
INDIAN ACADEMY OF PEDIATRICS
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TOPIC: ROAD MAP TO CYANOTIC CHD
CHAIRPERSON
DR ML GUPTA
MODERATOR
DR SANJAY KHATRI
PEDCCARDIOLOGIST MGMC
EXPERT
DR RAMBABU SHARMA
PRESENTER
DR. SHIPRA MAJUMDAR
DR. NISHTHA GUPTA

Patient Identification:

Name: Master AS

Age/Sex: 3 years / Male

Address: Mansarovar, Jaipur

Informant: Mother (reliable)

Chief Complaints:

1. Bluish discoloration of lips, tongue, and fingertips since 6 months of age
2. Episodes of exertional dyspnoea since 1½ years of age
3. Poor weight gain since infancy

History of presenting illness

Patient was apparently well till 6 months of age

BLUISH DISCOLORATION

- The mother first noticed that his lips, tongue, and fingertips turned blue.
- Initially, this blue discoloration appeared only when he cried excessively or during feeding.
- However, gradually over time, this bluish colour became more frequent and noticeable even when the child was calm or resting.

Mother noticed for last 2-3 months; the child had episodes of worsening cyanosis which were associated with transient loss of consciousness which got relieved by knee-chest position.

History of presenting illness

EXERTIONAL DYSPNEA

- Around one-and-a-half years of age, the mother started noticing episodes - child would suddenly develop difficulty in breathing, especially during active play, running, or when he was upset.

- During these episodes, the child seemed very restless and uncomfortable, breathing heavily and rapidly, and would sit down or crouch suddenly, hugging his knees tightly to his chest. After sitting like this quietly for a few minutes, his breathing would slowly improve, and he would resume his activities.

- Currently, these episodes happen about two or three times a week, lasting around five to ten minutes each. The mother also reports that the child gets tired very easily. He often stops playing early, preferring to sit down rather than run around like other children his age.

History of presenting illness

IMPROPER GROWTH

- Despite the child eating normally—similar meals as other children in the family—he does not seem to gain weight properly.
- Compared to his older sibling and friends of the same age, he appears thinner and smaller.

Negative history

No history of persistent cough, noisy breathing or recurrent admissions

No history of swelling over body, puffiness of face or pedal edema

No history of excessive sweating over forehead or during feeding

No history of abnormal body movements

No history of hemoptysis

No history of unexplained childhood deaths in family

Past history

- Child was growing well in the first 6 months of life.
- No history of any admissions in the past

Birth history

- Antenatal period – uneventful no history of fever or rash , no history of any unusual drug
- Booked case, full - term baby born via normal vaginal delivery, cried immediately after birth with birth weight of 2.8 kgs, institutional delivery with no perinatal complications
- Postnatal period – uneventful

Immunization History

- Immunized for age
- BCG mark present on left upper arm

Developmental Milestones

Gross Motor	Fine Motor	Social/Adaptive	Cognitive/Language
➢ Run well ➢ Alternate feet while going upstairs	➢ Makes tower of 9 blocks	➢ Dry by day	Knows his full name and gender

No gross delay in achieving milestones in all domains

Dietary History

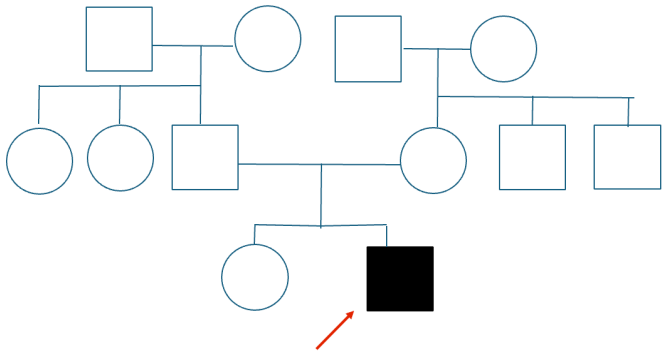
Calculated as per dietary recall history

	INTAKE	RECOMMENDED	% DEFICIT
CALORIE (Kcal/day)	850	1000	15%
PROTEIN (g/day)	11	12	8%

Time	Food Item	Quantity	Approximate Calories (kcal)	Protein (g)
8:00 am	Milk (cow's milk)	300 ml	200	4
9:30 am	Paratha with ghee	1 small	140	2
1:00 pm	Rice with moong dal	½ katori each	100 + 80 = 180	3.5
Few slices of cucumber	-	5	-	
4:00 pm	Banana (½ medium)	-	50	0.5
6:30 pm	Milk with Bournvita	150 ml	120	4
8:30 pm	Roti + potato curry	½ roti + ½ katori		

Family history

- Non consanguineous marriage
- No family H/o similar illness
- Siblings – 1 older female sibling – healthy.



Socioeconomic history

- Modified Kuppuswamy score - 9 (Upper Lower class)

Education of the head	Score
Profession or honours	7
Graduate	6
Intermediate or diploma	5
High school certificate	4
Middle school certificate	3
Primary school certificate	2
Illiterate	1

Occupation of the head	Score
Legislators, senior officials and managers	10
Professionals	9
Technicians and associate professionals	8
Clerks	7
Skilled workers and shop and market sales workers	6
Skilled agricultural and fishery workers	5
Craft and related trade workers	4
Plant and machine operators and assemblers	3
Elementary occupation	2
Unemployed	1

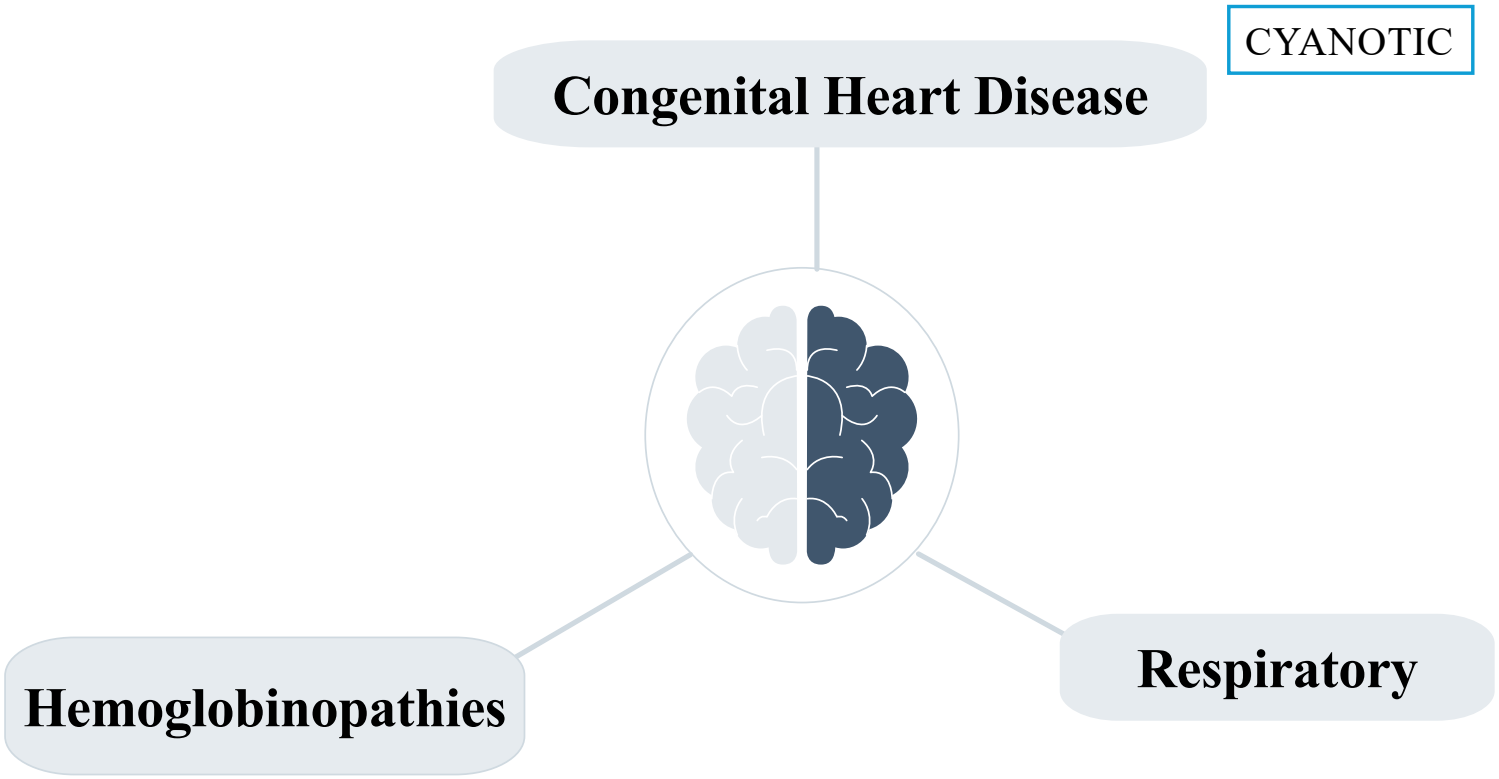
Updated monthly family income in rupees (2024)	Score
2,13,814 and above	12
1,06,850-2,13,813	10
80,110-1,06,849	6
53,361-80,109	4
31,978-53,360	3
10,703-31,977	2
≤10,702	1

Summary

AS, a 3-year-old boy, presents with a history of progressive bluish discoloration of lips, fingertips, and tongue since infancy associated with transient loss of consciousness, with progressive exertional dyspnoea relieved by crouching posture, and poor weight gain despite adequate nutrition.

Possibilities?

CYANOTIC


Congenital Heart Disease
Hemoglobinopathies
Respiratory

Cyanotic Congenital Heart Disease

↓ Pulmonary blood flow (R to L shunting)
• No history of recurrent LRTI
• Progressive cyanosis

• No signs of CHF present
  ○ Lethargy
  ○ Suck-rest-suck cycle
  ○ Profuse sweating over forehead and scalp

• Cyanotic spell and squatting

↑ Pulmonary blood flow (L to R shunting)
• History of recurrent LRTI

• Signs of CHF present
  ○ Lethargy
  ○ Suck-rest-suck cycle
  ○ Profuse sweating over forehead and scalp

CENTRAL CYANOSIS	PERIPHERAL CYANOSIS
• Oxygen saturation is <85%	• Oxygen saturation is normal
• Arterial blood desaturation	• Increased peripheral utilization
• Skin and mucous membrane involved	• Only skin involvement seen
• Skin - warm	• Skin – cold and clammy
• Clubbing/polycythemia +	• No clubbing/polycythemia
• No improvement with O_2/warming. Worsens with exercise	• Improvement with O_2 /warming / exercise
• Decreased PO_2	• Normal PO_2
• A-V O_2 difference is normal	• A-V O_2 difference > 12%

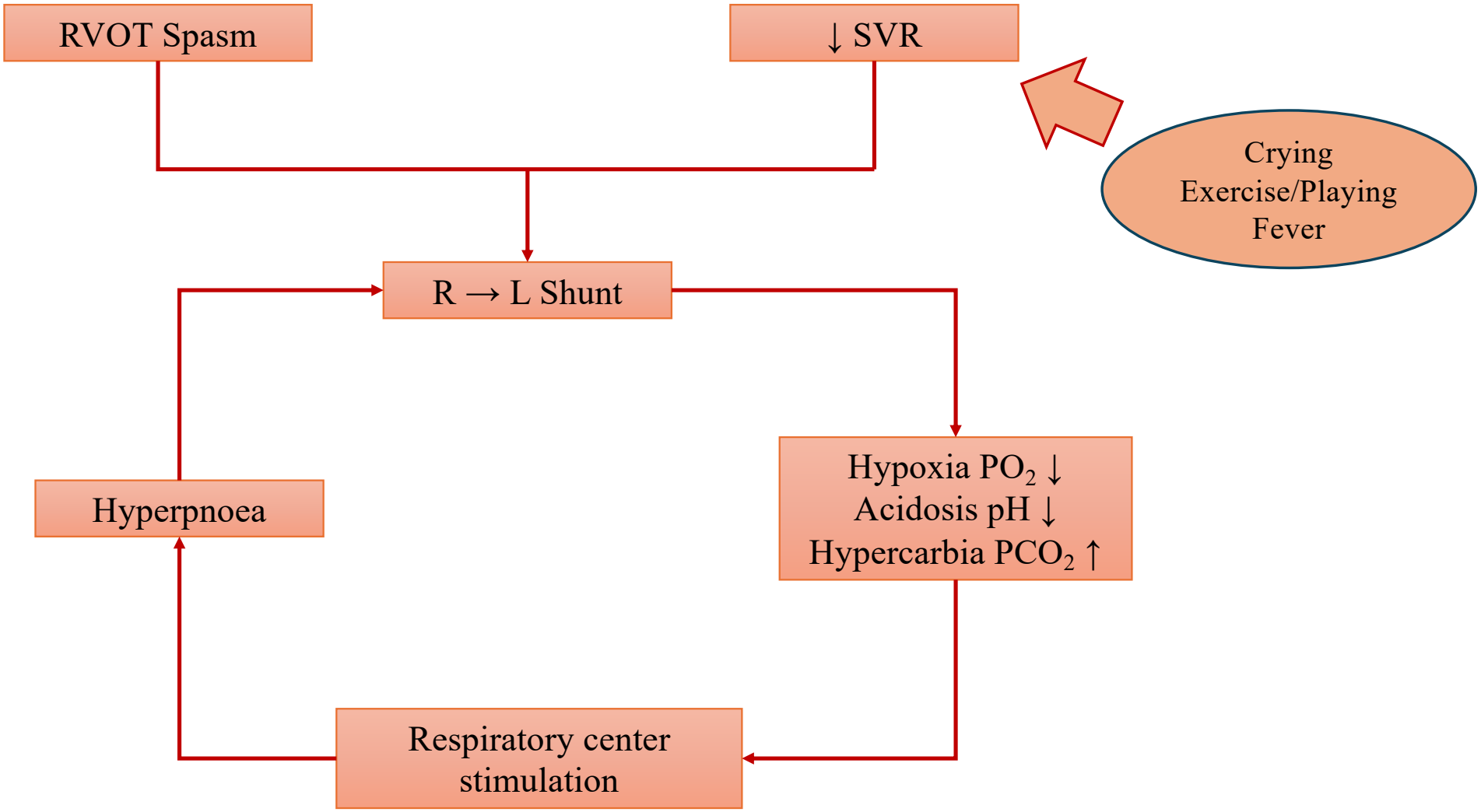
Questions

- **What is Cyanotic spell ?**
- **How squatting helps in Cyanotic spell ?**
- **What is differential cyanosis ?**

CYANOTIC SPELL

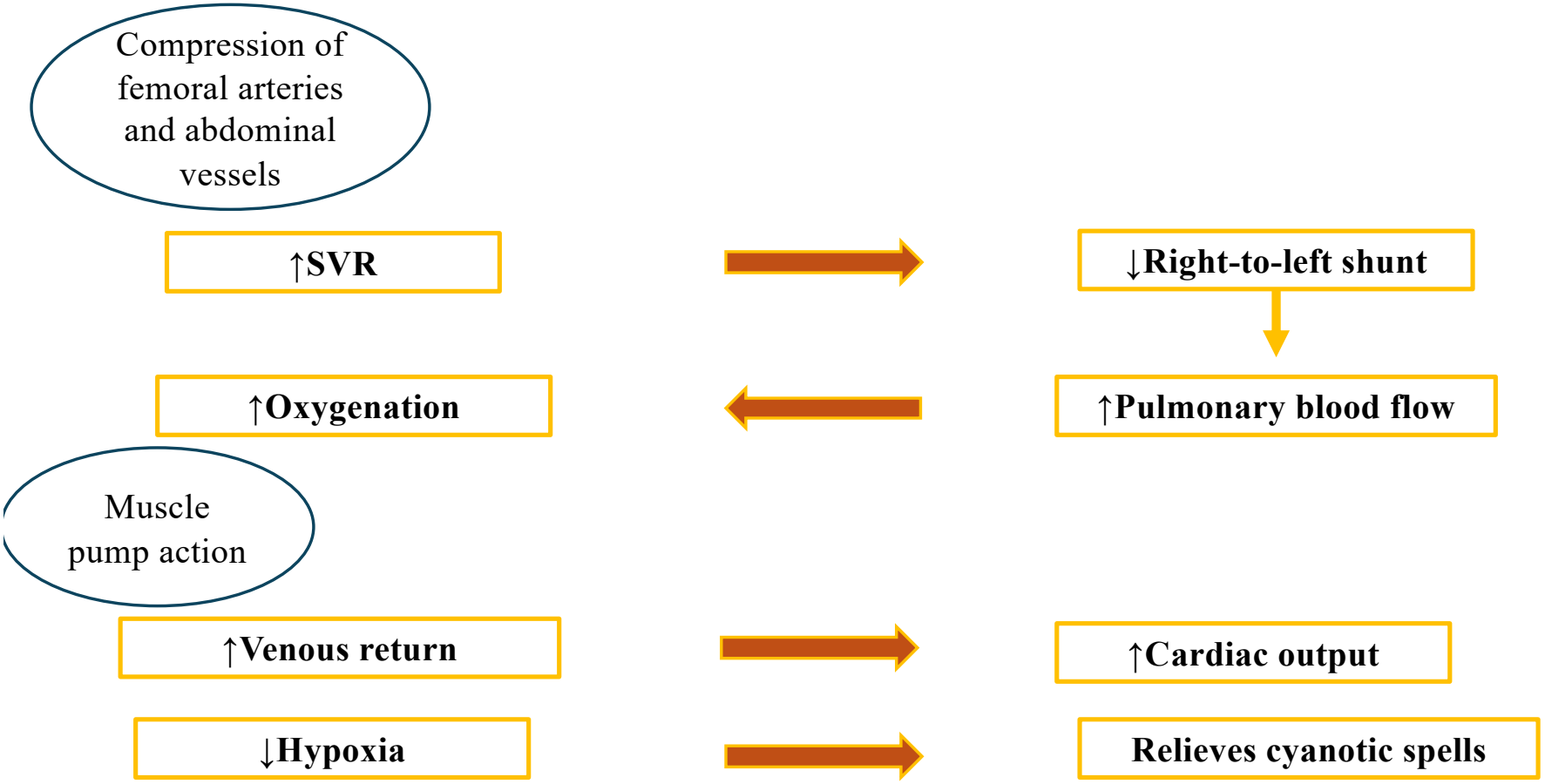
TRIAD:

- Worsening of cyanosis
- Hyperpnoea
- Disappearance of murmur

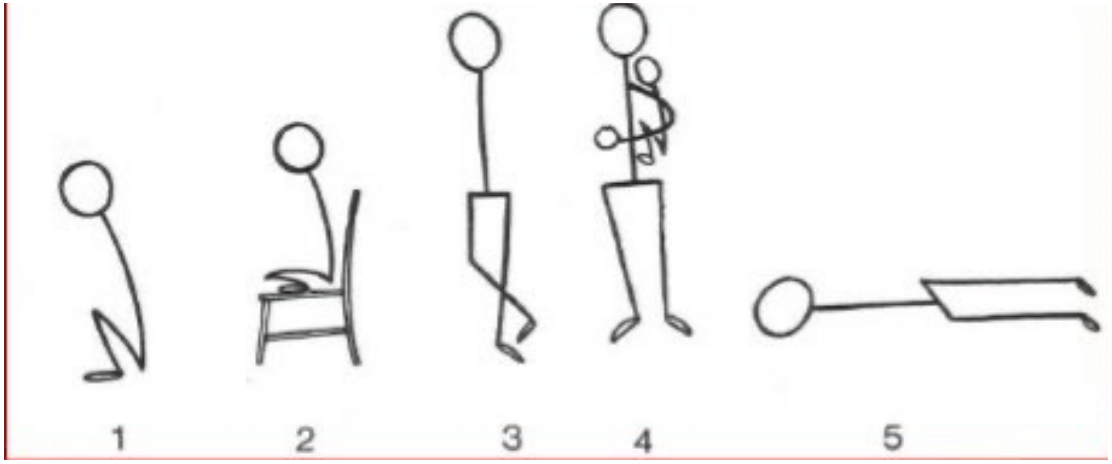

RVOT Spasm
↓ SVR
Crying
Exercise/Playing
Fever
R → L Shunt
Hypoxia PO_2 ↓
Acidosis pH ↓
Hypercarbia PCO_2 ↑
Hyperpnoea
Respiratory center
stimulation

HOW SQUATTING HELPS??

HOW SQUATTING HELPS??



SQUATTING EQUIVALENTS



1. Sitting down with legs drawn underneath
2. Sitting with legs drawn underneath
3. Legs crossed while standing
4. Holding the child with legs flexed up to the abdomen
5. Lying down

DIFFERENTIAL CYANOSIS

- **Upper limb - pink, lower limb - blue**
  - **PDA with reversal of shunt (PPHN)**
  - **Interrupted aortic arch with PDA**
  - **Severe CoA with PDA**

General physical examination

General physical examination

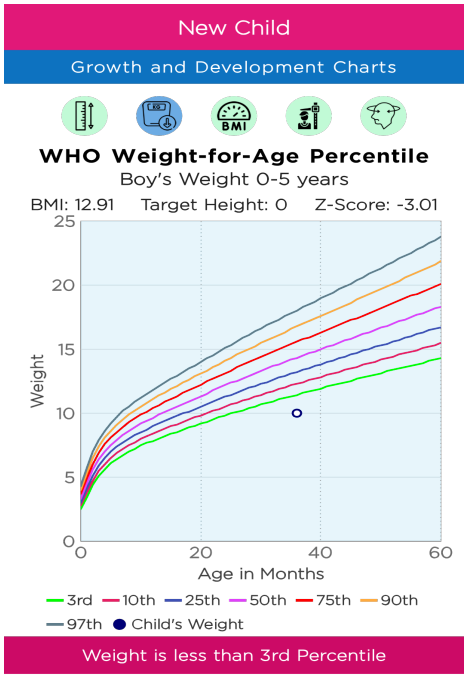
Child conscious, alert, cooperative, but appears small and thin for age.
Comfortably seated in mother's lap; no signs of distress at rest.

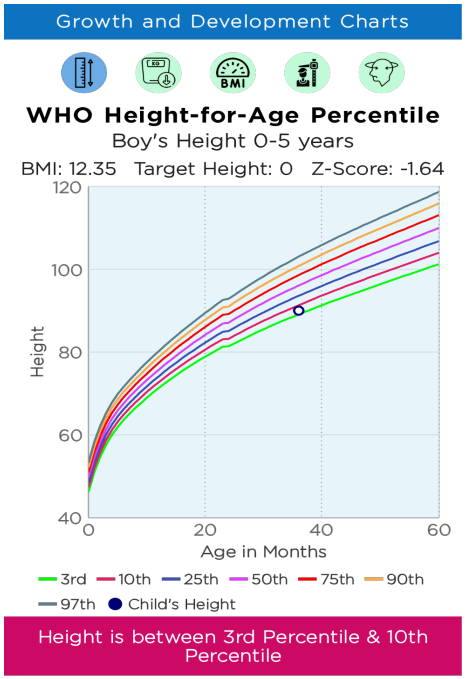
VITALS:

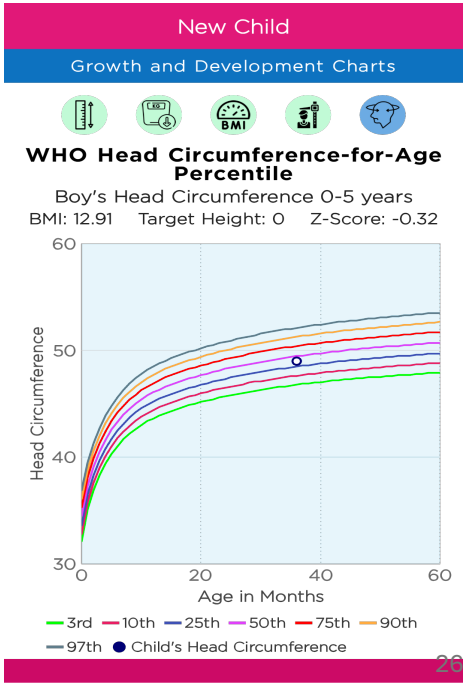
- Pulse: 96/min, regular, normal volume; no radio-femoral delay.
- Respiratory Rate: 28/min, regular, abdomino-thoracic breathing pattern.
- Blood Pressure: 88/60 mmHg (Right arm, supine with appropriate sized cuff).
- Temperature: Afebrile (37°C, in right axilla using a digital thermometer).
- SpO_2: 74% on room air (Right upper limb).
- CRT <3 seconds

Anthropometry

- Weight: 10 kg (<3rd percentile for age).
- Height: 90 cm (between 5th and 10th percentile).
- Head Circumference: 49 cm (normal for age).
- MUAC: 12.5 cm





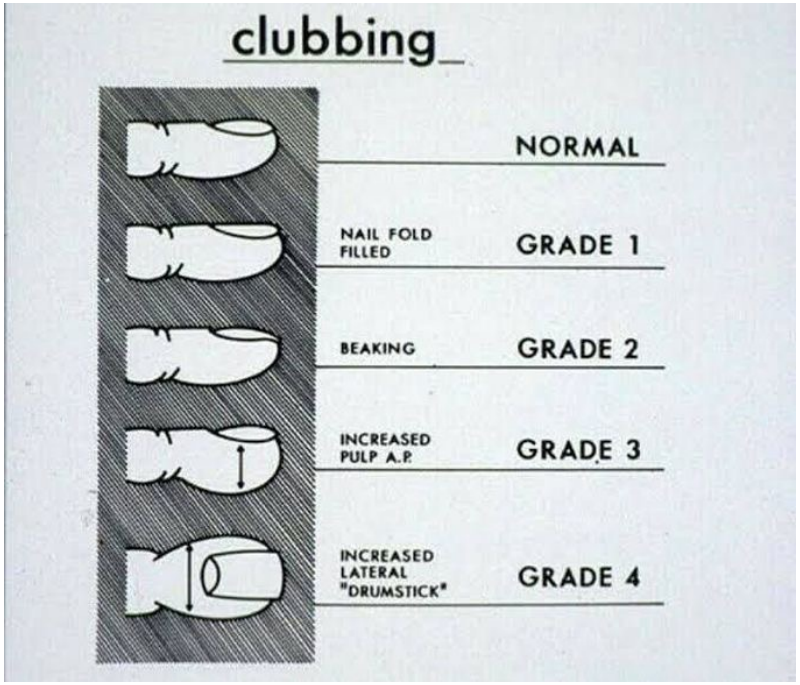


General Physical Examination

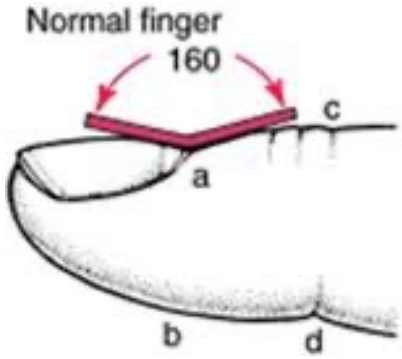
- Head & Face: Normal shape, no facial dysmorphism, eyes appear normal.

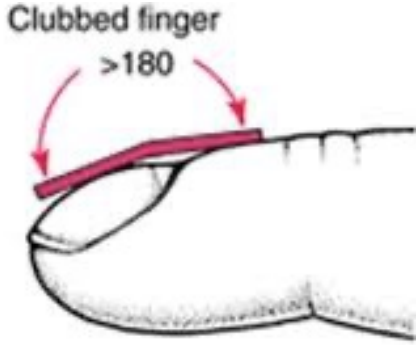
  Lips, tongue visibly bluish in colour.
- Eyes: suffused bulbar conjunctive. No icterus. No cataract.
- Mouth: Bluish discoloration of tongue, gums; good oral hygiene. No ulcers or dental caries noted. Dentition – 20.
- Hands & Feet: Clubbing present (grade II). Bluish discoloration noted on fingertips and nail beds.
- Lymph Nodes: No significant lymphadenopathy noted.
- Hydration Status: Adequate; no signs of dehydration.

GRADING

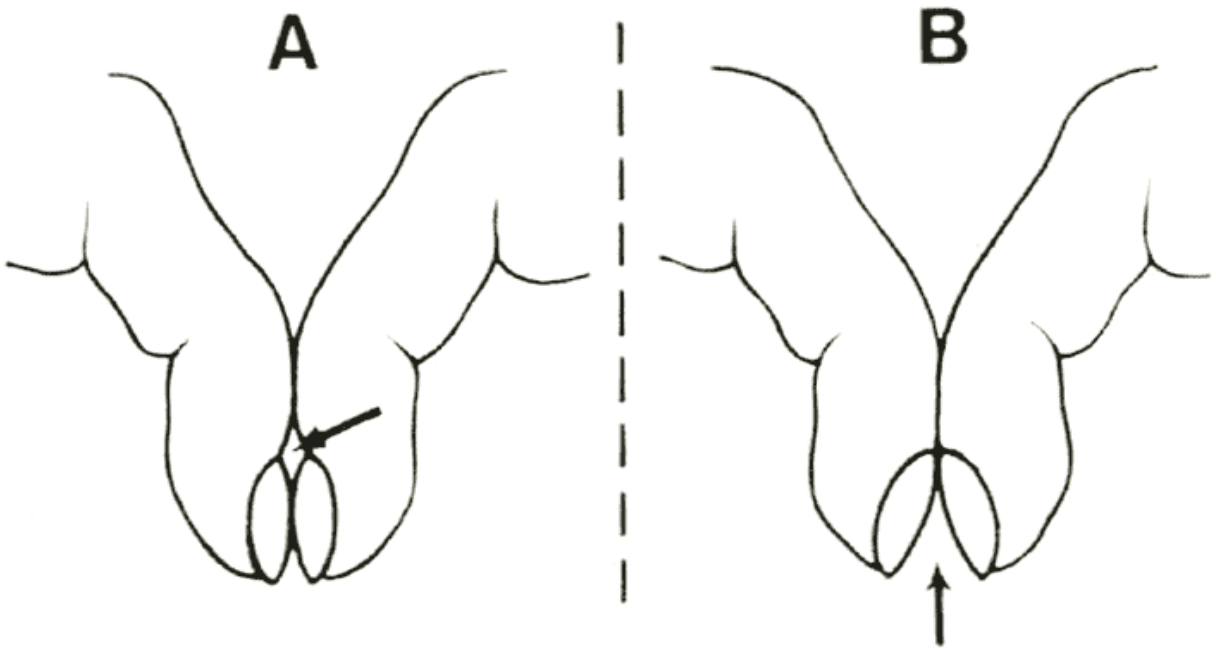


LOVIBOND ANGLE





SCHAMROTH'S SIGN



Systemic examination

CARDIOVASCULAR SYSTEM

Inspection	Palpation	Auscultation
• No precordial bulge/scar mark /venous prominence • No visible impulse over precordium	• **Apex beat** at left 4th intercoastal space, 1 cm inside mid clavicular line, diffused. • No left parasternal heave present. • Thrill absent. • **Epigastric pulsations** – absent.	• **Heart sounds:** S1 – Normal intensity, no split ➢ S2 – Single at left second intercostal space ➢ No additional sounds • **Murmurs:** Ejection systolic murmur at left 2nd intercostal space, grade 3/6, harsh, radiating towards back.

***REST ALL SYSTEMS WERE WITHIN NORMAL LIMITS**

Summary

3-year-old boy

Progressive cyanosis since 6 months of age,
History of cyanotic spell
Symptoms relieved by adopting the knee-chest position.

The child has poor weight gain and moderate undernutrition with chronic growth restriction.

Examination revealed central uniform cyanosis, digital clubbing, a diffuse apex beat (suggestive of right ventricular enlargement), a single S2, and an ejection systolic murmur at the left intercostal space radiating to the back.

DIAGNOSIS

Congenital Cyanotic Heart Disease with decrease pulmonary blood flow most likely Fallot's physiology:

- **Tetralogy of Fallot**
- **Tricuspid Atresia/VSD/PS**
- **DORV/VSD/PS**
- **TGA/VSD /PS**
- **Single ventricle /PS**
- **AVCD/PS**

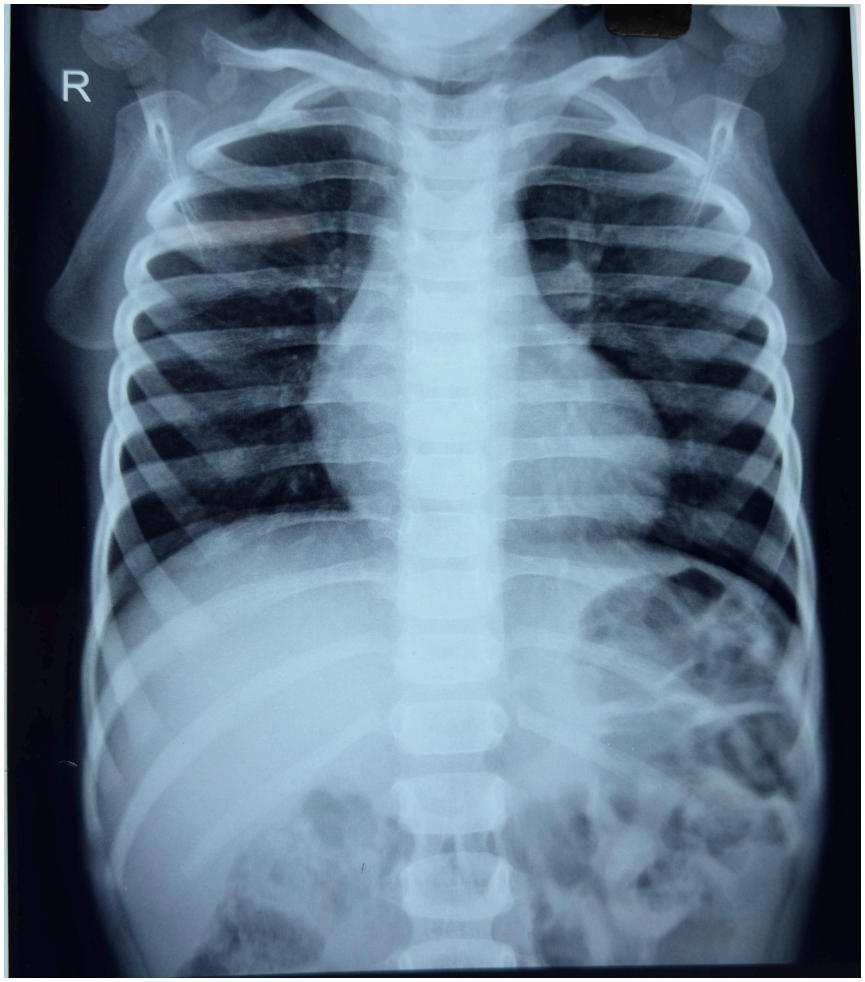
Investigations

Blood investigations

CBC	11/7/25
Hemoglobin	18 g/dl
HCT	56 %
MCV	75 fL
MCH	25 pg
MCHC	30.0 g/dl
RBC	6.80 X 10^6 /uL
WBC	9.60 X 10^3 /uL
Platelets	341 X 10^3 /uL

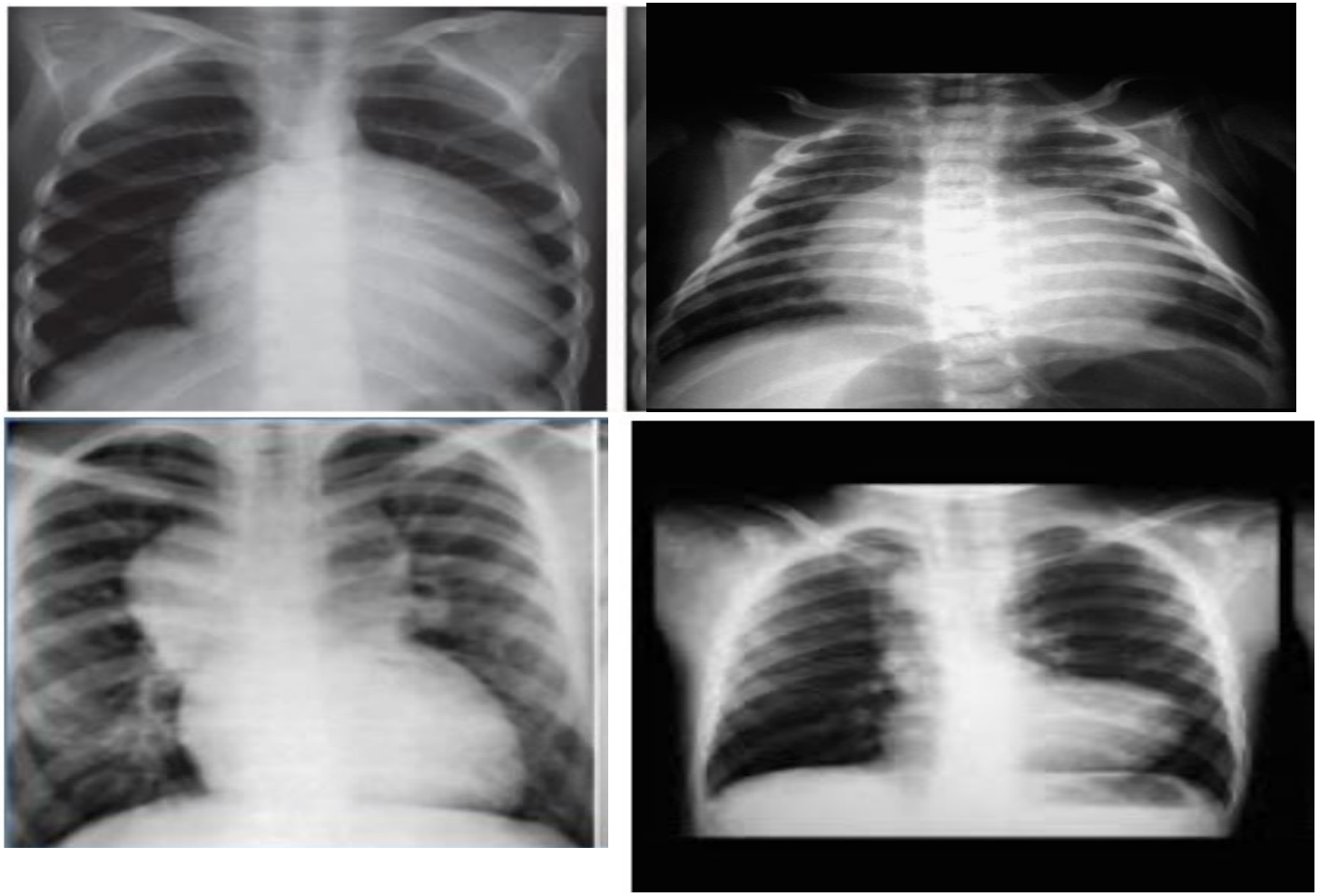
INVESTIGATION	11/7/25
Blood Urea Nitrogen	11.68 mg/dL
S. Creatinine	0.3 mg/dL

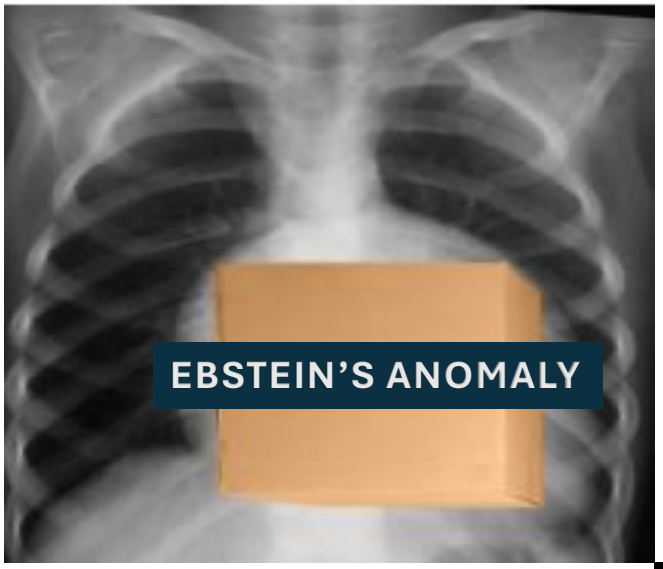
Radiological investigations

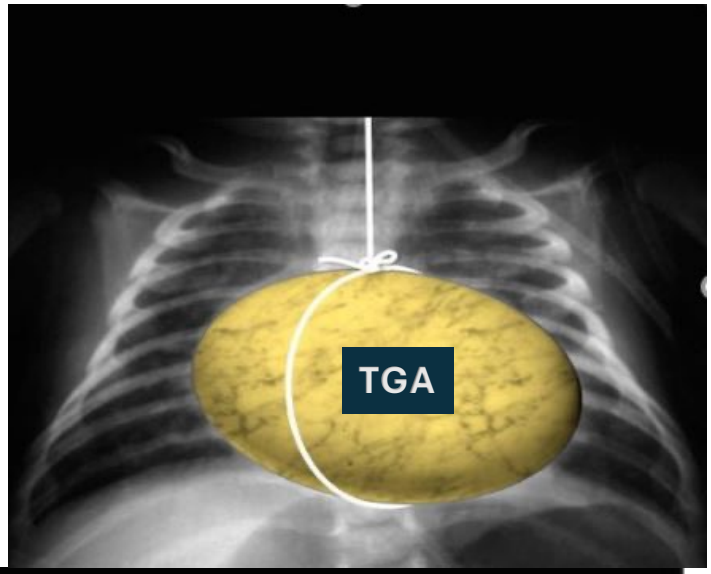


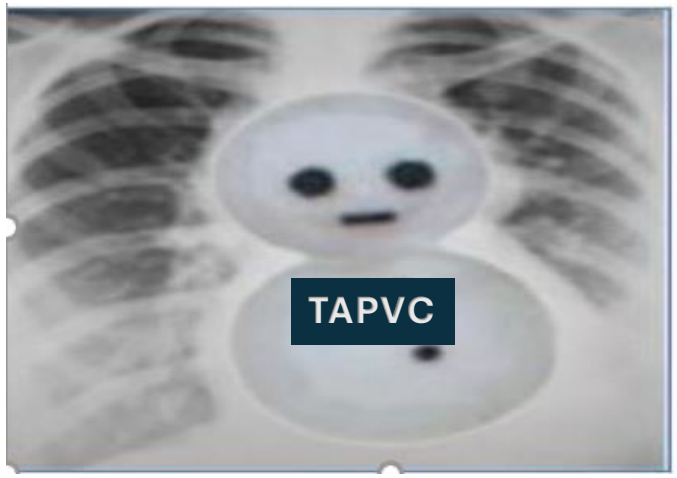
Classic chest X-Rays in CHDs

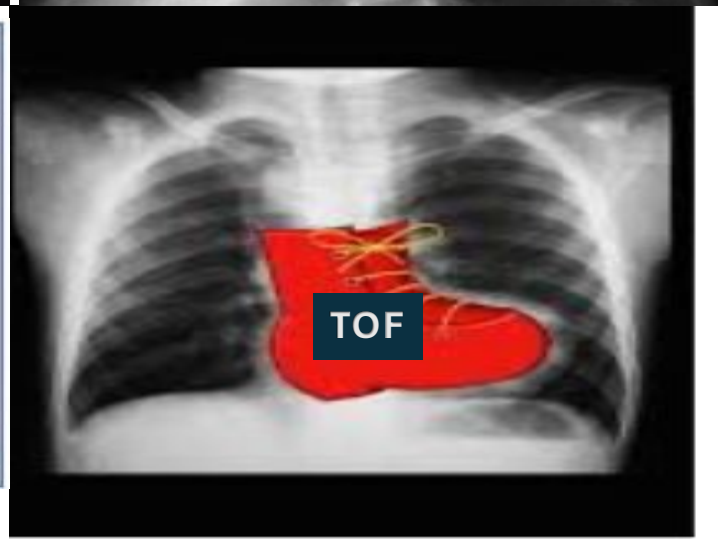
Chest X-Rays in CHDs

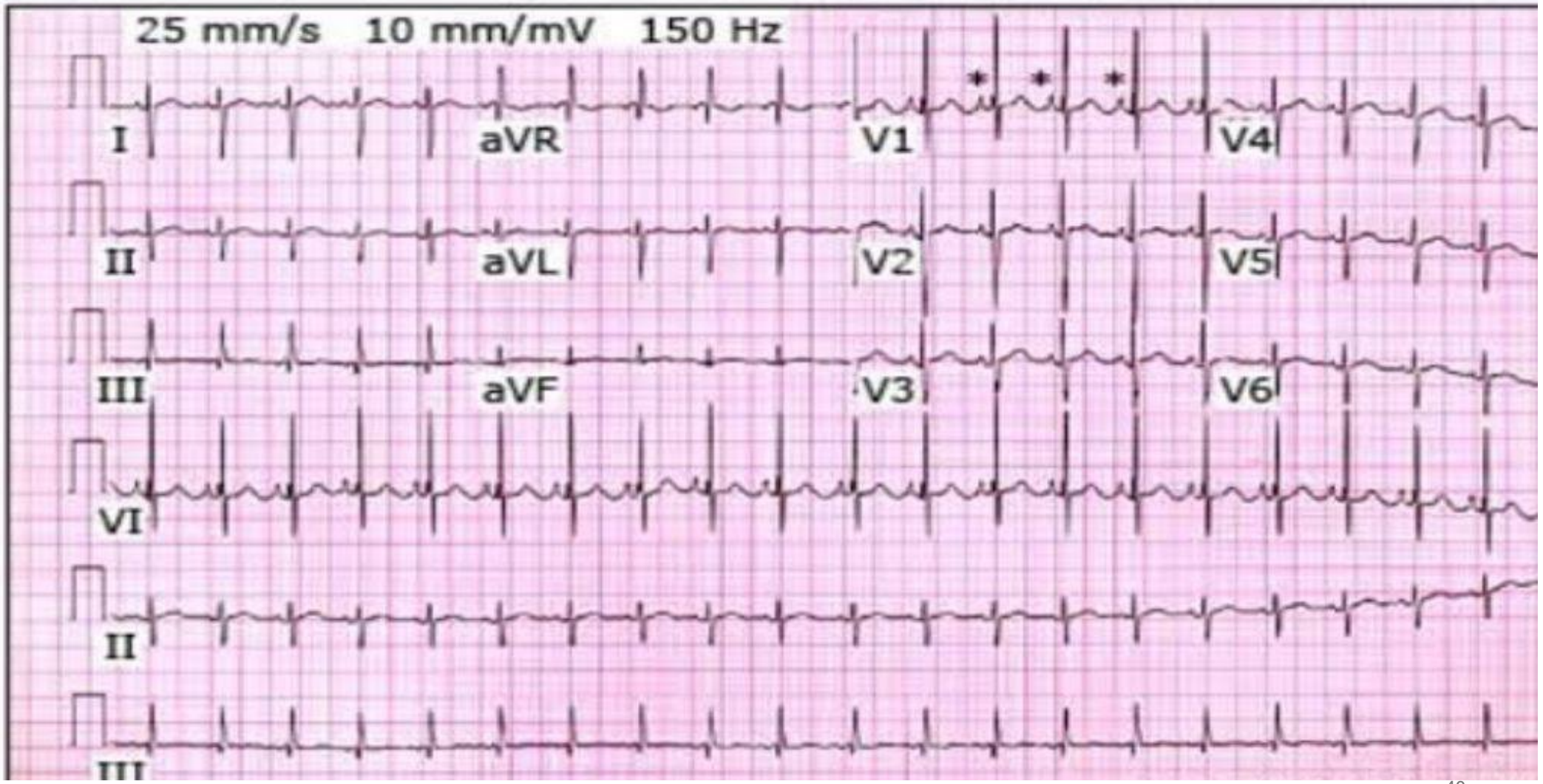


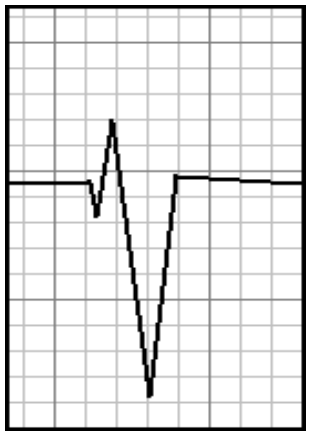

EBSTEIN'S ANOMALY


TGA


TAPVC


TOF


25 mm/s 10 mm/mV 150 Hz
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
VI
II
III



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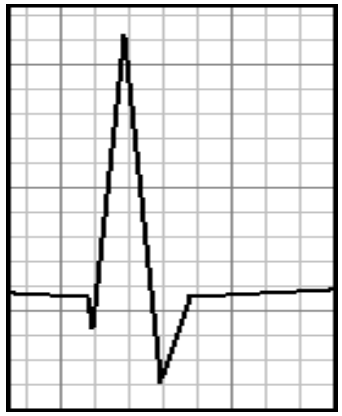
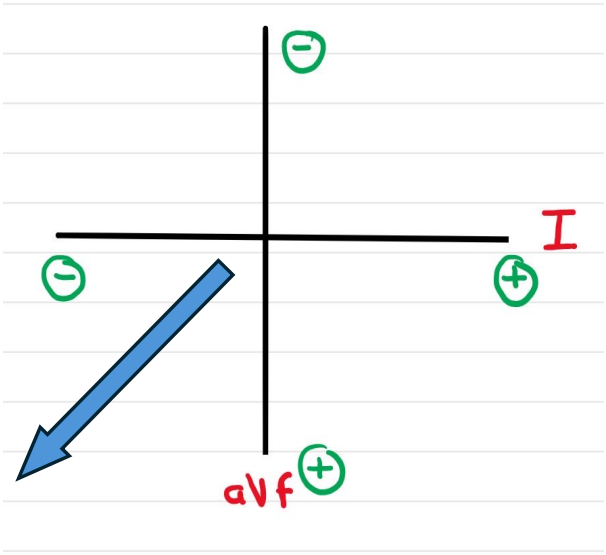


aVF



RAD

Tall R in V1

RVH

Sudden transition from V1 to V2

How can you differentiate Fallot physiology condition based on ECG alone?

TOF Physiology

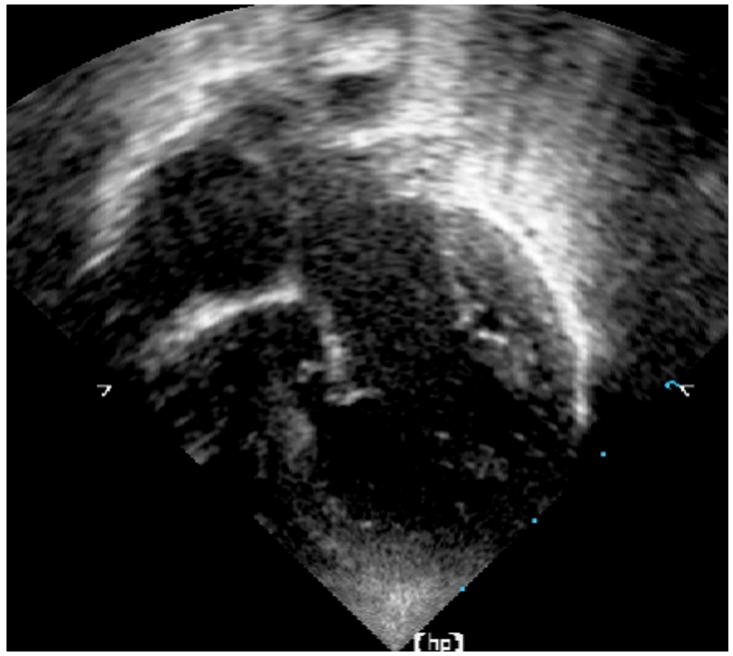
TOF

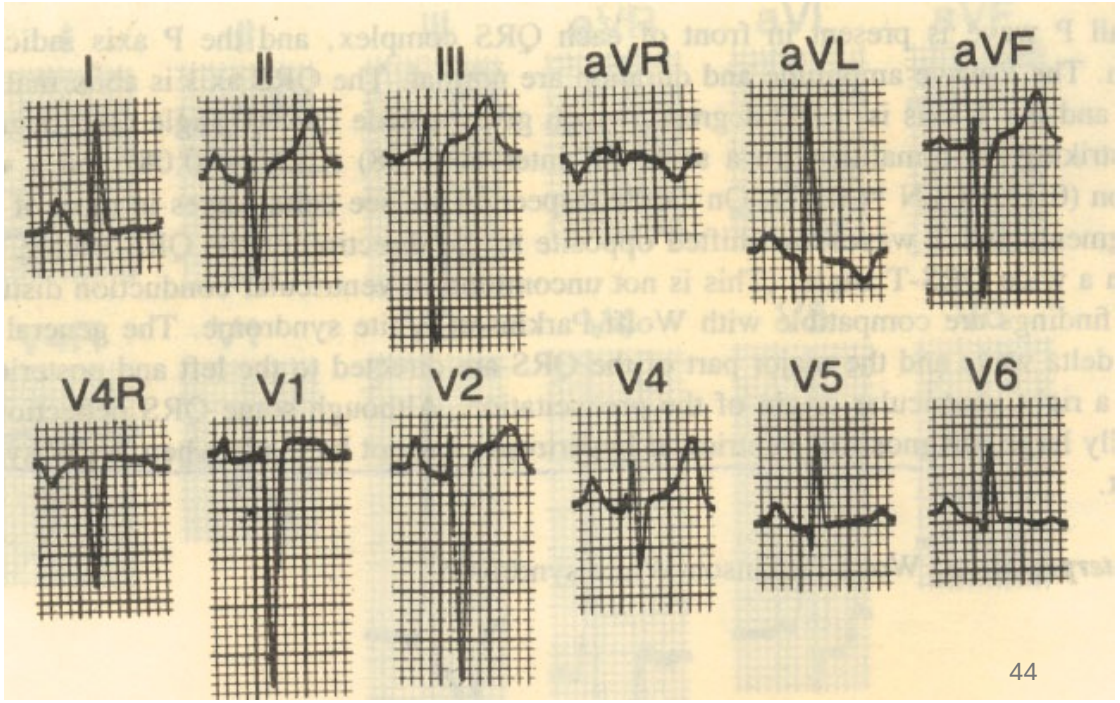
- **RAD upto +150**
- **RVH**
- **Abrupt change to rS pattern in V2**
- **Extreme Rt axis > 150**
  - **DORV / D-TGA ,VSD,PS**

TOF Physiology

Tricuspid atresia , PS

- LAD (- 30 to - 60)
- Rt atrial overload
- LVH

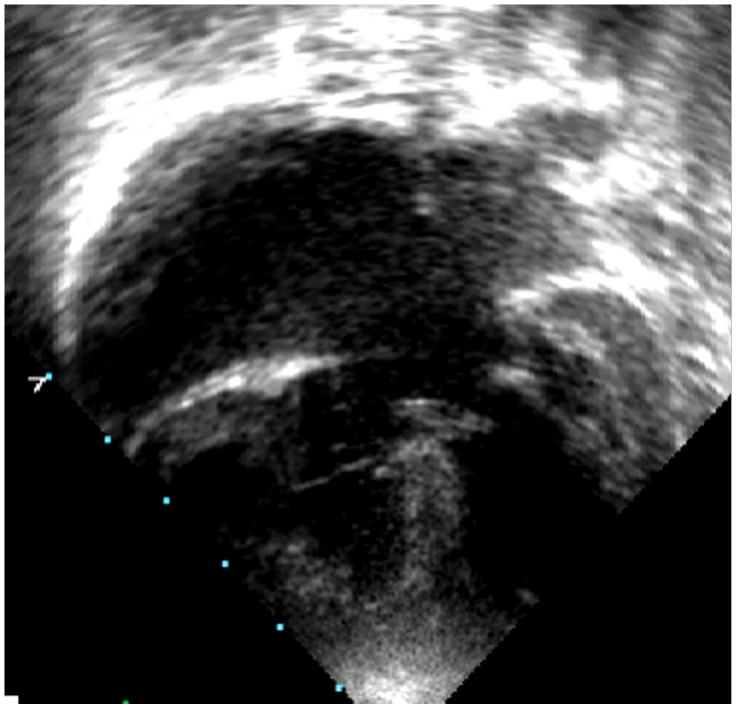


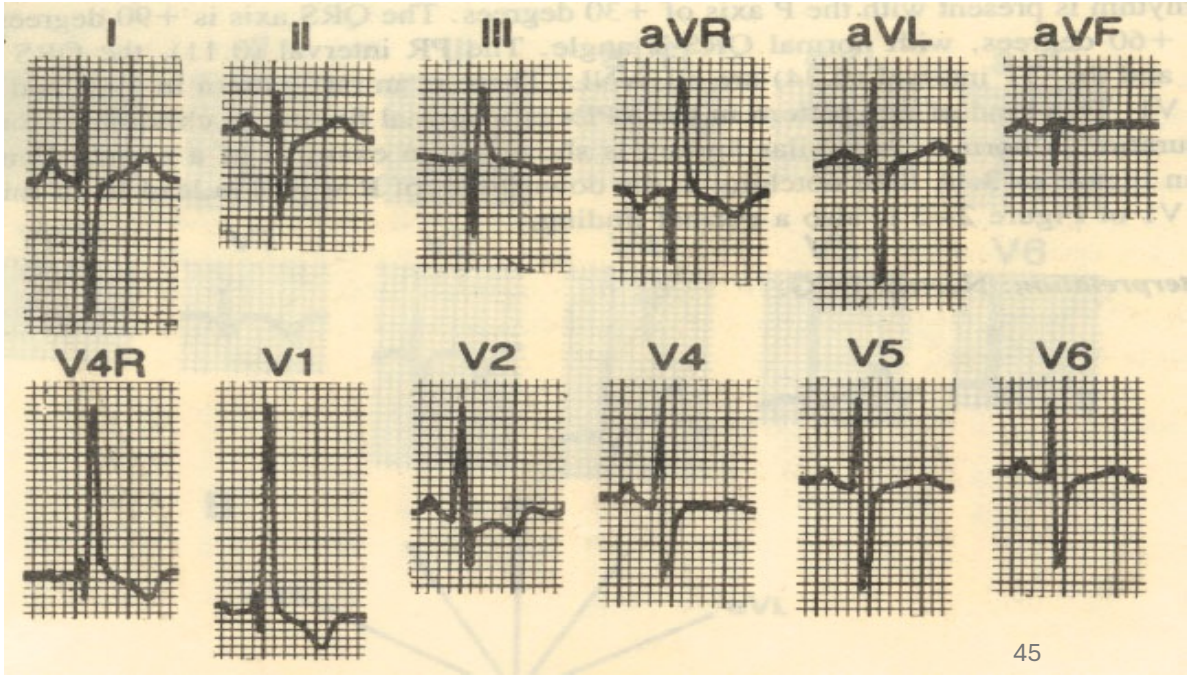


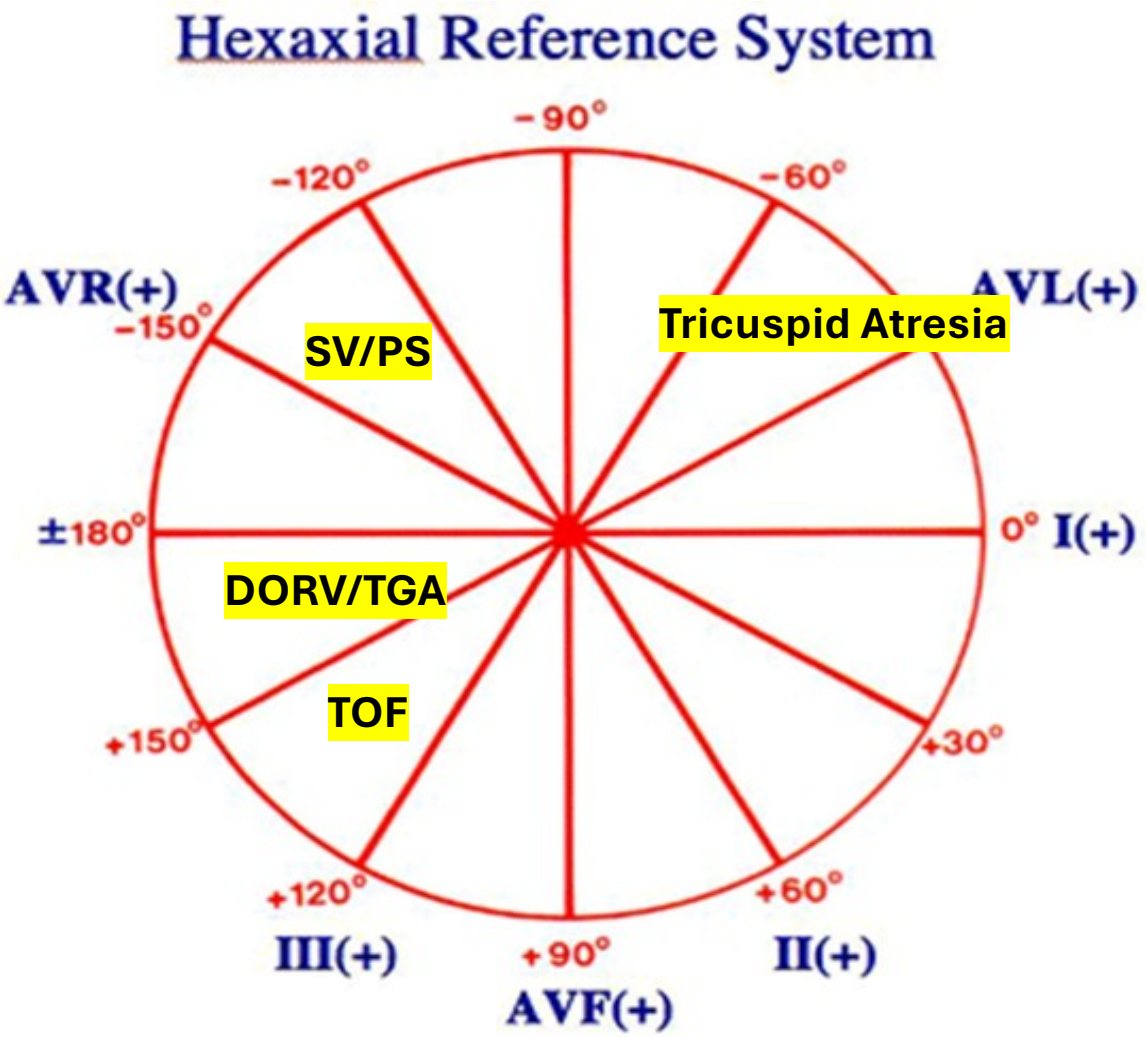
TOF Physiology

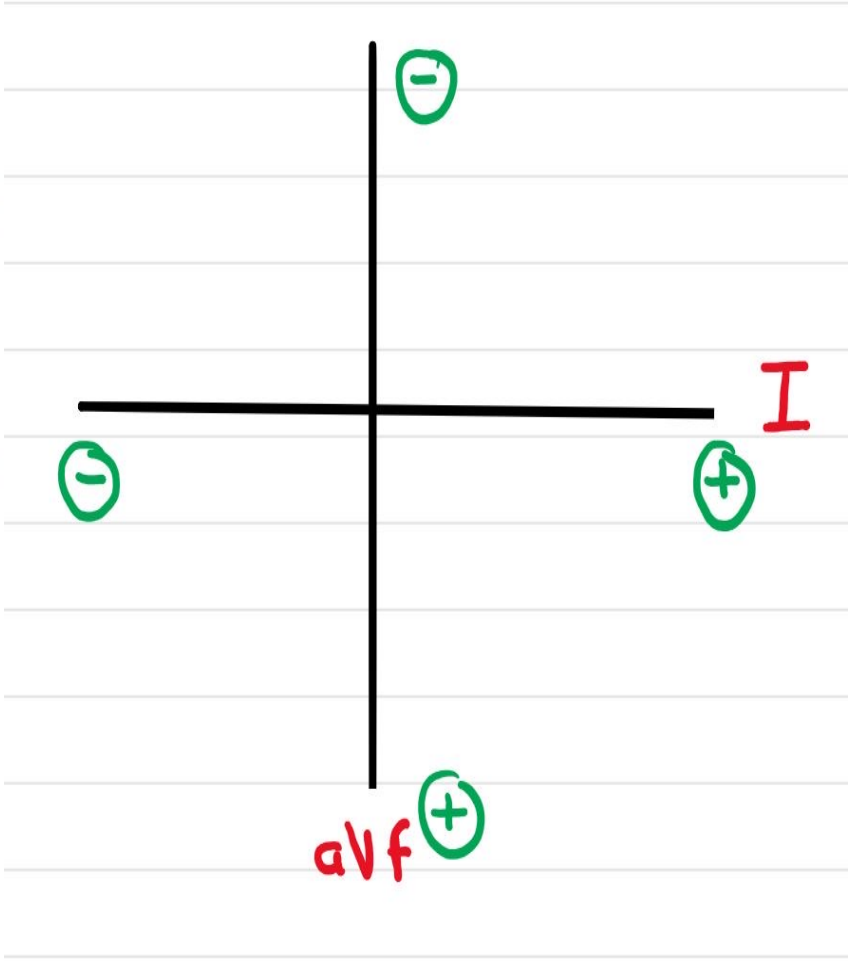
AVCD with PS

- **Extreme LAD,RVH**

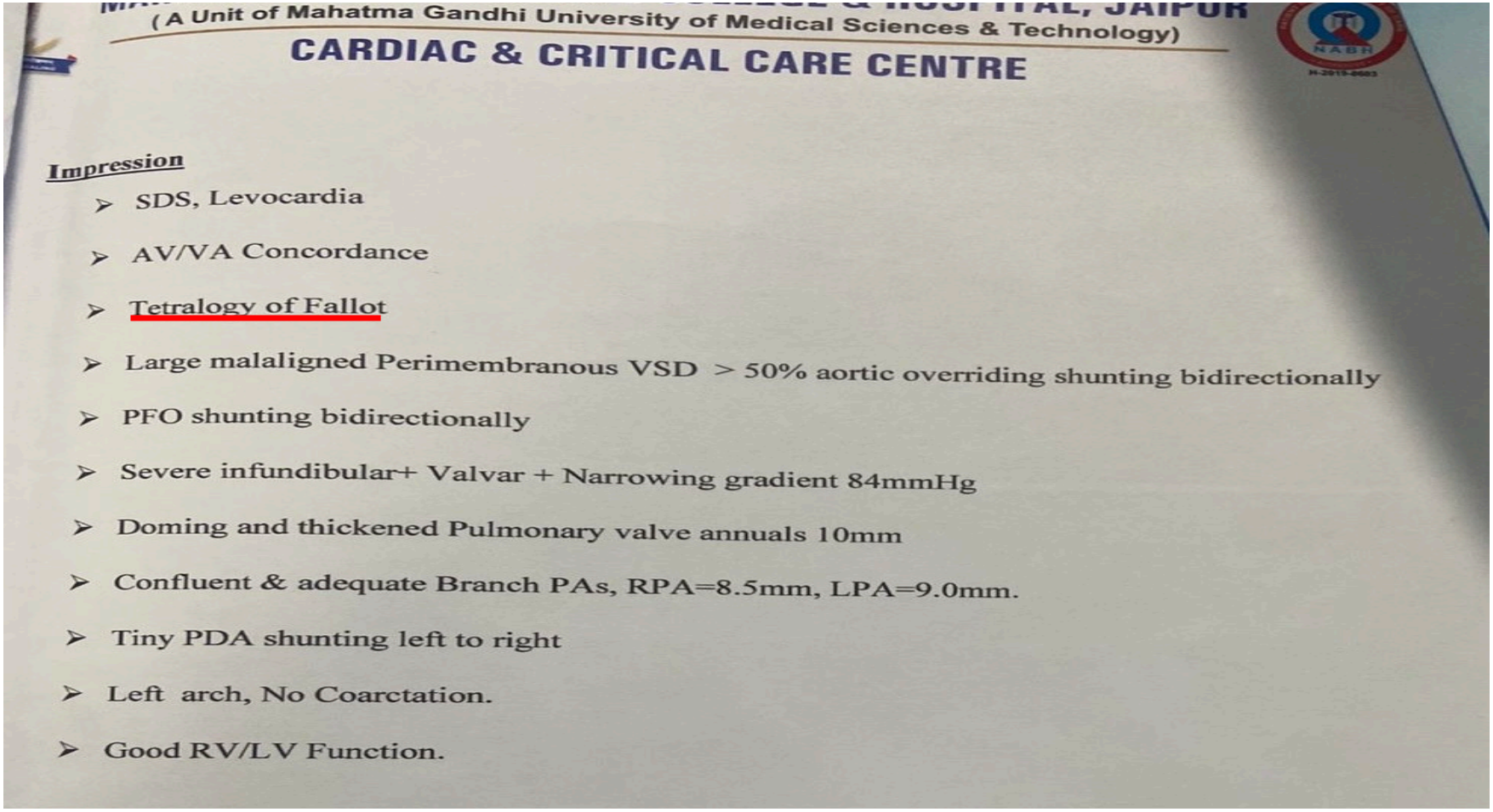





Hexaxial Reference System
-90°
-120°
-60°
AVR(+)
AVL(+)
-150°
SV/PS
Tricuspid Atresia
±180°
0° I(+)
DORV/TGA
TOF
+150°
+30°
+120°
+60°
III(+)
+90°
II(+)
AVF(+)


I
aVf

ECHO



(A Unit of Mahatma Gandhi University of Medical Sciences & Technology)

CARDIAC & CRITICAL CARE CENTRE

Impression

- SDS, Levocardia
- AV/VA Concordance
- Tetralogy of Fallot
- Large malaligned Perimembranous VSD > 50% aortic overriding shunting bidirectionally
- PFO shunting bidirectionally
- Severe infundibular+ Valvar + Narrowing gradient 84mmHg
- Doming and thickened Pulmonary valve annuals 10mm
- Confluent & adequate Branch PAs, RPA=8.5mm, LPA=9.0mm.
- Tiny PDA shunting left to right
- Left arch, No Coarctation.
- Good RV/LV Function.

FINAL DIAGNOSIS

- **Congenital Cyanotic Heart Disease with decreased pulmonary blood flow – Tetralogy of Fallot**
- **In sinus Rhythm, not in CHF**
- **No sign of infective endocarditis with chronic growth restriction**

MANAGEMENT

TREATMENT

Tet Spells (Acute)

1. **Non-pharmacological**
- Knee-to-chest/squatting
- Calm the patient
- Administer oxygen

2. **Fluids/Electrolytes**

3. **Pharmacological**
- Morphine(0.1mg/kg SC/IM)
- Metoprolol(0.1mg/kg IV or Infusion)
- Ketamine(0.25mg-1mg/kg IV/IM)
- Phenylephrine(5mcg/kg bolus, 1-4mcg/kg/min infusion)

Indications for surgery

1. Documented cyanosis (O_2 saturation <80%)
2. Recurrent cyanotic spells

Surgical timing

1. At 6 months if stable
2. Earlier if symptoms/ severe obstruction

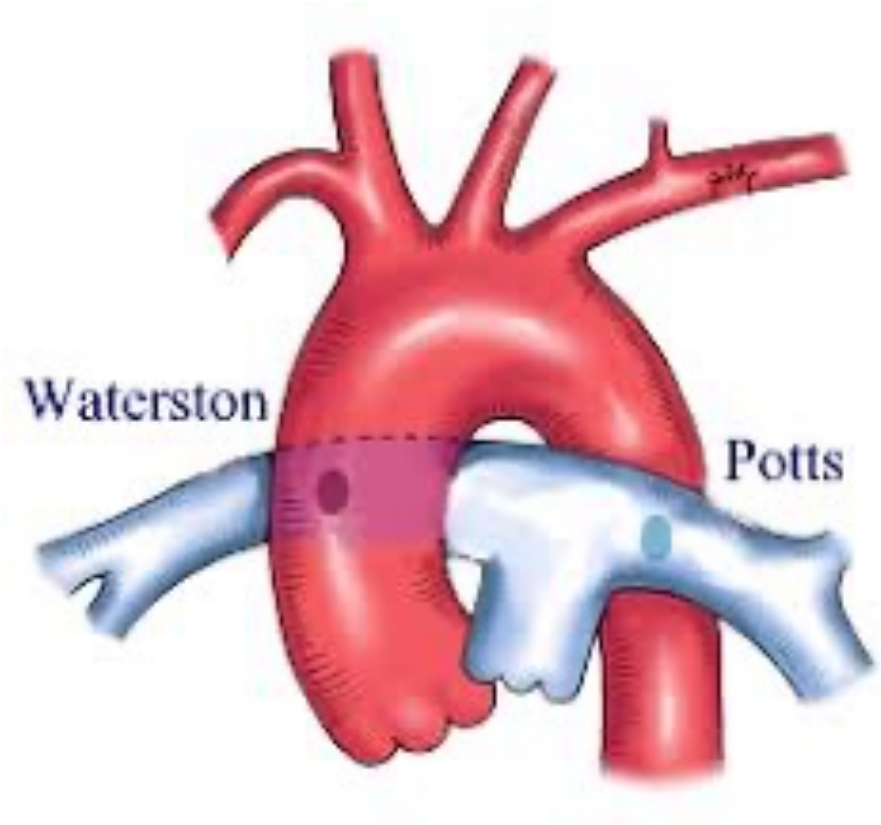
Post-op follow-up

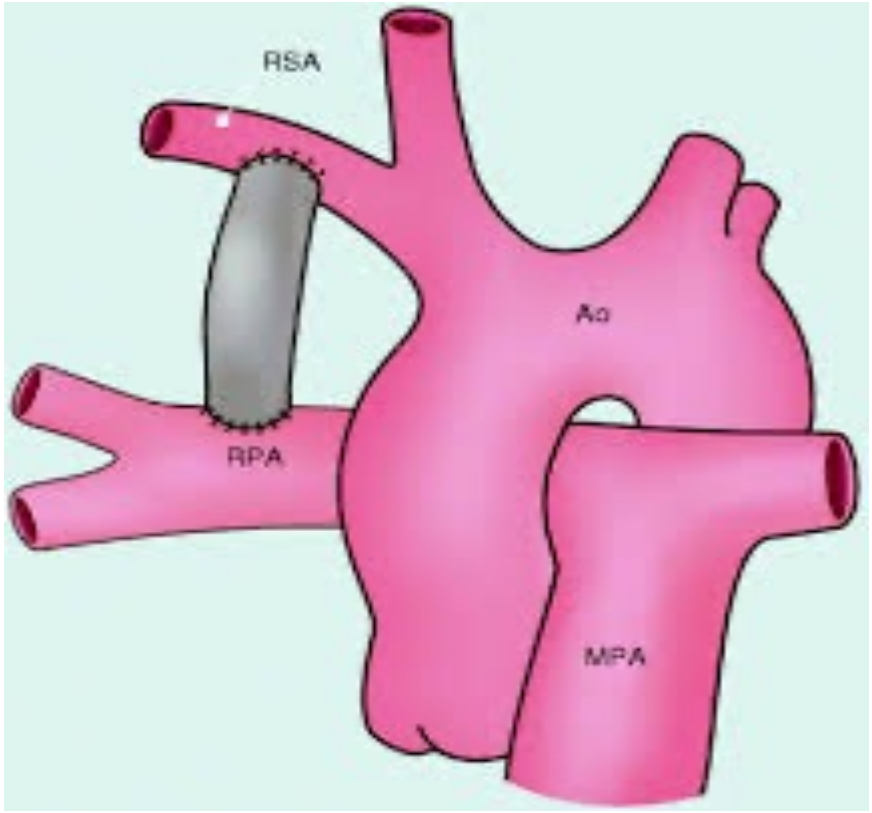
Lifelong surveillance ECHO,ECG,MRI, Exercise training, re-interventions as needed

Surgical Management

- **Palliative**

- **Corrective**


Waterston
Potts


RSA
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Thank You


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