# Approach to a baby with cyanosis

# Objectives

- Cyanosis : types
- Differentials: cardiac vs. non cardiac
- Approach
- Case scenarios

# Cyanosis

- Greek word "*kuaneos*" meaning dark blue
- Bluish discolouration of skin, nail beds, and mucous membranes.
- Depends on absolute concentration of reduced haemoglobin (> 3 g/dl in arterial blood and >5 g/dl in capillary blood)

# Types of cyanosis

**ACROCYANOSIS**

Physiological upto 72 hrs
Large arterio-venous oxygen difference

**CENTRAL CYANOSIS**

Pathological
Requires immediate evaluation

**DIFFERENTIAL CYANOSIS**

Definitive congenital heart anomalies (right-to-left shunt through PDA)

# Differentials

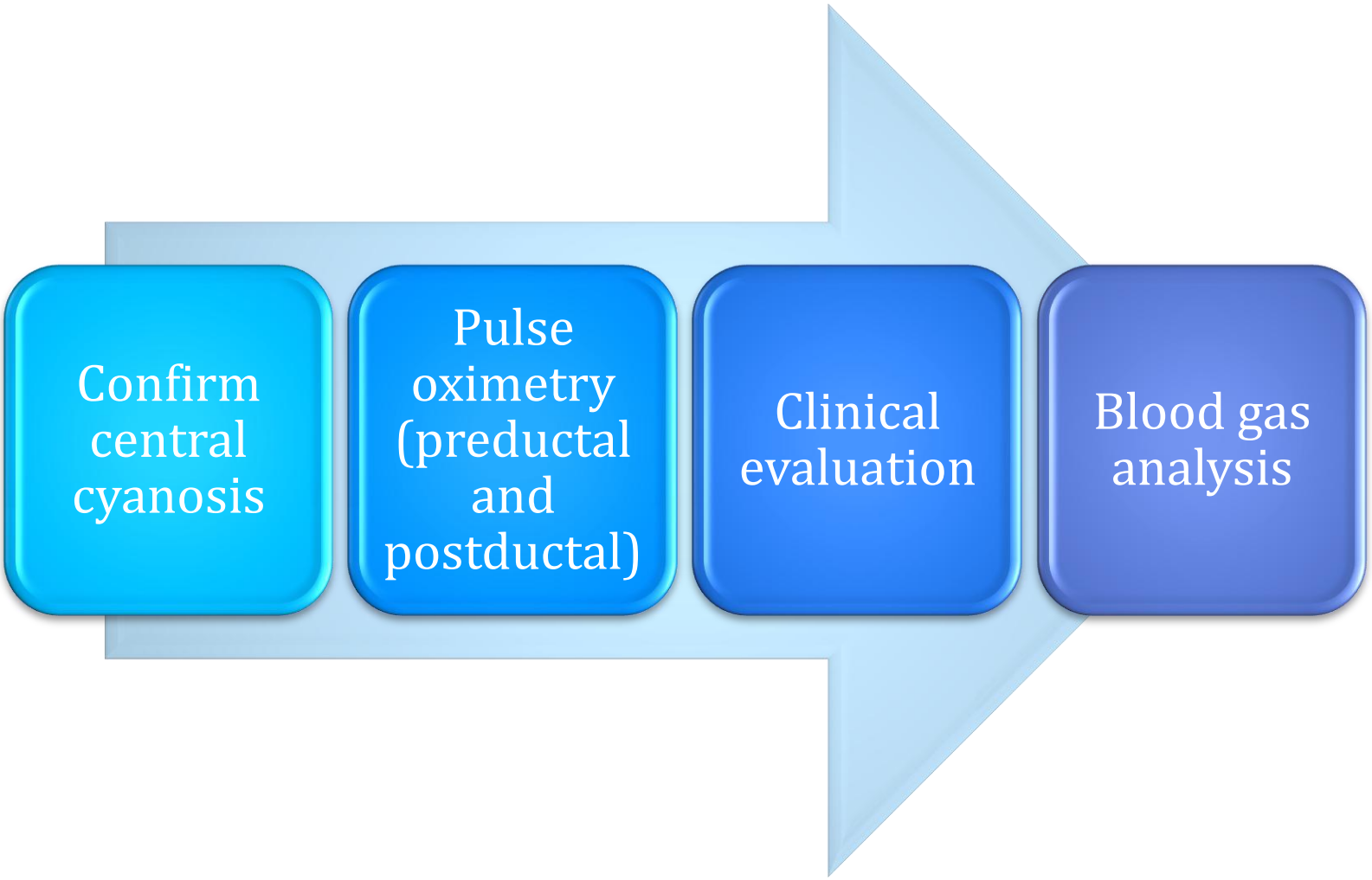
## Cyanotic heart disease

- Decreased pulmonary blood flow
- Increased pulmonary blood flow
- Severe pulmonary venous congestion

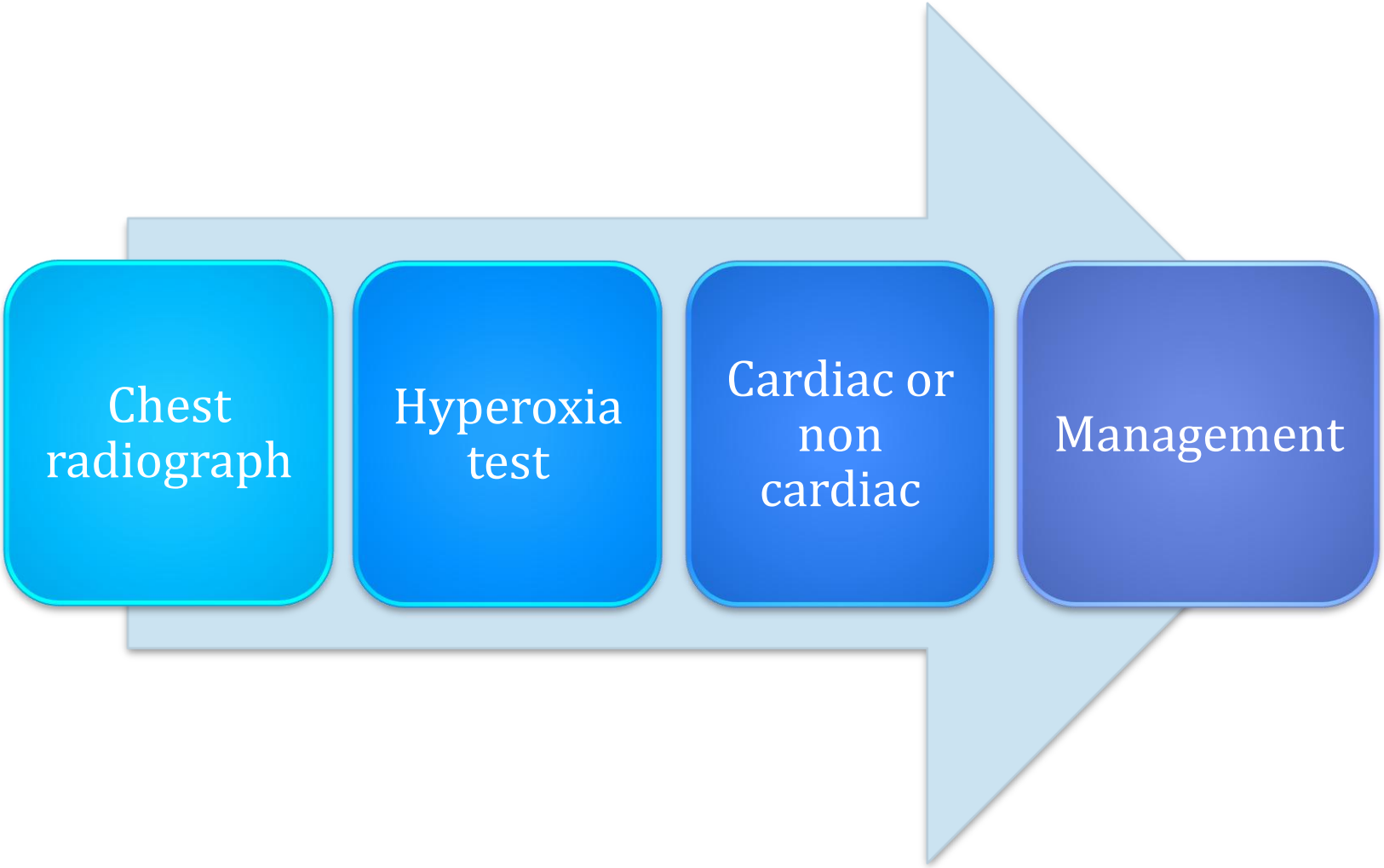
## Non cardiac causes

- Respiratory disorders
- Persistent fetal circulation
- Central nervous system disorders
- Miscellaneous

# Approach


Confirm central cyanosis
Pulse oximetry (preductal and postductal)
Clinical evaluation
Blood gas analysis

# Approach contd……



# Approach contd……
## Age at presentation of cyanosis

| 0-7 days | 7-28 days | >28 days |
|---|---|---|
| TGA | Truncus arteriosus | TOF like physiology |
| PS +IVS | TAPVC | TGA, ASD |
| HLHS | TGA,VSD | Truncus Arteriosus |
| Severe Ebstein Anomaly | TOF | PPHN group |
| TAPVC (obstructed) | | |

# Approach contd……
## Pulse oximetry

- Simultaneous measurements from the right hand and a foot: flow patterns through the ductus arteriosus.
- Avoid left hand.
- Confirms/ rejects central cyanosis
- R → L ductal shunting if differential cyanosis

# Clinical evaluation: some pointers

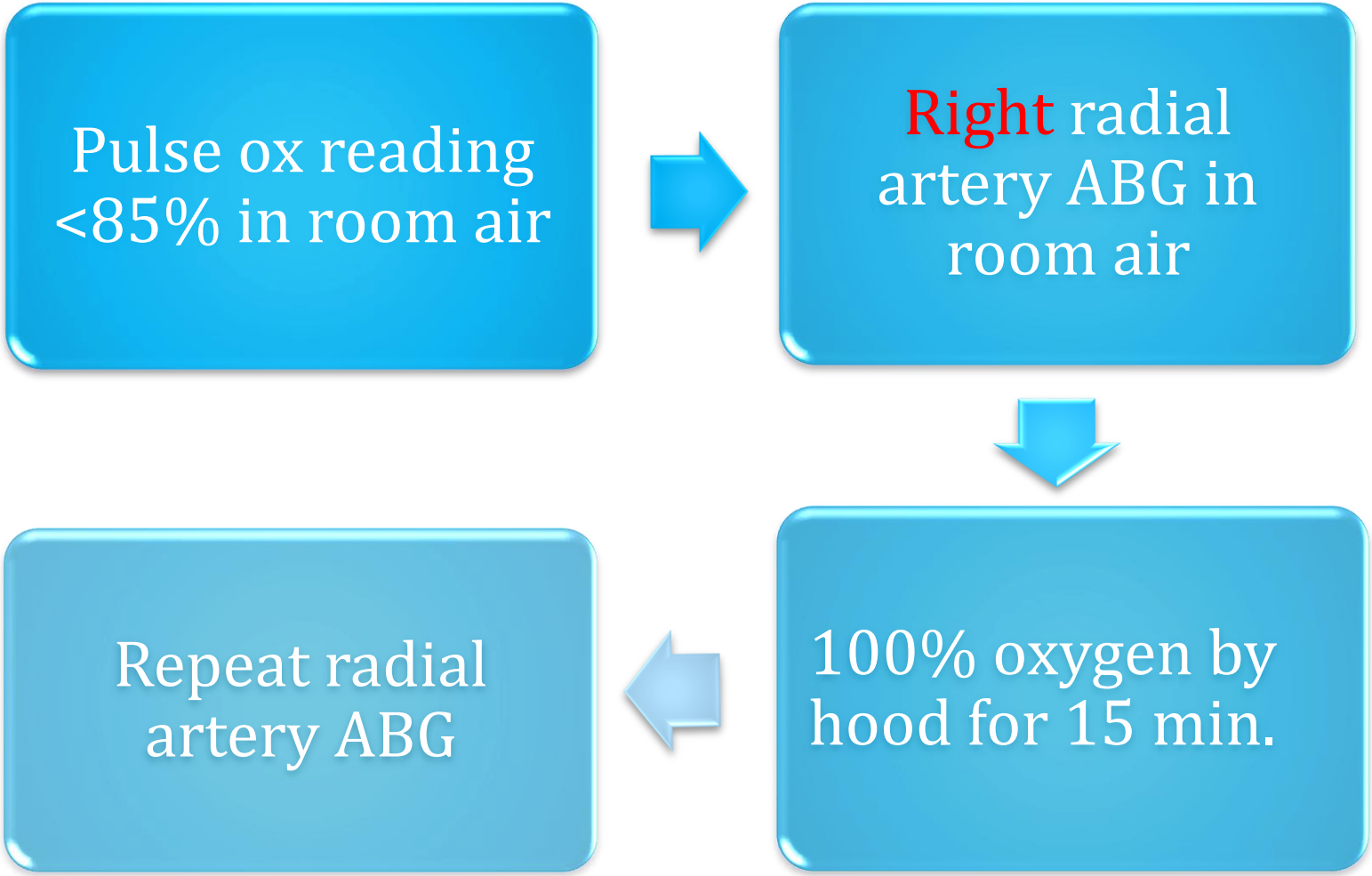
- Tachypnea with distress
- Crepitations +
- Cyanosis mild/uniform
- Responsive to oxygen
- Improves with crying
- Age: usually at birth

**NON CARDIAC**

- Tachypnea, no/ less distress
- Crepts -, except with PVH
- Cyanosis variable/ uniform
- No/ minimal response to oxygen, Worsens with crying
- Usually after 24 hrs

**CARDIAC**

# What next? Hyperoxia Test




Jones, 1976

# Interpret? Blood gas analysis

- Low pH
- Elevated PaCO2
- PaO2 >250 mm Hg after hyperoxia test (passed hyperoxia test)
- Respiratory acidosis predominantly

**NON CARDIAC**

- Low pH
- Normal or low PaCO2
- $PaO_2$ < 100 mm Hg/ Rise <10-30 mm Hg (failed hyperoxia test)
- Metabolic acidosis predominantly

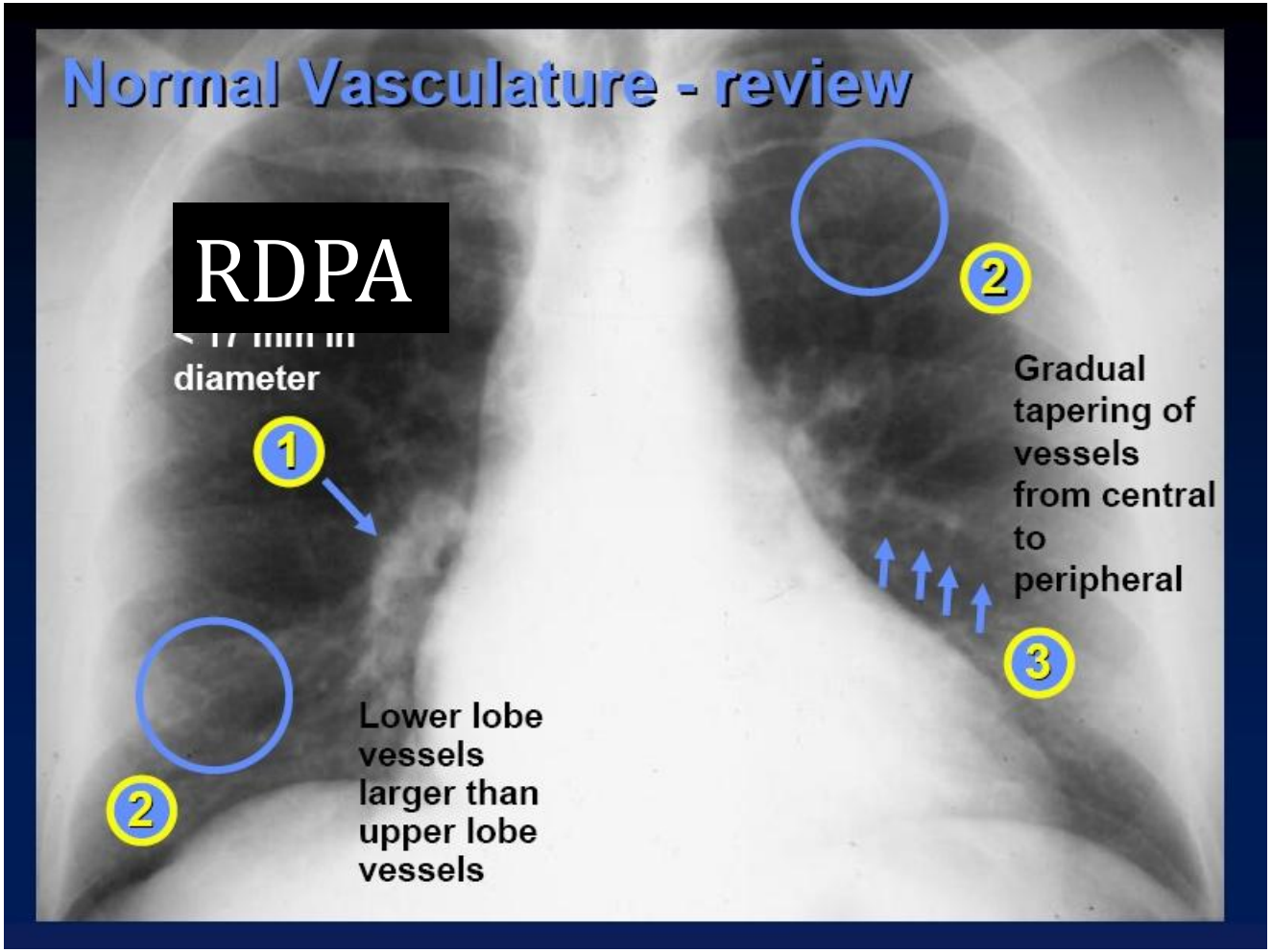
**CARDIAC**

# If still in dilemma?
## Hyperoxia Hyperventilation Test

- Intubation & hyperventilation

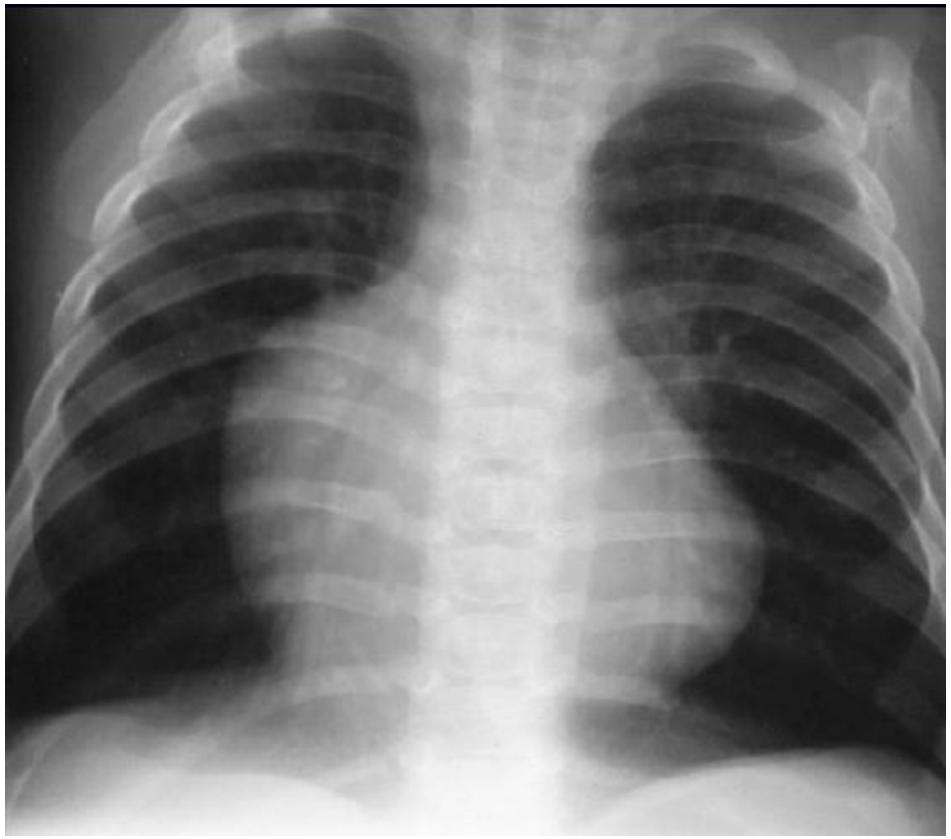
**Rationale**: Pulmonary vasodilation, decreases right to left shunt at atrial or ductal level

# Approach contd……. (X Ray)
# Pulmonary vasculature (Normal)



# X-Ray: Decreased vascularity

- Dark Lung Field
- Thin peripheral vessels
- Small Hila



# Cyanotic heart defects with decreased vascularity (examples)

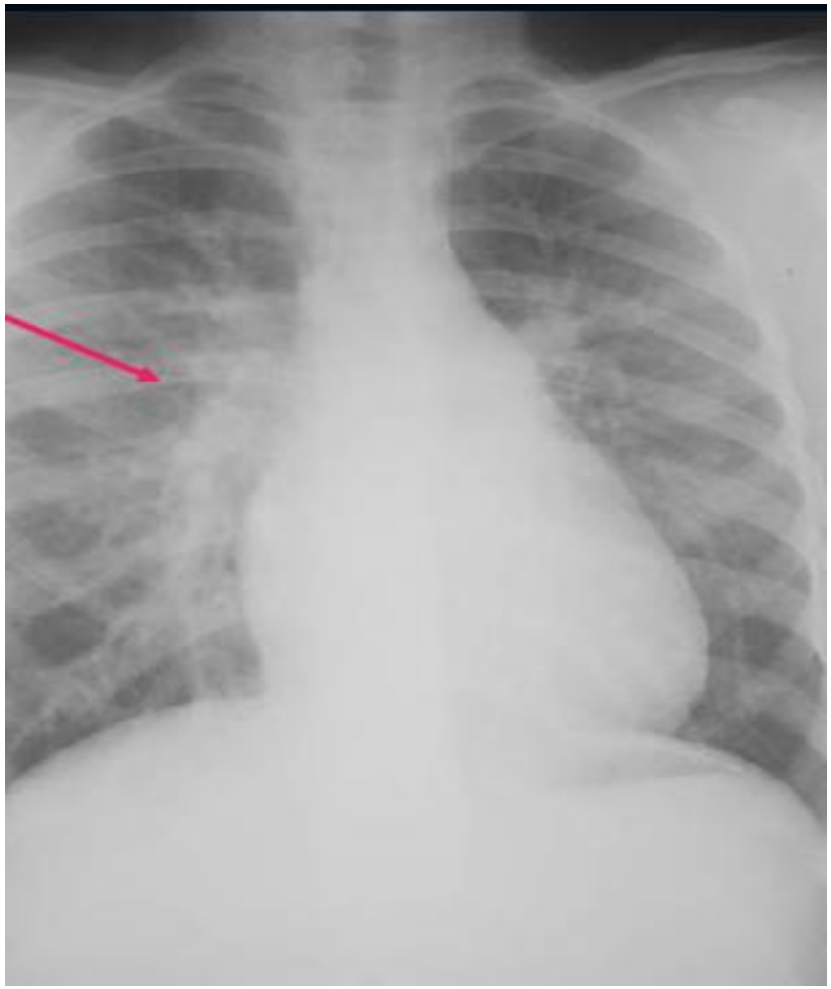
Critical Pulmonary stenosis/pulmonary atresia with intact ventricular Septum

Tetralogy of Fallot physiology

- TOF (VSD/ PS)
- DORV/ VSD/ PS
- AVSD/ PS
- TGA/ VSD/ PS
- Single ventricle/ PS
- Tricuspid atresia with restrictive VSD and/ or PS

# Increased vascularity

- Right des. PA dilated
- Prominent hilar vessels
- Pulm. vasculature traced till lateral 3rd of lung field
- End on vessels >4 in one lung field



# Cyanotic heart defects with increased vascularity (examples)

**Transposition physiology**

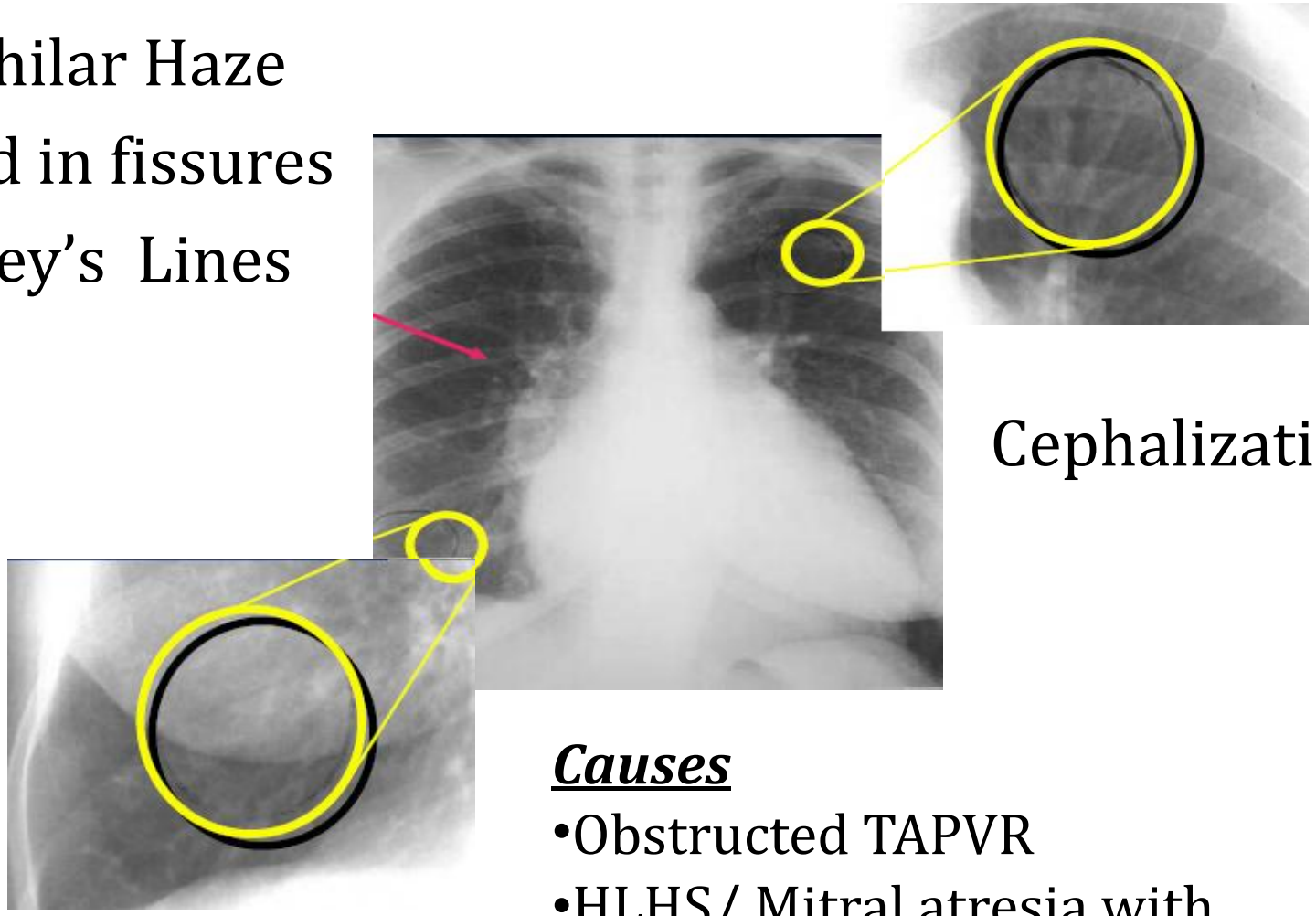
- Complete TGA
- DORV/ subpulmonic VSD (Taussig Bing)

**Admixture physiology without PS**

- At systemic or right atrial level: TAPVR, Mitral/ Aortic atresia with IVS
- At left atrial level: Tricuspid atresia
- At ventricle/ great artery level: Single ventricle, Complete AVSD with straddling AV valve, DORV/ subaortic or inlet VSD, Persistent truncus arteriosus

# Pulmonary venous hypertension

- Perihilar Haze
- Fluid in fissures
- Kerley's Lines



Cephalization

***Causes***

- Obstructed TAPVR
- HLHS/ Mitral atresia with restrictive ASD

# Questions which need to be answered

- Is there an imminent risk of death?
- What group of cardiac lesion?
- What further investigations?
- When to intervene?

*Making an exact diagnosis may not always be possible.*

# ECG: INTERPRETATION

- Axis : Leads I and aVF are used

1. P axis:P wave must be upright in leads I and aVF.

   0 to +90 degree = normal

   +90 to +180 degree = Atrial inversion

   0 to -90 degree = Ectopic atrial pacemaker/ AV junctional rhythm

2. QRS axis: QRS axis is perpendicular to lead with equiphasic QRS complex (R=S)

3. T axis: T waves must be upright in lead I and aVF

# Normal QRS axis

| Age | Mean ( Range ) |
|---|---|
| <1 wk | ard +135 |
| 1week -1 month | +110 (+30 to +180) |
| 1-3 months | +70 ( +10 to+125) |
| 3 month- 3 years | +60 ( +10 to 110 ) |
| >3 years | +60 ( +20 to 120 ) |
| Adult | +50 (-30 to +105) |

# Abnormal QRS axis

- LAD –QRS axis is less then lower limit of normal for age.
  (a) LVH (b) LBBB
  (c) Left anterior hemiblock
- RAD –QRS axis is greater then upper limit of normal for age.
  (a) RVH (b) RBBB
- Superior QRS axis: S>R in aVF
  (a) Endocardial cushion defect (ECD)
  (b) Tricuspid atresia
  (c) RBBB

# Further Evaluation

- Echocardiography: To confirm the type of lesion
- Cardiac catheterisation studies
- Angiography: confirmation, haemodynamics, oxygenation, intervention
- MRI: diagnostic for anomalies in pulmonary arteries, aorta, and vena cava

# Mangement: Role of PG E1

Indications:

- Cyanotic newborn suspected to have duct dependent lesion
- Echo proven duct dependent cardiac lesions

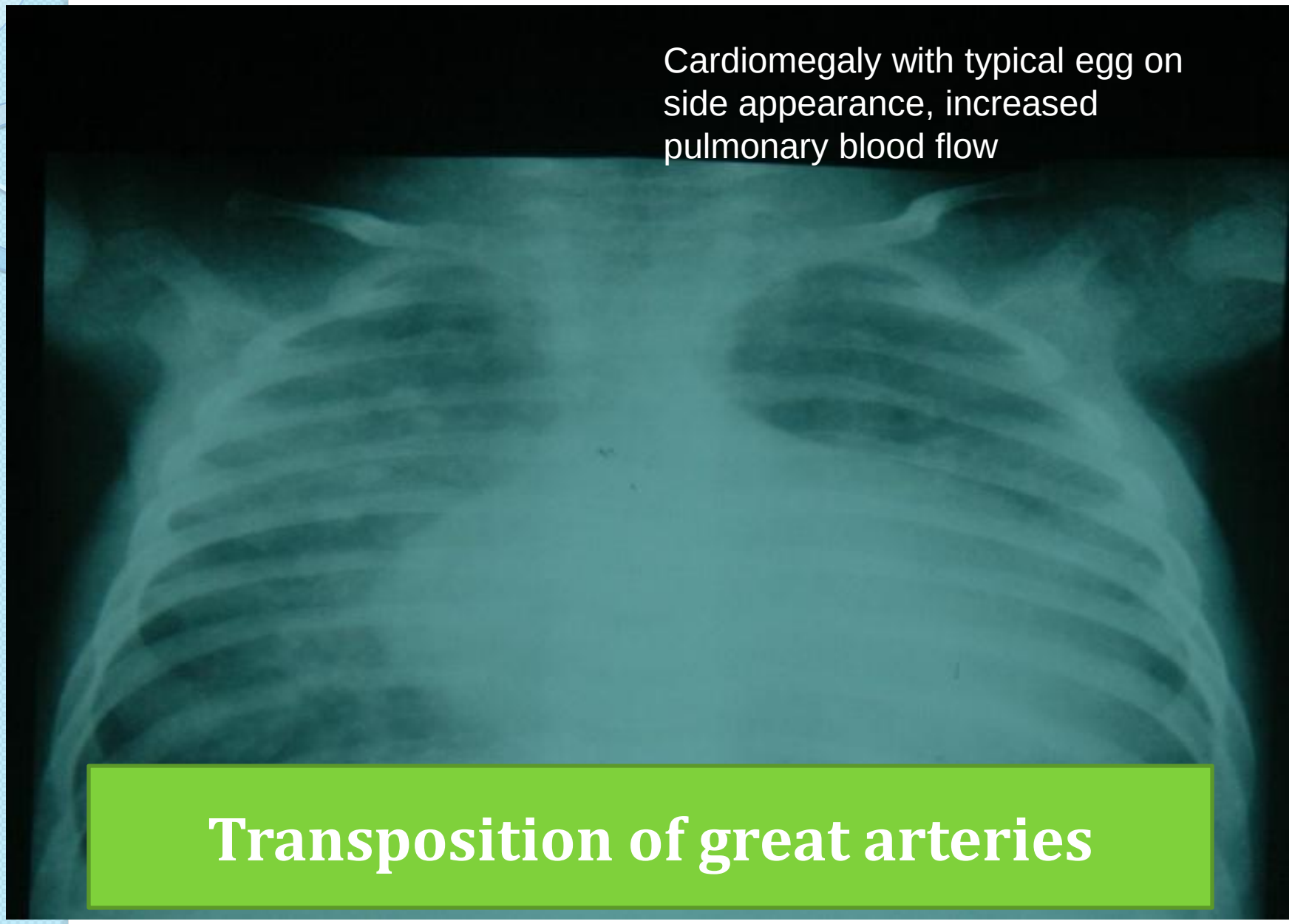
Dose: 0.01mcg/kg/min to 0.1 mcg/kg/min; gradually dec. to 0.025 mcg/kg/min before stopping (Neofax 2010)

Side effects: Apnea, pulmonary congestion, fever, hypotension, seizures, and diarrhea

# Case 1

A neonate is profoundly cyanosed and lethargic in his cot at 22 hours of life.

- Clinical examination reveals a soft systolic murmur heard at the left sternal edge and a single second heart sound
- Blood gas: unavailable
- ECG :normal neonatal pattern
- Chest X ray: available

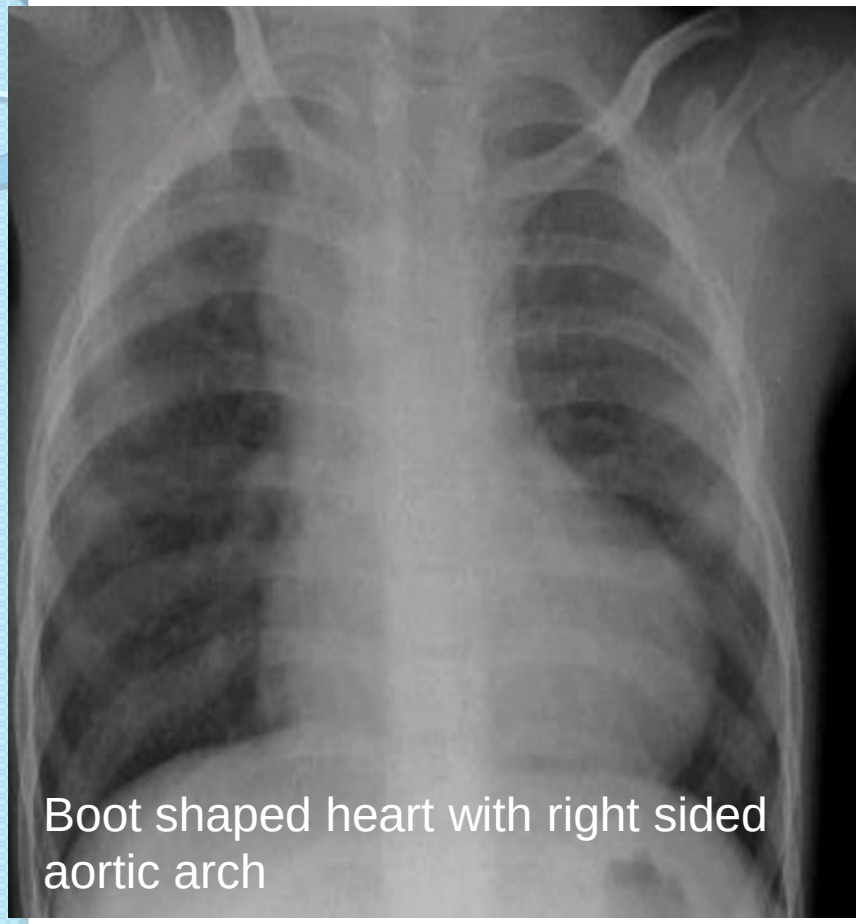



# Transposition of great arteries

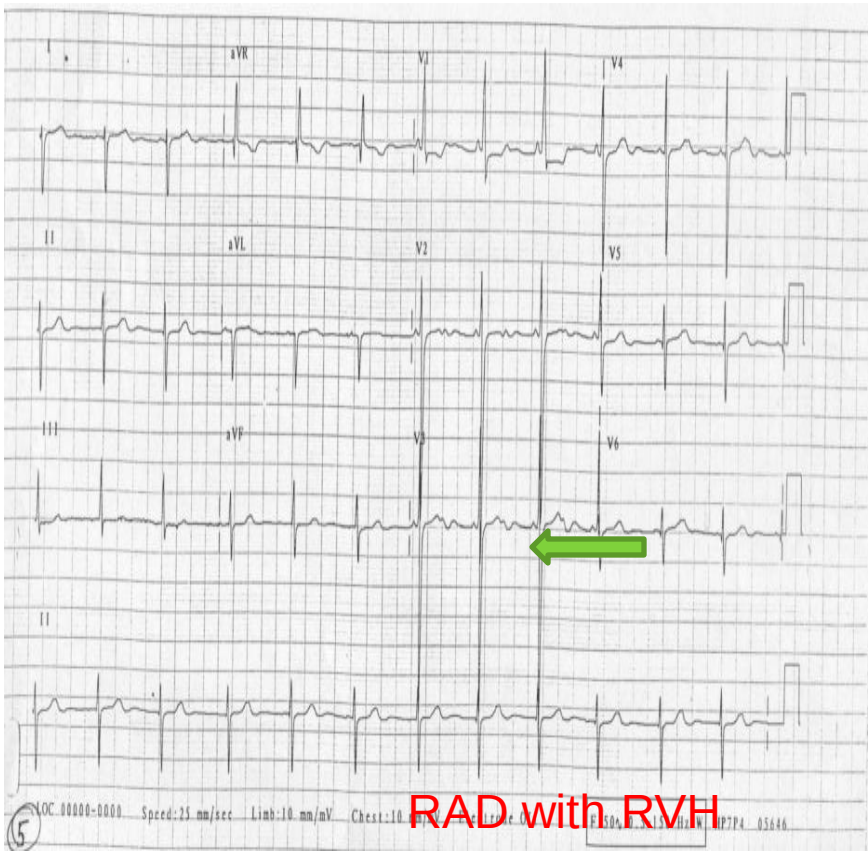
# CASE 2

A 3 mo infant presented with bluish discoloration of lips on crying since past 2 weeks

- No H/o suck-rest –suck cycle/ sweating/ cough or breathlessness
- Clinical examination reveals HR:110/min, RR:28/min. Central cyanosis+ worsening on crying. Apex beat in $4^{th}$ ICS inside MCL . ESM Grade 3/6 best heard in Pulmonary area. S1 N S2 single
- ECG and chest X ray is available



Boot shaped heart with right sided aortic arch

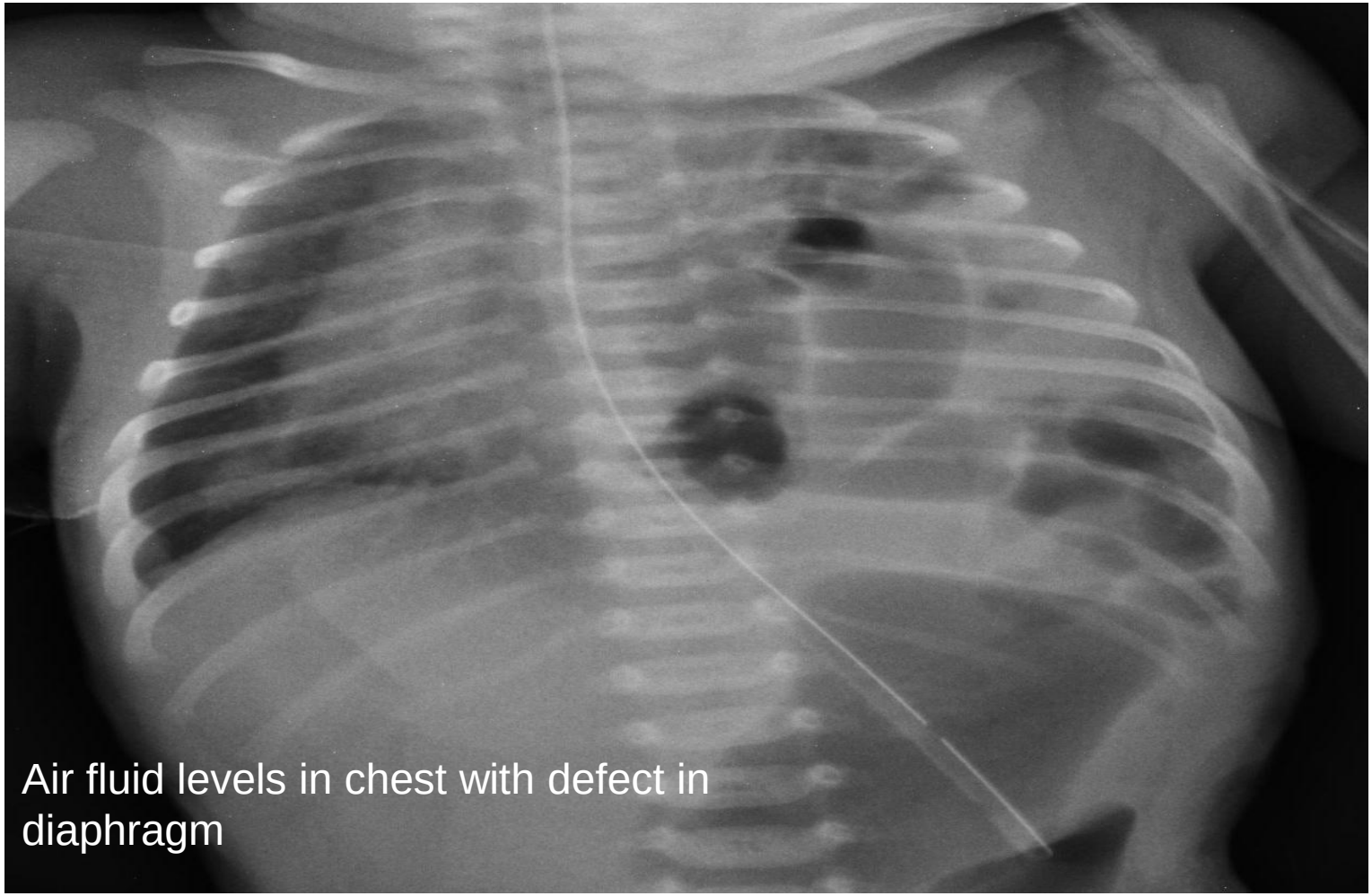



# Tetralogy of Fallot

# Case 3

Preterm (34 wks) neonate born by normal vaginal delivery with mild respiratory distress and cyanosis

- Put on CPAP
- Spo2 decreased from 95% on room air to 78% on 45% Fio2
- RR=60/min with Intercostal recession with decreased air entry on the left
- CVS: S1 S2 normal. No murmur




# Congenital diaphragmatic hernia