

# Paediatrics

Final year case templates

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# Contents

1. Diarrhoea - 4
2. Idiopathic thrombocytopenic purpura -19
3. Nephrotic syndrome - 28
4. Failure to thrive - 41
5. Tuberculosis - 53
6. Anaemia - 65
7. Asthma - 73
8. Developmental delay - 90
9. Haemophilia - 102
- 10.Neonatal jaundice - 111
- 11.Nephritic syndrome - 125
- 12.UTI - 135
- 13.Acute flaccid paralysis - 146
- 14.Constipation - 159
- 15.Fever with lymphadenopathy - 169
- 16.Haematemesis - 173
- 17.Heart failure - 180
- 18.Neonatal sepsis - 193
- 19.Pneumonia - 200
- 20.Arthritis in children - 211
- 21.Seizures
- 22.Meningitis
- 23.Congenital hypothyroidism

## Diarrhea in children

### History

#### General Information

- Age
  - Aetiology
    - < 2Yrs – Rota Virus, Adenovirus
    - > 2 Yrs – Astrovirus, Calcivirus
  - Complications - Very young children are more susceptible to complications
    - Dehydration
    - Secondary mal-absorption
    - Inter-susception – 5 months to 9 months
- Sex – Females are at increased risk of campylobacter infection
- Geography
  - Patient may be from an epidemic area
  - Low socio-economic status – Slum

#### Presenting complaint

- Loose stools
  - Blood and mucous (Dysentery)
  - Watery diarrhea
- Duration
  - > 14 days → Persistent diarrhea
  - > 1 month → Chronic diarrhea

#### History of presenting complaint

1. Establish diarrhoea
  - Normal bowel habits – Frequency & consistency
  - Describe the change of bowel habits
    - Frequency
      - Should > 3/day
      - More in large intestine infection compared to small intestine
    - Volume
      - More in small intestinal infection compared to large intestine
    - Consistency – Loose stools
    - Colour
      - Rice water stools – Cholera
    - Blood & mucous
    - Expansile
2. Other associated symptoms
  - Vomiting
  - Abdominal pain
  - Fever
  - Loss of appetite

3. Which part of the intestine involved? Small or Large intestine

- Small intestinal symptoms
  - Less frequent bowel opening but more volume loss
  - High risk of dehydration
  - Peri-umbilical or RIF pain not relieved by defecation
- Large Bowel Symptoms
  - More frequent bowel opening & less volume loss
  - Low risk of dehydration
  - Watery stools +/- blood & mucous
  - Pelvic pain relieved by defecation
  - Tenesmus
    - Painful sense of defecation but most of the time nothing passed or only some amount of blood or mucous
    - Child strain → Cry → Small amount of stools passes

4. Find out the cause for the infective diarrhea

- Contact history – family members, siblings, playmates, school mates
- Any other persons with similar symptoms including the mother
- Safety of foods – Whether there is a history of food ate from outside

5. Try to find out the aetiology for the infective diarrhea

- Viral
  - Watery diarrhea
  - Fever before the onset of diarrhea
  - Large volume loss
  - More chance of dehydration
- Bacterial
  - Water +/- Blood & mucous
  - Vomiting before the onset of diarrhea. Fever persisting for sometime after diarrhea
  - Small volume loss
  - Less chance of dehydration
    - Chloera
      - ✓ Profuse watery diarrhea
      - ✓ Mucous plates
      - ✓ Severe dehydration
    - Shigella(Bacillary dysentery)/Salmonella/E.coli (Shigella & E.coli strains can also cause blood & mucous diarrhea)
      - ✓ Frequent small volume mucoid stools
      - ✓ Abdominal cramps
      - ✓ Tenesmus
      - ✓ High fever causing febrile convulsions
    - Clostridium difficile → Pseudomembranous colitis
      - ✓ History of antibiotic therapy – mainly ciprofloxacin

6. Assess the complications

- Dehydration
  - Try to figure-out the amount of fluid loss
  - Oral fluids & food intake
  - Increased thirst
  - Assess the urine output. Reduced or normal
  - Child's general condition – Alert, restless or lethargic
- Malnutrition
  - More applicable to persistent & chronic diarrhea
  - Ask about the food intake of the child.
    - Chronic diarrhea
    - Weight loss
    - Vitamin deficiencies
      - ✓ Water soluble
        - i. Vitamin B
          - a) Anaemia
          - b) Neuropathy
            - ✓ Numbness
            - ✓ Unsteady gait/ataxia
        - ii. Vitamin C – Very rare
          - ✓ Fat soluble
            - i. Vitamin A
              - a) Night blindness
              - b) Gritty eyes, reduced tearing
              - c) Skin keratinization
            - ii. Vitamin D
              - a) Bone pain
              - b) Pathological fractures
              - c) Proximal myopathy
                - ✓ Unable to stand from squatting position
            - iii. Vitamin E – very rare
            - iv. Vitamin K – Bleeding tendency
    - Sepsis & severe systemic infection – High fever
    - Haemolytic uremic syndrome
      - Bleeding manifestations – haematuria
      - Acute renal failure – Oliguria
    - Hypernatremic dehydration – Convulsions

7. To find-out the non infective or extra-intestinal cause for the diarrhea

- Meningitis
  - Fever, Photophobia, neck pain
- UTI
  - Fever, dysuria, frequency

- Drugs
  - Antibiotics, PPI, Cimetidine, Propranolol, Cytotoxic drugs, NSAID, Digoxin, Laxative abuse
- Cow's milk protein allergy
- Inflammatory bowel disease
- Pancreatic insufficiency
- Chronic cholestasis

8. Investigations & treatment for the current episode of diarrhea

- Specially antibiotics
- Preparation & treatment with ORS
- IV fluids give or not

**Past medical history**

- Similar episodes in the past including investigations, treatment & hospital admissions
- Any chronic illness which reduce immunity

**Drug History**

- Antibiotics

**Allergic history**

**Birth history**

**Immunisation History**

- Rota virus vaccination – Not very effective

**Developmental history**

**Dietary History**

- Any history of taking meals from outside
- Who else has taken the same food

**Family history**

- Similar illness among family members
- IBD/IBS

**Social History**

- Socio economic status of the family
- Educational level of the parents
- Sanitary education
  - Water sealed toilet
  - Whether child is using it or not
- General hygiene of the child and family members
  - Washing hands with soap & water before meals and after using toilet
- Hygienic preparation of food
  - Who prepared?
  - Whether foods are covered from flees
  - Preparation of green leaves
  - Washing hands before cooking and preparing milk for child
  - Boiling of milk bottle & teat if the child is on formula feeding

#### Water source

- Drink boiled cooled water or not
- Knowledge of mother regarding the disease
  - How to prevent?
  - Value of oral rehydration

## Examination

### General Examination

- Anthropometric measurements
  - Weight
  - Height
  - OFC
- Describe the findings in centiles
- Apply to waterlow's or Gomez classification
- Malnutrition can be resulted from chronic diarrhea
- Bed side stool inspection
  - Watery
  - Blood & mucous
    - Shigella – Lot of blood
    - Salmonella – Less blood with more mucous
- Neck stiffness & kernig's sign/Brudzinsky sign to exclude meningitis
- General condition
  - Ill looking or not
  - Alert, restless, Lethergic or drowsy
- Assess the dehydration – see in management section
- Febrile or not
- Pallor – HUS
- Rashes – meningitis
- Lymphadenopathy – Infection, HIV
- Features of nutritional deficiencies
  - Subcutaneous fat – Reduction indicate malnutrition
  - Angular stomatitis, Glossitis, and koilonychia – Iron deficiency
  - Brittle hair, bitot spots on eyes, skin keratination, pallor, bleeding manifestations
- Quickly check the vital signs
  - Pulse
  - Respiratory rate
  - Blood pressure

### CVS

- Assess for hypovolaemia

### Respiratory system

- RR
- Respiratory distress

### Abdominal Examination

- Tenderness
- Organomegally
- Perianal excoriation
- Features of intestinal obstruction

### CNS

- GCS

## Investigations

 NOT VERY USEFUL (PLEASE DON'T MENTION INVESTIGATIONS IN CASES, SPECIALLY IF DR. SHAMAN PRESENT. EVEN BU/SE IS NOT USEFUL – Be alert to pick-up hypernatremic dehydration)

### Stool tests

- Stool full report
  - Blood
  - Ova and tropozoids of amoeba and giardia
  - Pus cells – bacterial infections
  - Epithelial cells
  - Fat globules
    - Presence of few pus cells and RBC doesn't indicate a bacterial infection
- Stool culture & ABST
  - If suspect Shigella
  - Stool cultures may be negative with prior antibiotic treatment
- Collection of stools in an young child – Pass stools on a Macintosh to avoid contamination with urine

### Blood tests

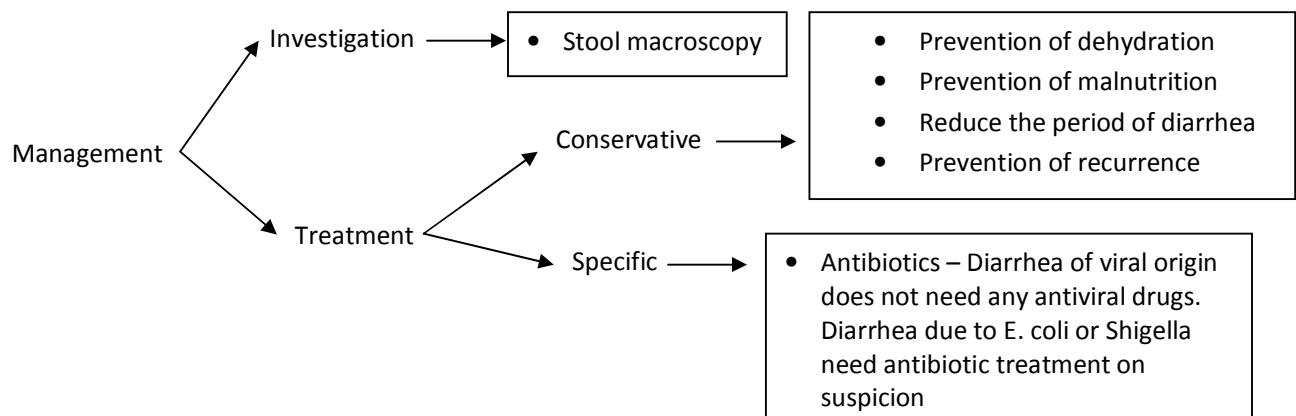
- FBC
  - Hb
  - PCV – Dehydration
  - WBC – Infection
- BU/SE
  - Useful in systemically ill child and in children with moderate to severe dehydration

### If features suggestive of meningitis

- Do urgent lumbar puncture after excluding raised intra-cranial pressure

# Management

## Management of diarrhea in children



## Assessment of a child with diarrhea

- Assessment of dehydration

&lt;5% of BW

5%-10% of BW

&gt;10% of BW

	No dehydration	Some dehydration	Severe dehydration
General Condition	Well, alert	Restless, Irritable	Lethargic, unconscious or floppy
Eyes	Normal	Sunken	Very sunken and dry
Tears	Present	Absent	Absent
Mouth & tongue	Moist	Dry	Very dry
Thirst	thirsty	Thirsty, drinks eagerly	Drinks poorly or not able to drink
Skin pinch	Goes back quickly	Goes back slowly	Goes back very slowly

\* Presence of 2 or more signs in each category will determine the degree of dehydration

\* Skin turgor should be checked midway between the umbilicus and the lateral edge of the body by pinching both skin & SC tissue.

### Estimation of child's fluid deficit

Assessment fluid deficit as % of body weight	Fluid deficit in ml/kg body weight
No signs of dehydration (<5%)	<50 ml/kg
Some dehydration (5-10%)	50-100 ml/kg
Severe dehydration (>10%)	>100 ml/kg

- Check pulse & BP

	No Dehydration	Some Dehydration	Severe Dehydration
Rate	Normal	Tachycardic	Reduced
Volume	Normal	Low volume	Weak
Blood Pressure	Normal	Normal	Reduced
	↓	↓	↓
	Plan - A	Plan - B	Plan - C

## Conservative Management

### Mx of dehydration

Goals -

- \* Correction of existing water & electrolyte deficit
- \* Replacement of ongoing losses

Provision of normal daily fluid requirement

#### PLAN –A

- Replace volume lost at each and every defecation and vomit(ongoing loss) – According to Guide lines

	Less than 2 yrs child	More than 2 yrs child
Small amount	Give 50ml for each	100ml for each
Large amount	Give 100ml for each	200ml for each

⊕ But in our wards volume lost is replaced by 10ml/kgBW for each stool or vomit & should be give within next hour after vomit or bowel opening.

- Replace the amounts given above little by little
- Continue normal diet and breast feeding
- Can give other fluids as well
- If the child vomits --> wait 10mins --> then give ORS more slowly
- Maintenance – Normal requirement

#### PLAN – B

- Correct the fluid deficit
  - 75ml/KgBW in 4 hours
- Review again after 4 hours. If still in the same situation repeat the same dose of ORS within 4 hrs
- If the condition persists even after 8 hrs of ORS start the same amount IV (75ml/KgBW Hartmann's or Normal Saline)
- What to review at ward rounds?
  - Ask from mother about frequency & consistency of diarrhea
  - How is the oral intake?
  - Does he vomit after oral intake?
  - Urine output
- Replace the ongoing loss as mentioned in “Plan A”
- Maintenance

### PLAN – C

- Medical Emergency
- Correct the deficit - Start IV Hartmann's or N.Saline

Give 100ml/Kg Hartmann's(Ringer lactate) or N.Saline divided as follows		
Age	1 <sup>st</sup> give 30ml/Kg in :	Then give 70ml/kg in :
<b>Infants (&lt;12months)</b>	1 hour	5 hours
	<b>6 hours</b>	
<b>Older</b>	30 minutes	2.5 hours
	<b>3 hours</b>	

- Doctor should be stay closer to the patient with monitoring
- Replace the ongoing loss as mentioned in "Plan A"
- Maintenance

### Maintenance

1<sup>st</sup> 10Kg ----> 100ml/kg  
 2<sup>nd</sup> 10Kg ----> 50ml/Kg  
 Next each 1 Kg ----> 20ml/Kg } For 24 Hours

### What to know about ORS ?

- Contents of new ORS solution (Reduced osmolality ORS)

Reduced osmolarity ORS	grams/litre
Sodium chloride	2.6
Glucose, anhydrous	13.5
Potassium chloride	1.5
Trisodium citrate, dehydrate	2.9
<b>Total weight</b>	<b>20.5</b>

Reduced osmolarity ORS	mmol/litre
Sodium	75
Chloride	65
Glucose, anhydrous	75
Potassium	20
Citrate	10
<b>Total osmolarity</b>	<b>245</b>

- What's the difference between the new & old ORS solution  
*WHO formula (old) – Osmolality 311 mOsm/L  
 Low osmolarity formula (New) – Osmolarity 245 mOsm/L  
 And new formula contains glucose to facilitate Na-glucose co-transporter*
- How to prepare
  - Use clean sterile container to prepare
  - Use boiled & cooled drinking water
  - Add the solution to 1L of water and stir well(1L = 2 & half of 400ml Elephant drink bottle)
  - Keep it closed to prevent contamination
  - Discard the solution after 24 hrs
- Importance of giving ORS
  - Replace the fluid & electrolyte
  - Increased absorption (In watery diarrhea brush border Na/Glucose co-transporter is intact. Glucose facilitates Na<sup>+</sup> & water absorption)
  - Reduce the stool output
  - Reduce the incidence of vomiting
  - Less expensive

- *Problems in ORS*
  - *Intolerance*
  - *Preparation problems*
  - *Storage problems*
  - *Perception of not a drug to cure diarrhoea among mothers*

#### ***What to know about Hartmann's solution***

- *What does it contain?*

Each 100ml contains

  - $\text{NaCl}$  : 0.6g
  - $\text{Na Lactate}$  : 0.312g
  - $\text{KCl}$  : 0.04g
  - $\text{CaCl}_2 \cdot 2\text{H}_2\text{O}$  : 0.027g
  - OSMOLALITY : 278mosm/Litre
- *What's the advantage of Hartmann's in diarrhea?*
  - It is similar to plasma in composition

#### **Prevention of malnutrition**

- How does it occur ?
  - Decreased food intake
  - Decreased nutrient absorption, because the contents remain in the GI tract for less period of time
  - Increased nutrient requirement
- Any food can be given
  - ✓ Solid foods – Meat, fish, eggs, rice, kottu, buriyani,
  - ✓ Fruits – any fruit in small amount(Fruits supply electrolytes, specially  $\text{K}^+$ )  
Papaw have to be given in small amount (Increased fibre ----> Induce diarrhea)
  - ✓ Fluids – Rice kanjee, Soup(can add meat, have to add little amount of salt), Fresh fruit juices without sugar(Orange, Lemon)
  - ✓ Youghurt
  - ✓ Milk with little sugar
  - ✓ Continue breast feeding
- What shouldn't be given in diarrhea ?
  - ✓ Artificially sweetened soft drinks (Cordials, Nectar)
  - ✓ Carbonated soft drinks
  - ✓ Coffee
  - ✓ Hard plain-tea
  - ✓ Chocolate
  - ✓ Dark green leaves ( absorbs more water to GIT)

#### **Symptomatic treatment**

- Anti-pyretics –PCM
- Anti diarrhoeal drugs are not recommended

- Anti emetics
  - Avoid metoclopramide & promethazine – Sedative
  - Use Domperidone IV tds – least central side effects

### Specific treatment

- No specific treatment for viral gastroenteritis
- Antibiotics
  - Not routinely recommended
    - Commonest cause is viral
    - Clinically difficult to differentiate bacterial or viral
    - If identified, still difficult to identify the sensitivity pattern of the organism
    - Add additional cost to treatment
    - Risk of adverse reactions
    - Development of resistant organisms
- Given if only bacterial cause is suspected – Shigella and E.coli
  - Oral route is preferred
  - Ciprofloxacin is not given to children <12 yrs (Risk of arthropathy)

#### Antibiotics used in the treatment of specific causes of diarrhoea

First line Antibiotics	Second line antibiotics	Alternative drugs
Furozolidone 2mg/kg/dose - 6 hrly	Mecillinam 10-20mg/Kg/ dose 8 hrly	Ciprofloxacin 5-10mg/kg/dose b.d oral
Nalidixic acid 15mg/kg/dose - 6 hrly oral	Cefuroxime 10-15 mg/kg/dose bd oral 25mg /kg/dose 8hrly (IV)	Cotrimoxazole
Gentamicin 2.5mg/kg/dose (IV) – 8 hrly	Cephalexin 12.5mg/kg/dose 8hrly	

### Complications of diarrhea

- Convulsions
 

Due to

  - ✓ Hyponatraemia
  - ✓ Hypernatraemia
  - ✓ Hypokalaemia
  - ✓ Hypoglycaemia
- Paralytic ileus
  - ✓ Hypokalaemia ----> No action potential generation in GUT ----> No peristalsis ----> Paralytic ileus ----> Present with distended abdomen, vomiting and absent bowel sounds
  - ✓ M<sub>x</sub> – Keep the child NBM  
Replace the K<sup>+</sup> IV
- Transient Lactose intolerance
  - Reduce milk intake
  - Continue breast feeding with control
  - Use lactose free milk if very severe.(Rarely used)

- Acute renal failure
  - ✓ Pre – renal – Give IV fluids
  - ✓ Renal type – They get acute tubular necrosis
    - Restrict fluids
    - Dialysis

### Zn supplementation

- Reduce the severity and the duration of diarrhea
- Reduce the stool bulk.
- Reduce the recurrence for 2-3 months
- Should have to start at the beginning of the illness
- And continue 10-14 days after diarrhea
- 10-20mg/day (5ml contains 22.2mg of Zn)

### Probiotics

- Live commensal micro-organisms in gut which prevents the growth of pathogenic organisms
- Lactobacillus GG is the commonly used
- Reduces duration of diarrhea and less protraction of cases

### Watch for complications especially in shigellosis

- HUS
  - Observe UOP, Bleeding manifestations, BP, Level of consciousness
  - Investigations
    - Blood pressure
    - BU/SE

### When to discharge?

When the child is not systemically ill & tolerating feeds

### Prevention

- Notification
  - If dysentery
- Personal hygiene
- Immunisation
  - Rota virus vaccine

### Parent education

- Knowledge about diarrhea
- How to prevent
- Oral rehydration
  - Rice kanjee, fruit juices, vegetable soup, king coconut water
  - Boiled cooled water
  - Give much as possible with ORS

## Discussion

### Other complicating conditions

#### **Hypernatremic dehydration**

- When give drinks that are hypertonic or salt – soft drinks, commercial fruit drinks (Nectar)
- Cause osmotic diarrhea and hypernatremia  $> 150 \text{ mmol/L}$

#### *Pathogenesis*



#### *Clinical features*

- They have thirst out of proportion to the dehydration. Other signs are minimal.
- Convulsions when sodium level  $> 165 \text{ mmol/L}$

#### *Management*

- Should be corrected slowly over 48 hours
  - 1<sup>st</sup> with N. Saline (To avoid rapid reduction)
  - Then with half normal saline
- Do not exceed the sodium reduction more than 10 mmol/L per 24 hours. If rapidly done → risk of cerebral haemorrhage and convulsions

#### **Severe dehydration & impending shock**

- ABC
- IV access
- Fluid bolus
  - 10 – 20 ml/kg of normal saline or heartmann
- Monitor hourly
  - Vital signs
  - Degree of dehydration and fluid accordingly
- When child can tolerate – give 5ml/kg/hour ORS

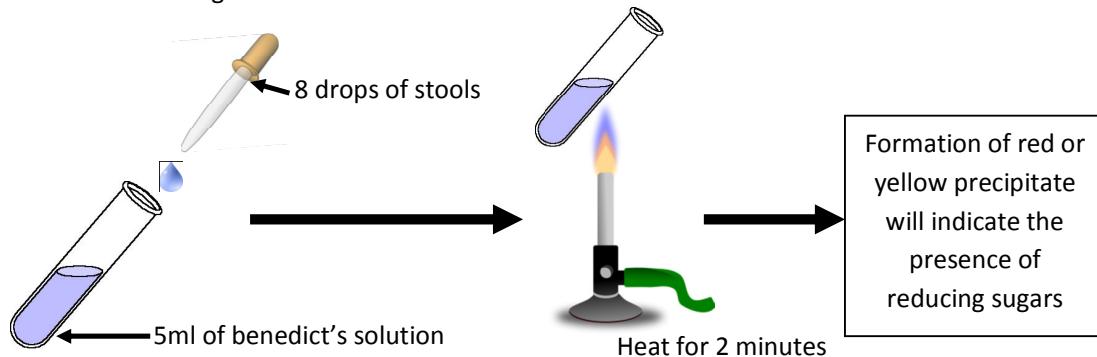
#### **Lactose Intolerance**

##### *Clinical Features*

- Profuse explosive watery diarrhea
- Abdominal distension
- Peri-anal excoriation

*Investigations*

- Stool for reducing substances – Benedict's test

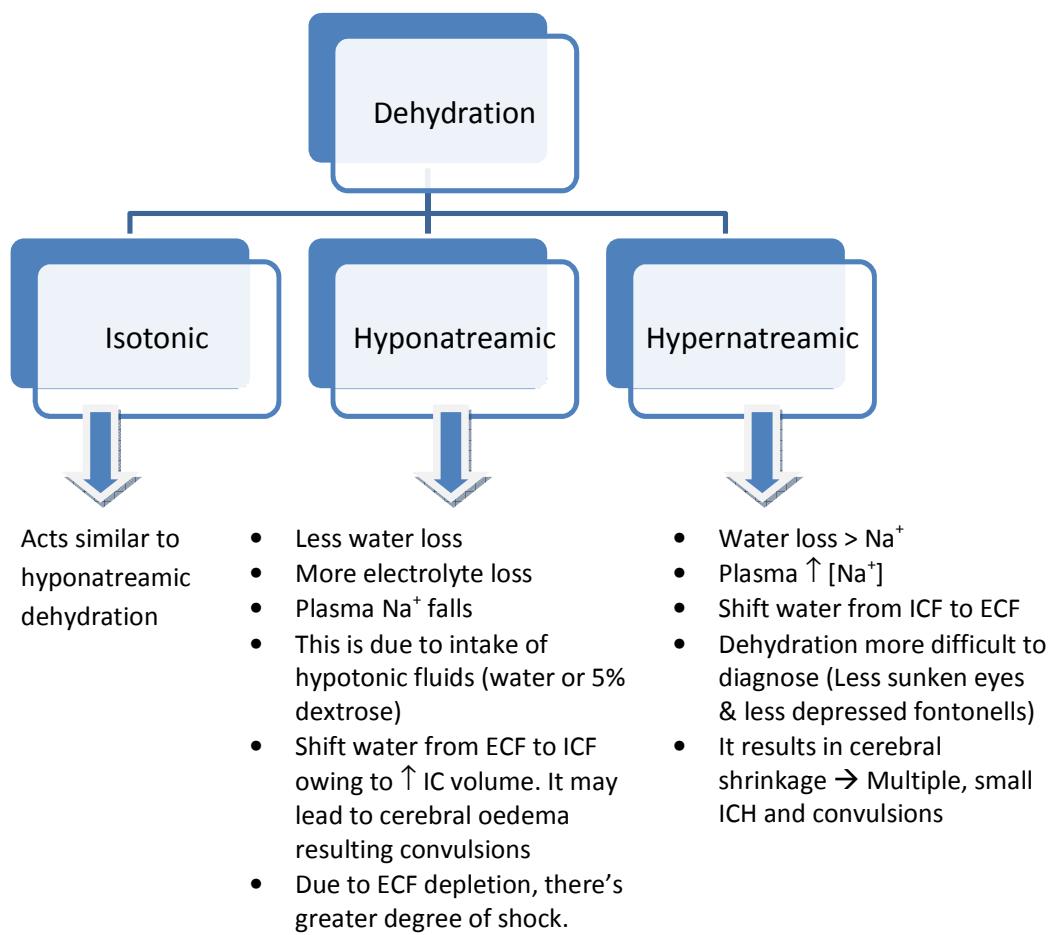


- If stools are semi solid no use of doing the test

*Management*

- Chart weight
  - No weight loss
    - Will resolve spontaneously within 4 weeks
    - Normal diet after that
  - Weight loss
    - Lactose free diet till brush border is recovered
      - ✓ Soya based formula
      - ✓ Cow's milk – Olac
      - ✓ Pre digested protein, fat and CHO – Progetamine
    - Continue breast feeding. But reduce the amount
    - Continue solids which are lactose free
- Treat peri-anal excoriation with gemon violet

## Types of dehydration



## Immune Thrombocytopaenic Purpura

### History

#### Introduction

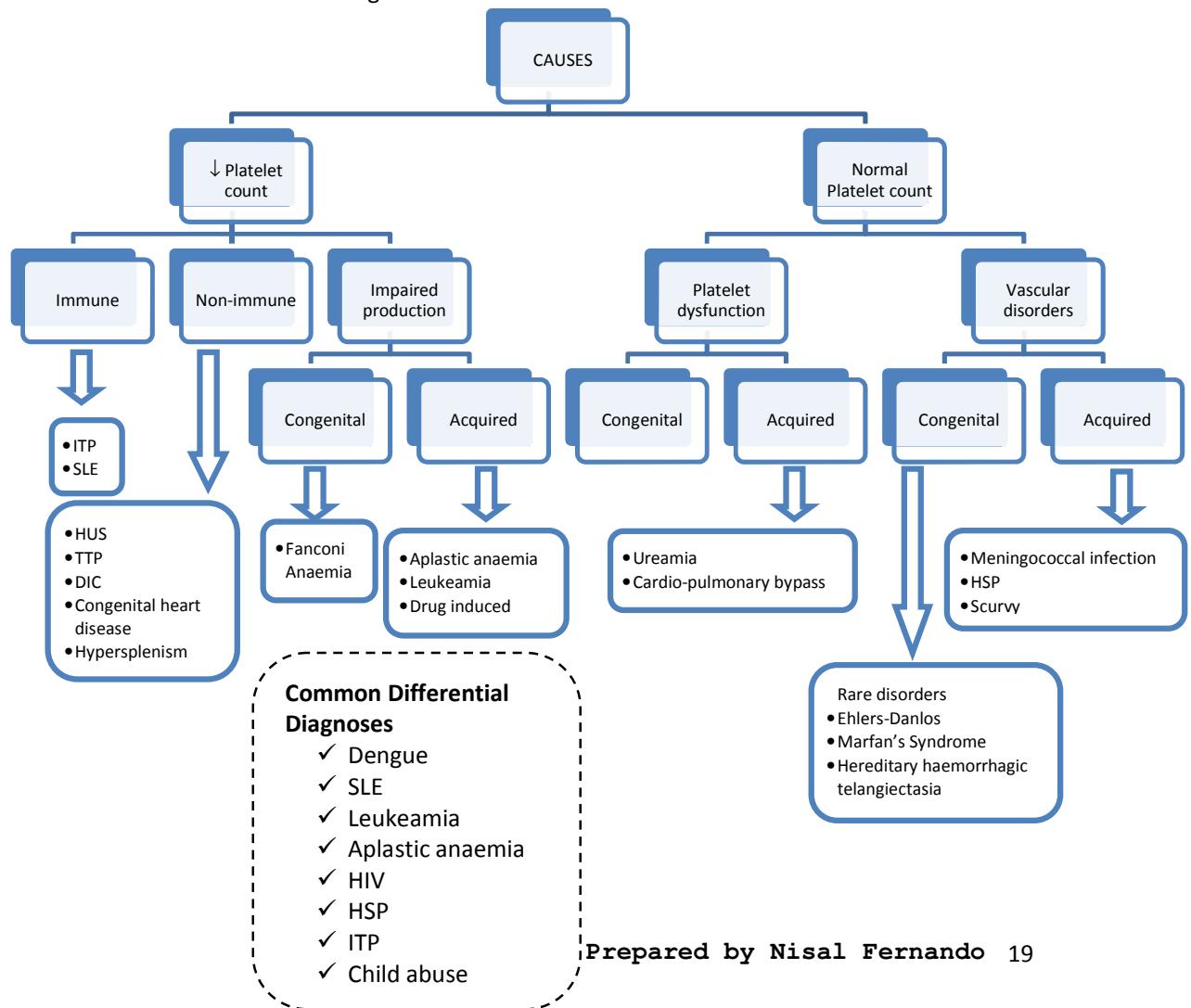
8 Year old boy who was apparently well previously, presents to the ward with gum bleeding while brushing teeth for 2 days. He also got easy bruising even in mild contact trauma.

#### Other presentations

- Superficial Bruising
- Mucosal bleeding such as epistaxis
- Profuse bleeding is uncommon

#### History of presenting complaint

1. Enumerate the presenting complaint
  - Onset of symptoms
  - Duration
  - Sites involved/Areas of bruising or purpura
  - Progression
  - Other associated features – Fever, Rashes
2. Exclude other differential diagnoses



- Dengue fever - Fever, Arthralgia, Myalgia, Retro-orbital pain, Petechiae
- SLE – Malar rash, Photo-sensitive rash, Alopecia, Oral ulcers
- Leukeamia
  - Fever, LOA, LOW
  - Pancytopenia – Recurrent infections, Features of anaemia, Bleeding tendency
  - Radiation exposure
  - Cytotoxic chemical exposure
- Aplastic anaemia
  - Anaemia, Recurrent Infection, Bleeding
  - Radiation & cytotoxic chemicals (Sulphonamide/Chemotherapy)
  - Toxin exposure – Benzene
- HIV
  - Recurrent infections
  - Lymphadenopathy
  - Chronic diarrhea
  - Maternal Hx of HIV or suggestive Hx
- HSP
  - Purpuric rash over the lower legs & buttocks
  - Arthritis of ankles or knees
  - Abdominal pain
  - Oedema
  - Haematuria (Gross haematuria only in 20-30%)
  - Proteinuria – Frothy urine
- **IT IS IMPORTANT TO EXCLUDE CHILD ABUSE**
  3. Ask questions to establish the diagnosis of ITP
    - Viral infection 1-2 weeks before the onset of the disease
    - Petechiae, Purpura, Epsitaxis, other mucosal bleeding
  4. Look for complications
    - Intracranial bleeding (Rare) – Focal signs/Paralysis

#### Past medical history

- Congenital heart disease
- Renal disease (GN) – HSP glomerular nephritis

#### Drug history

- Valproic acid, phenytoin, sulfonamides, and trimethoprim-sulfamethoxazole (**Drug induced thrombocytopenia**)
- Heparin-induced thrombocytopenia (and rarely, thrombosis) is seldom seen in pediatrics, but it occurs when, after exposure to heparin, the patient has an antibody directed against the heparin-platelet factor 4 complex. (**Drug induced thrombocytopenia**)

### Birth history

**Immunization history** – Similar illness following immunization  
Adverse effects following immunization

### Developmental history

**Family history** - Bleeding disorder  
SLE

### Social History

- Income/Family support
- Parental educational level
- How the disease affecting child's day to day life
- Over-protection

## Examination

### General Examination

- Growth – Height/Weight/Head circumference
- Pallor
- Petechial rash/Purpura/Multiple bruises
- Malar rash, Alopecia, Oral ulcers (SLE)
- Generalised lymphadenopathy – HIV, Leukeamia
- Short stature, abnormal thumb, Hyperpigmentation (Fanconi Anaemia)
- Fever, Flushed, Severely ill, Petechial rash –Dengue fever
- Purpura over buttocks & legs – HSP

### Abdomen

- Distension
- Tenderness (HSP)
- Hepatomegaly (Leukeamia/SLE/Dengue)
- Splenomegaly (SLE/Leukeamia/ITP) – Splenomegaly is rare In ITP
  - ➡ In case of hepato-splenomegaly, the picture is more suggestive of leukeamia
- Free fluid – Dengue fever

### CVS

- Pulse – Tachycardia, Low volume (Dengue)
- BP - ↓ Dengue, ↑ in HSP (Proteinuria, Oedema & Hypertension in HSP)
- Capillary refill time (< 2 secs)

### Respiratory

- Evidence of pleural effusions
- Crepts (LRTI)

### CNS

- Focal signs
- Fundus – Retinal haemorrhages

## Investigations

ITP is a diagnosis of exclusion

- Basic investigations
  - FBC
    - Thrombocytopaenia
    - Anaemia – Aplastic anaemia
    - Neutropaenia – Aplastic anaemia
  - Blood picture
    - Leucoerythroblastic blood picture – Leukeamia → Do bone marrow biopsy
- Bone marrow biopsy – If the history of hepatomegaly, marked lymphadenopathy, abnormal blood picture
  - ↑ Number of megakaryocytes – ITP
  - Predominant lymphoblasts → Leukeamia
  - Hypocellular, ↑ fat cells → Aplastic anaemia
- Exclude other DDs
  - SLE – DsDNA/ANA
  - UFR – Proteinuria & Haematuria(Macroscopic only in 20-30%) in HSP
  - S. Creatinine (In HSP → Deteriorating renal function)
  - Coombs test in case of un-explainable anaemia (Evans Xn → Haemolytic anaemia + ITP)

## Management

### *Acute ITP (Common in children- 80%)*

- Benign & self limiting
- Usually remitting spontaneously within 6-8 weeks
- Most children can manage at home
- Treatment is controversial
- Many do not need treatment even if there platelet count is <10,000

### *Indications for treatment*

- Evidence of major bleeding (ICH or gastro-intestinal haemorrhage)
- Persistent minor bleeding (Eg. Persistent oral bleeding)

### *Treatment options (Therapy should be aimed at controlling symptoms and preventing serious bleeding)*

- Oral prednisolone
  - Should only be given as short course, irrespective of the platelet count
  - 2mg/kg/day
  - Problems – Steroid SE

- IV-Ig
  - More rapid rise in platelet count than in steroids (within 1-2 days)
  - IVIG appears to induce a response by down regulating Fc-mediated phagocytosis of antibody coated platelets.
  - Expensive & time consuming
  - If bone marrow biopsy suggestive, it should be done prior starting IVig
  - Dose – 0.8 – 1 g/kg/day for 1-2 days
  - A/E - High frequency of headaches and vomiting, suggestive of IVIG-induced aseptic meningitis.
- Both are equally effective.
- Platelet transfusions are reserved for life threatening haemorrhage as they raise the platelet count only for a few hours.
- IV anti-D therapy
  - For Rh +ve individuals
  - Dose 50 $\mu$ g/Kg
  - Anti-D induces mild hemolytic anemia. RBC-antibody complexes bind to macrophage Fc receptors and interfere with platelet destruction, thereby causing a rise in platelet count. IV anti-D is ineffective in Rh negative patients

*Other measures*

- Parents need immediate 24-hour access to hospital treatment
- Child should avoid trauma as far as possible
- Avoid contact sports while the platelet count is very low.

*Chronic ITP*

- In 20% of children platelet count remains low 6 months after the diagnosis → Chronic ITP
- No treatment unless there's major haemorrhage.
- No long term steroid treatment

*Treatment is supportive*

- Avoid contact sports.
- Continue normal activities, including schooling.
- Need 24 hour access to the hospital
- Children with significant bleeding → Consultant care

*Splenectomy*

- In ITP, the spleen is the primary site of both anti-platelet antibody synthesis and platelet destruction. Splenectomy is successful in inducing complete remission in 64–88% of children with chronic ITP.
- Most effective treatment
- Significant morbidity & may be unsuccessful up to 25% of cases
- Before splenectomy, the child should receive Haemophilus influenza B, pneumococcal and meningococcal vaccines.
- After splenectomy, he or she should receive life-long penicillin prophylaxis.

- Indications
  - The older child (> 4 yr) with severe ITP that has lasted >1 yr (chronic ITP).
  - Whose symptoms are not easily controlled with treatment.
  - Splenectomy must also be considered when life-threatening hemorrhage (intracranial hemorrhage) complicates acute ITP.

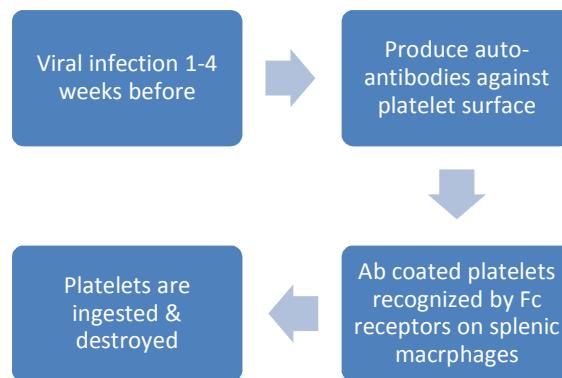
#### *Prognosis*

- Most remit within 3 years from the onset.
- Or stabilize with moderate, asymptomatic, thrombocytopenia.

When ITP becomes chronic, regular screening for SLE should be performed, as the thrombocytopenia may predate the development of auto-antibodies.

## Discussion

### *Pathophysiology of ITP*



### *Most common infectious viruses associated with ITP*

- Epstein-Barr virus - Usually of short duration and follows the course of infectious mononucleosis.
- HIV - Usually chronic.

### *Indications for bone marrow aspiration*

- Abnormal WBC count or differential or unexplained anemia.
- Findings suggestive of bone marrow disease on history and physical examination

### *Bone marrow aspiration findings in ITP*

- Normal granulocytic and erythrocytic series, with characteristically normal or increased numbers of megakaryocytes.
- Some of the megakaryocytes may appear to be immature and are reflective of increased platelet turnover

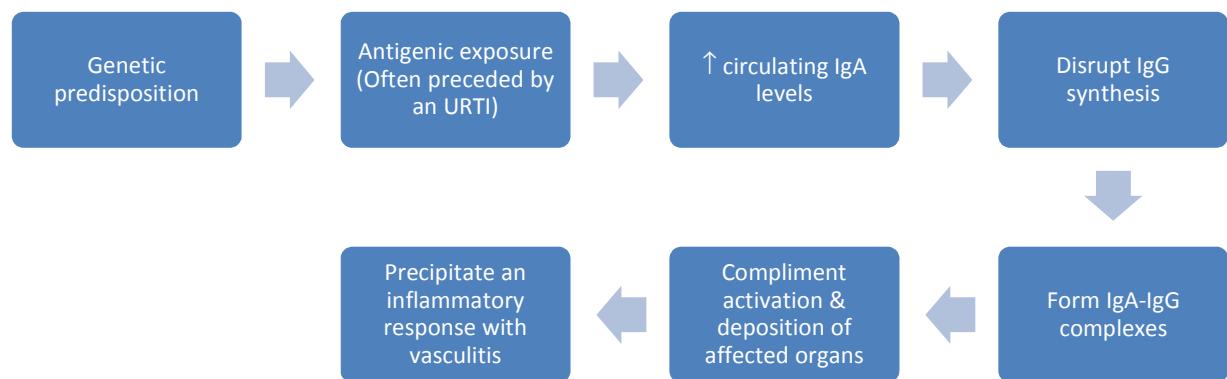
## Henoch-Schönlein purpura

- small vessel vasculitis characterized by
  - Purpuric rash
  - Arthritis
  - Abdominal pain
  - Glomerulonephritis
- HSP nephritis and IgA nephropathy demonstrate identical renal pathologic findings, but systemic findings are only seen in HSP



nephritis.

### Pathology & Pathogenesis



### Clinical features

- Usually occurs between the ages of 3-10 years.
- Twice as common in boys.
- Often precipitated by an URTI (1-4 weeks after)
- Gross hematuria (20–30% of cases)
- Patients may also present with
  - Rash (most obvious feature)
    - Non-blanching
    - Symmetrically distributed over buttocks, extensor surfaces of the arms & legs
    - Trunk is spared unless lesions induced by trauma.
    - Initially be urticarial → Becoming maculo-papular & purpuric
    - Characteristically palpable

- Recur over several weeks
- 1<sup>st</sup> clinical feature in 50%
- Joint pain (2/3 of patients)
  - Knees & ankles
  - Periarticular oedema
  - No long term damage & resolve before the rash goes.
- Colicky abdominal pain → If severe treat with corticosteroids
- Gastrointestinal Petechiae → Can cause haematemesis & malaena
- Colonic intussusceptions can occur
- HSP Nephritis (Common but rarely the 1<sup>st</sup> symptom)
  - Isolated microscopic hematuria
  - Hematuria and proteinuria
  - Risk factors for progressive renal disease are
    - ✓ Heavy proteinuria
    - ✓ Oedema
    - ✓ Hypertension
    - ✓ Deteriorating renal function
  - Usually make complete recovery

➡ All children with renal involvement are followed for a year to detect those with persisting urinary abnormalities.

### *Treatment*

- Uncontrolled studies suggest the potential value of *high-dose corticosteroid and cytotoxic therapy with cyclophosphamide or azathioprine in patients with crescentic glomerulonephritis or significant proteinuria*.
- Addition of dipyridamole and/or heparin/warfarin may provide additional benefit in patients with severe forms of nephritis.
- Some studies suggest that short courses of low-dose prednisone initiated at diagnosis reduce the subsequent risk of developing any clinical signs of nephritis.
- There are no controlled data suggesting that any therapy reduces the risk of progression to severe renal disease.
- Tonsillectomy does not appear to alter the course of HSP nephritis.
- Children with more severe forms of HSP nephritis remain at risk of chronic kidney disease in adulthood.

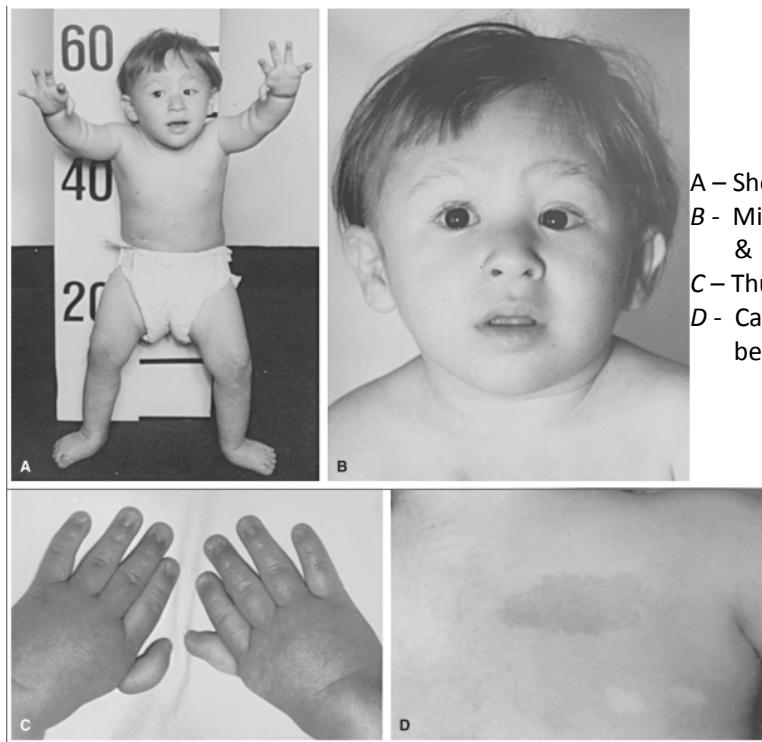
## **Fanconi(aplastic) Anaemia**

### *Aetiology*

- Autosomal recessive inheritance (in almost every case)
- At presentation, patients may have:
  - (1) Typical physical anomalies, but normal hematologic findings or
  - (2) Normal physical features, but abnormal hematologic findings or
  - (3) Physical anomalies and abnormal hematologic findings (classical phenotype - 39% of cases)
- There can be sibling discordance in clinical and hematologic findings, even in affected monozygotic twins.
- Approximately 75% of patients are 3–14 yr of age at the time of diagnosis.

### Clinical Manifestations

- Hyperpigmentation of the trunk, neck, and intertriginous areas, as well as café-au-lait spots and vitiligo, alone or in combination – Most common presentation.
- Short stature - Growth failure may be associated with abnormal growth hormone secretion, or with hypothyroidism.
- Absent radii and hypoplastic, supernumerary, bifid, or absent thumbs are common.
- Anomalies of the feet
- Congenital hip dislocation
- Leg abnormalities
- Males may have an underdeveloped penis
- Undescended, atrophic, or absent testes; and hypospadias or phimosis.
- Females can have malformations of the vagina, uterus, and ovary.
- Many patients have a Fanconi “facies,” including microcephaly, small eyes, epicanthal folds, and abnormal shape, size, or positioning of the ears.
- Approximately 10% of patients are mentally retarded. Ectopic, pelvic, or horseshoe kidneys are detected by imaging, as well as duplicated, hypoplastic, dysplastic, or absent organs.



A – Short stature & dislocated hips  
B - Microcephaly, a broad nasal base, epicanthal folds & micrognathia  
C – Thumbs attached by a thread  
D - Café-au-lait spots with hypopigmented areas beneath

# NEPHROTIC SYNDROME

## USUAL PRESENTATION

- Pre school children
- 3-10 years common
- Male:female-2:1

## PC

- Oedema
  - Frothy urine
  - UTI features
- $\left. \begin{array}{l} \\ \\ \end{array} \right\}$  1<sup>st</sup> episode or relapse

## DDs

- Nephrotic syndrome
- Nephritic syndrome
- CRF
- Heart failure
- Cirrhosis
- Protein losing enteropathy
- Agioedema

## HPC-

1. Describe the oedema
  - Describe the onset of the symptoms and how the mother noticed them
  - Describe the location of the oedema & most affected part
  - Describe the progression of the symptoms over the time
  - Diurnal variation of oedema
  - Aggravating and relieving factors of oedema
  - What the mother did after noticing the symptoms
  - Describe what was done in the ward
2. Ask specific questions to exclude the DDs of generalized oedema

Disease	Features
Nephrotic syndrome	Based on the progression and characteristics of oedema Starts on the peri orbital region and spreads downwards Ask for any change in urine
Nephritic syndrome	Ask for associated red coloured urine and documented elevated BP(ask the mother if she was informed about the elevated blood pressure) HT – Headache, visual blurring
CRF	Ask for weakness, fatigue(associated anaemia) Uremic symptoms Ask for past history of UTI
Heart failure	Past history of cardiac disease, Feeding intolerance, Nocturnal cough, PND, Orthopnoea. More dependent oedema worse towards evening Difficulty in breathing Poor exercise tolerance
cirrhosis	Hx of yellowish discolouration of the eyes Haematemesis, melena Evidence of hepatic encephalopathy
Malnutrition	Chronic diarrhea, allergy history

3. After establishing the diagnosis as NS if FX suggestive of 2<sup>nd</sup> NS try to find an etiology

- Joint pain, abdomen colicks, rashes = **HSP**
- Fever, mala rash, joint pain with morning stiffness = **SLE**
- Infections
  - Blood Tx, diagnosed with hepatitis - **Hep B**
  - Travel to malarial endemic areas - **Malaria**
  - Fever, malaise, generalized lymphadenopathy - **Hodgkin's lymphoma**
  - **Filaria, leprosy, HIV**
  - Drugs - Penicillamine, captopril, gold, NSAID, ethosuximide, methimazole, lithium, probenecid, procainamide, chlopropamide, phenytoin
  - Allergy - bee sting

Suspect of secondary NS when,  
HT  
Haematuria  
Renal dysfunction  
Rashes  
Arthralgia  
Prolonged ↓ of Serum complements(C3)

4. Ask for the complications of NS

- Flank pain with gross h'urea - **Renal vein thrombosis(RVT)**
- Calf pain+/- difficulty in breathing - **DVT and PE**
- Collapse and syncope - **Hypovolemia**
- Fever with abdominal pain - **Subacute bacterial peritonitis(SBP)**
- SOB, orthopnoea - **PE**
- Headache, visual disturbances, seizures - **HT, stroke**
- Severe vomiting - **Intestinal oedema**  
**Gastric irrigation due to oedema**
- Abdominal pain-Hypovolemia
  - SBP
  - Cellulitis
  - RVT

5. If a known pt come with a relapse

Key points-

- Describe the initial episode of oedema and how it was diagnosed
- What happened to the disease over time.DO NOT describe each relapse in detail. only the number.
- Describe the management
  - Mention the drugs used,(albumin given/not)amount
  - Ask for S/E of each medication(cushings)
- Aetiology if possible
- Social Hx-vry important

**PMHx-** Atopy (asthma)

**FHx** - SLE, Asthma Hx

## IMMUNISATION Hx - Pneumococcal

Hib

### SHx-

- Impact on the child
  - playing, amount of school missed, friends
  - Attitude towards the drugs and disease
  - Expectations
- Impact on parents
  - Socioeconomic impact
  - Impact of frequent hospital stays
- Impact on siblings
- Environment
  - Give a brief description of the environment of the household
- Support available
  - Family support
  - Extended family support
- Education of parents
  - Education of the disease
  - Knowledge about the drugs used and importance of proper compliance
  - Side effects of the medication
  - Method of urine testing
  - Knowledge on the diet and life style modifications
  - Identification of a relapse and when to bring child to a hospital
  - Awareness of the complications
- Psychological state and expectations of the patients

## Examination

### General

Fever, pallor-CRF  
Icterus- **cirrhosis**  
Oral ulcers- **SLE**  
Tonsils **infections**  
Cervical lymphadenopathy-**hodgkin's**

Look for features of **steroid toxicity**=

Cushinoid features  
Weight gain and obesity  
Hypertension  
Cataract

Establish the distribution of oedema

Vasculitic rashes in the skin-**SLE**

Weight  
Height  
BMI  
Surface area

### Pay specific attention to following

1. Weight
2. Accurate height
3. BMI
4. Fundoscopy
5. Evidence of infection
6. Evidence of underlying systemic disorder

### Abdomen

Hepatosplenomegaly

Free fluid in the abdomen-ascites, scrotal oedema, peritonitis

### Cardiovascular

Exclude cardiac diseases- pulse, JVP, cardiomegaly, basal crepts

Measure the blood pressure

## Respiratory

Asthma Fx  
Pleural effusions

## DISCUSSION

### Management of Nephrotic Syndrome

#### What's nephrotic syndrome?

- Edema
- Proteinuria >40mg/m<sup>2</sup>/hour or urine protein: creatinine ratio > 200mg protein/mmol creatinine
- Hypoalbuminemia (<2.5mg/dl) <25g/l
- Hyperlipidemia

#### Diagnosis

- ✓ Is based on the clinical presentation of the child and investigation findings
- ✓ The child will present with oedema which is initially noted in peri-orbital region And later involves the dependent areas of the body and is worse towards the afternoon
- ✓ Exclusion of other causes of generalized oedema

#### What are the investigations you will do?

- Investigations to confirm the diagnosis
  - **Urine ward test**(offers a qualitative assessment of the protein to creatinine ratio)  
Is usually more than >+3
  - **Urine full report**-proteinuria ≥2+  
Microscopic haematuria(transient haematuria 20-30%)  
Hyaline casts-NS  
Granular casts-nephritic
  - **24 urine collection for protein estimation**
  - **Serum proteins**- albumin <25g/L
  - **Lipid profile**-elevated total cholesterol, LDL, TG
  - **urine protein to creatinine ratio** -200mg/kg (early morning urine sample)
  - **Urine culture and ABST**

#### Other investigations

- Renal function tests and serum electrolytes
- **Serum complement (C<sub>3</sub>, C<sub>4</sub>)**
- **ESR, ANA**
- **Hep B surface antigen**
- Renal biopsy
- **HIV**

## Role of renal biopsy in nephrotic syndrome

### **Recommendations for renal biopsy.**

1. Onset < 6 months of age
2. Initial macroscopic haematuria (without infection)
3. Persistent microscopic haematuria with hypertension
4. Renal failure not attributable to hypovolaemia
5. Persistently low plasma C3, C4 levels
6. Steroid resistance

### **Preparation of a patient for renal biopsy**

1. Do the initial work up for the patient.  
Investigations-  
Serum creatinine, FBC, bleeding time and clotting profile,  
renal USS scan  
Discuss with the team and arrange a date  
Cross match blood  
Fasting for 6 hours  
2. post op  
Monitor vital parameters  
Collect urine samples  
Complete bed rest until haematuria settles

## **After diagnosis**

### **Classification**

Classification nephrotic syndrome

Primary nephrotic syndrome	Secondary nephrotic syndrome
Minimal change disease (85%)	Secondary to systemic diseases
Focal segmental GN	Infections-hepB, HIV
Membranous	Drugs - penicillamine, gold, captopril, ethosuximide
Mesangio-proliferative	CTD and vasculitis - HSP, SLE

### **DEFINITIONS**

#### **Nephrotic syndrome:**

Oedema, hypoalbuminaemia (<25g/l) and proteinuria > 40mg/m<sup>2</sup>/hour or protein/ creatinine ratio > 200mg/mmol

#### **Remission:**

Urinary protein excretion <4mg/m<sup>2</sup>/hour or reagent strip / sulphosalicylic acid test negative or trace for 3 consecutive days

#### **Relapse:**

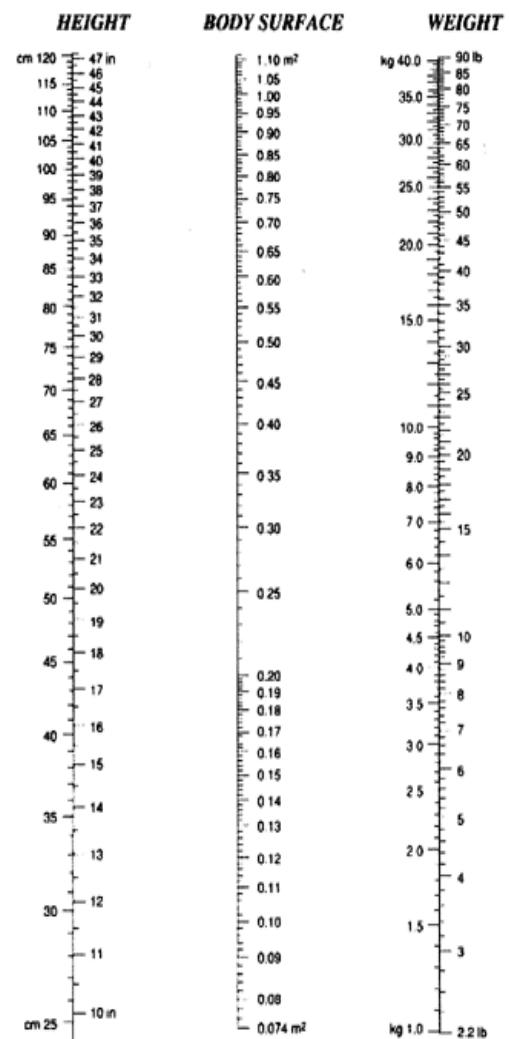
Urinary protein excretion >40mg / m<sup>2</sup>/hr or reagent strip / sulphosalicylic acid test ++ or more for 3 consecutive days or recurrence of proteinuria at any level with hypoalbuminaemia <2.5g/dl and/or oedema having previously been in remission.

## How to manage the 1<sup>st</sup> episode of NS?

### ➤ General management of the child

#### Monitor

- Daily weight & abdominal girth
- Input/put charts
- Urine protein
- Abdominal tenderness
- Fever chart
- BP tds PR and volume
- bed rest
- Protein restriction in the diet
- Fluid restriction
- Normal protein diet with restricted salt until resolution of this period
- IV fluids-
  - if no edema –no fluid restriction
  - if gross oedema
    - calculate the maintenance for 24 hrs and ask to take it orally
  - oliguria
    - measure the UOP and give the fluid thereafter as previous day UOP+ insensible loss (400ml/m<sup>2</sup>/day)
  - until u make a definitive diagnosis (NS/AGN you have to manage fluid cautiously)
- Antibiotics - Prophylactically - oral penicillin (250mg bd for 10 days) antibiotics if any obvious foci (Phenoxyethyl penicillin)
- Pneumococcal vaccine



### ➤ Steroid therapy

- Prednisolone 60mg/m<sup>2</sup>/d given as a single dose in the morning for 6 weeks. Then 40mg/m<sup>2</sup>/d EOD for 4 weeks.
- Then stop abruptly.
- EOD treatment should start on the next day after completion of daily treatment. (No gap)
- Calculate the body surface area by using a normogram which is available in the ward
- Usually pt. responds to steroid after 2-4 weeks.

### ➤ Remission

- Urine protein<4mg/m<sup>2</sup>/h
- Urine protein negative or trace for 3 consecutive days

### Advices you give to parents

- Tell about the disease-NS is a relapsing chronic disease and their support and understanding is extremely needed for the Mx of this disease
- Reassure-progression to CRF is extremely rare with adequate management
- Home based management
  - Give a normal diet with reduced fat and refined sugar
  - Urine ward tests and how to check at home
    - **Examination of urine should be done every morning during a relapse, during inter-current infection or if child has even mild peri-orbital oedema.**
    - **During remission → 2/3 a week**
  - Maintain a diary for protein testing, infections, management changes, school activities
  - Seek early attention for infections
- Educate parents about prednisolone.
- Possible S/E of prednisolone
  - Irritability with behavioral changes
  - Cushingoid features
  - Gastric irritation
  - Affects the growth-growth faltering, cataract, obesity
  - Emergencies-hypertension, hyperglycemia, addisonian crisis
  - Recurrent infections
- Importance of steroids and the risk of addisonian crisis if withdrawn abruptly. DO NOT stop when the child is having any kind of an infection
- Advice on vaccination
  - Avoid live vaccines 3 months after stopping of steroids.
  - During EOD treatment → Killed vaccine can be given
- Try to avoid crowded places due to risk of infection
- Admit to hospital if there's oedema or  $\geq+2$  protein for more than 2 days at home
- Follow up in clinic frequently with the urine protein testing records of the child. Look for any complications of drug therapy

### Method of checking proteins at HOME

- Collect urine to fill 2/3 of the test tube
- Heat the upper part of the tube
- Look for the formation of the turbidity. Add a few drops of Na salicylate/vinegar and see the turbidity disappears(phosphates)
- Quantify the amount of protein by holding the tube up against a newspaper

Nil	-No turbidity
Trace-Turbid	but no diff in reading print
1+	-Turbid. but can read
2+	-Turbid. can't read. but can notice black
3+	-Turbid. can't notice black
4+	-Precipitate

### **At discharge**

- Start the child on short course antihypertensive (in case of hypertension)
- Explain the disease
- Not to restrict the diet(normal diet)
- Get BP measured EOD by the GP
- Review in clinic in 1 week with a UFR

### **Progression of the disease and relevant management**

Condition	Definition	Treatment
Relapse	-Urinary protein excretion >40mg/m <sup>2</sup> /h <b>OR</b> -Urine testing shows 2+ or more for 3 consecutive days <b>OR</b>	-Relapses occur in 60-70% of children with NS -Prednisolone 60mg/m <sup>2</sup> /d (divided 2-3 doses)given as a single dose in the morning until urine is trace or nil for protein for 3 consecutive days. Then 40mg/m <sup>2</sup> /d on EOD for 4 weeks
Frequent relapses	2 or more relapses in the 1st 6 months after diagnosis <b>OR</b> 4 relapses at any 12 month period	- 60mg/m <sup>2</sup> /d until urine is trace or nil for protein for 3 consecutive days. - Then prednisolone is kept on as low as possible on alternate days for 6 months (dose should not exceed 0.5mg/kg EOD)

#### **Frequent relapses:**

Two or more relapses during the first 6 months after the initial episode or four or more relapses within any 12 month period.

#### **Steroid responsive:**

Remission achieved with steroid therapy alone.

#### **Steroid dependence:**

2 consecutive relapses occurring during corticosteroid therapy or within 14 days after its cessation.

#### **Steroid resistance:**

Failure to enter remission following 4 weeks of daily prednisolone at 60mg/m<sup>2</sup>/day.

## When do you consider alternative therapy?

- When the child requires more than 0.5mg/kg of prednisolone on alternative days to remain protein free
- If there's signs of prednisolone toxicity
- Steroid resistant NS

### Alternative therapy

Levamisole

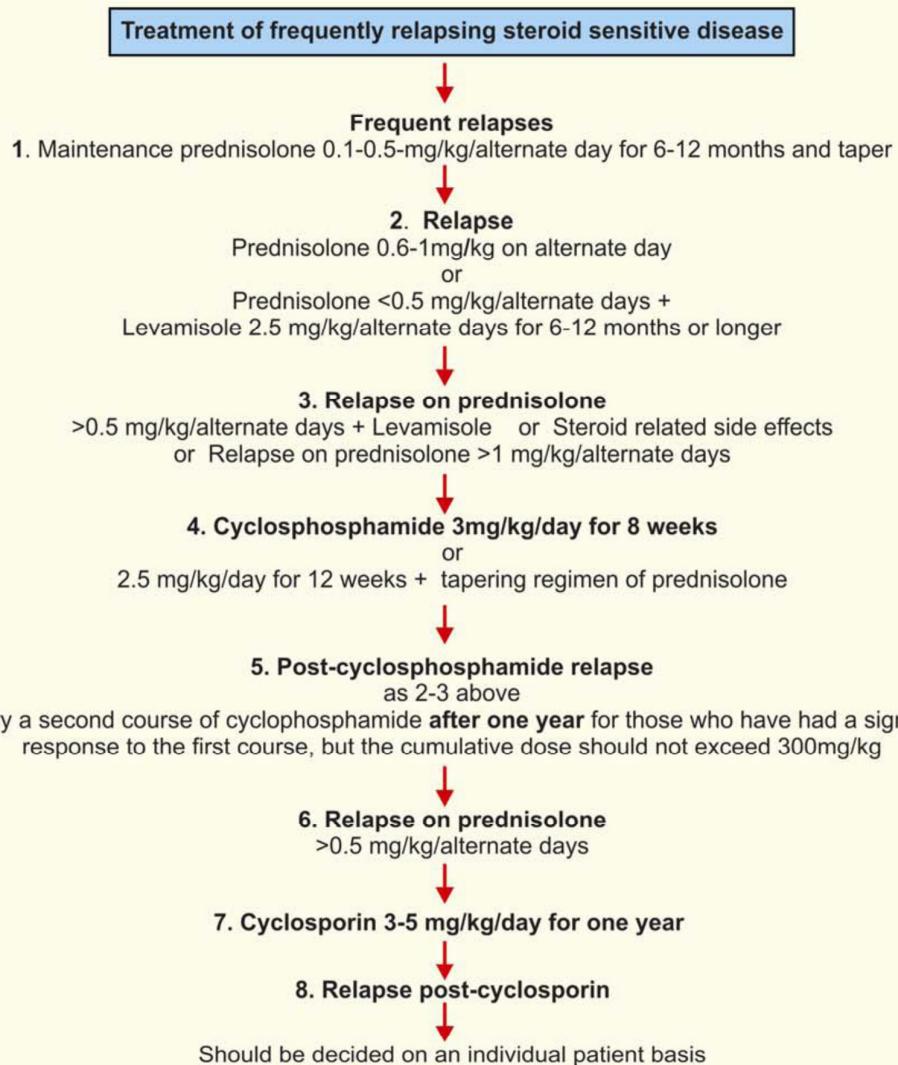
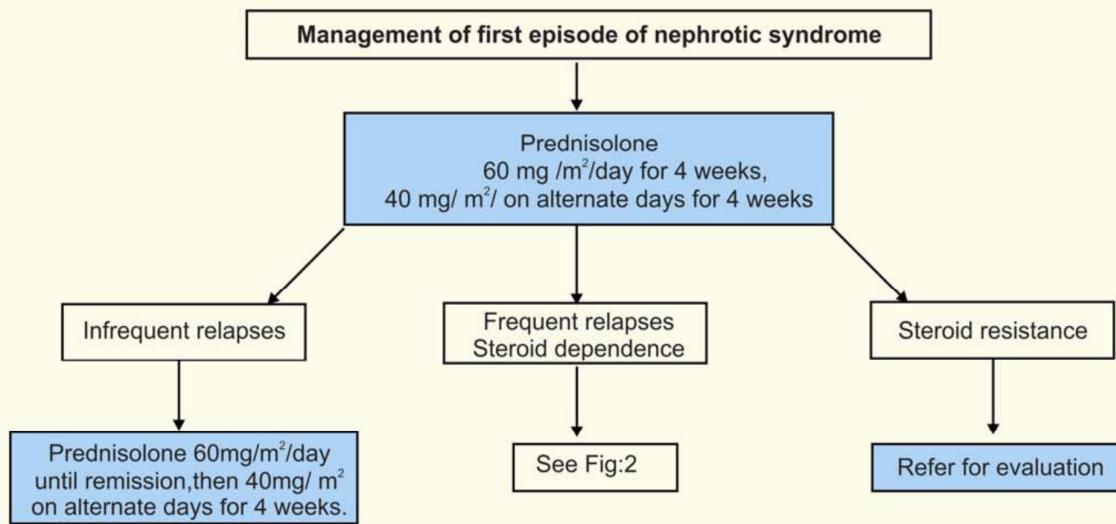
Cyclophosphamide

Chlorambucil

Cyclosporine

Drug and Dose	Monitoring	SE
<b>Prednisolone</b>	-Regular eye checkups(6monthly) -Regular assessment of Ht, Wt, Blood sugar, serum Ca <sup>2+</sup> (3-6 monthly) - Ca <sup>2+</sup> supplementation (can get hypocalcaemia due to hypercalciuric effects of steroids + vitamin D deficiency created by nephritic syndrome)	-Growth retardation -Avascular necrosis of femur -Corneal and scleral thinning, cataract -Peptic ulcer disease -Cushing's Xn -DM -Hypocalcemia
<b>Levamisole</b> 2.5mg/kg EOD for 6-12 months with prednisolone (duration of therapy is 1-3 yrs longer duration in selected Pt)	-WBC 2 weeks after Rx -WBC repeated monthly for 3 months -Then for 3 monthly	-Relatively less SE -Reversible neutropenia -Itchy skin rash and vasculitis * If above side effects occur, discontinue permanently
<b>Cyclophosphamide</b> 0.3mg/kg daily for 8 weeks <b>OR</b> 2mg/kg daily for 12 weeks	-Adequately hydrated before the commencement -WBC monitor weekly when oral Rx & monthly when IV - If leucopenia <3,000 or Neutropenia <1,500 → discontinue & recommence once counts recover to complete the 8 weeks. <b>(Use to induce remission in steroid resistant/dependant nephritic syndrome)</b>	- Neutropenia - Hemorrhagic cystitis - Disseminated varicella - Alopecia - Amenorrhoea - Azoospermia - ↑risk of malignancy
<b>Cyclosporine</b> 5mg/kg/day for 1-3 years	- Renal functions should monitor closely	

## Management of first episode of nephrotic syndrome



## Management of Complications of Nephrotic syndrome

### INFECTIONS

#### *Primary peritonitis*

Children with nephrotic syndrome are prone to infections particularly cellulitis and primary peritonitis. Should a child develop primary peritonitis the antibiotics recommended are parenteral penicillin and a third generation cephalosporin. (x)

#### *Pneumococcal infections*

Children with nephrotic syndrome have an increased risk of developing infections, especially pneumococcal infections. Prophylactic oral penicillin is recommended during treatment of relapses until proteinuria has cleared. Routine immunization with polysaccharide pneumococcal vaccine is currently not recommended.

#### *Varicella infections*

A child who develops overt disease while receiving immunosuppressive medications should be treated with intravenous acyclovir. Patients should be advised to avoid contact and in case of contact advised to seek medical advice immediately. Varicella zoster immunoglobulin, if available, is recommended within 96 hours of exposure, preferably as early as possible.(x)

Complication	Management
<b>Hypertension</b>	<ul style="list-style-type: none"><li>- Should evaluate hypertension carefully</li><li>- It may reflects hypovolaemia (may due to extreme vasoconstrictive response to hypovolaemia or due to the renal pathology itself)</li><li>- Nifedipine, hydralazine, atenolol</li><li>- Captopril is better for proteinuric hypertension</li></ul>
<b>Hypovolemic shock</b>	<ul style="list-style-type: none"><li>- UOP&lt;0.5ml/kg/hour</li><li>- Give normal saline bolus 20ml/kg within 20 minutes.</li><li>- Then start salt free 20% albumin 0.5-1g/kg/day (1g in 5ml)</li></ul>
<b>Peritonitis</b>	<ul style="list-style-type: none"><li>- Start antibiotics</li><li>- IV benzyl penicillin and IV cefotaxime</li></ul> <p><u>Prevention</u></p> <ul style="list-style-type: none"><li>- Oral penicillin 250mg bd during the episode</li><li>- Pneumococcal vaccine,Hib</li></ul>
<b>Thrombo-embolism</b>	<ul style="list-style-type: none"><li>- Anticoagulation initially with heparin and then continue with warfarin 6 months or longer only if there is clinical or radiological evidence of thrombosis</li></ul>
<b>Addisonian crisis</b>	<ul style="list-style-type: none"><li>- Patients on long term steroids</li><li>- Circulatory failure due to adrenal suppression during intercurrent illness</li><li>- Manage with IV fluids</li><li>- Correct blood sugar</li><li>- IV hydrocortisone</li></ul>

## How to manage acute conditions

### hypovolemic shock

#### Symptoms

- dizziness
- severe central abdominal cramps with or without vomiting or back pain
- reduced UOP(1ml/kg/hr)
- cold extremities
- low BP/**reactive HT**

#### Lab findings

- urinary sodium <5mmol/L
- raising PCV

#### Management

- Give O<sub>2</sub> via face mask
- Put a cannula 22G yellow
- Give rapid bolus of IV N/S 20ml/kg over 20 min
- Definitive Tx- infuse salt poor albumin at 0.5 to 1.0 g/kg/dose over one to two hours.  
If salt poor albumin is not available other volume expanders- 4.5% albumin/plasma  
10ml/Kg body wt
- Albumin is preferred over plasma
- Never use frusemide along with albumin/plasma
- Continue to monitor vitals PR,RR,BP,O<sub>2</sub> sat

#### Indications for albumin/plasma transfusions

Our aim should be minimum use of blood products

- Hypovolemia
- Control of intractable oedema

- Why nephritic children are more prone to spontaneous bacterial peritonitis?
  1. Ascites → Protein rich medium → Good source for bacterial growth
  2. Prednisolone (High dose) → Reduced immunity
  3. Increased urinary loss of immunoglobulin & prothrombin factor 'b'
- How to suspect SBP in nephritic children
  1. Fever & physical signs are minimal due to high dose steroid therapy
  2. Lipid catabolism is diminished because of reduced plasma lipoprotein lipase due to increased urinary loss
- What's the reason for hypercoagulable state?  
Nephrotic Xn

Increased prothrombotic factors

## Failure to thrive

- Exclude any organic cause for FTT
- Exclude PEM
- Identify any correctable cause for inadequate diet
- Features of micronutrient deficiencies.
- Complications of malnutrition.

### Definition

Change in the growth / weight crosses 2 major growth centiles.

### Causes

Organic	Non-organic
Inadequate intake <ul style="list-style-type: none"> <li>• Impaired sucking and swallowing</li> <li>• Neuromuscular disorder</li> <li>• Cleft palate</li> <li>• Cerebral palsy</li> <li>• Chronic illness leading to anorexia</li> <li>• Crohn's disease</li> <li>• CRF</li> <li>• Liver disease</li> </ul> Inadequate retention <ul style="list-style-type: none"> <li>• Vomiting</li> <li>• GORD</li> </ul> Malabsorption <ul style="list-style-type: none"> <li>• Coeliac disease</li> <li>• Cow's milk allergy</li> <li>• NEC</li> <li>• Short gut syndrome</li> </ul> Failure to utilize <ul style="list-style-type: none"> <li>• Congenital syndromes</li> <li>• IUGR/ extreme prematurity</li> <li>• Congenital infections</li> <li>• Congenital hypothyroidism</li> </ul> Increased requirement <ul style="list-style-type: none"> <li>• Thyrotoxicosis</li> <li>• Congenital heart disease</li> <li>• UTI</li> <li>• Malignancy</li> <li>• Chronic infection (HIV)</li> </ul>	Inadequate intake <ul style="list-style-type: none"> <li>• Inadequate availability of food</li> <li>• Feeding problems(BF, complementary f)</li> <li>• Unsuitable food</li> <li>• Lack of regular feeding time</li> <li>• Overfeeding</li> <li>• Intolerance of feeding behavior</li> </ul> Psychosocial deprivation <ul style="list-style-type: none"> <li>• Poor bonding</li> <li>• Maternal depression</li> <li>• Poor maternal education</li> </ul> Child neglect

## History

### Presentations

- 1) Poor weight gain
  - 2) Feeding problems
  - 3) Recurrent infections
    - ✓ UTI , diarrhea , RTI , TB
  - 4) Features of protein energy malnutrition
    - ✓ Marasmus
    - ✓ Kwashiorkor
  - 5) Other
    - ✓ Alopasia
    - ✓ Loss of subcutaneous fat
    - ✓ Reduced muscle mass
    - ✓ Dermatitis
- Mention the **duration**

### HPC

- Describe the presenting complaint in detail
  - ✓ Can ask about swallowing , nasal regurgitation
  - ✓ Vomiting after meals
  - ✓ Ask about steatorrhoea
  - ✓ Congenital infections leading to failure in utilization
  - ✓ Chronic disease : congenital heart diseases, UTI, asthma, Hypothyroidism, HIV
- Exclude marasmus or kwashiorkor ( ask the features given below)
- Find out the aetiology
  - ✓ Describe the detected causes in detail
  - ✓ Investigations done and treatment

### Past medical Hx

- Recurrent infections
  - ✓ Diarrhea , UTI, RTI,
- TB
- Measles
- Previous hospital admissions
- Feeding habits during illness

### Birth history

#### ANC

- Maternal age
- Febrile illness with rash
- Fever with lymphadenopathy
- Teratogenic drugs
- GDM ,PIH,
- Maternal renal diseases

Natal

- Term / preterm
- BW
- Asphyxia
- Any congenital abnormalities
- Breast feeding established.

Postnatal

- PBU admissions
- Neonatal sepsis
- Jaundice ,seizures
- Breast feeding , diarrhea after feeding (lactose intolerance)
- Failure to pass meconium
- Distended abdomen
- Pyloric stenosis→ projectile vomiting

Hurshprung's disease

**Developmental Hx**

- Delayed developmental milestones-CP

**Dietary history**

- ❖ **Depends on the child's age.**
- Breast feeding
  - ✓ Exclusive breast feeding up to which age?
  - ✓ Breast feeding habit after starting complementary feeding?
  - ✓ Assess the technique of breast feeding(position ,attachment, led down reflex)
  - ✓ Any problems with breast feeding (poor sucking→duration, continuously or stop in between)
  - ✓ What does the baby do after feeding?
  - ✓ How long the child sleeps after a feed (At least for 2 hrs)
  - ✓ Feeding frequency
  - ✓ Any maternal problems related to breast feeding?
- Formula feeding
  - ✓ When , why , by whom
  - ✓ What is the type, quality and quantity, frequency
  - ✓ Preparation
  - ✓ According to the instruction
  - ✓ Hygienic preparation
  - ✓ Whether a teat was used
- Diarrhea after feeds

- Complementary feeding
  - ✓ When ,what , how
  - ✓ Quantity, quality, frequency
  - ✓ Nutritive value and hygiene
  - ✓ How often breast feeding was done
  - ✓ Any complications
  - ✓ Any allergies
- Older children
  - ✓ Family diet
  - ✓ 3 main meals and 2 snacks
  - ✓ Assess the nutritive value
  - ✓ Quantity and frequency
    - Meat ,fish ,egg per week
    - 24 hour dietary recall

#### Immunization hx

- Any interruptions

#### Family Hx

- Consanguinity

#### Social Hx

- Size of the family ,age ,spacing, details of siblings
- Similar affected children
- Income
- Parents occupation

#### DD according to the age

- neonates
  - ✓ congenital syndromes
  - ✓ perinatal infections
  - ✓ lactation failure
  - ✓ improper formula preparation
  - ✓ psychological problems
- EARLY infancy
  - ✓ Congenital infections
  - ✓ Neurological problems
  - ✓ Recurrent infections
  - ✓ Maternal depression
  - ✓ Improper formula
  - ✓ Child neglect

Amounts of calories in food	
100g of any cereal	360-400KCal
100g of rice (1/2 tea cup)	120Kcal
1 yoghurt	80Kcal
Hoppers 1	80Kcal
4 stringhoppers	80Kcal
Fish /meat	minimum calories
1Egg	80 Kcal

- LATE infancy
  - ✓ Delayed/ inadequate complementary feeding
  - ✓ Recurrent infections
  - ✓ Food allergy
- AFTER infancy
  - ✓ Recurrent infections
  - ✓ Improper diet
  - ✓ Improper meal time

### **Examination**

#### Anthropometric measurements

- Weight
- Height
- OFC
- ❖ Categorize under Gomez and Waterlow classification

### **General**

- Hydration
- Ill/well looking
- Wasted
- Febrile
- Pallor (anaemia)
- Feature of marasmus
  - ✓ thin buttock ,rib buccal fat pads
- features of kwashiorkor
  - ✓ Skin rash , oedema , distended abdomen, thin and depigmented hair
- hypothermia (complication)
- features of other micronutrient deficiencies
  - ✓ angular stomatitis
  - ✓ glossitis
  - ✓ rickets
- features of chronic disease
  - ✓ jaundice
  - ✓ clubbing , cyanosis
  - ✓ CRF features
- Features of Down's Xn , CP
- Focus of infection
  - ✓ Ear discharge

## CVS

- Bradycardia
  - Hypotension
  - Features of CCF –gallop rhythm
  - Murmurs
- } As a complication

## RS

- Signs of infection- crepts

## Abdomen

- Hepatomegaly →non-tender : in Kwashiorkor ,  
→tender :CCF  
➔ congenital infections
- Splenomegaly  
✓ Infections

## Stools examination

## Investigations

### Blood

- **FBC**  
✓ Anaemia, infection, WBC counts (cyclical neutropenia)
- ESR /CRP  
✓ Infection , inflammation
- Random blood sugar
- Septic screen
- Serum electrolytes, Calcium , phosphate ,ALP
- Serum albumin levels
- Stool for parasites
- Liver function tests  
✓ Liver disorder
- Thyroid function tests  
✓ Congenital hypothyroidism
- Exclude HIV ( rapid antigen test)
- Immunoglobulins  
✓ Immune deficiency
- Anti-endomysial and anti-glandin antibodies  
✓ Coeliac disease

#### Urine

- UFR
- Urine culture

#### Stools

- Microscopy

#### Management

#### Calculation of Daily Calorie Requirement in a normal child

- 1<sup>st</sup> 10 Kg → 110 Kcal/Kg
- 2<sup>nd</sup> 10 Kg → 70 Kcal/Kg
- Each subsequent Kg → 30 Kcal/Kg

- **Organic FTT**
  - ✓ Treat the underlying cause
- **Non-organic FTT**
  - ✓ give advice to correct the identified factors which will lead to FTT
- Correct dehydration , hypoglycaemia
- Treat infections vigorously with broad spectrum antibiotics
- Start re-feeding
  - ✓ **Caloric requirement : 150Kcal/kg/day** (for present weight)  
(to maintain life caloric requirement is 110kcal/kg/day)
  - ✓ **Protein requirement : 4g/kg/day**  
(normal maintenance is 1g/kg/day)
- The **expected weight gain is 5-10g/kg/day**
  - ✓ Measure daily weight
  - ✓ Start micronutrient supplements
    - Zn
    - Vitamins
    - Iron is not started initially if the child is having an ongoing infection.
      - Some microbes utilize iron and grow.
      - Lactoferrin is saturated by iron, so the immunity is reduced.
  - ✓ Assess the development of the child
  - ✓ Encourage the activity.
  - ✓ Psychological support.

#### What are the therapeutic foods available in the hospital?

- F 100 (100ml → 100kcal) –milk powder
- F75 – milk powder
- BP100(biscuit ,one biscuit → 150kcal)
- Plumpy nut

#### What are the special formula available?

- Paediasure
- Isokal

### **What is the amount of calories in breast milk or cow's milk?**

- 100ml has 67kcal.

### **What are the complications of malnutrition?**

- Acute infections due to 2ry immune deficiencies
- Recurrent infections
- Dehydration
- Electrolyte imbalance (hypokalaemia)
- Hypothermia
- Hypoglycaemia

### **Prevention of FTT**

- Dietary
  - 3meals and two snacks.
  - Increase number and variety of food offered.
  - Increase the energy density of usual food by adding oil, margarine, butter
  - Give food which is thick in consistency
- Behavioural
  - Have meals at regular times, eaten with other family members.
  - Praise when food is eaten
  - Gently encourage the child to eat, avoid conflict.
  - Never force feed.

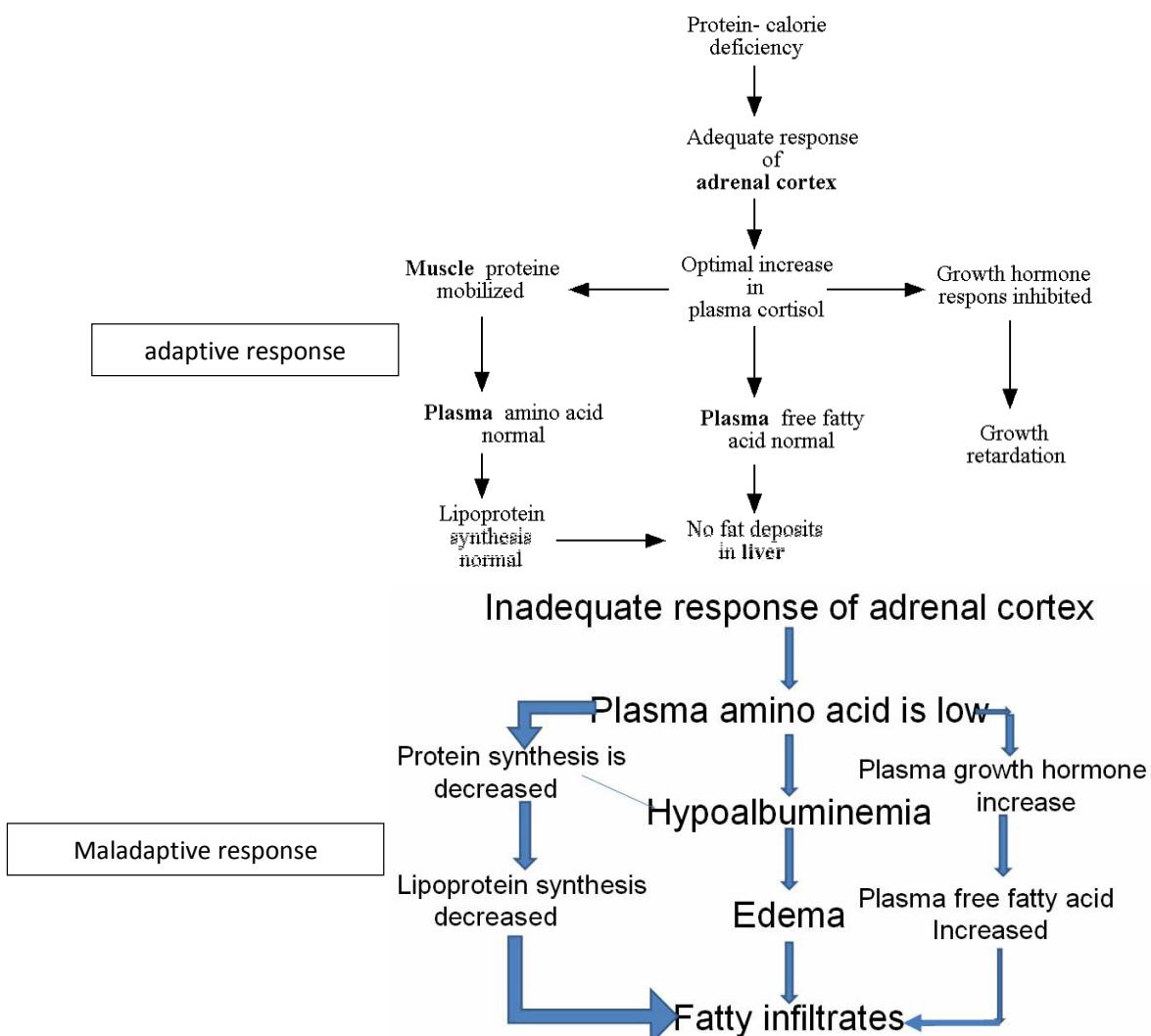
### **Malnutrition**

- Protein energy malnutrition.
- Vit.D deficiency
- Vit A deficiency ( night blindness ,corneal scarring)
- Iron deficiency

### **Assessment of the nutritional status**

- Anthropometry
  - Wt , Ht , mid arm circumference, skin fold thickness
- Lab
  - Low plasma albumin level
  - Low concentrations of specific minerals, vitamins
- Food
  - 24hr dietary recall
  - Dietary history
- Immunodeficiency
  - Low lymphocyte count

### Mechanism of protein energy malnutrition



### Kwarshiokor

#### Definition

- Manifestation of severe protein malnutrition in which body weight is 60-80% of expected and generalized oedema is present.

In addition, there may be;

- May initially present as vague manifestations that include lethargy, apathy, and/or irritability.
- A "flaky-paint" skin rash with hyperkeratosis
- Distended abdomen and enlarged liver (due to fatty infiltration)
- Angular stomatitis
- Hair which is sparse and depigmented.
- Diarrhoea, hypothermia
- Bradycardia, hypotension
- Low plasma albumin, potassium, glucose, magnesium



Flag sign

visuals unlimited



### Marasmus

Severe protein – energy malnutrition in children, with a weight less than 60% of the mean for age.

characterized by ;

- No oedema.
- Failure to gain weight and irritability,
- Looking for food and very alert.
- Followed by weight loss and listlessness until emaciation results.
- Gluteal thickness, hamstring muscle bulk is reduced.
- The skin loses turgor and becomes wrinkled and loose (as subcutaneous fat disappears).
- Loss of fat from the sucking pads of the cheeks often occurs late. thus, the infant's face is initially normal in appearance compared with the rest of the body, later the face become shrunken and wizened(old man's face)
- Infants are often constipated, but may have starvation diarrhea, with frequent, small stools containing mucus.
- The abdomen may be distended or flat, with the intestinal pattern readily visible.
- Muscle atrophy and hypotonia.
- Hypothermia , bradycardia.
- Skin fold thickness and mid arm circumference are markedly reduced.



### Classification of malnutrition

- ✓ Acute malnutrition
  - Wasting
  - The index is weight for height
  - The weight is reduced but the length is preserved.
  - Muscle mass and subcutaneous fat is reduced.
  - Buttocks and rib cage fat are first disappeared. Then buccal fat pad goes.
  - Reversible
- ✓ Chronic malnutrition
  - Stunting
  - Height for age index.
  - Irreversible
  - Body proportions may be normal.
- ✓ Acute on chronic malnutrition
  - Stunting and wasting both present.

### Gomez classification

- Weight of the child is compared to that of a normal child of the same age.
- 50<sup>th</sup> centile is taken for the comparison point.

$$\text{Percentage at } 50^{\text{th}} \text{ percentile} = \frac{\text{Child's weight}}{50^{\text{th}} \text{ centile weight of a normal child}} \times 100$$

- > 90% -normal
- 75-89% - grade I/ mild malnutrition
- 60-74% - grade II/ moderate malnutrition
- < 60% - grade III/ severe malnutrition

### Waterlow classification

- Assess both short and long term malnutrition
- Parameters : height for age  
weight for height

grade	Wasting (Acute malnutrition)	Stunting (chronic malnutrition)
• normal	• > 90%	• > 95%
• mild	• 81 - 89%	• 90 - 95%
• moderate	• 70 - 80%	• 85 - 89%
• Severe	• <70%	• < 85%

$$\text{Wasting} = \frac{\text{Weight of the child}}{\text{Median weight for height}} \times 100$$

Measure the child's height and plot it in the graph and find the age in the 50<sup>th</sup> centile. Then find the weight of the child according to that age by weight for age chart.

$$\text{Stunting} = \frac{\text{Height of the child}}{\text{50}^{\text{th}} \text{ centile height of same age}} \times 100$$

- If the child is **> 11 years** calculate the **BMI**.

## Tuberculosis

Age, living area

### PC

Chronic cough – for months

Low grade fever – for months

LOA and LOW – for months

+/-

Clouding of consciousness – hours to days

Sudden onset generalized convulsion – hours to days

Haemoptysis – for few days

### DD

- ❖ Tuberculosis
- ❖ Bronchial asthma
- ❖ Unresolving pneumonia
- ❖ Congenital heart disease

### HPC

1. Elaborate the PC

#### Cough

- Onset
- Duration – long duration over months
- Any diurnal variation
- Dry / productive
- Haemoptysis
- Associated features
  - Low grade fever
  - LOW
  - LOA
  - Wheezing
- Features of associated extra pulmonary complications
  - ❖ Meningitis
    - ✓ Fever with chills and rigors
    - ✓ Headache
    - ✓ Change in behaviour
    - ✓ Change in consciousness
    - ✓ Sudden onset seizures
    - ✓ Other neurological complains  
E.g.: squints, facial asymmetry
  - ❖ Arthritis
    - ✓ Joint swelling , pain & deformities
  - ❖ Renal
    - ✓ Painless haematuria
  - ❖ Intestine
    - ✓ Diarrhea
    - ✓ Features of obstruction – constipation, vomiting, abdominal distention

#### DD for fever with chills & rigors

- Meningitis
- Pneumonia
- Pyelonephritis
- Malaria
- Abscesses

Hx suggestive of any immunocompromised state

- Long term steroid intake – Nephrotic syn.
- Chronic illness – thalassaemia, haematological malignancy
- Childhood DM
- Cytotoxic drug intake

Ix & Rx done up to now.

## 2. Exclusion of DD

### Bronchial asthma

- Chronic history
- Nocturnal dry cough +/- wheezing
- No associated fever
- Diurnal variation + with an exacerbation in the early morning and with exercise/ playing
- Episodic Hx
- Presence of exacerbating factors – URTI, air pollutants,.....
- PMHx or FHx of eczema, allergic rhinitis, allergic conjunctivitis

### Unresolving pneumonia

- Chronic history
- Fever with chills and rigors
- SOB
- Productive / moist cough – purulent sputum
- Pleuritic chest pain
- Not responding to antibiotics

### Congenital heart disease

- Hx of a heart disease since birth
- Recurrent respiratory tract infections
- Associated LOW
- If severe – Features of heart failure

### **Antenatal Hx**

Maternal TB positivity during the antenatal period, Rx during pregnancy & breast feeding

### **Developmental Hx**

Developmental delay

### **Immunization Hx**

BCG – within the first 24hrs before hospital discharge, whether the scar is present

### **Nutritional Hx**

Malnutrition – *Predispose TB*

### **PMHx**

TB

HIV, Measles, Whooping cough - *Predispose TB*

### **PSHx**

**DHx**

Past Hx of anti TB drugs with a poor compliance

Current compliance to anti TB drugs

AE & any monitoring for AE

- Rifampicin
  - Hepatitis - *LFT*
  - Thrombocytopenia - *FBC*
  - Influenza like syn
  - Haemolytic anaemia
  - Renal failure
  - Red coloured urine
  - Flushing and itching +/- rash
- Isoniazid
  - Hepatitis
  - Peripheral neuropathy
  - Allergies – *Whether advised to avoid tuna fish*
- Pyrazinamide
  - Hepatitis
  - Hyperuricaemia
  - Arthralgia
- Ethambutol
  - Loss of visual acuity
  - Central scotomata
  - Peripheral vision loss
  - Red, green colour blindness

} Optic neuritis

**AHx**

**FHx**

Contact hx of TB – whether lived in the same house for several days

HIV

**SHx**

Poverty & poor socio- economic state

Overcrowding at home

Illumination & ventilation at home

Parental level of education

Social stigma



Edward S. Harkness Eye Institute  
Columbia University

Small pinkish yellow nodules  
in cornea with conjunctivitis



## Examination

Height  
Weight  
OFC }

Gomez classification / Waterlow classification

Level of consciousness  
Neck stiffness  
Kernig's sign  
BCG vaccination scar  
Cervical, supraclavicular lymph nodes  
Sign's of Horner's syndrome  
Mantoux test  
Erythema nodosum  
Phlyctenular keratoconjunctivitis  
Dactylitis – Inflammation of the fingers  
Joint swelling, redness, deformity

}

Manifestations due to immune activation  
or sero conversion

### RS

Evidence of consolidation – localized or diffuse  
Pleural effusion

### CNS

Focal neurological deficits  
Cranial nerve palsies

### Abdomen

Lump  
ascites



Swelling of the fingers - dactylitis

## Investigations

- ❖ FBC - Lymphocytic leukocytosis
- ❖ ESR – very high
- ❖ CXR
  - Apical consolidations and fibrosis
  - No cavitations
- ❖ Mantoux test/ Tuberculin test
  - 2 units (0.1 ml) of purified protein derivative (PPD) of tuberculin
  - Injected ID to the anterior aspect of the left forearm – to the interphase of upper 1/3<sup>rd</sup> & middle 2/3<sup>rd</sup>
  - Read in 72hrs – the transverse diameter of the induration is measured
  - Will be positive only 3-8 weeks after the exposure (as sero conversion takes time)
  - **Positivity only indicates** past infection of any Mycobacterium organism and not the active presence or extent of the TB
  - **A negative test** does not necessarily exclude active TB

- **Interpretation – If BCG is not given**
    - 10mm induration – Positive
  - **Interpretation- when BCG is given**
    - < 5 mm – No infection
    - 5 – 9 mm – Negative → from the BCG
    - > 10 mm – Positive for TB
    - > 15mm – strongly positive
  - **Conditions giving rise to false negativity**
    - Miliary TB
    - HIV
    - Malnutrition
    - Severe bacterial infection including TB
    - Cancer
    - Immunosuppressive drug intake – Steroids, cytotoxic drugs
    - Measles
    - Uncontrolled diabetes
  - **Conditions giving rise to false positivity**
    - Past infection of Mycobacterium tuberculosis or any other Mycobacterium
    - Previous BCG vaccination
- 
- ❖ Sputum for AFB & culture
    - ✓ In children < 5 yrs -1. Early morning gastric aspirate – 3 samples on 3 consecutive mornings with an empty stomach
      - 2. Sputum induction
    - ✓ In children >5 yrs –spontaneous expectoration like adults
  - ❖ CSF analysis
  - ❖ Other available Ix
    - TB PCR
      - NAA – Nucleic acid amplification test
        - Helpful in in the Dx of smear negative PTB & EPTB
      - IGRA – Interferone gamma releasing assay
        - For the Dx of latent TB
        - Until this Ix was available latent TB was solely Dxed using the Tuberculin test/ Mantoux test
        - IGRA is better than Mantoux as the result does not get interfered by the BCG vaccine.
    - Adenosine deaminase test
      - For the dx of TB in pleural, pericardial and peritoneal fluids
      - Also for the early Dx of PTB

## Management

### 1) Drug therapy

#### Pulmonary TB ( PTB)

- ❖ For 6/12
- ❖ Intensive phase – Rifampicin (R), Isoniazid (I), Pyrazinamide (P), Ethambutol (E) – **2/12**
- Ethambutol is not recommended in children < 6 years.**
- ❖ Continuation phase – Rifampicin, Isoniazid – **4/12**

Case definition	Treatment category	Treatment regime	
		Intensive phase	Continuation phase
New cases PTB smear positive PTB smear negative Extra pulmonary TB	CAT 1	<b>2 RIPE</b>	<b>4 RI</b>
Re treatment cases Relapse Treatment after failure	CAT 2	<b>2 RIPES</b> ( S – Streptomycin)	<b>5 RIS</b>

#### TB meningitis

- ❖ For 12 /12
- ❖ **2 RIPS + 10 RI**

#### BCG adenitis

- ❖ For 6/12
- ❖ **Isoniazid 10mg/kg**

### 2) Follow up & compliance

- Directly observed treatment
  - Recommended in the intensive phase of treatment at least for all sputum positive cases
  - Advantage – A pt who misses one attendance for DOT can be traced immediately, counseled & returned to treatment
- Monitor the response to Rx by sputum smear Ex – 2 samples should be collected at each follow up.

Category of Rx	When to do sputum smear
CAT 1 – Smear positive pulmonary TB	End of 2 <sup>nd</sup> month During the 5 <sup>th</sup> month At the end of Rx
CAT 1 – Smear negative pulmonary TB	End of 2 <sup>nd</sup> month At the end of Rx
CAT 2	End of 3 <sup>rd</sup> month (End of 4 <sup>th</sup> month if smear positive) During the 5 <sup>th</sup> month At the end of Rx

### 3) Optimize the nutrition

### 4) Notification

Health 816 – A notification form is used.

## 5) Contact tracing

- ✓ Household contacts should be screened for symptoms of TB (Adults & children > 5 yrs)
  - If symptoms + → Ix with CXR and sputum smear (irrespective of duration of symptoms)
- ✓ Children < 5 yrs – Screened with CXR and Mantoux irrespective of the presence of overt symptoms
  - Mantoux positive with symptoms – full Rx regime for 6/12
  - Mantoux positive w/o symptoms – Isoniazid for 6/12
  - Mantoux negative – Follow up

### Indications for chemoprophylaxis with INAH

- Breast fed infants of sputum smear positive mothers
- House hold contacts < 5 yrs of age of sputum smear positive pts who do not have evidence of active disease.

## Discussion

### ❖ How is a gastric aspiration done

- Done on 3 consecutive days - Early morning samples, with empty stomach.
- Diagnostic yield – 25- 50% only. Therefore a negative smear or culture does not exclude TB.
- Is a low risk procedure for TB transmission – Can be routinely performed at the bed side or in a routine procedure room
- CI
  - Thrombocytopenia
  - Bleeding tendency
- Procedure
  - Fast for 4hrs (Infants – 3hrs)
  - Position the child on his back or side
  - Measure the distance between the nose & the stomach – to estimate the distance to insert the tube
  - Attach a syringe to the NG tube (10 FG or larger)
  - Aspirate 2-5 ml of gastric contents using the syringe
  - If there is no aspirate
    - Confirm the position of the tube
    - Insert 5- 10 ml of sterile water or N. saline and attempt the aspirate again
    - Can repeat the above up to 3 times
  - Transfer the gastric aspirate from the syringe to a sterile sputum collecting container.
  - Add an equal amount of NaHCO<sub>3</sub> to the specimen (To neutralize the gastric acidity & therefore to prevent the destruction of Tubercle bacilli)
- After the procedure
  - Wipe the specimen container with alcohol/ chlorhexidine to prevent cross infection & label the container.
  - Transport the specimen to the lab in a cool box as soon as possible (within 4 hrs)
  - If the transportation is delayed store in 4- 8 °C.
  - Give the child normal food.

❖ How is sputum induction done

- Is an aerosol generating procedure
- Should be performed in an isolation room with infection control precautions.
- Risks
  - Coughing spells
  - Mild wheezing
  - Nose bleeds
  - Transmission of TB
- CI
  - Severe respiratory distress (rapid breathing, wheezing, hypoxia)
  - Intubated
  - Bleeding – low platelets, bleeding tendency, severe nose bleeds
  - Reduced level of consciousness
  - Hx of significant asthma (Dxed & Rxed by a Clinician)
- Procedure
  - Fast for at least 3 hours
  - Administer a bronchodilator (salbutamol) to reduce wheezing
  - Nebulize hypertonic saline (3% NaCl) for 15 min.or until 5ml is fully administered
  - Give chest physiotherapy if necessary.
  - If older children who can expectorate, instruct to do so.
  - For children unable to expectorate,
    - I. Suction of the nasal passage to remove nasal secretions
    - II. Nasopharyngeal aspiration to collect a suitable specimen

❖ About Tuberculosis

- ✓ Caused by Mycobacterium tuberculosis
  - An aerophilic
  - Gram -ve curved rod
  - Acid fast bacilli in ZiehlNeelsen stain
  - Culture is done on Lowenstein Jensen (L J) media
- ✓ Infectious form of the disease – Active pulmonary TB
- ✓ Main source of transmission – air borne droplets
- ✓ Children < 8 yrs are generally **not infectious**. As there is no cavitation formation in children.

❖ Main difference of TB among children and adults

Children	Adults
1 <sup>st</sup> TB	Post 1 <sup>st</sup> / 2 <sup>nd</sup> TB
No cavitations	Cavitations present
Therefore non infectious	Therefore infectious
High risk of developing EPTB ( 90% )	

❖ What is smear positive pulmonary TB

- I. At least 2 sputum smears positive for AFB by direct smear microscopy **OR**
  - II. At least 1 sputum smear positive for AFB by microscopy & chest x-ray abnormalities consistent with active pulmonary TB **OR**
  - III. At least 1 sputum smear positive for AFB by microscopy & sputum culture positive for *M. tuberculosis*
- 1 smear is not taken as positive as the *M. tuberculosis* is a fastidious organism.**  
**Therefore high risk of air borne contamination.**

❖ What is smear negative pulmonary TB

- I. At least 3 sputum smears negative for AFB by microscopy & CXR abnormalities consistent with active pulmonary TB & no response to a course of broad spectrum antibiotics & a decision by a Clinician to treat with a full course of anti tuberculous therapy **OR**
- II. Initial sputum smears negative for AFB but sputum culture is positive for *M. tuberculosis*.

❖ Why is culture important in TB in addition to the smear

- ✓ To identify the antibiotic sensitivity
- ✓ As there can be a positive culture w/o a positive smear

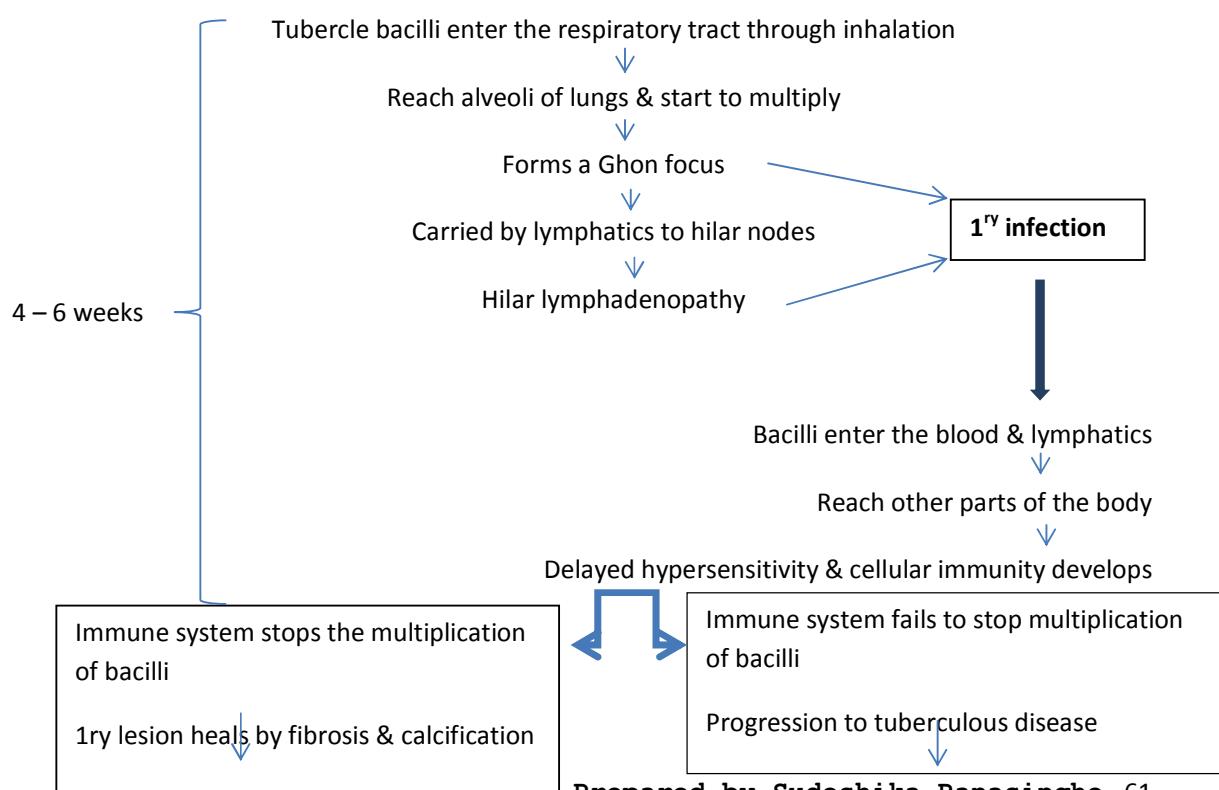
**Disadvantage of the culture**

Takes about 4/12

❖ What is extra pulmonary TB (EPTB)

TB of any organ of the body other than the lung parenchyma  
E.g. : pleural effusion, cervical lymphadenopathy, meningitis.....

❖ Pathogenesis of TB



❖ **Rationale for using multiple drugs**

- I. Different drugs have different actions against different sites
  - ✓ Isoniazid, Rifampicin, Ethambutol, PAS – Against metabolically active, continuously growing bacilli inside cavities
  - ✓ Rifampicin – Also absent against semi dormant forms which are extracellular
  - ✓ Pyrazinamide – Acts in an acidic environment inside cells
- There is no drug which can act against dormant bacilli**
- II. To prevent the development of drug resistant TB

❖ **What is multi drug resistance TB ( MDR TB)**

Resistance to at least Rifampicin and Isoniazid or more of the 1<sup>st</sup> line anti TB drugs.

Problem – Available drugs for the Rx are highly expensive and toxic

❖ **What is extensively drug resistant TB ( XDR TB)**

Resistance to at least Rifampicin and Isoniazid of the 1<sup>st</sup> line drugs

+ To any member of the Quinolone family

+ At least to one of the injectable 2<sup>nd</sup> line drugs ( Kanamycin, Capreomycin, Amikacin)

❖ **Anti TB drug induced hepatitis**

- ─ Caused by Isoniazid, Rifampicin & Pyrazinamide.
- ─ LFT should be done prior to the commencement of Rx & when pt present with hepatitis suggesting symptoms & signs
- ─ A mild transient rise in the serum transaminase levels (< 2-3 folds) may occur during the initial period
- ─ Diagnosis of drug induced hepatitis - > 3 fold rise in liver enzymes
- ─ Mx
  - ✓ All anti TB drugs should be stopped  
Exception - If the TB is so severe – Continue Streptomycin & Ethambutol until LFT is NL.
  - ✓ Admit to hospital
  - ✓ Repeat LFT after 1- 2 weeks
  - ✓ Introduce 1 drug in low dose (Isoniazid)
  - ✓ Gradually introduce full dose over 2-3 days
  - ✓ Monitor for symptoms & signs and do a LFT
  - ✓ If above are normal –Introduce the 2<sup>nd</sup> drug in low dose and gradually increase to full dose over 2-3 days ( Rifampicin)
  - ✓ Monitor & introduce the 3<sup>rd</sup> drug as above ( Pyrazinamide)
  - ✓ If any symptoms & signs develop or if the LFT is abnormal – the drug added last should be withdrawn and it should not be reintroduced. A suitable alternative drug regime should be introduced by a Chest physician.

❖ **Pregnancy and TB**

- ⊕ Dx – depend on sputum examination (as CXR is contraindicated – However if x-ray is essential it can be done with the abdomen covered with a lead apron)
- ⊕ Rx should be started as soon as the Dx is made
- ⊕ Full course of anti TB drugs given
- ⊕ Most 1<sup>st</sup> line drugs are safe in pregnancy, except Streptomycin. Streptomycin causes ototoxicity of the foetus.
- ⊕ Pyridoxine 10mg daily should be given with INAH – to prevent vitamin B<sub>6</sub> deficiency.
- ⊕ Vit. K is given to the baby at birth – As the risk of post natal haemorrhage is high with Rifampicin

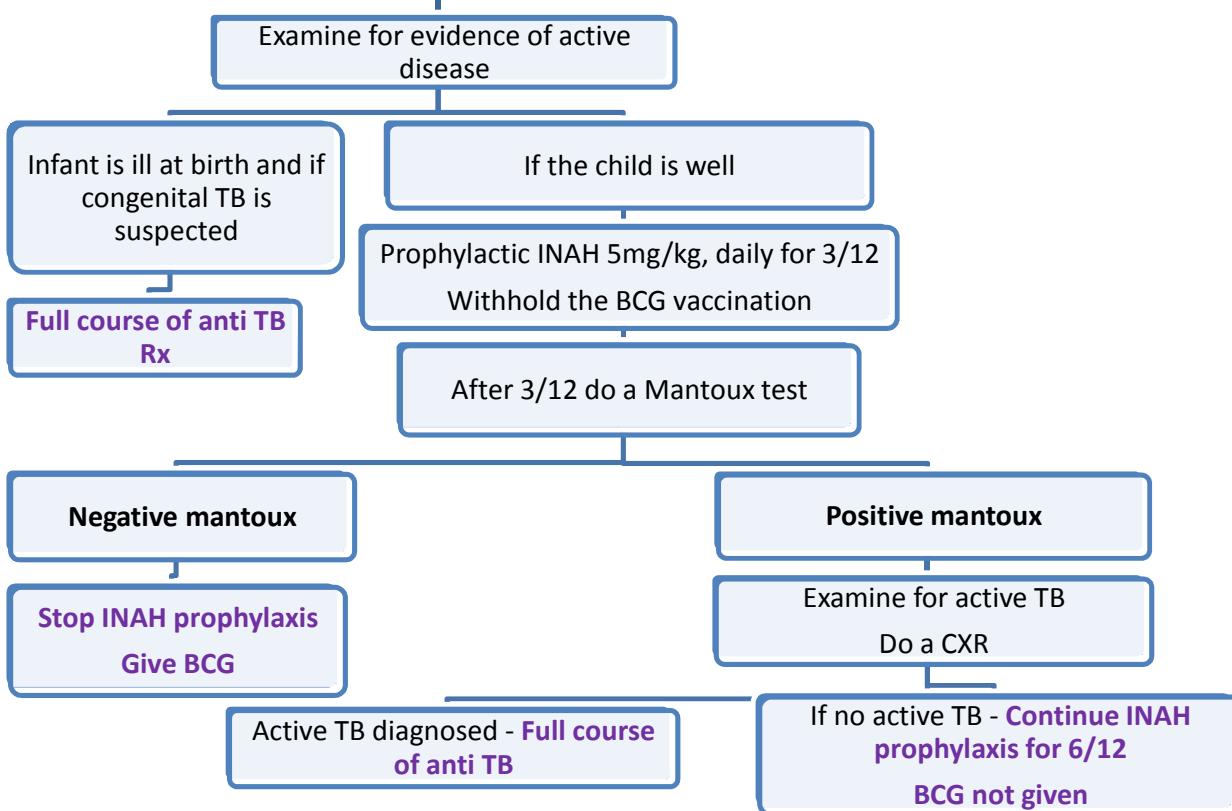
❖ **Rx during breast feeding**

- ⊕ Breast feeding should be continued even though the mother is sputum positive
- ⊕ All anti TB drugs are safe in BF

❖ **Mx of a new born child, of a mother with active TB**

- ⊕ Do not separate the child from the mother unless mother is acutely ill
- ⊕ **If the mother is smear negative & if the infant has no evidence of congenital TB (respiratory distress, lethargy, irritability, poor feeding)**
  - Only BCG is given
- ⊕ **If the mother is smear positive at the time of delivery ;**

**If the mother is smear positive at the time of delivery**



❖ **BCG vaccination**

- ✓ Protects young children from serious disseminated forms of TB
  - TB meningitis
  - Military TB
- ✓ Does not have an impact on the spread of the disease
- ✓ Does not protect the child from developing post 1ry TB in later life

❖ **What are the manifestations due to immune activation in 1ry TB ( manifestations of sero conversion**

- Erythema nodosum
- Phlyctenular keratoconjunctivitis
- Dactylitis
- Mantoux positivity
- Small pleural effusion

❖ **Role of steroids in TB**

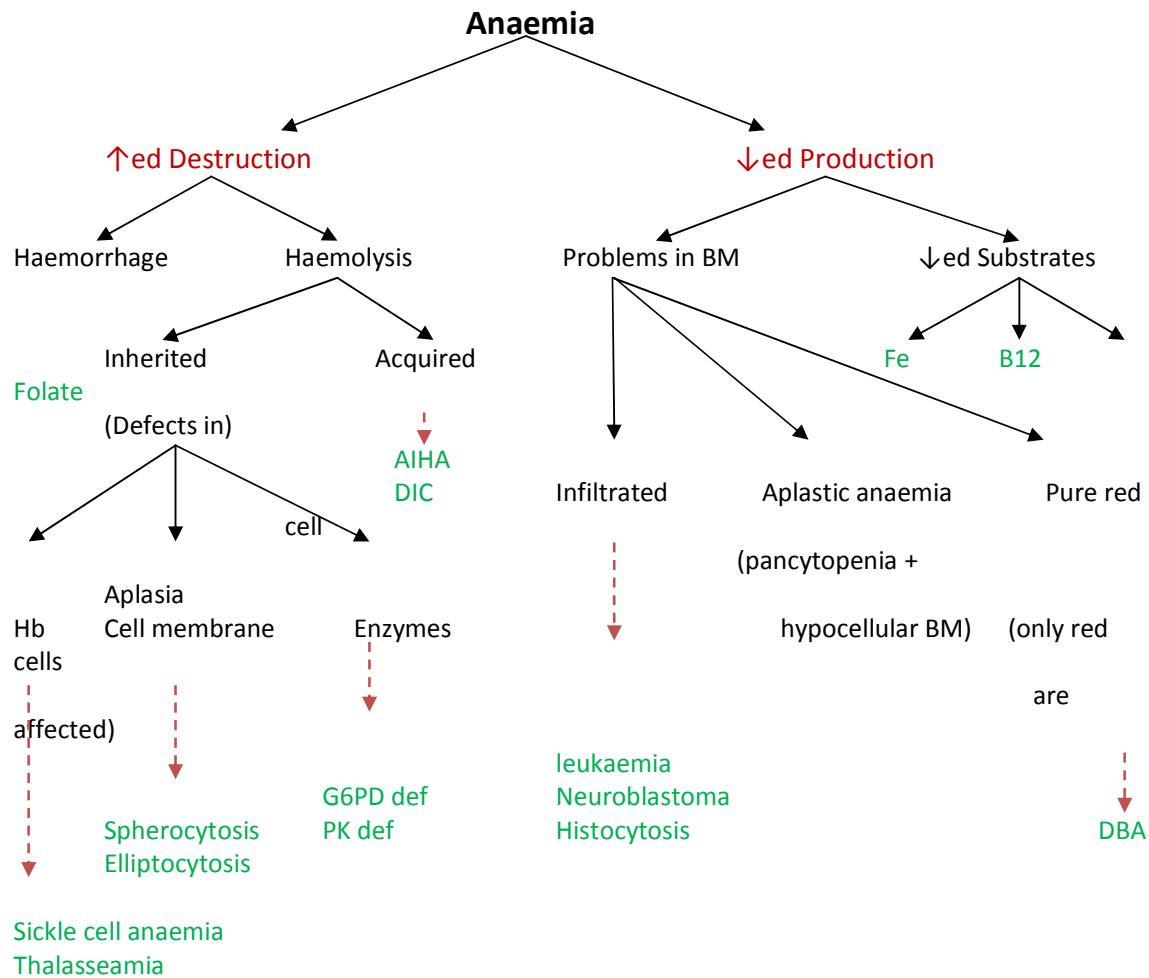
Help to minimize the fibrosis & organ dysfunction. Therefore improves the outcome.

Indications for steroid use in TB

- Adrenal insufficiency due to TB
- TB meningitis with altered level of consciousness and focal neurological signs
- Large and severe TB pleural effusion, which is not responding satisfactorily to anti TB drugs alone.
- Severe hypersensitivity reactions to anti TB drugs
- TB pericarditis, TB lymphadenitis causing compressive symptoms
- TB peritonitis
- Laryngeal TB with life threatening airway obstruction
- Lymphadenitis causing obstruction
- Renal tract TB ( In order to prevent renal scarring)
- Spinal TB with neurological involvement

## Anaemia

### Classification –



### Hx –

This case is usually not given as a separate one, but may be part of discussion in any case.

Child may present with Fx of anaemia – lethargy, poor exercise tolerance, exertional dyspnoea

Describe the onset & progression of symptoms

**Onset of symp.** – Rapid (over days/ 1-2wks) – acute bleeding/ acute leukaemia/ acute haemolysis  
 Insidious – usually – def./ aplastic anemia

**Establish the type of anaemia -**

■ Nutritional anaemia –

Detailed dietary Hx from mother –

Breast feeding

Weaning

24 hr dietary recall of the present diet

Present eating practices of the child

Fe def: Pica, dysphagia & sore tongue

Folate def: Irritable, fail to gain weight adequately, chronic diarrhea

May accompany kwashiorkor, marasmus, or sprue.

B 12 de: - Non specific manifestations such as weakness, fatigue, failure to thrive, or irritability.

pallor, glossitis, vomiting, diarrhea, and icterus.

Neurologic symptoms- paresthesias, sensory deficits, hypotonia, seizures, developmental delay, developmental regression, and neuropsychiatric changes.

Neurologic problems can occur in the absence of any hematologic abnormalities

Diseases of malabsorption – chronic diarrhea, steatorrhoea, LOW, poor growth

■ Chronic blood loss –

- Ask Hx of malaena
- Past Hx of worm infestation & Rx
- Menstruation in older children
- Ask for drug therapy – gastric irritant drugs

■ Anaemia of chronic disease-

Ask past Hx or symptoms of Δed diseases – eg: cardiac disease, CRF, JIA, chronic infections

■ Hemolytic anaemia –

- Ask for previous episodes of anaemia, yellowish discolouration of eyes & darkening of urine
- Past Hx of recurrent blood transfusions, jaundice & bld transfusions at birth
- Family Hx of recurrent bld transfusions & anaemia

■ BM disorder –

- Is this isolated anaemia or part of a pancytopenia?
- Hx of recurrent infections & bleeding manifestations which are associated with anaemia

**Family Hx** - AD – Hereditary spherocytosis (each generation affected)

X linked recess – G6PD

AR – thalasseamia.

Investigated for anaemia/bleeding tendency

**Social Hx** - – Income and education level of parents

Family support

Poverty & negligence

Effect on child's day-to-day activities and school performance

***Ex -***

GE - Growth parameters – Wt, Ht, OFC

III/well

Febrile/not

Pallor & icteric

Nutritional deficiency Fx – specially irondef: Fx

Glossitis

Angular stomatitis

Koilonychias

Facial Fx of Thalasseamia – bossing of frontal bone, enlarged maxilla

Dysmorphic Fx, short stature – in DMD

Lemon hue on face – pernicious anaemia

Purpura & petechiae – in pancytopenia

Lower limb ulcers – in sickle cell anaemia & thalasseamia

Abd – distension

Splenectomy scar

Epigastric tenderness

Hepatomegaly

Splenomegaly - haemolytic anaemia

CVS – systolic flow murmur

Evidence of heart failure – in severe anaemia

CNS – symmetrical sensory loss – touch, proprioception – feet, hands

Intact pain, temp

+ Babinski sign, absent knee jerk, absent ankle jerk – subacute cord degen.

Fundi – papilloedema, retinal haemorrhages, pallor of optic disc

## Discussion –

### ❖ Define anaemia –

Reduction in the Hb concentration below the normal range

Age	Hemoglobin (g/dL)	
	Mean	Range
Cord blood	16.8	13.7–20.1
2 wk	16.5	13.0–20.0
3 mo	12.0	9.5–14.5
6 mo–6 yr	12.0	10.5–14.0
7–12 yr	13.0	11.0–16.0
<b>Adult</b>		
Female	14	12.0–16.0
Male	16	14.0–18.0

### Morphological classification of anaemia –

Microcytic hypochromic – Fe deficiency }  
 β thal – trait  
 Pb poisoning  
 Sideroblastic anaemia  
 β thal – major  
 α thal – trait

Normochromic normocytic anaemia – Acute bld loss  
 Haemolytic anaemia  
 BM failure  
 Renal disease  
 Anaemia of chronic disease

Macrocytic → megaloblastic – eg: B12 / folate def  
 Non megaloblastic – eg: myelodysplasia, aplastic anaemia, pure red cell aplasia

### ❖ How would you investigate a child with anaemia ?

- FBC - Confirm anaemia by Hb concentration  
 Exclude a pancytopenia  
 Red cell indices – guide to classify anaemia based on morphology  
 RDW – quantitative assessment of the various sizes of RBC in blood

- Blood picture – Establish morphology as microcytic hypochromic, normocytic normochromic &
  - Macrocytic.
    - Identify various types of cells – help for Δ
    - ✓ Spherocytosis – microspherocytes
    - ✓ G6PD def – Bite cells
      - Blister cells / basket cells
      - Helmet cells
    - ✓ Sideroblastic anaemia – Microcytic hypochromic anaemia in BP (Ring sideroblasts in BM)
    - ✓ Pb poisoning – Basophilic stripling (blue spots in red cells)
    - ✓ DIC – fragmented red cells, thrombocytopenia
- Retic count - ↓ In Fe def anaemia  
↑ In haemolytic anaemia

**Subsequent Ix will be based on the morphology of anaemia**

Microcytic hypochromic – S. iron, TIBC, **S. ferritin**

Faecal microscopy – hook worm ova, whip worm

Hb electrophoresis

\*if there is ongoing infection, ferritin level is not reliable. Because it is an AFP

Macrocytic – S. folate/ RC folate, s.B12

Schilling test

Anti-parietal cell Ab/ anti-IF Ab

Normocytic normochromic – CRP / ESR

Liver profile

Renal profile

Auto immune profile

BM Ex – aspiration Bx → trephine Bx – aplastic anaemia, marrow infiltration

HA – 1<sup>st</sup> line → Reticulocyte count

UCB (indirect bil) ↑

S. LDH ↑

(Urobilinogen ↑)

Find the cause → coomb's test – indirect - AIHA

s. Haptoglobin ↓ }  
Urine Haemosiderin } IV haemolysis

❖ **Fe deficiency anaemia –**

Most common hematologic disease of infancy and childhood.

### How do you differentiate Fe def: & Thalasseamia?

	Fe deficiency	Thalasseamia
MCV	↓	↓
MCH	↓	↓
Hb	↓	↓
MCHC	↓	Normal
RBC count	↓	Normal / upper normal
RDW	↑	↓
Blood picture	Pencil cells Tear drop cells Target cells - less	More target cells
TIBC	↑	Normal
% saturation	↓	May be ↑
HPLC – HbA2	normal < 3.4%	↑ >3.4%

### How to treat Fe deficiency anaemia ?

#### 1) Fe supplementation –

Rx dose – 6mg/Kg/day

Prophylactic dose – 5mg/kg/day

Oral iron (elemental Fe) – eg: Mumfer, Orofer

Syrup – 5ml → 50mg

Dropper – 1ml → 50mg

Tablet – elemental Fe 60mg

	Tablet (mg)	Fe (mg)
Ferrous sulfate	200	65
Ferrous fumerate	200	65
Ferrous gluconate	300	35
Ferrous succinate	100	35

Rx duration – Hb back to normal + 3 more months (to replenish stores)

Ideally given before meals / 2 hrs after meals

Better to give with Vit C ½ tab & folic acid ½ tab

Brush teeth after giving Fe therapy

Shows a rapid response to Fe therapy

Retic count will go up in 6 hrs

Hb starts to ↑ in 48 hrs

With good compliance – Hb will ↑ in 1g/dl/wk

R/V in 1 month with FBC & bld picture → if there is a ↑ in Hb → R/V in 4 months

Blood transfusions – not I<sup>o</sup>ed, bcoz response to Fe is rapid

Parenteral Fe- causes anaphylaxis, so should not give unless there is a problem with absorption.

Should not give Fe in acute infections, bcoz

Bacteria can utilize Fe & grow faster

Lactoferrin (which is required for immune response) will get saturated with Fe

2) Dietary advice –

Take food rich with Fe (Heme Fe – ↑ absorption) – meat, eggs, fish (tuna, herring, salmon, dried

sprats & other dried fish)

pulses – (cowpea, mung, ulundu, soya )

(Non-heme Fe- ↓ absorption) – dark green leafy vegetables (thampala, sarana, kankun, gotukola)

Add sources of Vit C to the diet - ↑ absorption of Fe

Avoid giving tea to children - ↓ Fe absorption

3) Treat the underlying cause – eg: worm Rx

❖ **What is anaemia of prematurity –**

Anaemia which develops as a complication of prematurity

Occurs between 1-2 months of age

Causes –

Shorter RBC life span

Inadequate erythropoietin production (delayed e'poetin secretion stimulation)

Iatrogenic blood loss – frequent blood sampling while in hospital

Fe & folic acid deficiency – (after 2-3 months)

Rx – blood transfusion

Any premature child should start on,

Fe – from Day 28 -0.3 ml of Fe drops (prophylactic dose)

Should continue until weaning starts

Folic acid – if <34 wks

Multi vitamin – from Day 7

❖ **Congenital Hypoplastic Anemia (Diamond-Blackfan Anemia)**

Usually becomes symptomatic in early infancy - With pallor in the neonatal period

Occasionally may first be noted later in childhood

Over 90% of cases are recognized in the 1st year of life

Most characteristic hematologic features - macrocytic anemia, reticulocytopenia, and a deficiency or absence of red blood cell (RBC) precursors in an otherwise normal cellular BM

Majority of cases(80%) – sporadic

15%- dominant or recessive patterns of inheritance.

Corticosteroid therapy is beneficial in  $\frac{3}{4}$  of patients

Monthly red cell transfusions – for who are steroid unresponsive

# Asthma

- Wheeze
- Cough
- SOB

DD in wheezing child

- BA
- Post viral bronchitis
- Bronchiolitis-<1yr
- Topical pulmonary eosinophilia
- Loeffler's Xn
- Foreign body within bronchus
- Pressure from enlarged mediastinal node

## Diagnosis of asthma in children is based on [4]:

- the presence of key features and careful consideration of alternative diagnoses
- improvement with bronchodilators
- repeated assessment of the child, questioning the diagnosis if management is ineffective

Wheezing phenotypes in children with asthma:

- 1) Transient early wheezing
- 2) Non-atopic wheezing in the preschool child
- 3) IgE-mediated wheezing (atopic asthma).

### 1. Transient early wheezing –

Result from small airways being more likely to obstruct due to inflammation secondary to viral infections. Transient early wheezers have decreased lung function from birth, reflecting small airway calibre.

Main risk factors are the mother smoking during and/or after pregnancy and prematurity. **A family history of asthma or allergy is not a risk factor.**

Transient early wheezing is commoner in males than in females. It usually resolves by 5 years of age, presumably from the increase in airway caliber

### 2. Non-atopic wheezing

In contrast, non-atopic wheezers have normal lung function early in life, but a lower respiratory illness due to a viral infection (usually RSV) leads to increased wheezing during the first ten years of life. This phenotype seems to cause less severe persistent wheezing, and symptoms improve during adolescence.

### 3. IgE-mediated wheezing (atopic asthma).

Atopic wheezing is the usual perception of asthma. Lung function is normal at birth, but recurrent wheeze develops with allergic sensitization, with increased blood IgE and positive skin prick tests to common allergens. Atopic wheezers have persistence of symptoms and have decreased lung function later in childhood. Risk factors for the development of atopic wheeze (asthma) are family history of asthma or allergy and a history of eczema, while exposure to tobacco smoke or prematurity are not risk factors.

In history describe on,

1. Describe present episode in detail
2. Describe present state of disease
3. Past hx and progression up to now
4. Highlight the important events
5. Exclude other DDs and establish probable diagnose as asthma
6. Social hx

- Hx-presents with cough, SOB
- Confirm it is a wheeze and not stridor
- Is he/she is a known wheezer

**Present episode-**

- Onset, duration, progression of symptoms
- If starts with cough-did expose to any known allergen-smoke, wet plant, pollen, pets, food or drugs – aspirin, exercise induce symptoms
- Insidious onset – usually with coryza, symptoms of nocturnal cough, non productive →gradual ↑frequency and severity.
- Was there cyanosis, inability to speak, post jussive vomiting+/- affect feeding, cough in bouts excessive sweating
- Associated
  - Fever
  - Very high fever with SOB-bronchopneumonia
  - Abdominal pain
- Other symptoms - urinary, GI
- Fluid oral intake, UOP
- Activity in between cough like eating, sleeping
- **How it was managed?**
- What was done at home -broncodilators, steroids, nebulized
- What was done at OPD, GP, Ward-IV drugs, nebulize
- **About first episode**
- When diagnosed, where, by whom, age of onset, hospital admission
- Acute exacerbations and hospital admissions, ICU admissions
- **Assess severity**
- How many attacks per month
- Day time symptoms/Night time symptoms
- **Treatments given and compliance**, response to medications (relievers/prophylactics)
- Side effects of the medications given and follow
- Regular clinic follow-up

Disease	Important points in hx
BA	Symptom pattern Intermittent symptoms - child is well in between episodes Diurnal variation, definite trigger factors for episodes and good response to bronchodilators FH of atopy and BA
Structural abnormalities/congenital lesions of respiratory tract	This will excluded as the onset of symptoms is later on in life
TB	Contact hx of TB
Interstitial lung disease	Long standing hx of symptoms, FTT
Heart failure	Hx of cardiac disease, reduce exercise tolerance Orthopnoea (in older child)
GORD	Symptoms associate with meals, associate regurgitation
Recurrent aspiration	Risk factors for aspiration - BF position, Tracheo-oesophageal fistula, cleft palate
foreign body inhalation	Hx of FB inhalation
Rare causes-cystic fibrosis, dyskinesia, immune deficiency	Recurrent LRTI, chronic sinus infections

**PMHx-**

- Past hx of heart disease, urticarial, eczema, catarrh
- Contact hx of TB

**Birth history –**

- Prematurity, RDS, long term intubation, Positive pressure ventilation
- Maternal age at birth (<20yr↑risk)
- BW<2.5KG-↑Risk
- Maternal smoking during pregnancy

**Development Hx (chronic disease)-**

- Grade, school performance are good, development is age appropriate

**Immunization-**

- HIB
- Age appropriate
- Done at MOH office/school
- Adverse effects following immunization

**Dietary History**

- Any precipitating foods
- Restriction of foods
- Artificial foods with colouring – Precipitate asthma

**Drug hx-**

- Is on long term prophylactic or only during attacks
- Takes what - steroid, bronchodilator
- Take how-inhaler+/-spacer, nebulize, oral
- For how long
- Compliance of drugs - check the technique, Cleaning of spacer
- S/E of drugs- tremors, palpitations, ↑wt
- Any other drug which can predispose to asthma- NSAID's, β blockers, penicillin
- Theophylline – oral maintenance
- Steroids - dose

**FHx-** BA, atopy, eczema, catarrh/nasal polyp/allergic rhinitis

**Social hx-**

General introduction of family	Occupations, income-poverty-↑
Impact on child	Playing(exercise induced brocho-spasms), sports, school missing, diet-food restrictions/cow's milk intolerance, day to day life-interaction with others, sleeping (Nocturnal cough)
Impact on parents	Socio economic impact of disease, impact of frequent hospital stay
Impact on siblings	Care, parent hospital stay
Impact on normal family activities	Can family do what they did earlier
Environment	Urban, rural Describe the layout of the house, describe the surrounding environment-main roads, dirt and dust, factories, smokes from industries, bakery, massy lands House-floors how often swept and mopped Windows and available ventilation, how many people sleep in one room, windows close/open at night Sleep in bed/floor, polythene covers to pillow, cotton cover, mattresses, how often they change Using coils mats, book rack, carpets- how often dusted cooking fumes-firewood, kerosene kitchen-separated, attached, chimney smokes comes in to house smoking in house, out side also pets/cockroach ,cob webs soft toys of child environment of school nearest 24hr hospital and transport to OPD/GP with nebulization facility
Supports available	Family supports Extended family support Medical facilities available

Education of parents	Drugs and when to use them Difference between preventer and reliever medications Inhaler devices and how to use, clean them, washing mouth after use inhaler Myths regarding asthma-foods How to recognize acute exacerbation of asthma and what to do then When to bring child to hospital
Psychological state and expectations of the parents/patient	
Drugs	Stock, how they buy drugs-over the counter without prescription
Food	Restrictions, yellow dye-tetrasine/monosodium gluconate(msg)

#### **Examination-**

##### General examination

- Anthropometry- plot wt and height of the child on a centile chart.
- Ask mother for the CHDR- FTT as an alternative diagnosis to BA
- Position, drowsy, confused
- Dyspnea, cyanosis, dehydration, restless sweating, use of accessory muscles for respiration (nasal flaring, mylohyoid), can the child talk sentence, cannula
- Febrile/temp chart
- Look for growth faltering- steroids
- Other evidence of steroid toxicity- cushinoid features striae, truncal obesity, moon face
- Cataract- steroid
- Nose- polyp, nasal discharge
- Mouth and throat- oral thrush
- Cervical LN- URTI, TB
- Clubbing- indicate alternative diagnosis/cyanotic heart disease
- BCG scar
- Ankle/ sacral oedema- cardiac disease
- Skin rash-atopic eczema

##### **Respiratory-**

- Obvious chest deformity
- Evidence of respiratory distress- RR, IC/SC recessions
- Evidence of chronic hyper inflated lung fields- ↑AP diameter, barrel chest, Harrison's sulcus
- Impaired liver and cardiac dullness, liver pushed down, trachea, apex beat -shifted due to push, hyper resonance in percussion
- Rhonchi and crepitations
- Reduce intensity of breath sounds, VB/BB

##### **Inhaler technique of the child**

**CVS-**

- Pulse-tach;brady
- Bounding pulse - CO<sub>2</sub> retention
- Pulses paradox
- BP
- Murmurs

**Abdomen-** Liver pushed down-palpable

**PEFR**

**Complications-**

Due to disease-

- Life threatening asthma
- Respiratory failure- type 1 → type 2
- Pneumothorax
- Lung collapse

Due to treatment-

- Growth retardation-chronic disease/steroids
- Cushing's
- Social stigma
- Emotional problems
- Economical problems

**Investigations-**

- PEFR(<50% of normal)

**Diagnosis of asthma using PEF<sup>4</sup>**

$$\text{Percentage PEF variability (Amplitude \% best)} = \frac{(\text{Highest-lowest})}{\text{Highest}} \times 100$$

An example is illustrated below:

$$\text{Highest PEF} = 400 \text{ L/min}$$

$$\text{Lowest PEF} = 300 \text{ L/min}$$

$$\text{Amplitude} = 400 \text{ L/min} - 300 \text{ L/min} = 100 \text{ L/min}$$

$$\text{Percentage PEF variability (Amplitude \% best)} = \{100 / 400\} \times 100 = 25\%$$

- CXR(↑inflation, horizontal ribs)-
  - When diagnosis is doubted
  - Severe asthma not responding to Rx- pneumonia/pneumothorax as complication
- Allergic skin test-not available in SL
- Pulse oxymetry
- ABG
- WBC/DC - Infection
- ESR/manteaux

**Management-** If child has features across categories. Manage according to the severest.

#### Severity of an acute exacerbation

	Mild		Moderate		Severe	
	< 2 year	≥ 2 years	< 2 year	≥ 2 years	< 2 year	≥ 2 years
<b>Age</b>	< 2 year	≥ 2 years	< 2 year	≥ 2 years	< 2 year	≥ 2 years
<b>Activity</b>	Normal	Normal	Disturbed	Disturbed	Severely disturbed	Severely disturbed
<b>Feeding</b>	Normal	Normal	Disturbed	Disturbed	Severely disturbed	Severely disturbed
<b>Speech</b>	-	Normal	-	Halting	-	1-2 word dyspnoea
<b>Audible wheeze</b>	Nil	Nil	Present	Present	Marked	Marked
<b>Cyanosis</b>	Nil	Nil	Nil	Nil	Present	Present
<b>Respiratory rate/min</b>	< 50	< 40	50-60	40-50	> 60	> 50
<b>Use of accessory muscles</b>	Nil	Nil	Present	Present	Marked	Marked
<b>Chest in drawing</b>	Nil	Nil	Present	Present	Marked	Marked
<b>Air entry</b>	Good	Good	Good	Good	Poor	Poor
<b>Lung signs</b>	+	+	++	++	+++	+++
<b>Pulse/min</b>	< 110	< 100	110-130	100-120	> 130	> 120
<b>PEF</b>	-	70-90% of predicted	-	50 to 70% of predicted	-	< 50% of predicted
<b>SpO<sub>2</sub></b>	>92%	>92%	≥ 92%	≥ 92%	< 92%	< 92%

## MANAGEMENT

### Goals of asthma therapy in children [4,28]

Minimal, ideally no, symptoms during the day or at night

Minimal, ideally no exacerbations

Minimal use or no necessity for the use of reliever short acting  $\beta_2$  agonist  
FEV<sub>1</sub> and/or PEF over 80% of personal best or predicted normal

Minimal, ideally no adverse effects from medications

Normal activities and rare school absences

Optimum growth of the child

Minimal effects on other family members

#### Non pharmacological management



#### Primary prevention

- Breast feeding – most beneficial in children with maternal atopy.
- Hygiene hypothesis e.g. exposure to infections at early age reduces the risk.
- Maternal smoking in pregnancy increases the risk.

#### No clear benefits

- Avoidance of postnatal allergen exposure
- Modified infant formulae (hydrolysate of whey/casein or soy formulae)  
[29,30,31,32,33,34,35,36,37,38,39]

#### Pharmacological management See overleaf



#### Secondary prevention

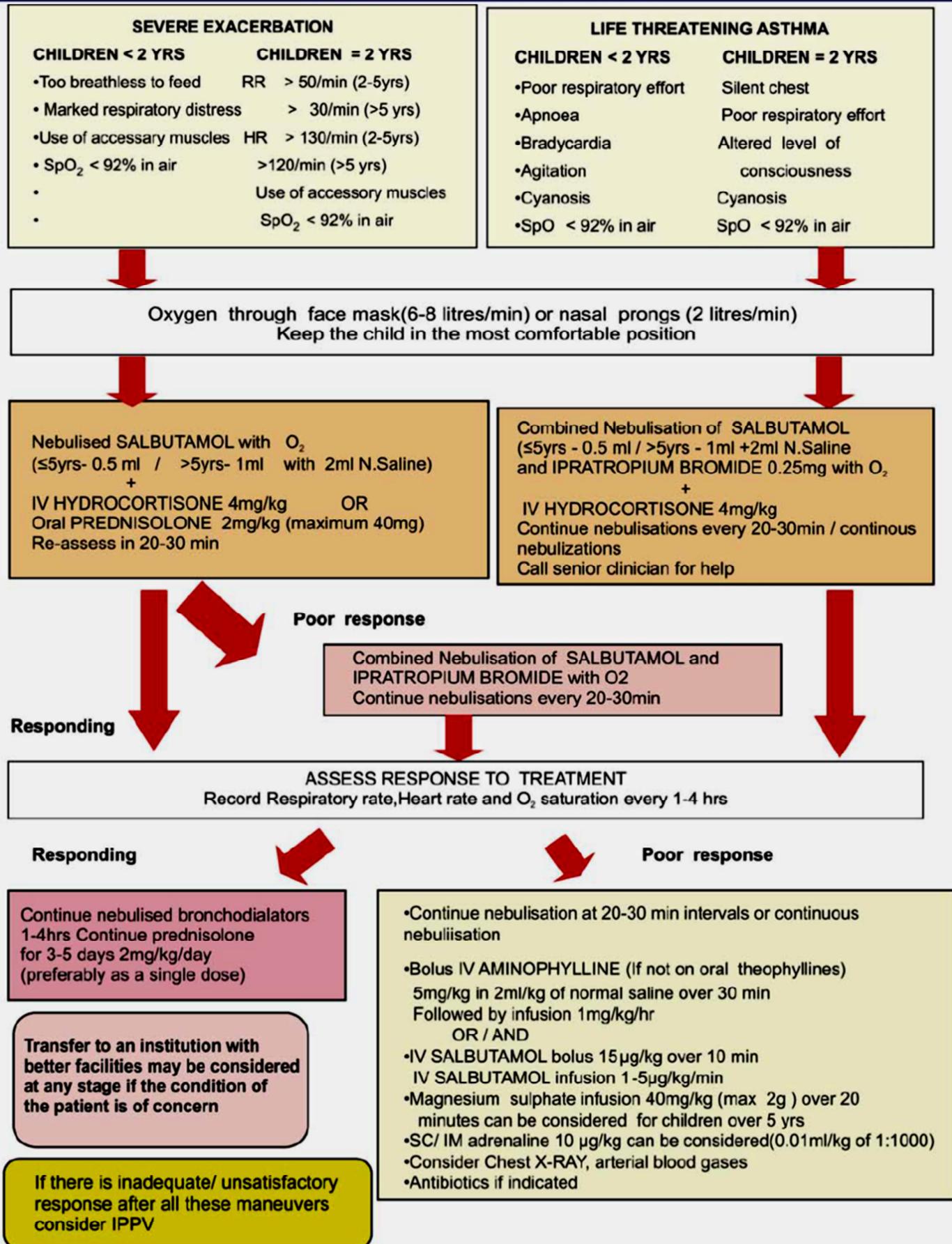
##### Avoid/minimize:

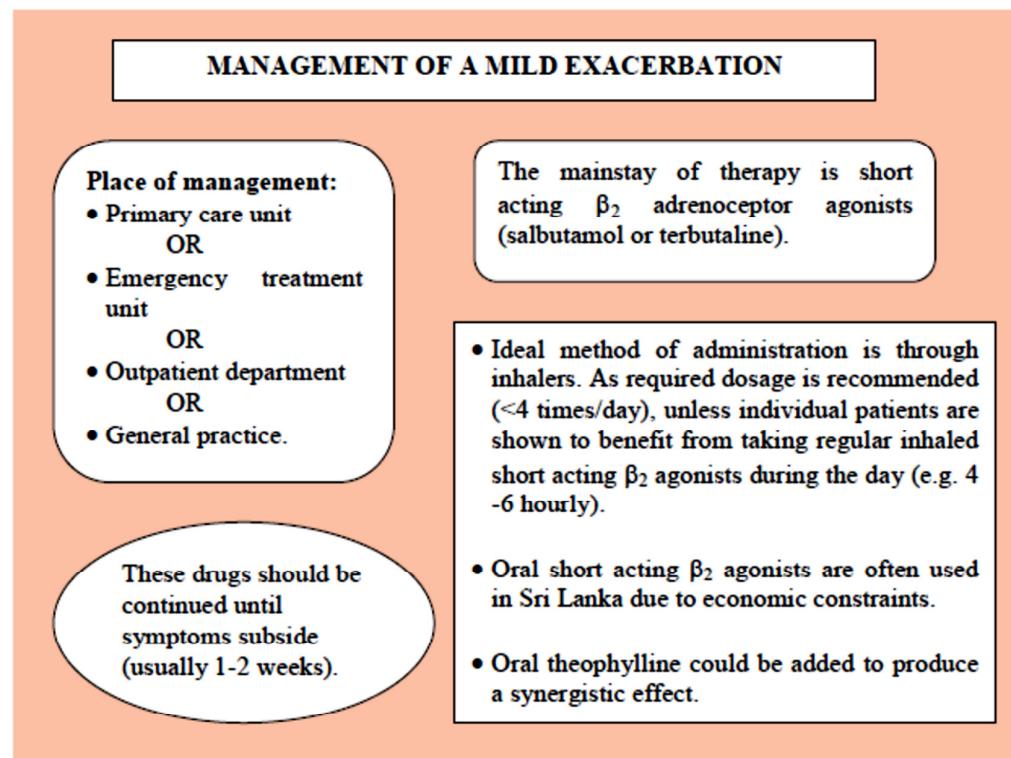
- identified allergens e.g. food/pollen
- Smoking      Active (teenagers)  
                    Passive
- Air pollution
- Obesity

##### No clear benefits

- House dust mite control
- Complementary or alternative medicine
- Generalized dietary restrictions (traditional Chinese medicine, acupuncture, homeopathy, hypnosis, yoga, Buteyko)
- Vitamin C
- Goat's milk  
[40,41,42,43,44,45,46,47,48,49,50,51,52,53,54]

# MANAGEMENT OF SEVERE AND LIFE THREATENING ATTACKS OF ASTHMA





#### Dosage of bronchodilator therapy for a mild exacerbation

Drug [55, 56]	Dosage		
Oral salbutamol	<2 yrs 100 $\mu\text{g}/\text{kg}$ tds/qds	2-6 yrs 1 to 2 mg tds/qds	> 6 yrs 2 mg tds/qds
Salbutamol inhaler	2-4 puffs every 4-6 hours		
Oral terbutaline	75 $\mu\text{g}/\text{kg}$ tds	75 $\mu\text{g}/\text{kg}$ tds	2.5 mg bd/tds
Theophylline	10 to 20 mg/kg/day divided tds/qds dosage		
Slow release theophylline	10 to 20 mg/kg/day two divided doses		

## MANAGEMENT OF A MODERATE EXACERBATION [4,55]

- **Place of management:** primary care unit / emergency treatment unit / outpatient department / general practice.

- **Inhaled  $\beta_2$  agonists via**

Metered dose inhaler (MDI) + spacer

*Dose: 2-4 puffs, increase by 2 puffs every 2 minutes up to 10 puffs according to response.*

### Nebulizers

*Dose: Salbutamol nebulizer solution (5mg in 1 ml)*

*<= 5 yrs - 0.5 ml salbutamol + normal saline 2 ml for 10 minutes*

*> 5 yrs - 1.0 ml salbutamol + normal saline 2 ml for 10 minutes*

- Prednisolone 2mg/kg/day 3-5 days, preferably given as morning single dose  
(no need to taper the dose at discontinuation)

Children under 3 years are likely to require a facemask connected to the mouthpiece of the spacer for successful drug delivery. Inhalers should be actuated into the spacer in individual puffs and inhaled immediately by tidal breathing.

*Reassess within one hour*

**Good response**

**Poor response**

Continue bronchodilators 1-4 hourly and could be discharged when stable on oral drugs or 6 hourly inhaler therapy. Further management should be reviewed as to whether the child requires long term treatment.

Repeat nebulization and the child should be referred to a hospital.

# Long term Management of Asthma

## STEP 1: Mild Intermittent Asthma

- Day symptoms(twice a week or less)
- Night symptoms(twice a month or less)
- Asymptomatic between exacerbations
- PEF <80% predicted

a. Daily medication: not needed.

b. Manage exacerbations

c. Avoid precipitants

d. Education: teach basic facts about asthma

## STEP 2: Mild Persistent Asthma

- Day symptoms more than twice a week but less than once a day
- Night symptoms more than twice a month
- Exacerbations may affect activity
- PEF <80% predicted

a. Preferred therapy - low dose inhaled steroids.  
Alternative therapy- sustained release theophylline, leukotriene receptor antagonist  
Ketotifen may be effective in younger children with atopy

b & c

d. Teach basic facts about asthma and proper technique of using inhaler. Discuss home management plan.

## STEP 3: Moderate Persistent asthma

- Day symptoms once a day
- Night symptoms more than once a week
- Daily need of  $\beta_2$  agonists
- Exacerbations affect activity, twice or more a week
- PEF 60%-80% predicted

a. Preferred therapy - medium dose inhaled steroids OR low dose inhaled steroids + long acting  $\beta_2$  agonists.  
Alternative therapy- Low dose inhaled steroids + either leukotriene receptor antagonist or theophylline  
Preferred treatment for patients with recurrent severe exacerbations: Medium dose inhaled steroids and long acting  $\beta_2$  agonist.

b & c

d: see step 2

## STEP 4: Severe Persistent Asthma

- Continuous symptoms
- Frequent night symptoms
- Limited physical activity
- Frequent exacerbations
- PEFR <60%

a. Preferred therapy – high dose inhaled steroids AND long acting  $\beta_2$  agonists  
May need oral steroid.

b & c

d. see step 2

- Patients at any level of grading can have mild, moderate or severe exacerbations
- Need to continue the therapy for a minimum of 1 year
- Attempt gradual withdrawal once the control is maintained.
- Always treat according to their most severe features across categories

Drug bd	Low dose (mcg)	Medium dose (mcg)	High dose (mcg)
Beclamethasone HFA	50-200	200-400	>400
Budesonide	100-200	200-600	>600
Fluticasone	100-200	200-400	>400

Combined medications	Steroid / LABA
Fluticasone+salmeterol	50/25, 125/25, 250/25
Fluticasone+salmeterol	100/50, 200/50
Budesonide+formeterol	80/4.5, 160/4.5
Leukotriene receptor antagonists	Daily oral dosage
Montelukast	4 mg oral granules once a day (12 months-5 years) 4 mg chewable tablet once a day (2-5 years) 5 mg chewable tablet once a day ( 6-14 years)
Zafirlukast	10 mg tablet b.d for 7-11 years

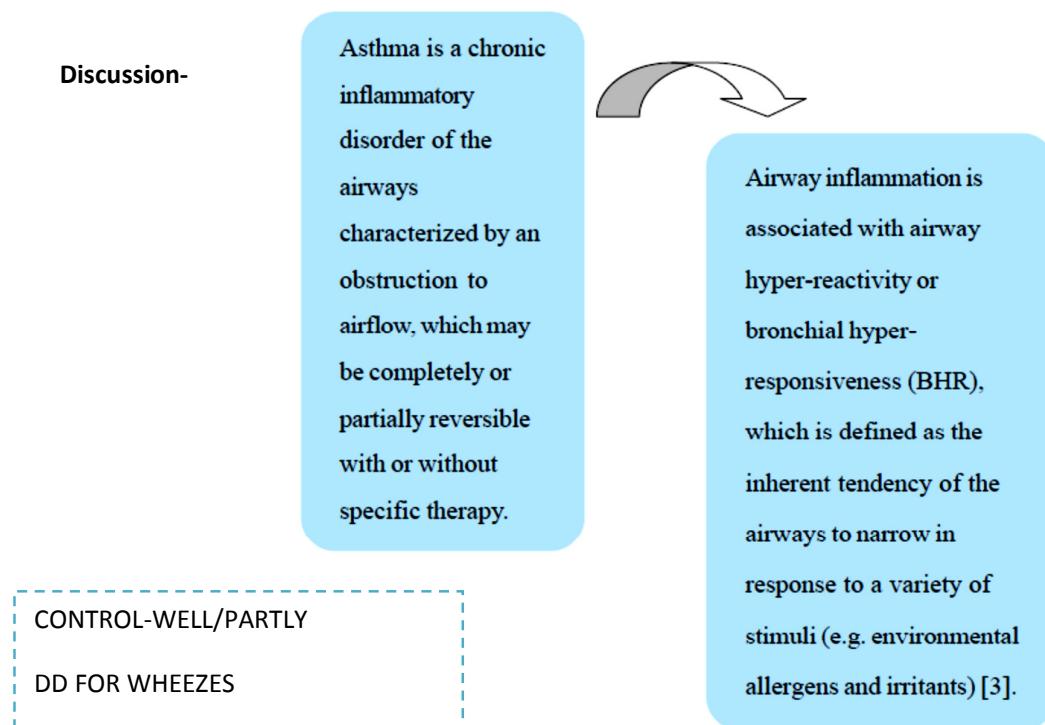
## Level of asthma control

Characteristic	Controlled (all of the following)	Partly controlled (any measure present in any week)	Uncontrolled
<b>Day symptoms</b>	None (twice or less/week)	>twice/week	Three or more features of partly controlled asthma present in any week
<b>Limitation of activity</b>	None	Any	
<b>Night symptoms/awakening</b>	None	Any	
<b>Need for relievers</b>	None (twice or less/week)	> twice/week	
<b>Lung function (PEF or FEV<sub>1</sub>)</b>	Normal	<80% predicted	
<b>Exacerbations</b>	None	One or more/yr	One in any week

## Inhaler devices

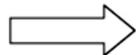
Age group	Device
<b>Up to 2 years</b>	Pressurized MDI + holding chamber
<b>2 – 3 or 4 years</b>	Pressurized MDI + spacer + face mask
<b>4-6 years</b>	Pressurized MDI + spacer
<b>&gt; 6 years</b>	DPI or breath-actuated MDI or Pressurized MDI + spacer

## DEFINITION OF ASTHMA



## PATOPHYSIOLOGY

Interactions between environmental and genetic factors result in airway inflammation



Bronchospasm, mucosal oedema, and mucus plugs

Airway obstruction

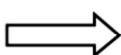
Alveolar hypoventilation

Hyperinflation

Increased resistance to airflow and decreased expiratory flow rates

Ventilation-perfusion mismatch.

In early stage, hypoxaemia without carbon dioxide retention occurs.  
With worsening obstruction carbon dioxide retention occurs



Respiratory alkalosis in the early stage and later result in metabolic and respiratory acidosis [3]

## Inhalers-

### Choosing the correct inhaler

Chosen according to the child's age and preference:

- Metered dose inhaler
- Breath-actuated metered dose inhaler, e.g. Auto-haler or Easi-Breathe
- Dry powder devices, e.g. terbutaline sulphate (Bricanyl Turbohaler) and salbutamol (Ventolin Accuhaler)

#### 1) Metered-dose inhaler (MDI)

Requires the greatest coordination and should never be used alone in children.

Using an MDI through a spacer device such as the Nebuhaler or aerochamber significantly increases the proportion of the drug reaching the airways, reduces impaction of drug on the throat and requires less coordination. In young children, a soft face mask can be attached to the spacer.

Ideally inhaled steroids should always be given by MDI and spacer, and spacers should be used in young children and for delivering beta agonists during acute asthma attacks. Spacers are very effective at delivering bronchodilators and inhaled steroids to the preschool child.



#### 2) Breath-actuated devices and dry-powder inhalers

Require less coordination than MDIs and can be used for delivering beta agonists in school-age children



#### 3) Nebulised treatment is now only given for severe life-threatening asthma, or rarely for children who need inhaled therapy but are unable to use any of these devices or require high doses

Metered dose inhaler	Metered dose inhaler with a spacer	Dry powder inhaler
Small and convenient to carry.	Less convenient to carry than a metered dose inhaler without a spacer.	Small and convenient to carry.
Doesn't require a deep, fast breath.	Doesn't require a deep, fast breath.	Requires a deep, fast breath.
Accidentally breathing out a little isn't a problem.	Accidentally breathing out a little isn't a problem.	Accidentally breathing out a little can blow away the medication.
Some inhalers require coordinating your breath with medication release.	A spacer makes it easier to coordinate your breath with medication release.	Doesn't require coordinating your breath with medication release.
Can result in medication on the back of your throat and tongue.	Less medication settles on the back of your throat and tongue.	Can result in medication on the back of your throat and tongue.
Some models don't show how many doses remain.	Some models don't show how many doses remain.	It's clear when the device is running out of medication.
Requires shaking and priming.	Requires shaking and priming and correct use of the spacer.	Single-dose models require loading capsules for each use.
Humidity doesn't affect medication.	Humidity doesn't affect medication.	High humidity can cause medication to clump.
Use of a cocking device generally isn't necessary.	Use of a cocking device generally isn't necessary.	May require dexterity to use a cocking device.

<b>Instructions for use of different inhaler types</b>	
<b>Type of device</b>	<b>Instructions for use</b>
<b>Pressurized aerosol metered dose inhaler (MDI)</b> Aerosol Evohaler	Remove the mouthpiece and shake the inhaler well. Hold the inhaler upright with the thumb on the base below the mouthpiece and the first finger on the metal canister. Breathe out as far as is comfortable then place the mouth piece between the teeth and close lips around it. Do not bite it. As you start to breathe in through the mouth, press on the top of the inhaler to release the medication whilst still breathing in steadily and deeply. Hold your breath, take the inhaler from your mouth and continue holding your breath for up to 10 seconds if possible. Wait 30 seconds prior to taking second puff. Use with spacer device to improve drug delivery. CFC-free inhalers need device washing every 2-3 weeks, as they can block.
<b>Spacer</b> Nebuhaler, Volumatic Aero Chamber, Able Spacer	Ensure spacer is compatible with patient's inhaler. Remove cap of inhaler and shake it. Insert it into end of spacer device. Place the other end of the spacer in the mouth. Press the inhaler canister once to release one dose of the drug. Take one deep breath in and hold, or take 3–4 steady breaths in and out. Repeat as indicated. Valve should rattle. Clean the spacer once a month with mild detergent, rinse, and air-dry. Replace after 6-12 months.
<b>Breath-actuated devices</b> Autohaler,Easibreathe	Prime the device. If an autohaler, remove the cap and lift the red lever; if an easi-breathe, open the cap. Insert device into mouth. Inhale slowly and deeply. Continue inhaling when the device ‘clicks’. Hold breath for up to 10 seconds if possible. Slowly breathe out. To take a second inhaled dose, lower the red lever and lift again. If an autohaler, close the cap and reopen if an easi-breathe and repeat the above sequence.
<b>Dry powder devices</b> <b>Accuhaler,Diskhaler</b> Turbohaler, Clickhaler,Twisthaler Cyclohaler, Aerocaps ,Spincaps	Prime the device. Turbohaler: remove the cap, twist the base as far as possible until the click is heard and then twist back again. Clickhaler: shake the device, remove the cap, click the top down and release. Twisthaler: remove the cap by twisting and the dose is then ready. Accuhaler: Open inhaler cover, mouthpiece facing you and push lever down to pierce the blister containing dose. Diskhaler: insert disc into device by opening and pulling out mouthpiece section. To prepare dose, lift up back of lid to 90° until the blister is pierced, then lower the lid. To use all the devices, hold them level, exhale fully, place the mouthpiece into mouth between teeth and inhale steadily. Hold your breath and remove the inhaler. For a second dose, repeat the above actions.
<b>Handihaler</b>	Flip lid and white mouthpiece open and insert capsule. Close the mouthpiece back down until it clicks. Pierce the capsule by pressing the green button at the side. Exhale as far as is comfortable. Place mouthpiece into mouth and breathe in slowly and deeply. Remove inhaler from your mouth and hold your breath for 10 seconds if possible. Slowly breathe out. Repeat if necessary to ensure all the powder from capsule is gone.

## Developmental delay

**Presentations** - <4yrs at 1<sup>st</sup> presentation – delayed milestones

- ❖ Delayed motor milestones
- ❖ Abnormal tone and posturing in early infancy – hypotonia/hypertonia
- ❖ Feeding problems –
  - Due to the spasticity of the tongue(tend to push food out), lips and pharynx
  - Recurrent aspiration
  - GORD
- ❖ Neurological/ motor weakness – abnormal gait, motor delay/asymmetry
- ❖ Developmental delay – language and social skills, lack of response to external stimuli
- ❖ Δased pt. coming with a complication – seizure, LRTI, fever
- ❖ Δased pt. coming for an Ix – CT brain, EEG

**PC** – Global/ focused to a specific area

Duration

**HPC** – start from the AN period to identify possible aetiological factors

**Birth Hx** – AN Hx – parity, mother's age, conception spontaneous / not

Rubella vaccination received, blood group and Rh, T1 fever with rash

Hx of PIH, IUGR

Hx of reduced fetal movements, abnormal CTG, failed induction – post dates

Perinatal – mode of delivery, prolonged labour, baby cried at birth, APGAR, any resuscitation

Meconium aspiration, PBU admissions

Postnatal – neonatal sepsis, meningoencephalitis, jaundice, hypoglycemia, seizures

### Disease manifestations in chronological order –

Was mother advised prior to discharge, when mother 1<sup>st</sup> noticed symptoms?

Spastic diplegia – Drag the legs behind during crawling, normal use of UL

Difficulty in sitting – parasternal mus.

Excessive adduction at hip – difficulty in clothing, posterior dislocation of the hip

Spastic hemiplegia – reduced spontaneous movements in one side of the body, early hand preference

Delayed walking, difficulty in hand manipulation – obvious by 1yr

Spastic quadriplegia – all 4 limbs affected, swallowing difficulty

Choreoathetoid – feeding difficulties, tongue thrust and drooling of saliva

DD – Cerebral palsy  
Degenerative diseases  
Muscular dystrophy  
Metabolic disorders  
Spinal cord tumours

### **Signs suggestive of CP in an infant**

Abnormal behavior
Excessive docility or irritability
Poor eye contact
Poor sleep
Oromotor problems
Frequent vomiting
Poor sucking
Tongue retraction
Persistent bite
Grimacing
Poor mobility
Poor head control
Hand preference before 2 years of age
Abnormal tone

<b>B</b>	<b>CP is likely if there is no</b>
Head control	3 months
Sitting	6 months
Rolling over	6 months
Walking	18 months

Complications –1) seizures  
                   2) FTT, GORD  
                   3) Recurrent LRTI, UTI  
                   4) Teeth problems – dental caries  
 Previous hospitalizations, indication, duration of stay

**Immunization Hx** – seizure following vaccination, any vaccine withheld Eg: JE

**Developmental Hx** – assess each domain

- 1) Gross motor and posture – delayed motor milestones, gait abnormalities  
                   Unable to maintain posture
- 2) Fine motor and vision – visual impairments – lack of eye contact, response to obj., Squints  
                   Visual assessment done – cortical blindness
- 3) Hearing & speech – hearing impairment, whether hearing assessment done – sensorineural deafness  
                   Speech – delayed, difficulty in articulation (due to muscle spasticity)  
                   Learning difficulties, mental retardation
- 4) Social behavior and play – incontinence, constipation, behavioural prb., lack of imitative play

**Dietary Hx** – feeding difficulties, weaning – difficult since child does not like to changeover to semisolids

Coughing bouts after feeds – aspiration → recurrent chest infections

#### **DHx and allergy**

**FHx** – consanguinity – agenesis of brain, any other child with similar prb., epilepsy  
 Neurological disorders in childhood

**SHx** – introduction to family – parents and siblings, occupation, income  
 Family support, social benefit available/not → 500/- for CP  
 Housing conditions – space available, accident prone areas – stairway, fireplace, close to main road  
 Affect on other family members – neglect  
 Rehabilitation – prosthesis – walking aids, feeding chairs, standing frames, teets  
                   Speech therapy, hearing aids  
                   Occupational therapy – for adductor thumb, ADL  
 Hygiene – presence of dental caries, bathing frequency  
 Knowledge about the disease and prognosis, family completed/not, family planning if completed  
 Nearest hospital and mode of transport

## Examination

### General

- Weight, height
- OFC – compared with height – microcephaly, hydrocephalus

Check the 50<sup>th</sup> centile Ht for his age  
 His Ht less than that → stunting  
 50<sup>th</sup> centile age for his height  
 Check the 50<sup>th</sup> centile OFC for above age – x  
 X > child's OFC - microcephaly

- Dysmorphic feature
- Hygiene – smell of stale urine – neglect, lack of bladder control
- Evidence of malnutrition – protein energy malnutrition – Kwashiokor/ marasmus features
- Posture - ophisthotonus, arm posture – flexed/ extended at shoulder/elbow
- Involuntary movements – choreo-athetoid
- Activities of daily living – age appropriate activities/ paucity of movement  
 (At the end of 2yrs only athetoid movements)
- Febrile
- Head – fontanelle – closed/ open, bulging/ sunken
  - Receding forehead, B/L dimpling due to poor brain growth – lIrry microcephaly
  - Ridging over skull – insult during labour/ just after birth (perinatal)
  - (Insult → brain stops growing → skull fusion while the skull bones are in overriding position)
- No ridges – in-utero insult
- Eyes – squints, follows the moving light → coloured object, cataract
- Hearing – moves the head to the sound of the rattle
- Oral cavity – Drooling of saliva
  - Oral hygiene
  - Markers of forceful feeding – contusions on either side of the mouth
  - Micronutrient def. – angular stomatitis, glossitis
  - Dental caries

Cranial N.	Observe for
II	Response to light , PERTL Presence of rotational nystagmus } cortical Fundus – optic atrophy } blindness
III, IV , VI	Eye movements
VII	Deviation of angle of mouth/ symmetrical facial expressions Can close both eye lids
VIII	Hearing – moves the head to the sound of the rattle
IX, X	Drooling of saliva, if opens mouth – uvula deviation
XII	Tongue deviation

CNS –

- UL / LL – presence of dynamic and static contractures – with the degree
  - Muscle wasting – esp. hamstrings and quadriceps
  - Tone, deep tendon reflexes
  - Clonus

## F Differences between spasticity & dystonia

	<i>Spasticity</i>	<i>Dystonia</i>
<i>Examination</i>	You feel	You see
<i>Tendon reflexes</i>	Increased	Generally normal
<i>Clonus</i>	Present	Absent
<i>Pathological reflexes</i>	Present	Rare

- Developmental assessment
  - Gross motor and posture – scissoring of legs – spastic quadriplegia
  - Fine motor and vision – fingers flexed/open, thumb adducted
  - Hearing and speech
  - Social behavior and play

Primitive reflexes – usually disappears in 3months. Persisting for >6months – abnormal

- 1) Asymmetric tonic neck reflex (ATNR) – if present cannot roll over
- 2) Grasp reflex

**Chest** – deformities – pectus excavatum/ carinatum

- AP diameter of chest wall
- Breathing – rate, depth – shallow

**RS** – features of LRTI (GORD, aspiration pneumonia, drooling of saliva)

**CVS** – PR, apex, heart sounds

**Abdomen** – fecal masses, palpable bladder – urine retention

**Hips** – Prominent greater trochanter, posterior dislocation of hip (due to extreme hypertonia)

**Back** – deformities – kyphosis, scoliosis

- Meningocele/ meningocele/
- Hair tuft, lipoma, skin discolouration, dermal sinus – spina bifida occulta

## Summary

### Problem list

#### Acute medical

- ◆ Current presentation – seizure, LRTI, UTI
- ◆ Constipation

#### Chronic medical –

- ◆ Microcephaly/hydrocephalus
- ◆ CP - type
- ◆ FTT
- ◆ Feeding problems
- ◆ Epilepsy
- ◆ GORD
- ◆ Vision and hearing problems

### Social problems

- ◆ Neglect of other family members
- ◆ Fear of having another child/ not practicing family planning
- ◆ Financial

## Discussion

### What is global development delay?

Condition where there is a delay in the acquisition of **2 or more** domains following a pathological process

### What is developmental regression?

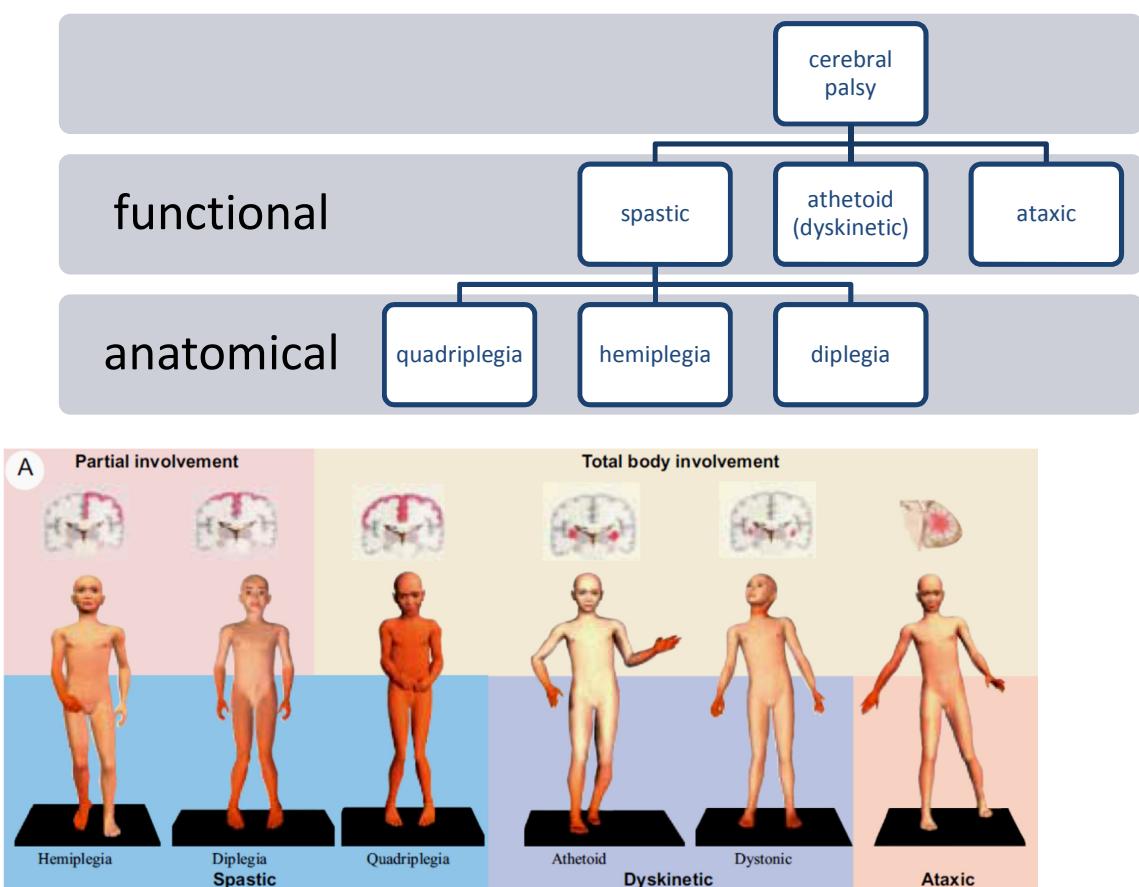
Acute/ gradual loss of a previously acquired skill

### Cerebral palsy

Definition: Disorder of **posture and movement** resulting from a **non progressive insult** to the **developing brain**

- Lesion is not progressive; yet the clinical manifestations change over time
- Incidence increased despite good perinatal and obstetric care – due to increased survival of premature and LBW baby

### Classification



<b>Spastic (70%)</b>		<b>Athetoid (dyskinetic, dystonic) -10%</b>	<b>Ataxic hypotonic (10%)</b>
Commonest type Lesion : cerebrum / pyramidal tract		Lesion : extra-pyrimidal (damage to basal ganglia + pathways)	Less common Lesion : cerebellum + connections
Limb tone ↑		Fluctuating tone – initial hypo --> ↑ tone, rigidity, dystonia	Early trunk and limb hypotonia
Brisk deep tendon reflexes		Freq. involuntary movements	Symmetrical signs
Extensor plantar reflex (+ babinski) B/L ankle clonus		Chorea Athetoid and dystonia	Poor balance and delayed motor development
		Feeding difficulty – tongue thrust, drooling	In co-ordinated movements
			Intention tremor
Quadriplegia	Hemiplegia	Diplegia	
All 4 limbs	One side	All 4	
UL > LL	Arm > leg	LL > UL	
Tip toe standing	Tip toe walking	Walking impaired	
Trunk –extens. posturing	Spared		
Poor head control	VII n. palsy common		Poor head control and marked head lag Oropharyngeal mus. Involve. – slurring of speech/↓ voice modulation Fits uncommon
Low central tone			
Ass. – fits microcephaly	Fits +(1/3 <sup>rd</sup> in 1 <sup>st</sup> yr/2)	Fits minimal	
Mod – severe intellect. ↓	Cognitive abn. +, MR	Intellect intact	Intellectual impairment
Supranuclear bulbar palsy			

Causes / risk factors - classified according to the time of injury

C	Time of brain injury
Prenatal period	Conception to the onset of labor
Perinatal period	28 weeks intrauterine to 7 days postnatal
Postnatal period	First two years of life

D	Manifestations of cerebral palsy
<i>Neurological</i>	<i>Associated problems</i>
Muscle weakness	Intellectual impairment
Abnormal muscle tone	Epilepsy
Balance problems	Visual problems
Loss of selective control	Hearing loss
Pathological reflexes	Speech and communication problems
Loss of sensation	Swallowing difficulty , Teeth problems
<i>Musculoskeletal</i>	Feeding difficulty, failure to thrive
Contractures	Respiratory problems
Deformities	Incontinence

Prenatal	perinatal	Postnatal
IU infections	Prematurity	Brain injury – cerebral oedema
Threatened miscarriage	Birth asphyxia	Postnatal asphyxia – cardiac arrest
APH	Perinatal strokes	Rapid changes in PaO <sub>2</sub> and CO <sup>2</sup>
PIH, maternal systemic illness	Prolonged fits	
IUGR		Neonatal hypoglycemia
Congenital anomalies of the brain		Neonatal sepsis, meningitis
		Kernicterus

A	Risk factors
<i>Prenatal</i>	
Prematurity (gestational age less than 36 weeks)	
Low birth weight (less than 2500 g)	
Maternal epilepsy	
Hyperthyroidism	
Infections (TORCH)	
Bleeding in the third trimester	
Incompetent cervix	
Severe toxemia, eclampsia	
Hyperthyroidism	
Drug abuse	
Trauma	
Multiple pregnancies	
Placental insufficiency	
<i>Perinatal</i>	
Prolonged and difficult labor	
Premature rupture of membranes	
Presentation anomalies	
Vaginal bleeding at the time of admission for labor	
Bradycardia	
Hypoxia	
<i>Postnatal (0-2 years)</i>	
CNS infection (encephalitis, meningitis)	
Hypoxia	
Seizures	
Coagulopathies	
Neonatal hyperbilirubinemia	
Head trauma	

Motor manifestations in CP

Primary deformity → secondary deformity → tertiary deformity

<b>Primary impairments</b> <i>(due to the brain lesion)</i>	
Muscle tone (spasticity, dystonia)	
Balance	
Strength	
Selectivity	Ability to selectively control antagonist & agonist mus.
Sensation	Loss of proprioception , movement sensation impaired
<b>Secondary impairments</b> <i>(due to the primary impairments causing the movement disorder)</i>	
Contractures (equinus, adduction)	
Deformities (scoliosis)	
<b>Tertiary impairments</b>	
Adaptive mechanisms (knee hyperextension in stance)	

D	Common sites for contracture		E	Common sites for deformity
	Upper extremity	Lower extremity	Spine	Scoliosis, kyphosis
Pronator		Hip adductor-flexor	Hip	Subluxation, dislocation
Wrist and finger flexor	Knee flexor		Femur & tibia	Internal or external torsion
Thumb adductor	Ankle plantar flexor		Foot	Equinus, valgus, varus

Visual impairments seen in CP	
Pathology	Clinical finding
Damage to the visual cortex	Cortical blindness
Damage to the optic nerve	Blindness
Loss of oculomotor control	Loss of binocular vision
Refraction problems	Myopia

G Teeth problems		D Oromotor dysfunction
Dentin	Primary or hyperbilirubinemia	Drooling
Malocclusion	Spasticity	Dysarthria
Tooth decay	Feeding, swallowing problems	Inability to chew
Gingival hyperplasia	Antiepileptic drug use	Inability to swallow

Reasons for failure to thrive	K Causes of inadequate food intake
Inadequate food intake	Difficulty chewing and swallowing
Recurrent vomiting	Hyperactive gag reflex
Aspiration	Spasticity of oropharyngeal muscles
High basal metabolic rate	Loss of selective control of oropharyngeal muscles
Recurrent infections, poor sensation → low appetite	Oesophagogastric reflux

Urinary problems	Causes of urinary problems
Enuresis	Poor cognition
Frequency	Decreased mobility
Urgency	Decreased communication skills
Urinary tract infections	Neurogenic dysfunction
Incontinence	

**Causes for recurrent RTI** – Palato-pharyngeal dysfunction, ↓ cough reflex → aspiration

Posture – orthostatic pneumonia

Hypoventilation

Poor nutrition - ↓ immunity

How will you manage?

- Clinical Δ at the end of Hx & Ex

- 1) MRI – location and extent of structural lesion  
Associated congenital malformations

Meantime...

- 2) Hearing assessment
- 3) Visual assessment
- 4) EEG – if fits
- 5) Feeding assessment – with a 24hr dietary recall and calculate the calorie requirement
- 5) Genetic studies – chromosomal abnormalities & metabolic disorders

## Management

- ✓ Multidisciplinary approach with the following members
  - Paediatrician, orthopaedic surgeon, neurologist
  - Nurses
  - Physio therapist
  - Occupational therapist
  - Social workers
  - Psychologist
  - Speech therapist
- ✓ Counseling and education
  - No cure
  - Possible cause
  - Teach how to work with their child in daily activities minimizing the effects of abnormal muscle tone  
Feeding, dressing, carrying, bathing and playing
  - Instruct to supervise exercises designed to prevent muscle contractures
  - Advise on another child, if not family planning
- ✓ treat conditions that are treatable
  - correction of visual problems – cataracts, squints
  - hearing – hearing aids
  - speech therapy – correct drooling
- ✓ physiotherapy – main stay of treatment, start early as possible
- ✓ occupational therapy – provide stimulation to the child
  - Teach normal day-to-day activities
  - Teach age appropriate activities
  - Correct oversensitivity related problems due to lack of stimuli to certain domains of sensation
- ✓ correct feeding – treat reflux, NG feeding
  - if severe reflux- feeding jejunostomy
- ✓ schooling – in special education units (esp. when intellect intact)

- ✓ Rx – 1) anticonvulsants
- 2) anti-spastics → Dantrolene Na
  - Benzodiazepines → cause sedation
  - Baclofen → ↓ seizure threshold
  - Intra-thecal form successful in selected pt.

Botulinum toxin injection - into spastic muscle ; good response in many pt.

Also control salivation

Anti cholinergics – control salivation

- ✓ Surgery – soft tissue procedures → ↓ muscle spasm around hips, adductor tetany
  - Spinal nerve root division
- ✓ Aids – walkers, standing frames
- ✓ Follow up

## Haemophilia

Common presentations: joint pain and swelling  
Bleeding manifestations

DD – joint pain and swelling	DD – bleeding manifestations
1) Rheumatic fever	1) vWD, Vit K deficiency, liver disease – coagulation disorders
2) JIA	2) HUS, DIC
3) Septic arthritis	3) BM failure
4) HSP/SLE	4) Thrombocytopenia – viral infections/Rx/ AI/ sequestration
	5) Vascular – HSP, scurvy, Ehler-Danlos

Hx – Already diagnosed/ 1<sup>st</sup> presentation  
Nearly always ♂

HPC – Onset, duration and progression of symptoms  
Preceding trauma/ spontaneous  
Associated discolouration of skin  
Joint deformity, unable to move the joint

What was done now – at home → hospital  
If haemarthrosis – more pain, more quickly than previous – joint arthropathy dev.

If 1<sup>st</sup> presentation exclude DD

Condition	Points in the Hx
Rheumatic fever	Fever, fleeting and flitting large joint involvement, subcutaneous nodules, carditis (new onset murmur), established valvular heart disease, chorea
JIA	PUO, large and small joint involvement, Rash with the height of fever Painful red eye – chronic anterior uveitis
Septic arthritis	Usually monoarthritis, child does not move the joint, high fever
HSP	Preceding URTI, palpable red rash over extensors and buttocks

### Hx of bleeding manifestations in the past

Infants – Bruises with knocks on the cot/ pick up, with crawling → painful joint swelling  
Bleeding – esp. ankle, circumcision  
Haematoma following vaccination

Children – severe bleeding following tooth extraction  
Haemarthrosis – esp. knee, elbow → deformities, disuse atrophy  
Early recognition – warm, tingling sensation  
Gum bleeding, epistaxis, frequent bruises, oozing from scars  
Abdominal pain  
Complications – Recurrent bleeds into same joint – target joint → chronic arthropathy  
ICH – focal fits, headache  
Haematuria, haematemesis, melaena  
Bleeding into iliopsoas – hypovolemia, vague groin pain, cannot extend hip

Δed pt. – past hospitalizations, F VIII/IX Tx, child's blood group, response to F VIII/IX – inhibitors  
Complications of blood Tx – reactions, anaphylaxis, Blood borne infec. – HBV, HCV, HIV, malaria  
Regular screening for infections

Birth Hx – Mode of delivery

Cephalhaematomas, ICH – seizures, umbilical bleeding  
(Factor VIII does not cross placenta, however during birth process VIII ↑ - since APP  
Factor IX physiologically low at birth)

Immunization – Hx of haematoma and bleeding following IM vaccination

Hep B vaccination given  
Vaccines given in the SC form

FHx – Consanguinity

Maternal uncles affected, any other siblings affected  
Mother – Menorrhagia (females can have bleeding due haemophilia – due to lyonization – random inactivation of the normal x chromosome, homozygous form – rare, die before birth, turners Xn, XY ♀)

SHx – Income and education level of parents

Family support  
Effect on child's day-to-day activities and school performance  
Overprotective parents  
Effect on other family members  
Knowledge about the disease, what to do during a bleed  
Nearest hospital with Tx facilities, transport available, time taken  
Awareness of the school teacher

## Examination

General – Is the child in pain

Fever – temperature chart (septic arthritis)  
Pallor  
Nose - epistaxis  
Dental caries  
LN - generalized  
Skin – bruises  
Child abuse – finger pulp contusions

MS – haemarthrosis – Painful/ tense swelling

Limitation of movement  
Deformities – disuse atrophy

Septic arthritis – Very warm, red and tender, no movement

Other joints and muscles – esp. iliopsoas – hip kept flexed and internally rotated

CVS – PR, pulse volume, BP – haemodynamic stability

ABD - I – Abdominal distension, bruises

P – Abdominal tenderness  
Hepatosplenomegaly – JIA, leukaemia, lymphoma

CNS – GCS

Pupils

Fundi – retinal haemorrhage

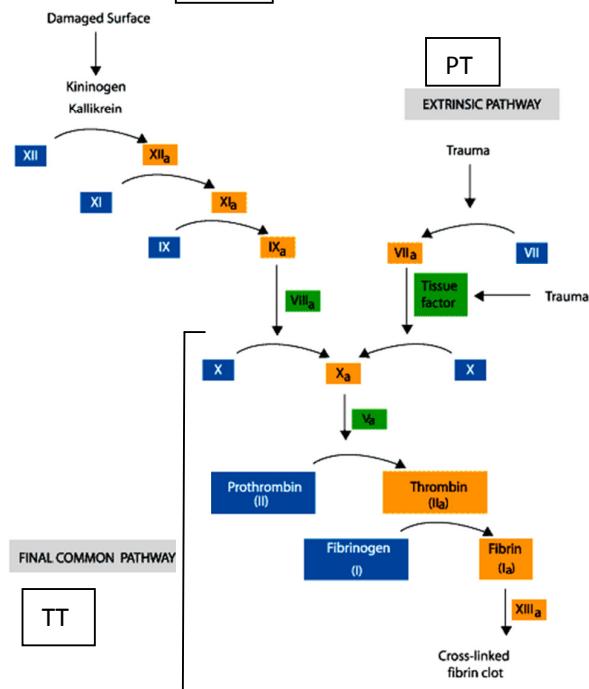
UL, LL – weakness

Ix

- FBC – Plt. count, Hb and other cell lines
- Blood picture – platelet morphology, presence of abnormal cells (leukemia, lymphoma)
- Clotting profile

	Plt. count	BT (7min) (Platelet & vWF)	Clotting time (Both extrinsic & intrinsic)	PT/INR(12s) (Extrinsic)	APTT (30-40s) (Intrinsic)
Haemophilia	N/L	N/L	↑	N/L	↑
vWF	N/L	↑ (plt dysfunc)	↑	N/L	↑
ITP	↓	↑	N/L	N/L	N/L
Liver disease	N/L	N/L	↑	↑	↑
DIC	↓	↑	↑	↑	↑

INTRINSIC PATHWAY APTT



Other Ix

- Factor assay

Haemophilia A – factor VIII

Haemophilia B – factor IX

(christmas disease)

Severity – severe <1% of normal + spontaneous bleeding

Moderate – 1-5% of normal + minor trauma

Mild >5% of normal + major trauma

- Inhibitor screen\*

Via factor correction 1:1

Corrected -no (-)      not corrected

(-) present

- Other

✓ DNA studies

✓ Prenatal Δsis – not in SL

Chorionic villous sampling

(8-10wks)

\* 1:1 mixing of pt's plasma with normal plasma → correction of APTT

In 25-35% due to presence of factor specific Abs/ inhibitors APTT is not corrected. They are directed against the active clotting sites

Quantitative Bethesda assay – find the Antibody titre

Acute management

Supportive care

- ♥ Pain relief – avoid aspirin and NSAIDs
- ♥ Intermittent cold compressions - ↓ leukocyte-endothelial interaction

DRUG	ADULT DOSE	PEDIATRIC DOSE
Acetaminophen (Paracetamol)	500mg -1 g every 4-6 hours	10-15 mg/kg every 4-6 hours (available as syrup)
If the pain is not relieved with acetaminophen alone then add any one of the following:		
Propoxyphene	65 mg every 4-6 hours	Not recommended for children
Codeine	180-200 mg every 4-6 hours	0.5 -1 mg/kg every 4 hours
Buprenorphine	0.8 mg every 6 hours Sub-lingual	Not recommended for children
Tramadol	50-100 mg every 6 hours	Not recommended for children

- ♥ Immobilize the limb in its functional Position – splint/sling – UL
- ♥ Gentle compression crape bandage/elastic Stocking around the Joint – Limit bleeding and Support the joint
- ♥ Rehabilitation after pain and swelling settles
  - Start physiotherapy – static exercises
  - Use walking aids
  - Daily exercises to improve muscle strength & maintain joint motion

#### Specific management

- Factor replacement therapy  
Based on the type of bleed and the haemostatic factor correction required

#### Technique of Ice Application

- Put ice cubes in a cloth and crush.
- Apply on the skin with a thick towel as a wet under-layer.
- Leave for 5 minutes.
- Remove for 10 minutes and repeat.

**Application of ice is not useful after 48 hours.**

**one unit of F VIII raises plasma level by**

- 2% (0.02U/ml)  $t_{1/2}$  - 12hrs

**one unit of F IX raises plasma level by**

- 1% (0.01u/ml)  $t_{1/2}$  - 24hrs

Dose to be infused = wt. (kg) x factor increment needed (%)

2

In haemophilia B divide by 2 – which is the percentage rise in plasma with one unit of F IX

Site of bleed	Haemostatic F level	Comment
Joint	30-50% minimum	Rest, immobilization, rehabilitation. Target joint will need several doses
Muscle	40-50% minimum	Calf / forearm bleeds Life threatening large blood loss in femoral / retroperitoneal bleeds
Oral mucosa	Initially 50% → Antifibrinolytic therapy	Antifibrinolytic therapy is critical
Epistaxis	Initially 80-100% → 30% until healing	Local measures ; pressure, packing/cautery useful for severe/recurrent bleeds
GIT	Initially 100% → 30% until healing	Endoscopy highly recommended to find lesion. Antifibrinolytic therapy useful

GUT	Initially 100% → 30% until healing occurs	Look for stones or UTI. Lesions usually not found. Prednisolone may be useful
CNS	Initially 100% → 50-100% for 10-14 days	Anticonvulsants. LP will require factor coverage.
Trauma or surgery	Raise up to 100% & maintain at 30-50% for up to 2 weeks to prevent 2ry haemorrhage	Pre & post op plan essential. Evaluate for inhibitors prior to surgery

- Bleeds into joint, muscle, oral & mucosal bleeding → require 50% factor correction
- Life threatening bleeds (CNS, GIT, GUT bleeding, Epistaxis & in surgery/trauma → require 100% correction

Eg: haemarthrosis –  $\frac{50\% \times \text{wt}}{2} = 25\text{IU/kg}$

Haemostatic level for factor VIII – 30-40%  
Haemostatic level for factor IX - >25-30%

1IU = amount of factor in 1ml of normal plasma

Types of factor concentrates →

- 1) Recombinant FVIII/IX – Expensive, not in gov. sector; **Adv.** – not plasma derived – no risk of infec.
- 2) High purity FVIII/IX – Plasma derived but virally inactivated via detergents/heat etc., available in SL
- 3) Plasma derived FVIII/IX – available in gov sector → risk of HBV, HCV, HIV
- 4) Cryoprecipitate – used in SL
- 5) FFP



F VIII 1 bottle – 250IU  
F IX 1 bottle – 600IU  
Cryoprecipitate – F VIII 150IU/pack  
Used for F VIII correction  
FFP – Used for F IX correction  
F IX – provides around 80IU/pack  
(Considering only 40% factor recovery with Tx)  
One pack – 250ml

If transfusing...

- ✓ FFP – should be ABO compatible. Rh not needed  
Contains no WBC → no risk of CMV Tx & GVHD
- ✓ Cryoprecipitate – multiple donor → ↑ risk of infec.  
Now single donor recommended – usually the father after DDAVP therapy (**No need of group specificity – Rh or ABO**)

TYPE OF HEMORRHAGE	HEMOPHILIA A	HEMOPHILIA B
Hemarthrosis	D1 -40 IU/kg factor VIII D2-5 → 20 IU/kg until joint function is N/L/ baseline. Consider additional Rx EOD for 7–10 days. Consider prophylaxis.	D1- 60–80 IU/kg D2-4 →40 IU/kg same
Muscle / significant SC hematoma	20 IU/kg factor VIII concentrate; may need EOD treatment until resolved.	40 IU/kg F IX concentrate <sup>[‡]</sup> May need treatment every 2–3 days until resolved.
Mouth, deciduous tooth, or tooth extraction	20 IU/kg factor VIII concentrate – one dose <b>antifibrinolytic therapy</b> ; remove loose deciduous tooth → cont. 5d after	40 IU/kg factor IX concentrate <sup>[‡]</sup> ; <b>antifibrinolytic therapy</b> <sup>[§]</sup> ; remove loose deciduous tooth.
Epistaxis	Apply pressure for 15–20 min; pack with petrolatum gauze <b>antifibrinolytic therapy</b> → fails 20 IU/kg factor VIII concentrate <sup>[*][*]</sup>	Same ↓ fails 30 IU/kg factor IX concentrate <sup>[‡]</sup>
Major surgery, life-threatening hemorrhage	50–75 IU/kg factor VIII concentrate → initiate continuous infusion of 2–4 IU/kg/hr to maintain factor VIII > 100 IU/dL for 24h → give 2–3 IU/kg/hr continuously for 5–7 days to maintain the level at > 50 IU/dL → an additional 5–7 days at a level of > 30 IU/dL.	120 IU/kg factor IX concentrate <sup>[‡]</sup> , → 50–60 IU/kg every 12–24 hr to maintain factor IX at > 40 IU/dL for 5–7 days → > 30 IU/dL for 7 days.
Iliopsoas hemorrhage	50 IU/kg factor VIII concentrate, then 25 IU/kg every 12 hr until asymptomatic, then 20 IU/kg every other day for a total of 10–14 days. <sup>[¶]</sup> <b>Confirmed – USS/CT</b>	120 IU/kg factor IX concentrate <sup>[‡]</sup> ; then 50–60 IU/kg every 12–24 hr to maintain factor IX at > 40 IU/dL until asymptomatic, then 40–50 IU every other day for a total of 10–14 days. <sup>[¶][¶]</sup>
Hematuria	Bed rest; hydrate well - 1½ × maintenance fluids; if not controlled in 1–2 days, 20 IU/kg factor VIII concentrate; → fails give prednisone (unless HIV-infected).	Bed rest; 1½ × maintenance fluids; if not controlled in 1–2 days, 40 IU/kg factor IX concentrate <sup>[‡]</sup> ; if not controlled, give prednisone (unless HIV-infected).
Prophylaxis	20–40 IU/kg F VIII concentrate EOD to achieve a trough level of ≥ 1%.	30–50 IU/kg factor IX concentrate <sup>[‡]</sup> every 2–3 days to achieve a trough level of ≥ 1%.

[‡] dose given for recombinant FIX. plasma derived FIX – 70% of above dose

### Other therapy

#### 1) Anti- fibrinolytic Rx

- used in mucous membrane bleeds (which occur due to fibrinolysis preventing clot stabilization)

Eg : Trenexamic acid – Best absorbed from the buccal mucosa

Dose – 20mg/kg – tablet dissolved in 10ml of water → solution kept in mouth  
as

long as possible → swallowed

#### 2) Prednisolone – In macroscopic upper tract haematuria

Dose – 0.5mg/kg x 5d → 0.25mg/kg x 5d

#### 3) Desmopressin (DDAVP)

I° - mild – moderate haemophilia A, type I vWD (Ineffective in haemophilia B)

Action – causes the release of vWF from endogenous stores in endothelial cells → ↑vWF and F VIII

Dose – IV 0.2 -0.3µg/kg in 50-100ml of N/S

Intra-nasal spray – 1 puff - <50kg

2 puffs >50kg

S/E – flushing, fluid retention, ↓ Na<sup>+</sup> → seizures (infants)

Adv. – cheap, no risk of infection

#### 4) Other – Ca alginate (seaweed derivative) I° - epistaxis

Danazole – short term, following CNS bleeds, target joint haemarthrosis, vWD - ♀

### Long term Mx

#### Parent education

- ✓ Nature of the disease – life long risk of bleeding, no cure
- ✓ Genetic basis – 'X' linked inheritance, maternal carriage, can be a spontaneous mutation

Risk to future pregnancies – 50% of boys will be affected

#### ✓ Preventive measures

<3yrs → 1) child should be supervised in bed/allow to sleep on a mattress on the floor  
2) Barricade any stairways  
3) Ensure toys have no sharp edges, prevent access to sharp instruments  
4) Protective gear – car seats, seat belt, helmets, protective pads

> 3yrs → 1) bicycle – stabilizers/guard wheels to prevent falls  
2) Avoid contact sports, encourage swimming, cycling

- Avoid dental caries, brush 2x/day
- Early presentation to hospital for acute bleeds
- First aid
- Avoid aspirin and NSAIDs
- Initiation of replacement therapy at home – Via indwelling venous access  
Not in SL
- Inform Dr/ dentist/ peers and teachers – Carry diagnostic card

#### ✓ **Avoid overprotection**

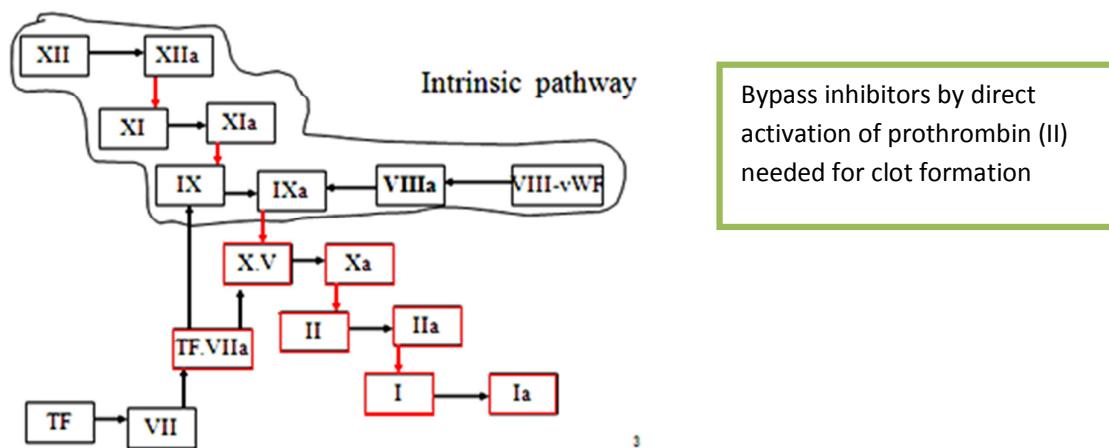
#### ✓ Early recognition of bleeds. Esp, life threatening bleeds

GIT – abdominal pain, melaena, haematemesis

CNS – early morning headache, altered consciousness

- ✓ Routine immunization  
Not via IM route  
All given subcutaneously via 26G needle → apply firm pr. over injection site for 3-5min  
HBV vaccination
- ✓ Maintain normal BMI

- ⊕ Long-term physiotherapy – promote strong muscle , protect joints and improve fitness
- ⊕ Regular joint and muscle Ex
- ⊕ Complication Mx
  - 1) Chronic haemophilic arthropathy & joint deformities  
chronic joint bleeds → target joints → chronic inflammation of the synovium → Loss of time from school, ↑ long term disability, Psychosocial consequences  
Mx – control the bleeding  
Synovectomy
  - 2) Contractures
  - 3) Disuse muscle atrophy
- 4) Inhibitor development
  - Low responding inhibitors & low Bethesda titres - **high dose factor & frequent or continuous replacement of factor (Desensitizing programs)**
  - Higher titres- **plasmapheresis/use porcine factor replacement**
  - Therapy for inhibitors
    - **By pass therapy** - Prothrombin complex concentrate (PCC) or activated PCC
    - **Inhibitor eradication**  
Immune tolerance - Daily or weekly use of factor + Cyclophosphamide/high dose gamma globulin / prednisolone



- 5) Life threatening bleeds
- 6) Blood borne infections

7) Psychological problems

8) Septic arthritis

■ Prophylactic factor correction

- Persons with moderate Haemophilia rarely develop chronic arthropathy
- Adequate prevention - by maintaining a minimum factor VIII or IX level at 1-5%

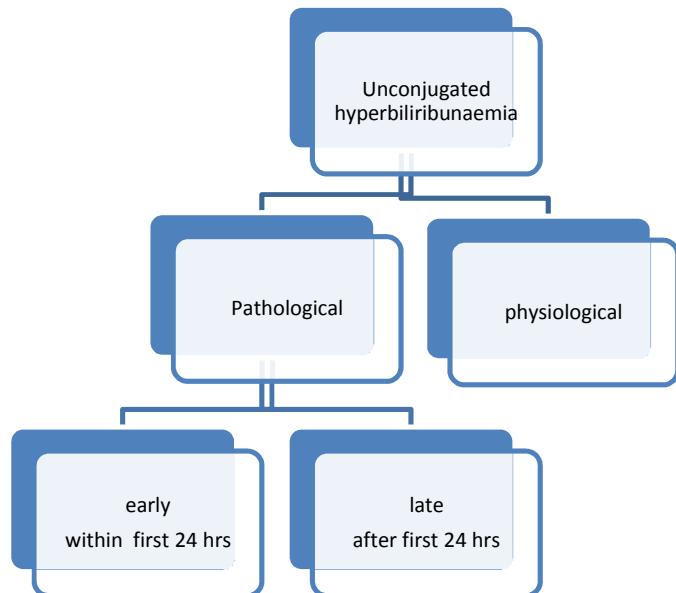
**Primary prophylaxis in severe disease**

- Factor replacement at 1-3 years
  - 25-40 units/Kg of F VIII three times a week & F IX twice a week
- Fewer Haemarthroses per year & normal joints

**Secondary prophylaxis**

- Initiated later due to early arthropathy - not as effective as primary prophylaxis  
X 4 increase in factor requirement

# Neonatal Jaundice



## JAUNDICE WITHIN 1<sup>ST</sup> 24 HOURS (DD)

- \* Haemolytic disease of newborn (HDN)
  - Rh incompatibility
  - ABO incompatibility
  - Rare blood group incompatibility
    - Kell
    - Duffy
- \* Haemolytic anaemia
  - G6PD deficiency
  - Pyruvate kinase deficiency
  - Congenital spherocytosis
- \* Sepsis

## Causes for prolonged jaundice (More than 2 weeks)

- Breast milk jaundice
- Sepsis - Septicemia
  - UTI
  - Congenital infections
- Endocrine - Hypothyroidism
- Metabolic - Galactosaemia

## JAUNDICE AFTER 24 HOURS

- Physiological
- Jaundice of prematurity
- Bilirubin production
  - eg. Bruising, Cephalhematoma, DIC, Polycythaemia
- Infections
- Infant of diabetic mother
- Persisting hemolytic disease
- Enteropathic circulation
  - intestinal obstruction
  - pyloric stenosis
- Congenital & metabolic
  - Down syndrome
  - Crigler Najjar syndrome
  - Gilbert syndrome
  - galactosaemia

## Introduction

- Baby of Mrs.....
- Age of the child
- Sex

## PC

- **Yellowish discolouration of sclera and body ..... duration**

## HPC

- Describe according to chronological order
- Start from the birth Hx.
- Antenatal Hx
  - In which pregnancy
  - Mother's blood group
  - Rubella immunization
  - Fever with a rash in the 1<sup>st</sup> trimester
  - VDRL status, scan results (hydrocephalus, heart defects)
  - Threatened miscarriages, APH , amniocentesis
  - Maternal blood transfusions
  - Maternal GDM, hypothyroidism
  - Maternal anti-thyroid drugs
  - Previous babies with jaundice
- Natal Hx
  - Pre-labour rupture of membranes
  - Number of VE done
  - How long had she been in the labour room
  - Maternal pyrexia at the time of delivery, antibiotics given /not
  - Mode of delivery
  - Forceps applied (cephalhaematoma)
  - At term or not
  - Birth weight, cried at birth, pink
  - Taken to PBU
  - Breast feeding was established/not
- Post natal
  - When did the mother first noticed jaundice (mention the exact day)
  - Up to which level did the jaundice extend in the body
  - Activity of the child
  - Crying
  - Sucking of the child/lactation failure
  - UOP and stool colour of the child

### Exclude other DD

- Sepsis
  - Fever, irritability, poor feeding, excessive crying
  - Floppy child
  - Discharge from the umbilical stump, ear discharge
- Lactation failure
  - Breast feeding position and attachment
  - Duration of a single feed
  - What does the baby do after a feed?
- **How do you assess the adequacy of breast feeding?**
  - UOP = 6 times /day
  - By the time of 5<sup>th</sup> day stool colour should change to yellow from meconium
  - Activity of the child
  - Frequency of breast feeding and duration of a single feed
  - Baby sleeps for about 2hours after feeding
- **How do you know the mother is producing adequate milk?**
  - Led down reflex (milk ejection reflex by Oxytocin)
    - ✓ When baby sucks from one nipple milk comes out from the other breast
    - ✓ Secretion of oxytocin by the posterior pituitary results in contraction of the myoepithelial cells surrounding the alveoli deep in the breast. This in turn, "squeezes" milk into the larger ducts, where it is more easily available to the sucking infant. When this "led down" or milk ejection reflex functions well, milk flows from the opposite breast as the infant begins to nurse.
    - ✓ Feeling of pins and needles from the breast
    - ✓ After feeding mother feels the emptiness of the breast

### Assess the complications

- Kernicterus : high pitched cry
- Floppy baby, seizures

### What had been done up to now?

- Blood Ix done
- Admitted to SCBU
- Phototherapy- single/double
- Exchange transfusion
- IV drugs, antibiotics received
- How the baby is fed
  - Expressed BM
  - Formula milk
  - IV infusions

### PMx

- Screening for STI

### Family Hx

- Consanguinity
- Family Hx of episodic jaundice, splenectomy

### Social Hx

- Maternal level of education
- Occupation
- Income
- Family support

### Examination

#### General

##### Do a normal neonatal examination, and specially concern about:

- Weight, length, OFC of the child (plot in centile charts)
- Posture : flexed /extended
- Hydration
- Scalp
  - Cephal haematoma
  - Caput succideneum
  - Suture lines
  - Fontenelles
- Pallor
- Icterus – up to what extent
  - Up to the level of the umbilicus needs phototherapy
  - Up to feet needs exchange transfusion
- Dysmorphic features (congenital infections)
- Neck lumps
  - Thyroid
- Umbilical stump
  - Erythema, discharge
- Skin rash
  - Sepsis
  - Congenital infections

#### CVS

- PR
- Palpate the femoral pulses
- Apex – cardiomegaly
- Comment on First and second heart sounds
- Murmurs

#### RS

- Air entry : equal /not
- Breathing : vesicular / bronchial
- Added sounds

## Abdomen

- Distension
- Umbilical hernia (congenital hypothyroidism)
- Visible veins, spider naevi
- Masses palpable : liver,spleen
- Kidneys
- Free fluid



## NS

- Eyes
  - For cataract and squints
  - Red reflex
- Tone
- Primitive reflexes (palmer, plantar, moro's reflex- tell whether it is symmetrical /not)

## Back examination

- Anus
- Genitalia

## Hip examination

- Otolarni and barlow's maneuvers

## Assess the technique of breast feeding

- Positioning
  - Whole body of the baby should be held in one horizontal line
  - Baby 's body shouldn't be covered with layers of clothes
- Attachment
  - Baby's whole body is facing his mother and close to her
  - Face is close up to the breast
  - Chin is touching the breast
  - Mouth is wide open
  - Lower lip is everted
  - More areolar is above the baby's upper lip and less areolar is below
  - Baby takes low deep sucks
  - Doesn't hear any noises when he sucks
  - Cheeks blow out
  - At the end of the feed baby is relaxed and happy
  - Mother doesn't feel nipple pain

## Summary

## Problem list

## DD

## Discussion

### Management

#### Investigations

##### Blood

- Blood grouping and DT
- FBC
  - o Hb
  - o WBC : may be high or low
- Serum bilirubin total and direct
- Septic screen
  - o CRP
  - o Blood culture & ABST
  - o Urine culture & ABST
  - o CXR
  - o Lumbar puncture
- Serum electrolytes if dehydration is suspected

#### If Indirect hyperbilirubinaemia is present

- Direct Coomb's test (if suspecting Rh /ABO incompatibility)
- Blood picture (if haemolysis is suspected)
  - o Hereditary spherocytosis : microspherocytes densely stained, smaller diameter than normal RBC
  - o G6PD deficiencies : fragmented RBC, bite and blister cells, Heinz bodies
  - o Sepsis : N'phil leukocytosis
- Retic count

##### Urine

- Bilirubin :low
- Urobilinogen :high

#### If direct hyperbilirubinaemia is present

##### Blood

- SGOT / SGPT
- Serum protein
- PT/INR
- Alkaline phosphatase

##### Urine (UFR)

- Bile salts positive
- Urobilinogen (negative)

To find the aetiology for direct hyperbilirubinaemia

USS abdomen

- Hepatosplenomegaly (neonatal hepatitis syndrome)
- Choledochal cyst (cystic dilatations of extrahepatic biliary system)
- Biliary atresia: absence of extra hepatic biliary tree

Liver biopsy and per-operative cholangiogram

TIBIDA scan of liver showing good hepatic uptake but no excretion in 24 hrs.

Biliary  
obstruction

Congenital infections

- Syphilis :VDRL
- Toxoplasosis : specific IgM
- Urine CMV
- Rubella IgM

Hepatitis B surface antigen

Hypothyroidism (check after 7 days)

- Serum TSH
- Free T<sub>4</sub>

## Treatment

### Indirect hyperbilirubinaemia

- 1) Phototherapy
- 2) Exchange transfusion
- 3) Treat the underlying cause

### Phototherapy

I<sup>o</sup>: serum bilirubin levels reaching the phototherapy levels

(Level is calculated according to the *birth weight, maturity, age, ill or well baby*)

**Mechanism of phototherapy** – Bilirubin in the skin absorbs the light energy and by reversible photo-isomarisation reaction converts bilirubin into an isomer which can be excreted via bile without conjugation. Other major substance is lumirubin which is excreted by urine without conjugation.

**Color of light-** Blue

**Wave length of that colour** - 450-460nm

**Maximum distance of baby and light source**- 18 inches

**Rate of SBR decrease following phototherapy**- 2mg/hr

### Complications of phototherapy

- ✓ Dehydration
- ✓ Hypothermia/hyperthermia
- ✓ Diarrhoea
- ✓ Erythematous skin rash
- ✓ Maternal anxiety
- ✓ Eye damage
- ✓ Bronze discolouration of skin if jaundice is conjugated



### Exchange transfusion

I<sup>o</sup>: serum bilirubin level reaching the exchange transfusion level

#### Method of exchange transfusion

- Double Volume exchange 160ml/Kg
- Rh -ve blood same as babies group
- Should be compatible with mother's blood group (therefore send a maternal sample)
- Fresh blood within 5 days
- Need an assistant
- Exchange transfusion set/3 way tap, feeding tubes
- Cut the cord longer if anticipate
- Done in Op. theatre/ICU
- Continuous cardiac monitoring
- Umbilical venous catheter – just to get continuous blood flow
- Hb, SBR pre exchange & after exchange
- Warm the blood before start
- Either 10ml/20ml aliquots are done
- Always take out the blood
- The average time for exchange is one hour
- **1ml Calcium gluconate** slowly after each 100ml to avoid hypocalcaemia
- Prophylactic antibiotics
- Monitor blood sugar & oxygen saturation
- Pre & Post transfusion BU/SE/S.Bilirubin/Hb/RBS are done

### **Effect of exchange with double volume**

- Removal of sensitized RBC > 85%
- 55 – 60% bilirubin
- After few hours SBR increases

### **Complications**

- Embolization (air/clots)
- Thrombosis (can later develop portal hypertension.)
- Cardiac arrhythmias
- Hyper/hypovolaemia
- Hypothermia
- Increase K<sup>+</sup>, Na<sup>+</sup>, decrease Ca<sup>++</sup> and acidosis
- Infection
- Hypoglycaemia

### Specific mx

- Cephalhaematoma : reassure
- Sepsis : IV antibiotics

## **Haemolytic disease of newborn**

### **Rh incompatibility**

- ✓ Pathophysiology
  - Arises when a mother is Rh-ve & baby is +ve
  - Rh positive fetal blood can be entered into Rh negative mother's circulation (feto-maternal haemorrhage) during pregnancy or during delivery (usually > 1ml)
  - Anti D IgM formation against the D antigen is initiated in the maternal circulation
  - Later the IgM is converted to IgG. This can readily cross the placenta and cause the haemolysis in subsequent pregnancies
- ✓ Clinical manifestations
  - Jaundice
  - Severe pallor with hepatosplenomegaly (due to compensatory hyperplasia of erythropoietic tissue)
  - Hydrops fetalis
    - Profound anaemia
    - Cardiomegaly, respiratory distress
    - Excessive and abnormal fluid in 2 or more fetal compartments(skin, pleura, pericardium, placenta, peritoneum, amniotic fluid)
  - IUD
- ✓ Prevention of Rh incompatibility
  - Rhogam injection

- ✓ Complication of Rh HDN
  - Kernicterus
  - Hypoglycaemia
  - Infections
  - DIC
  - Late anaemia
  - Folate deficiency
  - Obstructive jaundice

### Kernicterus

- Deposition of unconjugated (indirect) bilirubin in the basal ganglia and brainstem nuclei
- Clinical features
  - Early features may be reversible
  - Later become irreversible
- BBB permeable up to 7 -10 days

### ACUTE FORM

Phase 1 (1st 1–2 days): poor sucking, stupor, hypotonia, seizures

Phase 2 (middle of 1st wk): hypertonia of extensor muscles, opisthotonus, retrocollis, fever

Phase 3 (after the 1st wk): hypertonia

### CHRONIC FORM

First year: hypotonia, active deep tendon reflexes, obligatory tonic neck reflexes, delayed motor skills

After 1st yr: movement disorders (choreoathetosis, ballismus, tremor), upward gaze, sensorineural hearing loss

- Yellow discoloration of basal ganglia and hippocampus by UC bilirubin
- Conjugated bilirubin :no effect
- Increase risk of kernicterus in
  - Preterm infants
  - Rapidly rising bilirubin
  - Hypoalbuminaemia
  - Hypoxia, acidosis, hypoglycaemia, and sepsis
- Management
  - Early treatment will prevent complications
  - Exchange transfusion
  - Photo therapy

### ABO incompatibility

- Mother O +ve & baby A +ve commonly or B +ve
- About 90% of O +ve mothers have Ig M anti A & anti B
- 10% have Ig G anti A & anti B. They can cross the placenta and affect the child.
- 1<sup>st</sup> child can be affected
- Severe HDN rare
- Direct Coombs test weekly + ve or – ve

## Jaundice of prematurity

- Exaggeration of physiological jaundice
- TREATMENT
  - Phototherapy
  - Rarely exchange transfusion
- Needs treatment at lower levels of SBR

## Breast milk jaundice

- Commonest cause of prolonged jaundice
- Diagnosis of exclusion

### CLINICAL FEATURES

- Prolongation of physiological jaundice beyond 2/52
- Otherwise a normal baby
- Growing well
- Rarely kernicterus can occur in babies with BM jaundice.
- Due to inhibition of conjugation and increase enterohepatic circulation
- TREATMENT Reassurance & continue breast feeding

## Conjugated hyperbilirubinaemia

- Direct fraction > 20 % of total bilirubin level
- Dark green colour jaundice
- Hx of pale stool and dark urine
- Hepatosplenomegaly

### Causes

- Congenital infections
  - \* CMV \* Rubella
  - \* H. simplex \* Hepatitis B
  - \* Coxsackie B \* V. zoster
  - \* Treponema \* Toxoplasmosis
- Acquired infections
  - \* UTI
  - \* Septicaemia
- Decreased bile drainage
  - \* Biliary atresia/Hypoplasia
  - \* Choledocal cyst
  - \* Bile duct stenosis
- Metabolic
  - \* Galactosaemia
  - \* Alpha-1 antitrypsin deficiency
  - \* Cystic fibrosis
- Others
  - \* Idiopathic
  - \* Familial
  - \* Parenteral nutrition

## Neonatal hepatitis syndrome

### Causes

- Congenital infections
  - Rubella, CMV, Toxoplasma, Syphilis
- Inborn errors of Metabolism
  - Alpha 1 antitrypsin deficiency
  - Galactosaemia
  - Tyrosinaemia(type 1)
- Cystic fibrosis
- Total parenteral nutrition cholestasis
- Idiopathic neonatal hepatitis (sporadic or familial form with unknown origin)
  - has a familial incidence of ≈20%

DD: biliary obstruction

## Biliary atresia

- Occurs in 1 in 14000 live births
- It is a progressive disease in which there is destruction or absence of extra-hepatic biliary tree and intra-hepatic bile ducts
- Unlikely to recur within the same family
- This will lead to chronic liver failure and death unless surgical intervention is performed
- Babies will have normal birth weight but develop failure to thrive as the disease progress
- They are jaundiced and from the second day stools are pale, urine dark. Both jaundice and stool colour may fluctuate.
- Hepatomegaly is present and splenomegaly will develop as a result of portal hypertension

### Investigations

- Standard liver function tests are of little value in DD
- Fasting USS abdomen
  - May be normal or demonstrate contracted or absent gall bladder
  - Cystic dilatations: Choledochal cysts
- TIBIDA scan
  - Shows good uptake by the liver, but no excretion into the bowel
- Liver biopsy
  - Features of extra-hepatic biliary obstruction (fibrosis, proliferation of bile ductules)
- Intra-operative cholangiogram
  - To confirm the diagnosis
  - Failure to identify the normal biliary tree

### Treatment

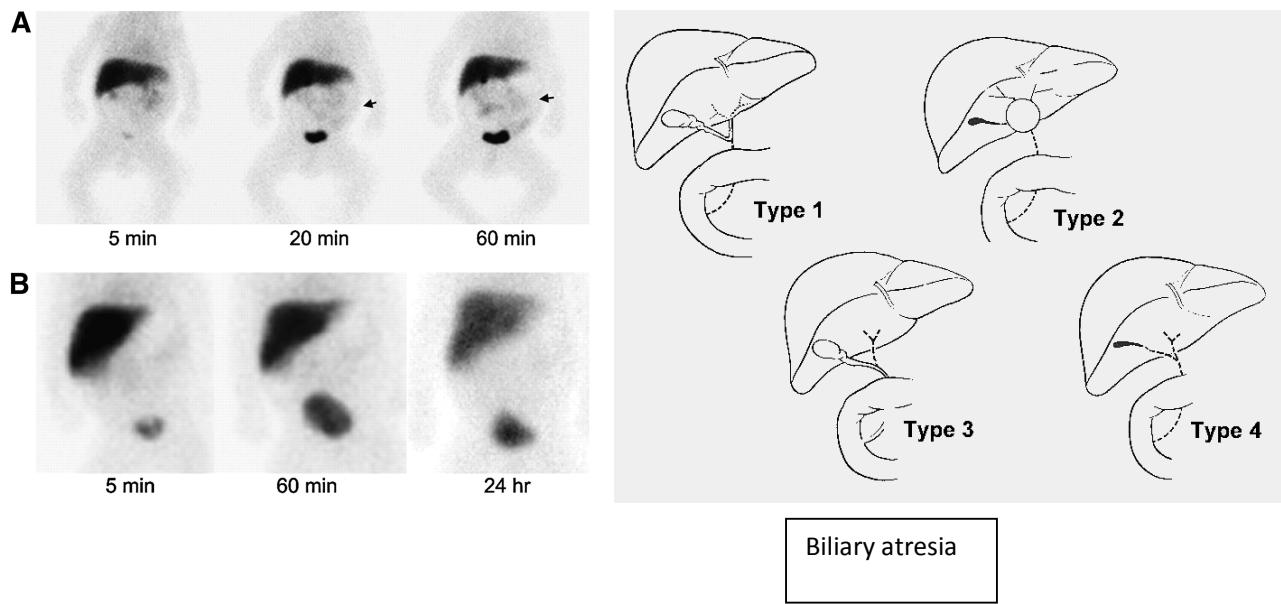
- Early recognition is very important
- Hepato-porto-enterostomy (Kasai procedure)
  - Jejunum is anastomosed to patent the cut surface of the porta hepatis
- If the Sx is done
  - <8/52 – Bile drainage is good (80%)
  - >12/52 – Prognosis bad



After Kasai procedure

#### Post-operative Complications

- Cholangitis
- Fat malabsorption
- Even the biliary drainage is successful. There will be progression to cirrhosis and portal hypertension
- Liver transplant has to be reconsidered.



Hepatocellular disease and biliary atresia. (A) Newborn girl with hepatocellular disease. Selected images from  $^{99m}\text{Tc}$ -disofenin scintigraphy at 5, 20, and 60 min reveal tracer elimination into bowel (arrows). (B) One-week-old boy with biliary atresia. Selected images from  $^{99m}\text{Tc}$ -disofenin scintigraphy at 5 min, 60 min, and 24 h do not show tracer in bowel. Large portion of tracer is eliminated by kidneys.

#### Child with jaundice (Wasantha madam)

**Findings** –Deep jaundice which is greenish yellow

Stools are greenish

Child is pale and irritable (due to itching)

No peripheral stigmata of CLF – spider naevi, clubbing, leuconychia

Child has severe failure to thrive with flattened buttocks, wasted limbs

#### **Abdomen**

It is markedly distended but flattened umbilicus. Sacral oedema absent

There is a clean cut well healed scar extending from the right loin → left hypochondrial region

Very likely it is a surgical scar. Not suggestive of explorative laparotomy or liver biopsy.  
Unlikely due to choledochal cyst drainage as the child is still jaundiced.

Obstructive jaundice

There is a gross hepatomegaly. Lower border of the liver is 5 finger breadths below the R/costal margin and is regular. Upper border is in the 5<sup>th</sup> ICS. Surface is finely nodular and it is hard and non tender

Spleen is moderately enlarged towards the LIF (seen in infants) firm.

Horse shoe dullness present

Conclusion – she is having features of obstructive jaundice with evidence of decompensated cirrhosis as evidenced by ascitis and evidence of portal hypertension showed by the splenomegaly. Possible underlying cause is a cholestasis most likely an extrahepatic one due to the attempted surgery.

### **What are the possibilities?**

Prolonged direct hyperbilirubinemia – 1) Intrahepatic biliary hypoplasia  
2) Extrahepatic – choledochal cyst  
Biliary atresia  
3) Neonatal hepatitis – idiopathic, viral – HBV, HCV  
4) α-1 anti-trypsin deficiency  
5) ↓ biliary excretion – Dubin Johnson Xn, Rotor Xn

### **What causes the itching?**

Deposition of bile salts

### **What are the bile salts?**

Primary bile acids → 1. cholic acid – conjugation with taurine (AA) – taurocholate  
Conjugation with glycine - glycocholate  
  
2. chenodeoxycholic acid → chenodeoxycholate

Secondary bile acids → 3. Deoxycholic acid

4. lithocholic acid

### **What are the problems in this child?**

- 1) Inability to absorb fat → lack of micelle formation. However able to absorb medium chain FA w/o micelle formation. Present in breast milk, progestamine (formula milk)
- 2) Inability to absorb fat soluble vitamins – given orally double the dose (A,D, E, K)  
Def. of vit K → Haemorrhagic disease – lack of clotting factors II,VII, IX,X → common pathway  
Prolonged clotting time, abnormal PT/INR
- 3) Biliary cirrhosis – improve bile excretion – cholestyramine – bile acid binding resin → prevents bile acid re-absorption. Increase faecal excretion  
Phenobarbitone (hepatic enzyme inducer) – aids conjugation of bilirubin

## NEPHRITIC SYNDROME

### PRESENTING COMPLAINT

- HAEMATURIA- AGN, UTI
- OEDEMA- NEPHROTIC SYNDROME
- HYPERTENSION

H/P/C

ELOBORATE ON ONSET, PROGRESSION, DURATION, AND ASSOCIATED FEATURES OF P/C

#### Haematuria

1. Is it haematuria or not?
  - Pseudo-haematuria
    - I. Red coloured urine
      - Preceding ingestion of red coloured food and beverages(eg; dyes)
      - **Drugs** : Rifampicine(red coloured), Metronidazole
    - II. Haemoglobinuria; cola coloured
      - **G6PD deficiency** : Intake of drugs like antimalarials, Antibiotics, Anti inflammatory, FMHx
    - III. Myoglobinuria ; plain tea coloured
  - Haematuria; pink/red
2. Duration
3. Associated with pain or not?  
Painless-RCC  
Painful - UTI, calculi
4. Time of the haematuria
  - ✓ Through-out - kidney, glomerular, ureters, bladder
  - ✓ Initiation - urethra
  - ✓ End of the urination-bladder neck, trigon
5. Recurrence and Progression ; persist or gradually falling?

#### Other causes of haematuria

1. **Bleeding disorders** – Past history of easy bruising, petechial haemorrhage, Family history
2. **Trauma** – Abdomen, renal angle or genitalia
3. **Drugs**
  - Cyclophosphamide – haemorrhagic cystitis
4. **UTI** : Fever +/- chills and rigors, abdominal pain, dysuria, frequency
5. **Stone** – Painful haematuria, Colicky abdominal pain, Loin to groin pain(Ureteric colic), Tip of the penis pain(Bladder neck or urethra)
6. **PSGN** : Recent history of sore throat or skin infection

**7. Immunological**

- I. **HUS** : Recent history of blood and mucous diarrhea
  - II. **HSP** : Joint pain, skin rash, abdominal pain
  - III. **SLE** : Arthralgia, skin rash, fever
  - IV. **IgA nephropathy** : Recent Hx of URTI or tonsillitis
  - V. **Infective endocarditis** : Congenital heart defects, Rheumatic fever
8. **PCKD** - Childhood polycystic kidney disease(AR), Loin pain, Haematuria, Family history
9. **Tumour** – Feeling of a mass in the abdomen (wilms tumour/Nephroblastoma <5 yrs)

**Ask about other features of AGN**

- **Urine out-put** – oliguria
- **Hypertension** - Headache, blurring of vision, seizures
- **Oedema-**
  - When it was noticed
  - Sites
  - Time of the day at which oedema is prominent
- Frothy urine/ proteinuria

**Ask about complication**

- Hypertension(encephalopathy) - Headache, drowsiness, seizures, blurred vision
- Acute LVF - dyspnoea, orthopnoea, PND
- ARF - Anuria

**What has done up to now**

- At home
- At hospital
  - Investigations-results
  - Treatments-response

**PMHx-**

- Similar illness in past
- Recent instrumentation, any abnormalities in urinary tract

**FMHx-**

- Deafness and haematuria(Maternal side) - alport syndrome(X linked)
- Family history of renal calculi – Hypercalcaemia, Cysteinuria
- Infantile PCKD(AR)- Associated with cysts in other places
- Kidney disease- renal transplant
- Vesico ureteric reflux (Familial)

**DHx**

- Anticoagulants

**SHx**

- Exclude child abuse
- Low socio economic class
- Hygiene
- Knowledge about disease

## Examination

- **General**
  - Activity, ill/well looking
  - Growth-wt/ht(plot) and also calculate the surface area
  - Hydration
  - Febrile or not
  - Anaemia and icterus - G6PD
  - Oedema – sacral ankle, peri-orbital
  - Evidence of skin infection - scabies
  - Bruising, petechiae - bleeding disorders
  - Skin rashes - SLE, HSP
  - Swollen tender joints - HSP
  - Throat - Evidence of infection
  - Peripheral stigmata of IE
- **CVS**
  - PR - Tachycardia
  - BP- hypertension
  - JVP- HF
  - Cardiomegaly-HF
  - gallop rhythm ( $3^{\text{rd}}$  / $4^{\text{th}}$  heart sound + tachycardia)
- **RS**
  - bilateral crepitations all over the lung fields - pulmonary oedema
- **Abdomen**
  - Supra pubic tenderness and bladder
  - Renal angle tenderness
  - Ballotable kidneys-PCKD
  - Palpable masses - Willm's tumour
  - Splenomegally- haemolytic anaemia, IE
  - External genitalia- oedema (NS)
- **CNS**
  - Features of hypertensive encephalopathy (confusion)
  - Deafness-sensory neural deafness in Alport's syndrome
  - Cranial nerve lesions, focal signs → In children with severe HT

## Investigations

1. **UFR**
  - RBC
  - Pus cells → In UTI pus cells > Red cells
  - RBC casts → suggestive of glomerular bleeding
  - proteinuria
  - Dysmorphic RBC-special request
2. **Mid-stream clean catch urine for culture and ABST**
  - $>10^5$  colony forming units/ml of single pathogen or  $10^{4-5}$ /ml + symptoms
  - catheter sample  $>105$  CFU/ml + positive UFR + symptoms
  - Supra-pubic aspiration → Any number

**3. FBC**

- Hb level
- WBC/DC → infection

**4. Urine ward test for protein → to exclude Nephrotic xn**

**5. USS KUB**

- Abnormal cortico-medullary demarcation suggestive of acute renal parenchymal disease → in AGN
- Bladder wall
- Cystitis →
- Dilated upper tract

**6. S.Creatinine , Bu/SE**

**7. X-RAY KUB → renal stones**

**8. Evidence of invasive streptococcal infection**

- ✓ ASOT- commonly elevated after throat infection (less commonly in skin infection)  
- > 200 is significant
- ✓ Anti-DNase B level-best single AB titre to identify skin (strep) infections
- ✓ Streptozyme test-detect AB to streptolysin O, DNase, hyaluronidase, streptokinase, NADase
- ✓ Throat swabs- may supportive (represent the carrier state)

**9. Serum complement levels**

- In AGN also C<sub>3</sub> C<sub>4</sub> levels reduced in acute phase
- But Return to normal 6-8 wks
- If not return to normal level → consider other causes of GN such as SLE

**When do you consider renal biopsy**

- ✓ In the presence of ARF >14 day in nephritic syndrome
- ✓ Also if haematuria, proteinuria, reduced renal function and or reduced C<sub>3</sub> persist > 2 months

**If suggestive of glomerular haematuria**

- ESR, complement levels and anti-DNA binding
- Throat swab and antistreptolysin O titre
- Hepatitis B antigen
- Renal biopsy if indicated
- Test mother's urine for blood (if Alport's syndrome suspected)
- Hearing test (if Alport's syndrome suspected)

## Management

### General measures

- ✓ Bed rest-not indicated unless HT emergency
- ✓ Fluid
  - Restriction is not indicated unless ARF
  - **Fluid requirement- previous day UOP+ insensible loss(400ml/m<sup>2</sup>/day)**
- ✓ Normal diet
  - Limit K<sup>+</sup> intake
  - Salt restriction+/-
  - Normal protein diet
  - Normal CHO , Lipid
- ✓ Input /output chart
- ✓ Check BP more regularly( initially 2 hourly)
  - BP> 95<sup>th</sup> centile for age, sex and height
  - If BP is more than 99<sup>th</sup> centile need immediate treatment
- ✓ Symptomatic management of fever, pain and vomiting if present
- ✓ Eliminate the carrier stage of strep infection - oral penicillin for 10 days
- ✓ Start anti-hypertensives
  - Diuretic – Frusemide (oral solution 1 hour to act and the peak action after 4 hours. If given IV act within 30 minutes)
  - Anti-hypertensives
    - Slow releasing Nifedipine (20mg or 10mg tablet) – Check BP before giving & give in seated position
    - Atenolol
    - Captopril – Exclude bilateral renal artery stenosis, Look for postural hypotension. Start with low dose.

### GIVE ANTI-HYPERTENSIVES AROUND THE CLOCK

### Management of complications

#### 1. Hypertensive encephalopathy

##### *Clinical features*

- ✓ Drowsy, confusion, altered level of consciousness, convulsion

##### *Management*

- ✓ BP should be reduced slowly
- ✓ Manage with IV anti hypertensives
- ✓ Preferably IV labitalol (not freely available)
- ✓ **IV hydralazine (0.2 mg/Kg)** – Arteriolar dilator→Reduce the peripheral vascular resistance
- ✓ In first 6 hours bring BP down by 1/3 ( otherwise cerebral perfusion pressure won't be maintained)
- ✓ Frusemide 2mg/Kg
- ✓ Control convulsions with anti-convulsants(diazepam/paraldehyde)

## 2. Management of acute renal failure

### Clinical features

- ✓ Rapid reversible reduction in renal function( $UOP < 0.5 \text{ ml/kg/day}$ ) for more than 6 hours with features of impaired renal functions.
- ✓ S.creatinine ↑

### Management

- ✓ Restrict fluid
- ✓ Add insensible loss + last 24 hour urine output
- ✓ Management of hyperkalaemia
  - Restrict  $K^+$  containing food
  - 10% calcium gluconate 0.5ml/Kg over 10 minutes
  - Nebulise with salbutamol
  - Insulin + dextrose infusion
  - $K^+$  binding resins
- ✓ Management of metabolic acidosis
  - Spontaneously corrected when the hyperkalaemia corrected
  - If not give 4.2 %  $\text{NaHCO}_3$  IV (If base deficit less than 10) – Double dilute the 8.4% solution & prepared
- Adverse effects
  - Irritable to veins
  - Hypernatremia
  - Exacerbate intra-cerebral acidosis
- ✓ In persistent ARF → Urgent peritoneal or haemodialysis is needed

## 3. Management of acute LVF

- ✓ Ix – X-ray telechest → Look for ventricular dilatation
  - A - Alveolar oedema
  - B – Bats wing appearance
  - C – Cardiomegaly, curly B lines
  - D – Upper lobe blood diversion
  - E - Effusion

### Management

- ✓ Prop-up
- ✓ High flow  $O_2$  via face mask
- ✓ IV frusemide

### Discharge

- BP should be normal
- Oedema should be settled
- Should be in polyuric phase

### Follow-up

- AGN doesn't need follow-up.
- Within 2 weeks hypertension settles.
- Macroscopic haematuria settles in 2 weeks.
- Microscopic haematuria can persist for one year.
- Complement levels should be normal within 6-8 weeks.
- If C3 persistantly low or haematuria persist → Further investigations needed such as ANA, Renal biopsy

### Cause for recurrent haematuria

- Alport's syndrome
- IgA nephropathy
- Thin glomerular basement membrane disease
- Idiopathic hypercalciuria

## Discussion

1. What are the diseases which commonly presents as acute nephritic syndrome?

- Post infectious GN
- IgA nephropathy
- Membranoproliferative GN
- HSP, SLE
- Wegener's granulomatosis
- Microscopic polyarteritis nodosa
- Goodpasture's syndrome
- HUS

2. Why family history is important in this type of patient?

Hereditary glomerular disease

- Hereditary nephritis (Alport syndrome:)
- Thin glomerular basement membrane disease
- IgA nephropathy

Other renal diseases with hereditary component

- PCKD
- Sickle cell diseases/trait

3. What are the complications of PSGN?

- Hypertension
- Hypertensive encephalopathy
- Acute LHF
- Hyperkalaemia
- Hyperphosphataemia
- Hypocalcaemia
- Acidosis
- Seizure
- Uraemia

### Acute Poststreptococcal Glomerulonephritis

- characterized by the sudden onset of gross hematuria, edema, hypertension, and renal insufficiency
- follows infection of the throat or skin by certain "nephritogenic" strains of group A β-hemolytic streptococci
- Acute glomerulonephritis may also follow infection with coagulase-positive and coagulase-negative staphylococci, *Streptococcus pneumoniae*, and gram-negative bacteria. Bacterial endocarditis may produce a hypocomplementemic glomerulonephritis with renal failure. Acute glomerulonephritis may occur after certain fungal, rickettsial, and viral diseases, particularly influenza

### **PATHOLOGY**

- Kidneys appear symmetrically enlarged
- All glomeruli appear enlarged and relatively bloodless and show diffuse mesangial cell proliferation with an increase in mesangial matrix
- Polymorphonuclear leukocytes are common in glomeruli during the early stage of the disease
- Crescents and interstitial inflammation may be seen in severe cases
- These changes are not specific for post-streptococcal glomerulonephritis
- Immunofluorescence microscopy reveals lumpy-bumpy deposits of immunoglobulin and complement on the glomerular basement membrane (GBM) and in the mesangium
- On electron microscopy, electron-dense deposits, or “humps,” are observed on the epithelial side of the GBM

### **CLINICAL MANIFESTATIONS**

- Most common in children aged 5–12 yr
- Uncommon before the age of 3 yr
- Typical patient develops an acute nephritic syndrome 1–2 wk after an antecedent streptococcal pharyngitis or 3–6 wk after a streptococcal pyoderma
- Renal involvement varies from asymptomatic microscopic hematuria with normal renal function to acute renal failure
- Depending on the severity of renal involvement, patients may develop various degrees of edema, hypertension, and oliguria
- Patients may develop encephalopathy and/or heart failure owing to hypertension or hypervolemia
- Edema typically results from salt and water retention; nephrotic syndrome may develop in 10–20% of cases
- Nonspecific symptoms such as malaise, lethargy, abdominal or flank pain, and fever are common.
- The acute phase generally resolves within 6–8 wk. Although urinary protein excretion and hypertension usually normalize by 4–6 wk after onset, persistent microscopic hematuria may persist for 1–2 yr after the initial presentation.

### **DIAGNOSIS**

#### Urinalysis

- Red blood cells (RBCs)
- Frequently in association with RBC casts
- Proteinuria
- Polymorphonuclear leukocytes

mild normochromic anemia may be present from hemodilution and low-grade hemolysis

serum C3 level is usually reduced in the acute phase and returns to normal 6–8 wk after onset

Confirmation of the diagnosis requires clear evidence of invasive streptococcal infection

- Positive throat culture report may support the diagnosis or may simply represent the carrier state
- Rising antibody titer to streptococcal antigen(s) confirms a recent streptococcal infection
- Antistreptolysin O titer is commonly elevated after a pharyngeal infection
- Antibody titer to document cutaneous streptococcal infection is the anti-deoxyribonuclease (DNase) B level

The Streptozyme test (Wampole Laboratories, Stamford, CT) is a useful and simple diagnostic test that detects antibodies to streptolysin O, DNase B, hyaluronidase, streptokinase, and nicotinamide-adenine dinucleotidase using a slide agglutination test.

DISEASES	POSTSTREPTOCOCCAL GLOMERULONEPHRITIS	IGA NEPHROPATHY	GOODPASTURE SYNDROME	IDIOPATHIC RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS (RPGN)
Clinical manifestations				
Age and sex	All ages, mean 7 yr, 2 : 1 male	10–35 yr, 2 : 1 male	15–30 yr, 6 : 1 male	Adults, 2 : 1 male
Acute nephritic syndrome	90%	50%	90%	90%
Asymptomatic hematuria	Occasionally	50%	Rare	Rare
Nephrotic syndrome	10–20%	Rare	Rare	10–20%
Hypertension	70%	30–50%	Rare	25%
Acute renal failure	50% (transient)	Very rare	50%	60%
Other	Latent period of 1–3 wk	Follows viral syndromes	Pulmonary hemorrhage; iron deficiency anemia	None
Laboratory findings	↑ ASO titers (70%)	↑ Serum IgA (50%)	Positive anti-GBM antibody	Positive ANCA in some
	Positive streptozyme (95%)	IgA in dermal capillaries		
	↓C3–C9;normal C1, C4			
Immunogenetics	HLA-B12, D “EN” (9) <sup>[*]</sup>	HLA-Bw 35, DR4 (4) <sup>[*]</sup>	HLA-DR2 (16) <sup>[*]</sup>	None established
Renal pathology				
Light microscopy	Diffuse proliferation	Focal proliferation	Focal → diffuse proliferation with crescents	Crescentic GN
Immunofluore	Granular IgG, C3	Diffuse	Linear IgG,	No immune deposits

DISEASES	POSTSTREPTOCOCCAL GLOMERULONEPHRITIS	IGA NEPHROPATHY	GOODPASTURE SYNDROME	IDIOPATHIC RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS (RPGN)
scence		mesangial IgA	C3	
Electron microscopy	Subepithelial humps	Mesangial deposits	No deposits	No deposits
Prognosis	95% resolve spontaneously	Slow progression in 25–50%	75% stabilize or improve if treated early	75% stabilize or improve if treated early
	5% RPGN or slowly progressive			
Treatment	Supportive	Uncertain (options include steroids, fish oil, and ACE inhibitors)	Plasma exchange, steroids, cyclophosphamide	Steroid pulse therapy

## UTI IN CHILDREN

**General information;**  
Name, Age , Sex, Residence

P/C

H/P/C

New born	Infancy & early childhood	Older children
Poor weight gain	Feeding problems(FTT)	Fever + chills & rigors
Prolonged jaundice	Excessive crying	Dysuria
Vomiting	Crying while urination	Frequency
Diarrhoea	Whining while urination	Urgency
Feeding refusal	Straining while micturition	Cloudy urine
Irritability	Offensive & cloudy urine	Offensive urine
septicaemia	Irritability	Haematuria
	Vomiting	Enuresis(secondary)
	Diarrhoea	Suprapubic pain
	Fever >38.5C + rigors	
	Febrile convulsions	
	Ballooning of prepuce	

Ask about onset, duration and progression.

### Risk factors

Predisposing conditions	Symptoms/signs
Urinary stasis	Infrequent voiding
Outflow obstruction(mechanical/functional)	Hesitancy Poor stream dribbling
Poor intake	
Constipation	
Poor personal hygiene	Wiping perineum back to front
Reduced host defence	Recurrent infections
Instrumentation/trauma	

### Also look for the any serious underlying pathology

- Recurrent UTI
- Voiding dysfunction and poor urine flow
- Evidence of hydronephrosis
- Palpable bladder
- Evidence of CRF

### Complications

- HT
- Pyelonephritis
- Septicaemia

(Ask about these symptoms also)

Type	Clinical features
<b>Lower urinary tract infection</b>	Dysuria, urgency, frequency, suprapubic pain, incontinence, and mal-odorous urine, usually no fever
<b>Pyelonephritis</b>	Abdominal or flank pain, fever with chills and rigors, malaise, nausea, vomiting, and, occasionally diarrhoea
<b>Asymptomatic bacteriuria</b>	

#### PMHx;

- Recurrent UTI
- Any urinary tract abnormality detected previously
- Constipation
- Recurrent infections
- Any neurological disease
- DM

#### Birth Hx

- Antenatal scan- renal anomalies
- Oligohydramnios- PUV

#### Developmental Hx

- Detailed

#### Immunization Hx

#### Drug/allergic Hx

- If the child had UTI previously → was the child on prophylactic drugs( Co-trimoxazole)
- Allergies- if planning for IVU
- Any long term drug-steroid

#### FMHx

- Renal disease(PUV)

#### SHx

- Urinary habits
- Bed wetting
- Hygiene

## Examination

#### GENERAL

- Fever
- Ill health, irritability
- Dehydration

#### CVS

- PR-bounding in septicaemia
- BP-record and plot

#### Abdominal

- Palpable bladder-
  - ✓ Acute retention
  - ✓ Neurogenic bladder
  - ✓ PUV
- Ballotable kidneys(not in PUV)
- Renal angle tenderness
- External genitalia-
  - ✓ Labial adhesions
  - ✓ Phimosis
  - ✓ Signs of inflammation

#### CNS

- Examine the back→ neural tube defects
- To look for associated features of neurogenic bladder
- Spinal defects

## Investigations

	Supportive	Specific
	<ul style="list-style-type: none"> <li>• Urinalysis           <ul style="list-style-type: none"> <li>✓ UFR</li> <li>✓ Dipstick</li> </ul> </li> <li>• FBC</li> <li>• USS-KUB</li> </ul>	Urine culture and ABST(usually take 3 day for the results to come)

BU/SE  
S.creatinine  
Blood culture  
LP(if indicated)

} ill or septic child

### Advice to parents on collection of CCMS urine sample

- Wash hands and genitalia with water - No antiseptics  
(Retract the prepuce of the older boys)
- Do not wash the urine culture bottle and do not leave the lid opened for a long time
- Send first few ml of urine out and collect a mid stream specimen directly into the sterile culture bottle without contamination
- Close the cap and hand over immediately

## Transportation

- Method and time\* of collection must be stated in the request form  
(\*to be written by the nursing officer who takes the sample)
- Send immediately to the lab
- If the specimen cannot be transported within 2 hours, refrigerate immediately at 4°C – **maximum time of refrigeration is 24 hours**

Investigation	
Urine culture and ABST	Gold standard CCMS→>10 <sup>5</sup> colony forming units/ml of single pathogen or 10 <sup>4-5</sup> /ml + symptoms catheter sample→>10 <sup>5</sup> CFU/ml + positive UFR + symptoms Supra-pubic aspiration→ Any number
UFR	Colour-red, straw Appearance- turbid Specific gravity Pus cells- >10 HPF(significant)(confirmatory than diagnostic) RBC->5 in un-centrifuged />10 in centrifuged sample Organisms- Protein-transient Casts – pus cell casts (pyelonephritis) Granular casts (tubular disease)
FBC	Leucocytosis in pyelonephritis
USS-KUB	In all with UTI To exclude hydronephrosis , size of the kidneys Urgent in neonates, complicated UTI , children with distended bladder, ballotable kidneys

## Management

### Admission criteria

1. Neonates and young infant
2. Ill, toxic and dehydrated child
3. Persistent vomiting
4. Symptoms and / or signs suggestive of obstruction or calculi

## Specific features suggesting upper tract involvement

- Unexplained fever ( $>38.5^{\circ}\text{C}$ )
- General ill health/ toxicity
- Loin pain / tenderness
- + CRP or ESR

### General

- QHT
- Control fever and pain - tepid sponging, PCM 0.15mg/kg 6hourly
- Input - output chart
- Avoid dehydration
  - If tolerate give orally
  - If not IV
- Vomiting - domperidone

### Specific management

#### Principles

- ✓ Valid urine sample prior to AB
- ✓ Empirical AB in correct dosage in suspected febrile UTI(pyelonephritis)(can delay till culture result in afebrile)
- ✓ Do not use urinary antiseptics to treat febrile UTI
- ✓ Use IV AB in neonates, ill and septic child and who are not taking orally
- ✓ Use aminoglycosides cautiously in children with renal impairment
- ✓ Duration is 7 days

## Oral antibiotics for treatment of UTI

Medication	mg/kg/day	Doses/day
Cefixime	8	2
Cephalexin	30-60	3
Co-amoxyclav	20-40 oral Amoxy content 25-45 oral Amoxy content	3 2 (duo)
Cotrimoxazole	6-10 trimethoprim or 30-50 sulphamethoxazole	2

## Parenteral antibiotics for treatment of UTI

Medication	mg/kg/day	Doses/day
Cefotaxime	100-150	2
Ceftriaxone	50-100	1-2
Cefuroxime	50-100	3
Co-amoxyclav	50-100	3
Gentamicin	7.5	3

Assess the treatment response clinically by;

- ✓ Fever
- ✓ Irritability
- ✓ Feeding
- ✓ Urinary frequency

If no response within 48 h

- ✓ Check the dose of the drug
- ✓ Check the compliance
- ✓ Check ABST
- ✓ Do a USS to exclude any predisposing urinary tract abnormality or any complication of UTI(Abscess)
- ✓ Repeat urine culture

### Discharge plan

Repeat U. culture before discharge

Educate on prevention

- ✓ Importance of increasing fluid intake maintaining good urine output
- ✓ Importance of continuing prophylaxis to prevent further attacks
- ✓ Hygiene-
  - Wiping perineum front to back
  - Frequent urination
  - Double micturition in children with VUR
- ✓ Avoid constipation-encourage fluid and dietary modification

### Start prophylaxis

- Drugs are given as a single dose in the night.
- Antibiotic prophylaxis is indicated for **all children below 5 years** following the **first attack of UTI until an USS of the kidneys is available**. Continuation of prophylaxis is decided according to following factors.
- **First attack <1 year**
  - a. Infants → with **afebrile UTI** → If the USS is normal → stop the prophylaxis and follow up without further investigations.
  - b. In infants → with **febrile UTI** → It is continued till recommended imaging studies are available or until their first birthday; whichever comes last.
  - c. Those with **structural abnormalities or recurrent UTI** need **prophylaxis till 5 years or longer**.
- **First attack between 1-5 years**
  - a. Children → afebrile or a simple febrile UTI → normal USS → will be followed up without prophylaxis or further investigations.
  - b. Structural abnormalities or recurrent UTI → need prophylaxis till 5 years or longer.

## Antibiotics for prophylaxis of UTI

Medication	mg/kg/dose	Remarks
Cephalexin	10	Recommended drug in first 1–3 months until trimethoprim is available in Sri Lanka
Cotrimoxazole	2 (trimethoprim)	Avoid in infants <1 month age
Trimethoprim	2	Recommended in neonates; but require specialist supervision
Nalidixic acid	12.5	Avoid <6 months
Nitrofurantoin	1–2	Avoid in infants <3 months age

### Follow up plan

R/V monthly at clinic with a culture

- ✓ Once a month in first three months
- ✓ Every 3/12 in infants and young children up to 1 year
- ✓ In older children if symptoms present only.

Clinical assessment-bowel and voiding habits, growth assessment

Continue prophylaxis-single dose daily at night

### DMSA

6 month after an acute attack **to detect scarring**

#### Indications

- Under 1 year of age
  - ✓ All children with febrile UTI
- Under 5 years of age
  - ✓ Clinical picture is highly suggestive of acute pyelonephritis other than fever
  - ✓ Second attack of febrile UTI before 5 years of age
  - ✓ Abnormal USS: hypoplastic / dysplastic kidneys, scarring
  - ✓ Other structural anomalies: VUR, duplex system, ureterocele etc.

If scar +ve followup with UFR, BP, growth while continuing prophylaxis

### MCUG

Has to be done after 6 wks except BOO (immediate)

Best investigation **to detect abnormalities of urethra and bladder** and it is also **useful in detecting and grading VUR**.

#### Pre procedure

- ✓ Recent U. culture (5 days before) must be negative
- ✓ Prophylaxis AB should be converted to therapeutic dose 3 days prior
- ✓ Exclude phimosis and labial adhesions
- ✓ Allergic Hx-important(IV hydrocotizone??

#### Indications

- Suspected BOO
- Dilated ureter / dilated ureter and dilated pelvicalyceal system in the USS under 5 years of age
- Abnormal DMSA scan:
  - Significant scarring under 5 years of age
  - Dysplastic kidneys
- First attack of febrile UTI under 1 year of age and DMSA scan is not available

- Second attack of febrile UTI under 5 years of age
- Clinical picture is highly suggestive of acute pyelonephritis of children below 5 years and DMSA scan is not available.

At least 3 films	Can detect
✓ Full bladder	✓ PUV
✓ Micturating	✓ VUR
✓ Post micturating	

#### DPTA

Suspected pelvi-ureteric/VU junction obstruction

#### X'ray KUB

UTI associated with recurrent microscopic or macroscopic haematuria

### Discussion

- Common in **female** (1<sup>st</sup> attack by the age of 5yrs in female, before 1yr in male)
- Common in uncircumcised
- important risk factor for the development of **renal insufficiency or end-stage renal disease** in children
- Virtually all UTIs are **ascending infections** Rarely, renal infection may occur by hematogenous spread(IE)
  - ✓ Simple and compound papillae in the kidney have an **anti-reflux** mechanism.
  - ✓ However, some compound papillae, typically in the **upper and lower poles** of the kidney, allow **intra-renal reflux**

The 3 basic forms of UTI are pyelonephritis, cystitis, and asymptomatic bacteriuria

#### Pyelonephritis

- Involvement of the **renal parenchyma** is termed *acute pyelonephritis*, whereas if there is no **parenchymal** involvement, the condition may be termed **pyelitis**
- Abdominal or flank pain, fever with chills and rigors, malaise, nausea, vomiting & occasionally diarrhea
- Newborn(nonspecific) symptoms such as poor feeding, irritability, and weight loss.
- Pyelonephritis is the most common serious bacterial infection in infants <24 mo of age who have fever without a focus.
- Acute pyelonephritis may result in renal injury, termed *pyelonephritic scarring*.
- **Acute lobar nephronia** (acute lobar nephritis) is a localized renal bacterial infection involving >1 lobe that represents either a complication of pyelonephritis or an early stage in the development of a renal abscess. Manifestations are identical to pyelonephritis
- **Renal abscess** may occur following a pyelonephritis or may be secondary to a primary bacteremia (*S. aureus*).
- **Perinephric abscesses** may be secondary to contiguous infection in the perirenal area (e.g., vertebral osteomyelitis, psoas abscess) or pyelonephritis that dissects to the renal capsule

#### Cystitis

- Indicates that there is bladder involvement;
- Symptoms include dysuria, urgency, frequency, suprapubic pain, incontinence, and malodorous urine. Cystitis does not cause fever and does not result in renal injury.
- Malodorous urine, however, is not specific for a UTI

### **Asymptomatic bacteriuria**

- Refers to a condition that results in a positive urine culture without any manifestations of infection.
- It is most common in girls.
- The incidence declines with increasing age.
- This condition is benign and does not cause renal injury, except in pregnant women, in whom asymptomatic bacteriuria, if left untreated, can result in a symptomatic UTI.

### **Aetiology**

- E.coli 70-90%
- Klebsiella aerogenes
- Proteus mirabilis
- Strep. faecalis
- Pseudomonas
- Viral – adenovirus acute cystitis
- TB

### **Risk factors**

- |   |   |
|---|---|
| <ul style="list-style-type: none"><li>✓ Female gender</li><li>✓ Vesicoureteric reflux</li><li>✓ Toilet training-voiding dysfunction</li><li>✓ Voiding dysfunction-infrequent/incomplete voiding</li><li>✓ Obstructive uropathy</li><li>✓ Urethral Instrumentation</li><li>✓ Poor hygiene- wiping perineum back to front</li></ul> | <ul style="list-style-type: none"><li>✓ Constipation-voiding dysfunction</li><li>✓ Anatomic abnormalities-labial adhesions</li><li>✓ Neuropathic bladder</li><li>✓ Immunosuppression</li><li>✓ Tight clothing</li></ul> |
|---|---|

### **Xanthogranulomatous pyelonephritis**

- Is a rare type of renal infection characterized by granulomatous inflammation with giant cells and foamy histiocytes.
- It may present clinically as a renal mass or an acute or chronic infection.
- Renal calculi, obstruction, and infection with *Proteus* spp. or *E. coli* contribute to the development of this lesion, which usually requires total or partial nephrectomy.

### **Acute hemorrhagic cystitis**

- Often is caused by *E. coli*; it also has been attributed to adenovirus types 11 and 21.
- Adenovirus cystitis is more common in males;
- it is self-limiting, with hematuria lasting approximately 4 days

### **Eosinophilic cystitis**

- is a rare form of cystitis
- The usual symptoms are those of cystitis with hematuria, ureteral dilation with occasional hydronephrosis, and filling defects in the bladder caused by masses that consist histologically of inflammatory infiltrates with eosinophils.
- Children with eosinophilic cystitis may have been exposed to an allergen.
- Bladder biopsy often is necessary to exclude a neoplastic process.
- Treatment usually includes antihistamines and nonsteroidal anti-inflammatory agents, but in some cases intravesical dimethyl sulfoxide instillation is necessary.

### Definitions

Significant bacteriuria	Colony count of $>10^5/\text{ml}$ of a single species in a midstream clean catch sample
Asymptomatic bacteriuria	Presence of significant bacteriuria in two or more specimens in a child with no symptoms.
Complicated UTI	Toxicity, persistent vomiting, dehydration, renal angle tenderness, renal impairment, clinical non response to treatment after 48 hours
Simple UTI	UTI with low grade fever, dysuria, frequency, urgency but none of the above symptoms

### Management of recurrent attacks of UTI

- Confirm the diagnosis, and obtain a urine sample via a SPA for repeat culture, in children not potty trained.
- Treat promptly with an appropriate antibiotic pending the culture report.
- Identify correctable risk factors e.g.: constipation, poor hygiene, inappropriate voiding practices etc
- Treat phimosis or labial adhesions appropriately. (They lead to false positive culture reports)
- Imaging studies as indicated.
- Check the compliance, especially if the urine culture yields organisms sensitive to the prophylactic antibiotic used.

### **Vesico Ureteric Reflux (VUR)**

VUR can be a risk factor for recurrent UTI. With bladder growth and maturation there is a tendency for reflux to resolve or improve.

#### **Management of VUR:**

- a. Prophylaxis is recommended for VUR till 5 years of age. Longer regimes are indicated for recurrent UTI.
  - b. Advice on double micturition.
  - c. Recurrent attacks of UTI need prompt treatment.
  - d. A repeat DMSA scan to assess new scar formation might be recommended in case of repeat attacks of febrile UTI
  - e. Repeat MCUG to assess the improvement of reflux is not usually recommended unless there is a plan for surgery.
- **There is no world wide consensus regarding the indications for surgical intervention in VUR. Each patient with reflux has to be assessed individually.**

#### **Definite indications**

- a. Recurrent break through infections

#### **Relative indications**

- a. Poor compliance for prophylaxis
- b. Recurrent infections in spite of prophylaxis.
- c. New scar formation
- d. Impaired renal function
- e. Persistent gross VUR (Grade IV - V) or persistent moderate VUR (grade III) with recurrent infections after discontinuation of prophylaxis

## Acute Flaccid Paralysis

**PC – B/L lower limb weakness [duration]**

### Differential Diagnoses

Poliomyelitis

GBS

Transverse myelitis

Channelopathies

Cord compression

**HPC-**

- Onset – sudden/ gradual
- Progression of the weakness- rapid or slow, ascending, proximal / distal
- Neurological impairment at each stage in chronological order based on the activities of the child-running, playing, walking ,sitting, standing up from sitting position, other day to day activities.
- Any sensory loss- sensory level, numbness, tingling
- Bladder & bowel incontinence

### Differential Diagnoses

- Poliomyelitis (Affects anterior horn cells)
  - ✓ Hx of fever, URTI, GI symptoms
    - ↓
    - Symptom free period for 2-3d
    - ↓
    - Recurrence of fever, muscle tenderness, neck stiffness, paralysis (usually asymmetric)
  - ✓ Vaccination against polio (both prevention & aetiology), Contact history
  - ✓ Management – Stool culture done

- GBS

#### **Symptoms**

- ✓ Acute onset
- ✓ Difficulty in standing up from sitting position or climbing stairs (proximal muscle weakness), unable to walk
- ✓ Initially weakness of legs, symmetrical
- ✓ Progresses upwards & may be weakness of all 4 limbs
- ✓ Rapidly progressive - usually 4 weeks
- ✓ Distal paraesthesia, tingling, burning sensation of extremities

#### **Complications**

- ✓ Autonomic symptoms
  - Incontinence or retention of urine & stools
  - Dizziness from sitting up – postural hypotension
  - Nocturnal diarrhea
- ✓ Tightness of chest, respiratory depression – respiratory muscle involvement
- ✓ Diplopia, drooling of saliva - cranial nerve involvement 3, 4, 6, 7

- ✓ Lower cranial nerve weakness - difficulty handling secretions and maintaining an airway

**Aetiology**

- ✓ Hx of respiratory infection (Mycoplasma, EBV, CMV, Varicella zoster) or blood & mucous diarrhea (Campylobacter jejuni) 1 – 3 weeks ago
- ✓ Hx of rabies or influenza vaccine

- Channelopathies

- ✓ Previous attacks-Periodic paralysis
- ✓ After heavy carbohydrate meal
- ✓ Trigger events
  - Potassium excretion - severe sweating (climbing mountain)
  - High temperature
  - Excitement
- ✓ Notices the symptoms on next day
- ✓ Difficulty in speech, swallowing & breathing
- ✓ Rapid improvement in young patients

- Transverse myelitis

- ✓ Bladder & bowel symptoms - incontinence
- ✓ History of viral infection – chicken pox, HIV
- ✓ History of CTD, SLE
- ✓ Radiation

- Paralytic rabies

- ✓ History of animal bite
- ✓ Fever, agitation, abnormal behavior
- ✓ Headache, malaise

- Cord compression/Trauma

- Low back pain
- Weakness of the legs
- Sensory loss below the compression, numbness
- Urinary symptoms - hesitancy, frequency, painless retention
- Bowel symptoms – constipation

What was done to the child up to hospital admission state the progression of the neurological symptoms

Ask the complications of the disease, specifically for bulbar involvement, respiratory muscle weakness

Describe specific Mx done – IVig

Present state of child, in regard to functional state

**Past medical and Surgical Hx-** recent respiratory /GI infection, similar episodes

**Birth Hx**

**Growth and development Hx**

**Immunization Hx-** Present state of immunization -Polio vaccination, Recent anti rabies vaccination

**Dietary history**

**FHx**

**SHx**

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**General introduction of family**

**Impact of disease on child**

**Impact of disease on parents**

**Impact of disease on siblings**

<b>Environmental factors</b>	(Paralysis-) steps to get in to house, mountain, accessibility to house Indoor-steps, obstacles and hazards to child Toilet-attached/separate ,distance, steps, commode, accessibility to wheel chair Method of traveling to school, location of class room, accessibility, type of toilets available in school, accessibility to physiotherapy
<b>Supports available</b>	
<b>Education of parents – disease, drug Rx</b>	
<b>Expectations</b>	
<b>Psychological state of child /parents</b>	

## Examination

- Weight, height
- General appearance – wasted/not
- Hygiene- kept clean/unclean
- Urine smell/fecal
- Pressure sores
- Nutritional status/features of nutritional deficiencies
- Hydration
- NG/cannula/catheter/sand bags
- Spinal surgery scar
- Dyspnoeic, cardiac monitor
- Febrile
- In pain
- Conscious

Disease	GBS	Transverse myelitis	Cord compression	Poliomyelitis
<b>Cranial nerves</b>	External ophthalmoplegia in Millar fisher syndrome B/L VII nerve palsy Bulbar weakness	Normal	Normal	Normal
<b>Muscle wasting</b>	Absent			
<b>Tone</b>	Flaccid	Flaccid	Initially flaccid Later spastic	
<b>Power</b>	Symmetrically ↓	↓	↓	Asymmetrical weakness
<b>Reflex</b>	Areflexic	Exaggerated	Exaggerated	
<b>Plantar reflex</b>	Down	Up	Up	Down
<b>Sensory</b>	Intact	Sensory level	Sensory level	
<b>Sphincter function</b>	Normal/impaired	Impaired	Impaired	Normal
<b>Cerebellum</b>	(+) ataxia	Normal	Normal	Normal

- Poliomyelitis
  - CNS
    - Weakness
      - Asymmetric involvement of LL, hypotonia
      - Proximal>distal weakness
    - Sensory - No sensory involvement
    - Can have associate bulbar weakness
- GBS
  - General
    - Dyspnoeic
  - Respiratory
    - Single breath count
    - Neck muscle weakness- ask to raise the head against resistance
    - Increase RR
    - Reduced chest expansion
  - CNS
    - Gait
    - Cranial nerves- 3, 4, 6, 7
    - Motor-
      - Power reduced
      - Tone reduced
    - Reflexes absent
    - Sensory
      - Distal numbness
      - Few sensory signs
    - Cerebellar signs
      - Ataxia- Miller- fisher syndrome (**Miller fisher Xn – Ataxia, Areflexia, Ophthalmoplegia**)

- Abdomen
  - Distended bladder
  - DRE – sphincter tone

## Investigation

- Poliomyelitis
  - Stool for virus isolation - Polio
- GBS (emergency medicine 153, 154pg) – Clinical diagnosis
  - CSF – raised protein, normal cell count (take 10 days to get the changes)
  - Nerve conduction studies/EMG (Demyelination/Axonal degeneration)

## Management

**Acute flaccid paralysis should be notified to MOH, RE, DDHS over the phone**

- GBS
  - Mainly clinical diagnosis
    - ✓ Nerve Conduction Study – slow conduction / conduction block
    - ✓ LP – CSF shows PROTEIN CELLULAR DISSOCIATION (Positive 10-14 days after the onset)
      - Raised protein 2 – 3 g/l ( normal 40mg/dl)
      - Normal cell count
  - Management
    - Monitoring bed
    - Monitor for complications
      - Respiratory muscle paralysis
        - Respirometer (Measure forced vital capacity)
          - ✓ >75ml/Kg – normal
          - ✓ <20ml/Kg – ICU admission needed
          - ✓ <15ml/Kg – Intubate & ventilate
        - Single breath count : >25 → Normal
        - Assess neck muscle weakness
        - RR, Pulse oxymetry (But changes occur at very late stage)
      - Autonomic instability
        - wide fluctuation in blood pressure, postural hypotension, and cardiac dysarrhythmias
        - So monitor PR, BP, Cardiac monitoring
          - ✓ 4 hourly in progressive stage (1<sup>st</sup> 4 weeks)
          - ✓ 6 hourly in plateau stage
    - Specific management
      - Plasmapheresis
      - IVIg - 0.4g /kg/day x 5 days } Equally effective
      - Steroid no benefit

- Supportive care
  - Pain relief – pains are self-limited and often respond to standard analgesics
  - Fluid
  - Nutrition
  - Bladder & bowel care
  - Prevention of bed sores
  - SC heparin prophylaxis for DVT
- Long term management – Physiotherapy & rehabilitation

**Notification-**

- Any child under 15 yr of age with acute flaccid paralysis should be notified immediately
- Notification to MOH of the area and RE by telephone  
(Investigation form EPID/37/1/R2004 Should be completed and returned to the epidemiological unit)

**Collection of stool samples - As in polio**

**Continuing management-**

- Management of muscle pain
- Bladder and bowel care chest physiotherapy for prevention of chest infections
- Supportive management while in the ward
- Passive physiotherapy-limbs
- Rehabilitation /occupational therapy
- GBS begins spontaneous recovery after 2-3 wks in inverse direction to the direction of paralysis

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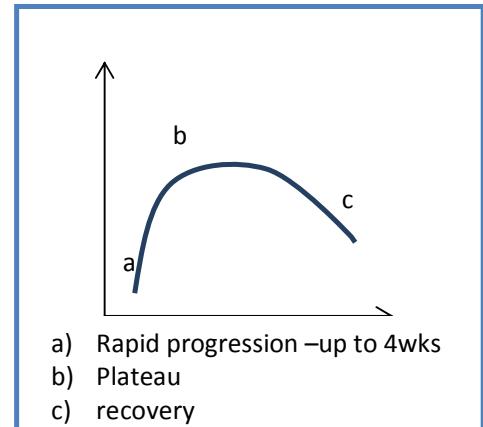
**Prognosis**

- Recovery begins(with or without) treatment between several days & 6 weeks from onset
- Approximately 85% of patients with GBS achieve a full functional recovery within several months to a year
- Improvement towards independent mobility is gradual over many months but may be incomplete
- 15% patients die or are left disabled
- Between 5 and 10% of patients with typical GBS have one or more late relapses; such cases are then classified as chronic inflammatory demyelinating polyradicular neuropathy (CIDP)

## Discussion

### ➤ GBS

- Acute inflammatory demyelinating polyradicular neuropathy
- Self limiting
- Picture
  - Rapid progressive onset
  - Plateau
  - Recovery
- Causes
  - Paralysis following infection
    - ✓ *Campylobacter jejuni* - severe GBS
    - ✓ Virus – CMV, EBV, VZV, HIV
    - ✓ Vaccines - Rabies, influenza
    - ✓ 40 % no cause

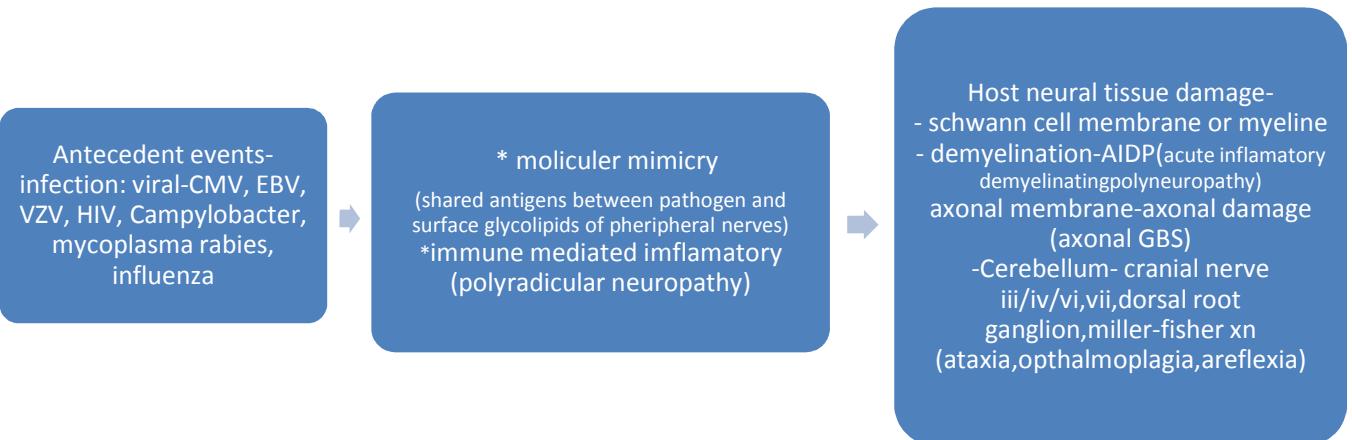


- Infective organisms induce antibody response against peripheral nerves
- Pathogenesis – Molecular mimicry
 

Shared antigens between pathogen & surface glycolipids of peripheral nerves.

Immune mediated inflammatory polyradiculo-neuropathy

The target epitopes – Myelin, Axonal membrane, Cerebellum, Cranial nerve nuclei (3,4,6,7), Dorsal route ganglion.



- Other neurological involvement
  - Cerebellum
  - Cranial nerves 3, 4, 6 dorsal root ganglia
  - Miller – fisher syndrome
    - ✓ Ataxia
    - ✓ Ophthalmoplegia
    - ✓ Areflexia

- Distal limb weakness or distal numbness
- Ascends proximally – days to 6 weeks
- Areflexia, sensory symptoms, few sensory signs
- 50 % facial weakness
- Severe
  - Respiratory & bulbar involvement
  - Need ventilation if vital capacity drops to 1L (<15ml/kg) or below
  - No signs of dyspnoea
- Protein – cellular dissociation
- NCS –slow conduction velocities or conduction block (MCQ)
- Changes best seen after 10 – 14 days
- GBS unlikely
  - Marked asymmetry of signs
  - Sensory level
  - Persistent bladder, bowel dysfunction
  - Fever at onset
  - CSF cells >50
- High dose IV IgG within first 2 weeks reduce duration & severity of paralysis
- Screen for IgA deficiency prior to Ig
  - Severe allergy reaction to IgG
  - Angina or MI can be precipitated by Ig

## Poliomyelitis- (last reported case in SL-1993)

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- Enterovirus : Viral subtypes 1, 2 and 3
- Transmission - Person to person via faeco-oral route
- Infection starts in the GI tract → invades the nervous system → damages the anterior horn cell
- Permanent asymmetric paralysis of (usually lower) limbs (incidence =1/200)
- Respiratory failure and death (5-10%)
- Time between infection and onset of paralysis – 10-21 days
- Minor illness-
  - Fever
  - Malaise
  - Sore throat
  - Vomiting
  - Abdominal pain
  - Loss of appetite
- Major illness - During initial stages this will resemble aseptic meningitis (LP done at this stage will show raised cell counts and raised protein)
  - 1) Spinal paralysis-
    - Cervical and lumbar segments are mostly involved,
    - Results in patchy asymmetrical paralysis of limbs.(very mild to severe paralysis)
    - Involvement of inter-costal muscles lead to breathing difficulty

- Involvement of cervical segments results in sudden worsening of respiratory distress
  - Sensory involvement can occur but rare
- 2) Bulbar paralysis-
- Associated with tonsillectomy
  - Difficulty in swallowing is the cardinal sign
  - Respiratory center and circulatory centers can be involved with fatal outcome
- 3) Combined bulbar and spinal paralysis

**Management-**

- Isolation with proper excretion of excreta
- Notification
- Stool samples (Viral excretion in stools maximum during first 2 weeks after initial symptoms)
  - Stool samples should be collected within 2 weeks from the onset of paralysis.
- Passive physiotherapy – start early
- Active physiotherapy once fever is settled
- Support paralytic limbs and splints
- Nutrition
- Monitor for evidence of respiratory failure/BP, pulse/Air way management

Follow-up at 60, 90 and 180 days (stool sampling)

**Notification-**

- On clinical diagnosis every case of AFP notified to the Epidemiologist immediately by telephone, telegram, fax or email
- to RE and MOH
- Notification done by MO in attendance (HO, MO or Specialist) – using special form
- Notification form filled and sent via fax/ post to Epidemiological Unit

**Stool sampling-**

- Stool sampling2 samples 24-48 hours apart, within 2 weeks of onset of paralysis
- 6-8 g (quantity of 2 thumbnails/ 2 tamarind seeds)
- Clean screw capped bottle; lid tightly closed to prevent leakage and drying(a special container)
- Clear correct labeling –introduction as in any sample, date of onset of paralysis, date of collection of sample , date of dispatch of stools, last date of polio vaccination
- Packed in a container with ice ,transport to MRI within 72hr of collection
- MOH-Stool samples also from 3-5 immediate contacts of a case
- One sample each from contacts

**Vaccination-**

- Vaccination Trivalent OPV (Sabin)
- IPV- killed polio virus (Salk)
- Live attenuated virus

**Outbreak response immunization**

- Limited outbreak response
- Day after the Ix
- House-to-house immunization of all children under age of the AFP case within 2 km radius of his/ her residence (250-300 people)

- Only one dose of OPV
- Contacts immunized after stool sampling

## Transverse myelitis

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- This term is used to describe a cord lesion and paraparesis(or paraplegia) occurring with viral infections, MS, mixed
- Connective tissue disease and other inflammatory and vascular disorders, e.g. HIV, sarcoid, syphilis, radiation myelopathy and anterior spinal artery occlusion.
- MRI is usually required to exclude cord compression.
- Transverse myelitis is a neurological condition caused by inflammation of the spinal cord resulting in axonal demyelination. It is often develops after viral infection or occur during autoimmune disease
- Varicella zoster, Herpes simples directly invade the spinal cord produce symptoms of transverse myelitis.
- Transverse myelitis is sometimes associated with other diseases, like systemic lupus erythematosus and sarcoidosis
- Usually B/L involvement

### Symptoms and signs-

#### Develop rapidly over a period of hours.

Symptoms include weakness and numbness of the limbs as well as motor, sensory, and sphincter deficits

- Abnormal sensations: Patients report sensations of tingling, numbness, coldness or burning below the affected area below the spinal cord.
  - Pain: Pain is sharp, shooting sensations begins suddenly in neck or back and radiate to legs, arm or abdomen depending on the part of spinal cord that is affected.
  - Weakness of arms or legs: Weakness to severe paralysis of arms and legs depending on the part of spinal cord that is affected.
    - May be total paralysis and sensory loss below the level of the lesion.
- if loss is only partial.
- Upper cervical cord is involved- all four limbs may be involved and there is risk of respiratory paralysis (segments C3, 4, 5 to diaphragm).
  - Lesions of the lower cervical (C5–T1) region will cause a combination of upper and lower motor neuron signs in the upper limbs, and exclusively upper motor neuron signs in the lower limbs.
  - Lesion of the thoracic spinal cord (T1–12) will produce upper motor neuron signs in the lower limbs, presenting as a spastic diplegia.
  - Lesion of the lower part of the spinal cord (L1–S5) often produces a combination of upper and lower motor neuron signs in the lower limbs.
- Bowel and bladder dysfunction
  - Muscle spasms
  - Headache
  - Fever

### **Complications of Transverse myelitis**

- Partial or total paralysis
- Spasticity of muscle
- Osteoporosis
- Depression.

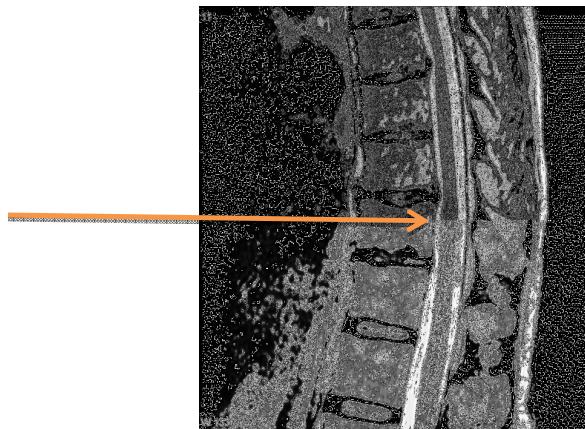
### **Diagnosis-**

- Diagnosis history, physical examination, lab tests and radiological imaging.
- Lumber Puncture: Shows high numbers of white blood cells or protein in the fluid, suggesting an infection or an inflammation.
- Blood test: Shows positive antibodies for neuromyelitis optica.????
- MRI: Shows brain and spinal cord may show inflammation

### **Management-**

- Hospital admission
- Intravenous steroid -IV dexamethasone
- Plasma exchange therapy
- Pain management: NSAID - aspirin, ibuprofen.
- Physical therapy: To increased the muscle power.
- Occupational therapy
- Other: some patients well responds to intravenous cyclophosphamide
- If Transverse myelitis is treated early prognosis is excellent

MRI showing lesion of Transverse myelitis (the lesion is the lighter, oval shape at center-right)



# Channelopathies-

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Diseases caused by disturbed function of ion channel subunits or the proteins that regulate them. These diseases may be either congenital (often resulting from a mutation or mutations in the encoding genes) or acquired (often resulting from autoimmune attack on an ion channel).

**Myotonias**- Characterized by continued, involuntary muscle contraction after cessation of voluntary effort. EMG is characteristic.

Patients with myotonia tolerate general anaesthetics poorly.

**Lambert–Eaton myasthenic–myopathic syndrome**

**Myotonia congenital**

**Hypokalaemic periodic paralysis**

**Hyperkalaemic periodic paralysis**

**Stiff person xn**

## Dystrophia myotonica (DM) or myotonic dystrophy (MD)

Autosomal dominant

Two different triple repeat mutations.

There is a correlation between disease severity, age at onset and approximate size of triple repeat mutations.

Evident between 20 and 50 years.

There is progressive distal muscle weakness, with ptosis, weakness and thinning of the face and sternomastoids.

Muscle disease is part of a syndrome comprising:

- Cataracts
- Frontal baldness
- Cognitive impairment (mild)
- Oesophageal dysfunction (and aspiration)
- Cardiomyopathy and conduction defects (sudden death can occur in type 1)
- Small pituitary fossa and hypogonadism
- Glucose intolerance
- Low serum IgG.

Tx- Phenytoin or procainamide

## Myotonia congenital (Thomsen's disease)

- An isolated autosomal dominant myotonia
- Usually mild, becomes evident in childhood.
- Myotonia, which persists, is accentuated by rest and by cold.
- Diffuse muscle hypertrophy occurs
- Patient has bulky muscles.

## Hypokalaemic periodic paralysis

- This channelopathy is characterized by
- Generalized weakness
- Including bulbar muscles
- Starts after a heavy carbohydrate meal or following exertion. Attacks last for several hours.
- First comes to light in the teenage years and tends to remit after the age of 35.
- Serum potassium is usually below 3.0 mmol/L in an attack.
- Weakness responds to (slow) IV potassium chloride.
- Autosomal dominant trait caused by mutation in a muscle voltage-gated calcium channel gene
- Mutations in the sodium channel and potassium channel
- Acetazolamide sometimes helps prevent attacks.
- Weakness can be caused by diuretics, thyrotoxicosis.

## Hyperkalaemic periodic paralysis

- Autosomal dominant
- Characterized by attacks of weakness, sometimes with exercise. Attacks start in childhood and tend to remit after the age of 20
- They last about 30–120 minutes.
- Myotonia may occur.
- Serum potassium is elevated.
- An attack can be terminated by IV calcium gluconate or chloride.
- There are point mutations in a muscle voltage-gated sodium channel gene
- Acetazolamide or a thiazide diuretic can be helpful.
- A very rare normokalaemic, sodium-responsive periodic paralysis also occurs.

## Stiff person syndrome

- Stiff person syndrome (SPS) is a rare disease, commoner in females, of varying muscular stiffness with abnormal posturing and falls.
- Attacks of stiffness are sometimes provoked by noise or emotion, but sometimes occur spontaneously.
- Between attacks, which last from hours to days or even weeks, the patient may appear normal.
- Widespread muscle stiffness is typical during an attack ; there are no other neurological signs.
- SPS has been mistaken for Parkinson's, dystonia and non-organic conditions.
- Anti-glutamic acid decarboxylase antibodies (anti-GAD) are found in more than 50%.
- Continuous motor activity is seen on EMG.
- Features of SPS remain lifelong. Treatment with diazepam
- Other muscle relaxants and IV immunoglobulin can be helpful during attacks.
- A form of SPS is also seen occasionally as a paraneoplastic condition associated with antibodies to synaptic protein amphiphysin, anti-gephyrin antibodies.

## Constipation

### DDs –

- Functional constipation
- Hypothyroidism – rare
- Hirschsprung's disease
- Spinal cord lesion
- Drugs – narcotics, psychotropics

### Hx -

#### PC – Abdominal pain

Abdominal distension  
Rectal bleeding  
Faecal soiling (overflow incontinence)  
Painful defecation

#### HPC –

- Time of onset of symptoms-
    - ✓ Usually 2-4 yrs of age
    - ✓ Had usual bowel habits before that /not
  - Frequency, consistency & volume of stools
  - Painful defecation, faecal incontinence
  - Straining on passage of stools
  - Associated features-
    - Abdominal pain, distension, anorexia, vomiting, blood stained stools, urinary symptoms
    - Crying while defecation – anal fissures
    - Timing of passage of meconium- passed within 1<sup>st</sup> 24 hrs/not
    - Hirschsprung disease-
      - Failure to pass meconium in first 24 hrs
      - Abdominal distension
      - Vomiting
      - Symptoms of enterocolitis-
        - Fever, foul smelling diarrhea
- In older children- chronic constipation
- Abdominal distension without soiling
  - Growth failure
- Toilet training –
  - Onset of potty training
    - ✓ Fear due to- sound of flushing
    - ✓ Fear of falling in to toilet
    - ✓ Violence during toilet training
  - With holding behavior, retention posturing
  - Any stressful family events just before the onset
    - Eg: Birth of a sibling, death of parents, sexual abuse
  - Hypothyroid Fx – lethargy, wt gain

**Birth Hx –** Hx of prematurity – (risk factor)  
Neonatal constipation  
Anorectal malformation  
Meningomyelocoele at birth

**Developmental Hx -** Look for developmental delay

**PMHx -** Aded pt with CP

**Dietary Hx -** Water & fibre intake

**Drug Hx -** Opiates, anticholinergics

**Social Hx –** Care givers

## **Ex –**

Assess growth & nutritional state - wt, Ht, OFC  
Febrile – (enterocolitis)  
Ill looking distressed child  
Goiter  
Hypothyroid Fx – coarse facies, dry scaly skin

**Abdominal Ex –** Distension  
Palpable masses – faecal mass  
Perianal – Abnormally placed anus  
    Perianal fistula  
    Perianal fissures  
DRE – After sedation of child, usually 5<sup>th</sup> finger  
    Hard stools – functional  
    In Hirshprung disease – Narrow segment  
                Gush of fluid stools & flatus with the withdrawal of examining finger

**Neurological Ex-** LL –in meningomyelocoele

## *Discussion-*

### **❖ Define constipation -**

Considered a symptom, rather than a disease  
Often interpreted as,  
    ✓ Infrequent motions  
    ✓ Passage of hard stools  
    ✓ Less than 3 bowel motions per week  
    ✓ Difficulty in passing stools.

Approximately 0.5% of school children have defecation frequency less than 3 per week and 0.3% have fecal incontinence. 20% have at least 1 clinical feature of constipation.

Therefore, use diagnostic criteria (Rome III criteria) based on multiple symptoms to define constipation.

**Table 1.** Pediatric Rome III Criteria for Constipation

Rome III criteria for neonates and toddlers
Must include 1 mo of at least two of the following in infants up to 4 yr of age:
1. Two or fewer defecations per week
2. At least 1 episode per week of incontinence after the acquisition of toileting skills
3. History of excessive stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large-diameter stools that may obstruct the toilet
Accompanying symptoms may include irritability, decreased appetite and/or early satiety.
The accompanying symptoms disappear immediately following passage of a large stool.
Rome III criteria for children and adolescents
Must include two or more of the following in a child with a developmental age of at least 4 yr <sup>3</sup> with insufficient criteria for diagnosis of irritable bowel syndrome:
1. Two or fewer defecations in the toilet per week
2. At least 1 episode of fecal incontinence per week
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large diameter stools that may obstruct the toilet

\*Criteria fulfilled at least once per week for at least 2 mo before diagnosis.

#### ❖ Causes & risk factors for constipation –

**Table 2.** Causes and Risk Factors of Constipation in Children

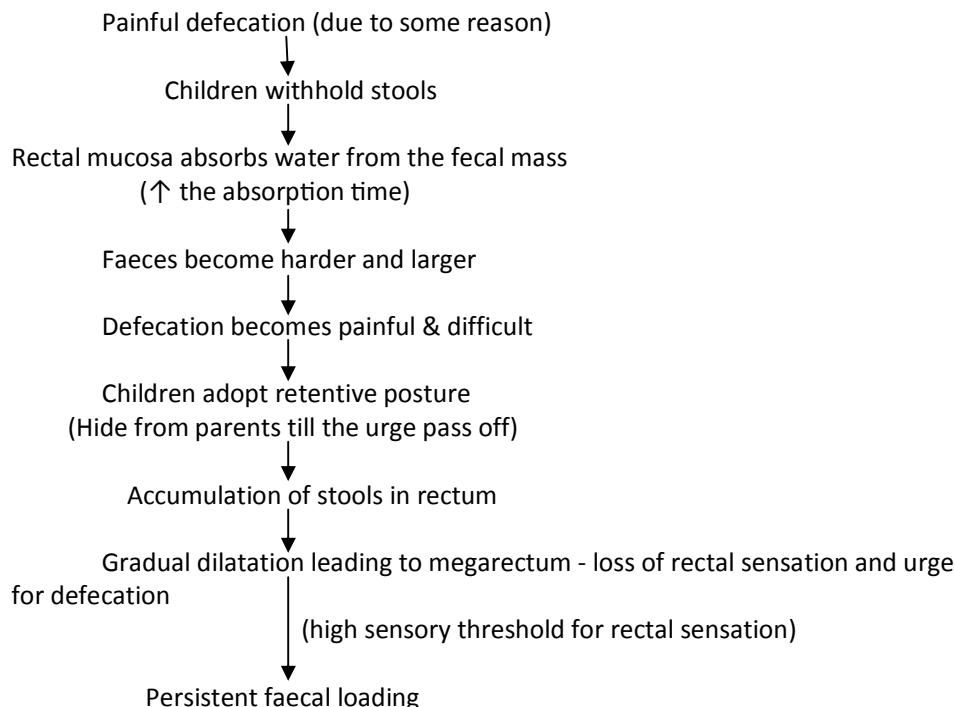
Intestinal causes	Hirschsprung disease Anorectal malformations Neuronal intestinal dysplasia
Metabolic/endocrine causes	Hypothyroidism Diabetics mellitus Hypercalcemia Hypokalaemia Vitamin D intoxication
Drugs	Opioids Anticholinergics Antidepressants
Other causes	Anorexia nervosa Sexual abuse Scleroderma Cystic fibrosis
Risk factors	Low fiber diet Psychological stress Cow's milk protein allergy Familial predisposition Prematurity Living in urban areas

- Functional constipation – 90%
- Low consumption of dietary fiber - Leading risk factors
- Psychological factors -
  - School-related stressful events
    - Separation from best friend
    - Bullying at school
    - Failure of exam
  - Family-related events
    - Severe illness of family member
    - Parents' job loss
    - Frequent punishment by parents

\* Psychological factors including emotional stress are likely to modulate colonic and rectal function, through the brain gut axis, leading to constipation.

- Children living in urban areas – due to high consumption of junk foods with low fiber content and sedentary life style

❖ **Pathophysiology of constipation –**



Contractions of full rectum inhibits the internal sphincter leading to fluid faeces to pass around hard faeces → overflow incontinence

❖ **How to investigate this pt?**

Laboratory investigations are rarely indicated in childhood constipation except in,

- ✓ Those with evidence of organic diseases from Hx & Ex
- ✓ Those who do not respond to adequate medical management.

**1) Plain abdominal radiograph -**

- Performed to identify the degree of fecal loading in the colon and rectum
- Useful in children who are not willing to undergo DRE due to pain & fear
- Very limited value in clinical assessment of constipation –
  - interpretation of the radiological findings is difficult & inconsistent
  - and
  - there is a poor correlation between clinical and radiological Δ

**2) Colonic transit studies -**

- The transit time of the colon is studied using radio-opaque markers
- Allow to differentiate constipation due to delayed segmental and pan-colonic transit from constipation with normal transit
- Beneficial in children with chronic treatment-resistant constipation to determine colonic transit abnormalities
- A tablet contains 24 pellets.
- No special dietary preparation is required
- No laxatives – lead to normal bowel habits
- Series of normal erect abdominal x-rays are taken
  - 24hrs – all pellets seen
  - 3 days – if 80% of pellets present (19 pellets) → constipation

**3) Anorectal manometry -**

- A collection of several tests that measure pressure changes in the rectum and the anal canal.
- It is often combined with surface electrode electromyography of the external anal sphincter and puborectalis muscle
- Provide details on rectal sensation, state of recto-anal inhibitory reflex, tone of anal sphincter and defecation dynamics.
- Most important benefit - to exclude Hirschsprung disease.
  - presence of recto-anal inhibitory reflex excludes Hirschsprung disease
- False positive results may occur due to immaturity of ganglion cells (in premature babies) and artefacts
- Therefore in cases with strong clinical suspicion of Hirschsprung disease, it is imperative to perform a suction biopsy to confirm or exclude the diagnosis.

**4) Colonic manometry -**

- Measures the intracolonic pressure using a multichannel manometry probe.
- Useful in patients with intractable constipation.
- Children with functional constipation show normal colonic motor activity
- Children with rare colonic muscle disorders demonstrate absent or weak colonic contractions.
- The gastro-colonic response is absent in colonic neuropathy
- An important investigation in children with chronic treatment-resistant constipation, who do not respond to maximum doses of combined laxative therapy

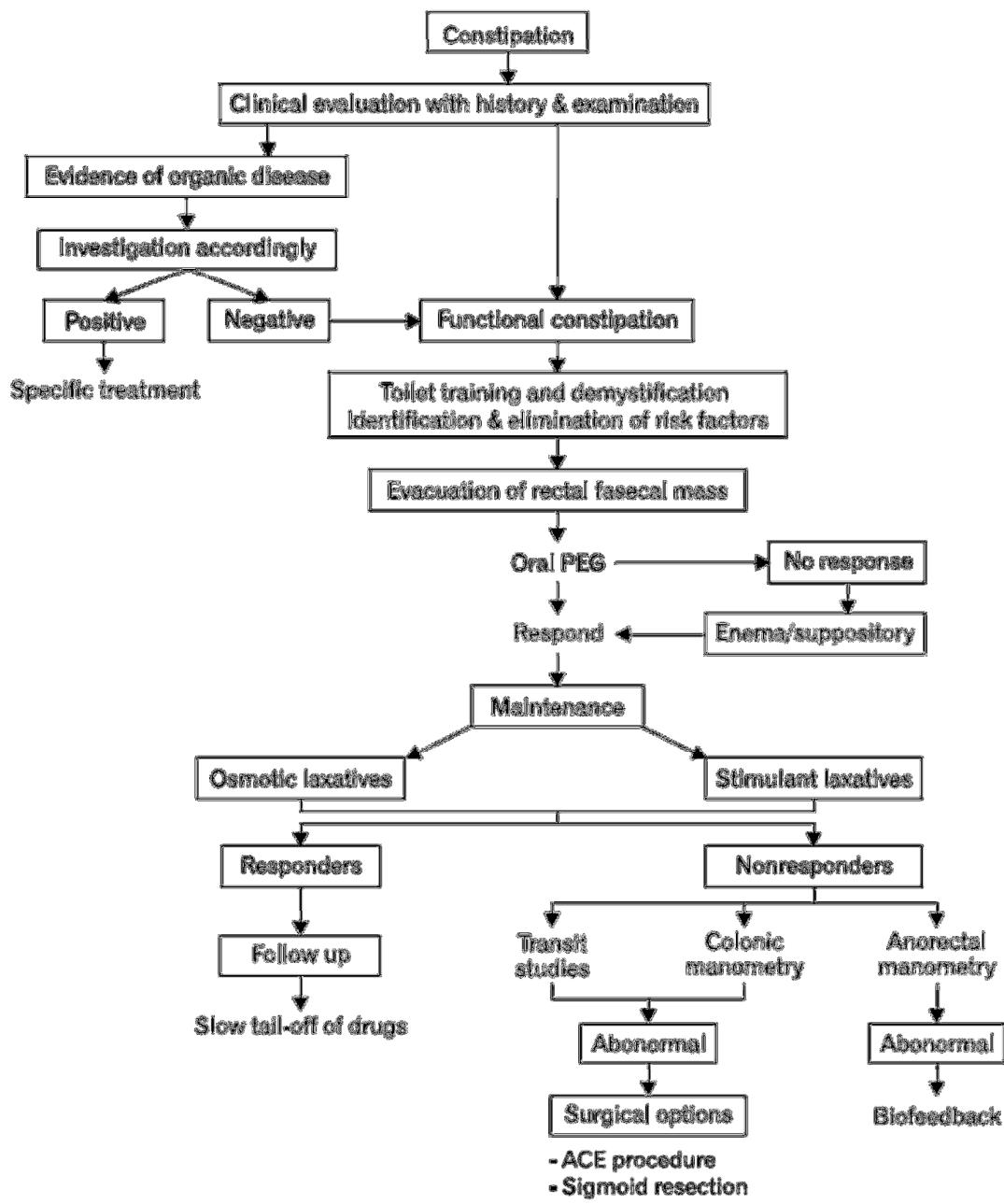
**5) Other investigations – (not valid for use in routine evaluation)**

- Fecoflowmetry - evaluates pressure changes in the rectum and anal canal during infusion of saline and also evacuation rates of saline from the rectum using uroflowmeter
- Pelvic ultrasonography – has been used to measure the diameter of the rectum in children with chronic constipation.
- Anal endosonography - reveal abnormalities in the sphincter complex in children with chronic constipation

**❖ How to manage this pt?**

The key steps in management

- 1) Education
- 2) Treatment of fecal impaction
- 3) Maintenance therapy
- 4) Close follow-up



#### Education –

- ✓ Educating the parents and patients about pathophysiology and precipitating factors
  - help to alleviate anxiety, minimize accusations and increase their involvement in management

- ✓ Behavioral Therapy
  - Regularize toilet routine
  - Discourage stool withholding
  - Psycho-education- Reduction of anxiety towards defecation using education and models
  - Child is taught of straining techniques
  - Finally the behavior is reinforced by motivation and reward system
- ✓ dietary modification - Adequate intake of dietary fiber (age + 5 in grams)

 **Disimpaction**

- Administration of enemas - relieve rectal fecal load.
- Oral polyethylene glycol – best
- If not respond – phosphate enema
- Use the rectal route, only when oral drugs have failed. Insertion of rectal enema may be extremely disturbing to the child who might already have anal fissures. Therefore, it needs to be given under sedation to minimize pain and psychological effects

 **Maintenance Therapy -**

- After achieving disimpaction - start daily oral laxatives to keep the stool soft – to prevent re-impaction.
- The duration of the maintenance may vary from months to years.
- Main pharmacological agents - osmotic and stimulant laxatives
  - ✓ Osmotic laxatives – lactulose, polyethylene glycol
  - ✓ Stimulant laxatives – bisacodyle (dulcolax)

 **Surgery**

- considered only when medical therapy fails in long standing constipation
- Children who do not respond to enemas may need manual evacuation.
- Sigmoid resection and removal of dilated megasigmoid – in severe constipation
- Antegrade colonic enema via appendicocaecostomy - in severe functional constipation

❖ **What are the types of drugs used in the Rx of constipation?**

- Stool bulking agents
- Osmotic laxatives
- Faecal softeners
- Stimulant laxatives

**Stool bulking agents**

- Rx of choice for simple constipation
- Increase the volume and lowering the viscosity of intestinal contents, ultimately forming soft bulky stools
- So it encourages normal reflex bowel activity
- Should be taken with liberal quantities of fluid(at least 2L daily)

### Osmotic laxatives

- Less absorbed
  - Reduce viscosity of intestinal contents, forming fluid stool
- a) Some inorganic salts
  - It retains water in the intestinal lumen
  - When given as hypertonic solutions, absorbs water from the body
  - Mild constipation : Mg(OH)<sub>2</sub>
  - Severe constipation – MgSO<sub>4</sub>
  - Both Mg salts act in 2-4 hrs
- b) Lactulose
  - Synthetic disaccharide
  - Taken orally
  - Remains unabsorbed in lumen & acts as osmotic laxative
  - Used in Rx of hepatic encephalopathy

\* Osmotic laxatives are frequently used to clear the colon for diagnostic procedures or surgery

To evacuate Enemas containing  
Distal colon PO<sub>4</sub><sup>-</sup> or citrate

Prepare for Oral preparations  
colonoscopy containing MgSO<sub>4</sub> +  
citric acid (*citramag*) or  
polyethylene  
glycol(*klean prep*)

### Faecal softners (Emollients)

- Useful in M<sub>x</sub> of anal fissures & haemorrhoids

Docusate sodium Lower the surface  
(dioctyl sodium tension in fluid in  
sulphosuccinate) bowel

↓

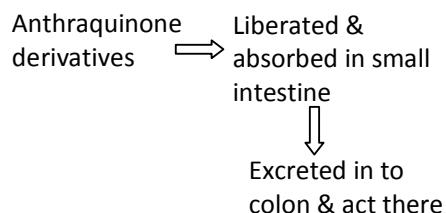
Allows more water to remain in faeces

- Acts in 1-2 days

### Stimulant laxatives

- Increase intestinal motility
- May cause abd. Cramps
- Caution in pregnancy
- C.I in suspected intestinal obstruction

- a) Bisacodyl
  - Stimulates sensory endings in the colon by direct action on the lumen
  - Effective orally in 6-10h
  - As a suppository acts in 1hr
- b) Sodium picosulfate
  - Similar action
  - Used to prepare bowel prior surgery
- c) Glycerol
  - Has a mild stimulant effect on the rectum when administered as a suppository
- d) The anthraquinone group
  - Includes senna, danthron, cascara, rhubarb and aloes



### Suppositories & enemas

- Bisacodyl, Glycerin
- To obtain an action within 1hr
- Produce defecation by
  - \* softening faeces
  - \* distending bowel
- used in surgical preparation, radiological examination & endoscopy
- Preparations with  $\text{Na}_3\text{PO}_4$  ----> poorly absorbed ----> retains water in gut ----> so it is generally used

# FEVER AND LYMPHADENOPATHY IN CHILDREN

## Regional

- Infective
  - pharyngitis, dental abscess, otitis media, actinomycetes
- Lymphoma
  - Hodgkin lymphomas, non-Hodgkin lymphoma

**DD**  
Local infection  
IMN  
Lymphoma  
Leukemia

## Generalized

- **Leukemia**
  - Acute lymphoblastic leukaemia
  - Acute myeloid leukaemia
  - Lymphomas (Hodgkin's, non-Hodgkin's)
  - Neuroblastoma.
  - Histiocytoses.
- **Infective**
  - IMN-(commonest)
  - Measles-
  - Rubella-
  - Cat scratch fever

## PC-fever with lymphadenopathy

### HPC-

#### Duration

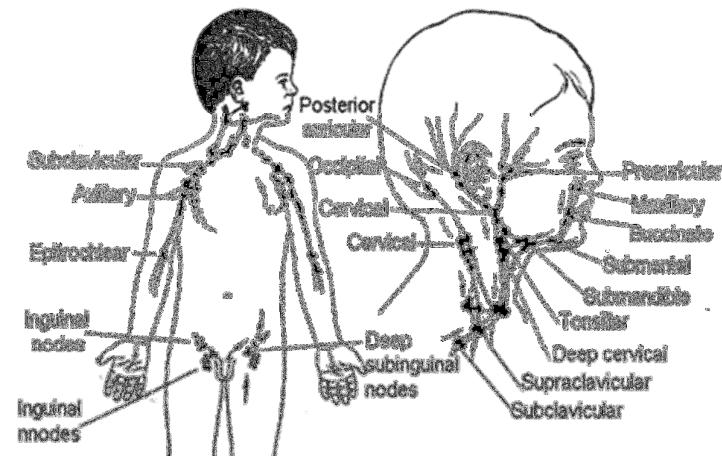
- ✓ Short (< 2 weeks) - likely to be infectious
- ✓ Long (> 2 weeks but < 1 year) - likely to be infectious, malignancy, autoimmune, drug reaction
- ✓ Very long (> 1 year) likely to be pathologic but not malignancy

#### Location

Localized - likely to be infectious

Regional - likely to be infectious

Generalized - more likely pathologic (e.g. malignancy, autoimmune, IMN etc.)



- 1) Head and Neck - likely infectious
- 2) Mediastinal - likely pathologic
- 3) Abdominal - likely pathologic
- 4) Inguinal - likely infectious

**Associated symptoms–**

**IMN**

- Fever-low grade
- Malaise
- Tonsillapharyngitis- cough
- Ocular muscle pain, photophobia
- Chest pain

**Rarely**

- Arthralgias
- Myalgias

**TB adenitis**

TB features

Lymphadenopathy mainly in the posterior triangle (51%) and deep upper cervical (48%).

In the majority of cases lymphadenitis is unilateral 3.

**Typhoid fever**

- Abdominal discomfort and diarrhea.
- Non-specific symptoms: Chills, sweating, headache, loss of appetite, sore throat, dry cough, constipation, muscle pains, weakness

**Measles**

- Fever, catarrh, conjunctivitis and harsh dry cough,
- Koplick spots- Typical rash start to appear on day3 starting behind the ear and spread along hair line.
- Rash (maculopapular rash) spread to face, trunk and then to limbs very rapidly.
- Rash start to fade by 3 days
- IP 7 – 14 days
- Prodromal stage occur 3 – 5 days before the eruption of rash

**Rubella**

- Cervical, post auricular, occipital nodes are usually involved

**Leukemia**

- Commonest ALL(adult note)

**Lymphoma**

- Children- NHL-B symptoms
- Adolescents-HL

**B symptoms**

- Fever
- Night sweats
- LOW, LOA
- Pruritus

**PMHx** – Dental caries, malignancy, cat scratches, local trauma, epilepsy, TB, syphilis/VDRL done, RA  
Previous treatments (such as antibiotics and how patient responded)

LN biopsies

Rx from Maharagama

Contact with infected people-kissing etc

**DHx** – phenytoin, carbamazepine

**FHx** -leukaemia

**SHx**- Pets - especially cats for Cat Scratch Disease

Travel - including Tuberculosis exposure

## **Examination**

GE -

Pallor – primary malignancy conjunctivitis, Jaundice - IMN  
Mouth – tonsillar enlargement – IMN, NHL  
Neoplastic tumours and ulcers  
Palatal petechiae& pharyngitis - IMN  
Dilated neck veins, venous engorgement of face – SVC obstruction (mediastinal LN)  
Linear scratches – lymphoma, cat scratch disease  
Bruising,rashes,SLE

LN-

Infection –	<0.5cm, discrete, mobile B/L, non-tender
Reactive hyperplasia	No associated Inflammation
LN infection	Larger,matted, warm,red,tender
TB	Non tender, matted, attached to skin, sinuses
Malignancy	>2cm, non-tender, hard, discrete/matted, fixed to skin/underlying tissue
Lymphoma	Asymmetrical, Non tender, firm, rubbery, matted
ALL, CLL	symmetrical

## Adult lymphadenopathy note

### **Abdomen**

Testicular enlargement – leukaemia(ALL)  
Hepatomegaly  
Splenomegaly – IMN, lymphoma, CLL, acute leukaemia

Respiratory

– Pleural effusion – HL

CNS

- Cranial n. palsies – TB,  
Focal neurological signs, meningism  
Peripheral neuropathy

## **Investigations**

### **leukemia,lymphoma-other note**

IMN

White blood cell (WBC) count - will be higher than normal

Monospot test - will be positive for infectious mononucleosis.

Antibody titer - tells the difference between acute and convalescent observations

## Management

Conservative

- Drink plenty of fluids.
  - Gargle with warm salt water to ease the sore throat.
  - Get plenty of rest.
  - Take paracetamol or ibuprofen for pain and fever.

Corticosteroids –if airway is compromised

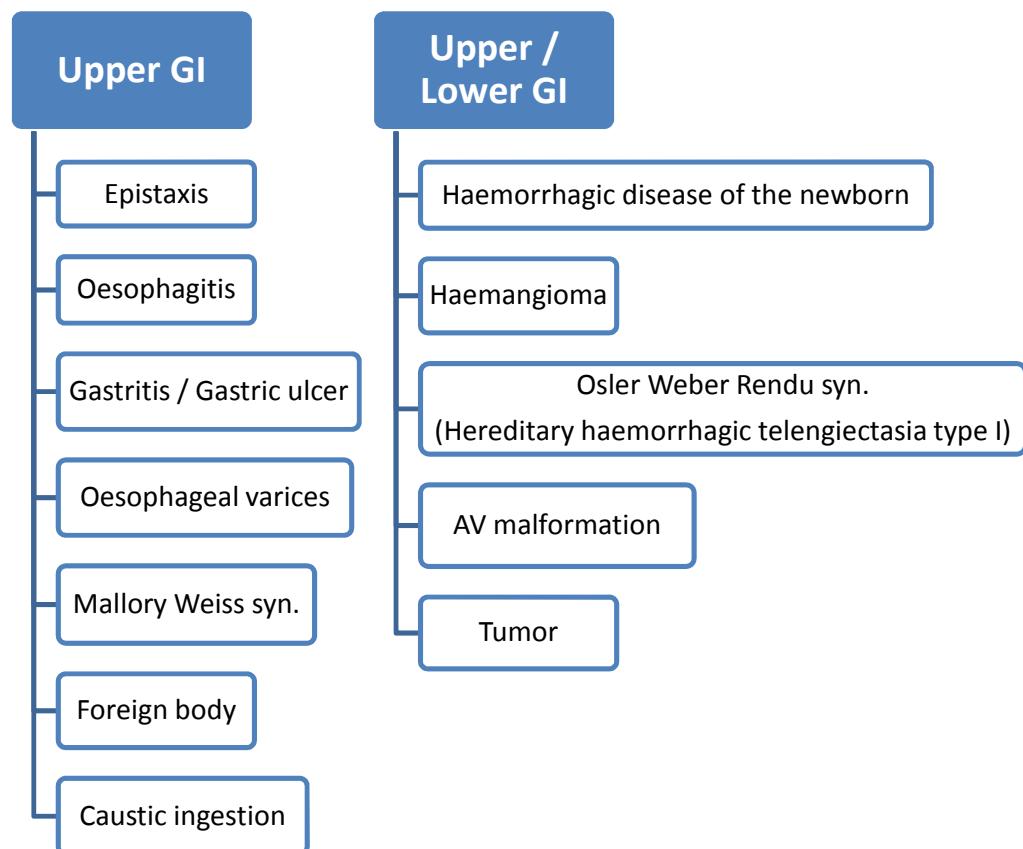
## **Avoid Amoxicillin→ Drug induced rash**

### **Complications of IMN**

1. Bacterial infection of the throat
2. Hemolytic anemia
3. Inflammation of the testicles (orchitis)
4. Nervous system problems (rare), such as:
  - Guillain-Barre syndrome
  - Meningitis
  - Seizures
  - Temporary facial paralysis (Bell's palsy)
  - Uncoordinated movements (ataxia)
5. Spleen rupture (rare; avoid pressure on the spleen)
6. Skin rash (Gianotti-Crosti syndrome - acrodermatitis of childhood)

## Haematemesis

Neonate	Child	Adolscent
<ul style="list-style-type: none"> <li>• Swallowed maternal blood</li> </ul>	<ul style="list-style-type: none"> <li>• Gastric ulcer / Gastritis</li> <li>• Swallowed epistaxis</li> <li>• Oesophageal varices</li> <li>• Oesophagitis</li> <li>• Coagulopathy</li> <li>• Mallory Weiss syn.</li> <li>• Haemangioma</li> <li>• Haemolytic uraemic syn.</li> </ul>	<ul style="list-style-type: none"> <li>• Gastric ulcers / Gastritis</li> <li>• Oesophageal varices</li> <li>• Oesophagitis</li> <li>• coagulopathy</li> <li>• Mallory weiss syn.</li> <li>• Telangiectasis (Angiodysplasia)</li> </ul>



**PC:** vomiting of blood for \_\_\_ duration

**HPC:**

**1) Elaborate symptoms**

Onset

Duration

Fresh / altered blood

Amount

How many times/day & the progression of symptom

Whether effortless vomiting or with effort

Associated melaena

Whether related to meals

Any past history of similar episodes

Any Ix & Rx done up to now

**2) Exclusion of DD**

**A. Gastric ulcer / duodenal ulcer**

- Epigastric burning/ aching/ dull abdominal pain. Sometimes peri umbilical pain
- Pain last for minutes to hours
- Periodic Hx ( 3-4 episodes/ month & the presence of symptom free periods)
- Precipitate with food – **Gastric ulcer**
- Relieve with food – **Duodenal ulcer**
- Relieve with antacids
- Nocturnal pain awakening the child from sleep – **Duodenal ulcer**
- Associated N, V, melaena, feeding difficulty, crying episodes
- Aetiology
  - Drugs – NSAIDs, Aspirin
  - Stress – sepsis, shock, intracranial lesions (**Cushing's ulcer**), severe burns (**Curling's ulcer**)
  - Genetic
  - Smoking
  - Alcohol abuse
  - H. pylori – poor hygiene, low socio economic status

**B. Oesophageal varices**

- Features of chronic liver disease
  - Yellowish discolouration of eyes, malaise, LOA, R. hypochondrial pain
  - Associated melaena
  - Growth retardation
  - Bleeding tendency – gum bleeding, bleeding into skin
  - Hx of blood transfusions or needle prick injuries or maternal hx of Hep. B at the time of delivery
- Features of fulminant hepatic failure
  - Drowsiness, altered sleep pattern, confusion, irritability
- Ix up to now & the results

**C. Mallory Weiss syn.**

- Hx of repeated forceful vomiting – with the initial vomitus w/o any blood

**D. GORD / Oesophagitis**

In infants –

- Regurgitation
- Irritability
- Failure to thrive
- Recurrent chest infections due to aspiration pneumonia
- Wheezing
- Features of anaemia

In older children & adolescents –

- Regurgitation
- Retrosternal burning chest pain radiating upwards
- Symptoms precipitate with heavy fatty meals & in lying down position.
- Soreness in the mouth
- Burping / belching
- Associated wheezing
- If severe –dysphagia, odynophagia

**E. Dengue haemorrhagic fever**

- High grade fever
- Bleeding into skin and gum bleeding
- Malaise and weakness
- Haemetemesis & melaena

**F. Foreign body ingestion**

- Cough
- Choking attack
- Stridor – noisy breathing on inspiration
- Odynophagia
- Retrosternal pain
- Excessive salivation

**G. Corrosive ingestion**

- Alkali – more harmful

**H. Bleeding disorder**

- Bleeding from gums & bleeding in to skin
- FHx of a bleeding disorder

**I. AV malformation**

- Presence of AV malformations in other sites
  - Skin – port wine stain
  - Brain – seizures

**Antenatal Hx**

Mode of delivery, birth weight

Any antenatal complications

Post natal complications – Jaundice, PBU admission, Ix, Rx,

Hx of neonatal omphalitis, sepsis, dehydration, or umbilical vein catheterization – Cause for prehepatic portal hypertension

Umbilical stump bleeding

### **Developmental Hx**

### **Immunization Hx**

### **Nutritional Hx**

#### **PMHx**

Local trauma, peritonitis (pyelophlebitis), hypercoagulable states, and pancreatitis – Pre hepatic portal hypertension

#### **PSHx**

#### **DHx**

#### **AHx**

#### **FHx**

#### **SHx**

## **Examination**

Height  
Weight      }  
OFC

Pallor

Icterus

Nasal bleeding, gum bleeding

Petechiae, purpura, ecchymosis

Port wine stain

Increased pigmentation

Scratch marks

Clubbing

Flapping tremors

Leukonychia

Palmar erythema

### **Abdomen**

Caput medusae

Shrunken impalpable liver – cirrhosis

Hepatomegaly

Splenomegaly

Ascites

## Investigations

FBC

- Hb

Blood grouping & cross matching

PT/ INR

Upper GI endoscopy

- To find the site of bleeding & to arrest bleeding

Liver function test

### Management of an acute upper GI bleeding

- Correct hypovolaemia
- Correct anaemia
- Stop bleeding
- Prevent recurrence
- Diagnose the aetiology
- Apply specific therapy

#### Mx of Oesophageal varices

- Stop bleeding
  - Endoscopic variceal band ligation or sclerosis
  - IV Octreotide 30 µg/m<sup>2</sup>/ hr
  - Surgical variceal ligation
  - selective venous embolization
  - TIPS (transjugular intrahepatic portosystemic shunt)
  - OLT (orthotopic liver transplantation)
- Prevent recurrence
  - Propranolol - oral

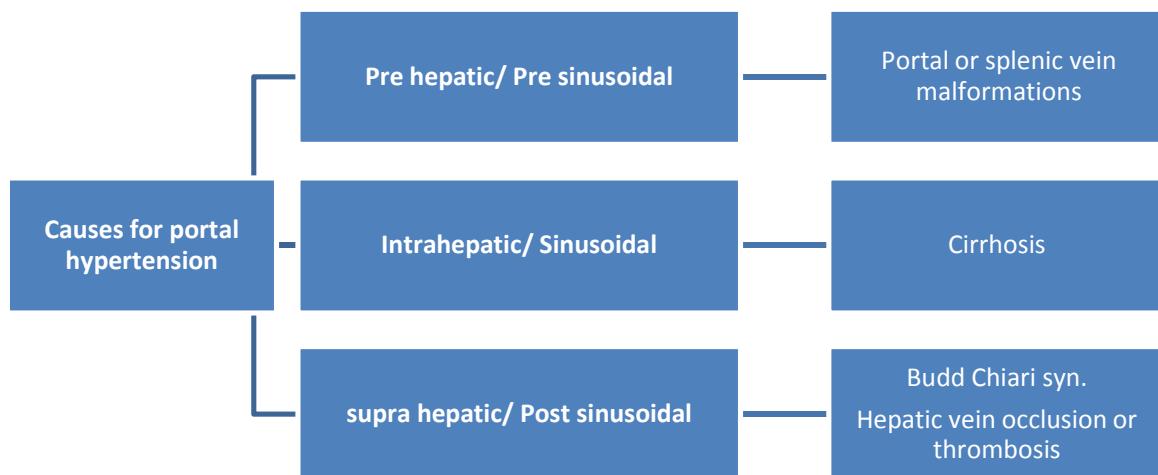
#### Mx of gastritis/ peptic ulcer disease/ oesophagitis

Omeprazole

H<sub>2</sub> blockers

Antacids

## Discussion



### Causes of Portal Hypertension

EXTRAHEPATIC PORTAL HYPERTENSION	
Portal vein agenesis, atresia, stenosis	
Portal vein thrombosis or cavernous transformation	
Splenic vein thrombosis	
Increased portal flow	
Arteriovenous fistula	
INTRAHEPATIC PORTAL HYPERTENSION	
1) Hepatocellular Disease	
Acute and chronic viral hepatitis	
Cirrhosis	
Congenital hepatic fibrosis	
Wilson disease	
$\alpha_1$ -Antitrypsin deficiency	
Glycogen storage disease type IV	
Hepatotoxicity	
Methotrexate	
Parenteral nutrition	
2) Biliary Tract Disease	
Extrahepatic biliary atresia	

Cystic fibrosis  
Choledochal cyst  
Sclerosing cholangitis  
Intrahepatic bile duct paucity

**3) Idiopathic Portal Hypertension**

**4) Postsinusoidal Obstruction**

Budd-Chiari syndrome  
Veno-occlusive disease

## Heart failure in children

### P/C

- Difficulty in breathing
- Poor feeding
- FTT
- Recurrent chest infection

### DIB

- LUNG
- HEART

#### Lung

- Respiratory distress syndrome
- Pneumonia
- Meconium
- Pneumothorax
- Transient tachypnoea of new born
- Congenital diaphragmatic hernia

	Symptoms & signs	Causes
RDS	Symptoms prior to 4 hrs of age Persist for 4 hrs Continued 4 hrs Tachypnoea Dyspnoea Grunting Cyanosis ↓ air entry Tachycardia Inactivity hypotonia	Intra partum asphyxia Pulm.infection Pulm.haemorrhage Meconium aspiration Cong.diaphragmatic hernia
pneumonia	Respiratory distress Pyrexia Low apgar score Hypotonia Jaundice hypoxia	Prolong rupture of membranes Maternal fever + high vaginal swab 7 days prior to the Delivery Purulent vaginal discharge Offensive liquor
Meconium aspiration Xn	Persistent pulmonary HT of the new born	Intrauterine distress Passage meconium in to the amniotic fluid Placental insufficiency Maternal HT Pre eclampsia Oligohydroamnios Maternal drug use
pneumothorax	Signs of respiratory Distress Affected side hyper resonance, ↓ AE	Over vigorous resuscitation at birth Meconium aspiration RDS Artificial ventilation

Transient tachypnoea of new born	Resp.distress soon after birth Lasts 24-48 hrs	LSCS is the main risk factor
Cong.diaphragmatic hernia	Resp.distress soon after birth Left sided hernia commoner Apex shifted, ↓AE Bowel sounds in the chest Scaphoid abdomen	

## Heart failure in children

### Symptoms

- Breathlessness ( particularly on feeding/ exertion)
- Sweating
- Poor feeding
- Recurrent chest infection

### Signs

- Poor weight gain/ FTT
- Tachypnoea
- Tachycardia
- Heart murmur
- Gallop rhythm
- Enlarged heart
- Hepatomegaly
- Cold peripheries

### Heart failure in neonates

Results from left heart obstruction  
 ↓  
 Obstructed systemic circulation /duct dependent circulation  
 ↓  
 If the obstruction is very severe  
 ↓  
 Arterial perfusion mainly by  
 ↓  
 Right to left flow of blood via the arterial duct

1. Hypoplastic left heart syndrome
2. Critical aortic valve stenosis
3. Severe co-arctation of the aorta
4. Interruption of the aortic arch

### Heart failure in infants

Due to left to right shunts  
 ↓  
 During the 1<sup>st</sup> few weeks of life  
 ↓  
 Pulm. Vasculature resistance falls  
 ↓  
 There is progressive increased pulm.blood flow  
 ↓  
 Symptoms of heart failure ↑up to the age of 33 months  
 ↓  
 May subsequently improve as the pulm.vasculature resistance ↑ response to the left to right shunt

1. VSD
2. Atrioventricular septal defect
3. Large persistent ductus arteriosus

Cause	Clinical Fx	Ix	Mx
Aortic stenosis	<p>Most are asymptomatic Severe stenosis ↓ exercise tolerance, chest pain on exertion, syncope In neonates – severe heart failure/a duct dependent systemic circulation leading to shock Physical signs Small volume, slow rising pulse Carotid thrill(always) Ejection systolic murmur maximal at the upper right sternal edge radiating to the neck Delayed &amp; soft A2 Apical ejection click</p>	<p>CXR NL/ prominent LV with post stenotic dilatation of the ascending aorta ECG LVH – deep S wave in V2 &amp; tall R wave in V6 (&gt;45mm total) Down going T wave suggests LV strain &amp;severe aortic stenosis</p>	<p>Regular clinical &amp; Echo assessment Children with symptoms on exercise → balloon valvotomy In older children → balloon dilatation In neonates Sx is more difficult &amp; dangerous</p>
Coarctation of the aorta	<p>Asymptomatic Always systemic HT in the right arm Ejection systolic murmur at upper sternal edge Radio – femoral delay</p>	<p>CXR Rib notching → development of large collateral intercostals arteries running under the ribs posteriorly to bypass the obstruction '3' sign → with visible notch in the descending aorta ECG – LVH</p>	<p>Stent insertion / surgical repair</p>
Interruption of the aortic arch	<p>Severe form of co-arctation No connection B/W the aorta proximal &amp; distal to the arterial duct A VSD is usually present Almost in the neonatal period</p>		<p>Complete correction with closure of the VSD &amp; repair of the aortic arch (performed within 1<sup>st</sup> few days of life) Risk of death is higher than simple coarctation Ass. with other conditions DiGeorge Xn – absence of thymus Palatal defects Immunodeficiency Hypocalcaemia</p>
Hypoplastic left heart Xn	<p>Under development of the left heart Mitral valve is small Ascending aorta is small</p>		
Ventricular septal	30% of all cong, heart defects	CXR – NL	

defect	Defects anywhere in the ventricular septum Usually perimembranous (adjacent to tricuspid valve) Or muscular Signs Thrill over lower sterna Loud pansystolic murmur at lower sterna edge Quiet P2 Defects are same size / bigger than the aortic valve  Symptoms HF with breathlessness + FTT Recurrent chest infection Signs Soft pansystolic murmur / no murmur Apical mid diastolic murmur ↑flow across the mitral valve after the blood has circulated through the lungs Loud P2 (pulm HT) Tachypnoea, tachycardia Enlarged liver	ECG – NL Echo – precise anatomical defect	
Large VSD		CXR Cardiomegaly Enlarged pulm.arteries ↑pulm.vascular markings Pulm.edema ECG Pulm.Ht	Drug therapy for heart failure Diuretics + captopril
Atrioventricular septal defect			
Large persistant ductus arteriosus – connects pulm.artery to descending aorta (in term infants it normally closes shortly after birth)  Fail to close by a month post term, due to defect in the constrictor mechanism Blood flow from aorta to pulm.artery	Continous murmur beneath the left clavicle (murmur continuous Pressure in the pulm.artery<aorta through out the cardiac cycle) Collapsing / bounding pulse (pulse pr.↑)	CXR,ECG – NL But if PDA is large CXR & ECG similar to large VSD Crossectional ECHO assisted by Doppler	Infants with asymptomatic PDA→closure to ↓risk of IE Closure done with a coil/ occlusion device introduce via a cardiac catheter At the age of 1yr

## Failure to thrive

- A. Inadequate intake
  - a) Non organic/environmental
  - 1. Inadequate availability of food
    - Feeding problems
    - Insufficient breast milk, poor technique, incorrect preparation of formula
    - Insufficient/unsuitable food offered
    - Lack of regular feeding times
    - Infant difficult to feed – resists feeding or disinterest
    - Conflict over feeding
    - Problems with budgeting, shopping, cooking food
    - Low socioeconomic status
  - 2. Psychosocial deprivation
    - Poor maternal infant interaction
    - maternal depression
    - poor maternal education
  - 3. neglect or child abuse
- b) organic
  - 1. impaired suck/swallow
    - oro motor dysfunction, neurological disorders; CP
    - cleft palate
  - 2. chronic illness leading to anorexia
    - chrons disease, CRF, cystic fibrosis, liver disease
- C. inadequate retention
  - vomiting
  - severe GORD
- D. malabsorption
  - celiac disease
  - cystic fibrosis
  - cow's milk protein intolerance
  - short gut Xn
  - post necrotizing enterocolitis
- E. failure to utilize nutrients
  - syndromes
  - chromosomal disorders ; down's Xn
  - IUGR or extreme prematurity
  - Congenital infections
  - Metabolic disorders ; cong.hypothyroidism, storage disorders, amino & organic acid disorders
- F. Increased requirement
  - Thyrotoxicosis
  - Cystic fibrosis
  - Malignancy
  - Chronic infection; HIV, immunodeficiency
  - Congenital heart disease
  - CRF

Hx

- ❖ Detail dietary Hx
- ❖ Feeding, including details of exactly what happens at meal time
- ❖ Other symptoms; diarrhea, vomiting, cough, lethargy,
- ❖ Premature, IUGR at birth
- ❖ Growth of other family members, any illness in the family
- ❖ Childs development is NL
- ❖ Any psychosocial problems at home

## Recurrent infection

- FTT
- Heart diseases
- Imuunodeficiency

## Birth hx

- Antenatal Dx of cardiac abnormalities
- Anomaly scan
- Suspected Downs Xn
- Parents have had a previous child with heart disease
- Mother has congenital heart disease
- Whether the parents have been counselled
  
- Any maternal disorders
  - Rubella infection
  - SLE
  - DM
- Maternal drugs
  - Warfarin
  - Maternal alcohol intake
- Any chromosomal abnormalities detected antenatally
- At birth
  - Cried
  - Cyanosed
  - Sucked well
  - SCBU
  - Ventilated
  - Stayed at hospital for how long
  - Whether the parents have been informed any abnormality which detected at routine Ex
  - Clinic followup

Immunization

Any delay

Development

Ask questions specifically to check all 4 domains

Diet

Detail dietary Hx

Feeding, including details of exactly what happens at meal time

PMHx

PSHx

DHx

FHx

- Cong, heart disease
- Any syndromes
- Consanguinity

Social Hx

- Family support
- Income
- Parental education
- Closest hospital
- Transport
- Other children care

O/E

Growth – height, weight

Pale

cyanosis

Hydration

Dyspnoeic/ not

Fx of nutrient def

Fx of chronic malnutrition

Tachycardia

Tachypnoea

Labored breathing

ICR, SCR, flaring of alae nasi

Hepatomegaly

Cardiomegaly

Gallop rhythm

↑JVP

Hepatomegaly

Basilar rales

Oedema

Cardiomegaly

Gallop rhythm

Pan systolic murmur

## Heart failure in childhood

### ***Definition***

Inability of heart to deliver adequate cardiac output to meet metabolic needs of body

### ***Aetiology***

1. Based on mechanism
  2. Age of presentation
    1. Based on mechanism
      - a) Volume overload
        - Regurgitant valve
        - High output status
      - b) Pressure overload
        - Systemic HT
        - Outflow obstruction
      - c) Loss of muscles
        - Post MI
        - Chronic ischaemia
        - CT disorders
        - Infections
        - Poisons – Co, Doxorubicin
      - d) Restricted filling
        - Pericardial diseases
        - Tachyarrhythmias
        - Restrictive cardiomyopathy
    2. Age of presentation

 Fetal

- Severe anaemia
- SVT / VT
- Cong.heart block
- Severe Ebstein anomaly
- Myocarditis

 Premature neonate

- Fluid overload
- PDA
- VSD
- Cor pulmonale (chronic lung disease)
- Hypertension
- Myocarditis
- Genetic cardio myopathy

Full term neonate

- Asphyxia cardiomyopathy
- A-V malformation
- Coarctation of the aorta
- Hypoplastic left heart Xn
- Single ventricle
- Truncus arteriosus
- Myocarditis
- genetic cardiomyopathy

infant toddler

- left to right cardiac shunts
- anomalous left coronary artery
- genetic / metabolic cardiomyopathy
- acute hypertension (AGN)
- SVT
- Kawasaki
- Myocarditis

Child – Adolescent

- Rheumatic fever
- Acute hypertension
- Myocarditis
- Thyrotoxicosis
- Hemochromatosis/ hemosiderosis
- Cancer therapy
- Sickle cell anaemia
- Endocarditis
- Cor pulmonale
- Genetic/ metabolic cardiomyopathy

***Pathophysiology***

**Systemic oxygen transport = CO x systemic O<sub>2</sub> content**

**Cardiac output = SV x HR**

Stroke volume depends on

- After load (pressure work)
- Pre load (volume work)
- Contractility (intrinsic myocardial function)

O<sub>2</sub> content depends on

- O<sub>2</sub> carrying capacity of blood ( $\downarrow$  in anaemia, hypoxia)
- $\uparrow$ O<sub>2</sub> demand
- Hyperventilation
- Hyperthyroidism
- Hypermetabolism

***Compensatory mechanisms***

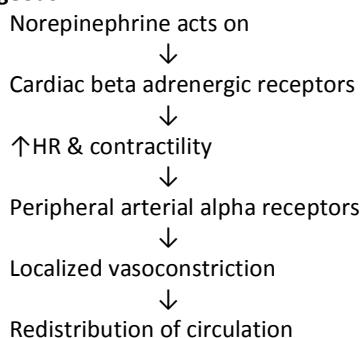
Molecular / cellular level

Regulation of metabolic pathways components leading to changes in efficiency of O<sub>2</sub> transport / utilization

**Neuro-hormonal**

- ✓ Renin angiotensin system
- ✓ Sympatho – adrenal axis

**Initially advantageous**



**Disadvantages of chronic sympathetic activation**

- Hypermetabolism
- ↑ afterload
- Arrhythmogenesis
- ↑ myocardial O<sub>2</sub> requirements
- ↓ renal/ hepatic/ GI functions
- Cardiac receptor down regulation
- Direct myocardial cell damage

**Symptoms & signs develops due to**

- ❖ Pulmonary venous congestion
- ❖ Systemic venous congestion
- ❖ Sympathetic overload

	symptoms	signs
Infant	Poor sucking DIB while sucking Profuse perspiring Poor wt gain Irritability Weak cry	Tachycardia Tachypnoea Labored breathing ICR, SCR, flaring of alae nasi Hepatomegaly Cardiomegaly Gallop rhythm
Child	Fatigue Effort intolerance Anorexia Abd pain Dyspnoea/ orthopnoea cough	↑JVP Hepatomegaly Basilar rales Oedema Cardiomegaly Gallop rhythm Pan systolic murmur

**Investigations**

**CXR – A B C D E**

- Alveolar oedema (bat;s wings)
- Kerly B lines (interstitial oedema)
- Cardiomegaly
- Dilated prominent upper lobe vessels
- pleural Effusion

**ECG** ; useful to determine the underlying defect

- Chamber hypertrophy
- Ischemic changes
- Myocardial inflammatory changes
- Rhythm abnormalities

**ECHO**

- Fractional shortening =  $\frac{EDD - ESD}{EDD} (28-40\%)$
- EF = 55-65%
- Cardiac output

**Biochemical**

- ↓arterial O<sub>2</sub> %
- Metabolic acidosis
- Respiratory acidosis
- Hyponatraemia

**Management**

1. Treat the underlying cause
2. General measures
3. Dietary measures
4. Pharmacological therapies
5. Electrophysiological approach

**General**

- Adequate rest positive pressure ventilation
- Significantly ↓ total body O<sub>2</sub> consumption
- Reverse acidosis
- Correct anaemia

**Dietary measures**

- FTT
- Need to ↑ daily calories
- ↑ number of calories per ounce of formula (max.24cal/ ounce)

**Salt**

- Human breast milk is the ideal low Na nutritional source
- No added salt

**Pharmacological measures**

- Diuretics
- Adrenergic agonists
- Phosphodiesterase inhibitors
- Afterload reducing agents
- Beta blockers
- Digoxin

**Diuretics**

**Furosemide**

- 1-2mg/kg bolus
- 1-4 mg/kg/day in qds
- Inhibits Na<sup>+</sup>, Cl<sup>-</sup>, reabsorption in DCT & LOH
- Common A/E – hypokalemia, hyponatraemia

**Spironolactone**

- 2-3mg/kg/24h in qds
- Aldosterone inhibitor
- K sparing

**After load reducing agents**

Captopril

Nitroprusside

- Actions
- ↓HR & ↑myocardial perfomence
- Specially useful in cardiomyopathy, severe MR, AR, left to right shunt
- CI in left ventricular outflow tract obstruction ; AS

ACE inhibitor

- Vasodilation- ↓after load
- Venodilatation - ↓ pre load
- Aldosterone inhibition - ↓ salt & water retention
- Additional benefits – cardiac remodeling

A/E –

- Hypotension
- Hyperkalemia
- Maculopapular pluiritic rash
- Neutropenia
- Renal toxicity
- Chronic cough

Alpha & Beta agonists

- Dopamine
- Dobutamine
- Epinephrine

Use in ICU settings with continuous monitoring of BP, HR

Long term administration - ↑morbidity & mortality

Dopamine

- Beta agonist with some alpha adrenergic effects at higher doses
- Less chronotropic than dobutamine
- Selective renal vasodilatation
- Dose ; 2-30µg/kg/min

Dobutamine

- Derivative of dopamine
- Dose; 2-20µg/kg/min

Epinephrine

- Mixed alpha & beta agonists
- ↑peripheral resistance + cardiac effects

Phosphodiesterase inhibitors

**Milrinone**

- cAMP degradation inhibitor → increase cAMP
- positive inotrophi effects on heart
- peripheral vasodilatation
- dose; 50 µg/kg loading dose

- 0.25-1.0µg/kg/min infusion
- A/E hypotension secondary to peripheral vasodilatation

**Beta blockers**

CI in acute phase

Beneficial effects

- ↑exercise tolerance
- ↓hospitalisations
- ↓overall mortality

Metoprolol – beta 1 selective antagonist

Dose; 0.2-2mg/kg/day

Carvedilol – alpha & beta blocker ; scavenger effect

Dose ; 0.1-1mg/kg /day

## Neonatal sepsis meningitis

PC – Non specific – Fever

- Poor feeding
- Irritability
- Lethargy, drowsiness

DD – neonatal sepsis  
Neonatal meningitis  
Dehydration

Maturity – POA at delivery

Duration

1<sup>st</sup> 48hrs – early onset sepsis

After 48hrs – late onset

HPC

- AN Hx - \* Parity, EDD, age of the mother  
\* AN care – where  
\* T1 – Hx of fever with rash  
\* Follow up during the late prenatal period  
\* Hx of maternal fever 1/52 prior to delivery  
\* Maternal UTI, purulent vaginal discharge  
\* Hx of STD  
\* PIH, IUGR  
\* Prolonged rupture of membranes (dribbling) - >18hrs prior to delivery → ↑ risk  
    High vaginal swab taken/not  
    GBS prophylaxis given – O. erythromycin  
\* Premature rupture of membranes – Rupture before the onset of delivery  
    Maternal chorioamnionitis – Maternal fever  
    Uterine tenderness

Labour – spontaneous/ induced

No. of VE

Duration of labour

Delivery – vaginal/ forceps

Baby cried at birth, cyanosed → needed resuscitation, Apgar score, birth weight

Meconium aspiration

Ask for the CHDR

- ◆ PBU admissions
- ◆ Breast feeding established/not – hypoglycemia, dehydration
- ◆ BCG given
- ◆ Current prb. – onset, duration and progression of symptoms
  - Fever – temperature checked, fluctuations
  - Evidence of poor feeding – absence of good feeding – proper suckling
  - Sleep after feeds (2-3hrs)
  - UOP – 10xday
  - Bowel opening – 4-6x/day
- ◆ Associated symptoms
  - Lethargy, drowsiness
  - High pitched cry, irritability, seizures – meningoencephalitis
  - Vomiting, diarrhoea, abdominal distension
  - Apnoea, respiratory distress
  - Jaundice (colour of urine and stools), abdominal distension
  - Umbilical sepsis
  - Bleeding – coffee ground vomitus

- ◆ Action taken – SCBU admission – admission before symptoms/after – site of infection  
Incubator care, O<sub>2</sub>, ventilator used, Hx of SCBU infection
  - Any other contact hx of fever
  - Blood culture, LP done –results
  - IV AB given – how many days
  - How is the feeding
  - Cannulated, +/- CVP line – if so for how long
- ◆ Complications
  - 1) Respiratory distress
  - 2) Jaundice – phototherapy, exchange Tx
- ◆ Past Obs Hx – similar presentation, organism identified
- ◆ PMHx – mother – DM, immunocompromised, recurrent UTI
- ◆ Social Hx – maternal hygiene, visitors and over-handling

### Examination

General – weight loss >10%

OFC – hydrocephalus  
Activity – lethargic, irritable, high pitched cry, moving all 4 limbs/not  
Maturity – term/preterm/post term  
Colour – pale, cyanosed, jaundice  
Febrile/ cold  
Hydration, posture - opisthotonus

C – coloboma  
H – Heart defect  
A – atresia of the choanae  
R – Retardation of growth  
G – genital abn.  
E – ear prb.

#### Head to toe

- head – bulging fontanelles, sunken if dehydrated
- eyes – sticky, hyperaemia, discharge, cataract – caused by neonatal infections  
coloboma – ass. Syndromes (CHARGE) – neonatal sepsis, immunosuppression
- face – dysmorphism
- ear discharge
- throat – inflammation, cleft lip, cleft palate
- UL – nails – yellow green stain (meconium aspiration)
- Skin – sclerema neonatarum – thick hard skin (underlying sepsis)  
(Necrosis of subcutaneous fat due to intracellular precipitation of FA)  
Pustules, bleeding manifestations
- Breast abscess
- Umbilical sepsis – erythema >1cm, discharge, foul smell
- Back – meningocele
- Joint inflammation, IM abscess
- In-situ cannula, UV catheter/remnant – suture in the umbilicus

CVS – tachy;brady ( tachy - >160bpm brady- full term <80 preterm <120)

↓ BP

Murmurs – congenital heart disease

RS – tachypnoea >60/min

Labored breathing, apnoea, grunting, recessions  
Air entry, breath sounds, +/- bronchial breathing, crepts

Abd – Distended

Hepatosplenomegaly, ballotable kidney – hydronephrosis  
Persistently palpable bladder  
Poor urine stream in males

**Table 1. Glasgow Coma Scale Modified For Pediatric Patients<sup>50</sup>**

Eye Opening Response	< 1 year
4	Spontaneous
3	To shout
2	To pain
1	None
Verbal Response	0 to 2 years
5	Babbles, coos appropriately
4	Cries but is inconsolable
3	Persistent crying or screaming in pain
2	Grunts or moans to pain
1	None
Motor Response	< 1 year
6	Spontaneous
5	Localizes pain
4	Withdraws to pain
3	Abnormal flexion to pain (decerebrate)
2	Abnormal extension to pain (decorticate)
1	None

Nervous sys – Level of consciousness  
Posture – opisthotonus  
Tone – hypotonia  
↓ Reflexes

## Summary

DD – Early onset/ late onset neonatal sepsis  
Neonatal meningitis/ meningoencephalitis  
Dehydration – fever, lethargy, jaundice

How will you manage?

### 1) Resuscitation – if needed

If child presented with drowsiness, ↓ consciousness and evidence of respiratory distress

- Assess A,B,C – correct if any compromise
- Airway – Cyanosis, ↑RR/↓RR, snoring – obstruction above larynx, stridor  
Drooling of saliva  
Correct – Connect to pulse oxymeter – low sat.  
Head in neutral position and jaw thrust
- Breathing – Look, listen and feel for breaths  
Start O<sub>2</sub> via nasal prongs if no seizures/ head box if in PBU/ well fitting facemask  
Ambu bag ventilation
- Circulation - ↑HR, low pulse volume, ↓BP, CRFT<2s  
Gain IV access – wide bore cannula – 22G (yellow)  
Give fluid bolus – 10ml/kg of N/S over 10min  
Repeat after 1/2hr if necessary  
Inotropes if needed

### 2) Take blood and urine for investigation

- ❖ Full septic screen – CRP - >6mg/dl, takes 12-24hrs to become positive  
Blood culture and ABST – infant -3cc, neonate 1-2cc  
Urine culture and ABST  
CXR  
LP – delay if evidence of ↑ ICP (altered consciousness)
- ❖ Partial septic screen
- ❖ RBS – hypoglycemia - correct by establishing BF if able to suckle/ EBF  
Failed → start 10% dextrose bolus 2-5ml/kg followed by infusion
- ❖ SE
- ❖ FBC - ↓Hb, ↑PCV – dengue  
WBCC – Neutropenia/ ↑  
Normal – 18-22,000/mm<sup>3</sup> predominant cell – neutrophil (upto 3weeks)  
Plt - ↓ (<100,000/ml)
- ❖ Swabs from any other specific foci – ear, eye, throat, umbilicus  
Gastric aspirate – PROM – cells, gram stain, culture

CSF – normal values

	Cell count	Predominant cells	Protein (g/l)	Glucose (% of blood sugar)
<b>Neonates</b>	<30 /ml	neutrophils	0.4 – 1.2	2/3 <sup>rd</sup> (66%)
<b>Children</b>	<10/ml	lymphocytes	0.2 – 0.8	60%
<b>Adults</b>	<10/ml	lymphocytes	0.2 – 0.5	60%

CSF – pathological values

	<b>Bacterial meningitis</b>	<b>Viral meningitis</b>	<b>TB / cryptococcal meningitis</b>
Cell count / mm <sup>3</sup>	100-3000	10-500	100-500
Differential count	Mainly neutrophils	Mainly lymphocytes	Mainly lymphocytes
Protein g/l	0.5-3.0	0.5-1.0	1.0-6.0
Glucose % Blood sugar expected range	<60% (0.0-2.2mmol/l)	Normal	<60% (0.0-2.2mmol/l)

- ✓ If neutrophil ↑ → bacterial
- ✓ Lymphocyte ↑ → viral/TB

### 3) Supportive care

- ❖ Ensure minimal handling of the baby and hand washing prior to handling, isolate from other babies, barrier nursing – 1 nurse for the baby
- ❖ Maintain thermo-neutral environment – incubator care  
Cot care (under 100W bulb 18 inches above the cot)  
If baby not very ill kept warm with the mother
- ❖ Establish feeding – expressed BM – 3 hourly based on total fluid requirement  
200ml/kg breast feeds + regular top up feeds of EBM  
NG/ gavage feeds  
Check if feeds retained or vomited out  
At next feed aspirate to see if food still in stomach  
If vomiting – domperidone syrup  
→ Fails – IV fluids – 10% dextrose + electrolytes

IV fluids; Term – D1 – 60cc/kg }  
Preterm – D1 - 75cc/kg } ↑ by 15cc/kg/day upto a max. of 150cc/kg/day

- ❖ Electrolytes – start from D2 – Na+ - 13cc/Kg/day ← N/S  
K+ - 1cc/kg/day ← KCl  
Ca - 6% of total volume ← 10% Ca gluconate
- ❖ If bleeding manifestations – pt. Tx, ftx, packed RBC
- ❖ If jaundiced – SBR – require therapy – phototherapy/ exchange Tx
- ❖ ABG – correct underlying

### 4) Specific therapy

- ❖ Start empiric AB, change over once ABST available
- ❖ Duration – Neonatal sepsis – 14 days  
Neonatal meningitis – 21 days

	AB	Dose	Age <7days	Age >7days
1 <sup>st</sup> line	c. penicillin	50-100mg/kg/dose	12hly	8hly
	Gentamicin	2.5mg/kg/dose	12hly	8hly
2 <sup>nd</sup> line	Cefotaxime	30-60mg/kg/dose	12hly	8hly
	Amikacin	15mg/kg/day	12hly	8hly
	Netilmycin	2.5mg/kg/dose	12hly	8hly
Staph. (late onset sepsis)	Flucloxacillin			
Broadspectrum	Meropenam	20mg/kg/dose	<2kg -bds	>2kg - tds

- ❖ If cultures (-) and pt. is improving – omit IV AB in 48-72hrs
- ❖ On D<sub>7</sub> omit gentamicin since cannot continue for >7days – risk of ototoxicity, nephrotoxicity, reduced CSF penetration with reducing inflammation of the meninges
- ❖ Change over to cefotaxime
- ❖ Ceftriaxone not recommended in neonates → cholestatic jaundice
- ❖ Meningitis – ampicillin + cefotaxime if GBS isolated – high dose c. penicillin.

5) Monitoring – QHT, IP/OP, pulseoximeter monitoring - PR, BP and saturation  
KUO for complications – fits

6) Mx of fits – IV phenobarbitone 20mg/Kg slow IV bolus → 1mg/Kg/min as an infusion over 30min  
1 vial – 200mg in 1ml – diluted in 10cc distilled water  
Monitor – HR, PR, sedation, RR, BP

7) Mx of the mother – treat underlying maternal infections  
Educate on BM expression – fingers around areolar → make an inward movement  
→Then fingers together movement  
Advise not to squeeze the whole breast

8) Notification

9) Follow up  
Need to assess the following

- ✓ Vision
- ✓ Hearing assessment –
- ✓ Growth and OFC
- ✓ Developmental assessment
- ✓ Immunization – Hib

## Classification of neonatal sepsis

Early onset <48hrs -72hrs	Late onset >72hrs
Perinatally acquired – asc. From Cx Vertical Tx/transplacental	Acquired from care giving environment Horizontal Tx – through hands
Pathogen – GBS                    HSV, CMV E. coli Listeria monocytogens – meconium asp.	Staph aureus, Staph epidermidis (coagulase (-)) E.coli Klebsiella, pseudomonas, proteus Streptococcus faecalis Candida, MRSA
Commonly affects respiratory tract – Apnoea, resp. distress, Temp. instability	
Risk factors – LBW, PROM, chorioamnionitis, VE  Substandard sterility in LR Unskilled substerile resuscitation Prolonged labour Meconium aspiration PHx of GBS baby	LBW Delayed/failure to initiate BF Superficial infection – eye, skin, umbilicus IV lines, frequent needle pricks Any disruption of skin integrity Prolonged course of broad spectrum AB

Common organisms causing sepsis in SL?

- Klebsiella
  - Staph aureus
  - Coliform bacteria
  - Spore forming bacilli

### Complications of neonatal sepsis

What measure can prevent neonatal sepsis?

- 1) Maternal immunization – hepatitis, rubella, polio, VZV
  - 2) Maternal prophylaxis – GBS (*streptococcus agalactiae*) – at 32-36weeks –PROM  
O. penicillin, erythromycin
  - 3) Treat STD of mother
  - 4) Intra-partum chemoprophylaxis – HIV – Zidovudine
  - 5) Good obstetric care, clean LR, trained staff
  - 6) Improve postnatal care
  - 7) Minimize AB use
  - 8) Proper care of umbilicus
  - 9) Change cannula, indwelling catheters as soon as it has served its purpose
  - 10) prompt treatment of superficial infections
    - Staph. *Epidermidis/aureus* – neomycin eye ointment tds x 1/52
    - Chlamydia trachomatis* – tetracycline eye drops x 3/52 or o. erythromycin 2/52

## Neonatal meningitis

- Complicates 20% of early neonatal sepsis and 10% of late neonatal sepsis

Common organisms → Group B streptococcus  
E. coli and other coliforms  
Listeria monocytogenes

Complications

- 1) Cerebral abscess
- 2) ventriculitis
- 3) Hydrocephalus
- 4) Hearing loss
- 5) Neurodevelopmental assessment

Chemoprophylaxis for contacts

Hib – neonate – rifampicin 10mg/kg daily x 4days  
Meningococcus – neonate – rifampicin 5mg/kg x 12hly for 2days

## Pneumonia

### Presenting complaint

- Fever and cough
- Fever and hemoptysis
- Fever and chest pain

### Fever and cough

### History of presenting complaint

- **Fever duration-Acute /Chronic**
  - ✓ Acute-Pneumonia, epiglottitis
  - ✓ Chronic-TB
- **Severity of fever**
  - ✓ High grade fever, may have sweating, rigors-Bacterial pneumonia, Epiglottitis
  - ✓ If low grade fever - TB, Atypical pneumonia(Mycoplasma)
- **Type of fever-**
  - ✓ Intermittent fever-TB
  - ✓ Continuous fever-Lobar pneumonia
- **Symptoms prior to fever-**
  - ✓ Preceding URTI - Sneezing, Sore throat, Runny nose(Following viral infection) - *Streptococcus pneumoniae, Staphylococcus aureus*
  - ✓ Viral infection predispose to bacterial pneumonia by damaging the respiratory epithelium
  - ✓ Prominent prodromal features-malaise, Body weakness, Arthralgia, Myalgia- Atypical pneumonia
- **Cough:-**
  - ✓ Acute-Pneumonia(Productive cough)
  - ✓ Chronic(more than 3 weeks)-TB
  - ✓ Severe barking type cough-Croup
  - ✓ Absent or silent cough- Epiglotitis
- **sputum:-**
  - ✓ Frequency, quantity ,appearance of expectorated sputum
  - ✓ Blood stained sputum- Pneumonia (Rusty sputum in *Streptococcus pneumoniae*, *Klebsiella pueumoniae*-Blood stained sputum), TB
  - ✓ Purulent sputum-Pneumonia, TB
- **Chest pain:-**
  - ✓ Pleuritic chest pain-Pneumonia
  - ✓ Need to exclude cardiac causes-Chest pain on exertion, Orthopnea, Paroxysmal Nocturnal Dyspnea, Palpitations, oedema
- **SOB**
  - ✓ Acute SOB-Pneumonia, Other cardiac respiratory, renal causes exclude

### Differential Diagnosis

- Pneumonia
- Atypical pneumonia
- Tuberculosis
- Bronchiolitis
- Epiglottitis
- Laryngotracheitis

**Pneumonia (*Strep. pneumoniae, haemophilus influenzae, klebsiella, Staph aureus*)**

- ✓ Fever-Acute onset fever
- ✓ Cough-acute onset, productive
- ✓ Sputum-purulent/Blood stained cause hemoptysis
- ✓ Chest pain(Pleurisy)-Pleuritic type chest pain
- ✓ + or - breathlessness-acute onset

**Aetiology**

- ✓ Recent hospitalization(Pneumonia by gram -ve)
- ✓ Hx of aspiration-
- ✓ GORD - Regurgitation, acid reflux, Develop
- ✓ Period of loss of consciousness in past - Epilepsy
- ✓ Oesophageal obstruction - Dysphagia, past hx Strictures - any acid ingestions
- ✓ Myasthenia gravis-easy fatigability.

***Staphylococcus aureus***

- ✓ Very ill patient
- ✓ Usually get after proceeding viral infection of influenzal illness. CXR-Patchy consolidations, break down to form abscesses. May appear as cysts on x ray. Pneumothorax, Empyema, effusions common.

**Tuberculosis**

- ✓ Fever-Low grade nocturnal fever long standing
- ✓ Cough-chronic cough>3 weeks,
- ✓ Sputum:-Blood stained sputum
- ✓ Loss of appetite, Night sweats, Loss of weight

**Aetiology**

- ✓ PHx or contact hx of contact hx of TB

**Atypical pneumonia**

**Aetiology**

No response to usual antibiotics (Previous treatment has not answered)

***Legionella***

- ✓ Previously fit individuals staying in hotels, institutions or hospitals where the shower facilities or cooling system (A/C) have been contaminated with the organism.
  - ✓ Sporadic cases where the source of the infection is unknown: most cases involve passive smokers(middle aged and elderly men who are smokers-at home)
  - ✓ Outbreaks occurring in immunocompromised patients:- Corticosteroid therapy
  - ✓ Malaise, myalgia, headache, fever with rigors, GIT symptoms:-vomiting, diarrhea, abdominal pain, Hematuria
- Complication**
- ✓ hematuria, Oliguria, reduce urine output(ARF)

***Mycoplasma pneumoniae***

- ✓ Address- Boarding institutions
- ✓ Scanty chest signs

#### **Extrapulmonary features**

- ✓ Myocarditis, Pericarditis-Fatigue, dyspnea, chest pain, palpitation
- ✓ Rashes, Erythema multiform
- ✓ Haemolytic anaemia and Thrombocytopaenia→ Lethargy, faintishness
- ✓ Myalgia, Arthralgia
- ✓ Meningo-encephalitis→confusion, headache, weakness of body
- ✓ Other neurological signs
- ✓ Gastrointestinal symptoms
- ✓ Eg:-Vomiting, Diarrhea

#### ***Chlamydia Psittaci***

- ✓ Get from infected bids handle
- ✓ Symptoms:-Malaise, High fever, Muscular pain, Liver and spleen occasionally enlarged.
- ✓ Scanty rose spots may seen on abdomen

#### **Viral**

- ✓ Uncommon in adults
- ✓ Viral infection predispose to bacterial infections

#### **Systemic enquiry**

- ✓ **GIT**-Jaundice(Legionella, Mycoplasma),
- ✓ Epigastric+ RHC pain→Atypical(hepatitis) sub phrenic abscess, Amoebic liver abscess, Diaphragmatic pleurisy,
- ✓ Associate with dark urine and stools→Mycoplasma (haemolytic anaemia)
- ✓ Diarrhea-Legionella
- ✓ **Genito-urinary**→ Reduce Urine output-Sepsis- Pre renal ARF/ATN, ARF by legionella, dehydration, Haematuria
- ✓ **CVS**→Pericarditis(Commonly with left side pneumonia)→Precordial pain worse on lying down and relieved by sitting forward, Arrhythmias(SVT), Myocarditis-Mycoplasma
- ✓ Postural hypotension→sepsis, cardiogenic shock
- ✓ **Musculoskeletal system**→Polyarthritis-Mycoplasma, Septic arthritis-Sepsis
- ✓ **Nervous system**-Non specific headache,photophobia,fits-Meningoencephalitis(Mycoplasma)
- ✓ Confusion, reduce concentration,drowsy-atypical, alcohol withdrawal
- ✓ Focal signs-Slurred speech,weakness(septic emboli--cerebral abscess)
- ✓ **Skin**-Non specific rash-Mycoplasma, bleeding in to skin(DIC)

If patient was previously treated by a doctor or hospital need to mention it.

#### **Exclude other causes of fever and cough**

- Epiglottitis→High fever in an ill patient, intensely painful throat and prevents the child from speaking or swallowing(Saliva drools down the chin)  
Soft inspiratory stridor, child immobile, upright with an open mouth to optimize the airway.
- Bronchiolitis→Coryza symptoms, Dry cough, increasing breathlessness, associated fever
- Croup(Laryngotracheitis)→Onset over days, preceding coryza present, cough-severe barking type, no drooling saliva, able to swallow, fever, harsh stridor, Hoarse voice

**Immunization:-** Hib vaccination given

**PMHx:-** Recurrent pneumonia

### Social hx

- ✓ Contact hx of fever
- ✓ Bird handlers-Psittacosis, avian influenza
- ✓ Animal handlers-Coxiella burnetti

### Examination

#### General examination

- ✓ Dyspnea, Febrile, Ill looking
- ✓ Sputum pot→Purulent(TB/Pneumonia)
- ✓ Pale(Mycoplasma, TB)
- ✓ Icteric(Atypical pneumonia)
- ✓ Skin rash
- ✓ Maculopapular erythema multiform→Mycoplasma
- ✓ Echymosis, purpura, bruises, bleeding from puncture site
- ✓ B/L pedal oedema

#### Respiratory system

- ✓ Features of respiratory distress, →chest wall recessions, use of accessory muscles of respiration, Cyanosis and grunting
- ✓ Reduce movements on affected side→Pneumonia, TB
- ✓ Trachea in mid line
- ✓ Dull on percussion-stony dull if associate with Pleural effusion
- ✓ increase vocal fremitus, Reduced if Pleural effusion
- ✓ Reduce air entry in Pleural effusion
- ✓ Coarse crepitations→ Pneumonia, TB
- ✓ Tachypnea

Age	Time taken
<2months	Over 60 breaths per minute
2 months-12months	Over 50 breaths per minute
12 months to 5 yrs	Over 40 breaths per minute
More than 5 sing-	Over 20 breaths per minute

#### CVS

- ✓ BP- reduce in septic shock, myocarditis
- ✓ Increase JVP, Gallop rhythm->In heart failure
- ✓ Murmur-Infective endocarditis
- ✓ Pericardial rub

#### Abdominal examination

- ✓ Tender hepatomegaly - Legionella Hepatitis, Liver abscess
- ✓ Massive spleen + Hepatomegaly→Haematological malignancy, Myelofibrosis

**CNS**

- ✓ GCS-Confusion due to atypical pneumonia, sepsis, respiratory failure, severe pneumonia,
- ✓ meningitis
- ✓ Cerebral abscess
- ✓ Increase intra cranial pressure→Papilledema
- ✓ Focal signs→Meningo-encephalitis

**Problem list**

1. **Medical problem**
  - a) Acute
  - b) Chronic
2. **Psychological problem**
3. **Social problems**
  - a) Economic problem

**Discussion**

- **How will you investigate this patient?**
  - ✓ **Blood investigations**→ FBC, Blood culture, CRP, ESR  
When CRP rise→ end of 1<sup>st</sup> day (24 hours), Fever of one day(RR will be normal. But you can do a CRP level and see the rise.)  
CRP-Increase in infections (It is an acute phase protein)
  - ✓ If Pleural effusion →pleural fluid analysis if significant pleural effusion present
  - ✓ Radiological investigations→CXR

	<b>Viral</b>	<b>Bacterial</b>	<b>Atypical pneumonia (Mycoplasma)</b>
<b>Clinical</b>	<ul style="list-style-type: none"> <li>• Low grade fever</li> <li>• Respiratory rate normal or slightly raised,</li> <li>• wheezing,</li> <li>• Marked chest wall recessions</li> <li>• Hyperinflation</li> </ul>	<ul style="list-style-type: none"> <li>• High grade fever(&gt;38.5)</li> <li>• Respiratory rate high</li> <li>• No wheezing</li> <li>• Chest wall recessions</li> </ul>	<ul style="list-style-type: none"> <li>• Low grade fever</li> <li>• Associated wheezing</li> <li>• Prolonged disease course</li> <li>• Prominent headache</li> <li>• Arthralgia, Myalgia,</li> <li>• Extrapulmonary manifestations</li> </ul>
<b>Investigation</b>	Usually no neutrophil leukocytosis or CRP	Neutrophil leukocytosis >150 000 WBC CRP elevated>35 to 60mg/l	Special investigations Serology Cold agglutination test
<b>Chest X ray</b>	Hyperinflation and lobar collapse	Consolidation, Pleural effusion, Special finding may also indicate the aetiology→Pneumatoceles, Cavitation(Staph aureus)	<ul style="list-style-type: none"> <li>• Reticulonodular opacification of the lower lobe</li> <li>• Hilar lymphadenopathy</li> <li>• Interstitial infiltrates</li> </ul>

Age	Organisms
Neonates	<b>Bacterial</b> → <i>Group B streptococcus, E coli, Klebsiella, Listeria monocytogenes</i>
1 month – 1 year	<b>Viral</b> → RSV, Para-influenza <b>Bacterial</b> → <i>Streptococcus pneumonia, Staphylococcus aureus, Chlamydia trachomatis, Bordetella pertussis</i>
1-5 years	<b>Viral</b> → RSV, Para-influenza, Influenza, Adenovirus <b>Bacterial</b> → <i>Streptococcus pneumonia, Staphylococcus aures, Haemophilus influenza b, Mycoplasma, MTB</i>
>5 years	<b>Bacterial</b> → <i>Streptococcus pneumonia, Mycoplasma, Chlamydia pneumonia, MTB</i>

- ✓ In infants and <5 years old children pneumonia is commonly caused by viruses
- ✓ In children >5 years of age Streptococcus pneumonia is the common bacterial aetiology.

- How will you assess the severity of pneumonia?

Age	Mild severe	Very severe
Infants	Temperature <38.5C, RR <50 cycles/min Mild recession Taking full feeds	Temperature >38.5 C RR>70breaths/min Moderate to severe recession Nasal flaring Cyanosis Intermittent apnea Grunting respiration Not feeding
Elderly children		Temperature >38.5 C RR>50 breaths/min Severe difficulty in breathing Nasal flaring Cyanosis Grunting respiration Signs of dehydration

- How TB cause broncho pneumonia, in a TB patient?

When there are paratracheal LN, it can perforate in to the bronchus discard all organisms and cause pneumonia. If go to a main bronchus then will go to that lung

- What is the drug of choice in atypical pneumonia?

Erythromycin (Macroloides)

- Causes of recurrent pneumonia?

1. Tracheo-oesophageal fistula
2. GORD
3. Immunosuppressive individuals

## Management

### Mx the acute state

- ✓ Assess the ABC
- ✓ Airway → oxygen supply via face mask,
- ✓ Oxygen therapy should be considered if saturation is less than 92%
- ✓ Circulation → Obtain IV access and take blood for investigations → FBC, CRP, blood culture
- ✓ Consider IV fluids if patient cannot take orally
- ✓ Mx fever and pain with PCM
- ✓ Start monitoring → PR, RR, BP, Oxygen saturation
- ✓ Feeding of the child → Try to avoid insertion of NG tube for feeding

### Antibiotic management

<b>0-3 months</b>	<b>IV Penicillin /Ampicillin and gentamycin as first line</b> Cefotaxime/ Co-amoxiclav 2 <sup>nd</sup> line
<b>3 months-1 year</b>	If the child is not ill: <b>Oral amoxicillin</b> If ill/lobar infiltrates on chest X-ray : <b>IV ampicillin</b> If no response within 48 hrs → use 2 <sup>nd</sup> or 3 <sup>rd</sup> generation cephalosporins ( <b>Cefuroxime/Cefotaxime</b> ) If suspecting Staph → IV cloxacillin
<b>Children 1-5 years</b>	If the child is not ill: <b>Oral amoxicillin</b> If ill/lobar infiltrates on chest X-ray : <b>IV penicillin</b> If no response within 48 hrs → use 2 <sup>nd</sup> or 3 <sup>rd</sup> generation cephalosporins ( <b>Cefuroxime/Cefotaxime</b> ) If suspecting Staph → IV cloxacillin <b>Macrolides</b> if suspecting Atypical pneumonia
<b>Children &gt;5years</b>	If the child is not ill → Macrolides → <b>Erythromycin, Clarithromycin</b> If ill/toxic → treat as for 1-5 years (Penicillin/Cephalosporins)

- ✓ Mycoplasma needs macroloids
- ✓ IV treatment at least till temperature is settled and no respiratory difficulty for 24 hrs. Then converted to oral
- ✓ Altogether duration 5-7 days

- **Why we need oral IV?** To get the Minimum inhibitory concentration quickly

- **Other aspects of management**

- ✓ Control temperature
- ✓ Hydration
- ✓ Nutrition

- **Indication for hospital admission**

1. Severe disease
2. Needing supportive treatment eg:-needing oxygen
3. Intensive care/High dependency unit
4. Not tolerating oral antibiotics
5. Social issues

- **In daily ward round what would you look in this patient?**
  - ✓ Look at the general condition of the child – fever, tolerating oral feeds/not
  - ✓ Examine the respiratory system of child
  - ✓ Look at the monitoring chart
  - ✓ Look at the latest investigations

## Discussion

- **What causes the pneumoniae due to opportunistic infections?**

Cytomegalovirus/ M.tuberculosis/M.avium-intracellulare/ L.pnuemophila/ Cryptococcus/Pyogenic bacteria/ Kaposi's sarcoma/ Lymphoid interstitial pneumonia/ Non specific interstitial pneumonia

- **What is pneumonia?**

Inflammation of lung tissues with accumulation of cells and secretions in alveolar spaces usually following an infection

- **How pneumonia is categorized according to aetiology?**

- a. Community acquired pneumonia
- b. Hospital acquired pneumonia
- c. Aspiration pneumonia
- d. Immunocompromised

- **What are the causes of community acquired pneumonia**

- ✓ Bacteria pneumonia-Streptococcus pneumoniae, Klebsiella, Staphylococcus aureus, Haemophilus influenza
- ✓ Atypical pneumonia-Mycoplasma, Legionella, Chlamydia, Viruses

- **What are the pulmonary complications of pneumonia?**

- a. Pleural effusion
- b. Empyema
- c. Lung abscess
- d. Respiratory failure

- How to perform the agglutination test in detection of mycoplasma?

### Cold agglutinin ward test<sup>3</sup>

*This often forgotten bedside test can be very useful in resource poor settings.  
It is positive in half the cases.*

1. Take 0.5 ml of blood to test tube.
2. Add 0.5ml of sodium citrate.
3. Place it in an ice bucket or refrigerator for 20 minutes.
4. Tilt the tube and look for layering of a film of clots from below.
5. The “grains of sand effect” appear on the glass portion of the tube.



Cold agglutinin ward test

### Treatment<sup>5</sup>

Macrolides are used if *Mycoplasma* or *Chlamydia pneumoniae* are suspected [D]. Because mycoplasma pneumonia is more prevalent in older children macrolide antibiotics may be used as first line empirical treatment in children aged 5 and above with community acquired pneumonia. [D]

Erythromycin 40mg/kg/ day orally 6 hourly for 7-10 days.

Azithromycin 12mg/kg as a single daily dose for 5 days

Clarithromycin 15 mg/kg day 12 hourly for 10 days.

The newer macrolides are better tolerated and have lesser dosing frequency.

Antibiotic prophylaxis for household contacts is not routinely recommended. However, if there are high risk household contacts, consider prophylaxis.

## *The scheme for management of acute epiglottitis*

- Toxic
- High fever
- Irritable
- Dyspnoeic
- Abnormal posture
- Muffled voice
- Chest indrawing



**Epiglottitis**



**Give 100% oxygen. Do not disturb the child. Do not attempt to examine the throat. It may precipitate total airway obstruction**



**Get help. Call the consultant anaesthetist, ENT surgeon and the paediatrician**



**Assessment of the airway by the anaesthetist or ENT surgeon**



**Intubate and secure airway. Failing which do the tracheostomy**



**Check the adequacy of the ventilation**



**Check circulation, gain intravenous access**



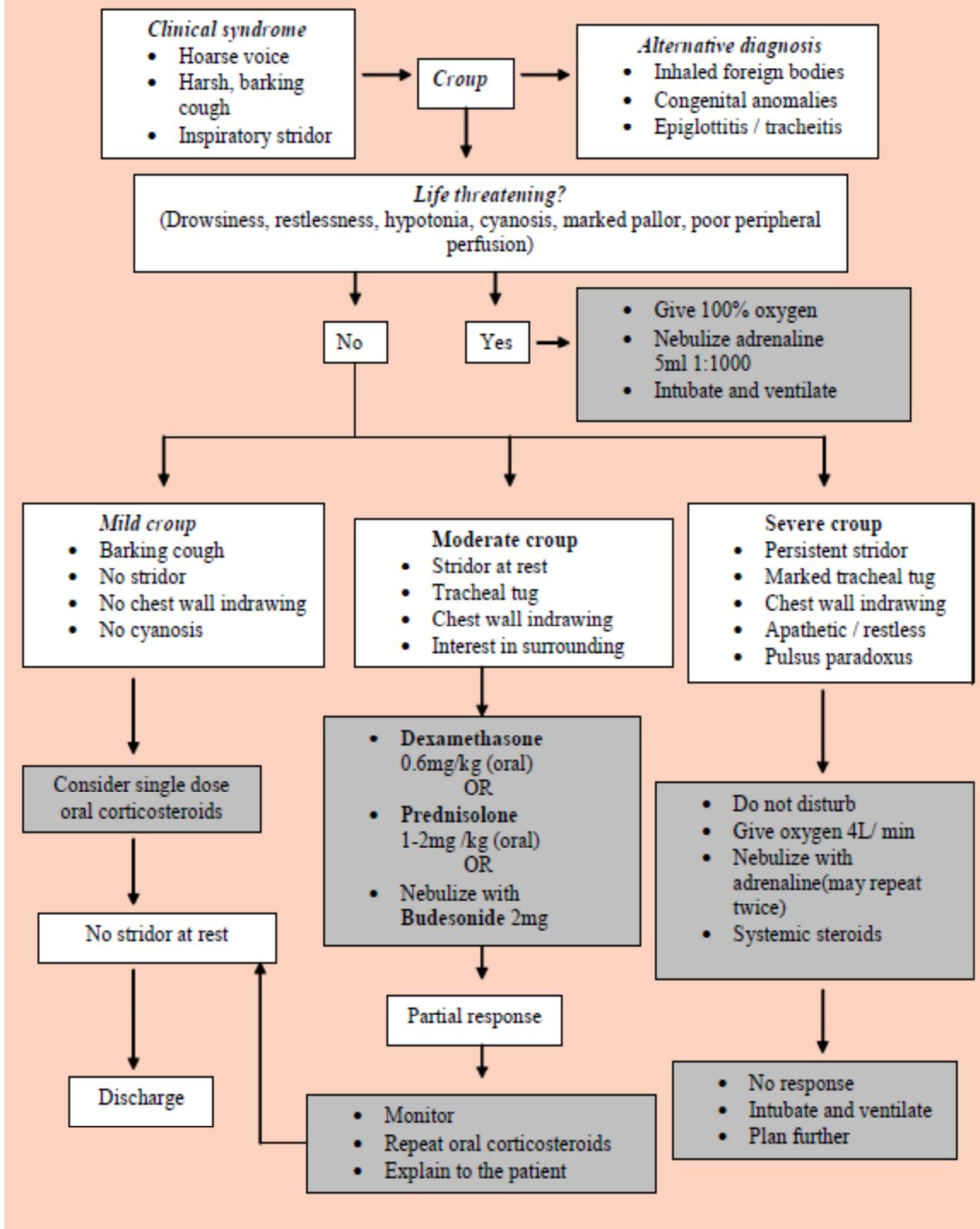
**Do the blood culture**



**Start iv ceftriaxone or iv cefotaxime**

*If these antibiotics are not readily available, use chloramphenicol 100mg /kg/d in divided doses.*

## The scheme of management for croup

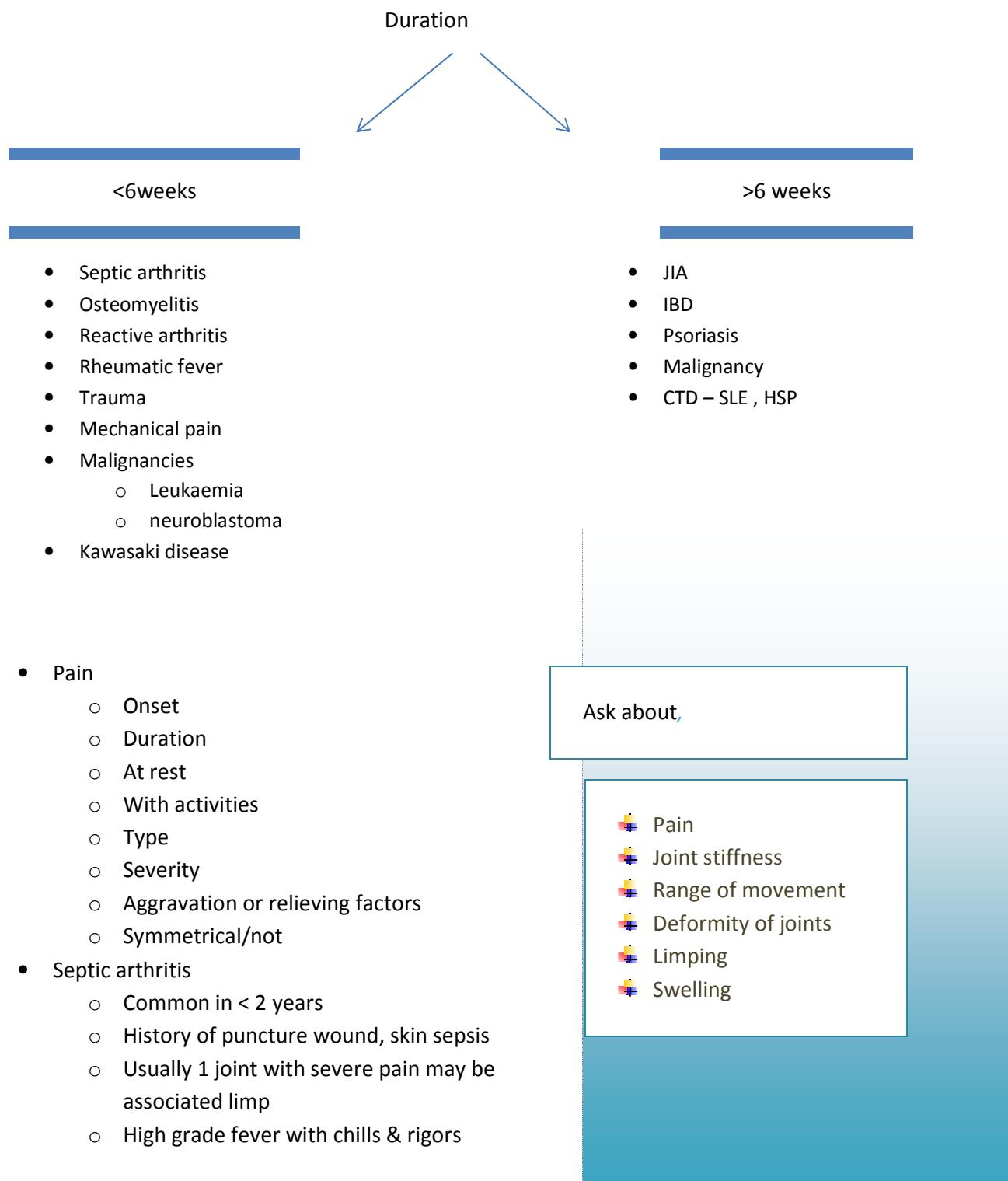


## Arthritis in Children [JIA]

### Introduction – age

PC – pain & swelling of joint/s

What joints



- No movements due to pain
  - Hib vaccine ( young children *H. influenza*)
  - Immunodeficiency
- Osteomyelitis
  - Painful immobile limb with swelling over the affected joint
  - Painful movement
  - Fever
  - Back pain in vertebral infection
  - Groin pain in pelvic infection
- Reactive arthritis
  - The most common form of arthritis
  - Transient joint swelling
  - History of Dysentery (*Salmonella*, *Shigella*, *Campylobacter*)
  - History of rubella, hand foot & mouth disease , *Mycoplasma*(Pneumonia)
  - Following Lyme disease
    - Fever, headache, fatigue
    - Skin rash
    - Migrating arthritis
    - Only one joint at a time
    - Large joints
- Rheumatic fever
  - Refer rheumatic fever note
- History of trauma
- Malignancy with bone marrow occupying carcinoma
  - Neuroblastoma
    - Age <5 years
    - Bone pain
    - Recurrent infections
    - Bleeding manifestations
    - Anemic symptoms
    - Weight loss
    - Malaise

Bone marrow suppression

- Acute lymphoblastic leukaemia
  - Joint pain & arthritis
  - Severe bone pain which awaken the child from sleep
  - Bleeding manifestations – abnormal bruising
  - Anemic symptoms
  - Recurrent infection
  - Malaise, anorexia
  - Headache, vomiting [ due to CNS infiltration]
- Kawasaki disease
  - Fever >5 days
  - Rash
- Psoriatic arthritis
  - **Skin rash comes first**
  - Joint pain, stiffness, swelling – commonly DIPJs, asymmetrical involvement
  - Hx of remission
  - Skin rash & arthritis may come at the same time & disappear
- IBD
  - Asymmetrical arthritis – predominantly LL
  - Symptoms during flare of IBD
- JIA
  - Age <16 years
  - Arthritis 6 weeks ago
  - Insidious or abrupt onset
  - Morning stiffness or stiffness after long period of sitting/inactivity( gelling phenomenon)
  - Arthralgia, sometimes no pain
  - A morning limp that improves with time
  - Contractures, reduce range of movements – due to stop using the joint
    1. Oligoarthritis (persistent)
      - 1 – 4 joints
      - Knee , ankle, wrist common
      - Chronic anterior uveitis
    2. Oligoarthritis (extended)
      - >4 joints involved after first 6 months
      - Asymmetrical involvement
        - Large & small joints
      - Chronic anterior uveitis

3. Polyarthritis

- 5 or more joints
- Symmetrical large & small joints
- Marked finger involvement.
- Cervical spine/temporomandibular joints may be involved.
- Low grade fever.
- Rheumatic factor +ve & -ve.;

4. Systemic arthritis

- Acute illness
- Arthralgia, Myalgia. Initially no arthritis
- Spiking fever once or twice daily.
- Oligo / polyarthritis
- Malaise
- Salmon pink rash, macular rash
- Lymphadenopathy, hepatosplenomegaly
- Chest pain, SOB due to pleural effusions
- Pericarditis

5. Psoriatic arthritis

- Asymmetrical large & small joints
- Dactylitis
- Psoriasis
- Nail pitting
- Chronic anterior uveitis

6. Enthesitis arthritis

- Lower limb large joint arthritis initially
- Mild lumbar spine or sacro-iliac involvement later-on
- Axial joint involvement – buttock & back pain
- Evening or post exercise pain

• SLE

- >8 years
- Fever, anorexia, weight loss
- Rash over face & photosensitive rash
- Oral ulcers
- Arthritis, joint stiffness in the morning
- Pleurisy, pericarditis
- Glomerulonephritis – oedema, haematuria, frothy urine
- Seizures
- Anemic symptoms

- HSP
  - Arthritis (knee, ankle), peri-articular oedema (no long-term damage to the joints)
  - Low grade fever ,fatigue
  - Skin rash in buttock, extensor surface
  - Abdominal pain
  - History of URTI
  - Haematuria
  - Hepatosplenomegaly, lymphadenopathy
  - CNS involvement
- ❖ Previous episodes
- ❖ First presentation
  - When
  - How
  - What was done
- ❖ Remission & relapsing
- ❖ Whether on treatment or not
  - Drugs
  - Duration
  - Complications
- ❖ Complications of the disease
  - Limp – pain, limb length discrepancy
  - Deformities
  - Growth failure
  - Contractures
  - Disability
  - Blind vision
- PMH
  - Sickle cell disease,
  - IBD
  - Psoriasis
- PSH
  - Joint surgeries
- DHx
  - Immuno compromising drugs like steroids, cyclophosphamide

➤ **FHx**

- RA, OA
- Psoriasis
- Gout
- IBD
- Ankylosing spondylitis

➤ **SHx**

- Level of education of the parents
- Socioeconomic status
- How does the illness affect the child
  - School absence/severity
  - School performance
  - Involve in sports activities
  - Mental state

➤ **Examination**

- Height
- Weight
- General appearance
  - Ill looking or not
- Septic arthritis
  - Very ill
  - Febrile
  - Wounds or infected skin lesions
  - Erythematous, swollen, warm, tender joint
  - Reduced range of movements- no movements
  - Joint effusion
- Osteomyelitis
  - Not very ill
  - Swollen tender erythematous joint
  - Some movement
  - Joint effusion
- Reactive arthritis
  - Large joints
  - Arthritis features
  - Lyme disease
    - Skin rash – erythema migrans at the site of tick bite
    - Febrile
    - 1 joint affected

- Rheumatic fever
  - Refer rheumatic fever note
- Leukaemia
  - Pallor
  - Bruises, petechiae
  - Lymphadenopathy
  - Hepatosplenomegaly
  - Cranial nerve palsies – CNS infiltration
- Neuroblastoma
  - Pallor
  - Bruises , petichiae
  - wasted
  - Abdominal distension
  - Abdominal lump ( crosses the midline)
- Kawasaki disease
  - febrile
  - Red, dry cracked lips
  - Strawberry tongue
  - Conjunctival injection
  - Cervical lymphadenopathy
  - Skin rash
  - Peeling of palms & soles
  - Gallop rhythm
- JIA
  - Gait
  - Joint examination
    - Joint swelling
    - Tender, warmth, erythema, limited ROM
- 1. Systemic arthritis
  - Ill looking, febrile
  - Pale
  - Evanescent salmon pink macular rash in trunk & extremities
  - Lymphadenopathy
  - Muscle tenderness
  - Hepatosplenomegaly
  - Pleural effusion
  - Pericardial effusion – pericardial rub, muffled heart sounds

2. Oligoarthritis

- Involvement in <4 joints
- Gait – walk with a limp
- Muscle atrophy
- Flexion contractures
- Slit lamp examination of eye for anterior uveitis

3. Polyarthritis

- Rheumatic factor +ve
  - Rheumatoid nodules
  - Look for both large & small joints
  - Symmetrical
  - Slit lamp examination of eye for anterior uveitis
- Rheumatic factor -ve
  - Febrile
  - Look small & large joints, hand, cervical spine, temperomandibular joint

4. Psoriatic arthritis

- SLE
  - Pallor
  - Febrile
  - Red eyes
  - Malar rash
  - Discoid lesions – in face & head
  - Oral ulcers
  - Lymphadenopathy
  - Raynaud's, nail fold vasculitis
  - Purpura, urticaria
  - BP
  - Pericardial rub, muffled heart sounds
  - Pleural effusion
  - Joint swelling, tenderness, reduced range of movements, effusion
- HSP
  - Febrile
  - Palpable maculo papular purpuric rash over calf, buttocks (over the extensor surfaces)
  - Angio oedema
  - Generalized edema, BP
  - Joint swelling, tenderness usually knees & ankles
  - Lymphadenopathy
  - Hepatosplenomegaly

- **Investigations**

Disease	FBC findings
Septic arthritis	Neutrophil leukocytosis
Osteomyelitis	Neutrophil leukocytosis
Leukemia	Neutropenia, thrombocytopenia, lymphocytosis, anaemia
JIA	Leukocytosis, anemia, thrombocytosis,
SLE	Leukopenia, thrombocytopenia, lymphopenia, anaemia
HSP	leukocytosis, thrombocytosis

- ✓ Blood picture
  - Abnormal blast cells in ALL
  - Normochromic normocytic anemia in JIA
- ✓ ESR, CRP
  - Elevated in
    - JIA
    - Septic arthritis
    - osteomyelitis
    - HSP
    - High ESR & normal CRP in SLE
- ✓ Septic arthritis
  - Joint aspiration for culture & ABST – definitive
  - USS scan – joint effusion
  - X ray of the joint
    - Joint space widening
    - Soft tissue swelling
    - Other feature are normal initially
    - Exclude trauma & bone diseases
  - Blood culture
    - Start antibiotics after this
  - Prolong antibiotics
    - Initially IV
  - Washing out of the joint (Arthroscopic washout) or surgical drainage
- ✓ Osteomyelitis
  - Blood culture
  - X ray
    - Initially normal, only soft tissue swelling
    - Take 7- 10 days for sub-periosteal bone deposition to occur
    - Hypo dense area in metaphyseal area of the bone
  - Antibiotics for several weeks to prevent bone necrosis, discharging sinuses, deformity, amyloidosis

- Resting in a splint initially. Then mobilized
  - IV antibiotics till clinical recovery & CRP become normal
  - Then oral for several weeks.
  - Atypical or immunocompromised
    - Aspiration
    - Surgical decompression
  - If not responding for antibiotics rapidly
    - Surgical drainage
- ✓ Kawasaki disease
- ESR elevated, ↑ Platelets
  - Other than above Ix,
    - Echo – coronary artery aneurysm
  - IV IG within first 10 days
  - Aspirin
- ✓ SLE
- ANA, anti dsDNA +ve
  - Low C3, C4
  - Hypergammaglobulinemia
- ✓ HSP
- Skin biopsy – leucocytoclastic angitis
  - UFR – if renal involvement
    - RBC
    - Protein
  - Supportive treatment
    - Hydration
    - Balanced diet
    - Analgesia for pain
    - Monitor BP, PR, IP/OP
- ✓ JIA
- S.Creatinine
  - BU/SE
  - ANA – elevated in oligoarthritis
  - RF +ve / -ve
  - HLA B27 +ve in enthesitis arthritis
  - X ray
    - Early – soft tissue swelling, periostitis
    - Late – subchondral bone erosions, bone destruction & fusion

- USS scan – joint effusions
- Multidisciplinary team approach
- No definitive cure. Control disease
- Pharmacology
  - Pain relief – NSAIDs (naproxen, Ibuprofen)
  - Methotrexate with folic acid (weekly) – preferred drug
  - Sulfasalazine
  - Cyclophosphamide
  - Eternacept (TNF alpha antagonist)
  - Steroids – acute exacerbation
- Non pharmacology
  - Nutrition
  - Physiotherapy – improve joint mobility & walking
  - Occupational therapy –writing
- Monitor FBC, LFT
- Refer the patient to an ophthalmologist for slit lamp examination of the eye
- Because anterior chronic uveitis is difficult to identify clinically. it lead to permanent visual impairment or blindness

## DISCUSSION

### ➤ Cause of arthritis

Infection	Bacterial – septic arthritis, osteomyelitis, TB Viral – rubella , mumps, adeno virus, coxsackie B, parvo Other - mycoplasma, Lyme disease, riccketsia Reactive – GI infection, <i>streptococcal</i> infection Rheumatic fever
IBD	Crohn's disease, ulcerative colitis
Vasculitis	HSP, Kawasaki disease
Haematologic	Haemophelia, sickle cell disease
Malignant	Leukaemia, Neuroblastoma
CTD	JIA, SLE, mixed connective tissue disease, poliarteritis nodosa
other	Cystic fibrosis

### Septic arthritis

- Infection of the joint space lead to joint destruction
- Common in <2 years
- Resulting from haematogenous spread or infected skin lesions
- Usually only one joint
- Age >neonates common organism is *Staph. Aureus*
- Young children prior to Hib vaccine it is *H.influenzae*
- Immunodeficiency & sickle cell disease are predisposing conditions

### Osteomyelitis

- Infection of the metaphysis of the long bones
- Distal femur & proximal tibia is the commonest sites
- Usually due to haematogenous spread or directly from infected wounds
- Commonest is *Staph. Aureus*
- In sickle cell anemia - high risk of staphylococcal & salmonella osteomyelitis
- Chronic infection – abscess formation (Bordetella abscess)
- Joint effusion of the adjacent joint is usually sterile effusion

### JIA (Juvenile chronic arthritis, Juvenile Rheumatoid arthritis)

Diagnostic criteria:

- Arthritis
- Persisting at least >6 weeks
- Age of onset <16 years
- Other causes of arthritis is excluded

### Types of JIA

Sub types	onset	Sex ratio F : M	Articular pattern	Extra articular features	Laboratory findings
<b>Oligoarthritis (persistent)</b>	1-6 yrs	5:1	1-4 joints, knee, ankle or wrist common	Chronic anterior uveitis, leg length discrepancy, prognosis excellent	ANA +
<b>Oligoarthritis (extended)</b>	1 - 6yrs	5:1	>4 joints, after 6 months. Asymmetrical distribution of large & small joints	Chronic anterior uveitis Asymmetrical growth Moderate prognosis	ANA +
<b>RF negative polyarthritis</b>	1 - 6yrs	5:1	Symmetrical large & small joint Finger involvement Cervical, temperomandibular joint	Low grade fever Chronic anterior uveitis, late reduction of growth rate	

<b>RF positive polyarthritis</b>	10 – 16yrs	5:1	Symmetrical large & small joints, marked finger involvement	Rheumatoid nodules Poor prognosis	RF +
<b>Systemic arthritis</b>	1 – 10yrs	1:1	Oligo – polyarthritis Arthralgia, myalgia	Acute, high fever daily Malaise, Macular salmon pink rash, lymphadenopathy, hepatosplenomegaly serositis	Anaemia Neutrophil leukocytosis Thrombocytosis High CRP
<b>Psoriatic arthritis</b>	1 – 16yrs	1:1	Asymmetrical large & small joints, dactylitis	Psoriasis, nail pitting, dystrophy Chronic anterior uveitis	
<b>Enthesitis</b>	1 – 16yrs	1:4	Initially LL large joints Later mild lumbar spine, sacro iliac joints	Often Achilles enthesitis Acute anterior uveitis	HLA B27 +

Undifferentiated 1- 16yrs 2:1 overlapping of articular & prognostic variable

Extra articular features of

2 or more subtypes