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Pulmonary Hypertension Specialist

Learning outcomes

Define	• Define what is cardiac versus non cardiac syncope.
Identify	• Identify what are the etiology of cardiac syncope in children.
Distinguish	• Distinguish cardiac from non-cardiac-sounding syncope.
Distinguish	• Distinguish innocent versus pathological murmurs in children and develop an assessment strategy.

Definition

A sudden, brief loss of consciousness associated with loss of postural tone from which there is spontaneous recovery.

Syncope = a symptom

Syncope has 3 components:

1. Loss of consciousness

2. Loss of postural tone

3. Recovery and return to baseline

Key questions

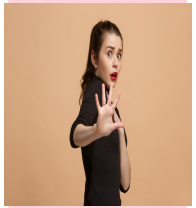
Is it syncope? Or could it be something else?

Is the underlying cause serious or life threatening ? (i.e. cardiac dysrhythmia, MI, GI bleed, Pulmonary embolism)

Is the child at high risk for a cardiovascular event, or death?

Types of syncope

Vasovagal



Situational

Cardiovascular

Orthostatic

Aetiology of syncope

Autonomic	Cardiac	Other
• Vasovagal syncope (also called neurocardiogenic) • Orthostatic hypotension • Postural orthostatic tachycardia syndrome • Breath-holding spells	• Brady/tachyarrhythmia • Long QT syndrome • Brugada syndrome • Wolff-Parkinson-White syndrome • Structural abnormalities (e.g. aortic stenosis, hypertrophic cardiomyopathy)	Functional disorder Hypoglycaemia Seizure Migraine Anaemia Narcolepsy Toxic exposure (e.g. carbon monoxide, clonidine)

Vasovagal syncope

- prodrome of nausea,
- presyncope & sweating;
- triggered by pain, anxiety, distress, or
- by prolonged standing or kneeling in a warm and/or crowded place

Situational syncope

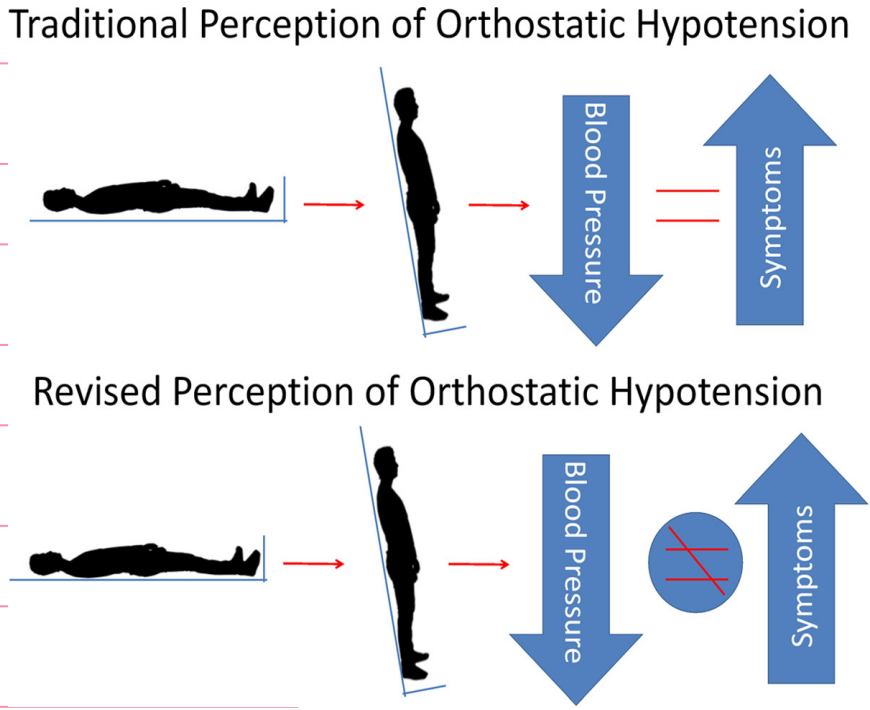
occurs with micturition, cough, defecation; or by carotid sinus pressure

(i.e. turning head, shaving), or

subclavian steal (i.e. during arm exercises)

Orthostatic syncope

- Triggered by BP meds,
- Dehydration, alcohol, &
- Autonomic dysfunction
- CNS: rarely the cause of syncope as the only symptom



Cardiovascular syncope

a. dysrhythmia, structural heart disease or ischemia;
b. clues are family history of sudden/unexplained death,
c. exertional syncope and
d. no prodrome

RED FLAGS:

- lack of prodrome
- palpitations or chest pain
- exercise-induced syncope
- past cardiac history
- family history of early cardiac death, arrhythmia or sudden death

3 arm approach to syncope

3 Arm approach to syncope work up

- History
- Vital signs and Examination
- Additional tests – ECG, Chest X-Ray and Echocardiogram

Physical Exam Pearls

- **Look for injuries:** serious head injuries are possible. Also look for evidence of seizure (bitten tongue, cheeks, multiple bruises).
- **Measure orthostatic vitals**
  - They have higher yield for older pts (1) who may have orthostatic syncope, however, abnormal findings do not rule out other causes of syncope.
- **Do a careful cardiac exam**
  - auscultate for valvular murmurs (i.e. mitral or aortic valve disease).
  - **Any outflow murmur that increases with Valsalva in syncope is hypertrophic cardiomyopathy (HCM) until proven otherwise!**
  - **If you can feel the Second heart sound – Pulmonary Hypertension**

Case 1

- 8-year-old boy was playing in the pool with his siblings
- Whilst playing he was walking from one end of the pool to the other and the then got a fright after he missed his footing in the pool and passed out in the pool
- His sisters pull him to the side of the pool and his father performs CPR for 3 mins before there is a return of circulation

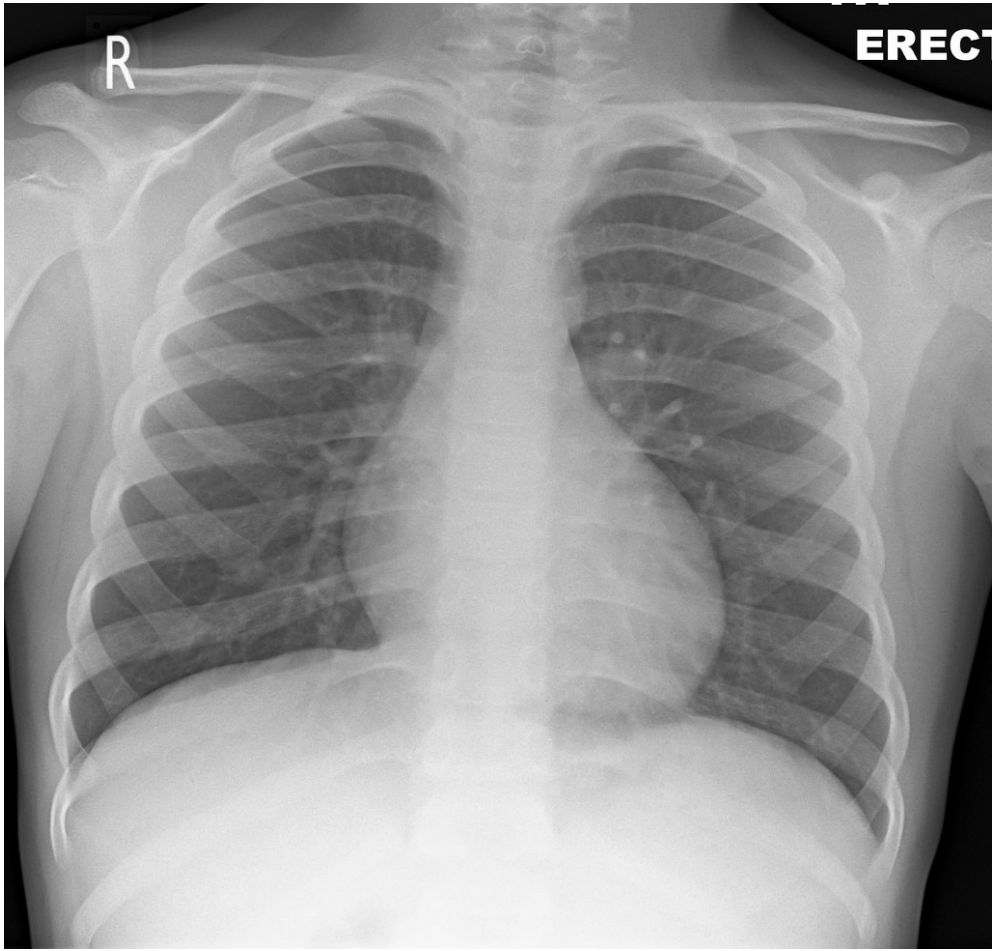
- What is your differential diagnosis?

Examination

- Weight 25kg, well in himself
- Normal S1 and S2, no murmurs
- Vesicular breath sounds
- No hepatomegaly, normal pulses

- Investigations including EUC, FBC, LFT, Ca/Mg/PO normal

- What test would you like to do Next?


R
EREC

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Vent. Rate	74 bpm
PR Interval	174 ms
QRS Duration	76 ms
QT/QTc Interval	448/473 ms
P/QRS/T Axes	52/85/64 deg
QTc:Hodges	

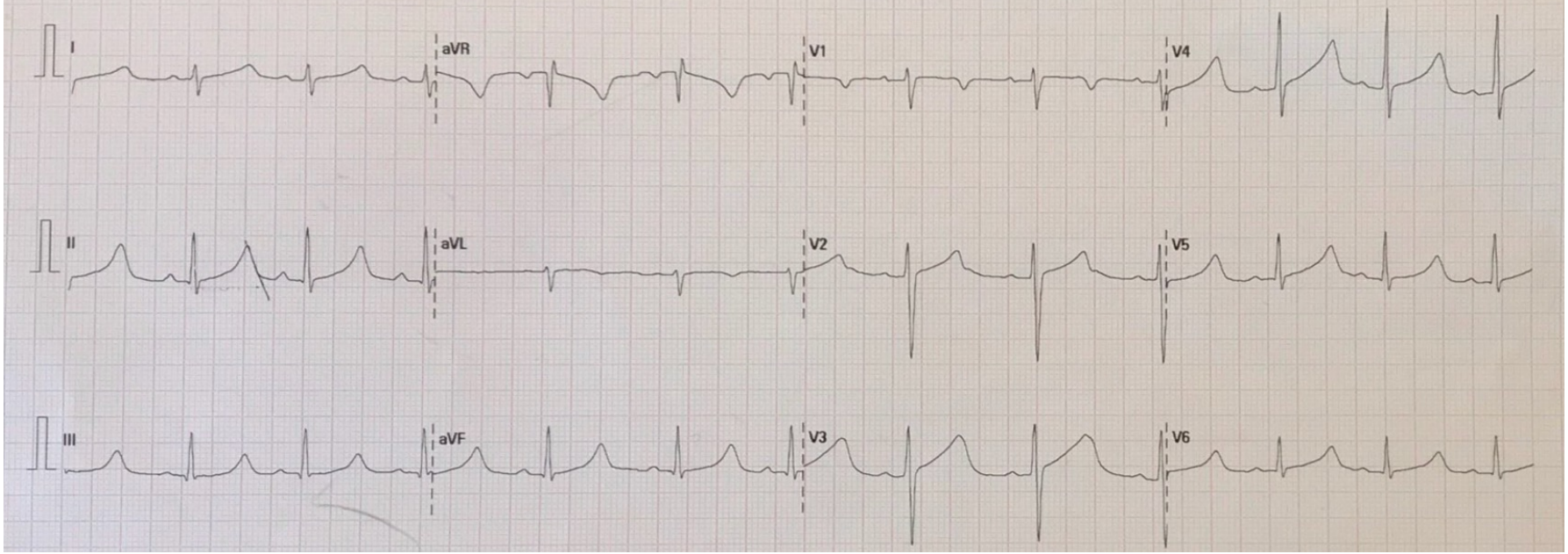
--- Interpretation based on pediatric criteria ---
Sinus rhythm with borderline 1st degree A-V block
Borderline prolonged QT interval

Borderline ECG

Unconfirmed Diagnosis

HR 75.

QTc 492
0.53.



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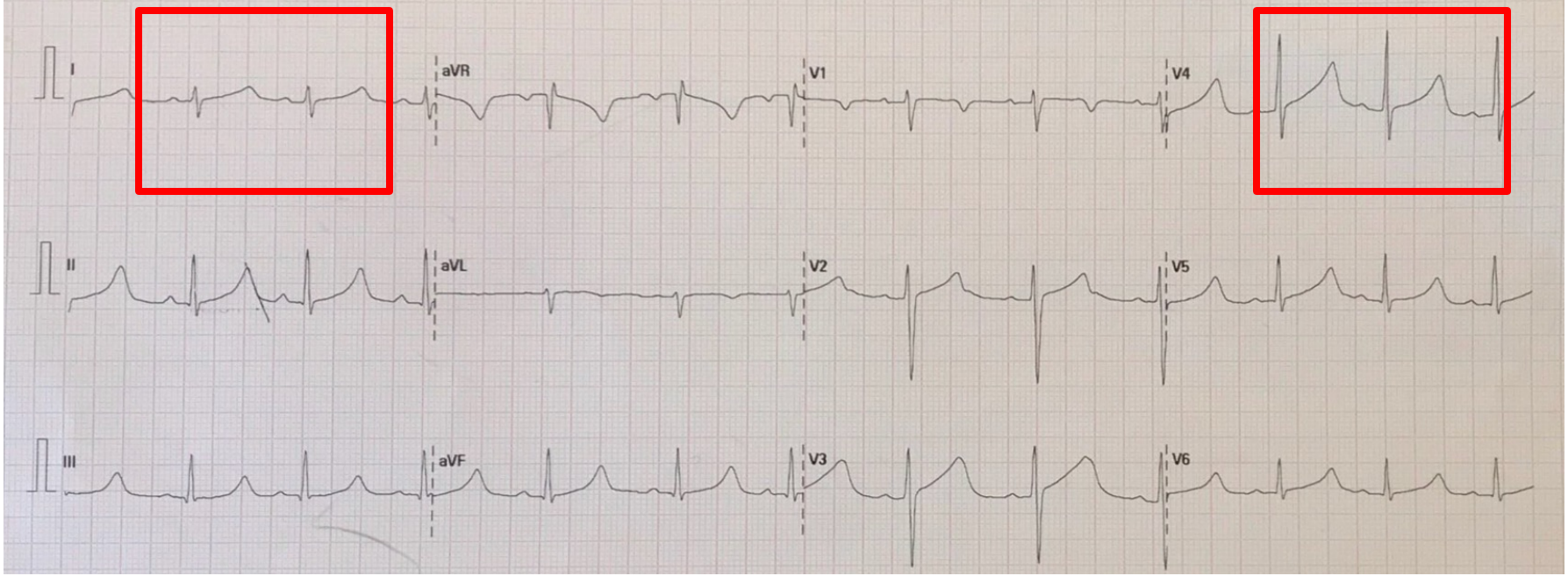


CLUES Family history red flags suggestive of cardiac disease

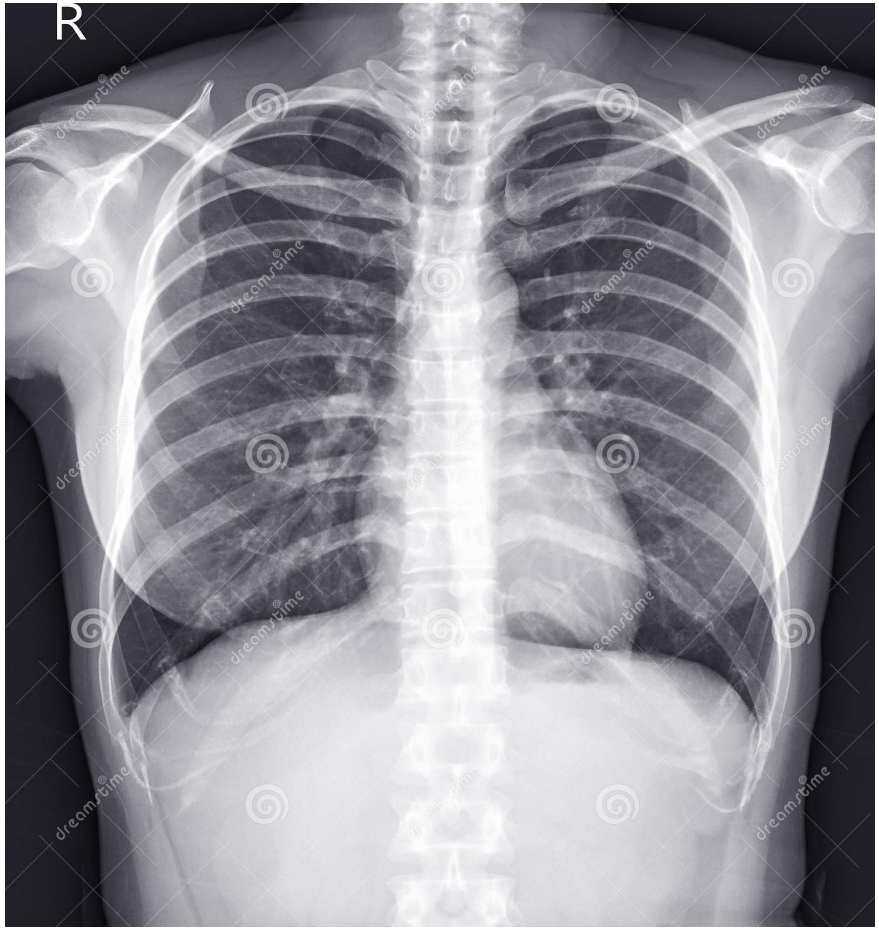
- Sudden cardiac death under the age of 50
- Any unexplained death
- Pacemakers/implanted defibrillators
- Cardiomyopathy
- Prolonged QT syndrome
- Congenital deafness
- Congenital heart disease

Case 2

- 15-year-old female athlete
- Recently had knee injury during gymnastics and recovering from a recent viral illness
- Presents to the emergency department with 12-hour history of chest pain
- This is her third presentation with chest pain and she could not sleep secondary to the chest pain so she presented to emergency
- The first episode of chest pain started following her meningococcal vaccine, when she complained of 7/10 chest discomfort, relieved by Omeprazole
    - Associated with dyspnoea in the morning
    - Palpitations occasionally
    - Pain relieved by Nurofen and Panadol
    - No syncope and she has returned to playing water polo

Examination

- Normal S1 + S2 with no murmurs
- Vesicular breath sounds bilaterally
- No hepatomegaly
- JVP not elevated
- No peripheral oedema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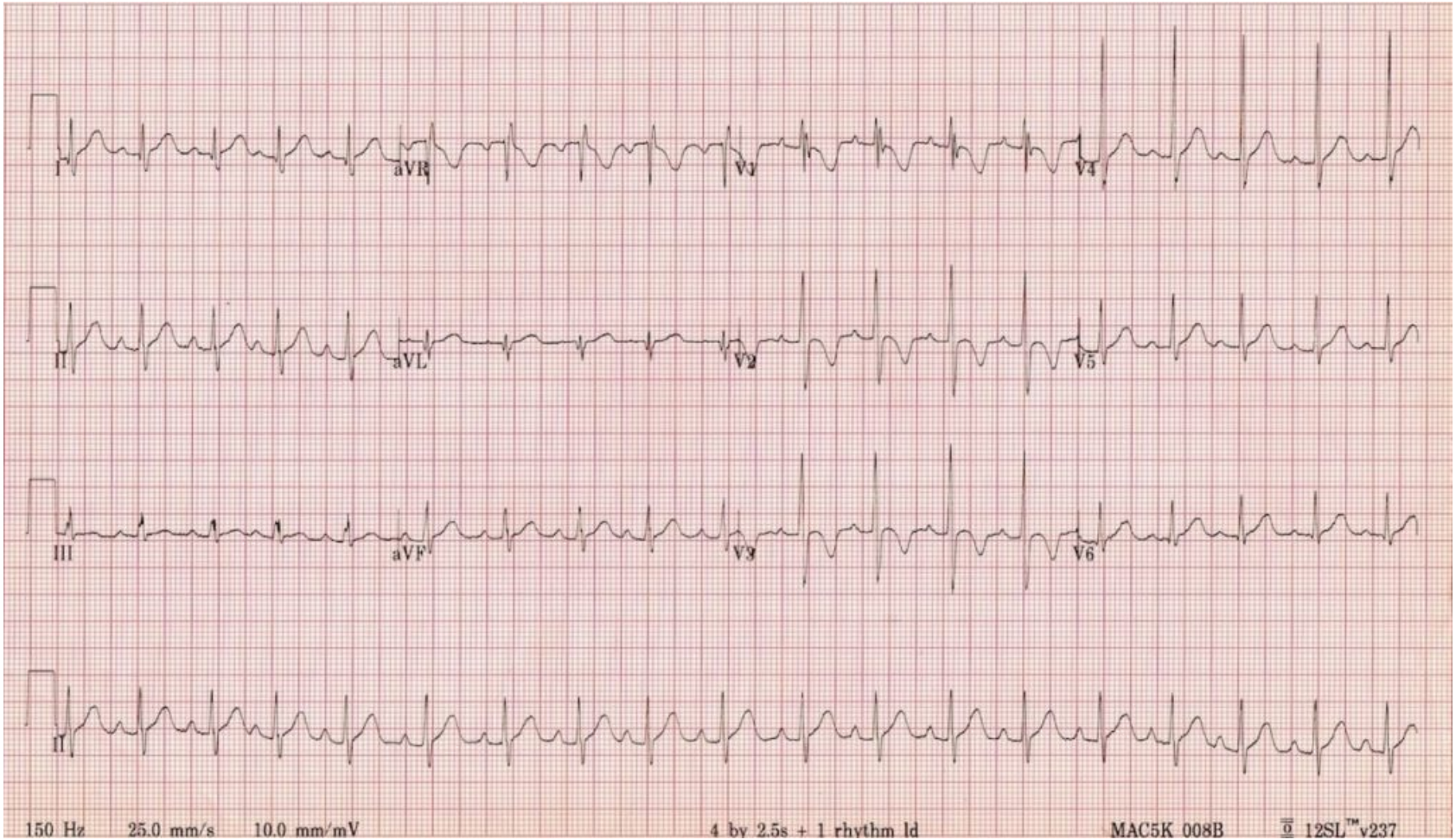

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
150 Hz
25.0 mm/s
10.0 mm/mV
4 by 2.5s + 1 rhythm ld
MAC5K 008B
12SL™v237

‘RED’ signs and symptoms	Consider…
Post cardiac surgery < 2 weeks	• Pericarditis/Post pericardiotomy syndrome • Pericardial or pleural effusion • Repair site complication • Infection/endocarditis • Pneumothorax
Connective tissue disorder eg. Marfan’s disease	• Dissection or rupture • Mitral valve prolapse
Known Kawasaki patients with known coronary pathology	• Coronary artery thrombus
Family history of Cardiomyopathy or conduction disorders (first degree relative)	• Risk factor for sudden death

‘AMBER’ Signs & Symptoms	Consider…
Chest pain associated with exercise	**Left heart obstruction or myocardial ischaemia**
Radiation to the jaw or left arm	**Coronary ischaemia**
Radiation to the left shoulder tip	**Pericarditis**
Associated with palpitations	**Pathological arrhythmia**
Associated with syncope	**Left heart obstruction or arrhythmia**

‘GREEN’ Signs & Symptoms	Possible causes
Chronic pain	Unlikely to be cardiac
Superficial tenderness, worse on movement	Musculoskeletal
Associated with cough or bronchospasm	Respiratory
Associated with eating or posture	Gastrointestinal
Anxiety and hyperventilation	Psychogenic

Case #3

- A 9-year-old boy had an out-of-hospital cardiac arrest at flip-out.
- He bumped his head at flip out and then started feeling a ‘fast heart rate.’
- He got to the bottom of the slide, and as he was walking, he collapsed.

- A bystander gave him 4 cycles of chest compressions. An ambulance was called. He had started to froth around the mouth, and then he became more alert and his colour returned to his face. His eyes deviated to the side, and by the time ambulance arrived, his Glasgow Coma Scale was 15. His initial Blood sugar was 4.0, and he was taken to the Hospital.

- What further information would you like to know?

Further information

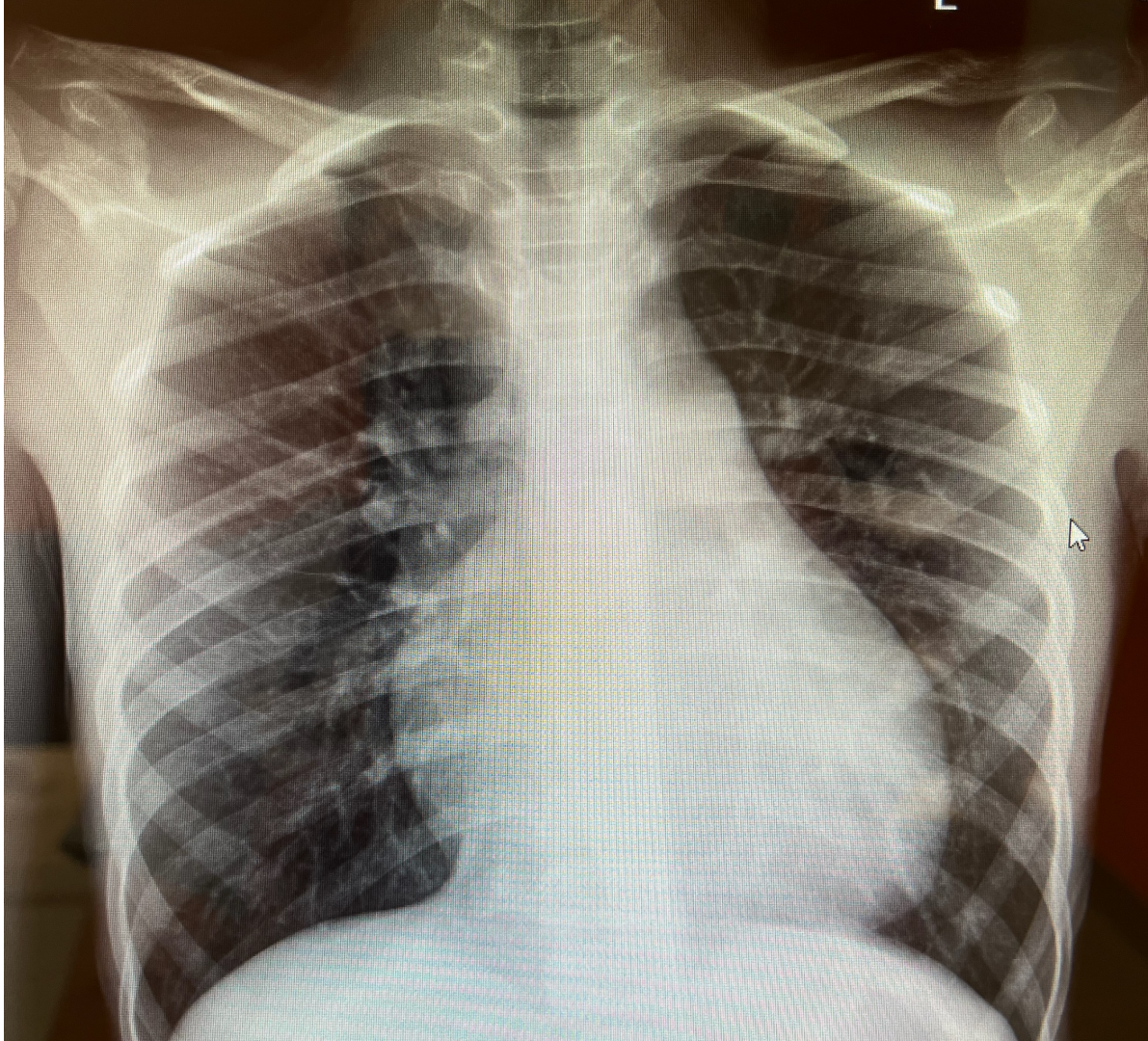
- No family history of cardiac disease. No personal history of seizures.
- No premature deaths or drownings.
- However, he has had a 2-year history of chest pain with exertion, dizziness and palpitations.

- What would you like to do next?

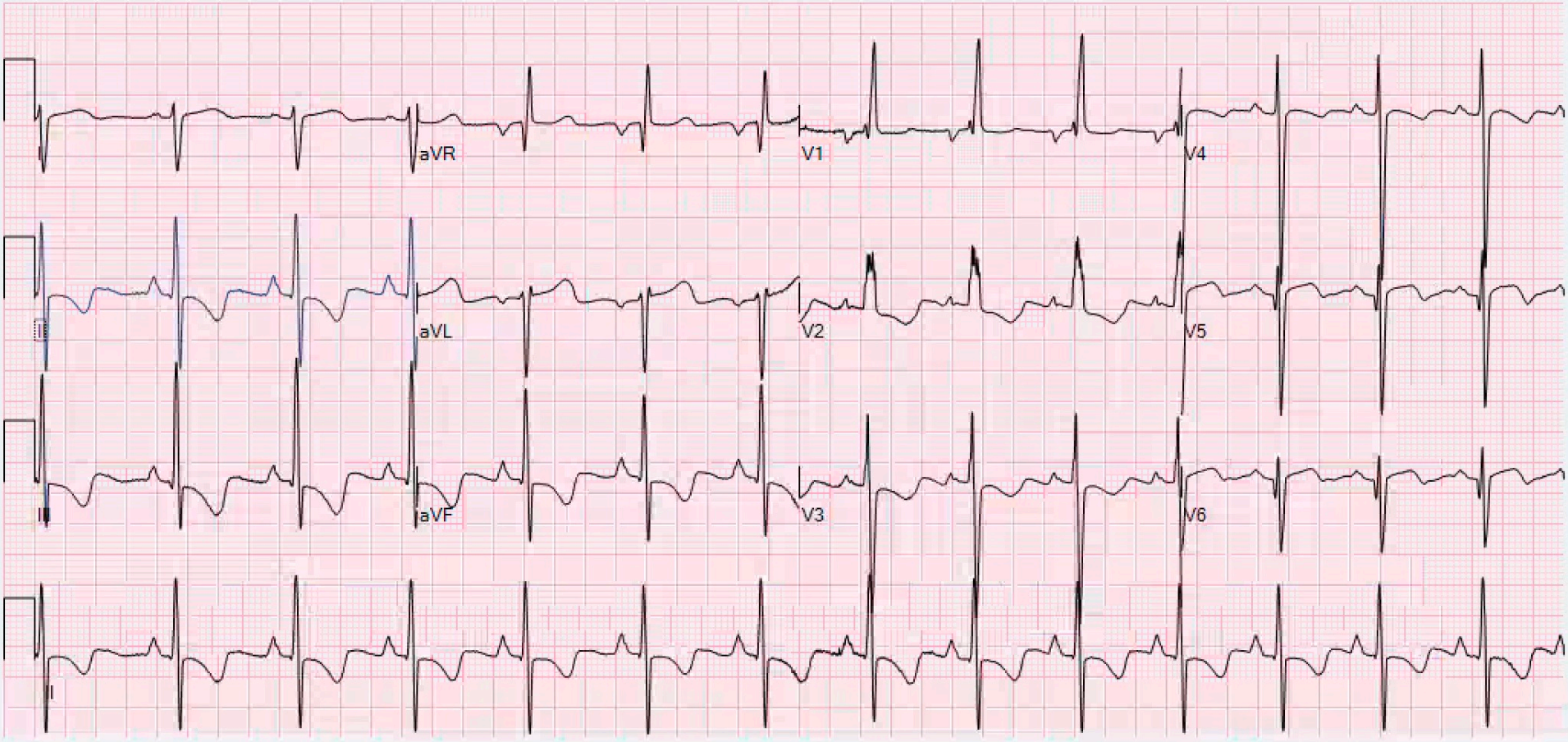
Examination

- Well, alert, GCS 15
- Oxygen saturation 99% in room air
- Systolic BP 90mmHg
- Normal S1 and Loud second heart sound
- Normal air entry bilaterally
- No peripheral oedema

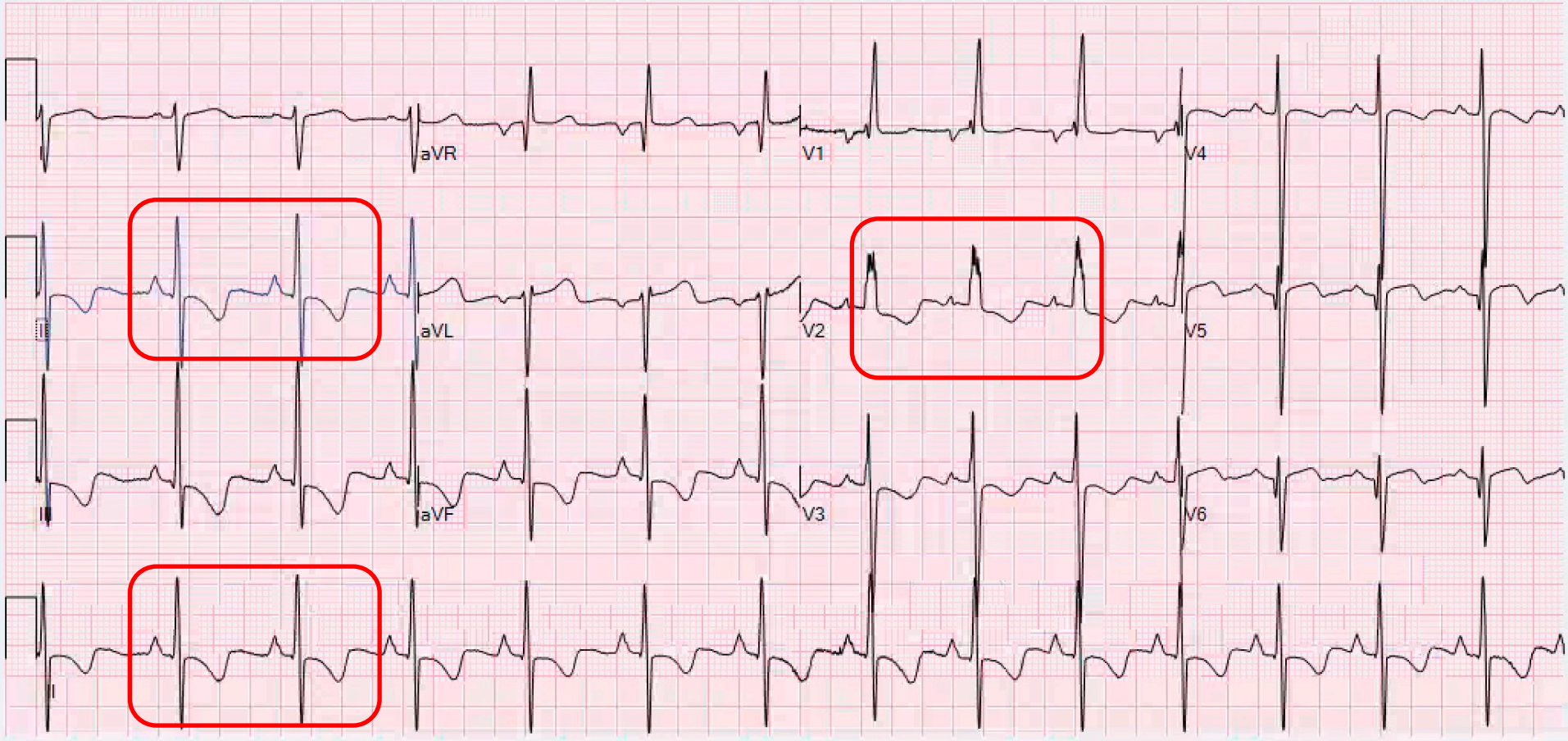
CHEST X-RAY WAS PERFORMED

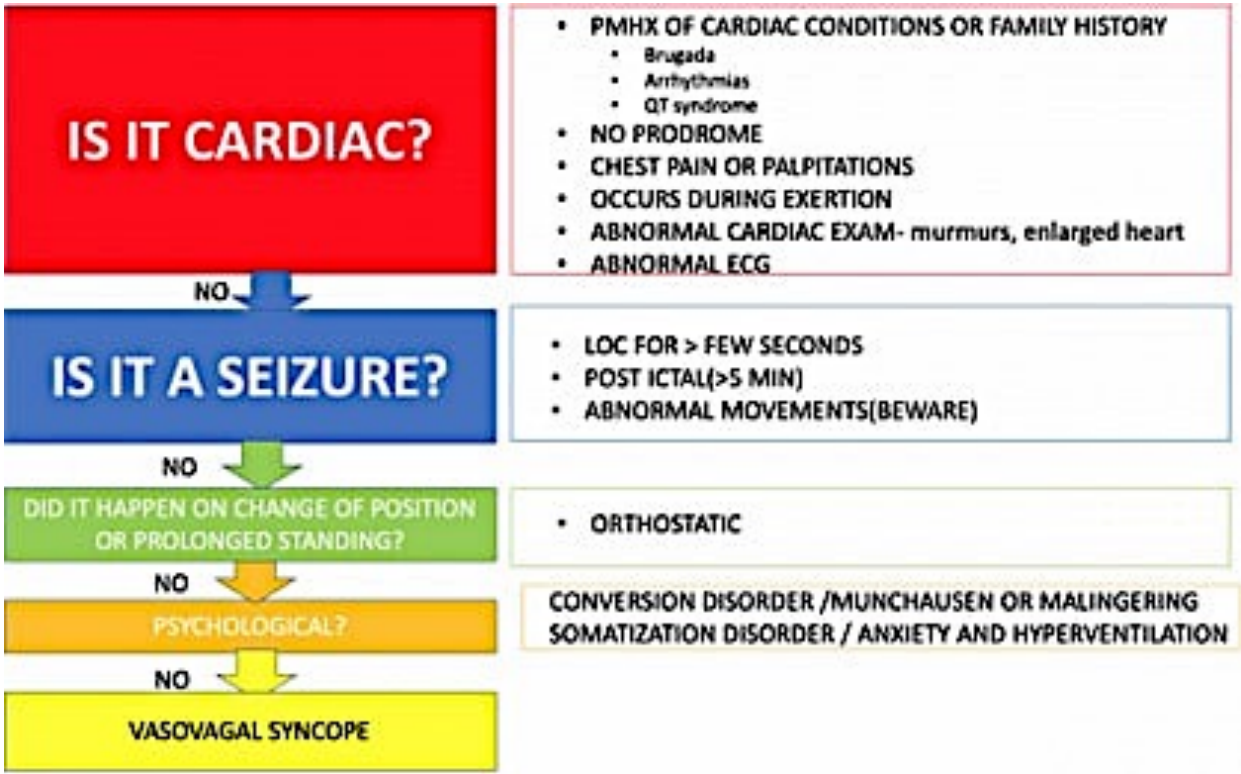


12 lead ECG



12 lead ECG

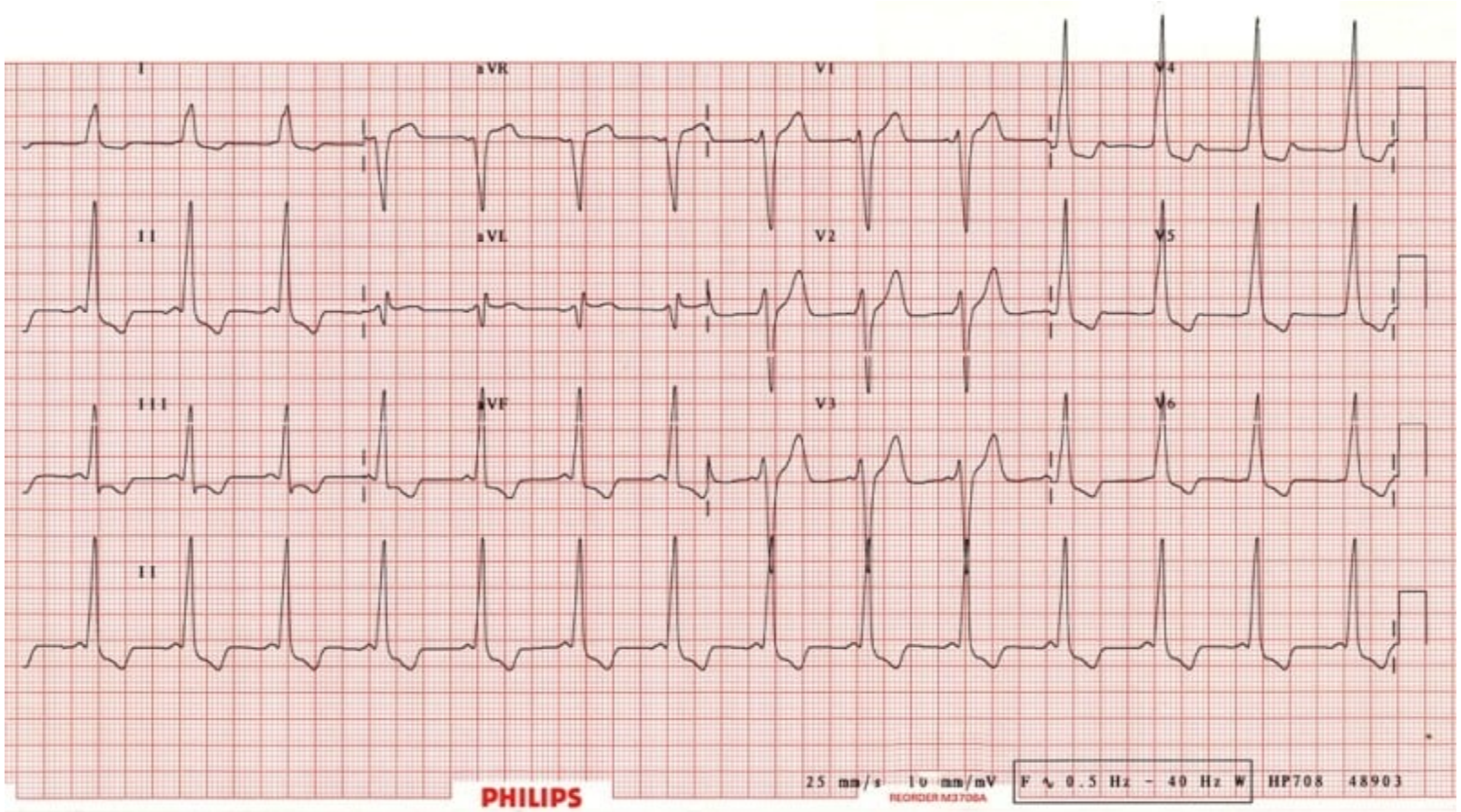



IS IT CARDIAC?
PMHX OF CARDIAC CONDITIONS OR FAMILY HISTORY
Brugada
Arrhythmias
QT syndrome
NO PRODROME
CHEST PAIN OR PALPITATIONS
OCCURS DURING EXERTION
ABNORMAL CARDIAC EXAM- murmurs, enlarged heart
ABNORMAL ECG
NO
IS IT A SEIZURE?
LOC FOR > FEW SECONDS
POST ICTAL(>5 MIN)
ABNORMAL MOVEMENTS(BEWARE)
NO
DID IT HAPPEN ON CHANGE OF POSITION OR PROLONGED STANDING?
ORTHOSTATIC
NO
PSYCHOLOGICAL?
CONVERSION DISORDER /MUNCHAUSEN OR MALINGERING
SOMATIZATION DISORDER / ANXIETY AND HYPERVENTILATION
NO
VASOVAGAL SYNCOPE

ECG clues – 'WOBBLER'

	Abnormality	ECG section
W	Wolff Parkinson White	P, PR
O	Obstructed AV pathway	PR
B	bifascicular block	QRS
B	Brugada	ST
L	Left ventricular hypertrophy – Aortic Stenosis and Hypertrophic obstructive cardiomyopathy	QRST
E	Epsilon Wave	ST
R	Repolarisation abnormality (Long QT, Short QT)	QT

Wolff Parkinson white syndrome -



Murmurs

Murmurs

Innocent murmur	Pathological murmur
○ Low intensity – often Grade 1 or 2 ○ Short duration ○ Often early systolic ejection murmurs ○ Commonly present with a vibratory or musical quality ○ Typically, louder when supine ○ Associated with a normal S2	• Are commonly either diastolic or pansystolic • Long duration • High intensity – often Grade 3 or higher • Commonly associated with other physical or auscultatory findings • Pathologic murmurs are often harsh in quality • Associated with an abnormal S2

Systolic murmurs may or may not be pathologic (depending on the other symptoms and signs) but diastolic murmurs are always pathologic!

Mnemonic for features of innocent murmur

- Sensitive (changes with position or respiration)
- Short duration (not pansystolic)
- Single (no clicks or gallops)
- Small (murmur is limited to a small area, non-radiating)
- Soft (low intensity)
- Sweet (not harsh)
- Systolic (limited to systole)

Pathological murmurs – do not miss!!

- Failure to thrive - increased metabolic demand
- Dyspnea
- Syncope
- Cyanosis - hypoxemia
- Lethargy

Types of Lesions

- CHD is the commonest of all congenital defects
- Incidence is 1 in 10
- Increasingly being diagnosed on prenatal screening

Differences between acyanotic and cyanotic

Acyanotic	Cyanotic
With acyanotic lesions, the majority of the blood entering the right ventricle is pushed towards and oxygenated by lungs and pulmonary circulation.	With cyanotic lesions, there is insufficient oxygenation of blood and deoxygenated blood from the right side of the heart is able to mix with arterial blood causing both central and peripheral cyanosis.
• VSD (30-35%) • ASD (8-10%) • PDA (10-11%) • Coarctation 4% • PS 6% • AS 4%	• Tetralogy of Fallot • Transposition of Great Arteries • Truncus arteriosus • Total anomalous pulmonary venous return • Tricuspid Atresia • Others • Hypoplastic left heart syndrome, Pulmonary atresia

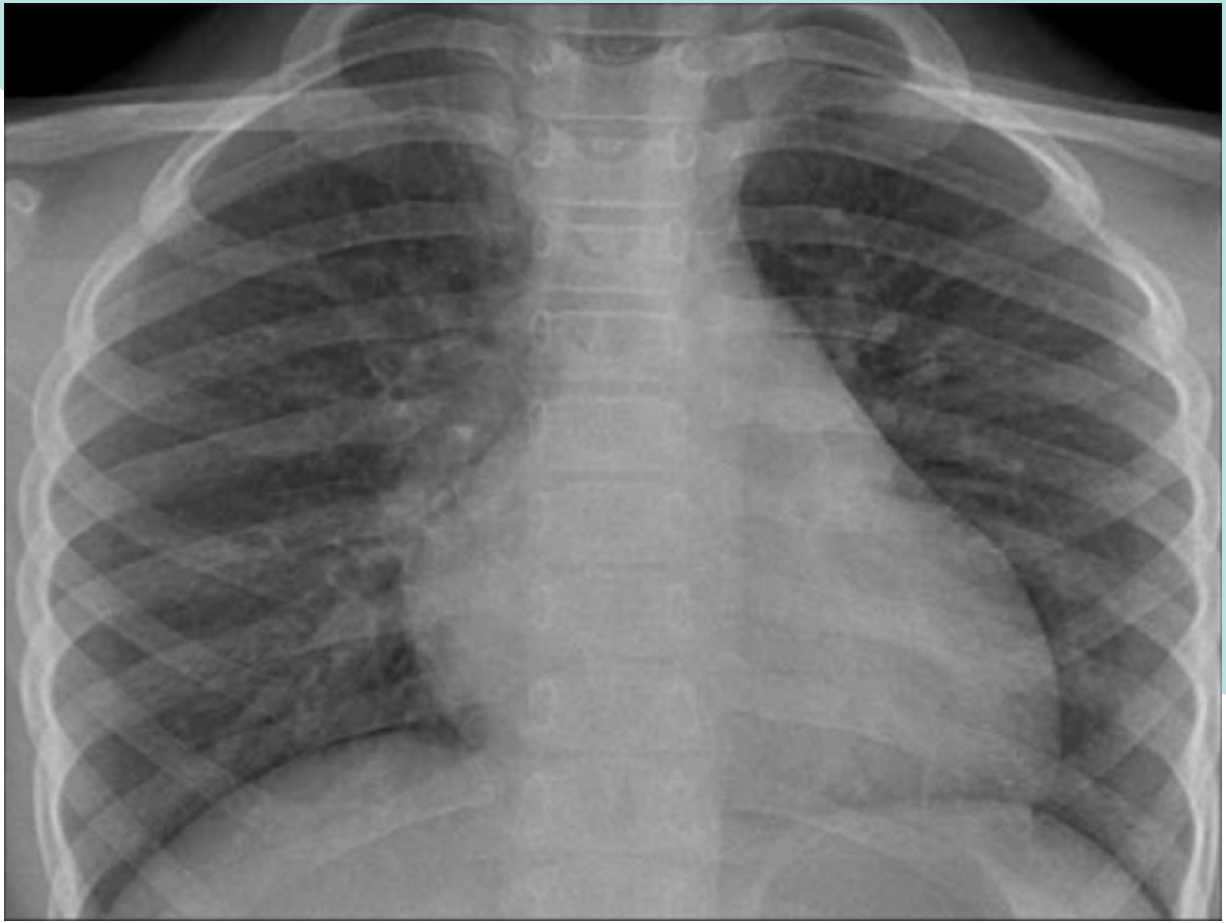
Case 4

- 4-year-old boy referred with a murmur
- The family denies any symptoms
- He often eats what the family is eating but does not gain weight very well

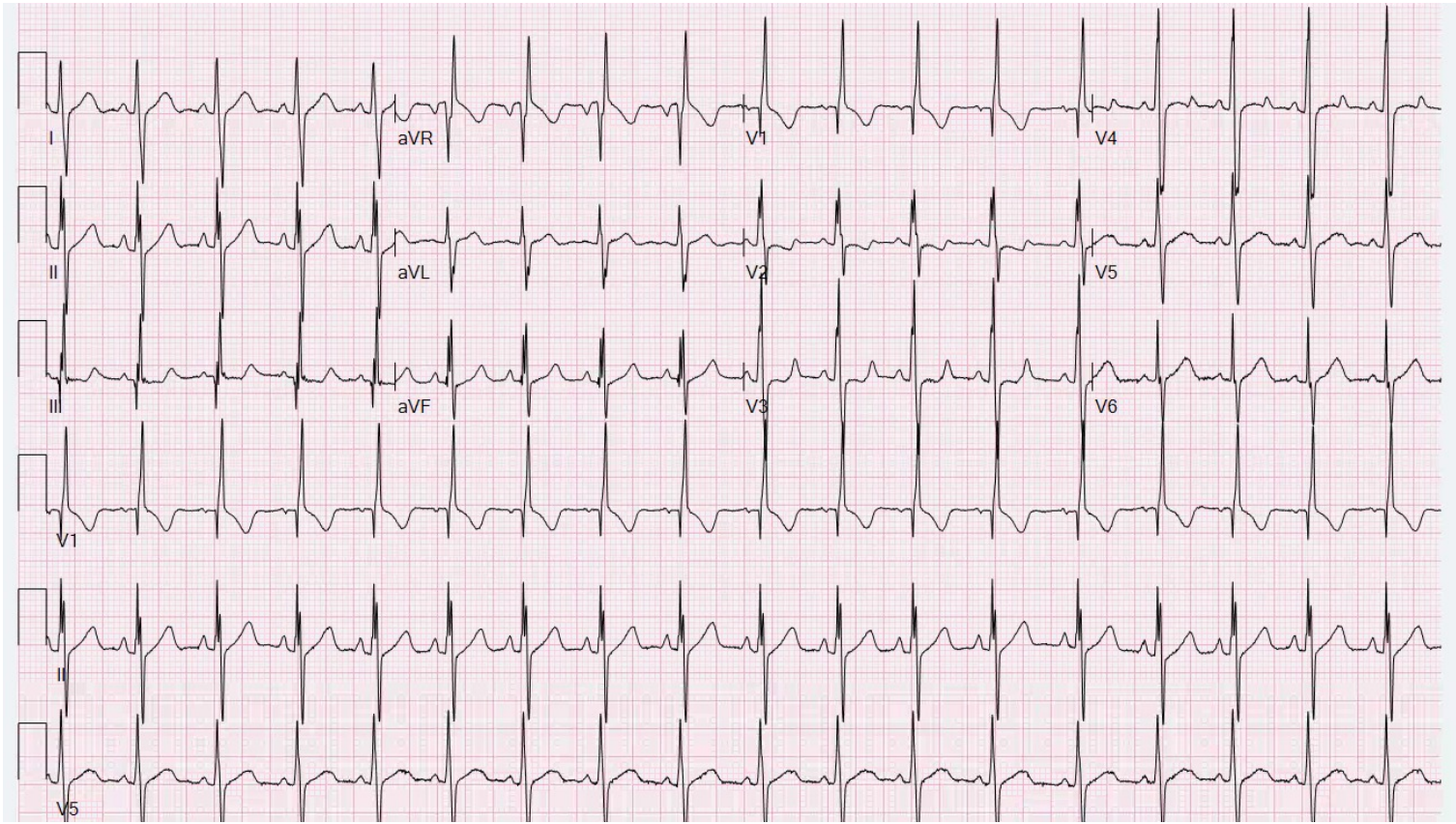
Examination

- 4-year-old boy, height 100cm weight 14.8kg
- Blood pressure 100/61, HR 108 beats/min
- SpO2 98% and RR 26 b/min
- Normal S1, fixed split S2, 2/6 ESM at LUSE
- No hepatomegaly

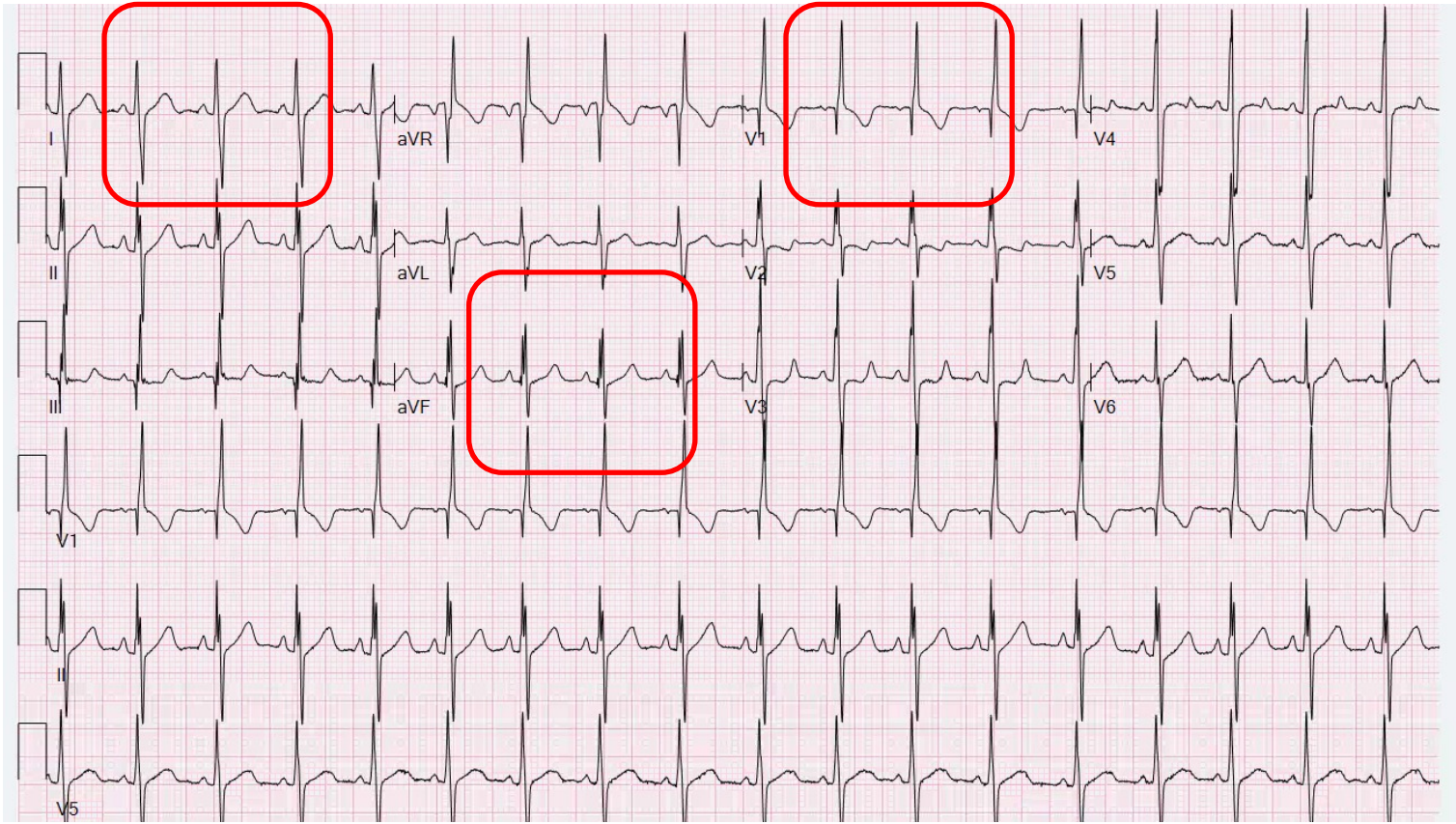
Chest X-ray



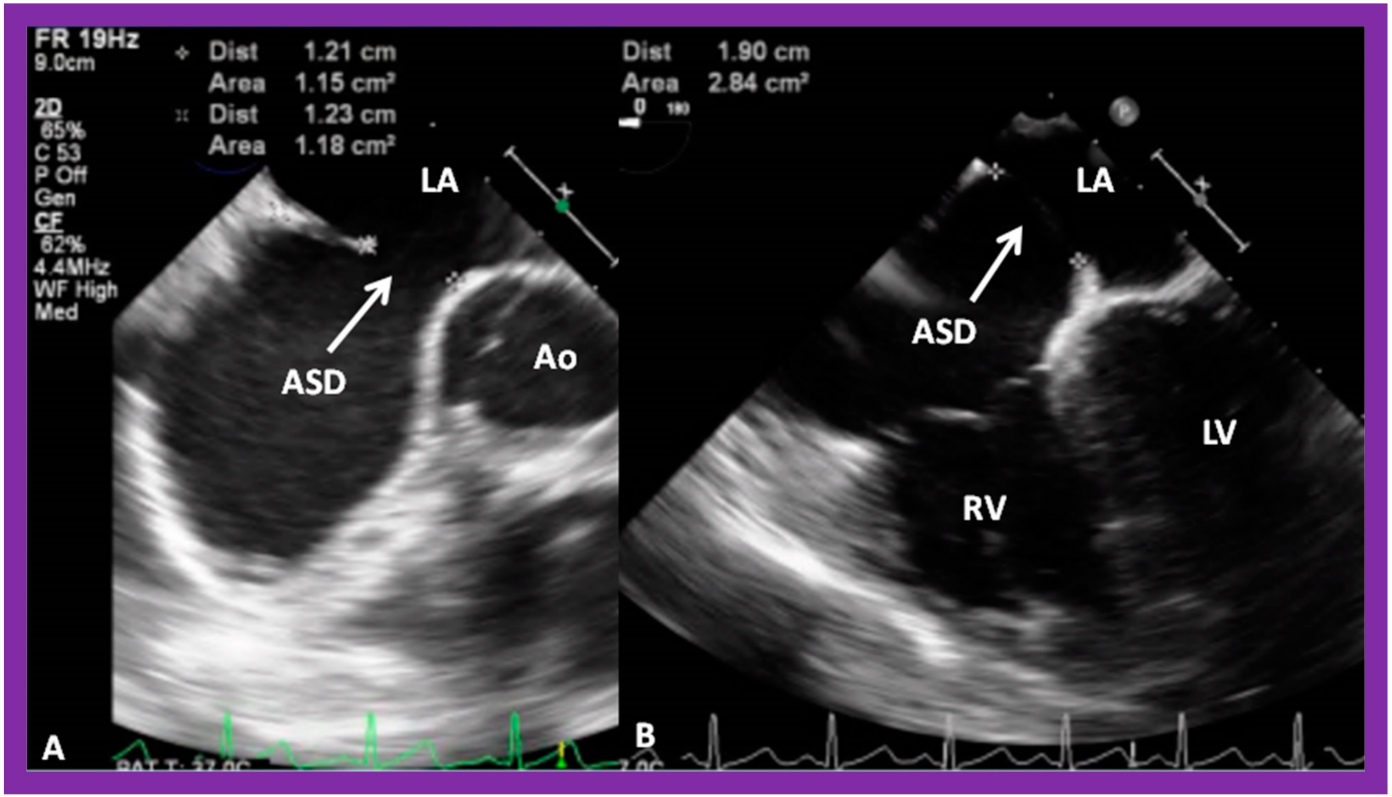
12 lead Electrocardiogram



12 lead Electrocardiogram



Echocardiogram of Atrial Septal Defect



Case #5

Mary is a 2-month-old infant who was referred to you from a walk-in clinic because a friend of the family noticed a murmur. You are a practice GP, and you have a GP registrar with you for the day who is eager to practice their physical exam skills. They would like to take history and start doing the clinical examination.

Before sending you in, she asks, “which of the following characteristics would raise concern for a pathological murmur?”

Red flags for murmurs

RED FLAGS

- respiratory difficulties,
- poor feeding,
- cyanosis,
- poor growth,
- Syncope

Note: diastolic and pansystolic murmurs are always pathological, whereas systolic ejection murmurs may or may not be pathological

Additionally, the following characteristics could also raise your suspicion for a pathological murmur:

- **family history of congenital heart disease or sudden cardiac death**
- **diminished or absent femoral pulses**
- **increased murmur intensity in upright position**
- **abnormal S2 (i.e. not physiologically split)**

Case #5

- Parents have no concerns about Mary other than her a viral upper respiratory tract infection last week, which has since resolved. Once or twice she has seemed to turn **slightly blue in the lips when crying very hard**; these episodes resolved spontaneously.
- PMHx: First child, uncomplicated pregnancy, spontaneous vaginal delivery at 38 weeks, went home at 3 days. Breastfeeding every 3 hours, generally well, but **sometimes takes a long time to feed**. There are no concerns about growing or gaining weight. Immunizations are up to date. No surgeries. No medications.
- Development: Meeting milestones. Gross motor – lifts head in prone, starting to steady her head when held. Fine motor – tracks objects, holds toys. Cognitive/social – recognizes mom's voice, smiles. Language – coos.
- FHx: Mom and dad are both healthy. No family history of murmurs, congenital heart disease, or sudden unexplained death.
- SHx: Mom is a teacher (currently on maternity leave) and dad is an architect. They have 2 dogs. Both maternal and paternal grandparents live nearby and are available to help.

Clues about significant lesion

- Prolonged feeding,
- diaphoresis with feeding, or an
- Infant needing to stop frequently to catch their breath while feeding.

There may be indicative of possible congestive heart failure, with pulmonary over-circulation, leading to shortness of breath and tachypnoea

Clues about significant lesion

- General Appearance: Mary is active and alert, appears well, no signs of distress
- Vitals: BP (systolic) 68, HR 116, RR 48, O2 saturation 85%, Temp 37.2 ° C
- Length 40th percentile, weight 30th percentile, head circumference 50th percentile
- Head/Neck: Normal
- Cardiac: normal peripheral pulses in all limbs, normal capillary refill 2 seconds, no thrills, RV heave, normal S1, single S2, Grade 3/6 harsh systolic crescendo-decrescendo murmur heard best along the left sternal border radiating to the axillae and back, liver is palpable 1cm below the costal margin.
- Pulm: Normal
- Abdominal: Normal
- Neuro: Normal
- Derm: Normal

Pediatric Vital Signs Reference Chart

Heart Rate (beats/min)		
Age	**Awake**	**Asleep**
Neonate (<28 d)	100-205	90-160
Infant (1-12 mos)	100-190	
Toddler (1-2 y)	98-140	80-120
Preschool (3-5 y)	80-120	65-100
School-age (6-11 y)	75-118	58-90
Adolescent (12-15 y)	60-100	50-90

Respiratory Rate (breaths/min)	
Age	**Normal**
Infant (<1 y)	30-53
Toddler (1-2 y)	22-37
Preschool (3-5 y)	20-28
School-age (6-11 y)	18-25
Adolescent (12-15 y)	12-20

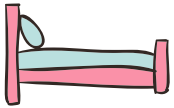
Reference: PALS Guidelines, 2015

Blood Pressure (mmHg)				
Age		**Systolic**	**Diastolic**	**Systolic Hypotension**
Birth (12 h)	<1 kg	39-59	16-36	<40-50
	3 kg	60-76	31-45	<50
Neonate (96 h)		67-84	35-53	<60
Infant (1-12 mos)		72-104	37-56	<70
Toddler (1-2 y)		86-106	42-63	<70 + (age in years × 2)
Preschool (3-5 y)		89-112	46-72	
School-age (6-9 y)		97-115	57-76	
Preadolescent (10-11 y)		102-120	61-80	<90
Adolescent (12-15 y)		110-131	64-83	

Reference: PALS Guidelines, 2015
For diagnosis of hypertension, refer to the 2017 AAP guidelines Table 4 & 5:
http://pediatrics.aappublications.org/content/early/2017/08/21/peds.2017-1904

2 Fast and 2 Big

Tachycardia

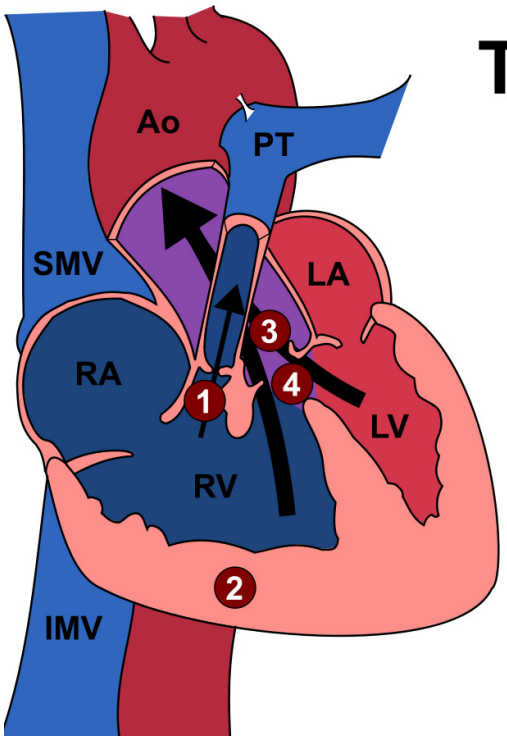


Cardiomegaly

Tachypnoea

Hepatomegaly

Features of tetralogy of Fallot



Tetralogy of Fallot

Major Defects

1. Pulmonary Stenosis
2. Right Ventricular Hypertrophy
3. Overriding Aorta
4. Ventricular Septal Defect

Lineage ©
Moises Dominguez

How to treat a Tet spell

Support measures

•The knees-to-chest position increases systemic resistance, reducing right to left shunting
•Having a caregiver hold the child prevents further agitation and may help him/her calm down

Medical interventions:

•Supplying oxygen is important because of the cyanosis (low oxygen saturation)
•Morphine can help calm the child and also reduce pulmonary vascular resistance

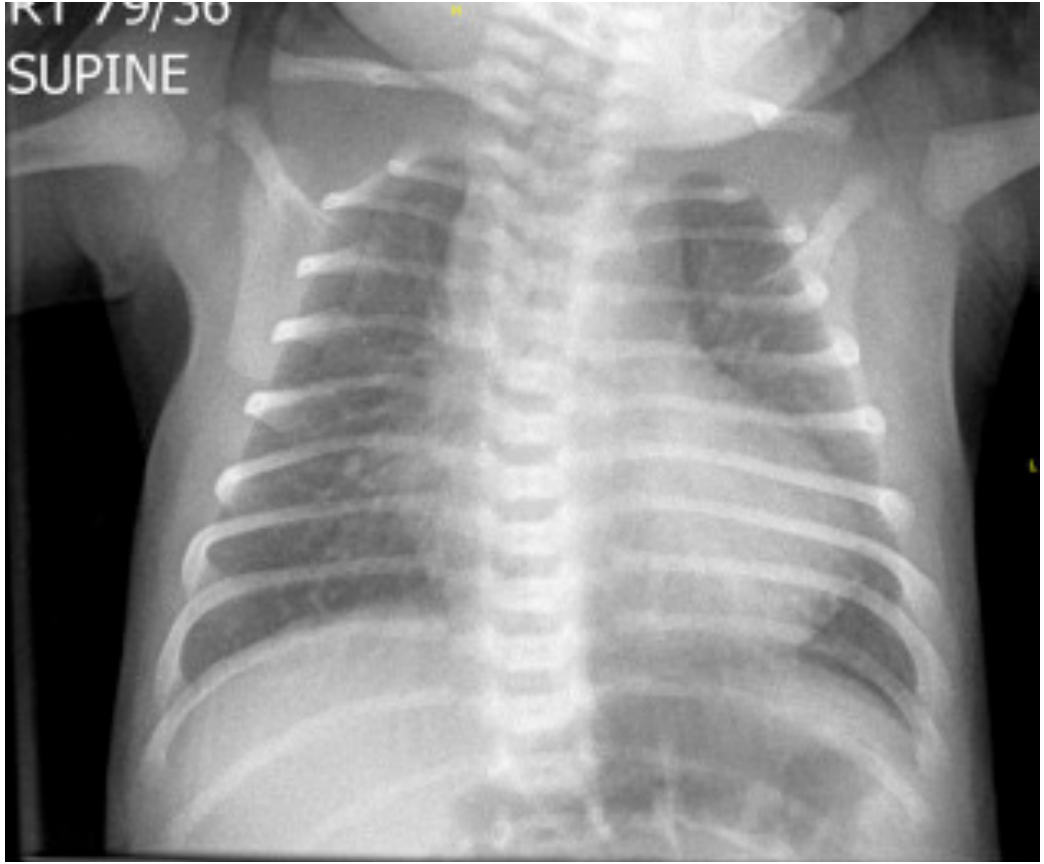
Case # 4

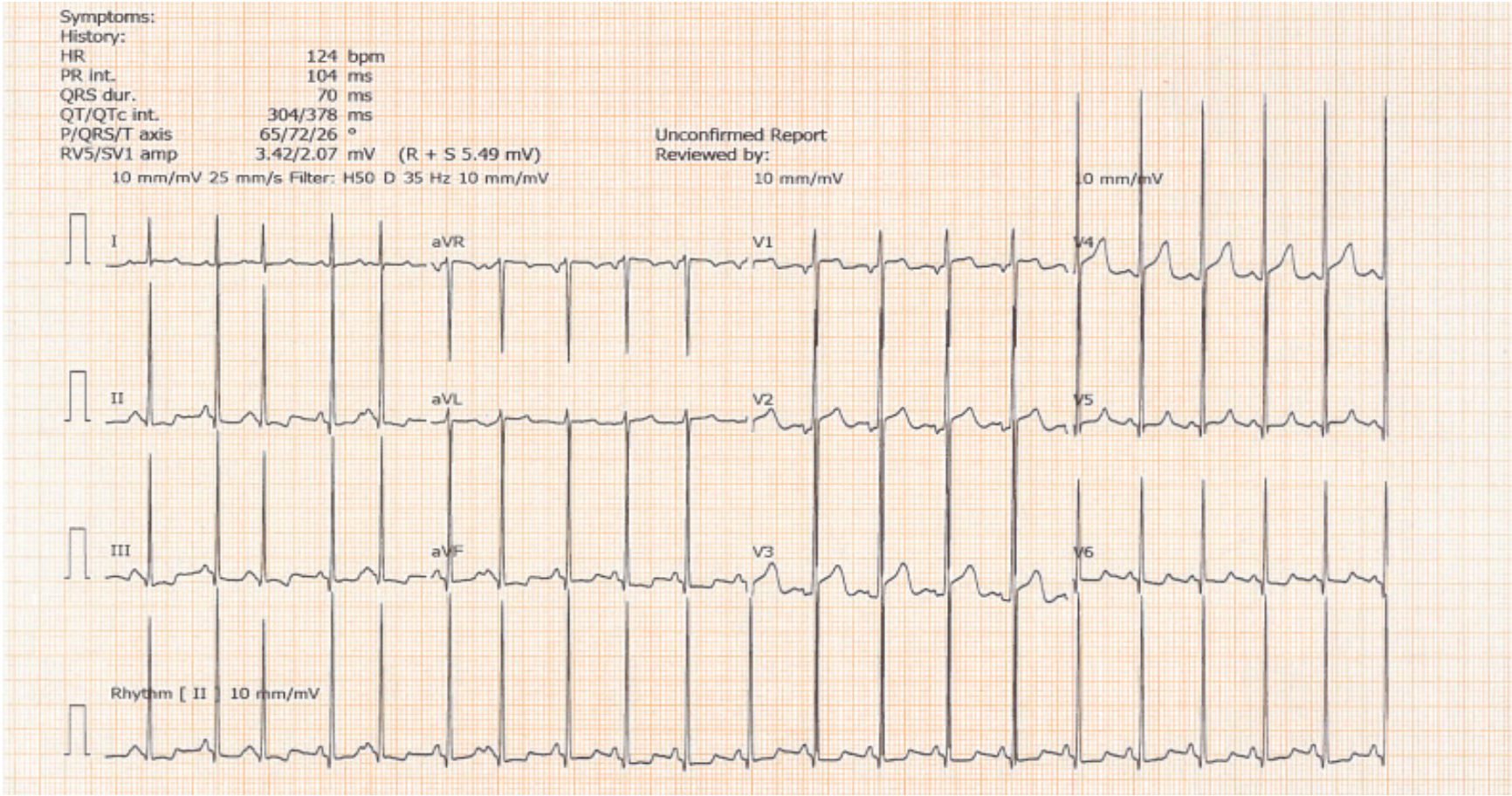
- 7 day old baby born at 38/40
- Referred to ED with feeding difficulties and weight loss
    - On discharge → 60mls q3h, now →15mls q2h
- Increased WOB and becomes tired, no sweating

Examination

- HR 184 b/min RR 80-90/min, SaO2 96%, BP – not picking up

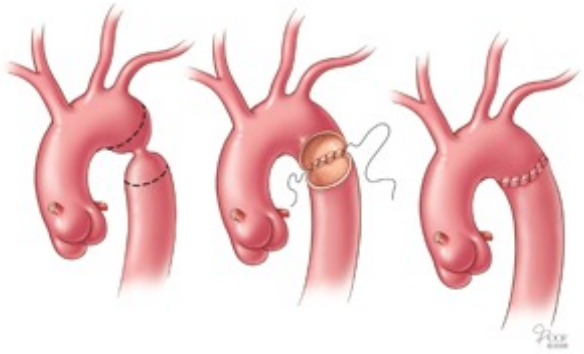
- No dysmorphic features, grey appearance, lethargic
- Tachypnoeic with subcostal recession
- Hands and feet are cool to touch
- Baby has a weak cry, radial pulses aren't easy to feel, brachial pulses are palpable
- Baby is lying in mum's arms and wrapped up
- Tachycardic…but the heart sounds are probably normal and there may be a soft ejection systolic murmur
- Femoral pulses are low volume

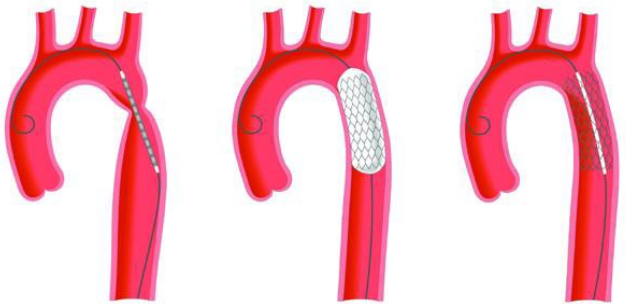

SUPINE


Symptoms:
History:
HR 124 bpm
PR int. 104 ms
QRS dur. 70 ms
QT/QTc int. 304/378 ms
P/QRS/T axis 65/72/26 °
RV5/SV1 amp 3.42/2.07 mV (R + S 5.49 mV)
Unconfirmed Report
Reviewed by:
10 mm/mV 25 mm/s Filter: H50 D 35 Hz 10 mm/mV
10 mm/mV
10 mm/mV
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
Rhythm [II] 10 mm/mV

Management

- Prostaglandin E1 infusion
- Manage heart failure
    - Inotropes, general supportive care (ventilation, correct acidosis)
- Definitive treatment
    - Balloon angioplasty or surgery





Location of murmurs

URSE (aortic area)	ULSE (pulmonary area)
AS supravalvular AS subvalvular AS	**ASD (relative PS)** **PS** **innocent pulmonary flow murmur (older chn)** **innocent pulmonary flow murmur (newborn)** pulmonary artery stenosis PDA COA AS PAPVR TAPVR
LLSE (tricuspid area)	Apex (mitral area)
VSD **AVSD** **vibratory innocent murmur (Still's)** TOF (VSD) TR HOCM	MR vibratory innocent murmur MVP AS idiopathic hypertrophic subaortic stenosis (IHSS)

Take home messages

Distinguish between benign and potentially life-threatening causes of

- syncope and
- cardiac murmurs in children

- Diagnose, manage and treat syncope and cardiac murmurs in children

Syncope summary

	Pearls	Pitfalls
Terms	Initial orthostatic hypotension is common in pediatric syncope	Classic Orthostatic is common in paediatric syncope
Position	Reflex syncope can rarely occur in sitting or supine position BUT…. Cardiac syncope should be evaluated in syncope of supine position	Reflex syncope cannot occur in sitting or supine position
Activity	Mid-exertional syncope = cardiac BUT ….post exercise syncope is more likely vasovagal syncope	All exercise-related syncope is cardiac syncope
Prodrome	Visual prodrome (i.e. blackening out or blurry vision) --> represents retinal hypoperfusion and that precedes brain hypoperfusion If no prodromal symptoms or absent = abrupt onset syncope	Because prodrome is an essential component of syncope, if there is no prodrome, syncope can be excluded
Way of falling	Flaccid falls without neurological impairment = syncope	A stiff fall = generalized tonic-clonic seizure
Movements	Convulsive movement is common in syncope Convulsive activity in syncope is characterized as brief, asynchronous movement that DOES NOT appear before falls	In children with transient loss of consciousness, convulsive movements strongly suggests seizures
Urinary incontinence	Urinary incontinence has no value in differentiating between syncope and epilepsy	Presence of urinary incontinence following loss of consciousness suggests seizures
Confusion	Postsyncopal symptoms, which are common and persistent for a while in Vasovagal Syncope, should not be mistaken for postictal confusion.	Post syncopal symptoms including persistent pallor, nausea and sweating are signs of confusion

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