

Long cases in paediatrics

Acute fever

Presenting complaint

- Fever
- State the duration

History of the presenting complaint

Description of the fever

Describe the following details of the fever in a chronological order

- Onset and any preceding symptoms
- Progression
- Height of the fever – documented or not
- Resolution of the fever and state of the child in between episodes
- Recurrence (comment on the fever pattern if possible)
- What the mother did at home? Especially important is the dose of paracetamol
- Associated factors

Associated features

- Ask for symptoms related to the important symptoms to try to identify a focus of infection and to think of a differential diagnosis

| Disease | Symptoms |
|--|---|
| Dengue fever | Headache, retro –orbital pain, arthralgia and myalgia, anorexia, nausea and vomiting Warning signs Abdominal pain, mucosal bleeding and other bleeding manifestations, lethargy and restlessness |
| Respiratory tract infection | Ask for Cough, sputum (if sputum is associated state the color and amount), rhinorrhoea, chest pain associated with breathing and difficulty in breathing |
| Ear infection | Ear pain and discharge |
| Pharyngitis | Ask for sore throat, pain on swallowing |
| CNS infection (Meningitis and encephalitis) | Headache, photophobia, altered behavior and loss of consciousness, seizures |
| GI infection | Ask for passage of loose stools |
| Hepatitis | Yellowish discoloration of the eyes, darkening of the urine |
| Leptospirosis | Exposure to muddy water/ possible contaminated water |

| | |
|---|--|
| Septic arthritis and osteomyelitis | Bone pain, joint pain and swelling |
| Urinary tract infection | Crying on passage of urine, frequency, hematuria |

History of exposure and epidemiological history of the fever

- Ask for history of contact with infected or otherwise ill persons
- Travel history if relevant
- History of cases of fever especially dengue fever in the community

Past medical history and surgical history

Other components of the history

Social history

Environment

- Describe the surrounding environment of the house especially with regard to possible mosquito breeding sites
- Ask if the garbage sites are cleaned regularly and ask if mosquito spraying is done regularly in the area
- Ask for the involvement of the MOH, PHI and other staff for dengue prevention in the area
- Ask for possible breeding sites within the house
- If the child is attending montessori or school inquire about the environment

Impact of the disease on the child and on the parents

- Inquire about the amount of school missed by the child
- Impact on the parents as the child has to stay in the hospital
- Concern of the parents

Other general factors on the family background

- Occupation of the parents
- Social circumstances of the family
- Economic status of the family
- Extended family support

Examination

General impression of the patient

- Look at the appearance of the patient
- Look at the alertness and activity of the child
- Examine the vital signs of the patient
 - Pulse rate and volume
 - Capillary refill time
 - Blood pressure and pulse pressure
 - Respiratory rate and signs of respiratory distress

General examination

Do a complete examination from head to toe

- Look for skin rashes
- Eyes
- Conjunctivae for pallor and the sclera for icterus
- Photophobia and neck stiffness
- Ear discharge
- Sinus tenderness
- Lymphadenopathy
- Open the mouth and look at the general oral hygiene
- Examine the throat and inspect the pharynx and tonsils

Respiratory system

- Look for evidence of pneumonia
- Look for pleural effusion – pneumonia, dengue

CVS

Abdomen

- Examine for hepatosplenomegaly
- Other masses
- Free fluid in the abdomen

Nervous system

- Should be assessed completely in a patient with suspected CNS infection

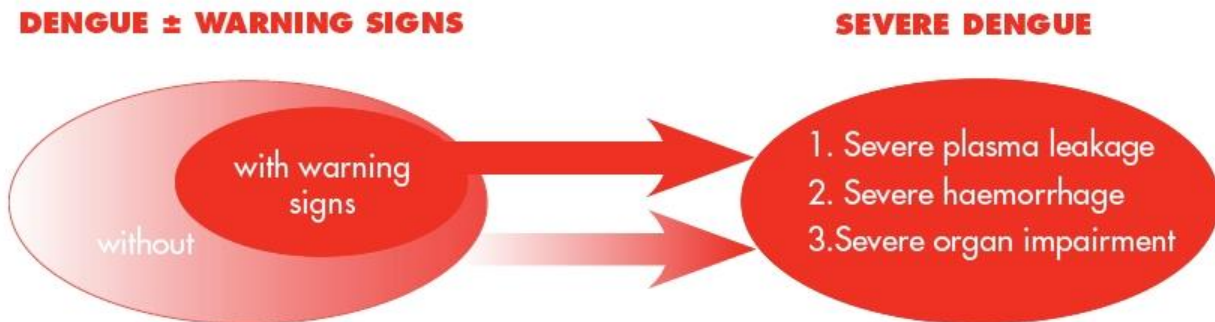
Musculoskeletal – Look for joint swelling and other features of acute inflammation of the joint

Discussion

Dengue fever

What is the diagnosis?

- The first step in a long case of acute fever is to make a diagnosis and classify the severity
- The most common case of acute fever given for the exam is dengue fever



| Probable dengue | Dengue with warning signs | Severe dengue |
|-----------------------------|--|--|
| Living in an endemic area | Persistent vomiting | Severe plasma leakage |
| Fever | Abdominal pain or tenderness | Shock |
| Two of the following | Lethargy, restlessness | Fluid accumulation with respiratory distress |
| Nausea, vomiting | Mucosal bleeding | |
| Arthralgia and myalgia | Clinical evidence of fluid accumulation | Severe bleeding |
| Rash | Liver enlargement >2cm | |
| Positive tourniquet test | | Severe organ involvement |
| Leucopenia | Dropping platelets and rising hematocrit | Hepatitis |
| | | Myocarditis |
| | | Encephalitis |

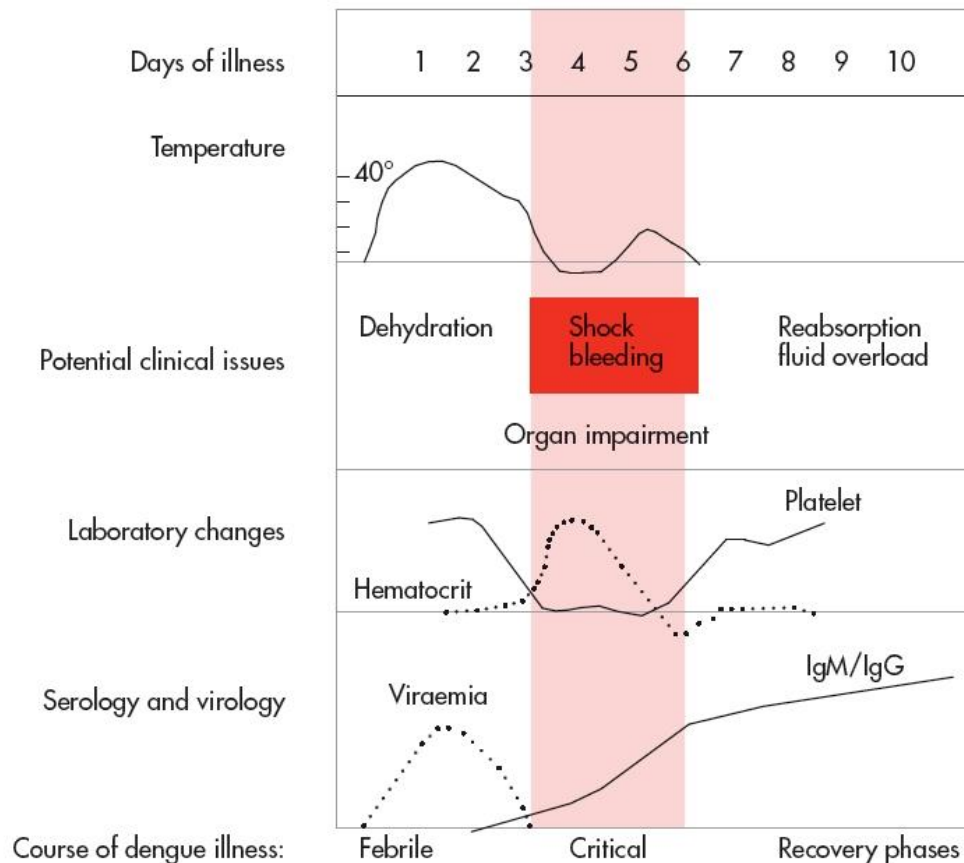
How would you perform the tourniquet test?

Tourniquet test

- The technique may be asked during the discussion
- First measure the systolic and diastolic blood pressures
- Maintain the blood pressure at a point midway between the systolic and diastolic blood pressure for a duration of 5 minutes
- Observe for petechial hemorrhages and draw a 1"x1" square in the area of maximum petechial hemorrhages

- The tourniquet test is positive when the number of petechial hemorrhages within this square exceeds 10

What is the natural history of dengue?



What is the pathogenesis of severe dengue? /DHF

- There are 4 serotypes of the dengue virus
- Primary infection from 1 serotype causes lifelong immunity to that particular serotype
- However during a secondary infection from another serotype these antibodies promote viral replication and increase the viral load
- There is also an exaggerated immune response and a cytokine storm which causes endothelial dysfunction, plasma leakage and platelet destruction and dysfunction

How would you manage a patient with dengue?

- The management of dengue is based on the natural history of the disease. This is shown in the following diagram
- Therefore the management will differ according to the stage of the disease

- The management of dengue will be discussed based on three clinical stages

| Febrile phase | Critical phase | Recovery phase |
|------------------------|--|-----------------------------------|
| Lasts 2-7 days | Is a period of 48h usually from the 3 rd day of fever | General improvement of well being |
| Fever | Fever subsides | Good appetite |
| Flushed | Leakage of fluid occurs | Diuresis |
| Arthralgia and myalgia | Leucopenia | |
| | Rise in hematocrit with drop in platelets | |
| | Can have complications | |

Discuss the management of a case of probable dengue in the febrile phase

- This can be managed on an outpatient basis
- Educate the parents about the warning signs of dengue and when to admit to the hospital
- Ensure good diet and hydration. If food is refused advise the parents to give fluids such as ORS, fruit juice and milk
- Advise them not to give the child any colored substances to eat or drink
- Prescribe paracetamol for the fever (10-15mg/kg, 6 hourly. Maximum daily dose is 60mg/kg)
- Domperidone may be prescribed for severe vomiting

How would you manage a patient in the critical phase of dengue?

- Admit the patient
- Remember that the most intensive management should be done in the critical phase of dengue as this can be complicated by shock and major bleeding manifestations

Monitoring

- Start the dengue monitoring process
- Monitor the following parameters – Pulse rate and volume, blood pressure and pulse pressure, cold peripheries, CRFT
- Frequency of monitoring varies according to the clinical condition
- Laboratory parameters – hematocrit, platelet count (hematocrit may be monitored in ward every 6h)

- Other investigations – ALT (hepatic dysfunction), PT/INR, serum electrolytes, serum albumin (can be low due to the plasma leakage)
- Blood should also be taken for grouping and cross matching

Antipyretics

- Give paracetamol for the fever (dose stated above). A regular dose is not given as this may alter the fever pattern

Fluid management

- The maximum amount of fluid which can be given in the critical phase is calculated by the following equation
- **Maximum amount of fluid = Maintenance + 5% Deficit (50ml/kg) over 48 hours.** This is known as the fluid quota
- Maintenance is calculated as follows

| Body weight (kg) | Maintenance fluid (M) per 24 hours |
|------------------|---|
| < 10 | 100 ml/kg |
| 10-20 | 1000 ml + 50 ml for each kg in excess of 10 |
| > 20 | 1500 ml + 20 ml for each kg in excess of 20 |

Halliday & Segar formula

- This is used as a guide and care is taken not to exceed this. There is also no rule to complete the quota. Fluid is given according to the clinical condition of the patient. Therefore the rate of fluid administration should be reduced as time progresses
- Fluid may be given as oral fluid (ORS, milk, fruit juice) or IV fluids (0.9% saline) or as a combination of the two

The patient is admitted with drowsiness and weak pulses with cold extremities. How would you manage?

- The earliest manifestations of shock would be prolonged CRFT, cold peripheries, rising diastolic blood pressure with normal systolic blood pressure (reduction in the pulse pressure) and tachycardia. This is termed compensated shock
- With time a drop in the systolic blood pressure is noted. This is termed hypotensive shock
- Fluid boluses should be administered to a patient in shock. The amount and rate is as given below
- **Bolus = 10ml/kg over 1 hour.** (Remember that this fluid volume should be deducted from the total fluid quota)
- If the patient does not improve give up to 3 repeat boluses with the last being a colloid (hetastarch)

- The fluid quota is calculated as shown above but is given over 24 hours due to the assumption that the patient has already been in the critical phase for 24 hours.
- **If the patient does not recover after this management consider the possibility of an internal bleed**

Management of the convalescent phase

- The most important aspect of this phase is that there is a risk of fluid overload. Therefore the patient should be assessed for features of fluid overload and pulmonary edema
- Proper fluid management in the critical phase of dengue should prevent severe fluid overload. But if this occurs discontinue fluid supplementation and frusemide 0.1mg/kg may be given IV or oral

What are the other complications of dengue? What is the management?

Hemorrhagic complications

- A major bleed may be suspected in the following clinical scenarios
- Persistent and/ or severe bleeding in an unstable patient regardless of the hematocrit
- Refractory shock
- Hypotensive shock with low hematocrit prior to fluid resuscitation

Management

- The definitive life saving procedure would be to transfuse blood. Fresh packed cells or fresh whole blood are the preparations of choice
- Give packed cells as 10ml/kg
- Continue monitoring the patient

Hepatic encephalopathy

- This can be due to the virus itself or due to paracetamol overdose
- **A,B,C**
- Investigations – AST/ALT, PT/INR, RBS, renal function tests
- Avoid hypoglycaemia
- Lactulose (check dose)
- IV antibiotics – IV metranidazole or IV cefotaxime
- IV N-Acetyl cysteine 75mg/kg 6 hourly if available
- IV vitamin K for 3 consecutive days
- IV ranitidine for gastrointestinal bleeding

As a house officer how would you assess the patient on your daily ward round?

History

Ask for the general condition of the child

Look for any warning signs

Ask about the appetite of the child

Look for the intake of fluid by the child

Look for the urine output – should be more than 0.5ml/kg/h

- **Examination**

PR and volume, CRFT, blood pressure

Signs of fluid overload

- **Management**

Check the adequacy of fluids

Look for the latest reports which are available

When would you decide to discharge the patient?

- Fever free for 48 hours
- Improvement of the clinical status (General well being, appetite, hemodynamic parameters and urine output)
- Out of shock for at least 2 days
- Rising trend in the platelet count (>50,000) with hematocrit responding to IV fluids

Other aspects of management

- Notification
- Education of the parents on elimination of mosquito breeding sites in the immediate vicinity and community

Prolonged fever

Presenting complaint

- Fever
- State the duration

History of the presenting complaint

Description of the fever

Remember that the details should be stated in a definite chronological order

- Describe the onset of the fever and state if there are any specific preceding events
- Describe how the fever was assessed and the value of the height of the fever
- The exact duration of the fever
- Describe the response of the fever to antipyretics and the duration taken for the resolution of the fever
- Describe the dose, route of administration and frequency that the child was given paracetamol
- If there is a recurrence of the fever state the time at which the fever comes back
- Describe the state of the child in between episodes of fever
- Are there associated chills and rigors
- Describe the pattern of fever as intermittent, remittent or continuous (however this is unreliable with the use of antipyretics)

| Fever pattern | Description | Clinical examples |
|---------------------|--|---|
| Intermittent | High spiking fever which reach the baseline | Pyogenic infections TB, lymphoma, systemic onset JIA |
| Remittent | Fluctuating fever which does not reach the baseline | Viral infections, IE, lymphoma |
| Continuous | Sustained fever with little or no fluctuation | Typhoid, typhus |
| Relapsing | Febrile episodes separated by one or more days without fever | Malaria, lymphoma |

The next step is to make a probable diagnosis. The list of differential diagnosis in a patient with prolonged fever is extensive but the common causes should be excluded in the history.

- The main categories of causes of prolonged fever should be dealt with. These are,
- Infective
- Inflammatory
- Connective tissue diseases
- Neoplasms
- Other rare causes

| Category | Diseases | Specific points in the history |
|--------------------------------|---|---|
| Infective Localized | Respiratory tract infections | Cough, sputum, nasal or ear discharge, sore throat |
| | Gastrointestinal infections and localized intra abdominal abscesses | Ask for alteration of bowel habits, recurrent episodes of abdominal pain |
| | Urinary tract infections | Dysuria, frequency, hematuria and other urinary tract symptoms |
| | Infections of the bones and joints | Ask for joint pain and swelling, limping, |
| Generalized | Infective endocarditis | Past history of heart disease, rheumatic fever with evidence of a predisposing event for bacteraemia |
| | IMN | Associated sore throat |
| | TB | Contact history of TB, chronic cough |
| | Typhoid fever | Ask for possible exposure to unhygienic food Initially presents with a slowly rising fever. Then during the 2 nd week of illness classically they have high fever, abdominal distension, "pea soup" diarrhoea, constipation. The 3 rd week of illness is characterized by complications – intestinal perforation |
| | Malaria | Visit to a malarial endemic area |
| | Other zoonotic infections | Contact history with animals |
| Inflammatory | JIA | Ask for a evanescent salmon pink maculopapular rash, associated joint pain and early morning joint stiffness |
| | SLE | History of facial rashes and joint pain |
| | Kawasaki disease | Ask for history of bilateral non purulent conjunctivitis, reddish oral mucosa, erythematous rash and peeling off of the skin, edema of the limbs |
| Neoplastic | Hematological malignancy | Evidence of bleeding, ask for the features of anaemia, history of bone pain, past history of recurrent infections |
| | Other malignancies | |
| Other | Drugs Factitious fever | Drug history |

- Ask for general associated symptoms such as loss of appetite and loss of weight and general malaise

Dietary history

- This is extremely important as children with prolonged fever tend to lose weight and should have a high protein and calorie diet

Past medical history

Past surgical history

Other components of the history

Social history

Describe the following factors in the social history

- General introduction to the family
- Impact of the disease on the child
- Impact of the disease on the parents
- Environmental factors – This is especially relevant if a diagnosis of typhoid fever is suspected
- Support available
- Education of the parents regarding the condition and future expectations
- Psychological state of the parents

Examination

General assessment of the patient

- General appearance of the child, activity and growth parameters. Plot the growth parameters in a centile chart

General examination

Skin

Examine the skin for lesions. The important skin lesions and their associations are given below

| Skin lesions | Associated diseases |
|--|---------------------|
| Malar rash | SLE |
| Salmon pink rash | JIA |
| Petichial rash, janeway lesions, osler's nodes | IE |
| Palpable purpuric rashes | Vasculitis |
| Erythema nodosum | TB, IBD, SLE |
| Eschar +/- erythematous rash | Typhus |
| Erythematous rash and desquamation | Kawasaki disease |

Head and face and neck

- Examine the eyes for conjunctivitis – non purulent conjunctivitis in **Kawasaki disease**
- Examine the fundi – **miliary tubercles in TB, other diseases can also have manifestations in the fundi – toxoplasmosis, roth spots in leukemia, Vasculitis**
- Examine the throat – pharyngitis and tonsillitis
- Examine the oral cavity and tongue – **reddened lips, strawberry tongue in Kawasaki disease**
- Look for palatal petichiae, tonsillar exudates in **IMN**
- Examine for lymphadenopathy – Generalized lymphadenopathy – **IMN, miliary TB, hematological malignancies, JIA, SLE**
Asymmetrical cervical lymphadenopathy in **Kawasaki disease**
- Examine the hands for clubbing and other stigmata of infective endocarditis

Cardiovascular system

- Examine the heart for murmurs – **IE**

Respiratory system

- Examine for the BCG scar
- Examine for evidence of consolidation/ pleural effusions

Abdomen

- Look for hepatosplenomegaly

- Any other palpable masses in the abdomen – **para aortic lymph nodes in lymphoma, Wilm’s tumor, neuroblastoma**

Musculoskeletal system

- Bone tenderness – **osteomyelitis, leukemia**

Discussion

What is the definition of PUO?

- No strict definition as in adults but should be suspected if the child has fever for more than 1 week

How would you investigate a patient with PUO?

- **FBC**
This is an important investigation. Look for the following
Anaemia – Is associated with chronic infections and inflammatory and connective tissue disorders
Pancytopenia – Is evidence of bone marrow suppression which may occur in leukemia
Thrombocytosis – Is known to occur in inflammatory diseases and Kawasaki disease
Look at the white cell count and the predominant cell types
- **Blood picture**
Look for any atypical cells which may suggest leukemia
- **Acute phase reactants**
ESR, CRP, serum ferritin
- **Blood culture**
- **Other investigations – renal, hepatic**
- **Urine full report and urine culture**
- CXR, mantoux test
- **Echocardiogram**
To look for vegetations, valvular dysfunction
- **Other investigations may be necessary – bone marrow biopsy**

Infectious diseases presenting with PUO

Infectious mononucleosis

Introduction

- Is caused by EBV
- Presents with prolonged fever and sore throat
- Examination may reveal cervical or generalized lymphadenopathy, palatal petichiae, membranous tonsillitis and splenomegaly
- Complications – splenic rupture, GBS

Investigations

- **FBC and blood picture** – may show an absolute lymphocytosis. The blood picture will show atypical lymphocytes
- **Paul- Bunnell test and monospot test** – Look for heterophile antibodies which agglutinate with sheep or horse blood. These have a low sensitivity
- **Specific EBV antibody test** – Done at MRI

Management

- This is usually supportive

Infective endocarditis

Infective endocarditis

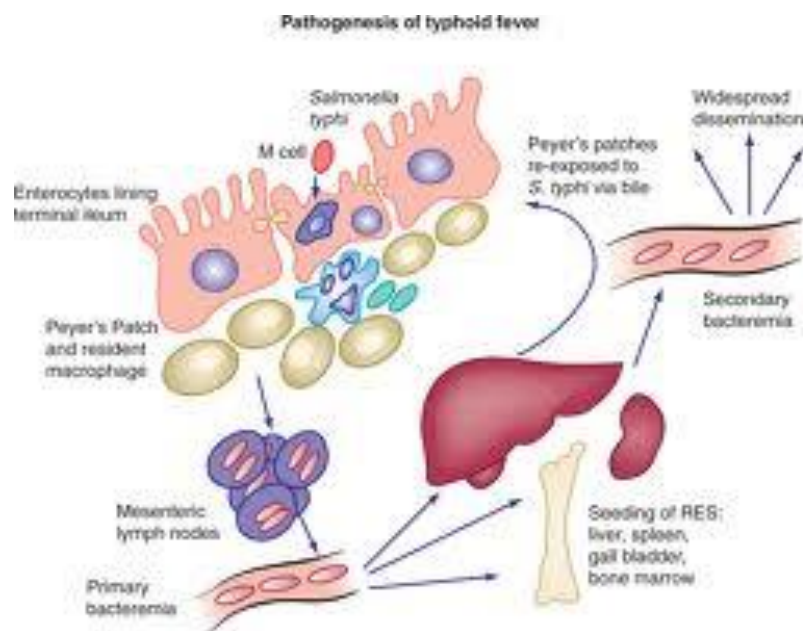
Diagnosis

- Diagnosis is based on the modified Duke's criteria
- **Major criteria**
 - Positive blood cultures
 - Evidence of endocarditis on echocardiography – oscillating intracardiac mass, new valvular regurgitation, abscess
- **Minor criteria**
 - Predisposing conditions
 - Fever
 - Embolic vascular signs
 - Immunological phenomena
 - Microbiological evidence not meeting the major criteria
- **Organisms causing IE**
 - Viridans group of streptococci*
 - Staphylococcus aureus*

Principles of management

- After obtaining the blood cultures the next step is to start high doses of empirical antibiotic therapy via the IV route
- This is done as high blood concentrations of the drug is required for penetration into the vegetations
- IV benzyl penicillin and gentamicin is the preferred combination as initial empirical therapy
- After receiving the reports of the blood culture and ABST antibiotics may need to be altered
- Continue antibiotics for about 4 -6 weeks
- Monitor the patient for the complications of the disease
Severe valvular dysfunction and heart failure
Myocardial abscesses
Systemic emboli
- Look for the following on the daily ward round
General condition of the child
Look at the fever chart and the response to antibiotics
Auscultate the murmur and note any change in the character or intensity
Look for features of heart failure
- Prophylaxis, proper dental care and oral hygiene

Typhoid fever



Diagnosis

- Presents with prolonged fever and the classical pattern of symptoms mentioned above in the section on history taking
- Incubation period 10-21 days

| Period of illness | Clinical features |
|-------------------|--|
| Week 1 | Non specific symptoms, headache, malaise and a step ladder type fever. The child may also have constipation |
| Week 2 | More ill looking, high temperature, relative bradycardia, rose spots on the abdomen, abdominal distension and splenomegaly |
| Week 3 | Continuous high fever, extremely ill, pea soup diarrhoea, systemic involvement |
| Week 4 | Begins gradual improvement. May develop a carrier state |

- **FBC** – mild leukocytosis may be seen initially but later leucopenia and neutropenia predominates
- **Blood culture**
- **SAT** – tests the antibodies against the H and O antigens of *Salmonella typhi*. Lacks sensitivity in endemic countries

Principles of management

Antibiotic therapy

- If the child is systemically ill 3rd generation cephalosporins are the drug of choice
- Other drugs
 - Amoxicillin
 - Chloramphenicol
 - Azithromycin
 - Co- trimoxazole
- Monitor for and manage complications

TB in children

Diagnosis

Is based on the following aspects

- History – PUO, chronic cough, FTT, contact history of TB
- Examination
- **Tuberculin skin testing**
5 TU (0.1ml) of the tuberculin PPD is injected intradermally into the skin of the forearm (exact site)
Inspection is done after 48-72h

Interpretation

Diameter of induration of ≥ 5 mm is considered positive in:

HIV-infected children

Severely malnourished children (with clinical evidence of marasmus or kwashiorkor).

Diameter of induration of ≥ 10 mm is considered positive in:

All other children (whether or not they have received BCG vaccination)

- **Bacteriological confirmation wherever possible**
This is extremely difficult to do in children as they cannot expectorate sputum but may be considered in children
- **Other investigations relevant to pulmonary or extra pulmonary TB**
CXR
Persistent opacification of the lung with enlarged hilar lymph nodes
Pleural effusion
Miliary shadowing
Apical infiltrates with cavitation are rarely seen in children
Other
- **HIV testing**

Drugs used in the treatment of TB in children

- Isoniazid
- Rifampicin
- Pyrizinamide
- Streptomycin
- Ethambutol

Cyanotic heart disease in children

Discussion of the management of cyanotic heart disease in children will focus on the following aspects

- Diagnosis and initial management (neonatal period)
- Further management
- Management of emergencies
- Management of other associated issues
 - Failure to thrive
 - Developmental delay
 - Socio economic issues

Diagnosis

- Presentation is usually with neonatal cyanosis. It is important to identify the other causes of cyanosis in neonates and how to differentiate between them

| Category | Important clinical examples | Key features |
|---|---|--|
| Central or peripheral nervous system hypoventilation | Birth asphyxia ICH Drugs Diaphragmatic palsy | Irregular, slow and weak respiration Associated CNS symptoms and signs |
| Respiratory disease | Upper or lower airway disease Upper airway Choanal atresia Congenital anomalies of the upper airway Lower airway RDS TTN Pneumothorax Infection Diaphragmatic hernia | Vigorous and labored respiration with tachypnoea Positive hyperoxia test Rise in the arterial partial pressure of oxygen after administration of 100% oxygen (>150mmHg) |
| Cardiac disease | With decreased pulmonary flow TOF Abnormal connections and mixing TGA TAPVD | Vigorous and labored respiration with tachypnoea Associated cardiac murmur (is not always present) Negative hyperoxia test |
| Other | Methhaemoglobinaemia Sepsis | |

- Investigations
2D echocardiography
CXR

Initial management principles

- **ABC** and adequate resuscitation in an optimal temperature
- Proper hydration of the baby
- Correction of acid base abnormalities, hypoglycaemia and electrolyte imbalance
- Administration of a prostaglandin infusion
- Transport the patient to a specialized center

Further management

- This will depend on the diagnosis made and the associated complications

Tetralogy of fallot

Background knowledge of the anatomy and pathophysiology

- Is due to abnormal deviation of the septum than separates the aortic and pulmonary outflow tracts
- Has 4 basic anatomical abnormalities. These are pulmonary infundibular stenosis, right ventricular hypertrophy overriding aorta and VSD
- The pulmonary infundibular stenosis causes right ventricular outflow tract obstruction and the severity of this determines the symptoms
- When the right ventricle contracts against the pulmonary stenosis blood is shunted across the VSD into the aorta

Diagnosis

Clinical – See short case on TOF

- Usually cyanosis is not present at birth unless the pulmonary stenosis is very severe
- With age there is increased RVOT obstruction and increasing cyanosis
- Central cyanosis, clubbing, ejection systolic murmur in the left mid sternal edge and soft P2

Investigations

- **CXR** -“Boot” shaped heart with pulmonary oligoemia
- **ECG** – Features of right ventricular hypertrophy
- **Echo** – For the confirmation of the diagnosis
- **Cardiac catheterization** – This will show the anatomy of the lesion and the state of the pulmonary arteries which is important in surgical intervention

Management

- As stated above TOF usually does not present with cyanosis in the neonatal period unless the degree of pulmonary stenosis is severe
- If there is neonatal cyanosis manage as stated above
- There are two options in the further management of these babies. These are,
Creation of a shunt from the subclavian artery to the pulmonary artery (modified Blalock – Tassing shunt)
Total correction
- Others should be carefully followed up and a date given for corrective surgery
- Complications may occur in these children
- **Hypercyanotic spells**
Place the child in the knee chest position
Administer high flow oxygen
Administer IV morphine – maximum dose 0.2mg/kg
Correction of metabolic acidosis with IV sodium bicarbonate

For resistant spells

IV propranolol 0.1 mg/kg

After management a date for early surgery should be given. The child is also given oral propranolol 0.5 – 1 mg/kg 6 hourly for prevention of hypercyanotic spells

Cerebral thrombosis

Cerebral abscess

Infective endocarditis

Transposition of great vessels

Background anatomy and pathophysiology

- In this lesion the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle
- Therefore unsaturated blood from the right ventricle reaches the systemic circulation via the aorta
- In order for these newborns to survive there should be a connection between the two sides of the heart. This may be via a PFO, PDA or VSD

Diagnosis

- Cyanosis and tachypnoea are observed in the first few hours of life
- Clinical signs are minimal on auscultation but may have a single, loud second sound. A murmur may also be audible if there is an associated VSD

- Echocardiogram is the investigation of choice for the diagnosis

Management

- Is an emergency
- Manage as given above. Especially the infusion of PG E1 is a critical component in the management as it keeps the ductus arteriosus open
- If there is poor response to the PG infusion an emergency balloon atrial septostomy should be performed
- Definitive surgery is by the arterial switch operation which should be performed within 14 days of life

The situation in Sri Lanka regarding the management of congenital heart disease

- Limited resources and long waiting lists
- Check social support available and funding

Recurrent wheezing in childhood – Asthma

Key points in the history – 5 key points to describe

1. Describe the present episode in detail

- Describe the onset, duration and progression of the symptoms
- Ask for any preceding triggering factors
- Describe what the mother did at home
- Assess the clinical severity of the episode and what was done in hospital

2. Describe the past history and the progression up to now

Highlight the following points and use a time line for the important events

- The first episode
- Acute exacerbations and hospital admissions
- Treatment given and the compliance
- Side effects of the medication given and follow up

3. Describe the present state of the disease

4. Exclude D/D's of recurrent wheeze and establish the probable diagnosis of asthma

| Cause | Important points in the history |
|---|---|
| Bronchial asthma | <p>Symptom pattern (most of these will have been described above)</p> <p>Intermittent symptoms (the child will be well in between episodes)</p> <p>Diurnal variation of symptoms may be present</p> <p>Definite trigger factors for the episodes and good response to medication</p> <p>Ask for family history of atopy and asthma</p> |
| Structural anomalies/ congenital lesions of the respiratory tract | This will be excluded as the onset of symptoms is later on in life |
| Tuberculosis | Ask for a contact history of TB |
| Interstitial lung disease | Long standing history of symptoms, failure to thrive |
| Heart failure | Ask for past history of cardiac disease, reduced exercise tolerance, orthopnoea (in an older child) |
| Gastro esophageal reflux disease | Ask if the symptoms are associated with meals and if there is associated regurgitation |
| Recurrent aspiration | Risk factors for aspiration |
| Foreign body inhalation | |
| Rare causes – cystic fibrosis, ciliary dyskinesia, immunodeficiency | Recurrent lower respiratory tract infections, chronic sinus infections, failure to thrive |

5. Get a very detailed social history as this is the most important component of the history

| | |
|--|--|
| General introduction to the family | |
| Impact on the child | Playing, amount of school missed, diet, bathing |
| Impact on the parents | Socio economic impact of the disease, impact of frequent hospital stays |
| Impact on the siblings | |
| Impact on normal family activities | Can the family do what they did earlier |
| Environment | <p>Describe the layout of the house</p> <p>Describe the surrounding environment Main roads, dirt and dust, factories</p> <p>Describe the house</p> <ul style="list-style-type: none"> • The floors and how often they are swept and mopped • Windows and available ventilation in the house, also ask how many people sleep in one room • Bed sheets, pillow cases, mattresses and how often they are changed • Ask how often the carpets in the house are dusted • Cooking fumes • Use of mosquito coils • Smoking in the house • Pets • Soft toys of the child • Describe the environment of the school |
| Support available | <p>Family support</p> <p>Extended family support</p> <p>Medical facilities available</p> |
| Education of the parents | <ul style="list-style-type: none"> • Drugs and when to use them • Difference between a preventer and a reliever medication • Inhaler devices and how to use them • Myths regarding asthma • How to recognize an acute exacerbation of asthma and what to do • When to bring the child to the hospital |
| Psychological state and expectations of the parents | |

Examination

General examination

- **Anthropometry**

Plot the weight and height of the child on a centile chart. Ask the mother for the CHDR of the child

This is important as FTT could indicate an alternate diagnosis to bronchial asthma. Also look for growth faltering as this could indicate steroid toxicity

- Look for other evidence of steroid toxicity - Cushingoid features
- Look for cataract
- Examine nose – nasal polyps
- Examine the mouth and throat – oral thrush
- Examine for cervical lymphadenopathy
- Look for clubbing – if present could indicate an alternate diagnosis
- Look for the BCG scar
- Ankle edema
- Skin for rashes – atopic eczema

Respiratory system

- Look for evidence of respiratory distress
- Look for the evidence for chronic hyperinflation of the lungs
Increased antero posterior diameter – barrel shaped chest
Harrison sulcus
Impaired liver and cardiac dullness
Liver pushed down
- Auscultate for ronchi and crepitations

Never forget to examine the inhaler technique of the child

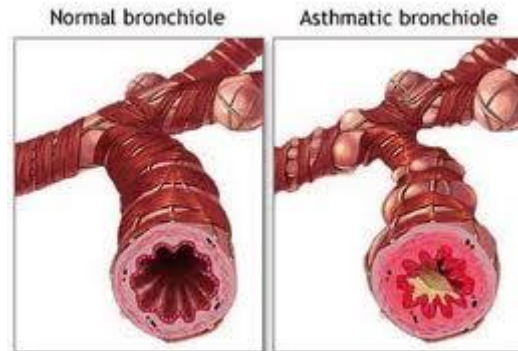


Bronchial asthma

Discussion

What is bronchial asthma?

- **Definition**
Asthma is a chronic inflammatory condition of the airways which is characterized by episodic reversible airway obstruction and airway hyper-responsiveness
- The diagnosis of bronchial asthma is a primarily clinical diagnosis in children based on the typical clinical features and the good response of these symptoms to bronchodilators
- Exclude other alternate diagnoses
- Other objective tests can also be carried out if the diagnosis is uncertain – FEV1/FVC ratio and reversibility of PEFR to bronchodilators



How severe is this child's asthma?

| Category | Days with symptoms | Nights with symptoms |
|---------------------|------------------------------|-----------------------|
| Mild intermittent | 2 or less per week | Less than 2 per month |
| Mild persistent | > 2 per week but < 1 per day | > 2 per month |
| Moderate persistent | Daily | > 1 per week |
| Severe persistent | Continual | Frequent |

If you were the house officer on admission how would you manage an acute exacerbation of asthma?

Management in the hospital

Acute severe asthma

- Focused history and examination
- **Recognition of acute severe asthma and life threatening asthma is the most important point as a house officer**

| | Acute severe asthma | Life threatening asthma |
|-------------|--|----------------------------------|
| History | Breathless at rest, cannot complete sentences in one breath | Drowsy or confused patient |
| Examination | Features of respiratory distress (Tachypnoea, use of accessory muscles of respiration, decreased saturation) | May have poor respiratory effort |
| | Ronchi and crepts on auscultation | Silent chest |
| | Tachycardia | Bradycardia |

- Place the child in the most comfortable position
- Give high flow oxygen via face mask at 6-8 litres/min
- Oxygen driven nebulization with salbutamol (0.5ml in children less than 5 years and 1ml in children more than 5 years) with 1.5 ml of normal saline
- An equivalent effect may be achieved with 10 puffs via the spacer device
- Reassess in 20 minutes
- Combine with ipratropium bromide 0.25mg
- Give IV hydrocortisone 4mg/kg or oral prednisilone 2mg/kg at this stage
- If the child is improving continue the nebulizations every 1-4 hours and oral steroids for 3-5 days

If there is no response

- Continue nebulization every 20 to 30 minutes.
- **IV aminophylline**
Give a bolus if not already on oral theophylline 5mg/kg in 2ml/kg normal saline over 30 minutes and follow up with an infusion of 1mg/kg/h
Connect the child to a cardiac monitor as aminophylline can cause SVT
- Contact your seniors
- **Other drugs**
IV salbutamol – requires potassium monitoring and continuous cardiac monitoring
IV Magnesium sulphate
IM/SC adrenaline – 0.01ml/kg of 1:1000

Other aspects of management

- IV fluids at 2/3 of maintenance
- Antibiotics

With improvement

- Wean the child off the nebulizations and recommence the usual inhaler medication. Consider the cause for the acute exacerbation

Once the child has recovered from the acute episode what will be the subsequent management?

This includes the following themes

- Control of factors contributing to asthma severity
- Patient education
- Asthma pharmacotherapy
- Regular assessment and follow up

Education of the parents

- Basic facts about asthma
- Importance of compliance to the medication and roles of the various medication
- Skills development in the use of the various devices and their care (revise the technique of use of these devices as it will be asked in the exam)
- Monitoring response by the use of a symptom diary
- Environmental modifications of asthma
- How to recognize an acute exacerbation of asthma and when to seek treatment

Control of factors which contribute to asthma severity

| Factors | Control measures |
|--------------------|---|
| Animal dander | Keep pets away from the child |
| Dust mite | <ul style="list-style-type: none">• Do frequent wet mopping of the floors and try to avoid dry sweeping while the child is in the house• Change pillow cases and bed sheets regularly• Sun drying the mattresses• Clean carpets and curtains regularly• Do not give the child any soft toys• Clean the fans frequently |
| Indoor mold | Adequate ventilation, avoid seepage of water through the roofs and walls |
| Cockroaches | Control |
| Chemical irritants | Stop smoking Avoid lighting of mosquito coils |

| | |
|------|---|
| | Keep cooking fumes to a minimum |
| Food | No restriction of the diet is made including cool drinks and ice cream, but asthma is known to be precipitated by some food colouring |

Asthma pharmacotherapy

- This has 2 aspects. These are
Long term management
Management of exacerbations of asthma
- The goals of pharmacotherapy are as follows
Minimal or no chronic symptoms at day or night
Minimal or no exacerbations
No limitations on activities
Minimal adverse effects of medication
- There are two categories of drugs which are used in the management of asthma. These are preventer medication and reliever medication

Available drugs

| Drug class | Name of the drug |
|------------------------|--|
| Beta 2 agonists | |
| Short acting | Salbutamol, terbutaline |
| Long acting | Salmeterol |
| Corticosteroids | Beclomethasone, fluticasone, budesonide |
| LTRA | Montelukast |

Stepwise therapy

| Step | Drugs used |
|----------------------------------|--|
| Step 1 Mild intermittent BA | Short acting inhaled beta -2 agonists – Salbutamol No daily medication |
| Step 2 Mild persistent BA | Preferred treatment Low dose inhaled corticosteroids Alternative treatment Sustained release theophylline LTRA |
| Step 3 Moderate persistent BA | Preferred treatment Medium dose inhaled corticosteroids OR Low dose inhaled corticosteroids and long acting |

| | |
|--|---|
| | <p>beta-2 agonists</p> <p>Alternative treatment Low dose inhaled corticosteroids and either theophylline or LTRA</p> <p>In recurrent episodes of severe exacerbations Medium dose inhaled corticosteroids and long acting beta-2 agonists</p> |
| <p>Step 4 Severe persistent BA</p> | <p>Preferred treatment High dose inhaled steroids and long acting beta-2 agonists Consider oral steroids</p> |

- At each step the other aspects of the management plan should be reinforced and short acting beta -2 agonists may be used for acute episodes

Indications for reliever medications in bronchial asthma

- Chronic persistent asthma
- After an episode of life threatening asthma
- Recent increase in the severity or frequency of acute exacerbations
- Nocturnal asthma which disturbs the child from sleep
- Frequent episodic asthma which interferes with normal life
- Severe exercise induced asthma
- Inaccessibility of medical care

Drug delivery devices in asthma

Selection of an appropriate device

| Age of the child | Suitable device |
|-------------------|---|
| Less than 2 years | Baby haler |
| 2-5 years | MDI with a spacer device (with a face mask up to 3 years) |
| More than 5 years | MDI with spacer/DPI |
| More than 8 years | MDI alone |

Use of an MDI with a spacer device

- Revise and practice the technique of the device. The most commonly asked will be the use of the mask spacer device.
- Remember to ask the patient to rinse the mouth after using a corticosteroid inhaler

Regular assessment and follow up

The following should be assessed at a routine asthma follow up

- Signs and symptoms of asthma
- Pulmonary function
- Quality of life and functional status
- Acute exacerbations during this period
- Adequacy of the management
 - Pharmacotherapy
 - Consider step up or step down every 3 months
 - Environmental modifications
- Assess for the side effects of the medication – especially steroids
 - Assessment of the weight and height
 - Measure the blood pressure
 - Encourage exercise
 - Adequate dietary calcium supplementation
 - Ophthalmological assessment

Now apply the above management principles to the problem list of the child. After the history and examination ask yourself the following questions

- Is this asthma?
- How is the control of asthma?
- Is there any indication to alter the medication?
- Are there any side effects?
- Are there any environmental risk factors?

If the child's asthma is poorly controlled what will you do?

- Are the drug and the dose adequate?
- Is there proper compliance?
- Are there any triggering factors in the environment which have not been corrected?
- Is diagnosis correct?

Pneumonia in children

Discussion

The first important point which will be asked in the discussion is how the clinical diagnosis of pneumonia was reached. This is based on the history and examination. Follow the points given below

How do you make a clinical diagnosis?

History

- Presents with fever and respiratory tract symptoms
- Classification of pneumonia should also be made based on the history into
- Community acquired pneumonia
- Hospital acquired pneumonia
- Pneumonia in the immunocompromised

Examination

- Febrile
- Tachypnoea – This is the most sensitive and specific sign of pneumonia in children
Definition (WHO)

| Age | Respiratory rate |
|----------------------|----------------------------|
| < 2 months | Over 60 breaths per minute |
| 2 months – 12 months | Over 50 breaths per minute |
| 12 months to 5 years | Over 40 breaths per minute |
| More than 5 years | Over 20 breaths per minute |

- Features of respiratory distress may be present such as chest wall recessions, use of accessory muscles of respiration, cyanosis and grunting
- Examination of the chest can reveal features of a lobar consolidation, pleural effusion and other diffuse respiratory signs

What are the investigations you would do?

- **Blood investigations**
FBC
Acute phase reactants
Serum electrolytes
- **Microbiological investigations**
Blood culture
Sputum culture – Difficult to obtain in most children

Pleural fluid analysis if significant pleural effusion present

- **Radiological investigations**
CXR

How do you arrive at a possible etiological diagnosis?

- This is made on the history, examination and investigations and is an important aspect to guide the treatment
- The age of the child is a good indicator to the aetiology

| Age | Microorganisms |
|-------------------|--|
| Neonates | Group B Streptococcus, E. coli, Klebsiella, Listeria |
| 1 months – 1 year | Viral RSV, parainfluenza Bacterial Streptococcus pneumoniae, Staphylococcus aureus Chlamydia |
| 1 – 5 years | Viral RSV, parainfluenza, influenza, adenovirus Bacterial Streptococcus pneumoniae, Staphylococcus aureus, Haemophilus influenzae b, Mycoplasma |
| > 5 years | Bacterial Streptococcus pneumoniae, Mycoplasma, Chlamydia |

- Remember the following basic points regarding the association of the age of the child and the aetiology of the pneumonia
- In infants and children less than 5 years of age pneumonia is commonly caused by viruses
- In children more than 5 years of age Streptococcus pneumoniae is the most common bacterial cause of pneumonia
- The clinical pattern of the illness and the investigation findings are also important in thinking of a possible aetiology

| | Viral | Bacterial | Atypical organisms (Mycoplasma) |
|-----------------|---|---|--|
| Clinical | Low grade fever (<38.5) Respiratory rate normal or slightly raised Wheezing Marked chest wall recessions | High grade fever (>38.5) Respiratory rate high No wheezing Chest wall recessions | Low grade fever Associated wheeze Prolonged disease course Prominent headache, arthralgia, myalgia Extra pulmonary |

| | Hyperinflation | | manifestations |
|-----------------------|--|---|--|
| Investigations | Usually no neutrophill leukocytosis CRP | Neutrophill leukocytosis >15,000 WBC CRP elevated > 35 to 60mg/L | Special investigations Serology Cold agglutinin test |
| Chest X ray | Hyperinflation and lobar collapse | Consolidation, pleural effusion Special findings may also indicate the probable aetiology – Pneumatocoeles, cavitation (S. aureus, klebsiella) | Reticulonodular opacification of the lower lobe Hilar lymphadenopathy Interstitial infiltrates |

How do you assess the severity of pneumonia?

Next you will be asked to assess the severity of the pneumonia based on the clinical assessment and the investigations

| | Mild | Severe |
|----------------|--|--|
| Infants | Temperature <38.5°C RR <50 breaths/min Mild recession Taking full feeds | Temperature >38.5°C RR >70 breaths/min Moderate to severe recession Nasal flaring Cyanosis Intermittent apnoea Grunting respiration Not feeding |
| Older children | Temperature <38.5°C RR <50 breaths/min Mild breathlessness No vomiting | Temperature >38.5°C RR >50 breaths/min Severe difficulty in breathing Nasal flaring Cyanosis Grunting respiration Signs of dehydration |

Diagnosis

The diagnosis involves the following details

- Clinical diagnosis of pneumonia
- Probable aetiological diagnosis

- Assessment of the severity of pneumonia

How would you manage this patient?

General management

- Assess the ABC
- Measure the oxygen saturation with the use of a pulse oxymeter
- Oxygen therapy should be considered if the saturation is less than 92%
- Obtain IV access and take blood for investigations – FBC, CRP, blood culture
- Consider IV fluids if the patient cannot take orally
Correct any dehydration/ deficits
Put the child on an IV drip at 2/3 of maintenance
- Manage fever and pain with paracetamol
- Start a monitoring chart. Include the PR, RR, BP, oxygen saturation and the respiratory signs and symptoms of the child
- Feeding of the child – try to avoid insertion of an NG tube for feeding as this can compromise the airway further

Antibiotic management

- There are several aspects which should be considered in the antibiotic management
- Whether to start antibiotics/not
In a patient with a clinical diagnosis of pneumonia empirical antibiotics should be commenced. However antibiotics should not be used in children with mild lower respiratory tract infections
- Choice of antibiotics

| Age | Empirical antibiotics |
|--|--|
| Children less than 5 years (excluding neonates) | Oral amoxicillin (if the child is not ill and can take orally) IV cephalosporins (used presently in the wards) IV ampicillin |
| Children > 5 years | Penicillin/ cephalosporins, Macrolides - erythromycin, clarithromycin (these can be used in combination) |

- Route of administration
IV should be considered if the child is extremely ill and refusing to take oral medication
- Duration of treatment
Usually treatment carries on for about 7-10 days
- With improvement the IV antibiotics may be switched from IV to the oral route

What would you look for in this patient on your daily ward round?

- Look at the general condition of the child
- Examine the respiratory system of the child
- Look at the monitoring chart
- Look at the latest investigations

Complications

Poor clinical response – the clinical response to treatment should take no more than 48 – 72 hours. If the child is still unwell after this period of time consider the following factors

Is the child receiving the appropriate dose of the appropriate antibiotics?

Assess the compliance to the medication – check if the child is receiving the antibiotics or has the child vomited the medication

Is the diagnosis correct?

Has the child developed any other complications of pneumonia – effusion, empyema, metastatic spread

Are there any host factors predisposing to the poor response?

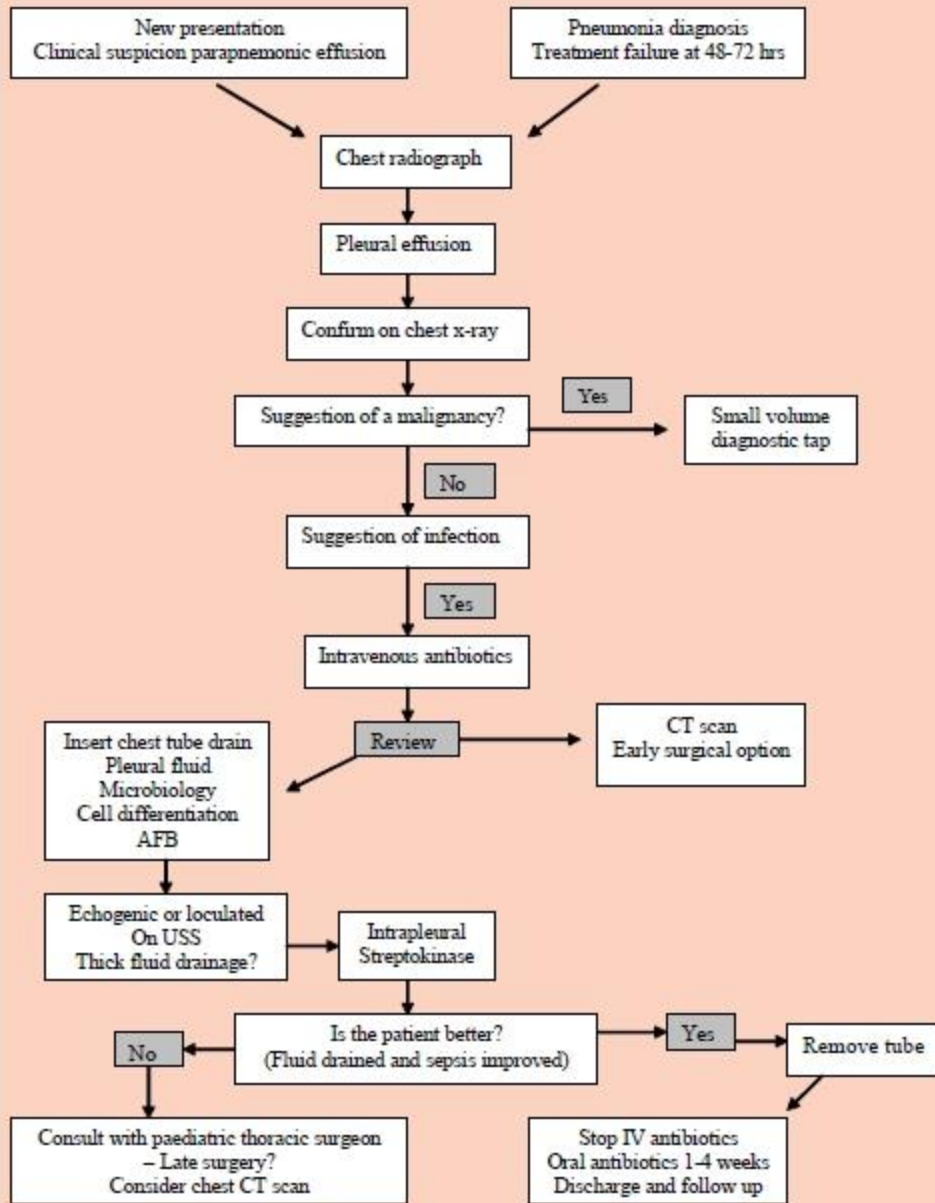
Other causes for the continuing fever

Assessment and further management

- Do a chest x ray and assess the patient
Look for pleural effusion, worsening of the infiltrates, foreign bodies, and features suggestive of atypical pneumonia
- If the chest x ray is unremarkable a septic screen may be indicated to look for disseminated infection
- Review the antibiotic therapy
- Second line investigations may be indicated if the problems persist

Management of pleural effusion

The scheme for management of pleural infection in children



Blood and mucus diarrhoea

Key points in the history

Presenting complaint

- Loose stools
- State the duration

History of the presenting complaint

Describe the key features of the diarrhoea

- Onset
- Duration
- Describe the characteristics of the stool – watery, mucoid or associated blood and mucus
- Describe the associated factors - Fever, nausea and vomiting with abdominal pain
- Try to quantify the amount
- Describe what the mother did at home
- Describe the progression of symptoms over time
- Describe what was done at the hospital

Exclude other conditions that can present with diarrhoea

| Category | Disease | Important points in the history |
|-------------------|---------------------------|--|
| Infections | Dengue | Ask about arthralgia, myalgia, headache and retro-orbital pain and bleeding manifestations |
| | Meningitis | Associated headache and vomiting, photophobia, irritability and altered behaviour |
| Surgical | Intussusception | Presents with blood and mucus diarrhoea (classic “red currant jelly” stools) Ask for screaming attacks in the child |
| | Acute appendicitis | Abdominal pain (periumbilical) |
| | IBD | Previous episodes of blood and mucus diarrhoea |

Ask for the risk factors of an infectious cause for diarrhoea

| | |
|-----------------------------|--|
| External factors | Was there any consumption of food from outside? |
| Maternal factors | Ask about the personal hygiene of the mother Does she wash hands after going to the toilet? Does she cut her nails? Food preparation Does she wash her hands before preparing food? Does she clean the vegetables and green leaves properly? Are the cooking utensils cleaned regularly? Is the food covered adequately after preparation? Where is it stored? Does the mother give bottle feeds? If so ask if she boils the bottles. Ask on the preparation of the formula milk Do they use boiled water? |
| Child factors | Hygiene of the child Does the child play with sand or dirt? Other playmates |
| Other family members | Personal hygiene in the other family members may also be relevant, especially of those who come in contact with the child |
| Environment | Give a brief account of any environmental risk factors for diarrhoea around the house – garbage collections, flooding, use of night soil as fertilizer |

Ask for the complications of diarrhoea

- Ask for the urine output of the child and for features of lethargy and drowsiness – Dehydration
- Seizures
- Can be due to the following causes – associated febrile convulsion, electrolyte imbalances, hypoglycaemia, HUS, shigella encephalopathy

Past medical history

- Ask for past history of episodes of diarrhoea/ dysentery

Other routine aspects of the history

Social history

- Most of this has been described in the history of the presenting complaint but present the social history in the usual order
- Give special emphasis to the education level of the mother and her knowledge of diarrhoea, preparation and administration of ORS

Examination

- The key point of the examination is to look for evidence of dehydration. Look at the following table

| | No dehydration | Some dehydration | Severe dehydration |
|-------------------|-------------------|-------------------------|------------------------|
| General condition | Well and alert | Restless and irritable | Lethargic, unconscious |
| Eyes | Normal | Sunken | Very sunken and dry |
| Tears | Present | Absent | Absent |
| Mouth and tongue | Moist | Dry | Very sunken and dry |
| Thirst | Thirsty | Thirsty, drinks eagerly | Drinks poorly |
| Skin pinch | Goes back quickly | Goes back slowly | Goes back very slowly |

- Examination of the vital signs is also extremely important as the child may present in shock
- Abdomen
Look for a distended abdomen – Intestinal obstruction, paralytic ileus due to hypokalemia, gas forming organisms
- Palpable masses – Intussusception

Discussion

Dysentery

What are the causes of dysentery?

| Bacillary dysentery | Amoebic dysentery |
|---|---|
| Is the most common cause of dysentery More faecal matter with less amount of blood | Rare Small amounts of faecal matter with larger amounts of blood |

What are the causes of bacillary dysentery?

- *Shigella* – *Shigella dysenteriae*, *Shigella sonnei*, *Shigella flexneri*, *Shigella boydii*
- *Escherichia coli*- Enteroinvasive and enteropathogenic
- *Campylobacter jejuni*

What are the differences between *Shigella dysenteriae* and *Shigella flexneri*?

- Compared to *Shigella flexneri*, *shigella dysenteriae* is highly infective and requires a smaller infective dose. However it survives for only a short period of time in the environment

How would you manage this patient?

Initial

- Admit the patient to the isolation room of the ward
- Obtain samples for stool smear and culture

Fluid management

- Follow the basic principles
- Total fluid requirement = Correction of deficit + maintenance + correction of ongoing losses
- **Correction of deficit**

| Degree of dehydration | Deficit | Replacement |
|-----------------------|---------------|---|
| Some dehydration | 50 – 100ml/kg | ORS 75 ml/kg over 4 hours |
| Severe dehydration | >100ml/kg | IV fluids 100ml/kg (preferred hartmann's) Age <12 months 30ml/kg in 1hour and 70ml/kg in 5 hours Age >12months 30ml/kg in ½ hour and 70ml/kg in 2 and ½ hours |
| Shock | >200ml/kg | Give boluses at 10ml/kg over 20 minutes |

- **Maintenance fluid calculation**
This is based on the Halliday and Segar formula
- **Correction of ongoing losses**
Give 50-100ml of ORS for each liquid stool or vomitus

Antibiotic therapy

- This is based on the local antibiotic sensitivity patterns. Several antibiotic options are available for the management. The patient should be given empirical antibiotic therapy should be based on the local sensitivity patterns. At present the drug of choice is furozolidone **2mg/kg/dose 6 hourly for 5 days**
- A common side effect is darkening of the urine. The mother should be warned of this

Dietary management

- Continue breastfeeding if the child is on breast feeding
- A special diarrhoea diet is given in the wards
- Rice kanjee and red rice kanjee – Prebiotic oligosaccharides
- Anamalu – has a property of forming the stools
- Yoghurt – Is a probiotic (living organisms that are colonizing organisms in the gut and prevent the invasion of pathogenic organisms)
- Rusk
- Lime juice

Zn therapy

- Has been shown to reduce the severity, duration and recurrences of diarrhoea. Give Zn 10-20mg/d for 10-14 days

Monitoring of the patient

Complications of Shigella

- **Local**
Intestinal perforation
Toxic megacolon
Proctitis and rectal prolapse
- **Systemic**
Disseminated infection
HUS
Neurological complications
Reactive arthritis

While in the ward the patient develops seizures. What are the possible causes?

- Febrile convulsion
- Electrolyte imbalances – hypernatremia, hyponatremia, hypokalemia, hypocalcaemia
- Hypoglycaemia
- HUS
- Shigella encephalopathy

How would you manage hypernatremic dehydration?

- These children present with thirst out of proportion to the degree of dehydration and seizures
- The main principle is not to drop the sodium rapidly as this can cause cerebral edema
- Correct slowly over a period of 12 hours

How would you manage a patient with HUS?

- Present 5-10 days after the onset of diarrhoea
- The 3 features are
Microangiopathic hemolytic anaemia
Thrombocytopenia
Acute renal failure
- Management is mainly supportive
Anaemia – Blood transfusion
ARF – Fluid management, management of electrolyte imbalances, mainly K+, antihypertensive therapy and dialysis in severe cases

What is the advice you would give to the patient on discharge?

- This should focus on the following themes
- Prevention of further episodes of diarrhoea with proper hygienic practices of the family
- What to do in an episode of diarrhoea
- Give the child more fluid than usual
Teach the mother about ORS and the technique of preparation of ORS. Also tell her about other fluids which can be used
Advise when to stop giving ORS
- Continue to feed the child
- Bring the child to the hospital especially if
High fever
Blood stained stools
Poor oral intake
Features of dehydration and over hydration

Edema and nephrotic syndrome

Key points in the history – 1st episode of edema

1. Describe the edema

- Describe the onset of the symptoms and how the mother noticed them
- Describe the location of the edema
- Aggravating and relieving factors for the edema
- Describe the progression of symptoms over time
- What the mother did after noticing the symptoms
- Describe what was done in the hospital

2. Ask specific questions based on the differential diagnosis of edema. The case which is usually given is generalized edema

| Category | Disease | Specific points in the history |
|------------------|----------------------------|--|
| Renal | Nephrotic syndrome | Usually based on the progression and characteristics of the edema Usually starts in the periorbital region and then spreads downwards Also ask for any change in the urine |
| | Nephritic syndrome | Ask for associated red coloured urine and documented elevated blood pressure (ask the mother if she was informed about elevated blood pressure) |
| | Chronic renal failure | Ask for weakness, easy fatigue(associated anaemia) and uremic symptoms Also ask for past history of UTI |
| Cardiac | Heart failure | Ask for past history of cardiac disease, difficulty in breathing and poor exercise tolerance |
| Gastrointestinal | Cirrhosis | History of yellowish discolouration of the eyes, hematemesis, malaena, evidence of hepatic encephalopathy Ask for chronic diarrhoea |
| | Protein losing enteropathy | |
| Other | Angioedema | Allergic history |
| | Drugs | |

3. After establishing that the most probable diagnosis is nephrotic syndrome try to find an aetiology

- Ask for evidence of systemic involvement – rash, joint pain and morning stiffness, fever
- Infections such as hepatitis B, malaria, HIV can also cause nephrotic syndrome

4. Ask for the complications of nephrotic syndrome

- Fever with abdominal pain – SBP
- Flank pain with gross hematuria – Renal vein thrombosis
- Calf pain +/- difficulty in breathing – DVT and pulmonary embolism
- Collapse, syncope – Hypovolaemia
- Abdominal pain in a patient with nephrotic syndrome
 - Hypovolaemia
 - SBP
 - Renal vein thrombosis
 - Mesenteric thrombosis
 - Fluid collection around the liver
 - Intestinal edema
 - Gastric irritation due to steroids

Key points in the history – Known patient with nephrotic syndrome with a relapse

- 1. Describe the initial episode of edema and how the diagnosis was made at the time**
- 2. Mention what happened to the disease over time. DO NOT describe each of the relapses in detail. Just mention the number**
- 3. Describe the management**
 - Mention the drugs used
 - Ask for the side effects of the medication
- 4. As given above ask for an aetiology for the condition**
- 5. Mention the complications**
- 6. Social history is extremely important**

| General introduction to the family | |
|------------------------------------|--|
| Impact on the child | Playing, amount of school missed |
| Impact on the parents | Socio economic impact of the disease, impact of frequent hospital stays |
| Impact on the siblings | |
| Environment | Give a brief description of the environment of the household |
| Support available | Family support Extended family support Medical facilities available |

Education of the parents

- Education of the mother on the disease
- Knowledge about the drugs used and the importance of proper compliance
- Side effects of the medication
- Method of urine testing
- Knowledge on the diet and lifestyle modifications
- Identification of a relapse and when to bring the child to hospital
- Complications

Psychological state and expectations of the parents**Examination****General examination**

- Anthropometry – Weight, height and BMI (Weight and height is used to calculate the body surface area – this is on which the dose is calculated)
- Look for features of steroid toxicity
 - Cushingoid features
 - Weight gain and obesity
 - Hypertension
 - Cataract
- Establish the distribution of edema
- Look for vasculitic rashes in the skin – secondary cause for nephrotic syndrome

Abdomen

- Look for free fluid in the abdomen

Cardiovascular

- Exclude cardiac disease
- Measure the blood pressure

Respiratory

- Pleural effusions

Management of nephrotic syndrome

What is nephrotic syndrome?

- Edema
- Proteinuria ($>40\text{mg}/\text{m}^2/\text{h}$ or urine protein to creatinine ratio $>200\text{mg}$ protein/ mmol creatinine)
- Hypoalbuminaemia ($<2.5\text{g}/\text{dL}$)
- Hyperlipidaemia

Diagnosis

- Is based on the clinical presentation of the child and the investigations
- The child will present with edema which is initially notes in the periorbital region and later involves the dependant areas of the body and is worse towards the afternoon
- Exclusion of other causes of generalized edema

What are the Investigations you will do?

Investigations to confirm the diagnosis

- Urine ward test (Offers a qualitative assessment of the urinary protein) – is usually $>+3$
- Early morning urine sample for estimation of the urine protein to creatinine ratio
- 24 urine collection for protein estimation
- Urinalysis is also an important investigation to look for microscopic hematuria and red cell casts which may be found in patients with a nephrotic/ nephritic mixed picture
- Serum albumin
- Lipid profile may also be done (Elevated total cholesterol, LDL and triglycerides)

Note

Proteinuria in children

| Transient proteinuria | Orthostatic proteinuria | Fixed proteinuria |
|---|---|---|
| Associated with fever, dehydration, exercise | Asymptomatic Increased protein excretion in the upright | Indicates renal disease. Can be due to glomerular or tubular disease |
| Usually does not exceed +2 and is normal on repeated daily measurements | Absence of protein on an early void sample for 3 consecutive days | Significant proteinuria on an early morning void sample on 3 consecutive days |

Other investigations

- Renal function tests and serum electrolytes
- Serum complement
- ESR, ANA
- Hep B surface antigen
- Renal biopsy

Role of renal biopsy in nephrotic syndrome

Recommendations

- Age of onset less than 6 months
- Initial macroscopic haematuria in the absence of infection
- Persistent microscopic haematuria with hypertension
- Renal failure not attributable to hypovolaemia
- Persistently low C3, C4 levels
- Steroid resistance

Preparation of a patient for renal biopsy

- Do the initial workup of the patient. This includes the following investigations – serum creatinine, FBC, bleeding time and clotting profile, renal ultrasound scan
- Discuss with the team and arrange a date
- Cross match blood
- Fasting for 6 hours

Post op

- Monitor vital parameters, UOP
- Collect all urine samples
- Complete bed rest until hematuria settles

After diagnosis

Classification

- Classification of nephrotic syndrome is in to idiopathic and secondary nephrotic syndrome

| Idiopathic | Secondary |
|------------------------------------|--|
| Minimal change disease (85%) | Secondary to systemic diseases |
| Focal segmental glomerulosclerosis | Infections |
| Membranous | Drugs |
| Mesangioproliferative | Connective tissue disorders and vasculitis |

How would you manage the first episode of nephrotic syndrome?

General management of the child

- Start daily weight chart and input/ output charts
- Bed rest is not recommended
- Protein restriction in the diet is also not recommended. Therefore give the child a normal protein diet. **(Salt restriction may be done until resolution of this episode)**. Fluid restriction is also not recommended
- Monitor the PR, volume and blood pressure
- IV fluids – initially start with ½ of the maintenance over 12 hours. Then measure the urine output and give the fluid hereafter as previous day UOP+ insensible loss
- Management of gross edema. Diuretics may be used only if hypovolaemia has been corrected. Drug of choice is frusemide 1-2 mg/kg/d. Use in conjunction with CPP. Start the CPP and give the frusemide mid transfusion
- Antibiotics – prophylactic oral penicillin 250mg bd for 10 days

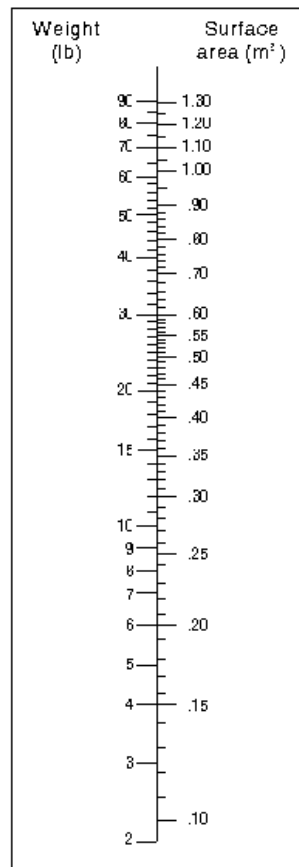
Steroid therapy

- Prednisolone 60mg/m²/d given as a single dose in the morning for 28 days. Then 40mg/m²/d on alternate days for 28 days
- Calculation of the body surface area should be done using the nomogram which is available in the ward
- Usually respond to steroids after 2-4 weeks

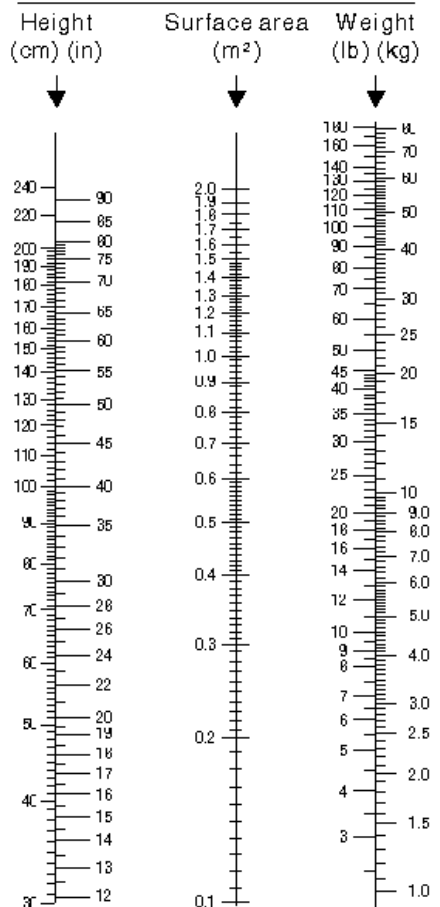
Remission

- Urine protein <4mg/m²/h
- Urine protein negative or trace for 3 consecutive days

FOR CHILDREN OF NORMAL HEIGHT AND WEIGHT



NOMOGRAM



BODY SURFACE AREA FORMULA (Adult and Pediatric)

$$BSA (m^2) = \sqrt{\frac{Ht (in) \times Wt (lb)}{3131}} \quad \text{or, in metric: } BSA (m^2) = \sqrt{\frac{Ht (cm) \times Wt (kg)}{3600}}$$

References

- Lam TK and Leung DT, "More on Simplified Calculation of Body Surface Area," *N Engl J Med*, 1988, 318(17):1130 (Letter).
 Mosteller RD, "Simplified Calculation of Body Surface Area", *N Engl J Med*, 1987, 317(17):1098 (Letter).

What is the advice you will give the parents?

- The first step is to inform the parents that nephrotic syndrome is a relapsing, chronic disease and that their support and understanding is extremely important to offer adequate management for the child. Also give them a basic idea of what happens in nephrotic syndrome
- Reassure them that progression to end stage renal failure is rare
- Explain the home based management of nephrotic syndrome
Give a normal diet to the child

Should contain all the nutrients but reduce fat and refined sugar

Ensure normal activity and school attendance in the child

Explain the method of urine protein testing at home – Frequency of checking

Method of checking for proteins at home

Collect urine to fill 2/3 of a test tube

Heat the upper part of the tube

Look for the formation of turbidity. Add a few drops of vinegar and see if the turbidity disappears (phosphates)

Quantify the amount of protein by holding the tube up against a newspaper

Nil – no turbidity, + - slight turbidity but can read the letters, ++ - cannot read the letters but can see the black color +++ - cannot see the print or the black color, ++++ - precipitate

Maintain a diary of the protein testing

Seek early medical attention for infections

- Educate the parents about prednisolone
- Importance of steroids and the risk of Addisonian crisis if withdrawn abruptly. DO NOT stop when the child develops an infection
- Educate the parents about the side effects of medication – especially prednisolone
Behavioural changes especially irritability
Increased appetite and weight gain
Cushingoid features
Gastric irritation – therefore give with meals
With long standing steroid use other side effects may occur – Growth faltering, cataract, obesity, hypertension, hyperglycaemia, osteopenia, recurrent infections
- Advise on vaccination – avoid live vaccines for 3 months after stopping steroids
- Try to avoid crowded places due to the risk of infection
- Ask them to admit the child if there is edema or >+2 protein for more than 2 days at home

Follow up

- Follow up with the urine protein testing records of the mother. Look for any complications of drug therapy

Future progression of the disease and the management

| Category | Definition | Management |
|--------------------------------------|---|---|
| Relapse | Urinary protein excretion >40mg/m ² /h OR Urine testing shows 2+ or more for 3 consecutive days OR Recurrence of proteinuria at any level with hypoalbuminaemia <2.5g/dL Having been in remission | <ul style="list-style-type: none">• Relapses occur in 60-70% of children with nephrotic syndrome• Manage the 1st and 2nd relapse as given below• Prednisolone 60mg/m²/d given as a single dose in the morning for 4 weeks. Then 40mg/m²/d on alternate days for 4-6 weeks |
| Frequent relapses | Two or more relapses in the first 6 months after diagnosis OR Four relapses in any 12 month period | <ul style="list-style-type: none">• Re induce remission as given above• Then give a maintenance dose of Prednisolone 0.1-1mg/kg EOD for 6 months and slowly withdraw over a 6-12 month period |
| Relapse while on Prednisolone | | <ul style="list-style-type: none">• Levimasole• Other drugs Cyclophosphamide |
| Steroid dependent nephrotic | 2 consecutive relapses while on steroids or relapse within 14 days of cessation of therapy | |

| Drug | Side effects |
|-------------------------|---|
| Levamisole | Reversible neutropenia (Check white cell count 2 weeks after treatment. Then monthly for the next month and 3 monthly thereafter) |
| Cyclophosphamide | Leucopenia, hemorrhagic cystitis, alopecia, nausea and vomiting |

Management of complications of nephrotic syndrome

| Complication | Aspects of management |
|---------------------------|---|
| Hypovolaemic shock | Give a 10ml/kg bolus of CPP and continue monitoring the vital parameters and the hematocrit |
| Peritonitis | Start antibiotics IV benzyl penicillin and IV cefotaxime Prevention Oral penicillin 250mg bd during the episode Pneumococcal vaccination |
| Thromboembolism | Anticoagulation, initially with heparin and then continue with warfarin |

Hematuria in children

History

Presenting complaint

- The patient will present with red coloured urine
- State the duration

History of the presenting complaint

- Describe the symptom carefully based on the following points – onset, duration and progression of the symptoms
- Red coloured urine in children is not always due to hematuria but an alternative cause should be considered only by exclusion
- Describe the characteristics of the red coloured urine. This can indicate the site of bleeding
Cola coloured and mixed throughout the stream – Glomerular bleeding
Fresh blood, associated clots and more towards the end of the stream – Lower urinary tract bleeding

Try to reach a differential diagnosis

| Cause | Key points in the history |
|---|---|
| Glomerular Acute nephritic syndrome | Ask for associated swelling of the body and decreased urine output Also ask the mother if she was told that the child had increased blood pressure Look for an aetiology Ask for preceding sore throat or skin sepsis a few weeks back (Post streptococcal glomerulonephritis) Ask for systemic features such as fever, malaise, joint pain and stiffness and skin rashes (Vasculitis and connective tissue disease (SLE, HSP)) |
| Mixed nephrotic and nephritic syndrome IgA nephropathy | Ask for history of recurrent gross hematuria. (Can occur 1-2 days after a URTI) |
| HUS | Ask for preceding history of AGE (5-10days back), fever, abdominal pain, seizures |
| Other rare glomerular disease | Family history of similar disease |

| | |
|---------------------------|---|
| Extra glomerular | |
| UTI | Ask for associated crying on micturition and fever |
| Stones | Associated abdominal pain and family history of urinary calculi |
| Trauma | |
| Bleeding disorders | Other sites of bleeding |

- Exclude other causes of red coloured urine
Consumption or red coloured food substances, drugs, associated features of jaundice and anaemia (haemoglobinuria)
- Describe what has happened to the child up to now

Ask for complications of nephritic syndrome (this will be the most probable diagnosis)

- Altered level of consciousness, seizures – hypertensive encephalopathy
- Acute renal failure
- Dyspnoea, poor exercise tolerance – heart failure

Take the other routine aspects of the history

Examination

General examination

- **Anthropometry** – This is extremely important in the management
- Look for pallor and Icterus – could indicate hemoglobinuria
- Note the distribution of edema
- Look for healed skin wounds, skin rashes suggestive of SLE or other types of vasculitis

Cardiovascular examination

- Measure the blood pressure
- Look for features of heart failure

Abdomen

- Palpate for masses – tumors of the renal tract, PCKD can present with hematuria

Neurological examination

- Examine the fundus for evidence of malignant hypertension

How would you investigate a patient with nephritic syndrome?

- Urine full report and microscopy
 - Glomerular hematuria** – Red cell casts, dysmorphic red cells (special microscopy) and proteinuria >100mg/dl
 - Hematuria from the tubules** – White cell casts, epithelial casts
 - Lower urinary tract** – Normal red cell morphology, proteinuria <100mg/dl
- Follow up a case of glomerular hematuria with the following investigations
 - FBC
 - BU/SC and serum electrolytes
 - Serum complement
 - ASOT
 - DNAase B
 - ANA
- Other investigations may be required if an extraglomerular cause is suspected – urine culture, USS of the abdomen

Management of AGN

General management

- The management of AGN is usually supportive
- Start a monitoring chart
 - Daily weight
 - Input output chart
 - Blood pressure
- **Fluid management**
 - Calculate the maintenance fluid requirement for the child and give $\frac{1}{2}$ of this amount over 12 hours. Then measure the urine output over this time and continue as
 - Fluid input = UOP + insensible loss
- **Management of edema**
 - Furosemide 1mg/kg
- **Diet**
 - Give a balanced diet with restricted salt. Do not give the child foods rich in potassium
- **Antibiotics**
- Monitor for complications
 - Hypertensive encephalopathy
 - Acute renal failure
 - Cardiac failure

Management of acute hypertension

- **Diagnose hypertension**
- Classify the severity of hypertension as hypertensive urgency or hypertensive emergency

| Diagnosis | Definition | Management |
|------------------------|--|---|
| Hypertensive urgency | Elevation of blood pressure without severe symptoms or evidence of target organ damage | Oral medication Oral nifedipine |
| Hypertensive emergency | Elevation of blood pressure with target organ damage | ABC IV antihypertensives – IV hydralazine Drop the blood pressure slowly |

Management of acute renal failure

Principles of management are given below

Fluid and electrolyte balance

- Give IV fluids as previous day's urine output + insensible loss based on the body surface area of the child
- Hyperkalemia
Salbutamol nebulization
IV calcium gluconate for stabilization of the myocardium
Insulin – dextrose infusion
Potassium binding resins
- Consider renal replacement therapy

When will you discharge the patient?

What is the advice you will give at discharge?

- The disease process will continue for 2-3 weeks. At the end of the natural course of the disease the child will develop polyuria. It is important to monitor the child during this period as well as the child can get hypovolaemic

At discharge

- Explain the disease to the mother
- Tell them not to restrict the diet if the child
- The child can be discharged on antihypertensive medication
- Explain the warning signs of hypertensive encephalopathy and tell them to come to hospital immediately
- Get the blood pressure measured EOD by a GP
- Review in the clinic in 1 week with a urine full report (may have microscopic hematuria)

Management of UTI in children

How would you diagnose UTI?

- This is based on the clinical assessment and the findings on the urine culture and ABST
- The child will present with the following symptoms based on the age. Look at the following table as a guide

| Age group | | Symptoms and signs | | |
|---|-----------|---|---|--|
| | | Most common | ↔ | Least common |
| Infants younger than 3 months | | Fever Vomiting Lethargy Irritability | Poor feeding Failure to thrive | Abdominal pain Jaundice Haematuria Offensive urine |
| Infants and children, 3 months or older | Preverbal | Fever | Abdominal pain Loin tenderness Vomiting Poor feeding | Lethargy Irritability Haematuria Offensive urine Failure to thrive |
| | Verbal | Frequency Dysuria | Dysfunctional voiding Changes to continence Abdominal pain Loin tenderness | Fever Malaise Vomiting Haematuria Offensive urine Cloudy urine |

- The clinical assessment should also indicate any serious underlying pathology which is associated with the infection. The points shown below are important in this regard
 - Recurrent UTI
 - Voiding dysfunction and poor urine flow
 - Evidence of hydronephrosis
 - Palpable bladder
 - Evidence of chronic renal failure
- The next step is to confirm the diagnosis. The investigation of choice is a urine full report and a urine culture/ ABST
- The urine culture/ ABST is considered as the gold standard of diagnosis for a UTI. Therefore the sample collection, transport and interpretation are extremely important
- These points are commonly asked at the exam

How would you collect urine for urine culture/ABST?

Methods of collection

- Clean catch midstream sample
- Supra pubic aspiration
- Catheter samples (usually not recommended except in failed SPA)

Advice to the parents on collection of CCMS urine

- Ask the mother to feed the child prior to the collection of the sample
- Wash hands and genitalia with soap and water. Retract the prepuce in older boys
- Open the cap of the special bottle once the child has started to pass urine. **Do not leave the lid open for a long time**
- Discard the first part of the urine and collect the midstream urine sample directly into the bottle. **Avoid contact of the bottle with the perineum of the child**
- Close the cap and hand over the bottle immediately

Transportation

- Fill out the request form and send immediately to the lab
- If the specimen cannot be transported within 2 hours refrigerate for a maximum of 24 hours

Interpretation of the culture report

- In a CCMS $>10^5$ would indicate a high probability of a UTI (80-95%). If it is between 10^4 and 10^5 an infection is likely
- Any number of colonies on an SPA culture would indicate a 99% probability of an infection

Other investigations

- Full blood count
- CRP
- Renal function tests
- Serum electrolytes

Describe the initial management of UTI in children

Initial management

General management

- Ensure proper hydration of the child
- Manage fever and pain with paracetamol

Antibiotic therapy

- Empirical antibiotic therapy
All cases of suspected febrile UTI should be started on empirical antibiotics. These should be started after collection of urine for culture
The selected antibiotics should cover the possible organisms – E. coli, Klebsiella, Proteus and Enterobacter
- Choice of antibiotics and route of administration
Oral – Cephalexin, Co amoxyclav, Cotrimoxazole
IV (should be given if the child is extremely ill or if the child refuses to take orally
Cefotaxime, ceftriaxone, cefuroxime
- Duration of treatment is usually 7 days

Follow up

- Review after 48 hours
- The child should show response to treatment usually within 48 hours. If the symptoms persist check the adequacy of the antibiotic treatment and the ABST
- Change or alter the dose of antibiotics if necessary

What are the further investigations necessary?

USS KUB

- USS should be performed in all children with a febrile UTI within 6 weeks of the attack. Look for
Gross structural anomalies of the renal tract
Features suggestive of acute pyelonephritis
Hydronephrosis and hydroureter
Bladder

Prophylaxis and further investigations

- Prophylaxis is indicated for all children < 5 years following the first attack of UTI until the USS report is available
- The continuing management is based on the following principles
- In a child with a febrile UTI – If the USS is normal continue the prophylaxis until the recommended imaging studies are available
- If structural anomalies are detected or in cases of recurrent UTI continue prophylaxis till 5 years or longer
- Start prophylaxis as given below
- Complete the course of antibiotics and start prophylaxis from the next day onwards. Repeat a urine culture at the 5th day of prophylaxis
- Antibiotics used for prophylaxis are given below. These drugs are given as a single dose at night

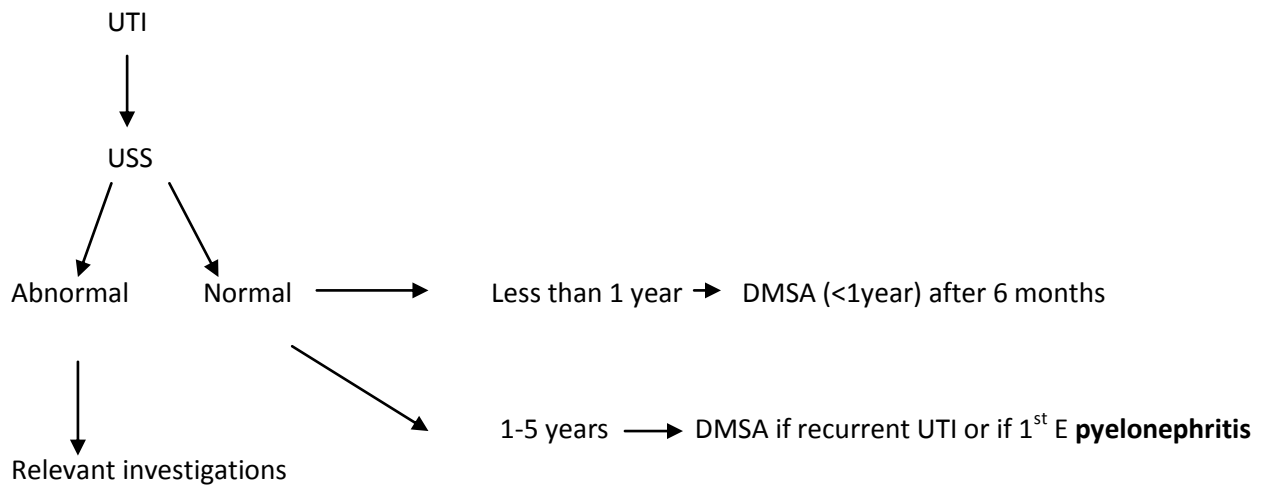
| Drug | Use |
|-----------------------|-------------------------------------|
| Cephalexin | Recommended in the first 1-3 months |
| Cotrimoxazole | Avoid in infants < 1 month of age |
| Nitrofurantoin | Avoid in less than 3 months |
| Nalidixic acid | Avoid in less than 6 months |

| Investigations | Indication | Uses |
|--|---|--|
| DMSA (Should be done after 6 months after the UTI) | <p>Under 1 year All children with a febrile UTI</p> <p>Under 5 years Clinical picture suggestive of acute pyelonephritis Recurrent febrile UTI Abnormal USS</p> | Looks for renal scarring |
| MCUG | <p>Suspected bladder outlet obstruction</p> <p>When the USS reveals hydronephrosis and hydroureter</p> <p>Recurrent UTI</p> | Used to diagnose posterior urethral valves and VUR |
| DTPA | Suspected PUJ or VUJ obstruction | <p>Used to diagnose PUJ and VUJ obstruction</p> <p>Can also give an idea about the differential renal function</p> |

- Preparation of the patient for the above investigations

Prevention of recurrent attacks of UTI – Advise the parents

- Educate the parents on the condition the child is having and the prognosis
- Teach them how to recognize the condition and when to bring the child to the hospital
- Educate them on the importance of giving the child the recommended prophylactic medication
- General
 - Good hydration
 - Avoiding constipation
 - Improve the hygiene of the child
 - Wiping of the perineum from front to back
 - Change the child's nappies frequently



Surgical conditions of the urinary tract

| Condition | Diagnosis | Management |
|----------------------------------|---|---|
| Posterior urethral valves | MCUG Dilated, elongated posterior urethra with a thick walled trabeculated bladder May have associated VUR | Cystoscopic ablation |
| VUR | MCUG is used for the diagnosis and the classification of VUR | Conservative medical With antibiotic prophylaxis Surgical intervention Indicated in recurrent UTI in |

| | | |
|------------------------|------|---|
| | | spite of prophylaxis, patients with impaired renal function, recurrent new scar formation and gross VUR |
| PUJ obstruction | DTPA | Reimplantation is performed Pyeloplasty |

Acute flaccid paralysis

Presenting complaint

- Weakness of the lower limbs
- State the duration

History of the presenting complaint

- Remember that in a neurology case the most important aspect in the history is the chronological order of development of the symptoms
- Describe the onset of the weakness as sudden onset or gradual onset. Then go on to describe the distribution of the weakness
- Then go on to describe the progression of weakness over time
- The description of the neurological impairment at each stage in the chronological order should be described based on the activities of the child such as running and playing, walking, sitting, standing up from a sitting position and other day to day activities
- Describe the other associated neurological symptoms
 - Altered level of consciousness or loss of consciousness
 - Change in behavior
 - Seizures
 - Diplopia
 - Deviation of the mouth, drooling of saliva from the side of the mouth
 - Vertigo
 - Dysphagia and nasal regurgitation
- Think of the possible differential diagnosis based on the site of the lesion and ask specific questions

| Site of the lesion | Disease | Specific questions in the history |
|-----------------------------------|-------------------------|--|
| Spinal cord | Spinal cord injury | Ask for history of trauma to the spine |
| | Transverse myelitis | Ask for associated backache, paresthesia, bladder and bowel incontinence |
| Anterior horn cell disease | Poliomyelitis | Ask for initial history of fever, upper respiratory and GI symptoms. This is followed by a symptom free period of a few days Then there is recurrence of the fever and muscle tenderness, neck stiffness and paralysis (usually asymmetric) |
| Peripheral nerve | Guillain barre syndrome | Ask for preceding history of diarrhoea or respiratory tract illness. This is |

| | | |
|-------------------------------|---------------------------------------|---|
| | | followed a few weeks later by paresthesia of the extremities and an ascending paralysis |
| | Other causes of peripheral neuropathy | Exposure to toxins, use of long term drugs, recent vaccines (post rabies virus) |
| Neuromuscular junction | Botulism | Possible ingestion of contaminated canned food Initial cranial nerve symptoms and descending paralysis |
| | Myasthenia gravis | Preceding history of drooping of the eyelids or weakness which is most prominent towards afternoon |
| | Snake venom and toxins | Ask for possible history of snakebite and exposure to toxins |
| Muscle | Myositis Periodic paralysis | Muscle pain, associated skin rashes |

- Then go on to describe what was done to the child up to hospital admission and state the progression of the neurological symptoms
- Ask for the complications of the disease, specifically for bulbar involvement, respiratory muscle weakness
- Describe any specific management which has been done to the child
- Describe the present status of the child. This is also in regard to the functional status

Past medical and surgical history

Birth history

Growth and developmental history

Immunization history

- This is very important. Describe the present state of immunization
- Describe clearly about the polio vaccinations of the child

Social history

- This is a critical component of the history
- Describe the following components of the social history
- General introduction to the family
- Impact of the disease on the child
- Impact of the disease on the parents

- Environmental factors
This is extremely important in a child with paralysis. Describe the environment around the house and accessibility to the house. Then describe the indoor environment of the house, obstacles and hazards to the child. The toilets are also important,
If the child is attending school describe how the child travels to school, the location of the classroom and accessibility. Also describe the type of toilets available in the school
- Support available
- Education of the parents regarding the condition and future expectations
- Psychological state of the parents

Examination

Nervous system

- Examine the nervous system starting from the lower limbs and go on to examination of the upper limbs and the cranial nerves
- As stated above the site of the lesion in acute flaccid paralysis can extend from the spinal cord level to the muscle
- Given below is a comparison of the physical signs of the most important pathologies causing acute flaccid paralysis

| | Spinal cord lesion | Poliomyelitis | GBS |
|---------------------------------|---------------------------------|--|---|
| Weakness | B/L weakness in the lower limbs | Asymmetric involvement of the lower limbs, hypotonia, proximal>distal weakness | B/L weakness of the lower limbs, hyporeflexia |
| Sensory | Sensory level | No sensory involvement | No sensory involvement |
| Other important features | Bladder and bowel incontinence | Can have associated bulbar weakness | Can have associated lower cranial nerve Involvement |

Respiratory system

- A proper assessment of the respiratory effort is essential
- Count the respiratory rate, look for features of respiratory distress
- Ask the child to perform a single breath count and look for the cough effort

Abdomen

- Look for a palpable bladder

Discussion

Guillain Barre Syndrome

Diagnosis

- The diagnosis of GBS is entirely clinical
- Investigations may be performed if there is any doubt about the clinical diagnosis
 - NCS
 - LP after 10 days of appearance of symptoms – shows cytoprotein dissociation

Initial management

- Book an ICU bed
- Assess the child and start monitoring the key clinical parameters. These are
 - Pulse rate and rhythm, BP** – Autonomic instability
 - Respiratory function** – Single breath count, features of respiratory distress, assessment of vital capacity (this is often difficult in children)
 - Neurological parameters** – State of paralysis and rate of progression with grading of the muscle power
- Admit the child to the ICU if the following are present
 - Rapidly progressive paralysis
 - Bulbar palsy
 - Respiratory involvement – Deteriorating SBC, features of respiratory distress, vital capacity <20ml/kg – **This is an indication for ventilation**
 - Autonomic cardiovascular instability
- Consider **IVIG therapy** if there is rapid progression of the neurological impairment
 - The recommended dose is 0.4g/kg for 5 days. The infusion rate should be slow initially as there is risk of anaphylaxis and later increased
- Continue monitoring the patient

Notification and other important investigations

- Any child under 15 years of age with acute flaccid paralysis should be notified immediately. The notification should be done to the MOH of the area and the RE by telephone
- The investigation form EPID/37/1/R2004 should be completed and returned to the epidemiological unit

Collection of stool samples

- Samples should be collected within 2 weeks of the onset of paralysis into the provided special container
- Two samples of stools should be collected 24-48 hours apart
- Sample should weigh 8-10g (the size of two thumbnails)

- Lid should be tightly closed and packed in ice
- The sample should be correctly labeled. The following details should be present
 - Introduction – as in any sample
 - Date of onset of paralysis
 - Date of collection of stools
 - Date of dispatch of stools
 - Last date of polio vaccination
- Samples should be transported to the MRI within 72 hours of collection
- The MOH should personally investigate the case of AFP and visit the community where the case is resident
- He should collect and dispatch one stool sample from 3-5 contacts and send to the MRI within 72 hours of collection
- The MOH is also responsible for initiating a program of limited outbreak response immunization. This includes administration of an extra dose of OPV to the children of the same age or below living around a 2km radius of the residence of the index case

Continuing management

- The patient should have supportive management while in the ward
 - Management of muscle pain
 - Bladder and bowel care
 - Chest physiotherapy for prevention of respiratory or chest infections
 - Passive physiotherapy

Rehabilitation

- Patients with GBS begin spontaneous recovery after 2-3 weeks in an inverse direction to the direction of paralysis
- Physiotherapy is extremely important
- Rehabilitation includes occupational therapy

Meningitis

Discussion

Diagnosis of meningitis in children

- The diagnosis of meningitis is made on the history, examination and investigations
- The important aspect is that the clinical presentation varies according to the age of the child
- Both bacterial and viral meningitis has a similar presentation but the latter usually has a milder course
- The table given below gives the presentations in various age groups

| | 0-2 months | >2 months | Older children |
|-------------|--|---|---|
| History | Usually no specific features Fever or hypothermia Irritability and high pitched cry, lethargy, poor feeding Seizures Apnoeic attacks Altered sleep pattern | Fever Irritability, lethargy Seizures | Fever Irritability, lethargy |
| Examination | Bulging fontanelle (Should be examined in the upright position when the child is not crying) Opisthotonus | Bulging fontanelle Brudzinkin's sign | Neck stiffness (more useful in children >3years) Kernig's sign Photophobia |

Describe the Initial management of the child

- The initial management should focus on the **A,B,C**
- Look for evidence of complications – increased intracranial pressure, sepsis, seizures
- Correct any abnormalities as you find it
- Obtain IV access and collect blood for investigations
 - Full blood count
 - Obtain blood for blood culture
 - C- reactive protein
 - Serum electrolytes
 - Renal function tests

- **Lumbar puncture and CSF analysis** for definitive diagnosis
Remember that even though the LP is used for the definitive diagnosis of meningitis there may be indications to delay the LP
- **Indications to delay the LP**
Symptoms and signs of increased intracranial pressure
GCS <13 or deteriorating level of consciousness
Recent (within 30 minutes) or prolonged seizures
Focal neurological symptoms and signs
Shock
Coagulopathy
Local sepsis
- The general rule should be to stabilize the patient before performing a LP
- Remember that in the case that the LP should be delayed **do not delay the 1st dose of IV antibiotics**
- The samples for LP should be taken as follows
CSF sugar (to be interpreted with a random blood sugar taken about 30 minutes before the procedure)
Protein
CSF culture
Full report, including gram stain
Other special investigations – bacterial antigen detection, mycobacterial, viral studies

Interpretation of the CSF report

| | Viral | Bacterial | Partially treated | TB |
|---------------------------|---------------------------|--------------------|--------------------------|------------------------------|
| Appearance | Clear | Turbid | Clear | Turbid, may clot on standing |
| Cells | 15-2000 | 10-10000 | 5-10000 | 10-500 |
| Differential count | Lymphocytes | Neutrophils | Monocytes or neutrophils | Lymphocytes |
| Glucose | >50% of BG | <50% of BG | Normal or decreased | <50% of BG |
| Protein | Normal/ slightly elevated | Elevated (100-500) | Elevated (100-500) | Very high |

Initial pharmacological management

- Pharmacological management should be initiated after the samples are collected for blood culture and CSF analysis
- Commencement of empirical antibiotic therapy is the most important management option in suspected acute bacterial meningitis

- Do not delay the antibiotics even if the LP is delayed
- Steroid therapy is also indicated in suspected acute bacterial meningitis. However there are specific criteria for the use of steroids

Age > 3 months

Should be administered before the first dose of parenteral antibiotics

- The recommended dose is 0.15/kg/dose IV every 6 hours. The first dose of steroids should be followed by the first dose of IV empirical antibiotics
- The recommended empirical antibiotics vary according to the age of the child and the antibiotic sensitivity patterns

| Age | Organisms | Recommended antibiotics |
|--------------------|---|--|
| Neonates | GBS, <i>E. coli</i> , <i>listeria</i> | Ampicillin or benzyl penicillin + cefotaxime |
| 1-2 months | Neonatal organisms, <i>haemophilus</i> , <i>pneumococcus</i> , <i>meningococcus</i> | Ampicillin or benzyl penicillin + cefotaxime |
| 2 months – 5 years | <i>haemophilus</i> , <i>pneumococcus</i> , <i>meningococcus</i> | Cefotaxime/ ceftriaxone |
| >5 years | <i>pneumococcus</i> , <i>meningococcus</i> | Cefotaxime/ ceftriaxone |

- The antibiotics may be altered according to the results of the cultures, based on the ABST
- Continue antibiotics for 10-14 days in an uncomplicated *Streptococcus pneumoniae* meningitis and 7-10 days for an uncomplicated *Haemophilus influenzae b* infection

As a house officer how would you assess a patient on your daily ward round?

- Maintaining a monitoring chart is extremely important. This chart should include the following data
 - QHT fever monitoring
 - Vital parameters – PR, RR, BP
 - Neurological assessment – GCS, pupillary reflexes, examination of the cranial nerves and limbs
 - Chart the OFC
- Maintain an input-output chart
- Keep a record of the daily investigations
- IV fluids – May be given in these patients at 1/2 - 2/3rds maintenance due to the risk of SIADH in normovolaemic and normotensive patients (But may be returned to normal if the serum sodium is normal)
- Manage dehydration and hypotension with 0.9% saline
- Identify and manage complications as they arise

What are the complications of meningitis?

Early neurological

- **Increased intracranial pressure** as a result of cerebral edema
 - Call and book an ICU bed
 - Nurse the patient with a 15-30 degree elevation of the head in the midline position
 - Temperature control
 - Appropriate fluid and electrolyte therapy – 1/2 – 2/3 maintenance after hypotension and deficits are treated
 - Seizure control
- **Specific measures**
 - Mannitol – 2.5 -5 ml/kg of 20% solution over 30 minutes
 - Frusemide – Can be used in combination with mannitol
- **Seizures**
 - ABC**
 - IV midazolam 0.15mg/kg or rectal diazepam 0.5mg/kg
 - For continuing seizures a bolus dose of phenytoin at 20mg/kg over 20 min may be given
- Stroke
- Acute hydrocephalus
- Cranial nerve palsies
 - Hearing impairment
- Subdural effusion

Other

- Disseminated infection and sepsis
- Electrolyte imbalance – SIADH
- Nosocomial infections

Late neurological

- Cognitive impairment

Chemoprophylaxis

- Recommended in all household contacts irrespective of the age when at least 1 unvaccinated contact is younger than 4 years of age
- Drug of choice is Rifampicin
- When index case is less than 2 years commence a full course of Hib vaccination regardless of the vaccination status

Follow up

- Brain imaging
- Assessment of the hearing of the patient
- Regular developmental assessment

Encephalitis

Not a common topic of discussion at the long case but the basic details given below should be known

Diagnosis

- Clinical
- Presents with a non specific prodromal period which is followed by CNS symptoms such as alteration of behavior, irritability, altered level of consciousness and seizures
- May be associated with meningitis
- Metabolic encephalopathies and post infectious encephalomyelitis should be considered as the differential diagnosis
- Causes
 - Viral**
 - HSV
 - Other herpes viruses – VZV, CMV, EBV
 - Enteroviruses
 - Arboviruses – JE
- **Investigations**
 - LP – CSF analysis typically shows a lymphocytic predominant leucocytosis with normal CSF glucose
 - EEG – Diffuse slowing or focal EEG changes
- **Management**
 - This is usually supportive
 - Manage seizures and increased intracranial pressure

Seizures in children

History

Presenting complaint

- The child will present with abnormal movements
- State the duration and number of episodes over this time (latest presentation)

History of the presenting complaint

- Describe the episode in detail. This includes the following details
- Pre ictal phase
- Ictal phase
- Post ictal phase
- What the mother did in response to the episode
- From these details the main objective is to identify the seizure pattern and to exclude seizure like events

Partial (focal) seizures

- Are of 3 types
- Simple partial, complex partial and partial seizures with 2ry generalization
- The 2 important types are described below

| Phase | Simple partial | Complex partial |
|-------------------|---|--|
| Pre ictal | An aura may be present | An aura may be present Can start with a simple partial seizure |
| Ictal | Consciousness is retained May present with motor symptoms – focal in origin with or without a Jacksonian march May also have features of head turning and conjugate eye movements – versive seizure Sensory presentations may also occur | Consciousness is impaired Automatisms are commonly associated – prolonged and repetitive lip smacking, chewing, swallowing and excessive salivation May also have gestural automatisms which involve alteration of behaviour May develop secondary generalization |
| Post ictal | Child is well after the seizure | Child is well after the seizure |

Generalized seizures

Important seizure types which could be given at the exam are given below

| Phase | GTC | Absence | Infantile spasm |
|------------|--|---------------------------------|-----------------|
| Pre ictal | No preceding aura | | |
| Ictal | Ictal cry Eyes rolling up Initial tonic state Subsequent clonic movements Urinary or faecal incontinence Tongue biting, frothing from the corner of the mouth | Transient loss of consciousness | |
| Post ictal | Have post ictal drowsiness | | |

Management of seizures and epilepsy in children

Evaluation of the first seizure

- In a child presenting with a seizure the first step is to make a clinical diagnosis based on the history and examination
- Look for a secondary cause (see history, examination and initial investigations)

Definition of epilepsy

Clinical condition characterized by recurrent unprovoked seizures

Diagnosis

- Is a clinical diagnosis
- The most important tool for the diagnosis is a firsthand witness account of the event

Classification of epilepsy in children

- Can be classified based on the seizure type and also by the epileptic syndrome. A syndrome of epilepsy is based on the age of onset, cognitive development, seizure type, findings on examination and the EEG findings
- About 50% of childhood seizures can be classified into a specific syndrome
- Classification based on the seizure type

Partial (focal)

Simple partial

Complex partial

Generalized

Generalized tonic clonic

Tonic

Clonic

Myoclonic

Atonic

Absence

Infantile spasms

Unclassified

- Classification of epileptic syndromes is complicated and is not asked at the long case discussion

| Name | Age | Seizure pattern | EEG pattern |
|---|-------------|--|---|
| Generalized epilepsies Infantile spasms | 4-6 months | Flexor spasms, clusters usually occurs on waking | Hypsarrythmias |
| Lennox – Gastaut syndrome | 1-3 years | Multiple seizure types, but mostly drop attacks, tonic seizures and atypical absences associated neurodevelopmental arrest or regression and behaviour | |
| Typical absence | 4-12 years | Absence seizures, child is developmentally normal. Episodes can be induced by hyperventilation | Generalized 3 per second spike and wave discharge |
| Juvenile myoclonic epilepsy | Adolescence | Myoclonic seizures, but generalized tonic clonic seizures and absence seizures may occur | Characteristic EEG |
| Focal epilepsy Benign rolandic epilepsy | 4-10 years | Simple partial seizures, tonic clonic seizures in sleep, abnormal feelings in the tongue and distortion of the face | Focal sharp waves in the centrotemporal area |

Investigations

EEG

- EEG Is an important investigation in a child with epilepsy. It is usually done after the second seizure
- Uses of EEG
 - Determination of the seizure type
 - Diagnosis of epileptic syndromes
 - Determination if further investigations are required
 - Prognosis

- About 40% of children with epilepsy will have a normal first EEG
- Other special methods may be utilized if the EEG is not conclusive. These are sleep EEG and video EEG (useful for evaluation of suspected pseudo seizures)

Neuroimaging

- **MRI**
May be used in special circumstances

A complete diagnosis in a patient with seizures includes the following details (Based on the ILAE recommendations)

- Seizure semiology
- Seizure type
- Seizure syndrome
- Impairment
- Aetiology

Management

Education

- The first aspect of the management is the education of the parents and caregivers of the child
- Education should include the following aspects
 - General information on epilepsy
 - Information of the first aid in an attack of seizures
 - Lifestyle modifications
 - Antiepileptic drugs and their side effects
 - Importance of proper compliance to the medication and how to administer the drugs
 - Psychosocial issues and social stigma

Antiepileptic drugs

- Starting of antiepileptic drugs should be done by a consultant pediatrician. It is usually initiated only in patients with recurrent seizures
- The choice of first antiepileptic drug depends on the seizure type/ syndrome, adverse effects, co morbidity, availability and cost
- Monotherapy is preferred over polytherapy
- The drug should be started at a low dose and gradually increased towards the maintenance dose

Pharmacology of antiepileptic drugs

| Drug | Mechanism of action | Pharmacokinetics | Adverse effects |
|------------------|---|---------------------------------|--|
| Carbamazepine | Blocks the voltage dependent sodium channels | Induces hepatic enzymes | CNS symptoms Diplopia, blurring of vision, dizziness and ataxia Other Skin rashes, blood disorders, hepatotoxicity |
| Sodium valproate | Decreases the breakdown of the inhibitory neurotransmitter GABA | Inhibitor of hepatic metabolism | CNS symptoms Hepatotoxicity (more in children less than 3 years) Other Weight gain, alopecia, blood disorders, pancreatitis |
| Phenytoin sodium | Membrane stabilizing effect | Inducer of hepatic enzymes | CNS Impairment of cognitive function, Diplopia, blurring of vision, dizziness and ataxia Other Skin rashes, coarsening of facial features, hirsutism, gum hypertrophy |
| Phenobarbitone | Barbiturate | | Behavioural changes, hyperactivity, sedation |
| Lamotrigine | Blocking of voltage dependant sodium channels | | Generally well tolerated but can cause cutaneous adverse effects – TEN, Steven – Johnson syndrome (risk is higher with the concomitant use of valproate) |
| Topiramate | Blocking voltage dependent sodium channels and enhances GABA activity | | Sedation, word finding problems, weight loss, acute myopia and raised intraocular pressure |
| Clonazepam | Benzodiazepine | | Drowsiness, insomnia |
| Vigabatrin | Structural analog of GABA | Does not induce liver enzymes | Visual field disturbances, confusion, psychosis |

Choosing an antiepileptic

The choice of a suitable antiepileptic is based on the following principles

- Efficacy
- Support by clinical guidance and research
- Side effects
- Predicted compliance to the medication
- Availability
- Cost

| Type of seizure | First choice antiepileptic drug | Other options |
|--------------------|-----------------------------------|---|
| Generalized | | |
| GTC | Sodium valproate | Topiramate Lamotrigine |
| Absence | Sodium valproate Ethosuximide | Topiramate Lamotrigine |
| Myoclonic | Sodium valproate Lamotrigine | Clobazam Clonazepam |
| Infantile spasms | ACTH Prednisilone | |
| Focal | Carbamazepine Sodium valproate | Lamotrigine Topiramate Clobazam Clonazepam |

Follow up

- Follow up of the child should be done based on the following principles
- Review the last attack of seizure
 - If seizures are continuing rethink the diagnosis
 - Confirm the seizure type
 - Check if the dose is adequate for the age of the child
 - Assess the compliance for the medication
 - Try increasing the dose of the existing anti epileptic
- **Use the principles of antiepileptic drug therapy**
- Remember that monotherapy is preferred over polytherapy
- If monotherapy in the maximal dose has failed introduce a second drug and monitor the response. Then gradually tail off the first drug and continue monotherapy with the second
- Emphasize the basic patient education on seizures
- Assess the for the side effects of the medication

- Assess the other parameters of the child, especially the development
- When the child is seizure free for 2 years or more consider tailing off the medication

Approach to anaemia in children

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

Definition of anaemia

- Is a reduction in the hemoglobin concentration of the blood below the normal range

Classification of anaemias

| | |
|--|---|
| Anaemias of inadequate production | Bone marrow failure syndromes and bone marrow aplasia Nutritional anaemias Anaemia of chronic disease |
| Hemolytic anaemia | Hereditary Membrane defects Enzyme defect Disorders of the structure of hemoglobin Acquired Immune hemolytic anaemia Non immune hemolytic anaemia |

Morphological classification

Microcytic hypochromic anaemia Normocytic normochromic Macrocytic

| | | |
|----------------------------------|----------------------------|-------------------------------|
| Iron deficiency anaemia | Hemolytic anaemia | Vitamin B12 deficiency |
| Beta Thalassemia major and minor | Anaemia of chronic disease | Folate deficiency |
| Anaemia of chronic disease | | Bone marrow failure syndromes |
| Sideroblastic anaemia | | |

Key points in the history

History of the presenting complaint

- Patients will present with the features of anaemia. These include lethargy, poor exercise tolerance, and exertional dyspnoea
- Describe the onset and progression of the symptoms

Is this an isolated anaemia or part of a pancytopenia?

- Ask for history of recurrent infections and bleeding manifestations which are associated with the anaemia

Try to establish the type of anaemia – given below are the key points which should be established in the history

| Nutritional anaemia (especially iron deficiency) | Hemolytic anaemia | Anaemia of chronic disease |
|--|---|---|
| <p>Get a detailed dietary history from the mother. Include the following</p> <ul style="list-style-type: none"> • Breast feeding • Weaning • 24h dietary recall of the present diet • Present eating practices of the child <p>Ask for history suggestive of chronic blood loss</p> <ul style="list-style-type: none"> • Ask for history of malaena • Past history of worm infestation and treatment | <p>Ask for history of episodes of anaemia, yellowish discolouration of the eyes and darkening of the urine</p> <p>Past history of recurrent blood transfusions and jaundice and blood transfusions at birth</p> <p>Family history of recurrent blood transfusions and anaemia</p> | <p>Ask for past history or symptoms of diagnosed diseases i.e Cardiac disease, CRF, JIA, chronic infections</p> |

- Menstruation in older children
- Ask for drug therapy with gastric irritant drugs

Diseases of malabsorption

Other routine components of the history

Social history

- This is extremely important. Take the usual social history but pay more attention on the living environment and socio economic status of the family

Examination

General examination

- Look for pallor and Icterus
- Look for the features of nutrient deficiency – especially iron deficiency
- Look for the facial features of thalassemia
- Look for other dysmorphic features on the general examination – these could indicate some other rare inherited causes of anaemia (i.e. Fanconi anaemia)
- Examine the skin for purpura and petichiae – pancytopenia
- Look for lower limb ulcers – sickle cell anaemia and thalassemia

Abdomen

- Look for hepatosplenomegaly – hemolytic anaemia

CVS

- Listen for a soft systolic flow murmur over the pulmonary area
- Look for evidence of heart failure in severe anaemia

Discussion

How will you investigate a child with anaemia?

| Investigation | Importance in the diagnosis |
|---------------|--|
| FBC | Confirmation of anaemia by the Hb concentration Excludes a pancytopenia |

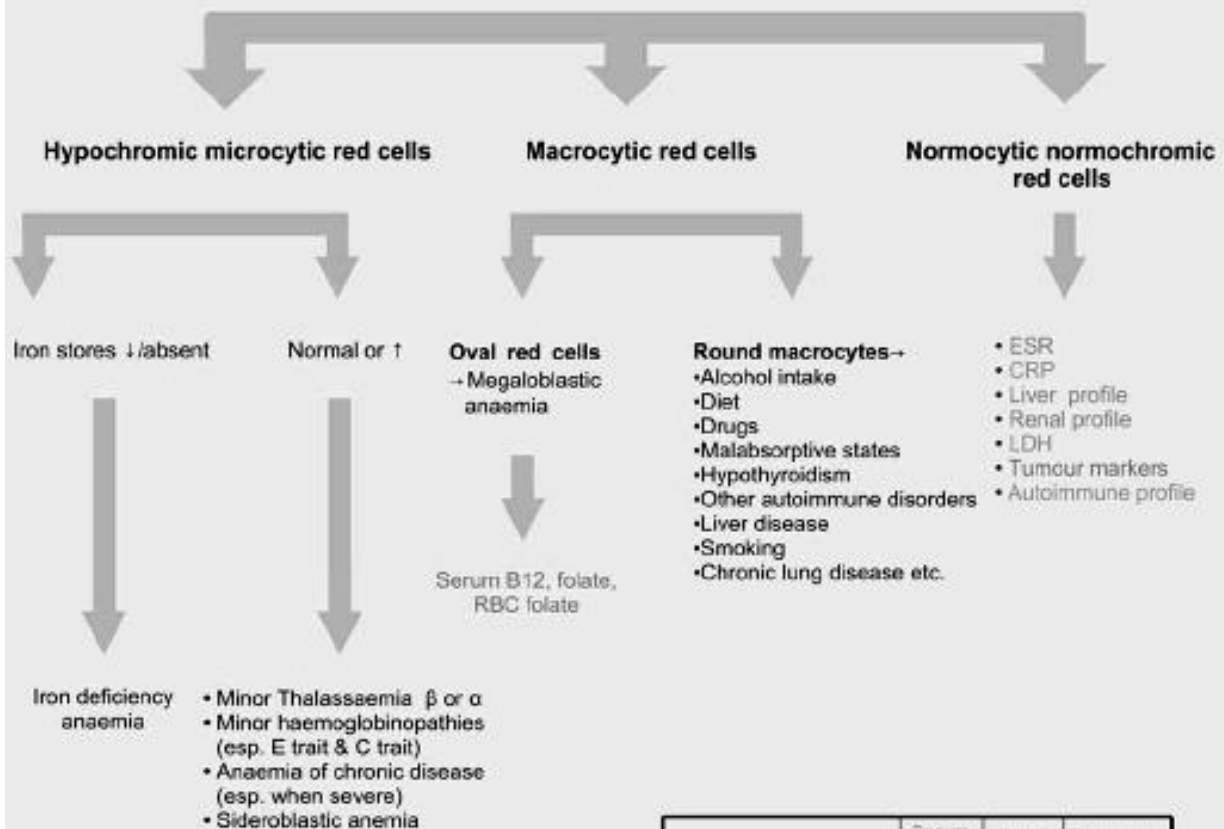
| | |
|-----------------------------|--|
| Red cell indices | Acts as guide to classify anaemia based on morphology |
| Red cell distribution width | Is a quantitative assessment of the various sizes of RBC in the blood |
| Peripheral blood smear | Establishes the morphology into microcytic hypochromic, normocytic normochromic and macrocytic Inspection of individual cells may also reveal the diagnosis |
| Reticulocyte count | Decreased in iron deficiency anaemia and increased in hemolytic anaemia |

The subsequent investigations will be based on the morphology of the anaemia

GUIDELINES TO INVESTIGATE NON HAEMOLYTIC ANAEMIA

First line investigations

- Full Blood Count (FBC) to be reported by a Medical Officer
- Blood picture
- Reticulocyte count



| | Serum ferritin | TIBC | Serum Iron |
|--|----------------|--------|------------|
| Iron deficiency | ↓ | ↑ | ↓ |
| Thalassaemia trait & minor haemoglobinopathies | Normal | Normal | normal |
| Anaemia of chronic disorder | ↑ | ↓ | ↓ |
| Sideroblastic anaemia | ↑ | normal | ↑ |
| Iron deficiency with inflammation | ↑ | ↑ | ↓ |

INVESTIGATION OF HAEMOLYTIC ANAEMIA

First line investigations

Full Blood Count with indices
Reticulocyte count / Absolute Reticulocyte count / Reticulocyte index
Urino urobilinogen,
Serum haptoglobin/ haemopexin/ Urine Haemoglobin & Haemosiderin
Blood picture
[to be reported by a medical officer]
Serum bilirubin

Spherocytes

DAT

Positive
• Investigate for Autoimmune Haemolytic anaemia & other causes

Elliptocytes

- Membrane studies
- Family screening

Agglutinates

- Direct coombs test
- Mono-specific test
- Cold agglutinin titre
- Look for aetiology
- Bone marrow if indicated

Negative

- Osmotic Fragility test
- Glycerol Lysis test
- Cryohaemolysis
- Cell membrane protein electrophoresis
- Family Screening

Haemoglobinopathy features

- Hb electroporesis [acid & alkaline]
- Sickling test
- HPLC
- Isopropanyl test.
- Heat stability test.
- Heinz bodies illustration
- Acid elusion test.
- Alkaline denaturation & Hb F estimation
- Quantitation of haemoglobin variants.
- Isoelectric focusing

Fragmented cells

G6PD deficiency

- Brewer's test (When Reticulocyte count is normal)

- Florescent screening test for G6PD Heinz bodies

PK deficiency

- Pyruvate kinase assay

Microangiopathic anaemias

- Coagulation screening

- D-Dimers/FDP

- Renal profile

- Liver profile

Drug induced & other acquired causes

Other causes

- Infections

- Physical/ Chemical / mechanical damage to red cells.

Management of iron deficiency anaemia

- After the diagnosis is made the following principles are used in the management
- **Treat the underlying cause**
 - Worm treatment
 - Management of chronic gastrointestinal bleeding
- **Consider blood transfusion if the anaemia is severe**
- **Dietary management**
 - Add iron rich foods to the child's diet. The following foods are a good source of iron in the Sri Lankan diet
 - Meat
 - Eggs
 - Fish – tuna, skip jack, hurulla, salaya, dried sprats and other dried fish
 - Pulses – Cowpea, mung, ulundu, bean sprouts, soya and soya based products
 - Dark green and other green leafy vegetables – thampala, sarana, kankun, mukunuwenna, gotukola
- **Other dietary advice**
 - Add sources of vitamin C to the diet as this increases the absorption of iron. Avoid giving tea to children as this can decrease the absorption of iron
- **Consider iron supplementation**
 - 4-6mg/kg of elemental iron in 3 divided doses daily
 - Various preparations of iron available – ferrous sulphate, fumarate, gluconate, iron polymaltose complex
 - Side effects mainly affect the gastrointestinal system – educate the mother
- **Follow up of the response**
 - Initially the reticulocyte count will pick up (peak at 5-7 days)
 - The Hb will return to normal after 4-30d
 - Stores will be repleted only after 1-3 months

Thalassemia

Key points in the history

Presenting complaint

- The most common reason for admission will be for routine blood transfusion

History of the presenting complaint

- When was the diagnosis made?
- What were the presenting features at that time?
- Describe the investigations performed on the child and also state any other special investigations such as genetic screening

Describe what has happened up to now in a chronological order

Blood transfusions

- State when the child was started on regular blood transfusions
- How has the frequency of blood transfusions changed over time? State the present frequency of transfusions
- Has the child developed any reactions to the blood transfusions?

Iron chelation therapy

- State when this was started and the indication if possible
- Describe the method of administration

Splenectomy

Complications of the disease over time

- Complications due to the disease itself
 - State any hospital admissions where the child has been admitted with severe anaemia +/- heart failure
 - History of bone pain and fractures
 - Recurrent infections and bleeding manifestations due to hypersplenism
- Complications of iron overload
 - Cardiomyopathy – Ask for symptoms of heart failure, palpitations and syncopal attacks
 - Liver disease – Ask for history of hematemesis and malaena and hepatic encephalopathy
 - Diabetes mellitus – Polyuria, polydipsia
 - Hypothyroidism – Ask for features of hypothyroidism
 - Reproductive – Ask if the menstrual cycles have commenced in if the patient is a girl

- Complications of iron chelation therapy
 - Fever, sore throat, diarrhoea
 - Rashes and allergic reactions
 - Poor vision and night blindness
 - Hearing impairment

Describe the follow up of the patient

- State when the last of the following investigations have been done
- Echo, FBS, thyroid profile, liver function tests, eye and ear referral

Family history

- Consanguinity
- Area of origin
- Family history of similar illness

Social history

- This should follow the usual format of taking a social history
- Impact on the child
- Impact on the parents
- Impact on the siblings
- Impact on the family life and social withdrawal of the family from society
- Socioeconomic details of the family and the living environment
- Support available
- Psychological state of the child and the parents
- Expectations for the future
- Family planning

Examination

General examination

- Anthropometric measurements – weight, height
- Pubertal classification – Tanner’s staging
- Face – Look for the typical thalassemic facies with frontal bossing, flat nasal bridge and maxillary hyperplasia
- Pallor and Icterus
- Look for the stigmata of chronic liver disease
- Look for pigmentation of the skin

Abdominal examination

- Look for scars – splenectomy and desferrioxamine injection scars
- Hepatosplenomegaly

Cardiovascular system

- Look for evidence of cardiomyopathy and heart failure

CNS

- Look for slow relaxing ankle reflexes which are associated with hypothyroidism

Neonatal sepsis

Diagnosis

History

The presentation of neonatal sepsis is usually non specific. Look for risk factors for sepsis in the history. These are

- Preterm
- IUGR
- PROM +/- ascending infection and chorioamnionitis
- Past history of GBS infection
- Infections in the mother especially STD

Following are the possible presentations of neonatal sepsis

- Poor feeding
- Decreased level of activity and lethargy
- Irritability
- Seizures
- Respiratory distress and apnoeic attacks

On examination

- Measure the temperature of the child and plot on a temperature chart – they can have hypothermia or hyperthermia
- Measure the weight, length and OFC of the child. The initial measurements are used as a baseline value
- Look at the general condition of the child and the colour
- Look at the vital parameters CRFT, heart rate and respiratory rate
- Examine the fontanelles
- Look for a focus of infection – eyes, ear discharge, umbilical discharge/ Erythema, rashes
- Examine the abdomen for hepatosplenomegaly

How would you investigate this child?

- **Full blood count**
Look for low platelets, high WBC with neutrophil leukocytosis (more than 25,000 total count) or low WBC (less than 7000) with neutrophil predominance
- **CRP**
- Blood culture
- Urine culture
- Lumbar puncture with CSF full report and culture

- Blood glucose
- Swabs may be taken if there is obvious discharge but are not routinely taken. Deep ear swab may be taken in fresh babies up to 24 hours

How would you manage this baby?

- Consider admitting the child to the SCBU or NICU based on the clinical condition