

# **MMed and DCH Lectures**

## **Congenital heart disease I**

July 5, 2021

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# Heart failure

## General

- Tachycardia >160
- Tachypnoea

## Congestive

- Right
  - Hepatomegaly
  - Oedema
  - Raised jugular veins
- Left
  - Tachypnoea, grunting
  - Fine crackles, signs of effusion

## Low output

- Hypotension
- Weak pulses
- Narrow pulse pressure
- Cold peripheries
- Skin mottling

# Causes of heart failure

- Structural congenital heart disease
  - Acyanotic
  - Cyanotic
- Valves
  - Rheumatic
  - Congenital
- Muscle
  - Myocarditis
  - Cardiomyopathy
- Cardiac conduction pathways
  - Arrhythmias
- Pericardial disease
  - Pericardial effusion / tamponade
  - Constrictive pericarditis
- Heart failure secondary to lung disease
  - Cor pulmonale
  - Pulmonary hypertension
- High-output cardiac failure
  - Severe anaemia
  - Beri beri
  - Thyrotoxicosis
  - Large A-V malformations

# 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...?

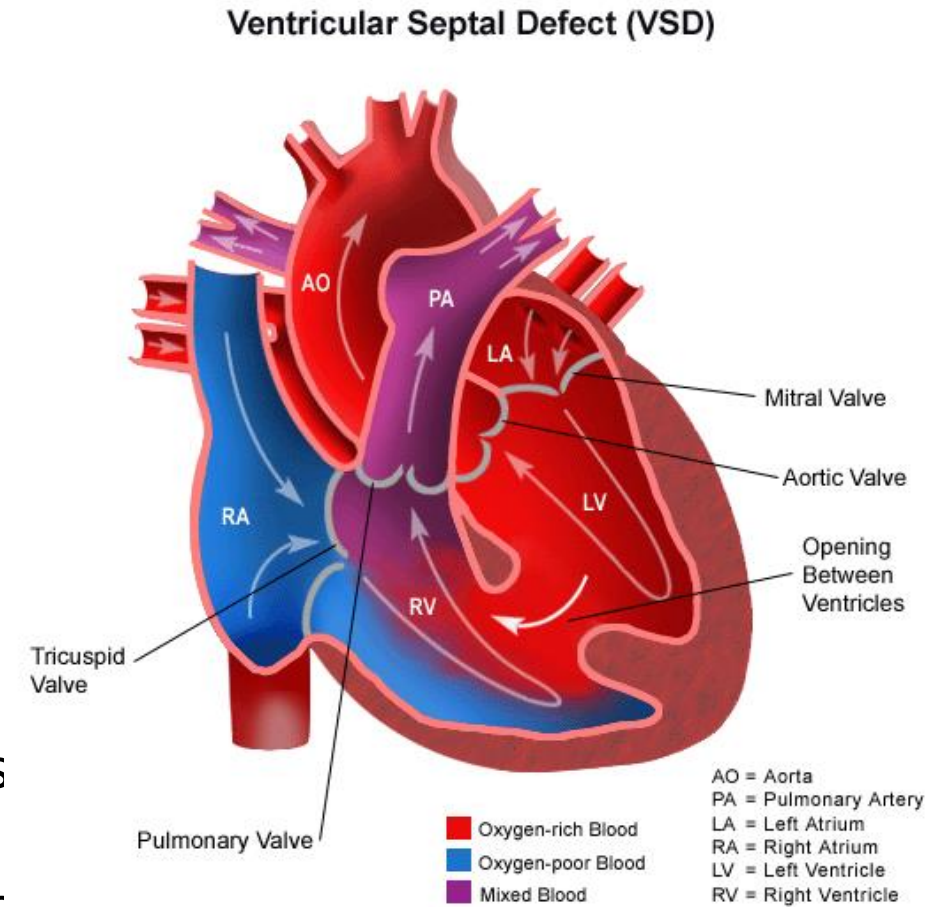


# Differential diagnosis

- 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

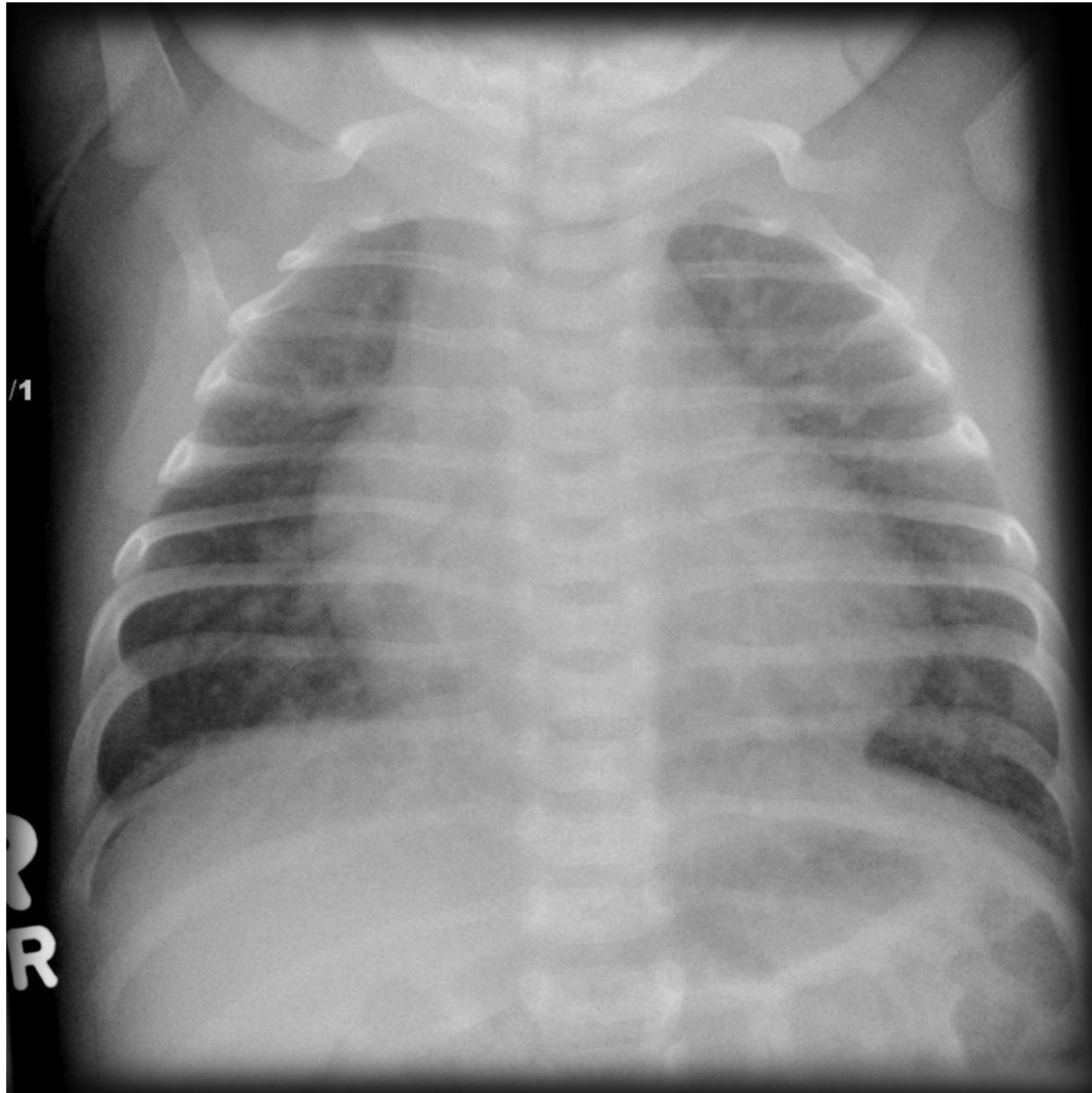
- 30% of all CHD
- Present 2-6 months of age
- Small: incidental murmur (loud)
- Large: congestive heart failure: failure to thrive, tachypnea and sweating on feeding
- Tachycardia, tachypnea
- Fine crackles on auscultation, hepatomegaly
- Cardiac auscultation
  - Parasternal heave (large LV), displaced 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

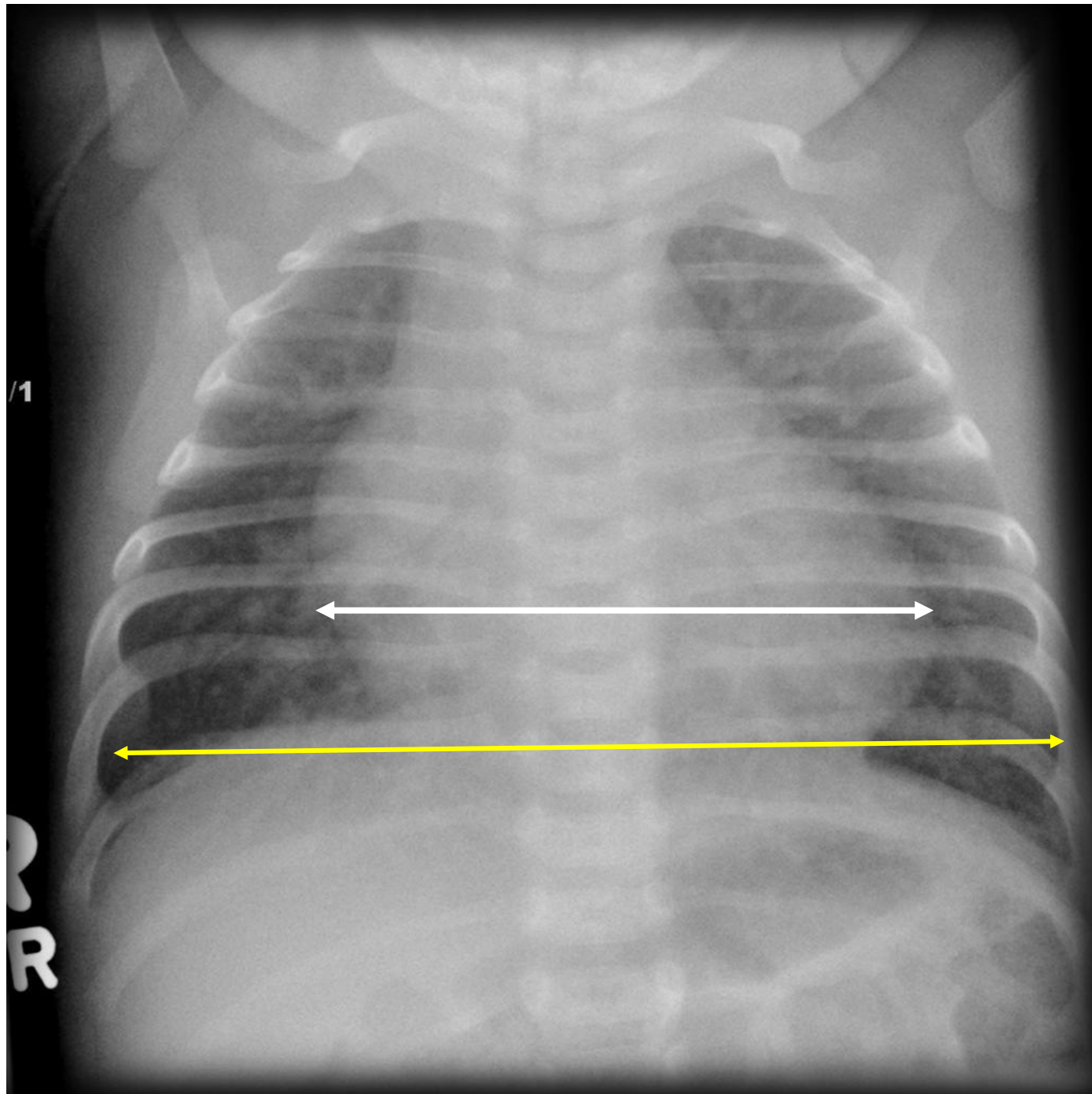
- Very small single mid-muscular (most common)
- Multiple
- Large
- Perimermbranous





/1

R R



CT ratio should be  $<0.6$  in children

The ECG and how it relates to electrical conduction and the cardiac cycle

Systole – contraction

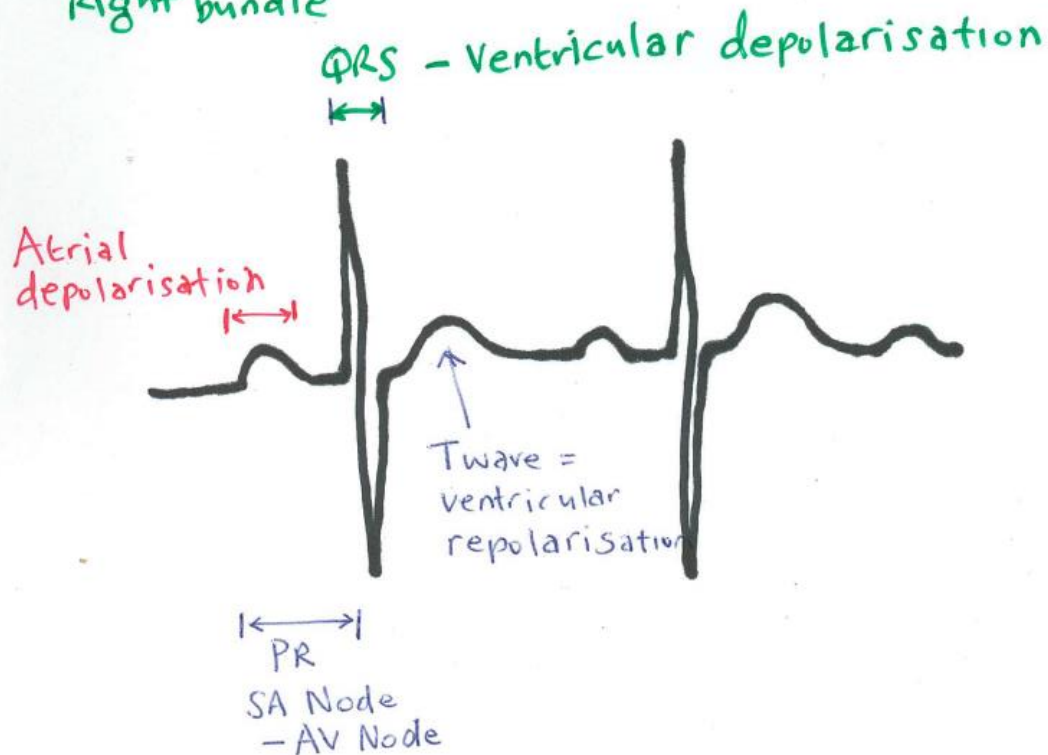
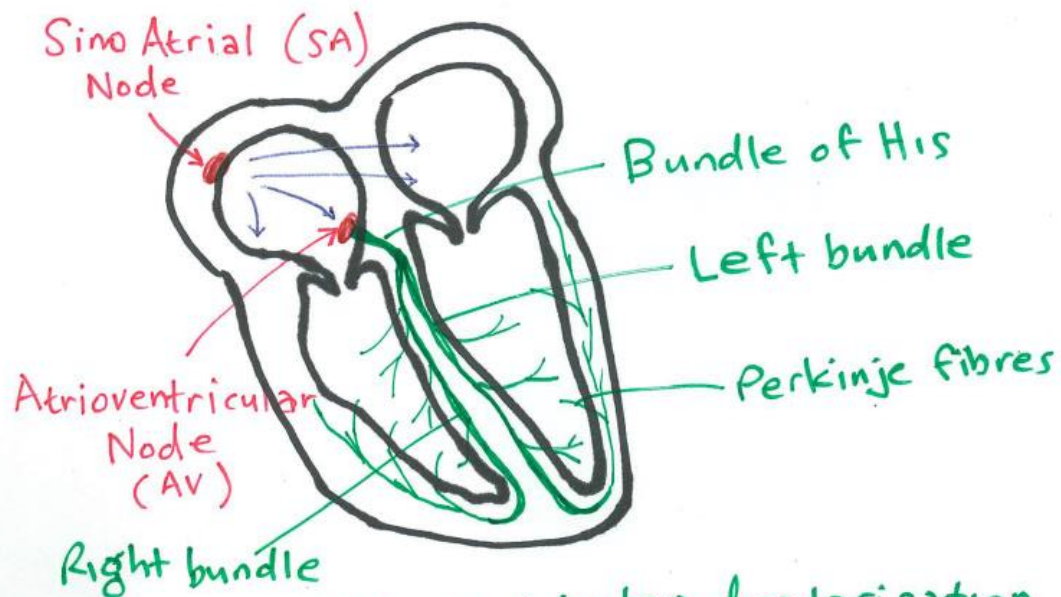
Diastole – relaxation

Atrial

Ventricular

Y-axis = Amplitude

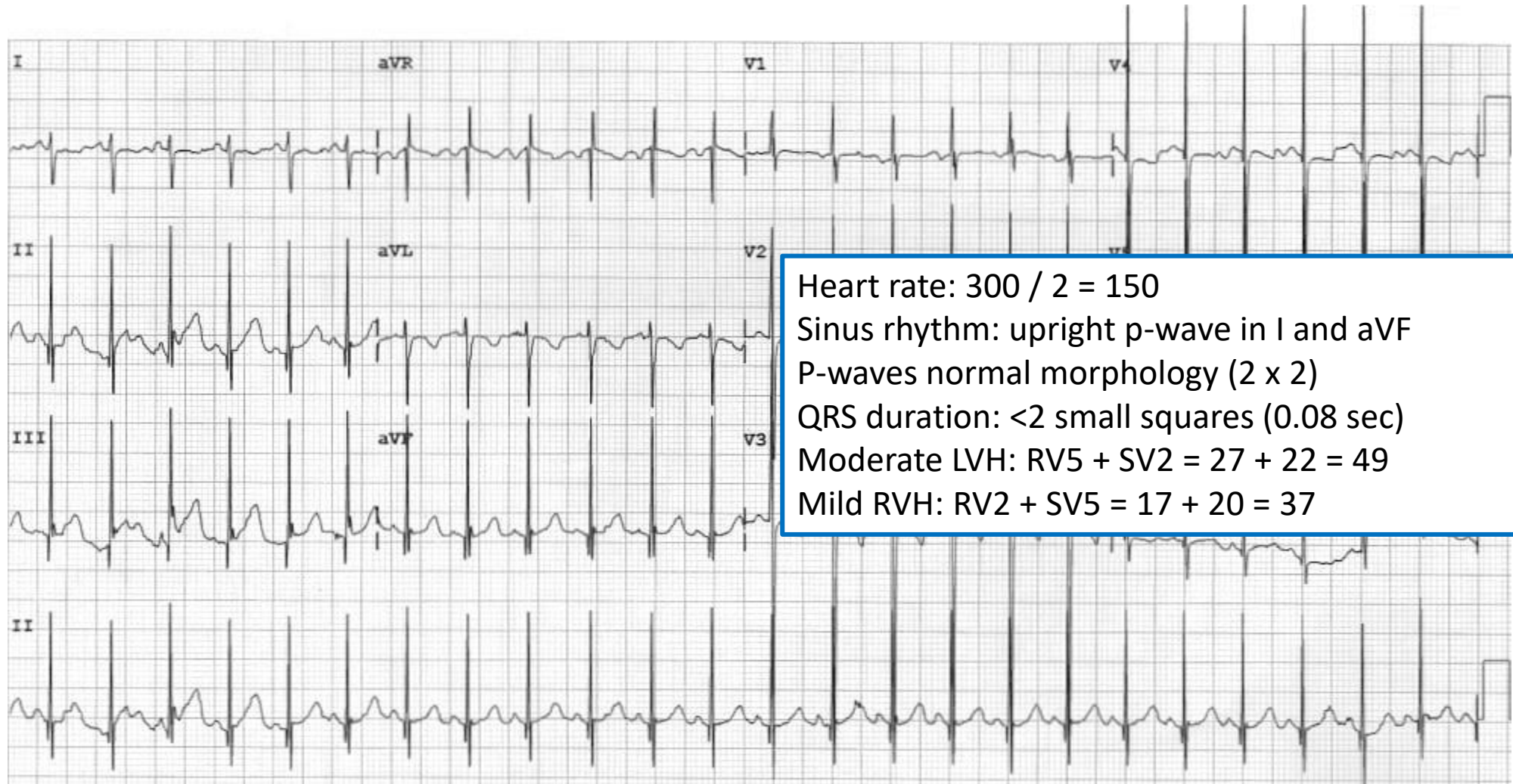
X-axis = Time

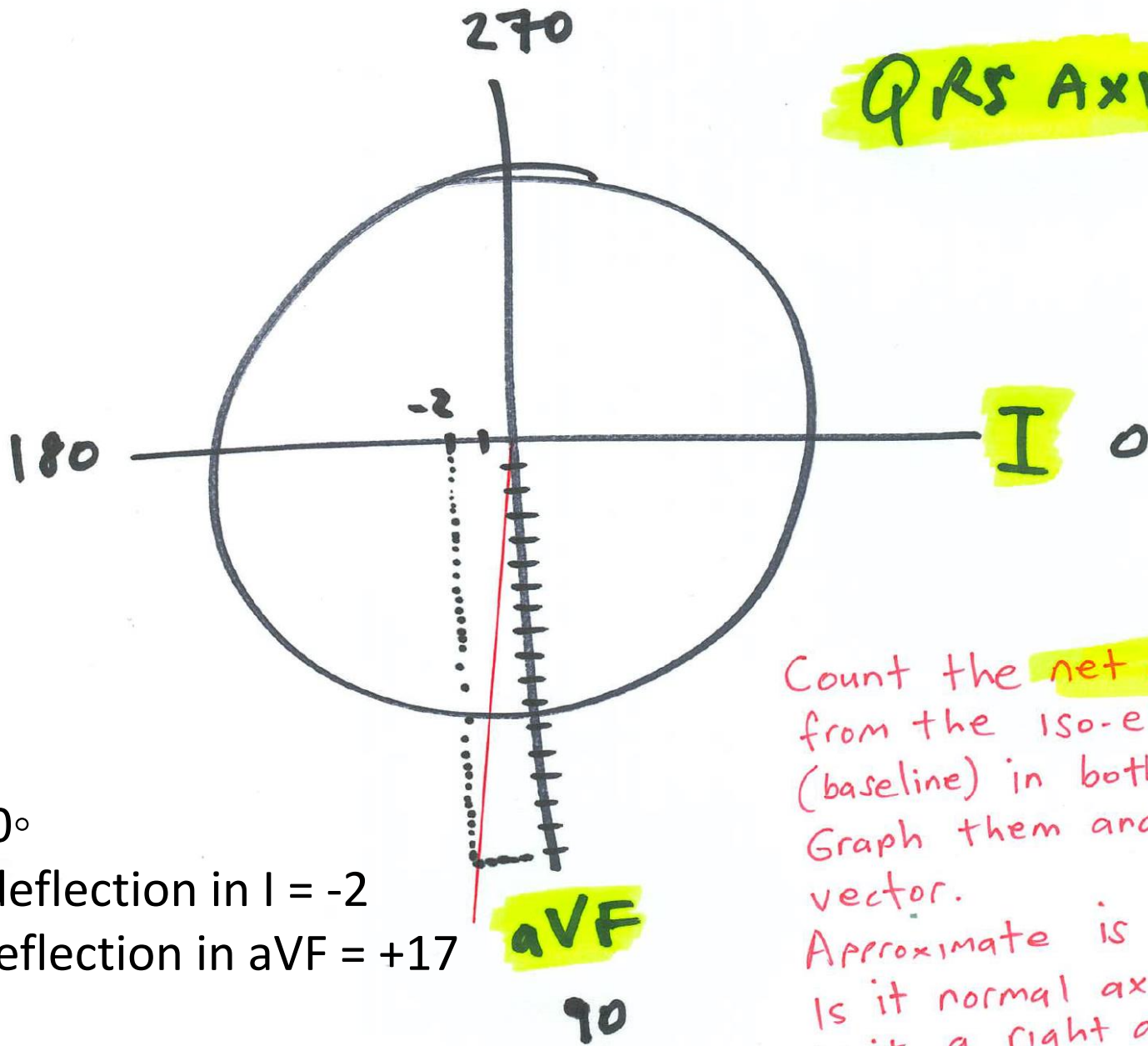


# How to read an ECG

1. **Heart rate:** 300 / large squares
2. **Rhythm:** sinus =
  - p before every QRS complex
  - upright p-wave in I and aVF
  - **PR interval** <0.16 (4 small squares)
3. **P-wave morphology:** 2 x 2mm – RAH=>2mm height, LAH >2mm width
4. **QRS duration:** <2 small squares (0.08 sec)
5. **QRS axis:** net deflection in I and aVF
6. **Ventricular hypertrophy**
  - LVH:  $RV5 + SV2 >35$
  - RVH:  $RV2 + SV5 >35$

# ECG of large VSD





QRS axis =  $100^\circ$

net negative deflection in I = -2

net positive deflection in aVF = +17

Count the net deflection from the 150-electric line (baseline) in both I and aVF. Graph them and add the vector.

Approximate is OK.

Is it normal axis?

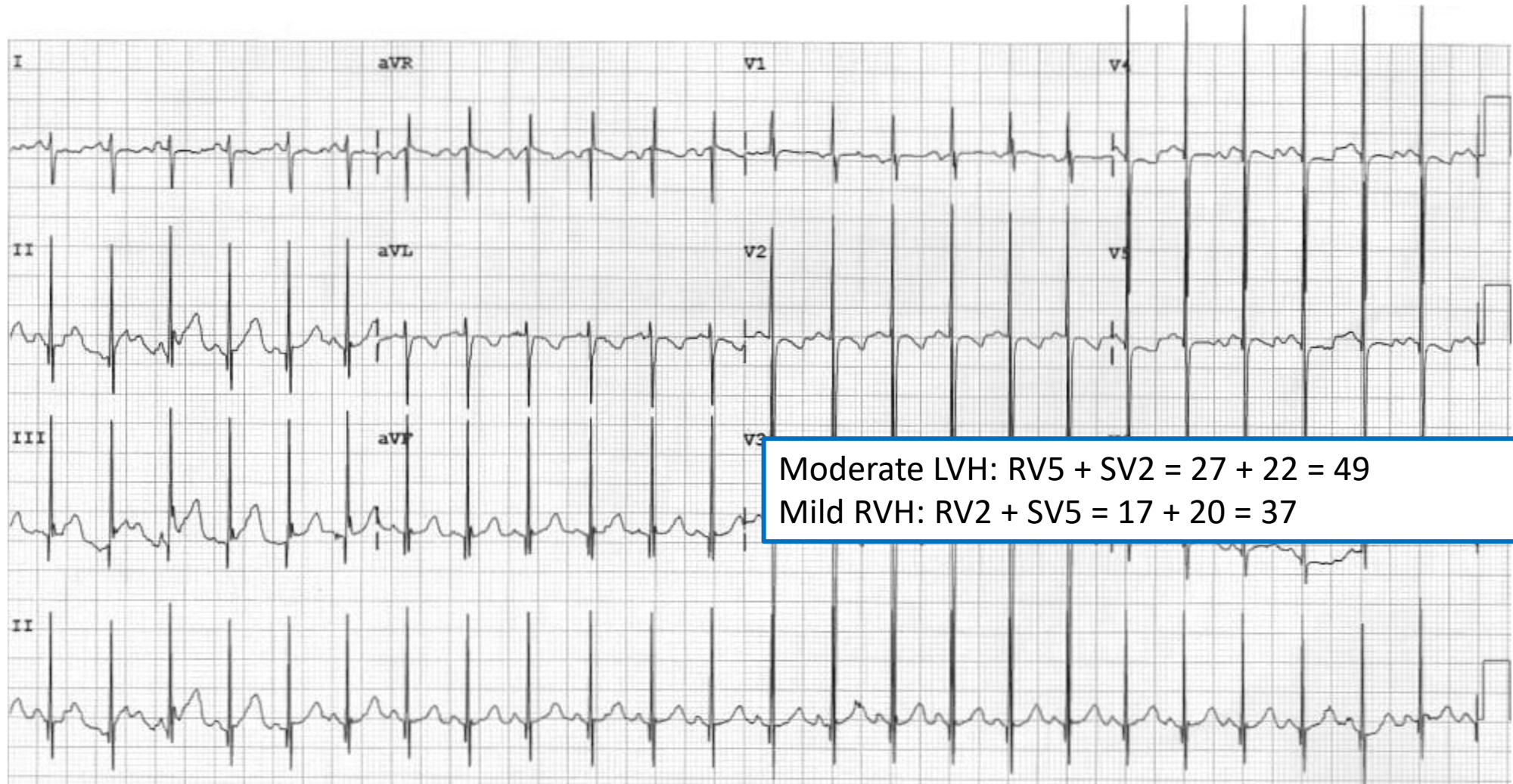
Is it a right axis deflection?

Is it a left axis deflection?

# QRS axis age changes

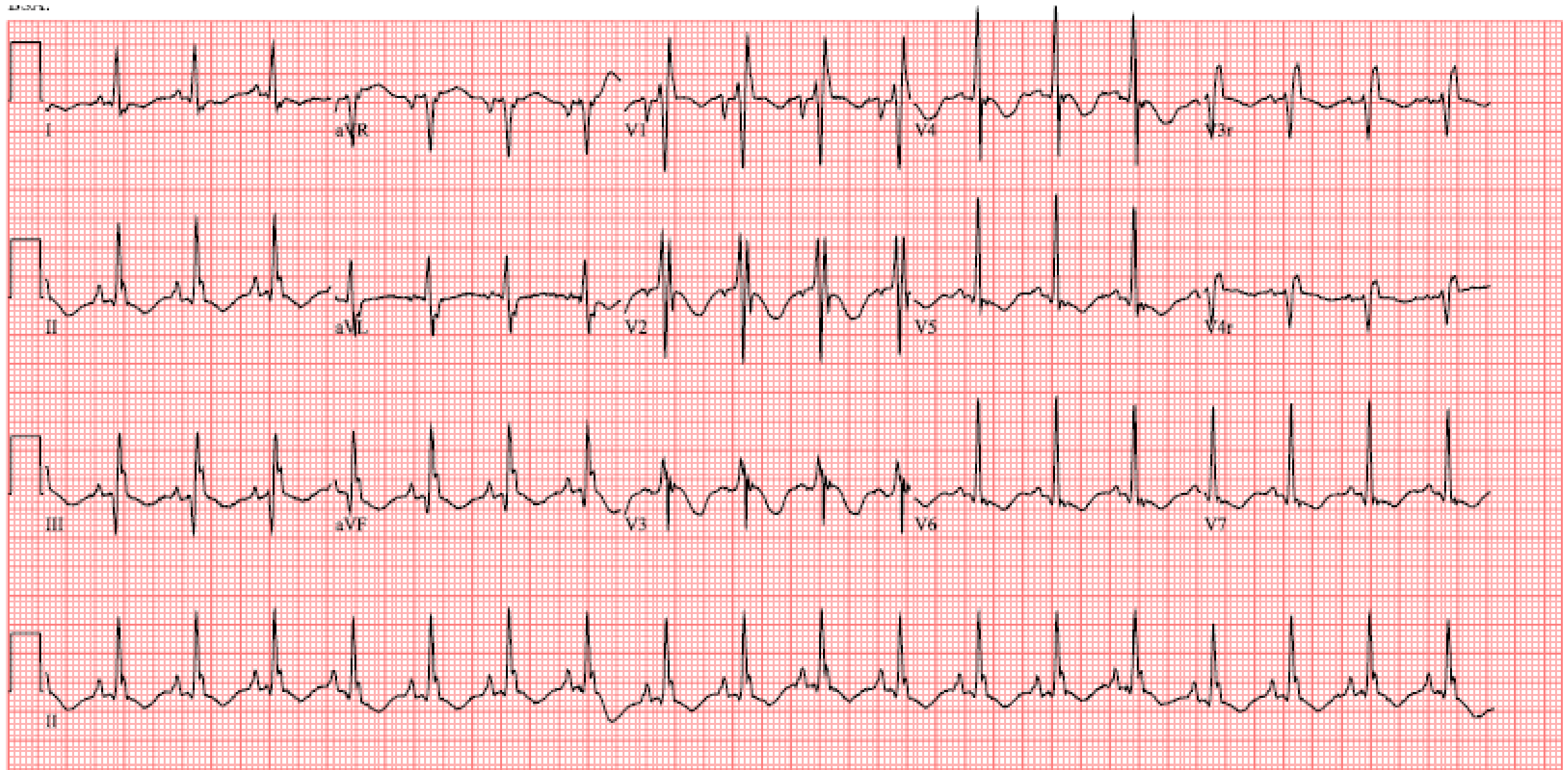
Age	Mean	Range
1 - 3 months	+70°	+10° to +125°
3 months - 3 years	+60°	+10° to +110°
3 years +	+60°	+20° to +120°
Adult	+50°	-30° to +105°

# ECG of large VSD: biventricular hypertrophy





2020.



# How to read an ECG

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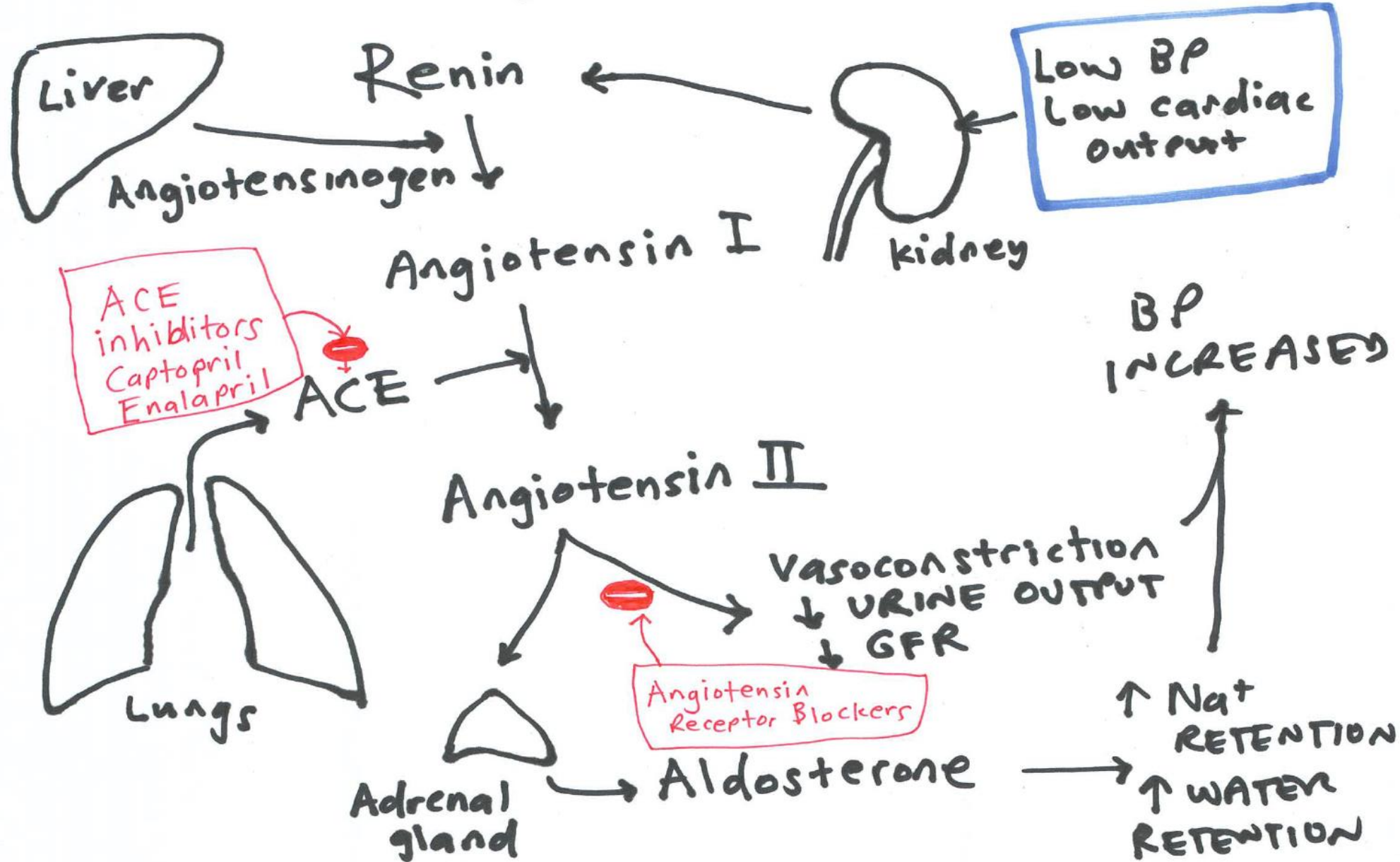
# Why do children with VSD go into heart failure?

- 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
- Digoxin
  - Rate control
  - AF

# Renin - Angiotensin - Aldosterone

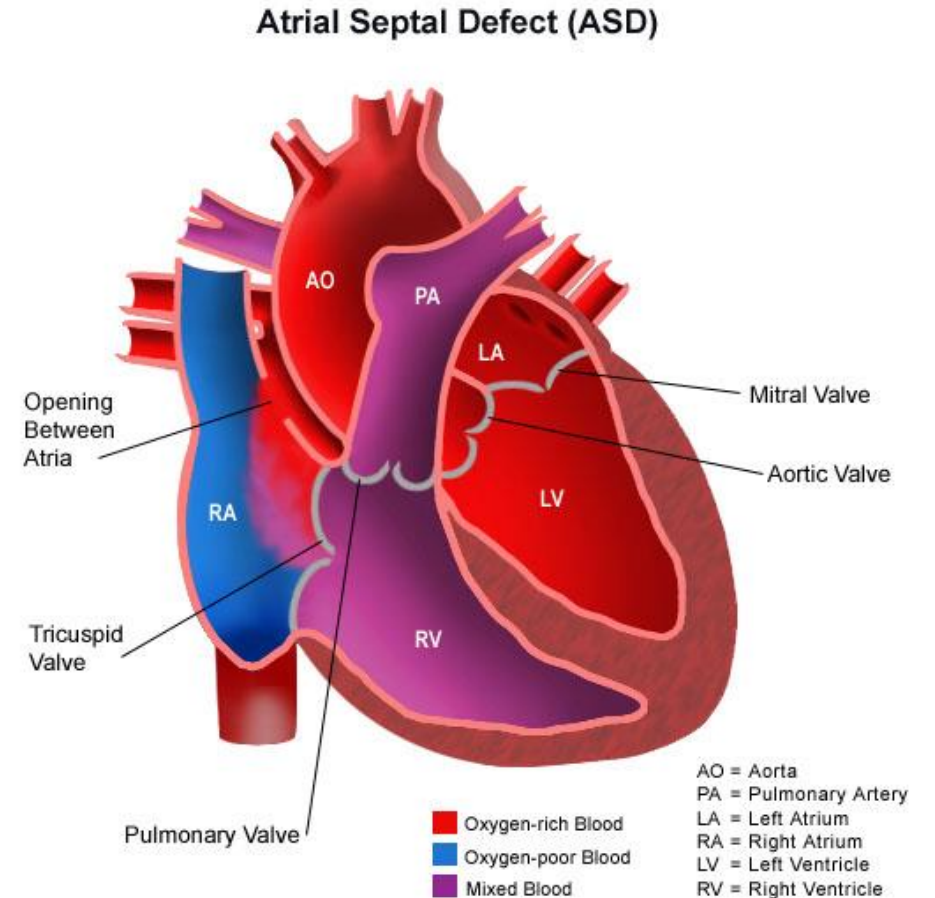


# VSD: complications and indications for surgery

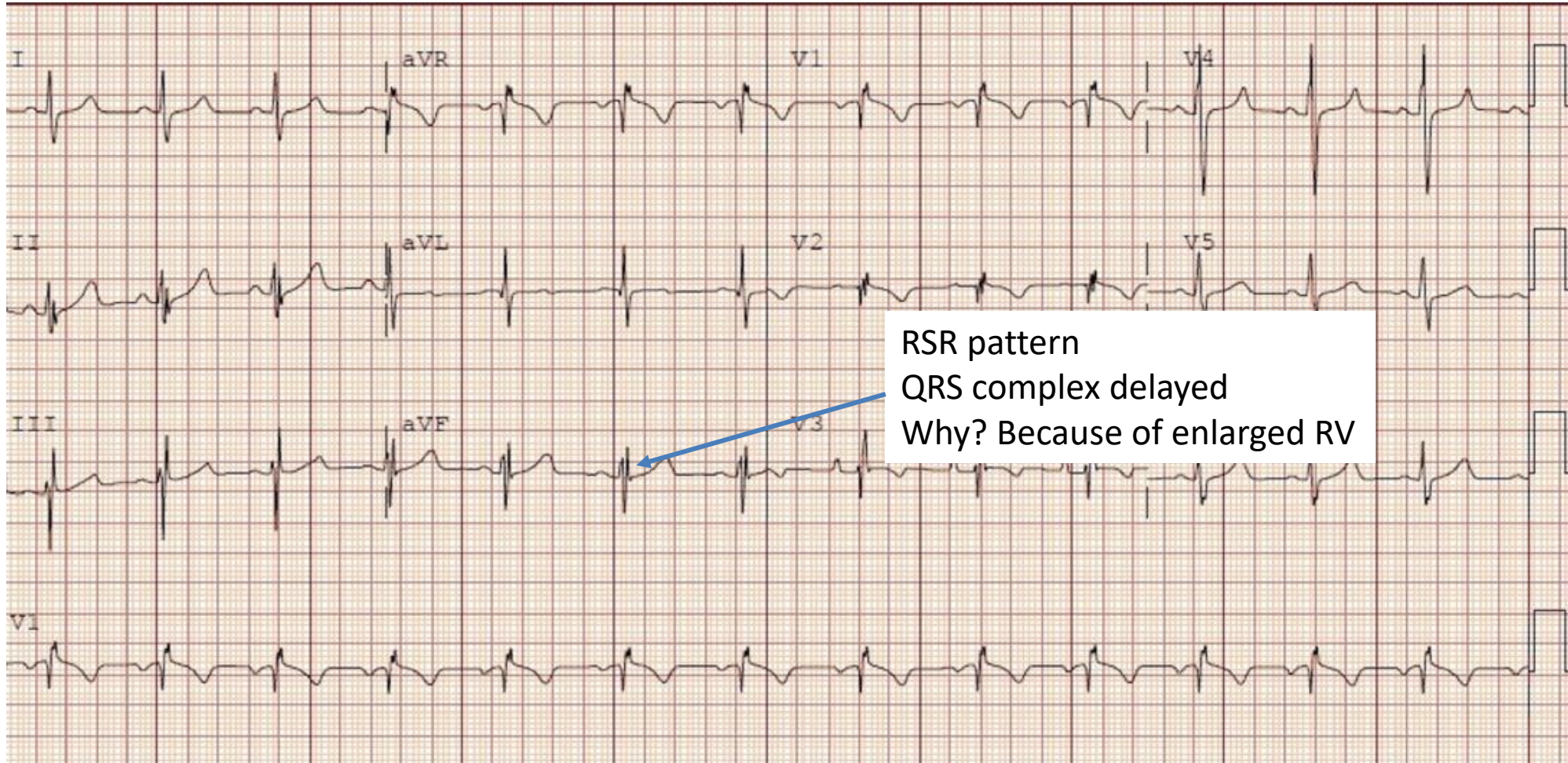
- Heart failure not controlled by medical management
- Failure to thrive, despite optimal medical management and nutrition
- Pulmonary hypertension (high QP: QS)
- Aortic valve prolapse into the VSD → aortic regurgitation
- Bacterial endocarditis

# ASD

- Primum ASD – low in the interatrial 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**



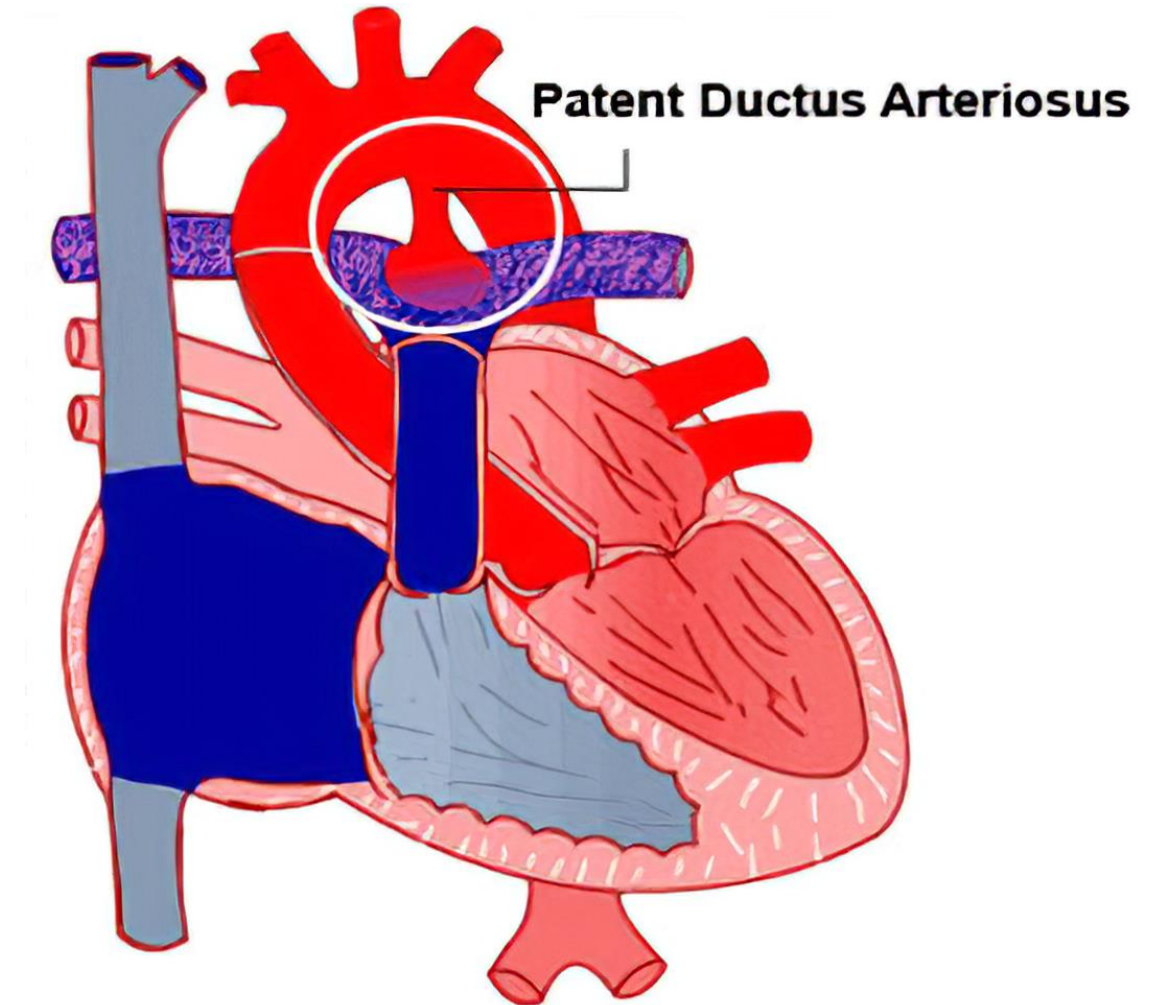
# ECG in ASD – sometimes right BBB





# 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

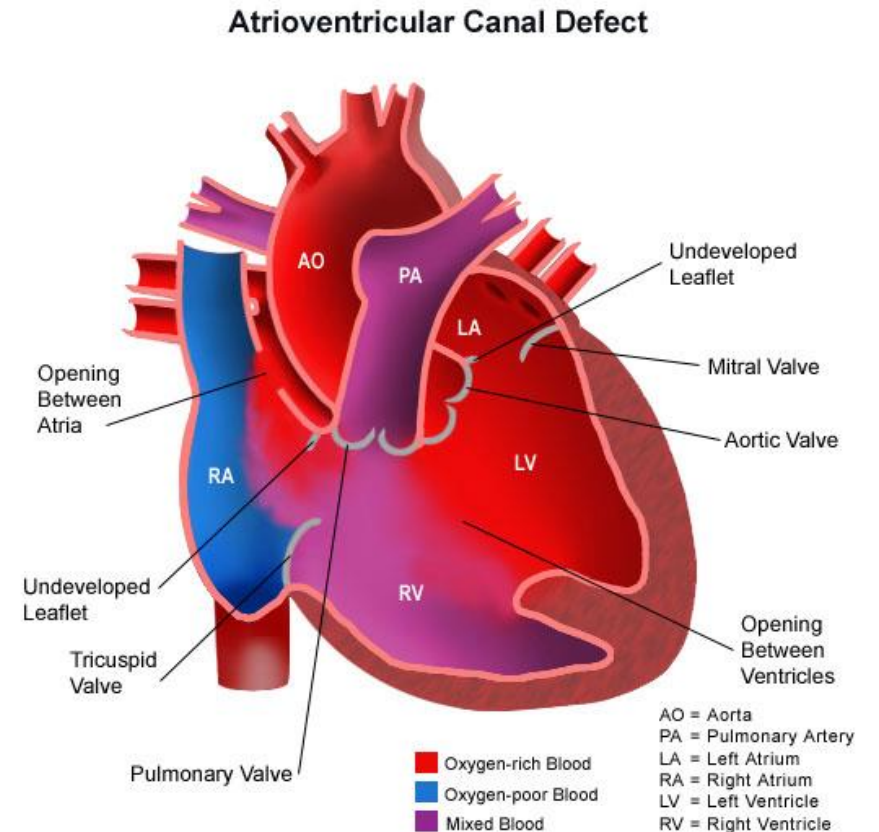


# Patent ductus arteriosus: treatment

- 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

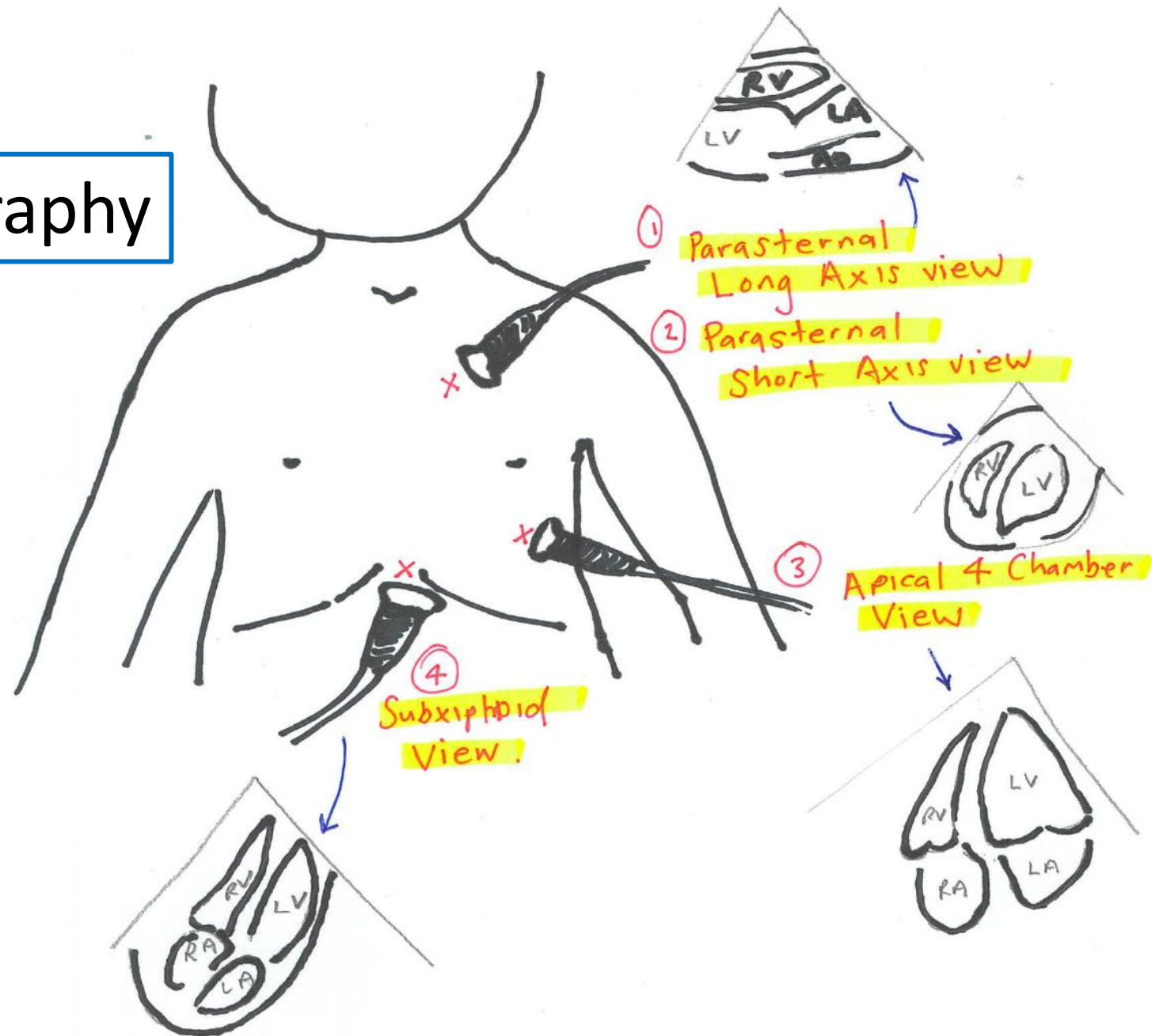
The newborn who is *well at birth*, discharged home, and returns in shock

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

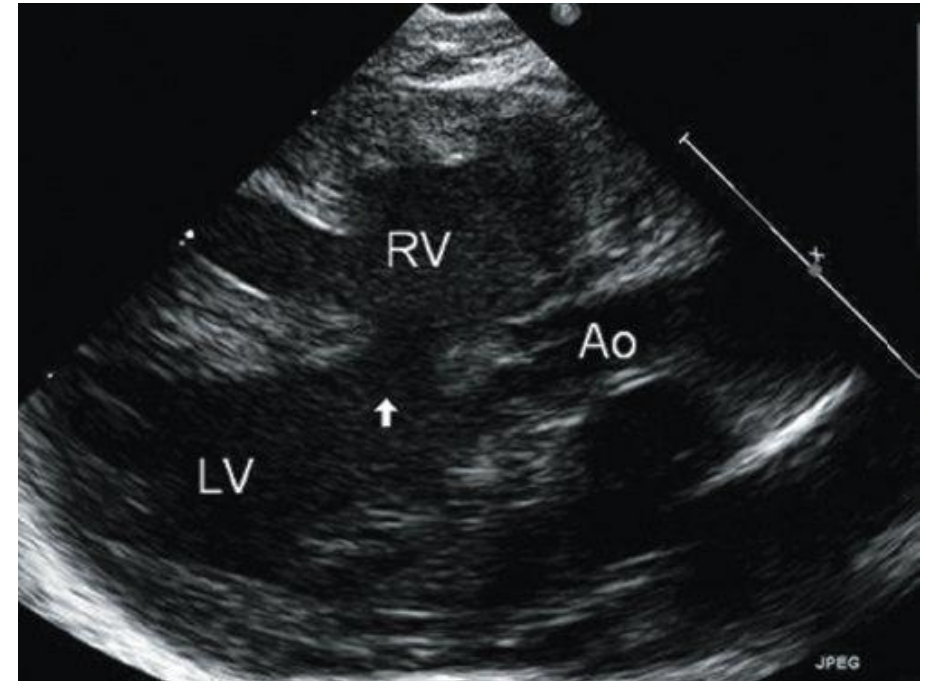
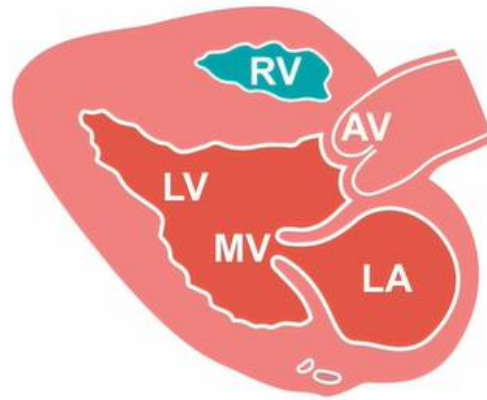
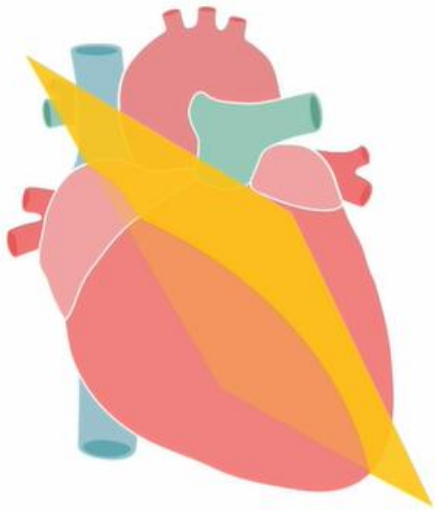
- Emergency treatment
  - Oxygen
  - Establish IV access (peripheral, umbilical)
  - Fluid resuscitation (10-20ml/kg IV)
  - Antibiotics – ceftriaxone, amikacin, cloxacillin
  - Adrenaline 0.05-0.1 mcg/kg/min
- Examination
  - Cardiac murmur, pulses
  - Signs of sepsis source
- Investigations
  - Chest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

# Echocardiography



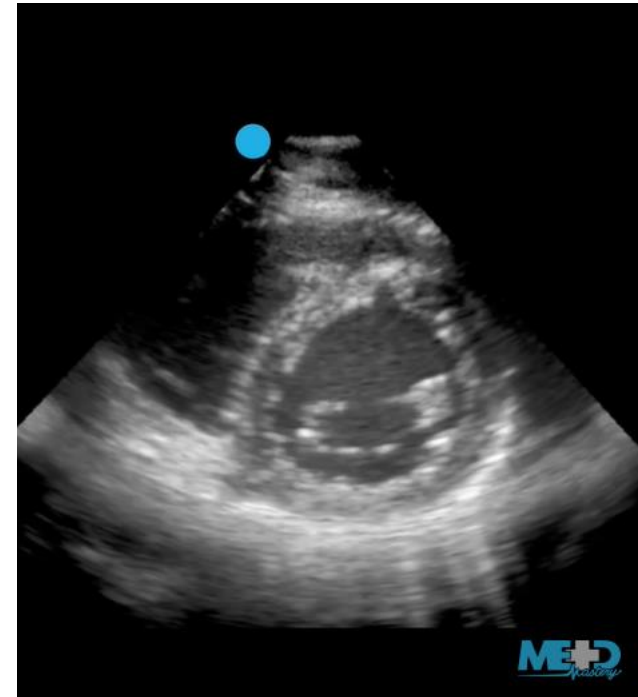
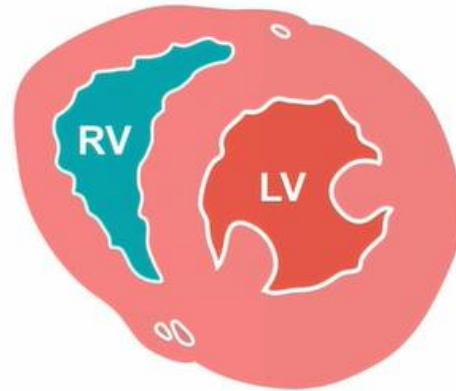
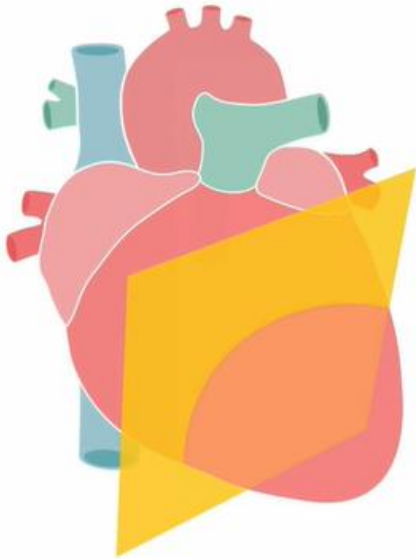


# 1. Parasternal long axis view



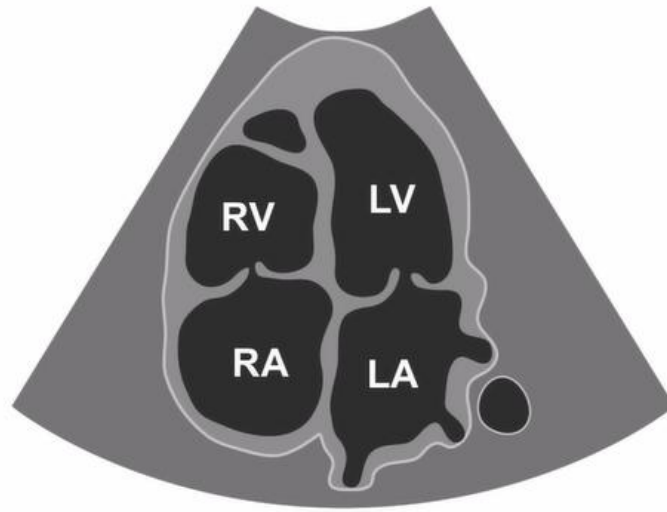
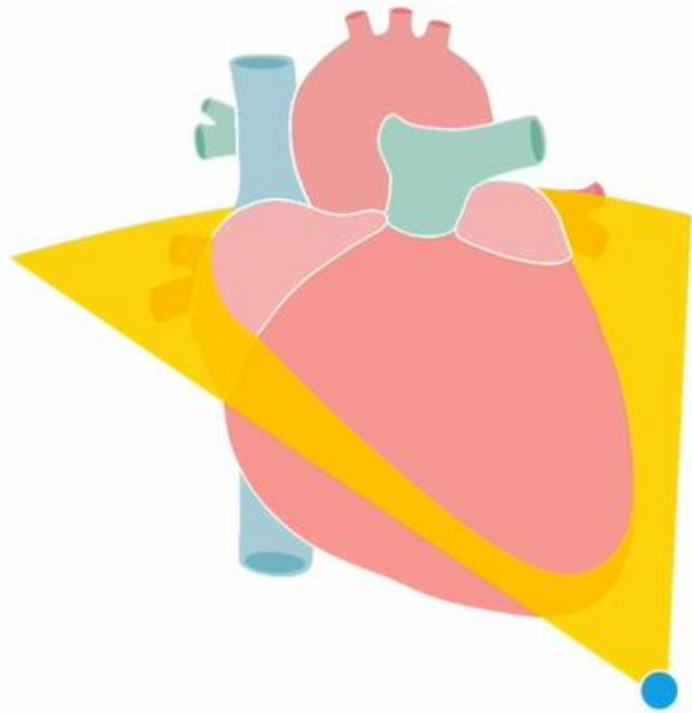
Probe placed to left of the lower third of the sternum 3<sup>rd</sup> / 4<sup>th</sup> ICS.  
Probe marker pointing towards **the right shoulder**

## 2. Parasternal short axis view



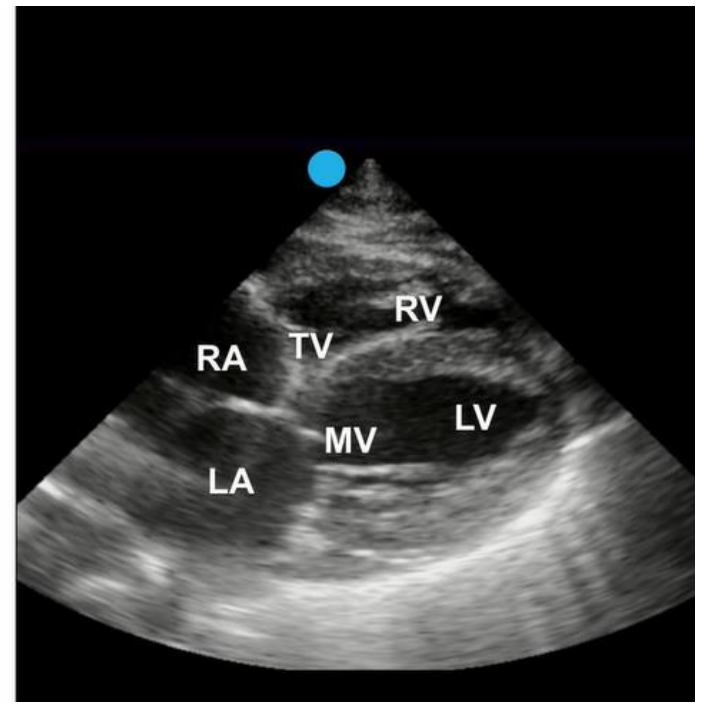
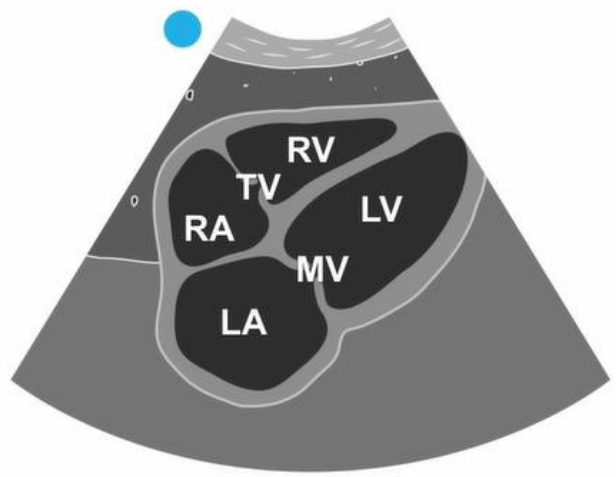
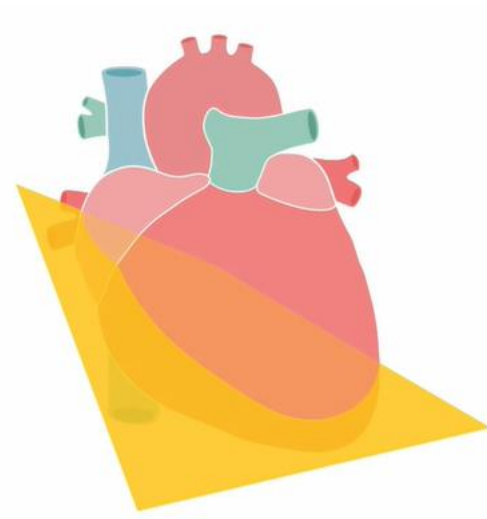
Probe placed to left of the lower third of the sternum 3<sup>rd</sup> / 4<sup>th</sup> ICS.  
Probe marker pointing towards **the left shoulder**

### 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