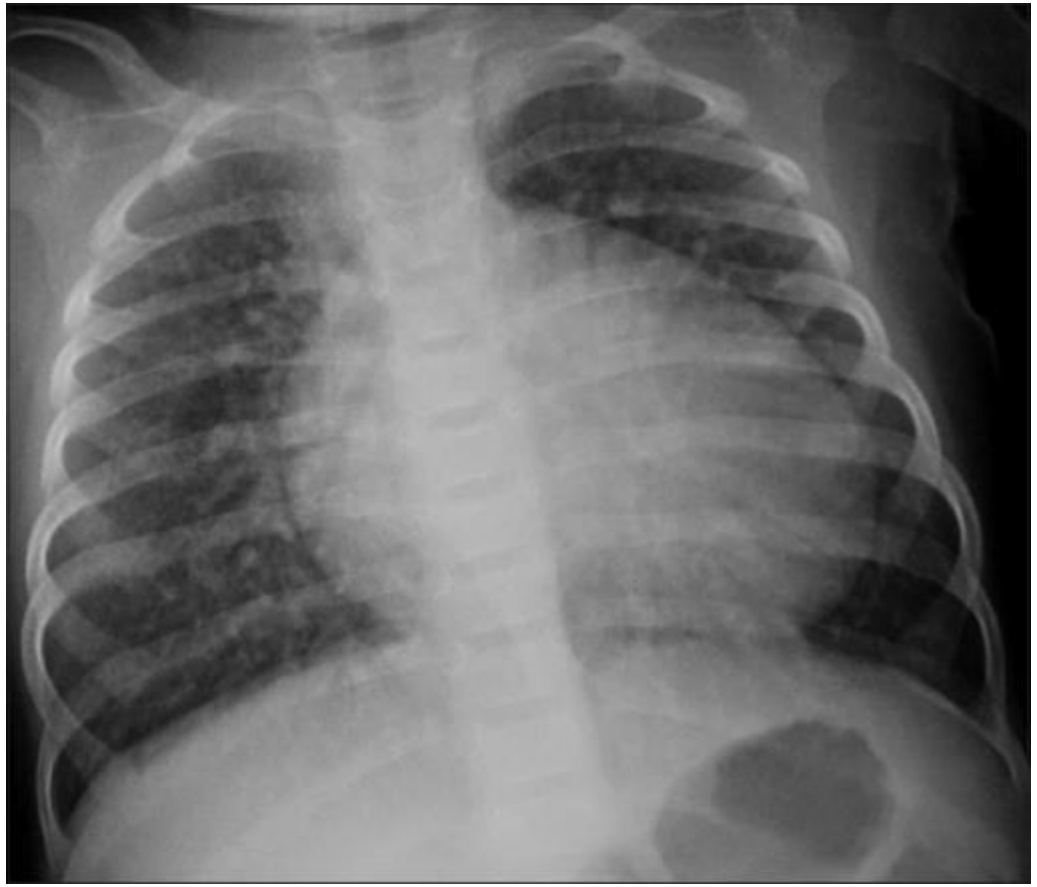
Prof Trevor Duke

Congenital heart disease: pathophysiology

- Acyanotic (75%)
  - Left to right shunts (VSD, PDA, ASD, AVSD)
  - Left ventricular outflow tract obstruction
- Cyanotic
  - Mixing lesions
  - Obstructed right ventricular outflow tract
  - Pulmonary vein anomalies

Case

- 3 ½ month old boy presents with failure to thrive and recurrent wheezing, sweating and fast breathing on feeding
- Chest x-ray
- Pulses hard to feel
- Chest auscultation: wheeze and crackles
- Hepatomegaly
- Differential diagnosis…?

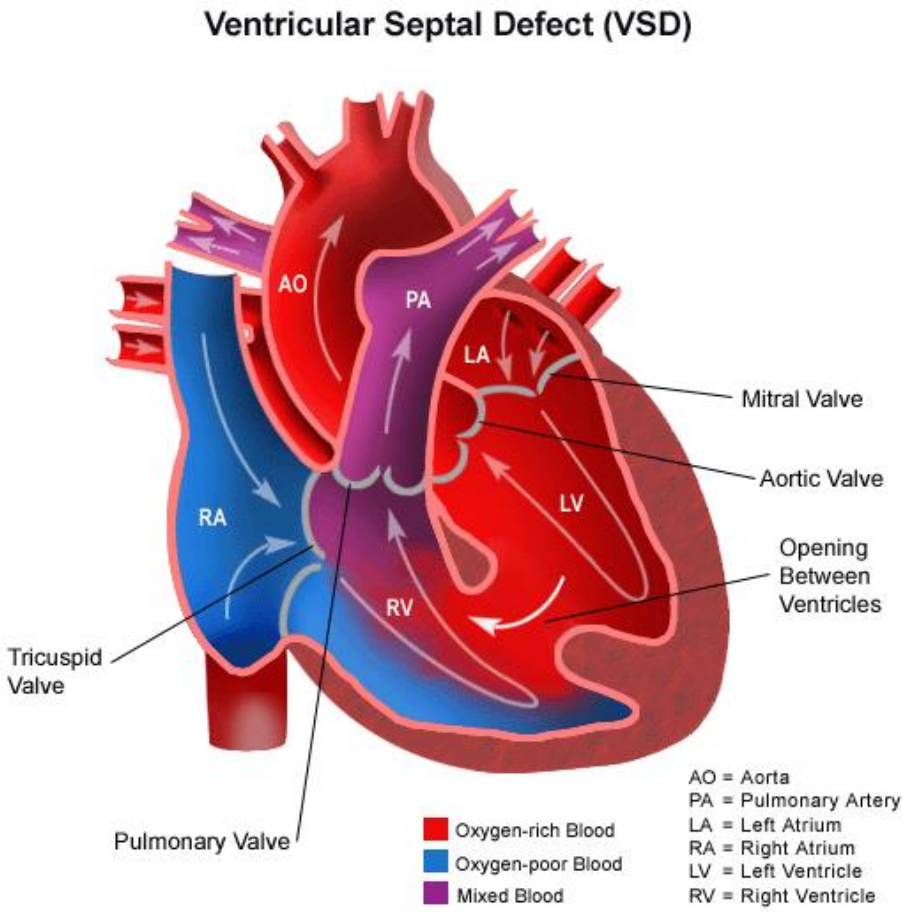


Differential diagnosis

- Bronchiolitis
  - Wheeze, crackles (course), ptosis of liver,
  - No murmur, no cardiomegaly, normal ECG
- Ventricular septal defect
  - Wheeze, crackles (fine), hepatomegaly
  - PSM LLSE, cardiomegaly, LVH on ECG
- PDA
  - Wheeze, crackles
  - Bounding pulses, wide pulse pressure
  - Continuous murmur

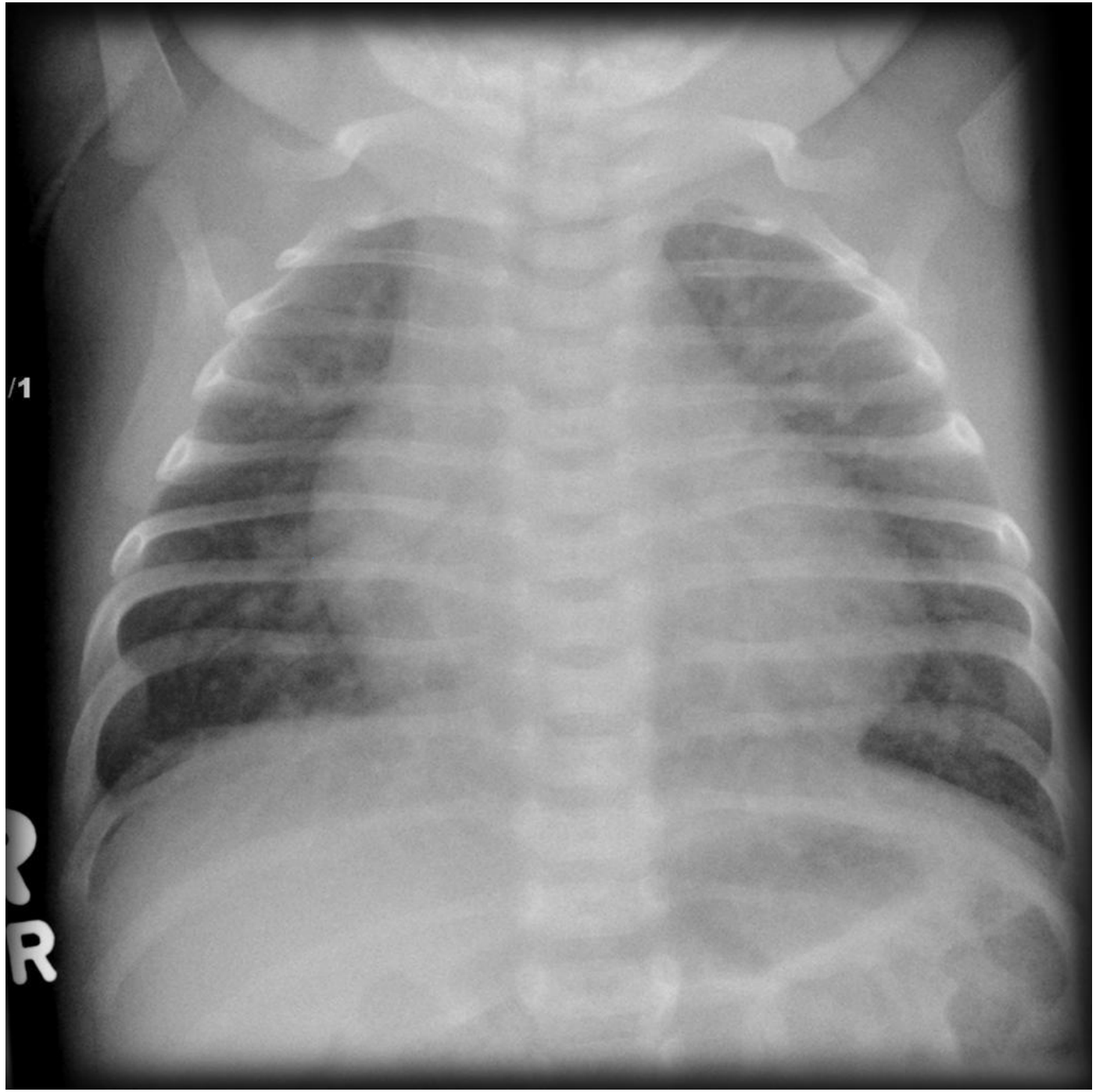
Ventricular septal defect

- 30% of all CHD
- Present 2-6 months of age
- Small: incidental murmur (loud)
- Large: congestive heart failure: failure to thrive, tachypnea and sweating on feeding
- Tachycardia, tachypnea
- Fine crackles on auscultation, hepatomegaly
- Cardiac auscultation
  - Parasternal heave (large LV), displaced apex
  - Pan-systolic murmur best heard at LLSE, radiates inferior to xiphoid
  - Loudness of murmur not indicative of defect size
  - Apical diastolic murmur (↑flow through MV)



VSD types

- Very small single mid-muscular (most common)
- Multiple
- Large
- Perimermbranous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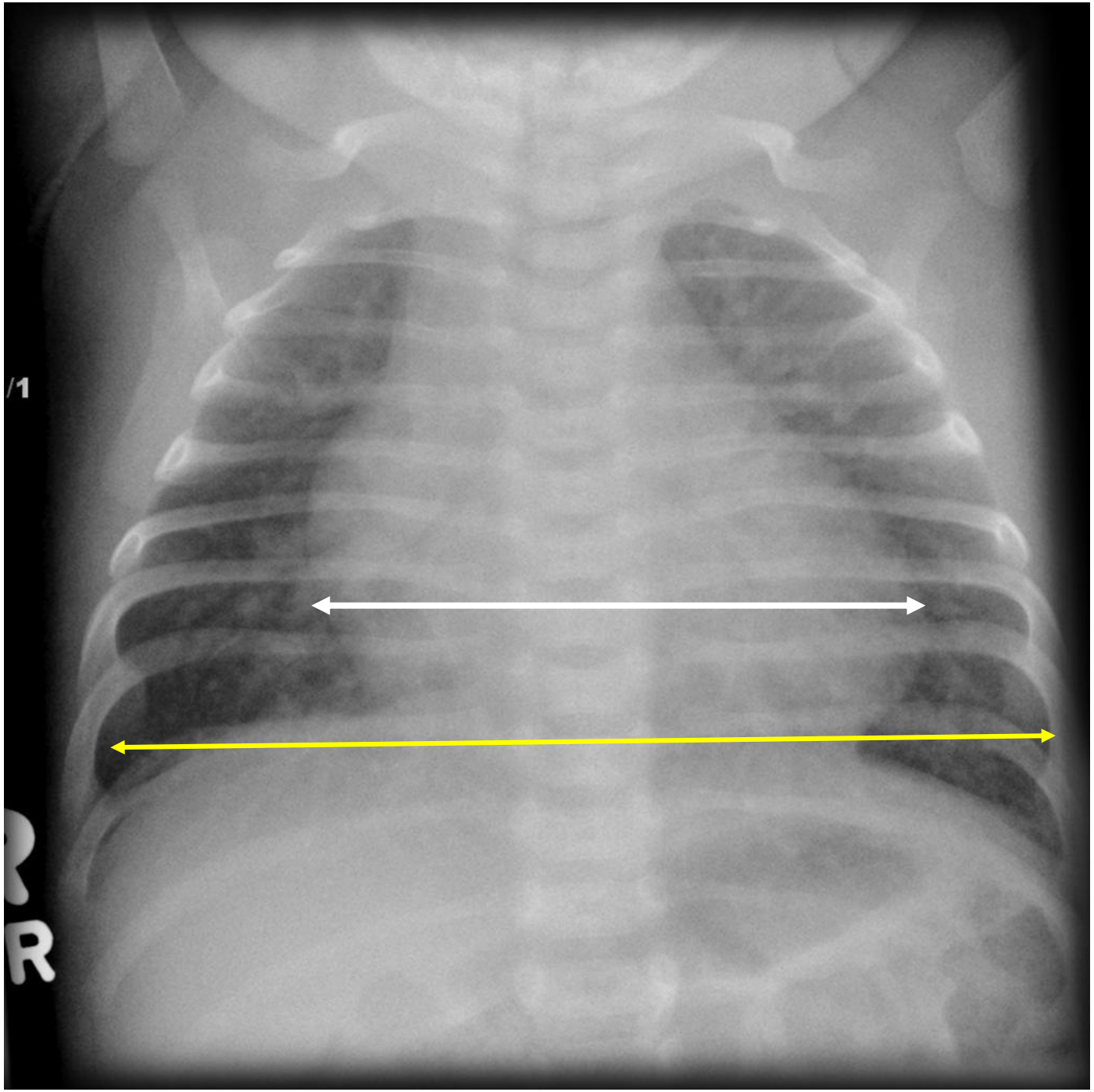
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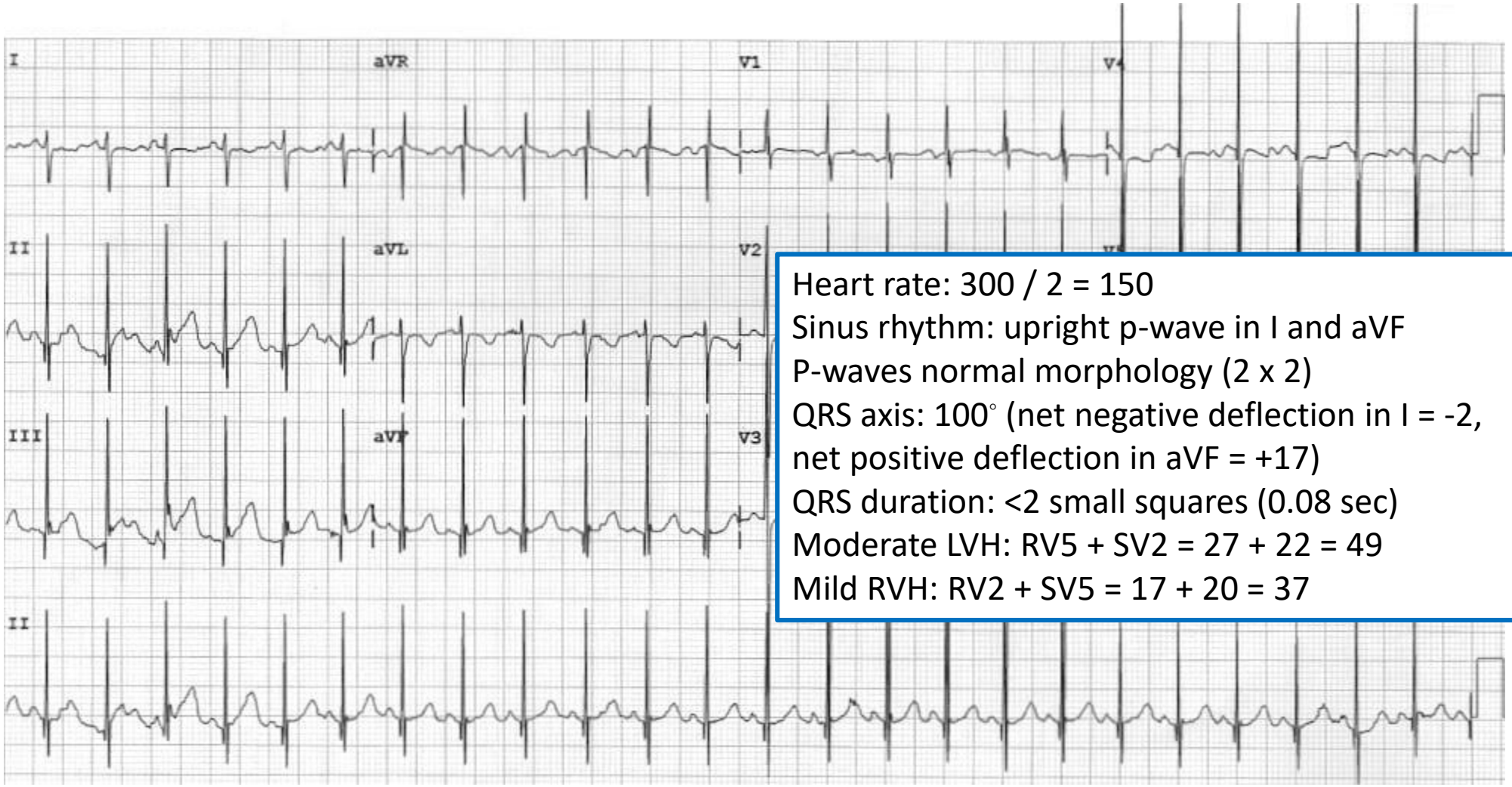
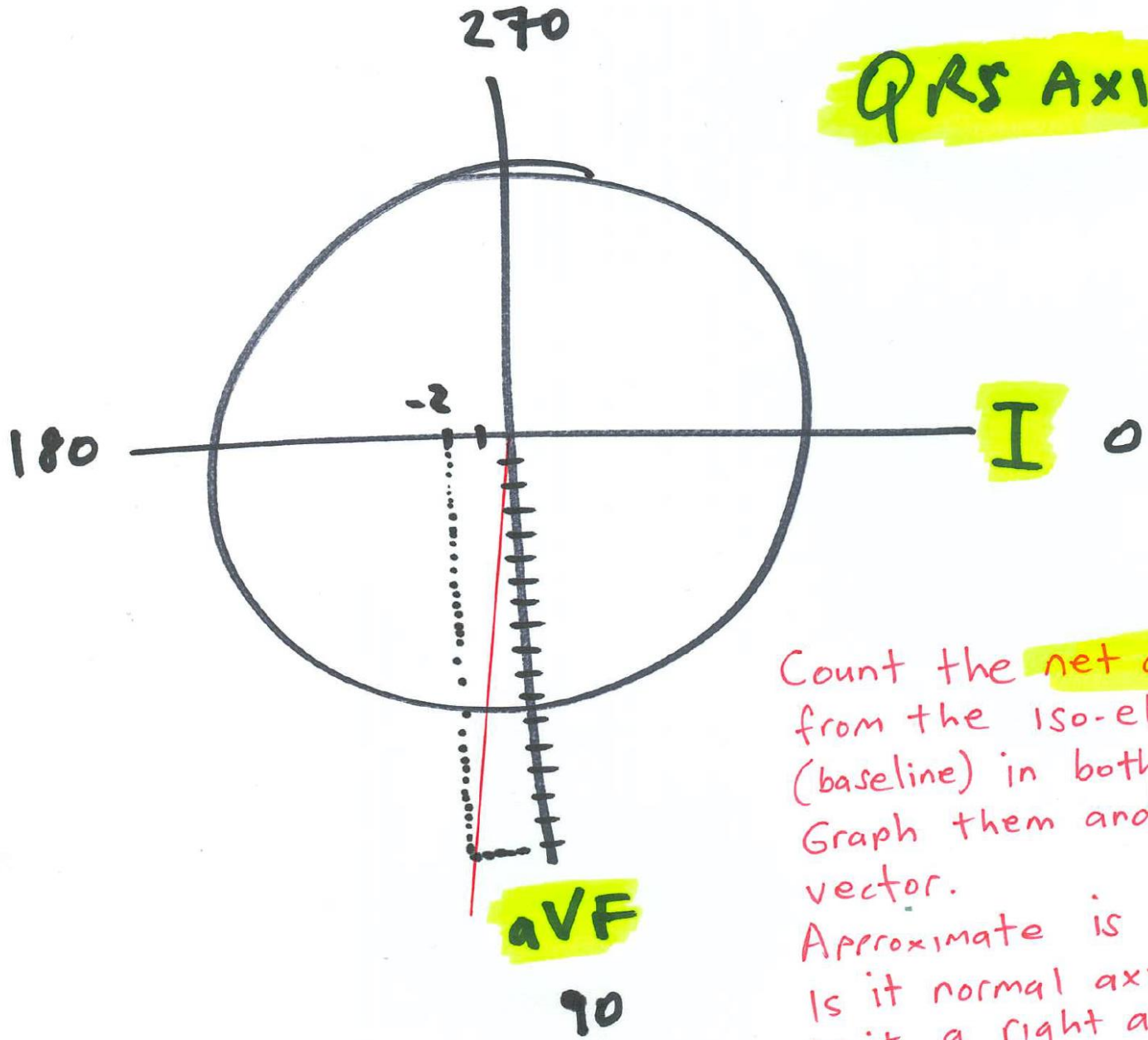
ECG of large VSD: biventricular hypertrophy



Heart rate: 300 / 2 = 150
Sinus rhythm: upright p-wave in I and aVF
P-waves normal morphology (2 x 2)
QRS axis: 100° (net negative deflection in I = -2, net positive deflection in aVF = +17)
QRS duration: <2 small squares (0.08 sec)
Moderate LVH: RV5 + SV2 = 27 + 22 = 49
Mild RVH: RV2 + SV5 = 17 + 20 = 37

QRS Axis



Count the net deflection from the iso-electric line (baseline) in both I and aVF. Graph them and add the vector.

Approximate is OK.

Is it normal axis ?

Is it a right axis deflection ?

Is it a left axis deflection ?

Why do children with VSD go into heart failure?

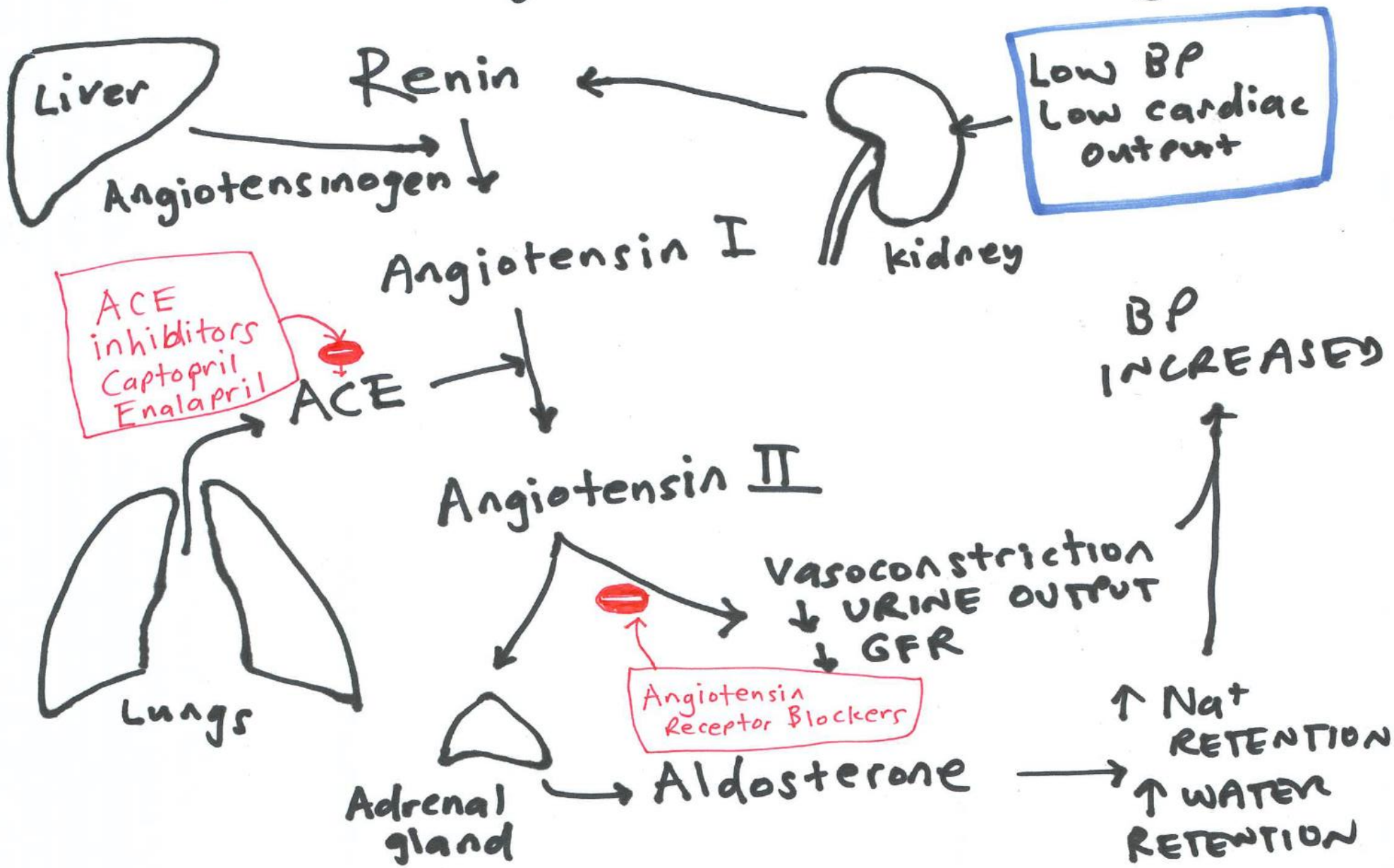
- Fall in pulmonary vascular resistance→ increased pulmonary blood flow
- What reduces PVR?
  - Anaemia →→ can reduce heart failure by increasing Hb
  - Oxygen →→ giving oxygen can worsen L→R shunt and worsen heart failure

- What increases PVR
  - Polycythemia
  - Hypoxia
  - Lung disease, e.g. pneumonia
  - Stress

Heart failure treatment

- Diuretics
  - ↓ salt and water retention
  - ↓ pre-load on the heart
  - ↓ pulmonary congestion
- Afterload reduction
  - Enalapril, captopril – ACE inhibitors

Renin - Angiotensin - Aldosterone

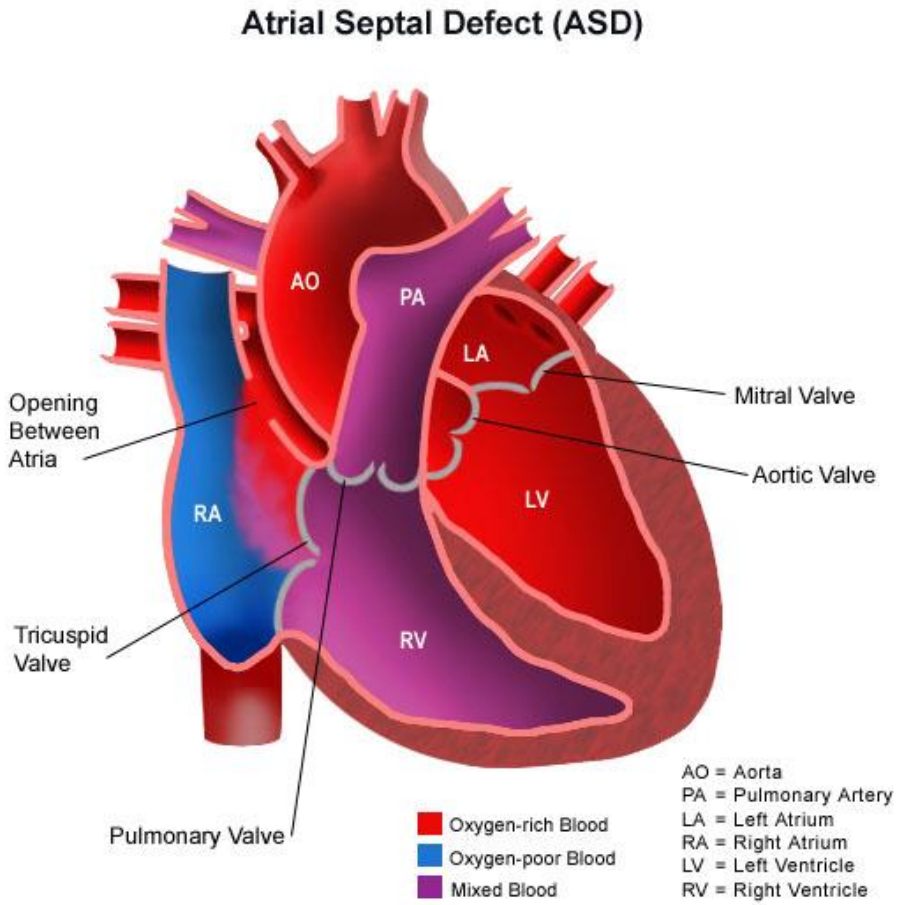


VSD: complications and indications for surgery

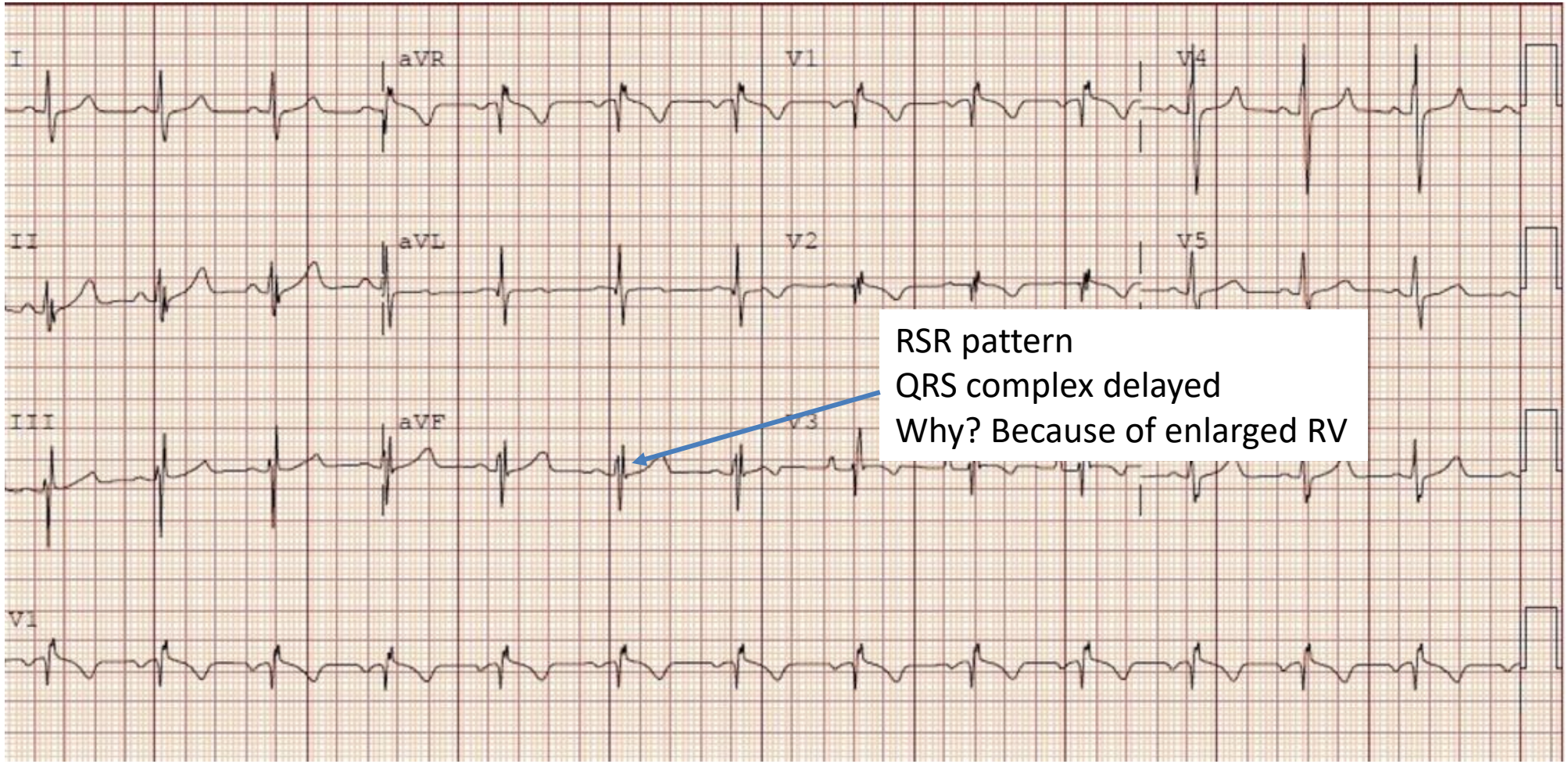
- Heart failure not controlled by medical management
- Failure to thrive, despite optimal medical management and nutrition
- Pulmonary hypertension (high QP: QS)
- Aortic valve prolapse into the VSD → aortic regurgitation
- Bacterial endocarditis

ASD

- Primum ASD – low in the interarial septum, often involving the AV valves
- Secundum ASD – mid-atrial septum (fossa ovale)
- Rarely cause problems in early childhood.
- May develop PHT if large, or arrhythmias
- Volume (flow) load on the right side of the heart, so dilated RV, RV heave, murmur of increased flow across the PV (ejection systolic)
- Delayed closure of the PV → wide and fixed splitting of P2

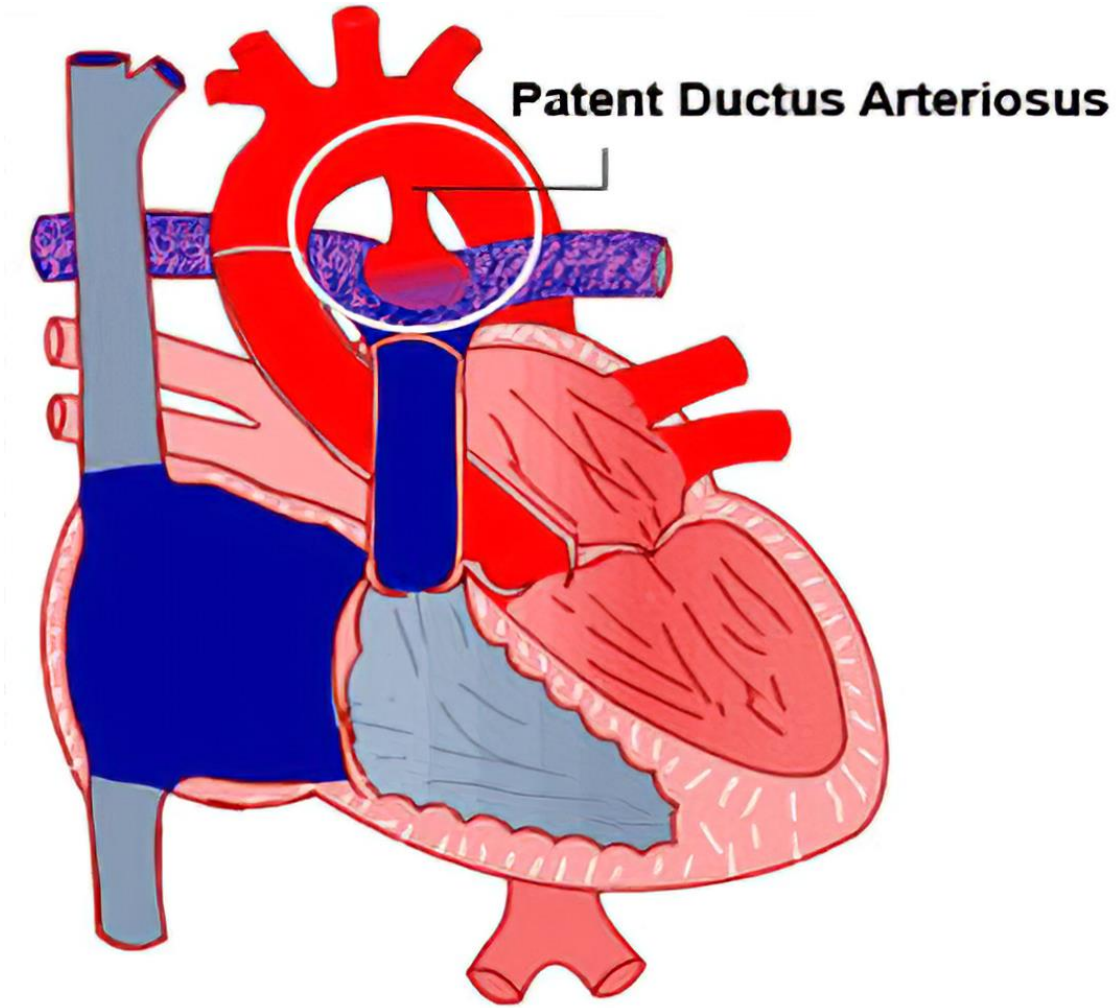


ECG in ASD – sometimes right BBB



Patent ductus arteriosus

- Prematurity or congenital anomaly
- Clinical features:
  - Poor feeding, tachypnea, poor weight gain
  - Pulses: tachycardia, bounding, wide pulse pressure (e.g. 55/18 in a preterm infant), hepatomegaly
  - “Continuous murmur” – through systole into diastole

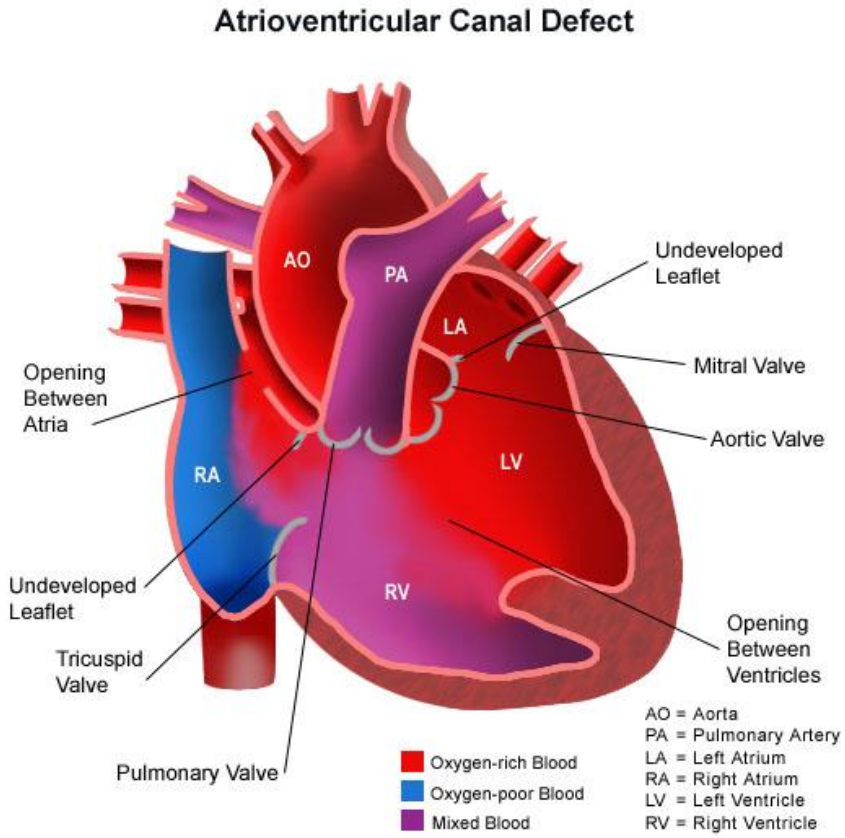


Patent ductus arteriosus: treatment

- Sometimes close spontaneously
- Diuretics, sodium and fluid restriction
- Prostaglandin inhibitor - Indomethacin (0.1 mg/kg Q8), or ibuprofen
  - Risks: NEC, renal impairment
- Correct anaemia (reduce trans-ductal gradient)
- Surgical closure if failure of medical treatment
  - device closure
  - ligation via left lateral thoracotomy

Complete AV canal / AVSD

- Atrial septal defect, ventricular septal defect, improperly formed mitral and/or tricuspid valves leading to regurgitation
- Commonest CH lesion in Down syndrome
- Severity and onset of symptoms depend on size of VSD and degree of AV valve regurgitation
- Early severe heart failure, cyanosis because of mixing of blood at ventricular level
- Pulmonary hypertension early and severe
- X-ray: cardiomegaly (like VSD)
- ECG: RBBB + Left axis deviation



Left ventricular outflow tract obstruction

- Aortic stenosis
  - Valvular (thickened bicuspid valve)
  - Subvalvular (fibrous stricture or muscular obstruction)
  - Supravalvular (Williams syndrome)
- Coarctation of aorta
  - Isolated lesion
  - Sometimes associated AS (e.g., bicuspid), VSD, MV abnormalities
- Hypoplastic left heart syndrome

Presentation of LVOTO

- Cardiac failure in neonatal period, often abruptly when the duct closes
- The neonate in shock:
  - Lethargy, cannot feed, pale, mottled, cyanosed
  - Tachypnea, cold limbs,
  - Poor or absent femoral and lower limb pulses
  - Severe lactic acidosis
  - Oliguria

Differential diagnosis of the neonate in shock

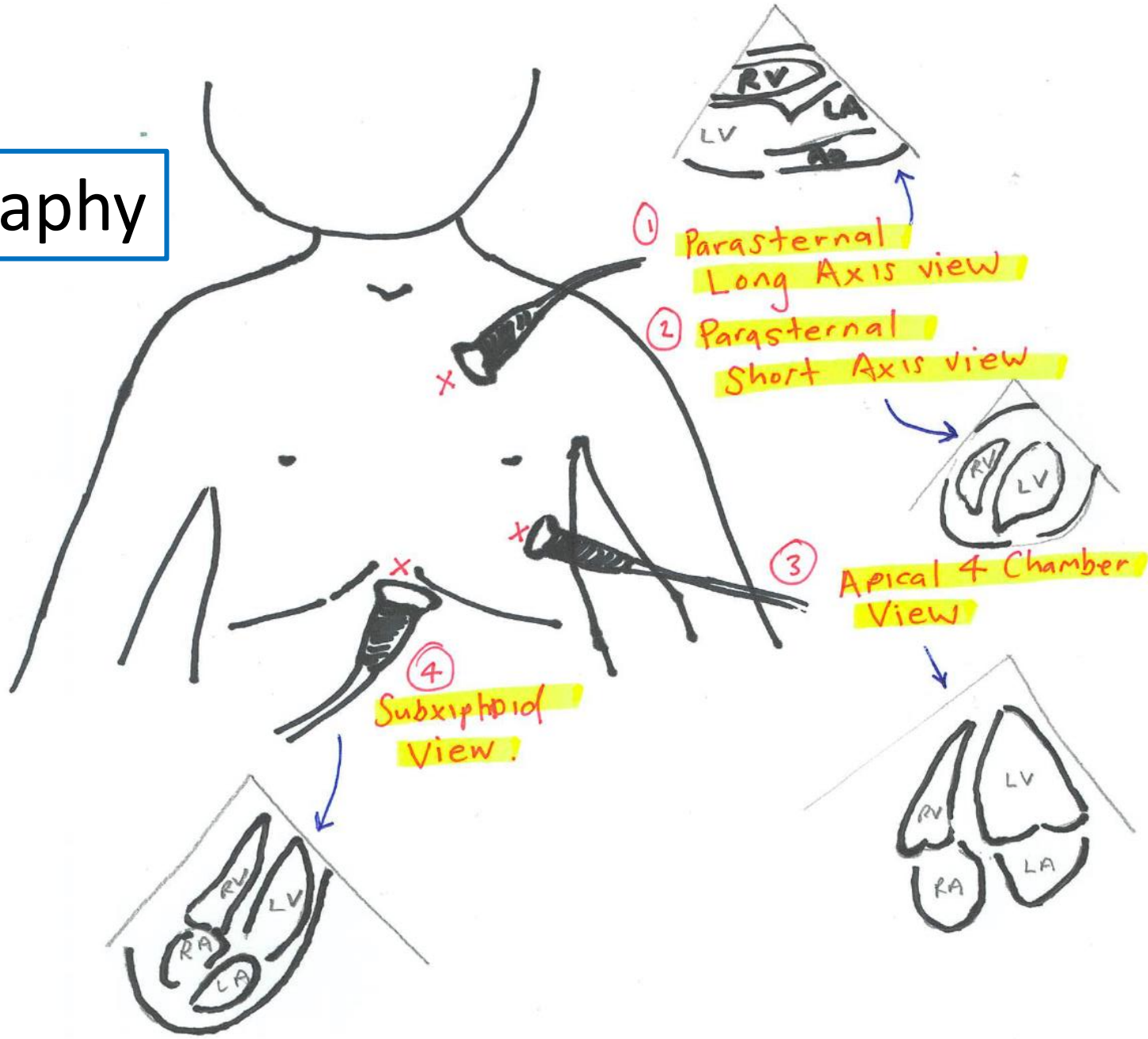
The newborn who is *well at birth*, discharged home, and returns in shock

1. Congenital heart disease (especially LVOTO such as a coarctation)
2. Sepsis
3. Inborn error of metabolism (e.g. hyperammonaemia)
4. Asphyxia (SIDS, other)

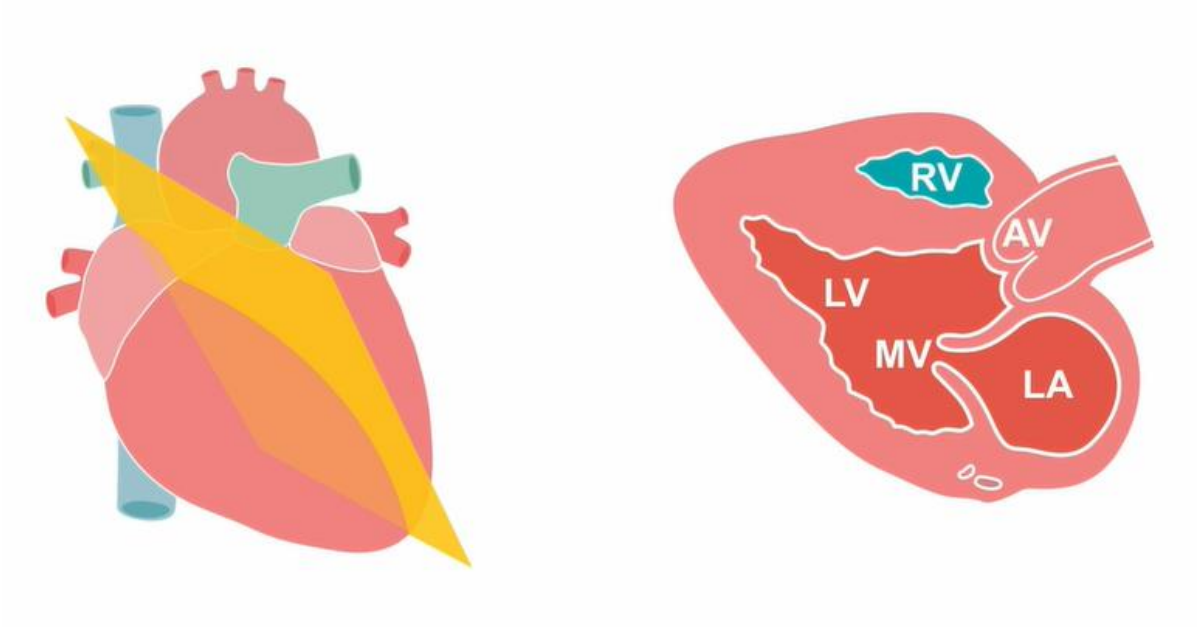
The shocked neonate: the golden hour

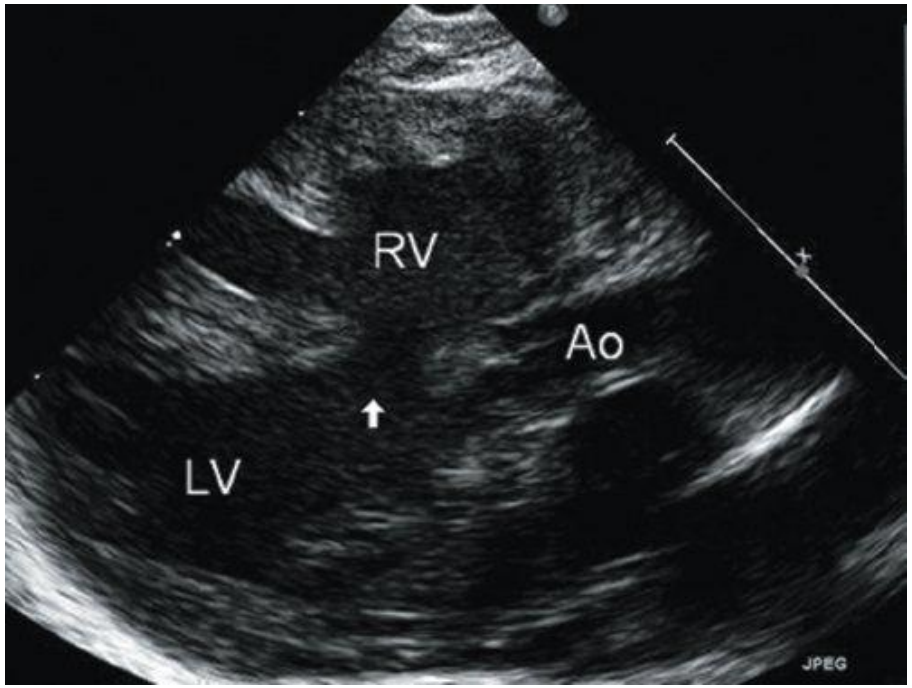
- Emergency treatment
  - Oxygen
  - Establish IV access (peripheral, umbilical)
  - Fluid resuscitation (10-20ml/kg IV)
  - Antibiotics – ceftriaxone, amikacin, cloxacillin
  - Adrenaline 0.05-0.1 mcg/kg/min
- Examination
  - Cardiac murmur, pulses
  - Signs of sepsis source
- Investigations
  - Chest x-ray
  - Blood culture
  - Blood gas, glucose, electrolytes
  - Echo

Echocardiography



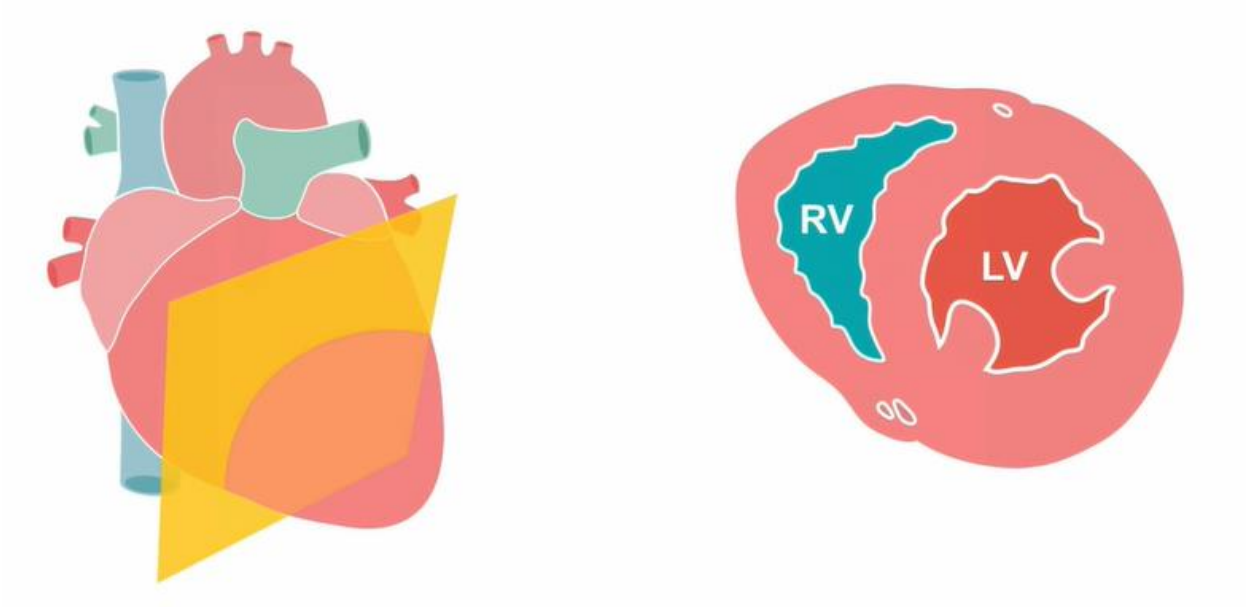
1. Parasternal long axis view

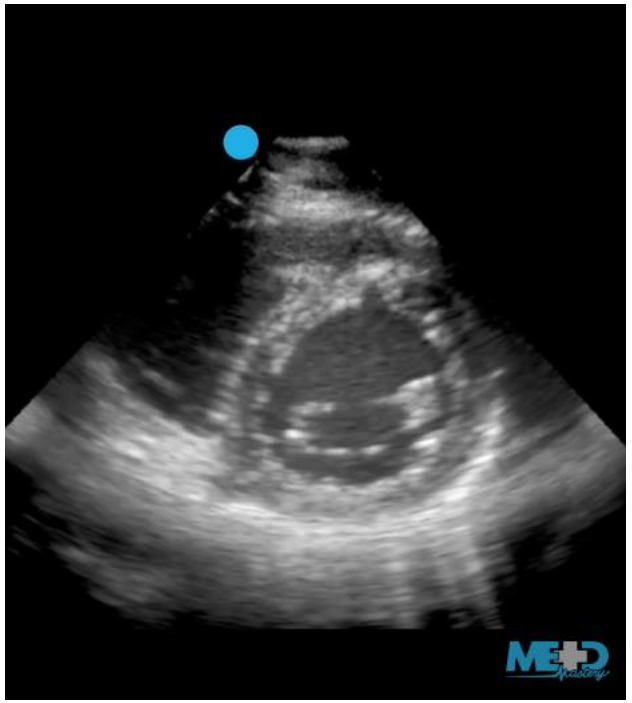




Probe placed to left of the lower third of the sternum 3rd / 4th ICS.
Probe marker pointing towards the right shoulder.

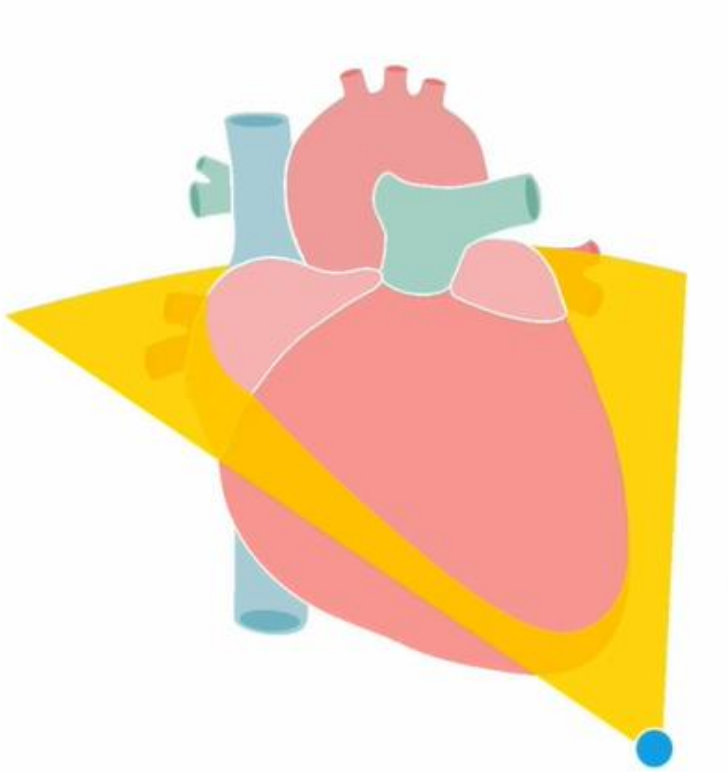
2. Parasternal short axis view

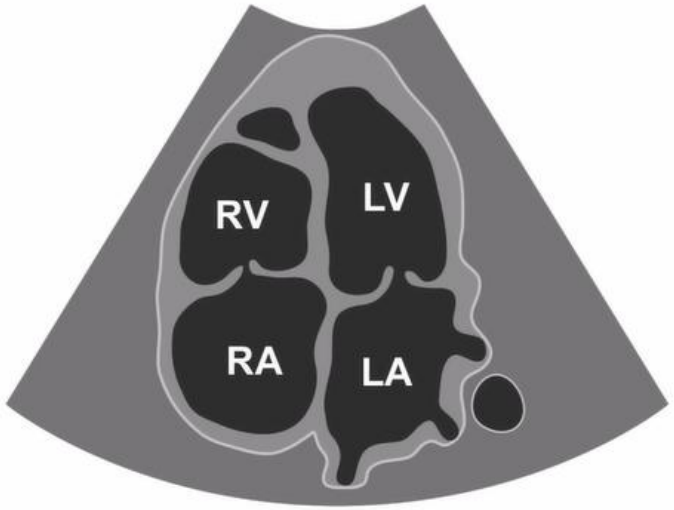


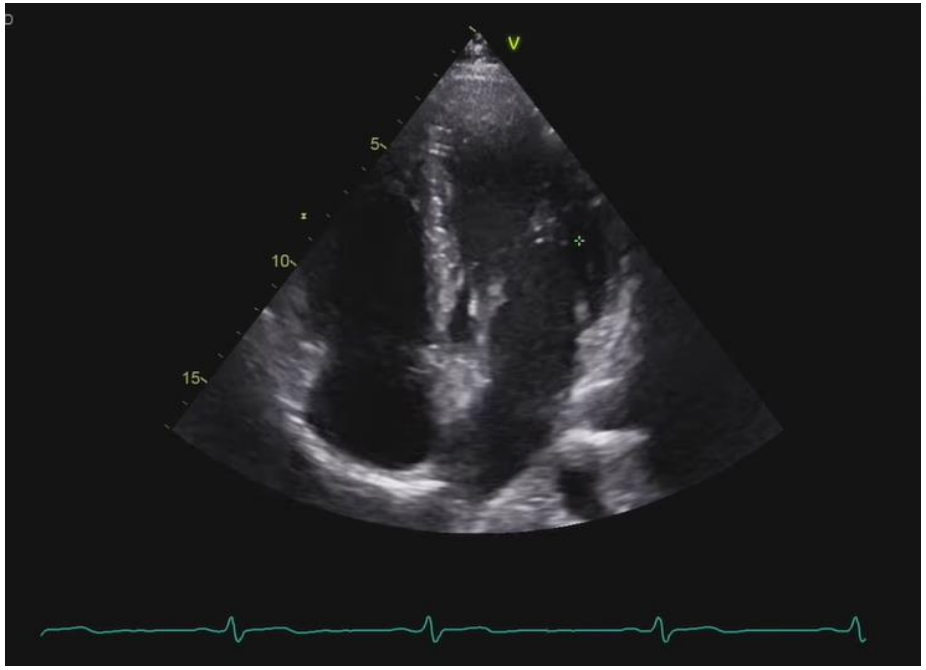


Probe placed to left of the lower third of the sternum 3rd / 4th ICS.
Probe marker pointing towards the left shoulder.

3. Apical 4-chamber view

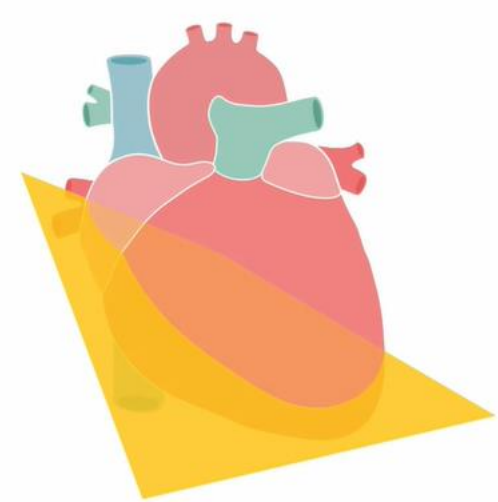


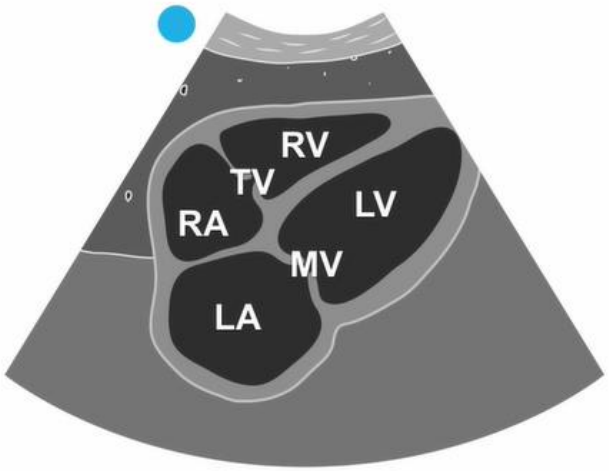


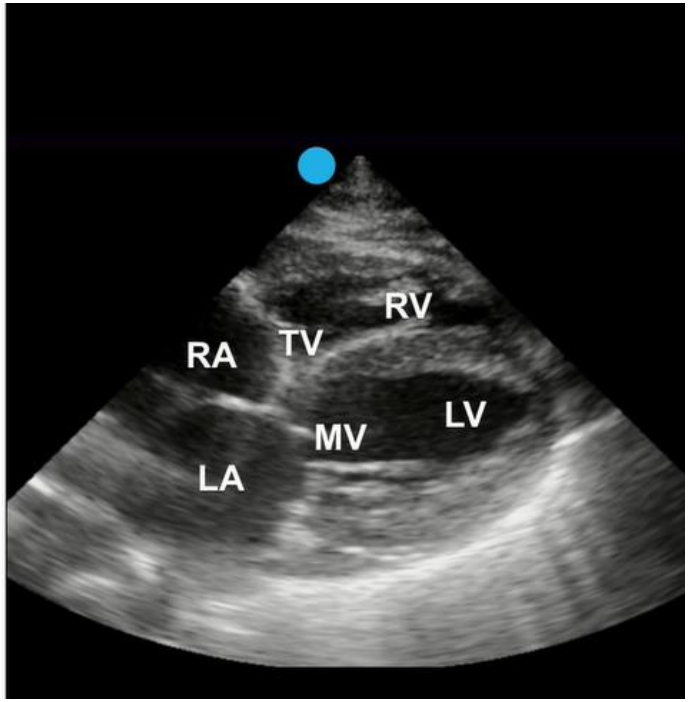


Place probe at apex, at an angle pointing towards the right shoulder.
The positioning marker on the probe faces the left shoulder

4. Sub-xiphoid view







Place probe under xiphoid process
Positioning indicator to right.