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

Conjuctivae 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

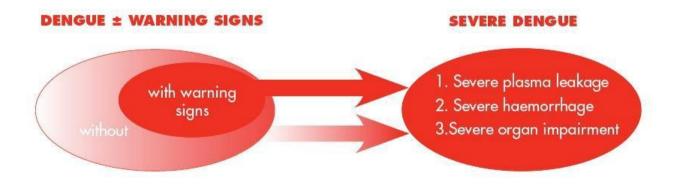
Musculoskeletal – Look for joint swelling and other features of acute inflammation of the joint

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 |
| Nausea, vomiting | Mucosal bleeding | respiratory distress |
| Arthralgia and myalgia | Clinical evidence of fluid | |
| Rash | accumulation | Severe bleeding |
| | Liver enlargement >2cm | |
| Positive tourniquet test | | Severe organ involvement |
| Leucopenia | | Hepatitis |
| | Dropping platelets and rising | Myocarditis |
| | hematocrit | Encephalitis |

How would you perform the tourniquet test?

Tourniquet test

The technique may be asked during the discussion

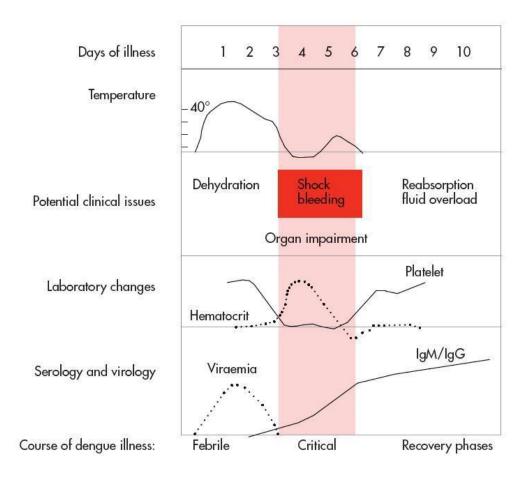
First measure the systolic and diastolic blood pressures

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 | General improvement of well being |
| Fever Flushed Arthralgia and myalgia | from the 3 rd day of fever Fever subsides Leakage of fluid occurs | Good appetite Diuresis |
| | 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 | Respiratory tract | Cough, sputum, nasal or ear discharge, sore |
| Localized | infections | throat |
| | Gastrointestinal | Ask for alteration of bowel habits, |
| | infections and localized | recurrent episodes of abdominal pain |
| | intra abdominal | |
| | abscesses | |
| | Urinary tract infections | Dysuria, frequency, hematuria and other |
| | | urinary tract symptoms |
| | Infections of the bones | Ask for joint pain and swelling, limping, |
| | and joints | |
| Generalized | Infective endocarditis | Past history of heart disease, rheumatic |
| | | fever with evidence of a predisposing |
| | | event for bacteraemia |
| | IMN | Associated sore throat |
| | ТВ | 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 | Contact history with animals |
| | infections | |
| 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 | Evidence of bleeding, ask for the features |
| | malignancy | of anaemia, history of bone pain, past |
| | Oth on weather a single | history of recurrent infections |
| Other | Other malignancies | D. Maria |
| Other | Drugs | Drug history |
| | Factitious fever | |

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 ferretin 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

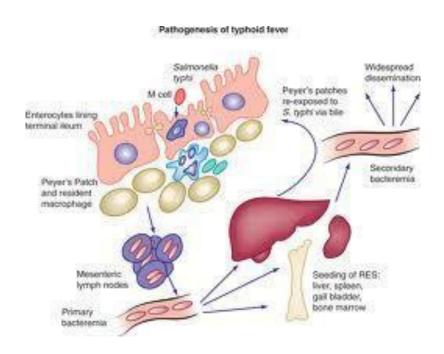
Systemic emboli

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 $\mathbf{3}^{\text{rd}}$ 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 oligaemia

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 | Symptom pattern (most of these will have been described |
| | above) |
| | Intermittent symptoms (the child will be well in between |
| | episodes |
| | Diurnal variation of symptoms may be present |
| | Definite trigger factors for the episodes and good response to medication |
| | Ask for family history of atopy and asthma |
| Structural anomalies/ congenital lesions of the respiratory | This will be excluded as the onset of symptoms is later on in |
| tract | 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, cilliary dyskinesia, | Recurrent lower respiratory tract infections, chronic sinus |
| immunodeficiency | 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 | Describe the layout of the house | |
| | Describe the surrounding environment | |
| | Main roads, dirt and dust, factories | |
| | Describe the house | |
| | 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 | Family support | |
| | Extended family support | |
| | Medical facilities available | |
| Education of the parents | 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 - Cushingnoid 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



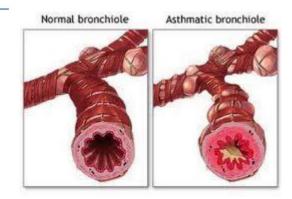
Bronchial asthma

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

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

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 | years) MDI with spacer/DPI | |

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 month s – 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 pnemoniae, Staphylococcus aureus | |
| | Chlamydia | |
| 1 – 5 years | Viral RSV, parainfluenza, influenza, adenovirus | |
| | | |
| | Bacterial | |
| | Streptococcus pneumoniae, Staphylococcus | |
| | aureus, Haemophillus 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 | High grade fever | Low grade fever |
| | (<38.5) | (>38.5) | Associated wheeze |
| | Respiratory rate normal | Respiratory rate high | Prolonged disease |
| | or slightly raised | | course |
| | Wheezing | No wheezing | Prominent headache, |
| | Marked chest wall | Chest wall recessions | arthralgia, myalgia |
| | recessions | | Extra pulmonary |

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

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

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

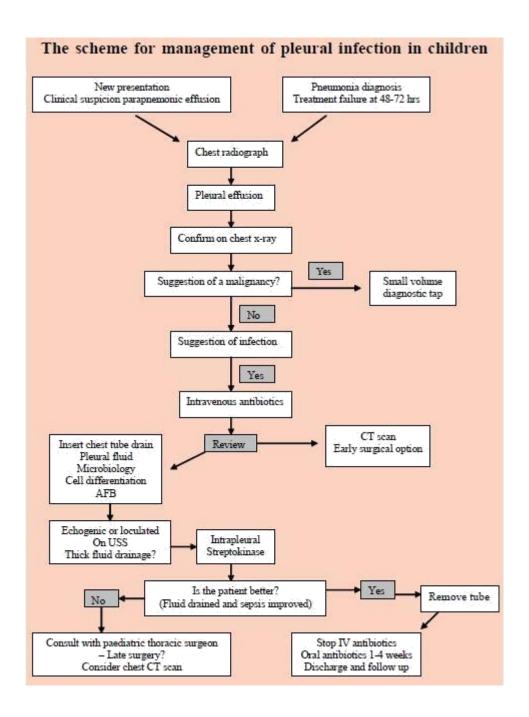
Look for pleural effusion, worsening of the infiltrates, foreign bodies, and features suggestive of atypical pneumonia

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



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 |
| | AAtttt. | A d |
| | 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 – Intusussception

Discussion

Dysentery

What are the causes of dysentery?

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

Prostitic and rostal prolon

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

i iigii icvci

Blood stained stools Poor oral intake

Features of dehydration and over hydration

Edema and nephrotic syndrome

Key points in the history -1^{st} 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 (whether edema worse in the evening)

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 |
| | | Ask for associated red coloured urine |
| | | and documented elevated blood |
| | Nephritic syndrome | pressure (ask the mother if she was |
| | | informed about elevated blood |
| | | pressure) |
| | | Ask for weakness, easy |
| | | fatigue(associated anaemia) and |
| | Chronic renal failure | 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

<u>Anthropometry</u> – 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 (>40mg/m²/h or urine protein to creatinine ratio >200mg protein/mmol creatinine Hypoalbuminaemia (<2.5g/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

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

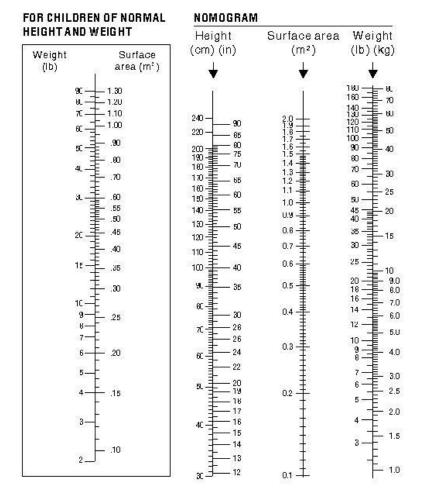
Calculation of the body surface area should be done using the normogram which is available in the ward

Usually respond to steroids after 2-4 weeks

Remission

Urine protein <4mg/m²/h

Urine protein negative or trace for 3 consecutive days



BODY SURFACE AREA FORMULA (Adult and Pediatric)

$$B\,SA\,(m^2) = \sqrt{\frac{Ht\,(in)\,\,x\,Wt\,(ib)}{3\,13\,1}} \ \, \text{or, in metric:} \,\, BSA\,\,(m^2) = \sqrt{\frac{Ht\,(om)\,x\,Wt\,(kg)}{3\,60\,0}}$$

References

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

Moreller, DD, "Simplified Calculation of Body Surface, Appl," N Engl, (Med, 1997).

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 prednisilone

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/m²/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 | Relapses occur in 60- 70% of children with nephrotic syndrome Manage the 1 st and 2 nd 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 Relapse while on Prednisolone | Two or more relapses in the first 6 months after diagnosis OR Four relapses in any 12 month period | 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 Levimasole |
| nelapse willie on Freumsolone | | Other drugs Cyclophosphamide |
| Steroid dependent nephrotic | 2 consecutive relapses while on steroids or relapse within 14 days of cessation of therapy | |

| Drug | Side effects |
|------------------|--|
| Levimasole | 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 is 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

Frusemide 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 | Oral medication |
| | without severe symptoms or | Oral nifedipine |
| | evidence of target organ damage | |
| Hypertensive emergency | Elevation of blood pressure with | ABC |
| ,, | • | |
| ,, | target organ damage | IV antihypertensives – IV |
| | • | IV antihypertensives – IV hydralazine |

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 or 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 5^{th} day of prophylaxis

Antibiotics used for prophylaxis are given below. These drug 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) | Under 1 year All children with a febrile UTI | Looks for renal scarring |
| , | Under 5 years Clinical picture suggestive of acute pyelonephritis Recurrent febrile UTI Abnormal USS | |
| MCUG | Suspected bladder outlet obstruction When the USS reveals hydronephrosis and hydroureter Recurrent UTI | Used to diagnose posterior urethral valves and VUR |
| DTPA | Suspected PUJ or VUJ obstruction | Used to diagnose PUJ and VUJ obstruction Can also give an idea about the differential renal function |

Prevention of recurrent attacks of UTI – Advise the parents

Educate the parents on the condition the child is having an 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

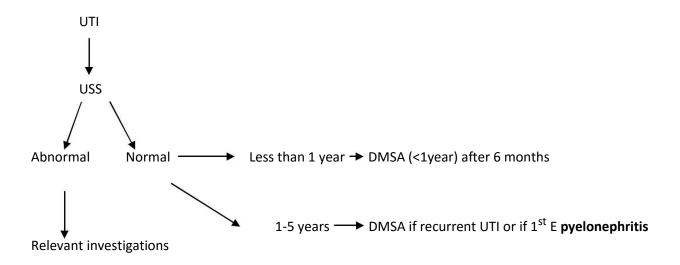
Good hydration

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 |
|-----------------|------|---|
| | | Reimplantation is performed |
| PUJ obstruction | DTPA | 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 Ask for associated backache, |
| | Transverse myelitis | 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 | Exposure to toxins, use of long term |
| | peripheral neuropathy | 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 | Muscle pain, associated skin rashes |
| | Periodic paralysis | |
| | | |

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 |

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 out break 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 of 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

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 Irritabilty, lethargy Seizures | Fever Irritabilty, lethargy |
| Examination | Bulging fontanelle (Should be examined in the upright position when the child is not crying) Opisthotonus | Bulging fontanelle Brudzinki'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 | ТВ |
|--------------------|---------------------------|-----------------------|---------------------------|------------------|
| Appearance | Clear | Turbid | Clear | Turbid, may clot |
| | | | | on standing |
| Cells | 15-2000 | 10-10000 | 5-10000 | 10-500 |
| Differential count | Lymphocytes | Neutrophils | Monocytes or neutrophills | 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, E. coli, listeria | Ampicillin or benzyl penicillin + cefotaxime |
| 1-2 months | Neonatal organisms, haemophillus, pneumococcus, meningococcus | Ampicillin or benzyl penicillin + cefotaxime |
| 2 months – 5 years | haemophillus, pneumococcus, meningococcus | Cefotaxime/ ceftriaxone |
| >5 years | pneumococcus, meningococcus | 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 *Haemophillus 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

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 | Consciousness is impaired |
| | 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 | Automatisms are commonly associated – prolonged and repetitive lip smacking, chewing, swallowing and excessive salivation May also have gestural automatisms which involve alteration of behaviour |
| | Sensory presentations may also occur | 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 | Transient loss of consciousness | |
| | Urinary or faecal incontinence Tongue biting, frothing from the corner of the mouth | | |
| 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 Hypsarrythmias usually occurs on waking | |
| 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 |
|------------------|--|------------------------------------|--|
| Carbamazapine | 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, hirsuitism, 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 | Topiramate |
| | Ethosuximide | Lamotrigine |
| Myoclonic | Sodium valproate | Clobazam |
| | Lamotrigine | Clonazepam |
| Infantile spasms | ACTH | |
| | Prednisilone | |
| Focal | Carbamazapine | Lamotrigine |
| | Sodium valproate | 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

| Iron deficiency anaemia | Hemolytic anaemia | Vitamin B12 deficiency |
|----------------------------|----------------------------|-------------------------------|
| Beta Thalassemia major and | Anaemia of chronic disease | Folate deficiency |
| minor | | |
| 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 |
|---|---|--|
| Get a detailed dietary history from the mother. Include the following | Ask for history of episodes of anaemia, yellowish discolouration of the eyes and darkening of the urine | Ask for past history or symptoms of diagnosed diseases i.e |
| Breast feeding Weaning | and darkening of the drine | Cardiac disease, CRF, JIA, chronic infections |
| 24h dietary recall of the present diet | Past history of recurrent blood transfusions and jaundice and blood transfusions at birth | |
| Present eating practices of the child | biood transfusions at birth | |
| Ask for history suggestive of chronic blood loss | Family history of recurrent blood transfusions and anaemia | |
| Ask for history of malaena | | |
| Past history of worm infestation and treatment | | |

Menstruation in older children

Ask for drug therapy with gastric irritant drugs

Diseases of malabsoption

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

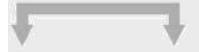
- · Full Blood Count (FBC) to be reported by a Medical Offier
 - Blood picture
 - * Reticulocyte count



Hypochromic microcytic red cells

Macrocytic red cells

Normocytic normochromic red cells







· Liver profile

· Renal profile

Tumour markers

. ESR

· CRP

· LDH

Iron stores 1/absent





Serum B12, folate, RBC folate

Round macrocytes-

- ·Alcohol intake
- Diet
- Drugs
- Malabsorptive states
- Hypothyroidism
- ·Liver disease
- Smoking

- •Other autoimmune disorders Autoimmune profile
- ·Chronic lung disease etc.

Iron deficiency anaemia

Minor Thalassaemia β or α

Normal or 1

- · Minor haemoglobinopathies (esp. E trait & C trait)
- · Anaemia of chronic disease (esp. when severe)
- Sideroblastic anemia

| | Serum femilin | TIBC | Serum Iron |
|---|------------------|--------|------------|
| Iron deficiency | 1 | 1 | 1 |
| Thalassaemia trait & minor haemoglobinopathies | Nomal | Normal | normal |
| Anaemia of chronic disorder | 1 | 1 | 1 |
| Sideroblastic anaemia | 1 | normal | 1 |
| Iron deficiency with inflammation | 1 | 1 | 1 |

INVESTIGATION OF HAEMOLYTIC ANAEMIA

First line investigations

Full Blood Count with indices
Reticulocyte count / Absolute Reticulocyte count / Reticulocyte index
Urine urobilinogen,

Serum haptoglobin/ haemopexin/ Urine Haemoglobin & Haemosiderin Blood picture

> [to be reported by a medical officer] Serum billirubin





DAT





• Family screening



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



- Hb electroporesis [acid & alkaline]
- · Sickling test
- · HPLC
- · Isopropanyi test.
- · Heat stability test.
- Heinz bodies
 Illustration
- · Acid clusion test.
- Alkaline denaturation & Hb F estimation
- Quantitation of haemoglobin variants.
- + Isoelectric focusing



G6PD deficiency

- Brewer's test (When Reticulocyte count is normal)
- Florescent screening test for G6PD Heinz bodies

PK deficiency

 Pyruvate kinase assasy

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.

Positive

 Investigate for Autoimmune Haemolytic anaemia & other causes

Negative

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

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

Diagnosis

History

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

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Preterm
IUGR

PROM +/- ascending infection and chorioamnionitis Past history of GBS infection
Infections in the mother especially STD
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Following are the possible presentations of neonatal sepsis

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Poor feeding

Decreased level of activity and lethargy
Irritability

Seizures

Respiratory distress and apnoeic attacks
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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

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Blood culture
Urine culture
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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