PAEDIATRIC CASED BASED QUESTIONS 01/09/20

CASE ONE

A 5 day old neonate is brought into the ED with poor feeding, rapid and abnormal breathing and pallor. She is a term neonate with no immediate perinatal complications and underwent standard antenatal care.

HR 200 SpaO2 75% (L arm) RR 60 BP 80/50 (R arm) 36.3 (rectal) central CRT 4s

- **What are your potential differential diagnoses?**

- Sepsis, cardiac, metabolic and electrolytes, trauma(NAI), other

- **How would you examine/investigate and manage this neonate?**

  - Get help (paeds, ED, ICU, Neonatal)

  - Initial supplement O2 accept sats 90% initially

  - IV access bloods and cultures, **Glucose**

  - Pre and post ductal saturations and 4 limb BP

  - Feel 4 limb pulses and auscultate HS ? murmurs and chest ? failure

  - ECG for treatable causes

    1. HR >200 and/or inverted p waves in I and aVF: susp SVT -> ? adenosine or cardioversion
    2. Big Q waves in lateral leads = ALCAPA: immediate surgery consult, ***very*** careful diuretics/inotropes
    3. LVH suggests tricuspid atresia ?

  - Treat for sepsis Abs and IVF if appropriate

- **You note a holosystolic murmur and poor pulses in left arm and lower limbs and cyanosis in the lower body. Observations and VBG:**

  BP R arm 82/50 L 45/30 VBG pH 7.05, Lactate 10, Gluc 4
  SpaO2 R arm 80% oa L 68%

  Discuss potential congenital cardiac causes for this presentation?

- Likely duct dependent obstructive systemic lesions : Coarctation, Ao interruption, AS

Three main ways they present with cardiac lesions:

- Shock: obstructed flow to body (e.g. coarct, AS)
- Blue: obstructed flow to lungs (e.g. tricuspid atresia)
- Heart failure (e.g. AV canal defect)

Duct dependent lesions:

- Ductal dependent *pulmonary lesion*: cyanosis/hypoxia with normal (or under-perfused) CXR
- Ductal dependent *systemic lesion*: shock, pre/post-ductal BP/Spo2 differential, congestion on CXR

- **How would you manage this neonate with likely duct dependent lesion?**
- Get help !
- Keep warm, tx for sepsis
- Keep isovolaemic, only small titrated IVF for shock
- Controlled O2 aim pre ductal sats 85 % is okay
- Prepare for the need to intubate with senior support
- Prepare for PGE infusion under Neonatal/PICU advice (apnea and hypotension common)
- Inotropes may be required choice dictated by likely lesion and degree of shock, avoid increasing afterload in systemic obstructive lesions
- Avoid hyperoxia as this will increase pulmonary blood flow and reduce systemic

Duct-dependent for Systemic Blood Flow

With severe left-sided obstructive lesions systemic blood flow is dependent on right-to-left flow through a patent ductus arteriosus, so these babies are duct-dependent. Examples: Hypoplastic Left Heart Syndrome, critical aortic stenosis, coarctation of aorta, interrupted aortic arch.

- **Insert a double lumen umbilical venous catheter**
- Commence a prostaglandin infusion at an initial dose of 10 nanograms/kg/min.
- Do not over-oxygenate the infant (over-oxygenation will result in increased pulmonary blood flow and reduced systemic blood flow).
- Accept oxygen saturations of 75% or above. Reduce inspired oxygen if saturations >85%.
- Contact the paediatric cardiologist on call between 12 midnight and 6.30 a.m.if saturations < 75%, otherwise inform on call paediatric cardiologist at 06.30 a.m.
- The baby is to remain nil by mouth.
- If the infant requires assisted ventilation, ensure that the baby is not over-ventilated. The aim should be to initially ventilate to keep a low-normal arterial pH. Sedation, muscle relaxation, and controlled hypoventilation to further reduce arterial pH may be necessary if there is excessive pulmonary blood flow and reduced systemic blood flow (oxygen saturations >85%, low MAP, tachycardia, cool peripheries).

Duct-dependent Cyanotic Lesions

These lesions are duct-dependent either to ensure adequate pulmonary blood flow (e.g. pulmonary atresia, critical pulmonary stenosis) or to ensure adequate mixing between the systemic and pulmonary circulations (transposition of the great arteries).

- Commence a prostaglandin infusion at an initial dose of 10 nanograms/kg/min.
- Babies with TGA should be nil by mouth until atrial septostomy has been performed. Babies who are duct dependent for pulmonary but not systemic blood flow (Pulmonary Atresia and Tricuspid Atresia) may receive EBM feeds if stable.
- Ensure that at least one extra IV leur is available in the event that the PGE1 infusion tissues.
- If the systemic oxygen saturation is below 75%, call the paediatric cardiologist on call.
- If the infant develops apnoea or the systemic oxygen saturation is below 75% despite prostaglandin, they should be ventilated.
- If the infant develops apnoea but has a systemic oxygen saturation of 75% or above, the dose of prostaglandin can be reduced (but not below 5 nanograms/kg/min). If apnoea continues, the infant should be ventilated.
- If the infant is delivered after midnight but is stable, the paediatric cardiologist should be contacted in the morning by 0700 hours. If unstable, contact the paediatric cardiologist on call.
- OBSTRUCTIVE LEFT HEART LESIONS: ie HLHS, Interrupted aortic arch, Co-arctation, Aortic or Mitral atresia - PICU within 24 hours after discussion with NICU/PICU/Cardiology consultants.

CASE TWO

A 5yr old Tongan boy is brought into the emergency department with history of fevers and joint pain; he collapsed at home but now is fully alert and relatively well looking. He has had some diarrhoea recently, which other family members in the household have also had. He also complains of significant low back pain prior to the collapse.

Obs HR 100, Temp 38.0, BP 98/65, Sats 99%

- **What are your differential diagnoses?**

- Broad range of potential
- In this setting always consider Rheumatic fever
- Be aware of atypical presentations which are not uncommon

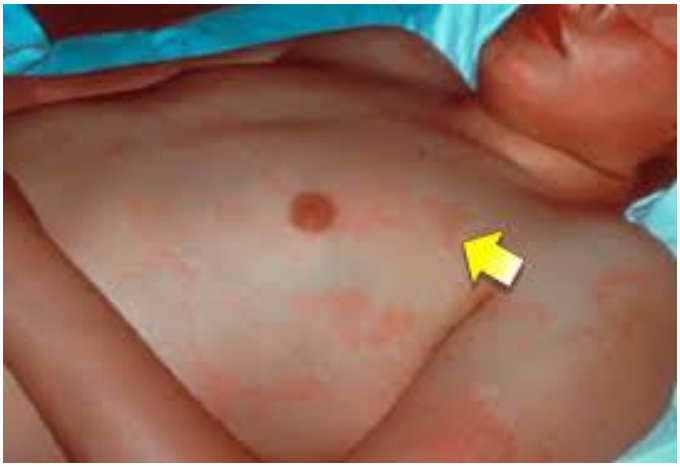
- **What important history do you want to ask?**

- Full social + pmhx + fhx (Rheumatic fever risks, cardiac hx)
- Details of the recent illness
  - Sore throat/skin infection/rash/chest pain/sob
  - Nature of the collapse ? syncope or seizure
  - Oral intake and losses
  - Systems review
- Immunisations
- Current medications and allergies
- Travel and contacts

- **What key examination findings are you looking for?**

- Thorough examination of all systems
- Skin rash, joint arthritis/arthralgia, red throat
- Murmurs
- Review nature of back pain - ? reproducible? bony? referred
- Neurologic exam

- **You find on examination a soft systolic murmur and tender bilateral ankles without obvious swelling, significant lumbar/SI pain worse with movement, and areas of erythematous rash (below). The rest of his exam is unremarkable. Discuss further investigations?**



- ECG ? myopericarditis, PR prolongation, AVB
- CXR – heart size
- Bloods and cultures : CRP/ESR, FBC, U+E, strep titres
- Consider Lumbar XR

- **What are the diagnostic criteria for Rheumatic fever?**

- See NZ Heart foundation guidelines (Nz specific take on Jones Criteria)

The presence of two major symptoms, or one major and two minor symptoms, in both cases with a prior GAS infection, are required to diagnose ARF.

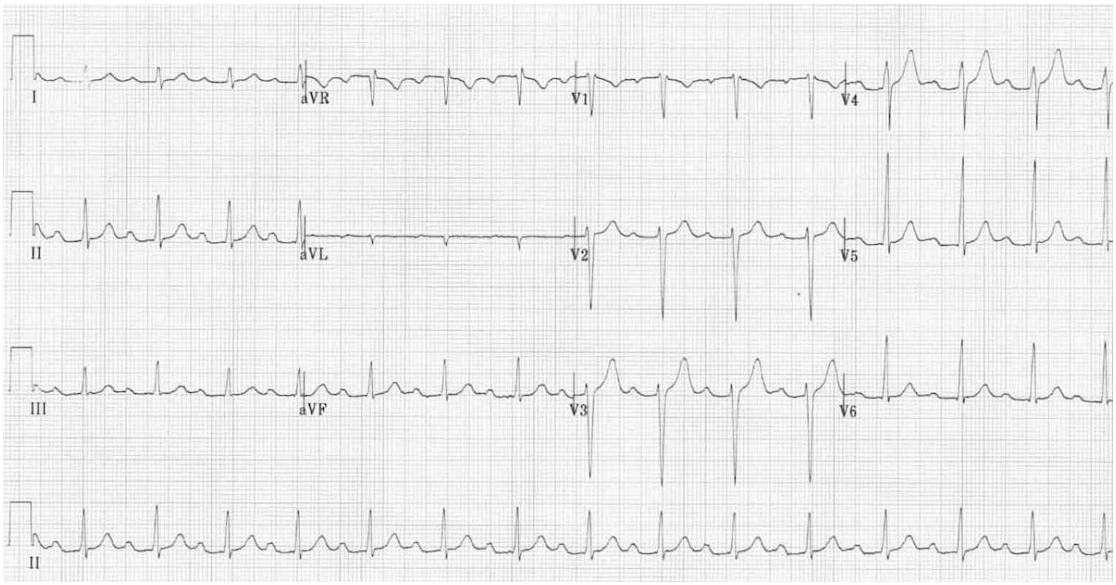
Major symptoms include:

- Arthritis, the most common symptom, occurs in 75% of first attacks, usually in the larger joints such as the knees and ankles[6]
- Carditis almost always affects the mitral and aortic valves and on presentation, a murmur may be heard.[6] In New Zealand, subclinical carditis confirmed by echocardiography is also considered a major symptom.
- Chorea (uncoordinated movements), often in adolescent females, especially affecting the hands, feet, tongue and face which disappear during sleep and may only affect one side of the body. Chorea may occur following a prolonged latency after streptococcus infection and generally resolves within six weeks.
- Erythema marginatum – rare (pink rings on the trunk and limbs)
- Subcutaneous nodules – rare, but highly specific to ARF

Minor symptoms include:

- Fever which accompanies most cases of ARF, except when chorea is present
- Joint pain/arthralgia
- Elevated CRP >30 mg/L or ESR >50 mm/h
- A prolonged P-R interval on ECG

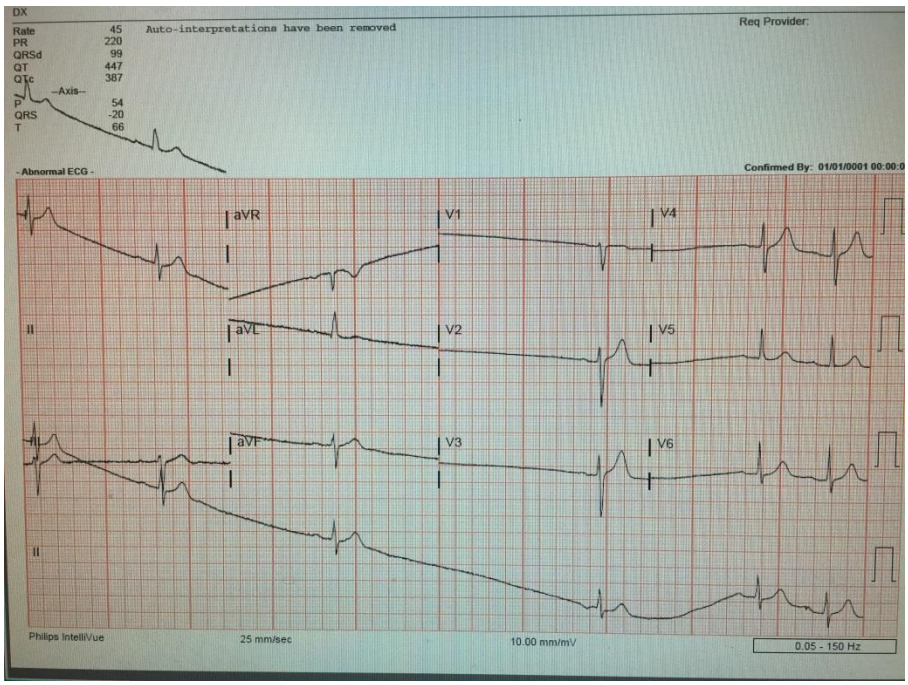
- **Below is his ECG please describe and discuss concerns?**



Markedly prolonged PR interval

- In context of hx of collapse consider possibility of intermittent higher degree block or sinister rhythm
- Suggestive of carditis

- **The child then develops episodes of being pale/clammy, and less responsive while in the assessment bed. These are recurrent lasting 10-20s. He is noted to have a palpated pulse that is very slow during these episodes and his BP is 80 systolic. Below is his ECG. Discuss your management of this child?**



SINUS ARREST/PAUSES

- Move to resus on monitoring and defib
- Get help ED/ICU/PAEDS/PAEDS CARDIO
- Get ready for potential need to intervene he develops sustained haemodynamic compromised state

Atropine 20mc/kg
Dilute adrenaline 0.5-1mcg/kg bolus titrated if required
Consider external pacing (sedation ?)

- Needs urgent paeds cardiology consideration transvenous pacing (not safe for transfer would need to come to patient ?)
- PICU involvement

CASE THREE

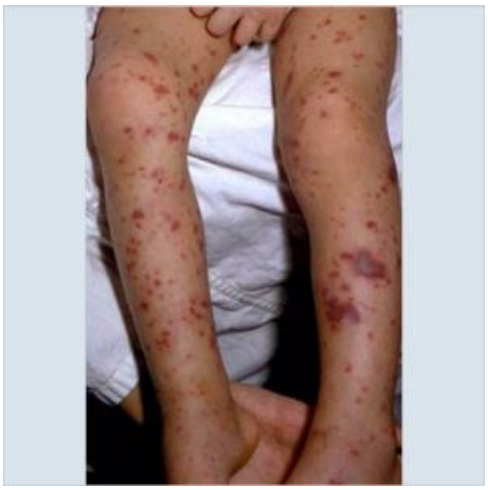
8yr old girl brought from home by her Mum with fevers for 1/7, headaches, and vomiting.

Obs HR 140 RR 20 BP 90/50 CRT 2s Sats 98% temp 38.5 GCS 15

- **What are your differential diagnoses and how would you manage this child?**

- Infection most commonly viral but consider bacterial sepsis
- Thorough history and examination
- Antipyretics and antiemetic
- Emla/ametop consideration need for further investigation
- ORT
- Consider establishing IV access immediately and empiric antibiotics if looking unwell

- **Within 20minutes of arrival she starts to look more pale and lethargic. A rash develops rapidly (as below). What is your concern and how would you manage this child?**



PURPURIC RASH

BACTERIAL SEPSIS/MENINGOCOCCAEMIA!

- Resus area and get immediate help!
- IV access x 2 if not already, IO if delay to IV and not able
- IVF bolus 10-20ml/kg N saline ideally warmed
- IV ceftriaxone 100mg/kg (max 2g) or cefotaxime (Firstline in < 12months) (risk reaction with hx of penicillin anaphylaxis < 1%)
- Ensure FBC/U+E/Coag/Grp&Hold/cultures/VBG
- Tx hypoglycaemia
- Keep warm (ongoing volume should be warmed)

- **Despite 40ml/kg boluses she remains shocked as per her observations and VBG/bloods. Discuss your on-going management?**

BP 70/40, HR 150, Sats 92% 4L Hudson, CRT 4s, GCS 13

pH 7.18 HCO_3 14 Na 130, K 3.5, Gluc 4, Lactate 8

Hb 90, WBC 30, Plats 80, Alb 18

APTT 80, PR 1.4, Fibrinogen 0.1 g/L

➢ RESISTANT SEPTIC SHOCK WITH EVIDENCE OF MULTI-ORGAN DYSFUCNTION

- Likely need Airway secured : planning/personnel and drugs
  Resuscitate before your intubate (HOP KILLERS)
- Will require pressors:
  Titrated dilute adrenaline boluses
  Titarted Metaraminol
  Ideally Norad via CVL

- On-going directed volume resuscitation using blood products
  4% albumin
  RBC
  CRYOPRECIPITATE (fibrinogen, von-willebrand factor, factor viii, factor xiii and fibronectin)
  PLATLETS
  FFP (all soluble clotting factors)
- $NaHCO_3$

- Disposition PICU

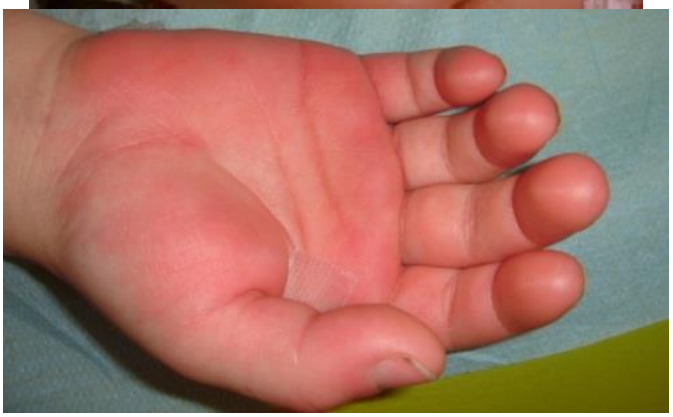
CASE FOUR

2 year old girl is brought to the emergency department with 4 days of high fevers and irritability. She has seen her GP and placed on amoxicillin for an ear infection. Her mother is concerned. She is immunised and has no past history.

- **Discuss how you would approach this patient's care?**

- Standard thorough history of events
- Examination top to toe: do they look unwell?
- Consider potential serious illness
- Is there a focus for infection?
- Consider investigations: Bloods/MSU/ECG/CXR

- **Below are some findings on the child. What other details/findings would you want to know and what other investigations would be useful?**



Consider Kawasaki Disease: multisystem illness with fever and rash, which occurs mainly in children less than 5 years old. **Within 3 days** of the abrupt **onset of fever**, the other characteristic features usually appear:

- Bulbar conjunctivitis (no exudate)
- Mucositis: red cracked lips, red mouth and throat, strawberry tongue
- Polymorphous generalized rash that can be morbilliform, maculopapular, scarlatiniform or may resemble erythema multiforme

- Induration of the hands and feet with red palms and soles
- Cervical lymphadenopathy (usually a solitary, unilateral node > 1.5 cm in size)
- BCG site reactivation (erythema around BCG scar, usually on left upper arm)

- **What are your differential diagnoses?**

- Viral infections e.g. measles, adenovirus, enterovirus
- Scarlet fever
- Staphylococcal scalded skin syndrome
- Toxic shock syndrome
- Polyarteritis nodosa
- Bacterial cervical lymphadenitis
- Drug hypersensitivity reactions
- Stevens-Johnson syndrome
- Leptospirosis
- Mercury hypersensitivity

- **What is Kawasaki Disease and what are the diagnostic findings?**

- Multi-system disease/vasculitis
- 20% of untreated patients may develop coronary artery aneurysm
- Desquammation hand/feet/digits is a late sign
- Pathway for incomplete criteria (SSH guidelines)
- Diagnosis can be made where there is **fever plus at least four of the five features:**

➢ Bulbar conjunctivitis (no exudate)
➢ Mucositis: red cracked lips, red mouth and throat, strawberry tongue
➢ Polymorphous generalized rash that can be morbilliform, maculopapular, scarlatiniform or may resemble erythema multiforme
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- **Other Findings:**

Extreme irritability, severe abdominal pain, diarrhoea and vomiting are common. Other features may include:

➢ Urethritis with sterile pyuria (70% of cases)
➢ Hepatic dysfunction (40% of cases)
➢ Arthritis or arthralgias (35%)
➢ Aseptic meningitis (25%)
➢ Pericardial effusion or arrhythmias (20%)
➢ Gallbladder hydrops (<10%)
➢ Carditis with congestive heart failure (< 5%). This can occur at any time in the first 3 weeks, and usually resolves by 6 to 8 weeks.
➢ Other arterial aneurysms (e.g. iliac, femoral, renal, axillary) may occur

- **Supplementary Laboratory Information:**

- Albumin < 30g/L
- Anaemia for age
- White cell count > 15 E+9/L
- Platelets after 7 days >450 E + 9/L
- Elevated ALT
- Urine White Cells > 10 per high powered field

- **Echo finds:**

- Positive Echocardiogram (any ONE of the following)

- Z score of RCA or LAD >2.5
- Japanese MOH criteria
- Any 3 suggestive features:

  - Perivascular brightness

  - Lack of tapering of coronary arteries

  - Z score > 2.0

  - Pericardial effusion

  - Mitral regurgitation

  - Impaired LV function

- **Late Manifestations:**

- The lips usually begin to dry, crack and fissure by day 6 of the illness
- The skin of the fingertips, palms ± soles begins to peel in weeks two and three
- Beau lines (transverse grooves in nailbeds) and temporary hair loss
- With no treatment, the average length of fever is 12 days, and when the fever resolves the child may remain irritable for a further 2 to 3 weeks.

CASE FIVE

5 weeks old male presented with 1 week of coryzal illness and poor feeding since yesterday. He last feed this morning - 10mls. He was taken to the GP and referred to hospital for dehydration. The GP noted a HR of 110bpm.

On arrival at ED his obs where noted as:

HR 270bpm, BP 100/89, RR65, CRT 3s , Alert

1. **What is you initial concern?**

- Likely tachyarrhythmia given HR > 200bpm
- 30-40% of new SVT present in the first few weeks of life

2. **Discuss your approach outlining important history, examination, and initial investigations?**

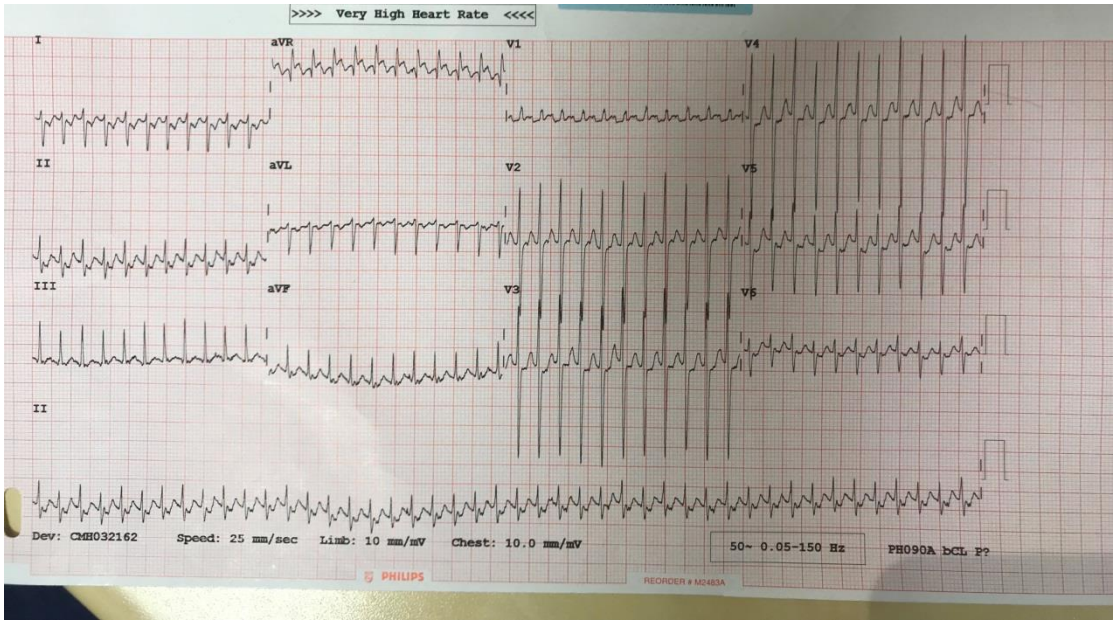
- Place in resus area with monitoring
- Remember the family, explain and ongoing information

- History – antenatal/perinatal/postnatal
- Known cardiac hx
- Feeding
- Breathing, apnoea, colour change
- Infective symptoms

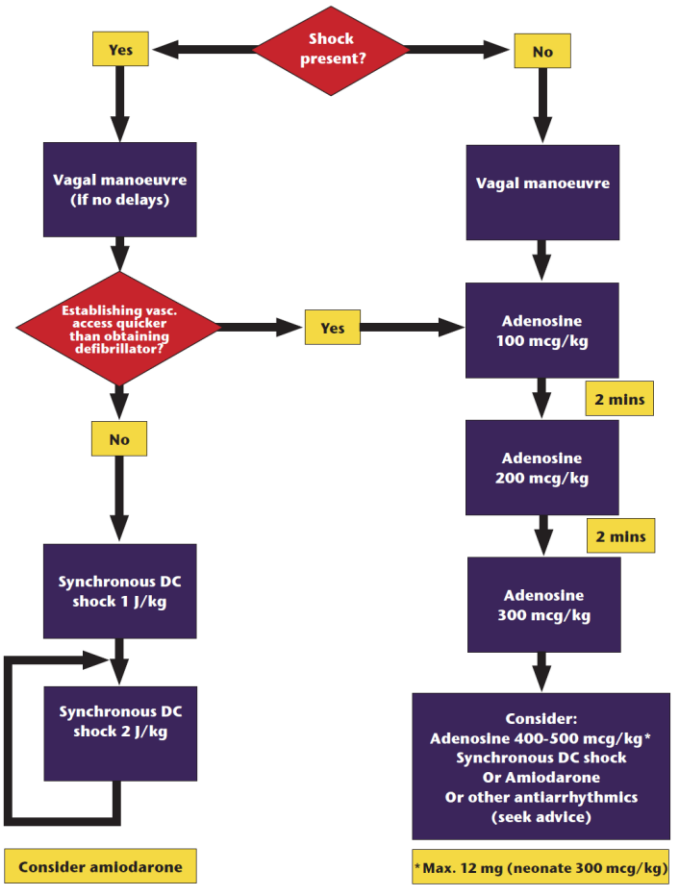
- Examination
- Pulses, BP, and saturations pre and post ductal
- Murmurs
- Signs failure: lungs, liver, edema
- Rashes
- WOB

- Investigations
- ECG
- BGL
- Basic bloods – electrolytes, TFTs

3. Below is his ECG discuss your diagnosis and management plan?



- Narrow complex tachyarrhythmia 270bpm : SVT
- Follow SVT algorithm

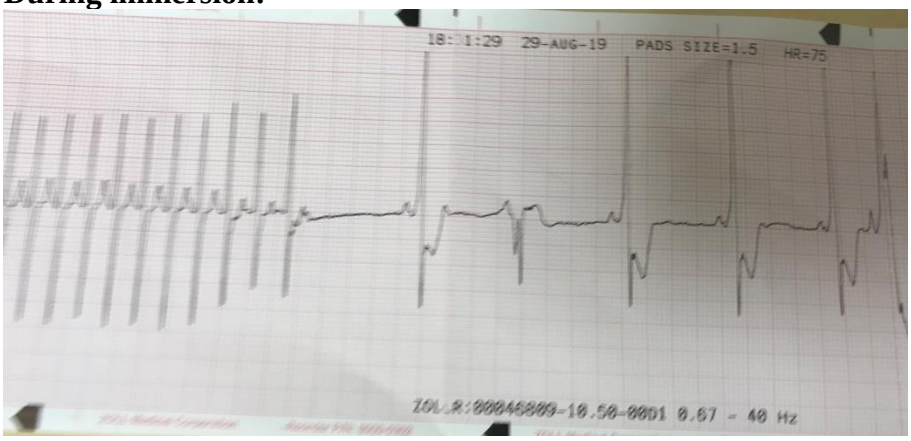


4. **Discuss how to carry out vagal stimulation in neonates/infants and approach with a toddler or older child?**

   - **Neonates and infants:** Facial immersion in ice water 10-15s. **This technique must not be used for infants in circulatory shock.** The baby is attached to a cardiac monitor, arms are wrapped in a towel, and the whole face is immersed in ice water slurry for five seconds. It is unnecessary to occlude the nostrils. This technique is safe and 90% effective in terminating a reentrant tachycardia. Explain carefully to the parents what you are doing. The baby will not drown!
   - **Toddlers:** Ice cold facecloth to the face 30s . Older infants resist being dipped into water as above, but this technique is almost as effective. Eyeball pressure is no longer recommended. Unilateral carotid sinus massage can be useful in older children, but it is often difficult to perform.
   - **School-aged children:** Valsalva technique; ask the child to blow on their thumb after full inspiration for 10-15 seconds. Demonstrate the technique and have the child copy you. There should be no air escape and the child should be seen to strain ("playing the trumpet silently").

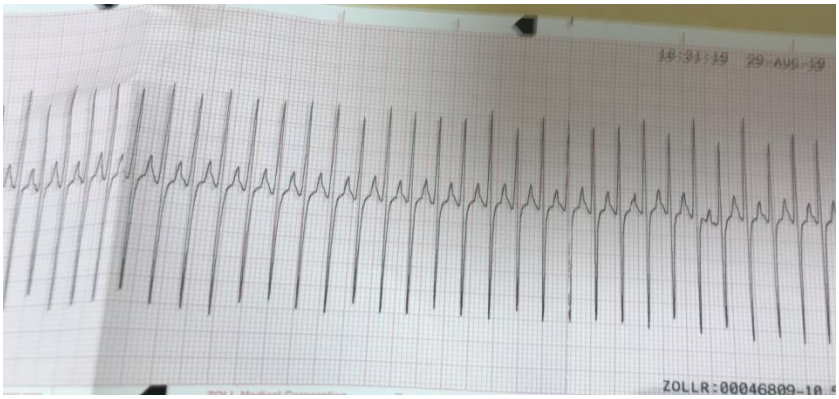
5. **Below is the ECG tracing during and after face immersion after two attempts. Discuss the ECG morphology and your next step in management?**

During immersion:



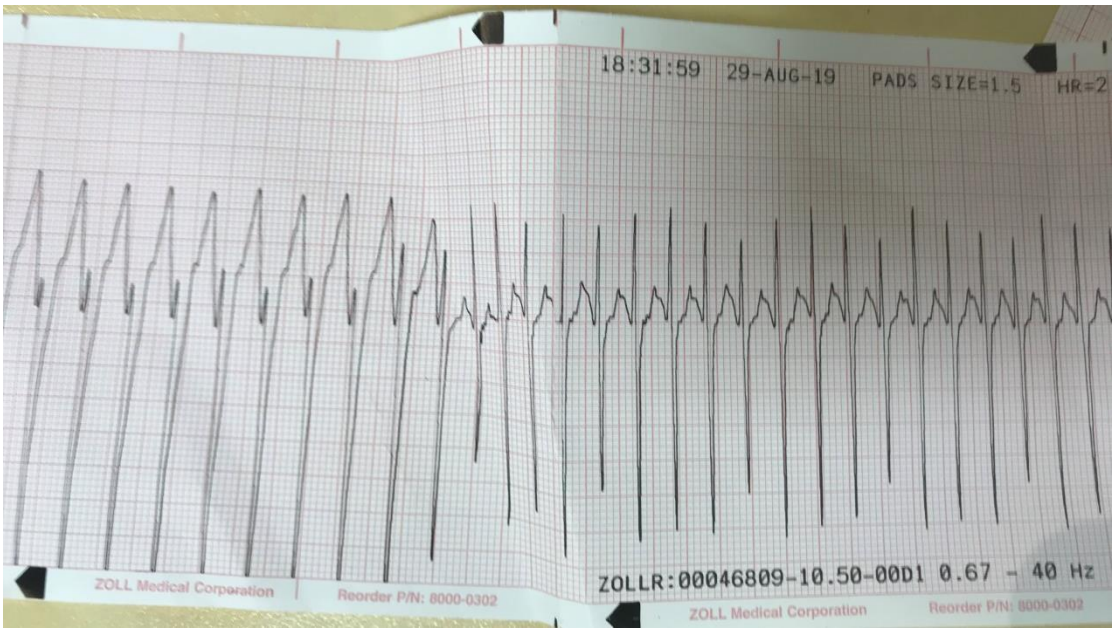
- Reversion to abnormal p-QRS-T morphology: short PR, possible delta wave

Post immersion



- Failed sustained reversion ? underlying pre excitation
- Discuss with cardiology/review tracings post reversion morphology vs pre excite

6. **Cardiology suggest adenosine starting 200mcg/kg bolus. You proceed with this. The below ECG tracing is seen. The child becomes shocked after the adenosine with a BP of 40 and CRT 4s. Discuss your approach?**



- Broad complex tachy likely VT with reversion to SVT

- Manage shock:
- Consider small IVF bolus if not in failure
- Prepare for DC cardioversion

7. **The babies BP and shocked state improves. What would you do from here?**

- Discuss with cardiology – options transfer to site with service and PICU
- Consider amiodarone under specialist advice
- If shocked DC cardioversion