

Contents

Exam (1) 20-6-2010 New System -----	2
Exam (2) 20-6-2010 Old System -----	16
Exam (3) 29-8-2010 New System -----	25
Exam (4) 29-8-2010 Old System -----	35
Exam (5) 3-7-2011 New System -----	45
Exam (6) 4-7-2011 Old System -----	56

Prepared by: Dr. Ahmed Badr

Edited by: Medad Team

Exam (1) 20-6-2010 New System

Q1) Describe the diagnosis and treatment of neonatal sepsis?

* Neonatal septicemia is a very common neonatal problem and the most common cause of death in neonates. According to onset, there are 2 types (early sepsis and late sepsis):

- 1) Early sepsis: which occurs in the first 72 hours of life, is usually due to nosocomial or hospital infection.
- 2) Late sepsis: occurs after 72 hours of life.

* The common pathogens of the neonatal period in Egypt are the gram negative organisms (Klebsiella, Pseudomonas, Proteus and Escherichia coli), then followed by gram positive organisms (Staphylococcus aureus and Group B Streptococcus).

* Diagnosis of neonatal septicemia needs a high index of suspicion, as the clinical manifestations are usually vague and non specific. It is important to know that diagnosis of neonatal septicemia is a clinical diagnosis, which can be confirmed by laboratory investigations. It is also important to note that positive blood culture is not necessary for diagnosis as it is only positive in 50% of cases.

* Clinical & laboratory findings of neonatal septicemia:

- Suggestive clinical finding >>

- 1) Weak suckling, vomiting and poor feeding (very important).
- 2) Unstable body temperature (fever or hypothermia).
- 3) Tachypnea, apnea or irregular breathing.
- 4) Early and prolonged jaundice.
- 5) Abdominal distension.
- 6) Mottling of the skin and poor refill time.
- 7) Sclerema (skin hardening).

- Confirmatory laboratory finding >>

- 1) Complete blood picture: leukocytosis or leukopenia and bandemia (high band count above 10%).
- 2) Positive C reactive protein (CRP) and high sedimentation rate (ESR).
- 3) Positive blood culture (only in 50% of cases).

- 4) CSF examination should be also done to exclude meningitis.
- 5) Swab from eyes, ear ,throat ,umbilicus and rectum as part of sepsis workout.

* Serious complication may occur in severe cases and can lead to death, as:

- 1) Spread of infection: meningitis, pneumonia, osteomyelitis and arthritis.
- 2) Septic shock: tachycardia, poor peripheral perfusion and hypotension.
- 3) Acute renal failure.
- 4) Disseminated intravascular coagulation (DIC): bleeding and purpura.

* Treatment of septicemia:

- 1) Urgent hospitalization.
- 2) Immediate investigations: CBC, CRP, ESR, CSF examination and blood culture.
- 3) Immediate parenteral combined antibiotic therapy:
 - While awaiting the result of the investigations, the ill baby should be started immediately on an antibiotic combination parenterally to cover most of gram negative and positive organisms.
1. Widely used combination are gentamicin and penicillin G in the first week of life.
2. Gentamicin and fluxacillin are used thereafter.
3. Third generation cephalosporins may be also used in severe cases.
- 4) When the result of the septic screen is known, the particular organism may be treated more thoroughly in accordance with the antibiotic sensitivities found.

Q2) List causes and differential diagnosis of acute cough in an infant.

* Acute cough duration less than 2 w.:

- Without respiratory distress: acute bronchitis, acute laryngitis and acute sinusitis.
- With respiratory distress: acute bronchiolitis, pneumonia and acute asthmatic attack.

1) Acute bronchitis:

- Clinical picture: it is mostly preceded by nasopharyngitis, few days before the onset.
- The onset is gradual with dry cough and chest discomfort. Fever is common in young children but it may be absent. Persistent high fever should suggest a bacterial origin.

- The course of illness can be divided into 3 stages, each for few days.
- 1. Early stage: during the first few days, the cough is dry, severe, metallic (brassy) and may be spasmodic (tracheitis). During this stage, chest examination is unrevealing and diagnosis depends entirely on the characters of cough.
- 2. Productive stage: during the next few days, cough becomes productive and less severe and the chest becomes rattling. Chest examination reveals palpable rhonchi. Rough breath sounds, expiratory ronchi and moist crepitations.
- 3. Convalescent stage: during the last few days, cough decreases in frequency and severity and chest signs disappear gradually. It is not unusual for simple bronchitis to remain as long as two weeks.
- Bronchitis should not be confused with other causes of cough.

2) Acute bronchiolitis:

- Clinical picture: the course of illness can be divided into 3 stages:
- 1. Prodromal stage: mild URI with nasal discharge and sneezing for few days.
- 2. Respiratory distress and expiratory wheezing: manifestations of respiratory distress (rapid respiration and retraction) with paroxysmal wheezy cough become evident. Chest auscultation reveals expiratory wheezing. This stage usually lasts for few days.
- 3. Rapid recovery usually occurs within few days. The case fatality rate is less than 1 %.

3) Pneumonia:

- Diagnosis of pneumonia: pneumonia should be a possibility in every case of respiratory distress especially when associated with fever and/or cough.
 - Grades:
 - 1) Grade 1: tachypnea (rapid respiration).
 - 2) Grade 2: intercostal and subcostal retractions.
 - 3) Grade 3: expiratory grunting.
 - 4) Grade 4: cyanosis appears.
 - Diagnosis of pneumonia should include the pathological type, the possible causative organism and associated complications.
 - Diagnosis of pneumonia:
- Pathological types:
1. Lobar pneumonia: bronchial breathing over the involved lobe or lobes is the main finding. Some dullness to percussion over the involved lobe may be also present.

2. Bronchopneumonia: fine bilateral consonating crepitation is the main finding.
3. Interstitial pneumonia: severe spasmodic cough and tendency to expiratory wheezing are the main findings.

4) Asthma:

- Diagnosis of asthma:

A) History >>

1. Recurrent episodes of cough and/or wheezes (mainly at night or early morning) that are relieved by bronchodilators.
2. Cough, wheezes, chest tightness precipitated by triggers.
3. Colds that go to chest & persists for 10 days or longer.
4. Other atopic manifestations (e.g. eczema or rhinitis)
5. Family history of atopy.

B) Physical examination.

C) Objective measurement of airflow obstruction (using spirometry).

D) Measurement of atopy (skin prick test, serum total or specific IgE).

E) Others: chest x-ray, eosinophilia.

Q3A) Mention the different types of milk formulas and their clinical use.

* Type of milk to be used: there are several commercially available formulas suitable for healthy or diseased infants. In most situations, the standard humanized formulas should be used. Special formulas for low birth weight babies and for particular diseases states are also available.

* Types of formulas:

1) Standard humanized infant formulas:

- Have cow's milk as a base. Energy content are the same as human milk (67 Kcal/100 ml). Fat is replaced with vegetable oils. Vitamins and minerals are added to approximate the nutritional content of human milk.
- Examples: S.26 milk, Nan milk, Bebelac 1 milk, Nestogen milk, Babysan 1 milk.

2) Lactose-free, soy-based formulas:

- Lactose is replaced by other sugars (sucrose) and proteins are replaced by soy.
- These formulas are used in the following situations:
 1. Lactose intolerance (following severe gastroenteritis).
 2. Galactosemia (in-born error of galactose metabolism).

3. Suspected cow's milk allergy.

- Examples: isomil milk, s.26 LF milk, nursoy milk, bebelac FL milk.

3) Elemental formulas (hypo-allergenic protein hydrolysate formulas):

- Used for infants who are allergic to both cow's milk and soy protein.
- Enzymes hydrolyze protein (casein or whey) to amino acids and small peptides.
- Fat sources vary; some may contain medium chain triglycerides (MCT).
- These formulas are very expensive, taste bad but are essential for some infants (malabsorption syndromes, chronic diarrhea, cystic fibrosis, some allergies).
- Examples: pregestimil milk.

4) Preterm infant formula:

- Used only until the infant achieves the desired weight for hospital discharge.
- These formulas provide more calories (80 Kcal/100 ml) and higher protein.
- Have low lactose (preterm have less lactase enzyme in their GIT).
- Examples: S.26 low birth weight milk, enfalac premature milk.

Q3B) Describe the management outline of a febrile infant or child.

***Management of a febrile infant or child:**

1) General measures:

1. Bed rest, easily digested food and excess fluids.
2. Supplementation with multivitamins in cases with prolonged dietary restriction.

2) Antipyretic measures:

1. Sponging with tap water: it can be effective in lowering the body temperature.
Cold compresses with iced water should be avoided as it increases the peripheral vasoconstriction.
2. Antipyretic drugs: one of several antipyretic drug can be described orally or rectally every 4-6 hours (see below). In case of hyperpyrexia (temperature above 41 C), I.V acetylsalicylic acid can be used.

Antipyretics drugs:

Acetaminophen(paracetamol)	10-15 mg/kg/dose	Oral or rectal
Ibuprofen (brufen)	10-15 mg/kg/dose	Oral or rectal
Acetylsalicylic (aspirin)	10-15 mg/kg/dose	Oral, rectal or parenteral
Metamizole (novalgin)	10-15 mg/kg/dose	Oral or rectal
Mefenamic (ponstan)	5 mg/kg/dose	Oral or rectal
Diclofenac (voltaren)	0.5-1 mg/kg/dose	Oral or rectal
Ketoprofen (ketofan)	0.5-1 mg/kg/dose	Oral

3) Specific treatment:

1. In simple infections (as tonsillitis, otitis media, sinusitis and bronchitis), one oral antibiotic can be used for about 5-7 days.
2. In serious infections (as pneumonia, meningitis, peritonitis and septicemia), parental combined antibiotic therapy (to cover both gram positive and gram negative infections) is essential. Duration of therapy is for at least 7-10 days.

Q4A) List the causes of persistent diarrhea and how to manage each.

* Persistent diarrhea: an acute attack of diarrhea usually lasts between 4-7 days, but sometimes diarrhea lasts longer and persists for more than two weeks. It occurs in 5-20% of acute diarrheal episodes in children. It commonly leads to growth failure, protein energy malnutrition and increase susceptibility to infections.

*** Causes:**

- 1) Sugar intolerance: this is mainly due to disaccharide intolerance. The infectious process may destroy the brush border of the intestinal villi where the disaccharide enzymes are located. Lactase enzyme is most affected, followed by sucrose then maltase.
 - Mechanism of diarrhea: malabsorption of disaccharides will lead to osmotic diarrhea with excretion of disaccharides in the stools. Fermentation of sugars by the intestinal flora will lead to formation of organic acids, which can also induce diarrhea.
 - Diagnosis: watery frothy stools, which contain sugar and acid.

- Treatment: transient elimination of disaccharides from the formula. This is made by prescribing one of the lactose-free formulas for few or several days. (See formula feeding).
- 2) Cow's milk protein allergy: the gastro-intestinal tract is impermeable to proteins after the early days of life. Damage to the intestinal wall by diarrhea can cause intestinal allergy and continuation of diarrhea. The stools characteristically contain mucus and occult or frank blood.
- Diagnosis: only by withdrawal of the offending protein and its reintroduction, this can cause recurrence of symptoms.
 - Treatment: by removing of the offending protein and its replacement by soy protein or protein hydrolysate (soy protein formula).
- 3) Overgrowth of bacteria in the upper small intestine:
- The upper part of small intestine is almost sterile. The fecal types of bacteria (mainly anaerobes and E.coli) do not exist high up in the intestine.
 - After acute diarrhea, the fecal types of bacteria may invade the upper small intestine and multiply leading to damage of the mucosa, diarrhea and sugar intolerance.
- * Mucosal injury and atrophy:
- The presence of offending agents in the intestine (disaccharides, milk protein, bacteria, etc.) will lead to continuous intestinal mucosal injury and atrophy in prolonged cases.
 - The mucosal atrophy will impair the various digestive and absorptive functions of the small intestine and leads to prolongation of diarrhea and a vicious circles is established, which leads to more mucosal injury and the development of malnutrition.
- * Management:
- Treatment of persistent diarrhea is mainly dietetic by:
 - 1) Removing the offending agents from the diet (e.g. Disaccharides and milk ptn).
 - 2) Supplying a lactose free hypo-allergic formula (isomil). Dietetic management should continue for some times after disappearance of diarrhea to allow for mucosal regeneration.
 - 3) Vitamins, especially vitamin A and trace elements are also needed.

Q4B) Describe clinical and laboratory differentiation of the causative agent of viral hepatitis.

* Identification of the causative agent:

1) Clinical differentiation: Accurate clinical differentiation between viral agents is almost impossible. However the following characteristics of different agents could be useful to suggest the causative virus:

- Hepatitis A: it is the most common form. The onset is acute and course is rather short. Fulminate hepatitis is rare and carrier state or chronic liver disease does not occur.
- Hepatitis B: it is characterized by insidious onset and prolonged course. Fulminate hepatitis may occur; carrier state and chronic liver disease are possible.
- Hepatitis C: it is similar to hepatitis B with the difference of being more insidious and more prolonged. Fulminate hepatitis is rare but chronicity occurs in 85% of cases.
- Hepatitis D: it cannot produce infection without concurrent hepatitis B infection. The illness is severer than hepatitis B but the chronicity is less.
- Hepatitis E: it is similar to A with no chronicity or carrier state. It produces fulminate hepatitis in (20%).

2) Laboratory differentiation: Hepatitis markers (antigens and antibodies) of different agents are the only reliable way for differentiation.

- Hepatitis A: anti-HAV antibodies belonging to the IgM class indicate acute disease, whereas anti HAV IgG antibodies persist after recovery.
- Hepatitis B: acute hepatitis B is heralded by appearance of HBsAg, followed by anti-HBc IgM. Recovery and development of immunity is noted by appearance of anti-HBs. in chronic infections, HBsAg persists and anti-HBc igG develops.
- Hepatitis C: anti-HCV antibody denotes exposure to infection but does not denote recovery or development of immunity.

Q5A) Mention the diagnostic work-up of a case with urinary tract infection.

* Diagnostic investigations:

1) Urine analysis (for detection of pyuria):

- Presence of more than 5 WBCs per high power field (HPF).

- Numerous cells are usually present in acute infection. However, it is unreliable because false positive and false negative results are common as:
 1. False negative results: urinary tract infection may be present without pyuria as with:
 - A. Closed infection.
 - B. Obstructive lesions.
 - C. With antibiotic therapy.
 2. False positive results: pyuria may be present without infection as with:
 - A. Febrile illnesses.
 - B. Dehydration.
 - C. Poststreptococcal glomerulonephritis.
- 2) Urine culture (for detection of bacteriuria):
 - It is the only reliable test.
 - Presence of more than one organism in culture indicates contamination.
- 3) Other investigations:
 - Abdominal ultrasound: with suspected pyelonephritis, pyonephrosis.
 - CBC and CRP: with suspected pyelonephritis.
- 4) Investigations of recurrent urinary tract infection:
 - Abdominal x-ray: to exclude radio-opaque urinary calculi
 - Abdominal ultrasound: to exclude obstructive uropathy
 - Intravenous pyelography (IVP): to exclude obstructive uropathy
 - Evaluation renal function: to exclude chronic renal failure.
 - Voiding cystourethrography (important): to exclude vesicoureteral reflux.

Q5B) State early and late manifestation of congenital hypothyroidism.

* Early manifestations (in neonatal period) include one or more of the following:

- 1) Prolonged gestational period.
- 2) Large anterior fontanel.
- 3) Prolonged physiological jaundice.
- 4) Constipation.
- 5) Prolonged sleep and little crying.
- 6) Feeding difficulties.

- 7) Abdominal distension and umbilical hernia.
- 8) Skin mottling.

* Late manifestations (in infancy) are summarized in the following table:

- Coarse features:

- 1) Large head, coarse hair, low anterior hair line, wrinkled forehead.
- 2) Swollen eye lids, depressed nasal bridge, thick protruded tongue.
- 3) Thick skin, broad hand and fingers.

- Other features:

- 1) Anemia, heart murmur.
- 2) Umbilical hernia.
- 3) Growth failure: The child keeps the infantile body proportions with long trunk and short legs.
- 4) Delayed motor development: As delayed head support, sitting and standing.
- 5) Mental retardation: As delayed smiling, laughing and recognition of mother. The infant is frequently apathetic and uninterested in his surroundings.

Q6A) Describe the clinical picture and therapy of immune thrombocytopenic purpura (ITP).

* Clinical picture:

The onset is abrupt with history of preceding viral infection 1-2 weeks before the onset 60-80% of cases.

- 1) Purpura: it is in the form of small pinpoint petechiae to large ecchymoses in skin of the trunk and limbs. It usually fades spontaneously in 2-3 weeks.
- 2) Bleeding: oozing gums, epistaxis, melena or hematuria may occur in severe cases. Serious intracranial hemorrhage occurs only in 1% of cases.
- 3) Anemia: if present, is only related to blood loss.
- 4) Liver and spleen are usually not enlarged.

* Therapy: Spontaneous remission occurs in about 85% of mild cases.

- 1) In mild cases (cutaneous hemorrhage only): Avoid trauma, salicylate and close follow-up
- 2) In moderate cases (muco-cutaneous hemorrhage):
 - 1. Prednisone: 2mg/kg/d. it inhibits antibody synthesis & phagocytic activity. It is continued until platelet count is elevated above 20.000/mm.

2. Intravenous immunoglobulin (IVIG): 0.8-1 gm/kg/d for 2 days, it causes rapid rise of platelet count.
- 3) In severe cases (severe muco-cutaneous hemorrhage or intra cranial hemorrhage):
 1. Platelet transfusion in very severe cases. Sometimes fresh whole blood is needed.
 2. Prednisone.
 3. IVIG.
 4. Plasmapheresis: when other measures fail. It has transient effect.
 5. Emergency splenectomy: when there is no response to other measures.
- 4) In chronic cases (more than 6 months):
 1. Careful evaluation for associated disorder (e.g. SLE).
 2. Trial of other measures (prednisone, IVIG).
 3. Splenectomy in chronic severe cases not respond to other measures.
 4. Immunosuppressive therapy (e.g. azathioprine).

Q6B) List feature of x-linked recessive inheritance and give examples.

*** Characteristic features:**

- 1) The carrier female transmits the abnormal gene to 50% of her daughter (to be carrier) and to 50% for each son (to be affected). This is sometimes referred to as vertical or 'knight's move pattern of inheritance.
- 2) The affected male will transmit the abnormal gene to all of his daughters who will be carrier and to none of his sons who will therefore not be affected, as male-to-male transmission cannot occur.
- 3) Females may be affected in 2 conditions:
 1. When she has inherited 2 copies of the abnormal gene, one from her carrier mother and other gene from her affected father.
 2. When she has turner syndrome (45, X).

*** Examples:**

- 1) Blood diseases: glucose-6-phosphate dehydrogenase deficiency, factor VIII deficiency (hemophilia A), factor IX deficiency (hemophilia B = christmas disease).
- 2) Neurological diseases: duchenne muscular dystrophy, lesch-nyhan syndrom.
- 3) Immunological diseases: bruton agammaglobulinaemia.
- 4) Ophthalmological diseases: red-green color blindness.

Q7A) Define acute respiratory failure and differentiate between its two types.

* Definition: failure of respiratory system to do any of its two jobs:

- 1) Oxygenation of the blood.
- 2) CO₂ elimination from the blood.

* Types:

	Type 1 Lung failure	Type 2 Pump failure
Basic defect	Oxygenation	Ventilation (CO ₂ elimination)
Causes	Causes of respiratory distress	1) Respiratory depression. 2) Respiratory paralysis. 3) Respiratory muscle fatigue (Severe type 1 RF).
clinically	1) Respiratory distress. 2) Chest signs of the cause.	1) Shallow &/or irregular breathing. 2) Coma or paralysis. 3) Severe respiratory distress.
ABG	1) Hypoxia (low PaO ₂) 2) +/- hypoventilation (high PaCO ₂). 3) Acute metabolic acidosis.	1) Hypoventilation (high PaCO ₂). 2) +/- hypoxia (low PaO ₂) 3) Acute respiratory acidosis.

Q7B) Describe the cerebrospinal fluid (CSF) findings in different types of meningitis.

	Normal M.	Bacterial M.	Viral M.	Tuberculous M.
Appearance	clear	cloudy	Usually clear	Opalescent
Cells/mm ³	0-5 lymphocytes	10-100.000 polymorphs	15-2000 lymphocytes	250-500 lymphocytes
Glucose (mg/dl)	40-80	low	normal	Very low
Proteins (mg/dl)	20-40	↑	Normal /mild increased	↑

* Culture & sensitivity study of the CSF are essential to identify the causative organism.

* Antibody & PCR for viral infection is done to exclude viral meningitis and encephalitis.

Q8A) Mention diagnostic features and management of patent ductus arteriosus (PDA) in children.*** Symptoms:**

- 1) Small defect are asymptomatic and a heart murmur is detected routinely or coincidentally.
- 2) Moderate to large defects may lead to:
 1. Recurrent chest infection.
 2. Chronic congestive heart failure (feeding difficulties, exertional dyspnea).

*** Cardiac examination:**

- 1) Loud, contentious murmur heard best below the left clavicle.
- 2) The pulmonary second sound is loud.
- 3) The heart is enlarged clinically with a prominent left ventricle in large defects.

*** Investigations:**

- 1) Chest x-ray shows the typical features of a left-to-right shunt.
- 2) ECG shows left ventricular hypertrophy.
- 3) The diagnosis is confirmed by echocardiography.

*** Treatment:**

- 1) If heart failure is present, medical treatment is necessary.
- 2) Surgery should be carried out as soon as the child's condition allows.

Q8B) Give reasons:

- 1) Mental changes occur constantly in cases of kwashiorkor.
 - Are due to niacin deficiency, disturbed amino acid metabolism and maternal deprivation.
- 2) Karyotyping is indicated for a girl with short stature.
 - Is indicated to diagnose Turner syndrome.
- 3) Checking the femoral pulse is essential during examination of every newborn.
 - So as not to miss coarctation of the aorta.

- 4) Delayed feeding is not recommended during management of acute gastroenteritis.
- As prolonged starvation affects intestinal mucosal cells integrity and delayed repair and may lead to persistent diarrhea.
- 5) Diagnosis of chronic renal failure requires a high index of suspicion.
- Because clinical presentations are usually nonspecific and not directly related to urinary symptoms.

Exam (2) 20-6-2010 Old System

Q1) Describe neonatal reflexes and their clinical significance.

- * There are several reflexes that can be normally elicited in the newborn. Eliciting these reflexes is important for evaluation of general condition, evaluation of vision and detection of focal neurological signs. The reflexes can be classified as tendon reflexes and primitive reflexes.
- * Primitive reflexes are peculiar to newborn and they usually disappear by 4-6 months. Persistence of these reflexes indicates a neurological problem.

1) Moro reflex:

- How to elicit it? Dropping the head with the examiners hand supporting the shoulder or making a loud noise near the baby's ear or sudden withdrawal of the blanket from underneath the infant.
- The response: Abduction of the arms at the shoulders and extension of the forearm at the elbow. The hands and fingers are held wide-open. The arms are then moved as in an embrace. A cry follows these movements and should be vigorous. There is a generalized extension of the trunk.
- Clinical significance of the reflex:
 1. Absence: it may be due to intracranial birth injury, cerebral depression by narcotics or anesthesia given to the mother just before delivery or, prematurity.
 2. Asymmetrical response: it may be due to brachial palsy or fracture clavicle.
 3. Persistence of the reflex: if it is persistent after 6 months of age it.

2) Tonic neck reflex:

- How to elicit it? It is elicited by placing the infant in a supine position and performs a lateral rotation of the head to one side.
- The response: Extension of the arm and leg towards which the head is turned and flexion of the limb on the opposite side. The response is obtained in infants from 1-7 months.
- Clinical significance:
 1. Absence: it may due to spinal cord disease.
 2. Abnormal response: indicates neuro-motor disorder of cerebral origin.
 3. Persistence of the reflex: if it persists, cerebral disease is a possibility.

3) Rooting and suckling reflexes:

- Rooting reflex: the reflex elicited in 4 areas, at both corners of the mouth and on the upper and lower lips and at the midline. The mouth opens or the head turns towards the side of stimulus.
- Suckling reflex: if you introduce the nipple or a finger in the infant mouth, he suckle it. The reflex is important for feeding. Good suckling indicates a good general condition. Absent reflex indicates a serious brain lesion or serious infection.

4) Blinking & papillary reflexes:

- Sudden exposure of the eye to bright light leads to blinking. The reflex is important for evaluation of vision. Again, exposure to bright light leads to papillary constriction while exposure to dim light leads to papillary dilatation. The reflex is present since birth and does not disappear.

5) Grasp reflex:

- If you put your finger in the infant palm, he grasps it. The reflex disappears by 4 months of age. Unilateral absence indicates a focal neurological lesion. Persistence of the reflex indicates an upper motor neuron lesion.

6) Landau reflex:

- When the infant is held prone in a horizontal position the body form a convex arc upwards with head. Trunk and hips flexes and shoulders drawn back. Extension of the head occurs after a month of age.

7) Stepping reflex:

- When the infant is held in upright position and lowered until the feet touch the table. Stepping movement is made.

8) Placing reflex:

- When the dorsal surface of the foot touches the edge of the table, the foot is elevated and placed on the table.

9) Babinski reflex:

- It is positive and disappears at about 1 year. Unsustained ankle clonus (5-6 beats) may normally be present until 2-4 months of age.

Q2) State the indicators of adequate breast milk intake and how to manage minor problems that may occur during breast feeding.

* Indicators of adequate breast milk intake:

1) Weight gain: Baby gains 200 gm/week or 250 gm/10 days in the first 4 months of life.

2) Infant behavior:

1. Baby feeds at least 8 times/day

2. Swallowing noises are heard during feedings.

3. Baby is satisfied between feedings.

4. Baby passes at least 4 soft, yellow stools/day and wets 6 or more diapers/day.

3) Breast fullness:

1. Mother notices that her breast felt full before a feeding and softer afterwards.

2. Some mothers experience a let-down sensation.

* Management of minor problems:

- Some problems may arise during the first few weeks. Sore nipple, mammary engorgement and breast-milk jaundice are the most common. Providing proper advice about these problems is important to ensure successful breast-feeding.

1) Sore nipple: It occurs due to improper attachment to nipple.

- Management is by:

1. Proper positioning and attachment.

2. Begin on less sore side and feed for short period.

3. After feeding, allow air drying of the nipple and apply lanolin cream.

4. Temporary pumping may be needed in severe cases.

2) Mammary engorgement:

- It occurs between 2-6 postpartum days and it may rapidly decrease milk supply. It occurs due to failure of complete evacuation of the breast, which may occur due to infrequent feeding, poor suckling, sore nipple or maternal or infant illness.

- Management by:

1. More frequent feeding.

2. Treatment of sore nipple.

3. Regular breast emptying (by hand expression or breast pumping) to remove excess milk.

3) Breast-feeding jaundice:

- It may be associated with inadequate intake of breast milk. Thus, it requires increasing the frequency of nursing. In severe cases, transient interruption of breast-feeding for 24-36 hours may be necessary. The mother's breast should be emptied (with an electric breast pump) during this period.

4) Insufficient breast milk:

- Management includes:

1. Apply moist hot towels to breast 3-5 minutes before feeding.
2. Massage breast before and during feedings.
3. Express or pump breast between feedings.
4. Mother should eat a nutritious diet and drink fluids.

Q3) Mention the definition, causes and types of cerebral palsy.

- * Cerebral palsy is a motor disability that results from malfunction of the developing brain centers and pathways. It is a non progressive, non-fatal, non-curable disorder.

- * Other manifestations of organic brain damage (as seizures, mental retardation, sensory and learning defects with behavior and emotional disturbances) may be also present.

Causes:*1) Pre-natal causes:**

1. Congenital malformations.
2. Congenital infections: cytomegalovirus, congenital rubella, toxoplasmosis.
3. Severe fetal anoxia (intrauterine).

2) Peri-natal causes (acquired during process of birth):

1. Hypoxic ischemic syndrome (HIS).
2. Birth injuries as intracranial hemorrhage.
3. Prematurity (asphyxia, respiratory complications and intracranial hemorrhage).

3) Post-natal causes:

1. Trauma: accidental or non-accidental.
2. Infections: encephalitis, meningitis.
3. Hypoxia: asphyxia, status epilepticus.
4. Metabolic: kernicterus.

Types:*1) Spastic cerebral palsy:**

- It accounts for 70% of cases.
- It is characterized by clasp-knife hypertonic, abnormally brisk tendon jerks, ankle clonus and extensor planter responses.
- According to distribution of spasticity, there are 3 types:
 1. Hemiplegic type: it affects one side of the body (hemiparesis) and the arms are usually more affected than the legs.
 2. Quadriplegic type: it affects both sides of the body and the arms is more affected than the legs.
 3. Spastic diplegia: both legs are spastic and the arms is less affected or not affected.

2) Atonic cerebral palsy (cerebral infantile hypotonia):

- It accounts for 10% of cases.
- It is characterized by severe hypotonia (floppy infant) with exaggerated tendon reflexes.

3) Dyskinetic cerebral palsy (dystonic or athetoid):

- It accounts for 10% of cases.
- It is characterized by irregular and involuntary movements of some or all muscles groups. Athetosis is the commonest form with purposeless movements.

4) Ataxic cerebral palsy:

- It accounts for 10% of cases.
- It is characterized by ataxia (uncoordinated movements and intention tremors), which is associated with hypotonia.

Q4A) Describe the clinical significance of the growth curve.

* Growth curve (growth charts) are standards for growth of normal infant, children and adolescents.

* They are available in percentile values where:

- 1) 50th percentile represents the average or the mean.
- 2) 25th, 10th and 5th percentile are low normal values.
- 3) 75th, 90th and 95th are high normal values.

- * Types: there are available charts for weight, height and head circumference.
- For each parameter, there are charts for boys and others for girls.
- There are also charts in relation to age, charts for the first 3 years and charts for 2-20 years.

*Uses: growth curve are useful in two ways:

1) With single measurement, values below 5th or above 95th percentile are abnormal.

Abnormalities in physical growth:

	Below 5 th percentile	Above 95 th percentile
Weight	Underweight	Overweight or obesity
Length or height	Short stature	Tall stature
Head circumference	Small head	Large head

2) With repeated or serial measurements, the growth rate can be assessed. Any normal child should follow his own percentile on serial measurements. So, any deviation from the own percentile is abnormal. Acceleration of growth (catch up) occurs with therapy.

Q4B) List non compulsory vaccines against viral infections.

1) Hepatitis A vaccine:

- Type of vaccine: it is a sterile suspension containing inactivated hepatitis A virus.
- Indications: it is given to adult and children above the age of one year especially in areas where hepatitis A infection is common.
- Schedule of vaccination: it consists of 2 doses. The second dose is given 6 month after the first. The dose in children between 1-15 years of age is 0.5ml, intramuscular. In adults, the dose is 1 ml, intramuscular.

2) Chickenpox vaccine:

- Type of vaccine: it is a varicella virus live-attenuated vaccine.
- Indications: it can be given to children above the age of one year in a dose of 0.5ml, I.M. or subcutaneous. The second dose at age 4-6 years.

3) Rabies vaccine:

- Type of vaccine: It is a live-attenuated vaccine.
- Indications: vaccination against rabies is directed to individuals who are subjected to unprovoked bite of domestic or wild animals. Dogs and cats are the main

offenders. When the offending animal is under observation, the vaccination can be withheld until the animal acts abnormally. On the other hand, if the biting animal ran away after an unprovoked bite, immediate vaccination is indicated.

- Schedule of vaccination: human diploid cell vaccine (HDCV) is available. It is given in 5 doses (each is 1ml intramuscular) at 0,3,7,14,28 days.
- Local treatment of the puncture wound is equally important.

4) Seasonal influenza vaccine:

- Type of vaccine: it is an inactivated vaccine, usually trivalent against influenza virus serotypes that are expected in the relevant season.
- Indications: it is indicated in children above the age of 6 months, and adults who are at risk especially those with cardiac diseases, pulmonary, metabolic diseases and immune-deficiency.
- Schedule of vaccination: the dose is 0.5ml intramuscular or subcutaneous for children above 3 years old and only 0.25ml for children between 1-3 years.

5) Rota virus vaccine:

- Type of vaccine: live attenuated vaccine (monovalent or pentavalent)
- Route of administration: oral
- Schedule: 2 doses at age 2 and 4 months (for mono-valent). 3 doses at age 2,4 and 6 months (for penta-valent). The first dose should not be given after age of 15 weeks and the series of vaccines should be completed before 8 months.

Q5A) Mention the characteristics and types of innocent murmurs.

* Innocent murmurs (soft, faint murmurs, mostly systolic, with normal chest x-ray, ultrasound and ECG).

- 1) Venous hum
- 2) Blowing, continuous murmur heard at the base of the heart, often just below the clavicles. It varies both with respiration and the position of the head.
- 3) Pulmonary flow murmur
- 4) Soft, systolic ejection murmur heard in the pulmonary area (second left space).
- 5) Vibratory murmur.
- 6) Short, systolic murmur heard at the left sternal edge or at the apex of the heart. It is variable and changes with position.

Q5B) Enumerate causes and laboratory findings of iron deficiency anemia.

*Causes of iron deficiency anemia:

1) Inadequate iron stores at birth:

1. In premature babies and twin pregnancies.
2. Severe iron deficiency of the mother.

2) Deficient dietary iron:

1. Prolonged breast feeding without supplementary iron-containing food (the commonest cause of iron deficiency anemia in infancy).
2. Protein calorie malnutrition and foods poor in iron are other causes.

3) Impaired absorption of iron:

1. Chronic diarrhea.
2. Malabsorption syndrome.
3. Prolonged use of cow milk.

4) Excessive demands for iron (blood loss):

1. Acute or chronic hemorrhage
2. Parasitic infections with blood loss (ankylostomiasis and bilharziasis).
3. During adolescence especially in girls (menstruation increases the demand).

*Characteristic laboratory findings of iron deficiency anemia:

- 1) Hypochromic microcytic anemia (color index is below one).
- 2) Normal or elevated reticulocytic count. It usually increases with initiation of iron therapy.
- 3) Low serum iron (normal level is 90-150 microgram/dl).
- 4) Low serum ferritin.
- 5) High iron binding capacity (normal level is 250-350 microgram/dl).
- 6) Hyperactive bone marrow (erythroid hyperplasia) with low iron stores.

Q6A) List the complications and cardinal laboratory findings of diabetic ketoacidosis.

* Complications:

1) Shock:

1. Hypovolemic: polyuria and vomiting.
2. Septic infection.

2) Brain edema:

1. Rapid correction of hyperosmolarity and hypovolemia.
2. Ischemic hypoxia (secondary to hypovolemia).

3) Pulmonary edema:

1. Hyperosmolarity.
2. Myocardial heart failure (secondary to acidosis).

4) Cardiac arrhythmias:

1. Hyperkalemia.
2. Hypocalcaemia.

* Cardinal laboratory findings:

- 1) Hyperglycemia (more than 300gm/dl)
- 2) Ketonemia.
- 3) Metabolic acidosis (PH less than 7.3, HCO_3 less than 15mEq/l).
- 4) Glucosuria and ketonuria.

Q6B) Give reason:

1) Chest x-ray is needed in almost every cases of pneumonia.

- Chest x-ray is essential for confirmation of diagnosis, identification of the pathological type and detection of possible complication (effusion or abscess).

2) Consequences of cholestasis include fat malabsorption and pruritis.

- Fat malabsorption Due to decrease bile delivered to intestine, and pruritis due to retention of bile acid.

3) Both parents of a galactosemia patient are expected to have affected gene.

- Because its pattern of inheritance is autosomal recessive, so they are expected to be carriers.

4) Intussusceptions may be confused with a diarrheal illness.

- Because it is associated with abdominal pain, vomiting and blood and mucus in stool with a peak incidence during infancy.

5) Fever in nephrotic syndrome patient can be alarming.

- Because nephrotic syndrome is a febrile disease so presence of fever indicates infection in an immune-compromised [patient with edematous tissue.

Exam (3) 29-8-2010 New System

Q1) Describe differential diagnosis and management of neonatal bleeding.

* Bleeding in healthy newborn:

- 1) Hemorrhagic disease in the newborn (most common).
- 2) Neonatal thrombocytopenia (maternal lupus, maternal IIP, maternal drugs).

* Bleeding in sick newborn:

- 1) Disseminated intravascular coagulations (DIC).
- 2) Neonatal septicemia, neonatal liver disease, hemophilias.

* Investigations of neonatal bleeding include:

- 1) Complete blood picture.
- 2) Prothrombin time, prothrombin concentration, partial thromboplastin time.
- 3) Liver function tests.
- 4) Fibrinogen and fibrinogen degeneration products (in DIC).

* Hemorrhagic disease in the newborn:

- It is the most common cause of bleeding in the neonate. It commonly presents between 2-7 days of life by bleeding from GIT, intracranial hemorrhage, or bleeding following circumcision or from injection sites.
- It occurs due to depletion of vitamin K dependant coagulation factors due to transient deficiency of free vitamin K (due to absence of bacterial intestinal flora normally responsible for its synthesis).
- Prophylactic I.M. vitamin K should be administration, transfusion of fresh frozen plasma or whole blood if needed.

Q2) Mention the differential diagnosis and complications of mumps.

* Complications:

- 1) Meningo-encephalomyelitis: it usually occurs 3-10 days after parotitis with higher incidence in males. There are fever, headache, vomiting, and stiffness of the neck and may be convulsions. Prognosis for recovery is good.

2) Epididymo-orchitis: it occurs in about 25% of adolescent and adults. The affected testis becomes swollen, tender and red. In 3-5 days, the swelling and pain subside. Atrophy and impaired fertility occurs in 13%.

3) Pancreatitis: there is sudden onset of severe epigastric pain and tenderness, vomiting and fever. Elevated serum amylase is characteristic.

* Diagnosis: it is mainly clinical. In acute pancreatitis, elevated serum amylase level will be helpful to avoid unnecessary surgical intervention.

* Differential diagnosis:

1) Cervical lymphadenitis: the cervical glands show a well-defined border, firm and lack the anatomic position of the parotid.

2) Other causes of parotitis as suppurative parotitis and recurrent parotitis.

3) Calculus obstruction or stenosis of the duct : usually the swelling is intermittent.

4) Mikulics syndrome: there is chronic bilateral parotid and lacrimal swelling associated with dry mouth and absence of tears.

Q3A) Describe the clinical features of fallot's tetralogy.

1) Central cyanosis:

- Bluish discoloration of lips, tongue and fingers.
- The onset of cyanosis is delayed (1-2 months after birth).
- In early cases, it is only exertional (only appears during crying).

2) Hypercyanotic spells:

- Attacks of deep cyanosis and respiratory distress for minutes or hours.
- Attack is precipitated by crying, infections or iron deficiency.

3) Clubbing of fingers:

- It is usually not observed before the age of 1-2 years (blue clubbing).

4) Cardiac signs:

- Left parasternal pulsations (denoting right ventricular hypertrophy).
- Systolic thrill over left second and third spaces (in 50% of cases).
- Ejection systolic or pansystolic murmur over pulmonary area.
- The heart is not enlarged (or only mildly enlarged).
- Heart failure is very rare.

5) Possible complications:

- Thromboembolic complications of polycythemia (as hemiplegia).
- Bacterial endocarditis.
- Brain abscess may occur.

Q3B) Describe hepatitis markers (antigens and antibodies).

Hepatotropic viruse	Antigens	Identified antibodies
Hepatitis A virus (HAV)	HAV	Anti-HAV Anti-HAV IgM
Hepatitis B virus (HBV)	HbsAg HbcAg HBeAg	Anti-HBs Anti-HBc Anti-HBc IgM Anti-HBe
Hepatitis C virus (HCV)	-----	Anti-HCV
Hepatitis D virus (HDV)	HDV Ag	Anti-HDV
Hepatitis E virus (HEV)	-----	Anti-HEV, Anti-HEV IgM
Hepatitis G virus (HGV)	-----	Anti-HGV

Q4A) Enumerate factor affecting physical growth.

- * There is a wide range of variations in physical growth among normal infants and children. The factor that explain these variations are:
 - 1) Genetic and hereditary factor: children inherit their height and body frame from their parents. It observed that some families are short and others are tall.
 - 2) Race: there is a racial difference in rate and pattern of growth so it is better for every country to use its own growth chart.
 - 3) Sex: girls grow faster than boys from 7 months to 4 years and also they start and end their puberty at younger age. There are growth curves for boys and others for girls.

- * There are other factors that affect growth and lead to abnormalities:
 - 1) Nutrition: under-nutrition leads to growth failure and over-nutrition leads to obesity. The first sign in protein energy malnutrition is declaration of growth .
 - 2) Socioeconomic factors: children of low socioeconomic classes have poor nutrition, poor hygiene and poor health compared to those of high socioeconomic classes.
 - 3) Chronic illness: chronic systemic diseases as renal, cardiac, gastrointestinal and endocrinal diseases cause growth failure.

Q4B) Describe why fresh animal milk is not suitable for feeding infants below one year of age.

* Fresh animal milk (as cow's milk and buffalo's milk) is not recommended for infants below one year of age for several reasons:

- 1) It has a very high protein content predominantly casein, which forms large curds causing gastric upsets, difficulty in digestion, and may cause intestinal blood loss.
- 2) Cow's milk protein is a famous allergen for predisposing infants.
- 3) Fat globules are larger and more difficult to digest. Animal milk contains a higher proportion of volatile fatty acids, which cause gastric upsets. The amount of essential fatty acids in animal milk is insufficient to maintain optimal neurological growth.
- 4) It lacks oligosaccharides, which are important in resistance against viral and bacterial pathogens.
- 5) Iron content is low in both human and cow's milk; but human milk is superior in the respect due to the presence of lactoferrin (higher bioavailability of iron).
- 6) Calcium content is high, but the Ca/P ratio is not adequate for absorption.
- 7) Sodium content is high, which affects the hydration status of the infant. This problem becomes even more pronounced following boiling (due to water evaporation).
- 8) Zinc is less bio-available than that in human milk.
- 9) It is prone to contamination with a variety of microorganisms, and lacks all the protective mechanisms of human milk.
- 10) It is expensive and liable to adulteration.

Q5A) List causes of macrocephaly.

* Large head or macrocephaly (head circumference above 95th percentile for age and sex).

1) Cranial (skull) causes:

Rickets, achondroplasia, chronic hemolytic anemia, familial.

2) Intracranial causes:

Hydrocephalus, subdural effusion, subdural hemorrhage, brain tumors.

Q5B) Describe laboratory and radiological investigations of pulmonary tuberculosis.

- 1) Tuberculin test: mantoux test is the most important immunological diagnostic tools.
- 2) Sputum examination: direct smear with Zeihl Nelson stains for tuberculous bacilli.
- 3) Stomach wash and gastric aspirations: for a direct smear or culture.
- 4) Biopsy of lymph nodes or pleura: for pathological study.
- 5) Radiological studies: Chest x-ray and chest computed tomography (CT scan)
- 6) Recent methods for diagnosis: usage of ELISA and PCR (polymerase chain reaction).

*** Tuberculin test:**

- 0.1ml purified protein derivative is injected intra-dermally in the skin of the flexor surface of the forearm. The reaction should be read at 48-72 hours. Induration (and not erythema) is measured in mms in the longitudinal as well as the transverse diameters and the mean reading is recorded.
 1. An induration less than 5 mm is considered negative.
 2. An induration measuring 5-9 mm is considered doubtful and should be repeated.
 3. An induration of 10 mm is considered positive.
- Positive tuberculin test: it indicates vaccination or infection:
 1. BCG vaccination: the reaction is never exceeds 15 mm induration.
 2. T.B. infection: it considered in 2 situations:
 - a. Positive reaction in a child less than 5 years who is not BCG vaccinated.
 - b. Strongly positive reaction (over 15 mm) in previously vaccinated child.
- False negative tuberculin test: may happen in the following situation:
 1. Subcutaneous instead of intradermal injection of tuberculin or use of outdated tuberculin.
 2. Recent use of corticosteroids or immunosuppressive.
 3. Intercurrent infections especially viral ones.
 4. Recent antiviral vaccine (especially measles and mumps).
 5. Advanced disseminated T.B. Chronic debilitating disease with cachexia and cell mediated immune deficiency.

Q6A) Describe laboratory diagnostic criteria of type 1 diabetes mellitus.

* Ordinary case:

- 1) Fasting blood glucose.
- 2) Venous sample more than 126 mg/dl.
- 3) Two hours post prandial.
- 4) Venous sample more than 200 mg/dl.
- 5) Random blood glucose sample.
- 6) More than 200 mg/dl (with presence of symptoms of diabetes).

* Diabetic ketoacidosis:

- 1) Hyperglycemia (blood sugar above 300 mg/dl).
- 2) Metabolic acidosis (low PH and bicarbonate).
- 3) Glycosuria and ketonuria.

Q6B) Describe clinical grading of shock.

Grade 1	Early shock	Tachycardia and poor peripheral perfusion
Grade 2	Established shock	Grade 1 + hypotension
Grade 3	Advanced shock	Grade 2 + multiple organ system failure (MOSF)
Grade 4	Irreversible shock	Grade 3 + refractory metabolic acidosis

Q7A) State features and give examples of multifactorial inheritance.

* Multifactorial (polygenic) inheritance:

- The common pattern of inheritance appears from the interaction of a genetic predisposition with adverse environmental factors. The genetic susceptibility is determined by the additive effects of many genes and hence is known as polygenic.

* Characteristic features:

- 1) These disorders are much commoner than single gene disorders.
- 2) The recurrence risk is much lower than that of single gene disorders (only 2-5%) and it declines sharply, as the relationship with the affected individual becomes more distant.

* Examples:

1) Congenital malformations of infancy:

1. Cleft lip/palate.
2. Pyloric stenosis.
3. Congenital heart disease.
4. Congenital dislocation of the hip and neural tube defects.

2) Acquired disorders of childhood and adult life:

1. Bronchial asthma.
2. Diabetes mellitus.
3. Epilepsy.
4. Hypertension.

Q7B) State investigations and treatment of recurrent urinary tract.

* Diagnostic investigations:

1) Urine analysis (for detection of pyuria):

- Presence of more than 5 WBCs per high power field (HPF)
- Numerous cells are usually present in acute infection. However, it is unreliable because false positive and false negative results are common.

2) Urine culture (for detection of bacteriuria):

- It is the only reliable test.
- Presence of more than one organism in culture indicates contamination.

3) Other investigations:

- Abdominal ultrasound: with suspected pyelonephritis, pyonephrosis
- CBC and CPR: with suspected pyelonephritis.

4) Investigations of recurrent urinary tract infection:

- Abdominal X-ray: to exclude radio-opaque urinary calculi.
- Abdominal ultrasound: to exclude obstructive uropathy
- Intravenous pyelography (IVP): to exclude obstructive uropathy
- Evaluation of renal function: to exclude chronic renal failure
- Voiding cystourethrography (important): to exclude vesicoureteral reflux.

* False negative results: urinary tract infections may be present without pyuria as with:

- 1) Closed infection.
- 2) Obstructive lesions.
- 3) With antibiotic therapy.

* False positive results: pyuria may be present without infection as with:

- 1) Febrile illness.
- 2) Dehydration.
- 3) Post streptococcal glomerulonephritis.

* Treatment:

1) Treatment of pyelonephritis:

1. Hospitalization and combined I.V. antibiotic therapy: Ampicillin (50-100 mg/kg/day) and gentamicin (4 mg/kg/days) is preferable.
2. Antibiotic therapy can be adjusted according to the results of urine culture-sensitivity studies.
3. Urine should be sterile within 48 hours of adequate therapy.
4. Antibiotic may be given orally after about 5 days of I.V. therapy, if the patient's general condition has improved with absence of vomiting.
5. The total duration of treatment should extend for 10-14 days.

2) Treatment of cystitis:

- These patients can be managed at home with one oral antibiotic for 7-10 days.

There are several choices:

1. Co-trimoxazole (trimethoprim-sulfamethoxazole) is effective against most strains of E.coli and many other gram negative organisms and is considered the initial drug of choice.
2. Amoxicillin (50 mg/kg/day) is also an effective initial oral therapy but it has no advantage over co-trimoxazole. Treatment can be also adjusted according to the result of urine culture-sensitivity.
3. A second generation cephalosporin (as cefclor, 50 mg/kg/day) may be also used.

3) Treatment of frequent recurrences:

- They needed long-term prophylaxis with oral drug as co-trimoxazole (trimethoprim-sulphamethoxazole). The dose in this case is only one third the therapeutic doses.

1. Broad-spectrum antibiotics as ampicillin are not useful for prophylaxis

2. Adequate fluid intake, frequent voiding, avoidance of constipation and proper hygiene are important measures to prevent UTI.
3. Surgical interference is indicated in presence of stones or vesicoureteral reflux.

Q8A) Compare between hypertonic and hypotonic dehydration.

Types	Hypernatremic (hypertonic)	Hyponatremic (hypotonic)
Serum Na (mEq/l)	More than 150	Less than 130
Incidence	15%	10%
Water loss /Na loss	+++ +	+ +
Water shift	IC TO EC	EC TO IC
Hypovolemia	+	+++
Cellular dehydration	+++	+
Tongue	Very dry	Rather moist
Skin turgor	normal	Markedly lost
Consciousness	irritability	Coma

Q8B) List clinical presentations and diagnosis of acute leukemia.

- * It is the most common pediatric malignancy. It is due to malignant clonal proliferation of white cell precursors (blast cells), which occupy and inhibit the function of the bone marrow. They are two main categories of acute leukemia:
 - 1) In acute lymphoblastic leukemia (ALL): the blast cells resemble primitive precursors of lymphoid origin. ALL account for 75% of childhood leukemias and it has better prognosis.
 - 2) In acute myeloid leukemia (AML): they resemble myeloid. Prognosis is worse and treatment is unsatisfactory.

* Clinical picture: the most common signs are the following:

- 1) Anemia (pallor).
- 2) Purpura (petechiae) or mucous membrane bleeding.
- 3) Fever.
- 4) Lymphadenopathy and splenomegaly are prominent features (important).
- 5) Bone pain and arthralgia can be also present.

* Laboratory diagnosis: It depends on the following:

- 1) Presence of anemia and thrombocytopenia in the peripheral blood.

2) Presence of blast cells (in ALL) OR myeloid precursors (in AML) in the bone marrow.

* Treatment: ALL is treated by the following regimen:

- 1) Induction of remission by 3 drugs (vincristine prednisone and asparaginase).
- 2) Intra-thecal treatment by 3 drugs (methotrexate, hydrocortisone, cytarabine).
- 3) Systemic continuation of treatment by 2 drugs (6 mercaptopurin, methotrexate).

Exam (4) 29-8-2010 Old System

Q1) Describe treatment options for un-conjugated neonatal hyperbilirubinemia.

* Causes of neonatal hyperbilirubinemia:

- 1) Decreased uptake of bilirubin from plasma: due to decreased Y protein.
- 2) Decreased conjugation: due to deficiency of uridine phosphate glucoronyl transferase activity.

* Pathologic indirect (unconjugated hyperbilirubinemia):

1) Hemolytic disease:

- This is the most common cause of jaundice in the first 24 hours of life.
- It is due to hemolysis of the newborn red cells by maternal antibodies.
- This occurs in cases of incompatibility between maternal and fetal blood, or due to defects of the newborn's RBCs. These include:

2) Rhesus incompatibility:

- Mother is Rh negative she is sensitized during pregnancy or more commonly during a previous delivery or miscarriage to fetal Rh positive blood, previous Rh positive blood transfusion. She reacts to fetal blood by producing antibodies. These globulin antibodies (normally IgG) cross the placenta and cause hemolysis of RBCs of the baby during pregnancy or after delivery.

3) ABO incompatibility:

- This is a common condition in which maternal blood group is O and fetal blood group is A or B, the mother produces anti-A and anti-B hemolysins which cross the placenta and cause hemolysis of RBCs of the baby during pregnancy or after delivery.

4) G6PD deficiency:

- X-linked disorder caused by deficiency of glucose-6-phosphate dehydrogenase enzyme to a variable degree resulting in hemolysis of RBCs on exposure to reducing substances such as salicylates, sulpha, or fava beans. It is a common disorder in Mediterranean countries and should be considered as a possible cause of neonatal jaundice.

5) Spherocytosis:

- Abnormality of the red cell membrane affecting its flexibility causing it to be more liable to hemolysis on passage through the spleen, of variable presentation. It may present as neonatal hyperbilirubinemia or it may present later on life.

6) Other causes:

- Several other conditions may lead to un-conjugated hyperbilirubinemia as congenital infection, septicemia, cephalohematoma, metabolic disorders (as galactosemia), hypothyroidism and pyloric stenosis.

7) Breast milk jaundice:

- Is a form of mild neonatal jaundice occurring in breast fed infants as a result of maternal hormones present in breast milk and competing with bilirubin for enzyme activity.

Q2) Mention the differential diagnosis of papulo-vesicular rash.**1) Chickenpox (varicella):**

- It is a common highly contagious viral disease of children caused by varicella virus.
- Epidemiology: it is most common between 2-10 years. 90 % of affected patients are under 10 years, but the disease may occur at any age including the neonatal period.
- Period of infectivity: it occurs 24 hours before and up to 7 days after the appearance of the characteristic rash. Dry scales are non-infective.
- Clinical manifestations: start after an incubation period of 2-3 weeks.
- Prodroma of mild fever and malaise precede the typical rash by 24 hours.
- The rash has the following characters:
 1. Successive crops: it appears in successive crops over 3-4 days. Each crop consists of small red papules that pass into vesicles (tear drops) on an erythematous base. Vesicle produces a crust that falls with no scar. The severity of the disease is variable.
 2. Centripetal distribution: it starts on the trunk and spreads to the face, scalp and extremities. Vesicles may occur in the mucous membrane of the mouth.
 3. Pleomorphic: at the peak of the disease, the rash consists of crusts (earliest crop), pustules (next crop) and papules and vesicles (latest crops).
 4. Pruritic: pruritis is usually present and may be distressing.
- Complications:
 1. Sepsis due to bacterial contamination.
 2. Neurological as encephalitis, myelitis and polyradiculitis.
 3. Children receiving corticosteroids are at great risk for severe, often fatal chickenpox.
- Management:
 1. Prophylaxis: a live attenuated varicella vaccine is recently available.

2. Treatment: it is mainly symptomatic:

- a. Local and systemic antipruritic agents for itching.
- b. Antipyretics for fever. Aspirin should not be used as it increases the risk of reye syndrome (acute encephalopathy and fatty degeneration of the viscera).
- c. Antibiotics are indicated only if secondary bacterial infections occur.
- d. Antiviral drugs (acyclovir, I.V) are effective for cases complicated by pneumonia or encephalitis. It is also used in immune-compromised children developing varicella.

2) Herpes simplex infections:

- Infection with this virus is extremely common.
- Herpes virus type 1: it spreads by infected saliva and it requires close contact. Primary infection may affect the mouth, skin or eyes.
- Herpes virus type 2: it is a genital infection that usually spreads by sexual contact. The infection has several presentations:
 1. Neonatal infection: it occurs if the mother has genital herpes infection (type2) at the time of delivery. It may present by lethargy, vesicular rash, hepatosplenomegaly, bleeding and neurological symptoms. The mortality rate is high. Elective caesarean section should be considered if active maternal genital herpes is diagnosed.
 2. Acute gingivostomatitis: it is a common disease, characterized by high fever, swollen bleeding gums and painful ulcers in oral mucosa, tongue and palate. Enlarged cervical lymph nodes may be evident. The illness lasts about 10-14 days.
 3. Kerato-conjunctivitis: it is associated with severe eyelid edema and dendritic corneal ulcers. Primary infection of the skin with vesicular lesion is common in older children. Treatment with topical idoxuridine is partially effective.
 4. Cold sores: recurrent herpes simplex infection is common and usually occurs as cold sores around the mouth.
 5. Meningoencephalitis: it may occur even in the absence of skin lesions. The illness is severe and mortality is high. EEG shows temporal and frontal lobes abnormalities. The diagnosis is confirmed by a rising antibody titer in C.S.F. acyclovir (antiviral drug) is the drug of choice.

3) Other cause of vesicular rash:

1. Herpes zoster: vesicles appear along the course of the nerves and are usually unilateral. The eruption is preceded neuralgia along the nerve. The disease is more common in adults.

2. Papular urticaria: no constitutional symptoms. The rash appears mainly in limbs and is usually papular but vesiculation may occur. The scalp and mouth are free.
3. Impetigo: it is a superficial bacterial infection of the skin. The vesicular lesion rapidly progresses to pustules, which are infectious. The mucous membranes are spared.

Q3) Mention the causes and clinical manifestations of pneumonia.

* Pneumonia is a common serious lower respiratory tract infection characterized by an acute inflammatory consolidation of alveoli, infiltration of interstitial tissue with inflammatory cells or a combination of both. Most cases are caused by bacterial or viral infections.

* Causes & pathological types:

- Causes:

1) Bacterial infections:

1. Gram-positive: pneumococcal (most common), streptococcal and staphylococcal.

2. Gram-negative: hemophilus influenza, klebsiella and pseudomonas. Tuberculous pneumonia.

2) Viral infections: Respiratory syncytial virus, adenovirus and giant cell pneumonia.

3) Other infections: Mycoplasma pneumonia and fungal infections.

4) Other causes:

1. Aspiration pneumonia (due to amniotic contents, food and/or gastric acid, foreign body).

2. Hypostatic pneumonia.

- Pathological types:

1) Lobar pneumonia:

Unilateral involvement of one lobe or more than one lobe in the same side. It is mostly bacterial. Other causes are mycoplasma pneumonia and tuberculosis. Chest x-ray shows lobar consolidation.

2) Bronchopneumonia:

Bilateral involvement of both lungs with small foci. It can be bacterial or viral.

Chest x-ray shows fine nodular or patchy infiltration.

3) Interstitial pneumonia:

Bilateral involvement of interstitial tissues. It is mostly viral. Chest x-ray shows dense parahilar shadow with radiating streaks.

- * Diagnosis of pneumonia: pneumonia should be a possibility in every case of respiratory distress especially when associated with fever and/or cough.
- Manifestations of respiratory distress include:
 - 1) Grade 1: tachypnea (rapid respiration).
 - 2) Grade 2: intercostals and subcostal retractions.
 - 3) Grade 3: expiratory grunting.
 - 4) Grade 4: cyanosis appears.
- Diagnosis of pneumonia should include the pathological type, the possible causative organism and the associated complications.
- Pathological types:
 - 1) Lobar pneumonia: bronchial breathing over the involved lobe or lobes is the main finding. Some dullness to percussion over the involved lobe may be also present.
 - 2) Bronchopneumonia: fine bilateral consonating crepitations are the main findings.
 - 3) Interstitial pneumonia: severe spasmodic cough and tendency to expiratory wheezing are the main findings.
- Causative organism: Differentiation between viral and bacterial pneumonia are important.
 - 1) Bacterial pneumonia:
 1. High fever and severe course suggests bacterial infections.
 2. Leukocytosis, raised ESR and raised CRP are commonly present.
 - 2) Viral pneumonia:
 1. Fever is usually not high.
 2. Laboratory tests (leukocytic count, ESR, CRP) are normal or only mildly affected.
- * Complications:
 - 1) Respiratory failure: the most serious complication and the main cause of death.
 - 2) Pleural effusion: with bacterial pneumonias especially pneumococcal, staphylococcal pneumonia.
 - 3) Lung abscess: with bacterial pneumonias especially staphylococcal pneumonia.
 - 4) Myocarditis and acute heart failure: especially in infants with severe bacterial pneumonia.
- Chest x-ray is essential for identification of the pathological type and detection of possible associated effusion or lung abscess.

Q4A) State the genetic type of Down syndrome.

1) Nondisjunction type (95% of cases):

- Total number of chromosomes is 47.
- Higher incidence with advanced maternal age (1/100 at 40 years 1/10 at 50 years).

2) Translocation type (4% of cases):

- Total number of chromosomes is 46.
- It has 2 subtypes:
 1. D/G 21 translocation: the extra chromosome is translocated into D group (13, 14 or 15).
 2. G/G 21 translocation: the extra-chromosome is translocated into G group (21 or 22).
- About 40% of D/G 21 and 10% of G/G 21 are inherited.

3) Mosaic type (1% of cases):

- Some cells are normal (46 chromosomes) and others are trisomic (47).
- Clinical features are less evident and mental retardation is mild.

* Recognition of the genetic type (by chromosomal karyotyping) is important for precise genetic counseling as some types are inherited with a high recurrence rate.

Q4B) List indications of adequate breast milk intake.

- 1) Weight gain: Baby gains 200 gm/week or 250 gm/10 days in the first months of life.
- 2) Infant behavior: Baby feeds at least 8 times/day.
- 3) Swallowing noises are heard during feeding.
- 4) Baby is satisfied between feedings.
- 5) Baby passes at least 4 soft yellow stools/ day and wets 6 or more dippers/day.
- 6) Breast fullness: Mother noticed that her breasts feel full before a feeding and softer afterwards. Some mothers experience a letdown sensation.
- 7) When breast milk is not sufficient (the baby is not growing properly), combined feeding is used where breast feeding is completed with one or more bottle feeds.

Q5A) Define macrocephaly and list its causes.

* Large head or macrocephaly (head circumference above 95th percentile for age and sex).

1) Cranial (skull) causes:

Rickets, achondroplasia, chronic hemolytic anemia, familial.

2) Intracranial causes:

Hydrocephalus, subdural effusion, subdural hemorrhage, brain tumors.

Q5B) Describe the prevention of infective endocarditis.

* Infective endocarditis is a well recognized complication of congenital as well as rheumatic heart disease. The risk is higher with lesions, which result in a turbulent jet of blood, such as a ventricular septal defect, coarctation and patent ductus.

* Causative organism: The endocardium becomes infected in the presence of bacteremia, which may occur during dental treatment or cardiac surgery. Two organisms are responsible:

1) Streptococcus viridians.

2) Staphylococcus aureus: it has become increasingly more common.

- In approximately 10% of cases blood culture is negative.

* Clinical features: clinical manifestations include one or more of the following:

1) Toxic manifestations:

1. Pale, toxic appearance.

2. Tachycardia out of proportion of fever.

3. Splenomegaly in 70% of cases.

4. Clubbing of fingers.

5. Cutaneous manifestations:

a. Osler's nodes: purplish, pea sized painful nodules in pads of fingers and toes.

b. Splinter hemorrhage: linear hemorrhage beneath the nails.

c. Janeway spots: painless erythematous lesions in the palms and soles.

2) Embolic manifestations:

1. Cerebral infarction: hemiplegia / aphasia / blindness.

2. Renal infarction: hematuria even microscopic.

3. GIT, pulmonary infarction.

4. Mycotic aneurysm as a result of occlusion of vasa vasorum of cerebral vs. it may rupture and ends in intracranial hemorrhage.

3) Cardiac manifestations:

1. Appearance of new murmurs or change of already presented murmur.
2. Rupture of the cusps with appearance of sea gull murmur.
3. Heart failure.

* Investigations:

- 1) Repeated blood culture: diagnosis depends on positive blood culture.
- 2) Echocardiography to detect valvular vegetation.
- 3) Acute phase reactant (leukocytosis, raised ESR and CRP).

* Treatment:

- 1) Parental antibiotic therapy: it is given in large doses for a period of 6 weeks.
 - The choice of antibiotics depends on the causative organism:
1. Streptococcus viridians: the combination of benzylpenicillin (300000 unit/kg/day) and gentamicin (4-6 mg/kg/day) is useful.
2. Staphylococcal infection: antistaphylococcal drugs as cloxacillin (200 mg /kg/day) should be added. Gentamicin can be substituted with amikacin (15 mg/kg/day).
- 2) Heart failure if presented should be treated.

* Prevention:

- Antibiotic prophylaxis: any child with a heart lesion, congenital or rheumatic (no matter how trivial) needs antibiotic prophylaxis with dental treatment (extraction, fillings) or before surgery (involving the middle ear, tonsils or adenoids). The usual practice is to give oral amoxicillin as a single large dose 1 hour before the treatment.

Q6A) Mention the prevention and treatment of bleeding esophageal varices.*** Treatment:**

- Therapy is directed at the presentation and/or management of variceal hemorrhage.
- Emergency therapy of bleeding varices: hospitalization and:
 - 1) I.V. fluids, blood transfusion.
 - 2) Nasogastric tube placement.
 - 3) Correction of coagulopathy by I.V. vitamin K, fresh frozen plasma, platelet transfusion.
 - 4) Vaso-pressin transfusion infusion if bleeding persists.
 - 5) Endoscopy: as soon as patient is hemodynamically stable, endoscopy and either sclerotherapy or band ligation of the varices should be done.
 - 6) Surgical treatment: transjugular intrahepatic porto-systemic shunt (TIPSS) or emergency surgical porto-systemic shunt.

*** Prevention of bleeding:**

It includes prevention of the first bleeding episode and prevention of rebleeding.

1) Prevention of the first bleeding episode: it includes the following:

1. Avoid aspirin and non-steroidal anti-inflammatory drugs.
2. Beta adrenergic blockers (propranolol).
3. Prophylactic sclerotherapy or band ligation.

2) Prevention of re-bleeding: it includes the following:

1. Beta adrenergic blockers (propranolol).
2. Endoscopic sclerotherapy or band ligation.
3. Surgical porto-systemic shunt.
4. Liver transplantation.

Q6B) List causes of hydrocephalus.

* Hydrocephalus is the dilatation of the ventricular system, due to imbalance between production and absorption of CSF, with or without concomitant increase in CSF pressure.

Hydrocephalus can be congenital or acquired and obstructive or communicating.

- 1) Obstructive hydrocephalus: there is obstruction to flow of the CSF within the ventricular system. There is dilatation of the ventricular system proximal to obstruction.
- 2) Communicating hydrocephalus: the flow of CSF within the ventricular system is intact (communicating ventricles) but there is interference with the absorption of the CSF in the subarachnoid space. All ventricles are dilated.

* Causes & types of hydrocephalus:

1) Obstructive hydrocephalus:

1. Aqueductal stenosis (congenital or acquired as with tumors, hemorrhage and infections) it is the most common congenital cause.
2. Dandy-walker malformation.
3. Malformation of the vein of galen.
4. Midline brain tumors (especially cerebellar tumors as medulloblastoma).
5. Posterior fossa subdural hematoma.

2) Communicating hydrocephalus:

1. Arnold-chiari malformation.
2. Post-meningitic (due to subdural adhesion).
3. Subarachnoid hemorrhage.
4. Overproduction of CSF as choroid plexus papilloma.

Exam (5) 3-7-2011 New System

Q1A) List causes of cyanosis with respiratory distress in a newborn infant.

- 1) Cardiac disorders - cyanotic congenital heart disease.
- 2) Respiratory disorder e.g. surfactant deficiency, meconium aspiration, pulmonary hypoplasia.
- 3) Persistent pulmonary hypertension of the newborn (PPHN) - failure of the pulmonary vascular resistance to fall after birth.
- 4) Infection - septicemia, group B streptococcus and other organisms.
- 5) Inborn error of metabolism- metabolic acidosis and shock.
- 6) Polycythaemia.

Q1B) Mention neonatal problems associated with maternal diabetes mellitus. Discuss, briefly their management.

- 1) Hypoglycaemia : transient hypoglycaemia is common during the first day of life from fetal hyperinsulinism, but can often be prevented by early feeding. The infant's blood glucose should be closely monitored during the first 24 hours and hypoglycaemia treated.
- 2) Respiratory distress syndrome (RDS): more common as lung maturation is delayed.
- 3) Hypertrophic cardiomyopathy: hypertrophy of the cardiac septum occurs in some infants. It regresses over several weeks but may cause heart failure from reduced left ventricular function.
- 4) Polycythemia (venous haematocrit more than 0.65): makes the infant look plethoric. Treatment with partial exchange transfusion to reduce the haematocrit and normalize viscosity may be required.

Q2A) Enumerate the common bacterial causes of septicemia in infant and children. Discuss its clinical feature.

* Bacteria may cause focal infection or proliferate in the blood stream, leading to septicemia, in septicemia, the host response include the release of inflammatory cytokines and activation of endothelial cells which may lead to septic shock. The

commonest cause of septic shock in childhood is meningococcal infection, which may or may not be accompanied by meningitis. Fortunately, its incidence in the UK has fallen markedly since immunization was introduced. Pneumococcus is the commonest organism causing bacteraemia, but it is unusual for it to cause septic shock. In neonates, the commonest cause of septicemia is group B-streptococcus or gram-negative organisms acquired from birth canal.

* Clinical features of septicemia:

- 1) History: Fever, poor feeding, miserable, lethargy, history of focal infection e.g. (meningitis, osteomyelitis, gastroenteritis, cellulitis predisposing conditions, e.g. sickle cell disease, immunodeficiency).
- 2) Examination: Fever, purpuric rash (meningococcal septicemia), irritability, shock, multiorgan failure.

Q2B) Describe diagnostic workup (investigation) of tuberculosis in children.

* Diagnostic investigations:

- 1) Tuberculin test: mantoux test is the most important immunological diagnostic tools.
- 2) Sputum examination: direct smear with Zeihl Nelson stains for tuberculous bacilli.
- 3) Stomach wash and gastric aspirations: for a direct smear or culture.
- 4) Biopsy of lymph nodes or pleura: for pathological study.
- 5) Radiological studies: Chest x-ray and chest computed tomography (CT scan)
- 6) Recent methods for diagnosis: usage of ELISA and PCR (polymerase chain reaction).

* Tuberculin test:

- 0.1ml purified protein derivative is injected intradermally in the skin of the flexor surface of the forearm. The reaction should be read at 48-72 hours. Induration (and not erythema) is measured in mms in the longitudinal as well as the transverse diameters and the mean reading is recorded.
- 1. An induration less than 5 mm is considered negative.
- 2. An induration measuring 5-9 mm is considered doubtful and should be repeated.
- 3. An induration of 10 mm is considered positive.
- Positive tuberculin test: it indicates vaccination or infection:

1. BCG vaccination: the reaction is never exceeds 15 mm induration.
2. T.B. infection: it considered in 2 situations:
 - a. Positive reaction in a child less than 5 years who is not BCG vaccinated.
 - b. Strongly positive reaction (over 15 mm) in previously vaccinated child.
- False negative tuberculin test: may happen in the following situation:
 1. Subcutaneous instead of intradermal injection of tuberculin or use of outdated tuberculin.
 2. Recent use of corticosteroids or immunosuppressive.
 3. Intercurrent infections especially viral ones.
 4. Recent antiviral vaccine (especially measles and mumps).
 5. Advanced disseminated T.B. Chronic debilitating disease with cachexia and cell mediated immune deficiency.

Q3A) Discuss the cytogenetic of Down's syndrome .

- * The extra chromosome 21 may result from non-disjunction, translocation or mosaicism.
- 1) Non-disjunctions (94%):
 - Most cases result from an error at meiosis.
 - The pair of chromosome 21s fails to separate, so that one gamete has two chromosome 21s and one has none.
 - Fertilisation of gamete with two chromosome 21s gives rise to a zygote with trisomy 21.
 - Parental chromosomes do not need to be examined.
 - 2) Translocation (5%):
 - When the extra chromosome 21 is joined onto another chromosome (usually chromosome 14, but occasionally chromosome 15, 22 or 21), this is known as an unbalanced robertsonian translocation. An affected child has 46 chromosomes but three copies of 21 materials. In this situation, parental chromosomal analysis is essential since one of the parents carries a balanced translocation in 25% of cases. Translocation carriers have 45 chromosomes, one of which consists of the two joined chromosomes.
 - In translocation Down's syndrome:
 1. The risk of recurrence is 10-15% if the mother is the translocation carrier and about 2.5% if the father is the carrier.

2. If a parent carries the rare 21:21 translocation, all the offspring will have Down's syndrome.
3. If neither parents carries a translocation (75% of cases), the risk of recurrence is less than 1%.

3) Mosaicism (1%):

- In mosaicism some of the cells are normal and some have trisomy 21. This usually arises after formation of the zygote, by non-disjunction at mitosis. The phenotype may be milder in mosaicism.

Q3B) List prenatal, perinatal and postnatal causes of developmental delay and learning disability (mental retardation).

* Prenatal:

1) Genetic:

1. Chromosome/DNA disorders e.g. Down's syndrome, fragile X syndrome.
2. Cerebral dysgenesis e.g. microcephaly, absent corpus callosum, hydrocephalus, neuronal migration disorder, vascular occlusion.
- 2) Metabolic: Hypothyroidism, phenylketonuria.
- 3) Teratogenic: Alcohol and drug abuse
- 4) Congenital infection: Rubella, cytomegalovirus, toxoplasmosis.
- 5) Neurocutaneous syndrome: Tuberous sclerosis, neurofibromatosis.

*Perinatal:

- 1) Extreme prematurity: Intra-ventricular hemorrhage/peri-ventricular leucomalacia.
- 2) Birth asphyxia: Hypoxic-ischemic encephalopathy.
- 3) Metabolic: Symptomatic hypoglycemia, hyperbilirubinemia.

* Postnatal:

- 1) Infection: Meningitis, encephalitis.
- 2) Anoxia: Suffocation, near drowning, seizures.
- 3) Trauma: Head injury-accidental or non accidental.
- 4) Metabolic: Hypoglycemia, inborn error of metabolism.

* Other: Unknown (about 25%).

Q4) List the causes and describe the pathogenesis of rickets.

* Nutritional (primary) rickets- risk factors:

- 1) Living in northern latitudes.
- 2) Dark-skinned people.
- 3) Decrease exposure to sunlight e.g. in some Asian children living in the UK
- 4) Maternal vitamin D deficiency.
- 5) Diets low in calcium, phosphorus and vitamin D e.g. exclusive breast feeding into late infancy or rarely toddlers on unsupervised dairy free diets.
- 6) High phytic acid diet e.g. chapattis.
- 7) Macrobiotic, strict vegan diets.
- 8) Prolonged parenteral nutrition in infancy with an inadequate supply of parenteral calcium and phosphate.

9) Intestinal malabsorption:

- Defective production of 25(OH) D3 - liver disease.
- Increase metabolism of 25(OH) D3 - enzyme induction by anticonvulsant.

* Defective production of 1, 25(OH) 2D3

- 1) Hereditary type 1 vitamin D-resistant (or dependent) rickets (mutation which abolishes activity of renal hydroxylase).
- 2) Familial (X-linked) hypophosphataemic rickets (renal tubular defect in phosphate transport).
- 3) Chronic renal disease.
- 4) Fanconi syndrome (renal loss of phosphate).

* Target organ resistance to 1, 25(OH) 2D3.

* Hereditary vitamin D- dependent rickets type 2 (due to mutations in vitamin D receptor gene).

* Pathogenesis of rickets: Deficiency of vitamin D > decrease Ca absorption from gut > initial, slight and transient decrease in serum Ca > 2ry hyperparathyroidism > mobilization of Ca and P from bone and inhibition of P re-absorption from kidney leading to the classic 3 changes that occur in rickets:

- 1) Normal serum calcium (N 9-11 mg %).
- 2) Low serum phosphorus (N 4.5-5.5 mg %).
- 3) High alkaline phosphatase (early finding).

Q5) Describe the treatment of hypercyanotic spells in infant's with fallot's tetralogy.

* Hypercyanotic spells are usually self-limiting and followed by a period of sleep. If prolonged (beyond about 15 minutes), they require prompt treatment with:

- 1) Squatting (knee chest position).
- 2) Sedation and pain relief (morphine is excellent).
- 3) Intravenous propranolol (or an alpha adrenoceptor agonists), which probably works both as a peripheral vasoconstrictors and by relieving the subpulmonary muscular obstruction that is the cause of reduced pulmonary blood flow.
- 4) Intravenous volume administration.
- 5) Bicarbonate to correct acidosis.
- 6) Oxygen.
- 7) Prophylactic iron therapy.

Q6) Discuss bronchodilator therapy in childhood asthma.

* Bronchodilator therapy:

- Inhaled b2-agonists are the most commonly used and most effective bronchodilators. Short acting b2-agonists (often called relievers) such as salbutamol or terbutaline have a rapid onset of action, are effective for 2-4 hours and have few side effects.
 - They are used as required for increased symptoms, and in high doses for acute asthma attacks.
 - In contrast, long acting b2-agonists (LABAs) such as salmetrol or formoterol are effective for 12 hours and are used in conjunction with regular inhaled corticosteroids. They are not used acute asthma, and should not be used without inhaled corticosteroids. Long-acting b2 agonists are useful in exercise induced asthma.
 - Ipratropium bromide, an anticholinergic bronchodilator, is sometimes given to young infants when other bronchodilators are found to be ineffective, or in the treatment of severe acute asthma.
- * Slow release oral theophylline: is an alternative, however it has a high incidence of side-effects (vomiting, insomnia, headaches, and poor concentration) and blood levels need to be monitored, so it is now rarely used in children.

Q7) Discuss the etiology, clinical features and investigations of acute post infectious polyneuropathy (Guillain-Barre syndrome).

* Presentation is typically 2-3 weeks after an upper respiratory tract infection or campylobacter gastroenteritis. There may be fleeting abnormal sensory symptoms in the legs, but the prominent features is an ascending symmetrical weakness with loss of reflexes and autonomic involvement. Sensory symptoms, usually in the distal limbs, are less striking than the paresis but can be unpleasant. Involvement of bulbar muscles leads to difficulty with chewing and swallowing and the risk of aspiration. Respiratory depression may require artificial ventilation. The maximum muscles weakness may occur only 2-4 weeks after the onset of illness. Although full recovery may be expected in 95% of cases, this may take up to 2 years. The CSF protein is characteristically markedly raised, but this may not be seen until the second week of illness. The CSF white cell count is not raised. Nerve conduction velocities reduced.

Q8) Describe, preferably as a table, the clinical assessment of different grades of dehydration.

	Moderate dehydration	Severe dehydration
Body weight loss	5-10%	More than 10%
General appearance	Thirsty, drowsy.	Drowsy, limp, cold, sweaty, cyanotic extremities.
Respiration	Deep, may be rapid	Deep and rapid
Eyes	Sunken	Grossly sunken
Tears	Reduced/absent	Absent
Mucous membranes	Dry	Very dry
Capillary refill time	Prolonged (more than 2 sec)	Prolonged (more than 2 sec)
Tissue turgor	Retract slowly	Retract very slowly
Blood pressure	Normal or low	Low
Radial pulse	Rapid and weak	Rapid, thready may be impalpable
Anterior fontanelle	Sunken	Very sunken
Urine output	Reduced	Marked oliguria

Q9) Describe the laboratory diagnosis of acute viral hepatitis in children.

* Confirmatory investigations of acute liver injury:

- Investigations to confirm the presence of acute liver injury include serum bilirubin level, serum transferases, serum albumin and prothrombin time. Blood ammonia and serum electrolytes are also important.

* Laboratory evidence of acute liver injury:

1) Direct or mixed hyper bilirubinemia :

Mild (2-6 mg /dl), moderate (6-10 mg/dl) or severe (more than 10mg/dl) in fulminate hepatitis, peak levels above 20 mg/dl occur.

2) Raised serum transferases: (levels between hundreds and thousands are common)

1. Raised aspartate aminotransferase (AST) formally known as (SCOT).

2. Raise alanine aminotranferase (ALT) formely known as (SGPT).

3) Evidence of acute hepatic failure (in fulminate hepatitis):

1. Raising bilirubin level (above 10mg/dl).

2. Low serum albumin level (below 3 gm/dl).

3. Prolonged prothrombin time (more than 20 seconds).

4. High blood ammonia level (above 150 mcg/dl).

5. Hypokalemia, hyponatremia and metabolic acidosis.

* Laboratory differentiation:

- Hepatitis markers (antigens and antibodies) of different agent are the only reliable way for differentiation

1) Hepatitis A: anti-HAV antibodies belonging to the IgM class indicate acute disease, whreas anti-HAV IgG antibodies persist after recovery.

2) Hepatitis B: acute hepatitis B is heralded by appearance of HbsAG, followed by anti-HBc IgM. Recovery and development of immunity is noted by appearance of anti HBs, in chronic infections. HbsAG persists and anti-HBc IgG develops.

3) Hepatitis C: anti-HCV antibody denotes exposure to infection but does not denote recovery or development of immunity.

Q10) List causes of congenital hypothyroidism. Describe its clinical features.*** Causes of congenital hypothyroidism:**

- 1) Mal-descent of the thyroid and athyrosis - the commonest cause of sporadic congenital hypothyroidism. In early fetal life, the thyroid migrates from a position at the base of the tongue (sublingual) to its normal site below the larynx. The thyroid may fail to develop completely or partially. In mal-descent, the thyroid remains as a lingual mass or a unilobular small gland. The reason for this failure of formation or migration is not well understood.
- 2) Dyshormonogenesis, an inborn error of thyroid hormone synthesis, in about 5- 10 % of cases, although commoner in some ethnic groups with consanguineous marriage.
- 3) Iodine deficiency, the commonest cause of congenital hypothyroidism worldwide but rare in the UK. It can be prevented by iodination of salt in the maternal diet.
- 4) Hypothyroidism due to TSH deficiency - isolated TSH deficiency is rare (less than 1% of cases) and is usually associated with panhypopituitarism, which usually manifests with growth hormone and adrenocorticotrophic hormone (ACTH) deficiency before the hypothyroidism becomes evident.

*** Clinical features of hypothyroidism:**

- 1) Failure to thrive.
- 2) Feeding problems.
- 3) Prolonged jaundice.
- 4) Constipation.
- 5) Pale, cold, mottled dry skin.
- 6) Coarse facies.
- 7) Large tongue.
- 8) Hoarse cry.
- 9) Goiter (occasionally).
- 10) Umbilical hernia.
- 11) Delayed development.

Q11) Describe complications of sickle cell disease, and their management.**1) Infection:**

- All have marked increase in susceptibility to infection from encapsulated organisms such as pneumococci and haemophilus influenza. There is an increased incidence of osteomyelitis caused by salmonella and other organisms. This susceptibility to infection is due to hyposplenism secondary to chronic sickling and micro infection in the spleen in infancy. The risk of overwhelming sepsis is greatest in early childhood.

2) Painful crises:

- Vaso-occlusive crises: causing pain may affect all organs of the body with varying frequency and severity. A common mode of presentation in late infancy is the hand-foot syndrome, in which there is dactylitis with swelling and pain of the fingers and feet from vaso-occlusion.
- The bones of the limbs and spine are common sites whereas cerebral and pulmonary infection are less common but serious. The commonest presentation of cerebral infarction is an acute stroke. With hemiparesis from blockage of the medium to large arteries, unlike vaso-occlusion in the small blood vessels elsewhere in the body. A vascular necrosis of the femoral heads may also occur, dehydration, excessive exercise or stress, hypoxia or infection.

3) Acute anemia (sudden drop in hemoglobin from):

1. Hemolytic crises- associated with infection.
2. Aplastic crises- haemoglobin may fall precipitously. Parvovirus infection causes complete, though temporary, cessation of red cell production.
3. Sequestration crises- sudden splenic enlargement, abdominal pain and circulatory collapse from accumulation of sickled cells in spleen.

4) Short stature & delayed puberty.**5) Long term problems:**

1. Cardiac enlargement: from chronic anemia.
2. Heart failure: from uncorrected anemia.
3. Renal dysfunction: may exacerbate enuresis because of inability to concentrate urine.
4. Pigment gallstones: due to increased bile pigment production.

Q12) List the causes and investigations of hematuria in children.*** Causes of hematuria:****1) Non-glomerular:**

1. Infection (bacterial, viral, TB, schistosomiasis).
2. Trauma to genitalia, urinary tract or kidneys.
3. Stones.
4. Tumors.
5. Sick cell disease.
6. Bleeding disorders.
7. Renal vein thrombosis.
8. Hypercalcinuria.

2) Glomerular:

1. Acute glomerulonephritis (usually with proteinuria).
2. Chronic glomerulonephritis (usually with proteinuria).
3. IgA nephropathy.
4. Familial nephritis.
5. Thin basement membrane disease.

*** Investigations of hematuria:****1) All patients:**

1. Urine microscopy (with phase contrast) and culture.
2. Protein and calcium excretion.
3. Kidney and urinary tract ultrasound.
4. Plasma urea, electrolytes, creatinine, calcium, phosphate, albumin.
5. Full blood count, platelets, clotting screen, sickle cell screen.

2) If suggestive of glomerular hematuria:

1. ESR, complement level and anti-DNA binding.
2. Throat swab and antistreptolysin O titer.
3. Hepatitis B antigen.
4. Renal biopsy if indicated.
5. Test mother's urine for blood (if Alport's syndrome suspected).
6. Hearing test (if Alport's syndrome suspected).

Exam (6) 4-7-2011 Old System

Q1) Discuss the clinical features and management of neonatal respiratory distress syndrome

*** Clinical features:**

- 1) Tachypnoea more than 60 breath/minute.
- 2) Laboured breathing with chest wall recession (particularly sternal and subcostal indrawing) and nasal flaring.
- 3) Expiratory grunting in order to try to create positive air way pressure during expiration and maintain functional residual capacity.
- 4) Cyanosis.
- 5) The characteristic chest X-ray appearance.

*** Treatment:** with raised ambient oxygen is required, which may need to be supplemented with contentious positive air way pressure (delivered via nasal cannulae) or artificial ventilation via a tracheal tube. The ventilatory requirements need to be adjusted according to the infant's oxygenation (which is measured contentiously) chest wall movements and blood gas analyses. Artificial ventilation may be synchronized as far as possible with the infant's respiration, or the infant's breathing may be partially or completely suppressed with sedatives and muscle relaxants. Mechanical ventilation may be with intermittent positive pressure ventilation or high frequency oscillation.

Q2) Discuss advantages of breast feeding to the infants.

- 1) Provides the ideal nutrition for infant during the first 4-6 months of life.
- 2) Is life saving in developing countries.
- 3) Reduces the risk of gastrointestinal infection, and in preterm infants of necrotising enterocolitis.
- 4) Improves cognitive development.
- 5) Enhances mother-child relationship.
- 6) Reduces risk of insulin-dependent diabetes, inflammatory bowel, sudden infant death syndrome (unproven) in later life.

Q3) Enumerate the bacterial causes of meningitis according to age. Discuss the laboratory investigations of a case suspected of bacterial meningitis.

- 1) Neonatal (3 months): Group B streptococcus, E.coli and other coliforms, listeria monocytogenes.
- 2) 1 month - 6 years: Neisseria meningitides, streptococcus pneumonia, haemophilus influenzae.
- 3) More than 6 years: Neisseria meningitides, streptococcus pneumoniae.

* Investigations:

A lumbar puncture is performed to obtain CSF confirm the diagnosis; identify the organism responsible, and its antibiotic sensitivity. If any of the listed in figure 14.16 are present a lumbar puncture should not be performed, as under the circumstances the procedure carries a risk of coning of the cerebellum through the foramen magnum. If necessary, a lumbar puncture can be performed once the child's condition has stabilized. Although by this stage the organism will rarely be grown, the cytological and biochemical abnormalities of bacterial meningitis will still be present for several days after starting treatment. Even without a lumbar puncture, bacteriological diagnosis can be achieved in at least 50% of cases from the blood by culture, rapid antigen screen or PCR. A throat swab should also be taken. Scrapings from a purpuric skin lesion may also be cultured. A serological diagnosis can be made on convalescent serum 4-6 weeks after the presenting illness.

Q4) Discuss the diagnosis and treatment of large ventricular septal defects.

* LARGE VSD: these defects are the same size or bigger than the aortic valve.

* Clinical features:

1) Symptoms:

1. Heart failure with breathlessness and failure to thrive after 1 week old.
2. Recurrent chest infection.

2) Physical signs:

1. Active precordium.
2. Soft pansystolic murmur or no murmur.
3. Apical mid-diastolic murmur (from increased flow across the mitral valve after the blood has circulated through the lungs).

4. Loud pulmonary secondary sound (P2) - from raised pulmonary arterial diastolic pressure.

5. Tachypnoea, tachycardia and enlarged liver from heart failure.

3) Investigations:

1. Chest X-ray:

- Cardiomegaly.
- Enlarged pulmonary arteries.
- Increase pulmonary vascular markings.
- Pulmonary edema.

2. ECG:

- Biventricular hypertrophy by 2 months of age and signs of pulmonary hypertension.

3. Echocardiography:

- Demonstrates the anatomy of the defect, hemodynamic effects and severity of pulmonary hypertension.

* Management:

1) Drug therapy for the heart failure is with diuretics often combined with captopril. Additional calorie input is required. There is always some degree of pulmonary hypertension in children with large left-to-right shunt. This damages the lungs as increased pulmonary blood flow and pulmonary hypertension will ultimately lead to irreversible damage of the pulmonary capillary vascular bed. This pulmonary vascular disease usually become established in the second year of life, but Eisenmenger's syndrome with cyanosis due to intra-cardiac shunting from right to left, rarely evolves until the second decade.

2) Surgery is usually performed at 3-6 months of age in order to:

1. Manage heart failure and failure to thrive.
2. Prevent permanent lung damage from pulmonary hypertension and high blood flow.

Q5) Describe the clinical features and treatment of a case of hemophilia A.

* It is an X-linked recessive disorder. This occurs due to reduced plasma factor activity.

* The incidence is 1/14000 males and it represents 80% of hemophilias.

* Clinical features:

1) Bleeding: it occurs early in life after injection or circumcision.

- 2) Easy bruising and even large hematomas: they occur as ambulation starts.
- 3) Spontaneous hematuria: it may occur in severe cases.
- 4) Hemarthrosis: It occurs with minor trauma or spontaneous and it is the hallmark of severe hemophilia A. repeated hemorrhages inside joint spaces produce degenerative changes and ultimately a fixed unstable joint.
- 5) Intracranial hemorrhage: it represents a serious life-threatening emergency.

* Treatment:

- 1) Cold compresses: it can be beneficial in mild cases. However when major bleeding episodes occurs, replacement therapy is very essential.
- 2) Replacement therapy: it aims at increasing factor in the plasma to a level that secure homeostasis. This can be done by the following:
 1. Intravenous infusion of fresh frozen plasma.
 2. Intravenous infusion of cryoprecipitate (plasma concentrate of factor) it is preferred, Because of small volume. And it is given in a dose of 25-50 unit/kg and should be repeated every 8-12 hours. Since cryoprecipitate carries the risk of transmission of viral infections especially hepatitis B and C and HIV virus. It is recommended to vaccinate hemophiles against hepatitis B
 3. Intravenous infusion of purified factor concentrates 25-50 unit/kg.
 4. Physiotherapy: it is important to strength muscles especially after long periods of immobilization, as muscles are very important for joint stability.
 5. Desmopression (antidiuretic hormone): it increase factor production and may be beneficial in mild and moderate cases.

Q6) Describe the clinical features and differential diagnosis of roseola infantum.

- * Clinical manifestations: start after an incubation period of about one week.
- * Onset: sudden by high fever (39-41c) with no sign to explain it. The fever falls by crisis on the fourth day and a maculopapular eruption appears simultaneously.
- * The rash: appears on the trunk and spreads to the neck, the arms the face and the legs. It fades within 24 hours only.

* Maculopapular rash:

- 1) Infections: Common exanthems (rash is essential for diagnosis):
 1. Measles: high fever and rash for 6 days.

2. German measles: mild fever and rash for 3 days.
3. Roseola infantum: high fever and rash for one day.
4. Scarlet fever: high fever and rash for 3-7 days.
- 2) Other infections: (rash may or may not appear):
 1. Typhoid fever: fever, splenomegaly and may be a rash.
 2. Infectious mononucleosis: fever, pharyngitis, lymphadenopathy and Splenomegaly.
 3. Enteroviral infection: nonspecific rash.

* Rheumatic diseases:

- 1) Rheumatoid arthritis: fever, chronic arthritis and may be a rash.
- 2) Systemic lupus erythematosus: fever, butterfly rash and renal disease.
- 3) Dermatomyositis.

* Skin & allergic diseases:

- 1) Sweat rash: fine papular rash mainly in neck, trunk and napkin area.
- 2) Urticarial rash: itchy, erythematous raised skin lesions (wheals).
- 3) Drug rash: nonspecific rash.

Q7) List the early and late signs of shock in pediatric patients.

* Early (compensated):

- 1) Tachypnoea.
- 2) Tachycardia.
- 3) Decrease skin turgor.
- 4) Sunken eye and fontanelle.
- 5) Delayed capillary refill (more than 2 sec).
- 6) Mottled, pale, cold skin.
- 7) Core-peripheral.
- 8) Temperature gap (more than 4C).
- 9) Decrease urinary output.

* Late (decompensated):

- 1) Acidotic (Kussmaul) breathing.
- 2) Bradycardia.
- 3) Confusion / depressed cerebral state.
- 4) Blue peripheries.
- 5) Absent urine output.
- 6) Hypotension.

Q8) Discuss the diagnosis and management of recurrent non-organic abdominal pain.

* Dysfunctional recurrent abdominal pain:

- Fortunately, more than 90% of children with recurrent abdominal pain belong to this group. Other terms as (nonspecific or psychogenic) recurrent abdominal pain can be used. Reassurance is important.

- 1) Causes: the true mechanism is unknown but several factors may be responsible. Stressful event as family problem, loss of one parent, delivery of new sibling and school phobias are all important factors. Some children complain to gain more love and sympathy and others may imitate an adult with recurrent abdominal pain.
- 2) Site of pain: the pain is usually periumbilical and the child can often locate the site even with one fingers. It is usually vague, not severe (not interfering with activity) and subsides spontaneously in less than 20 minutes. As the pain is not severe, parents usually come to complain after several weeks of onset.
- 3) Associated complaints: there are no other associated complaints and the child appears normal and healthy.
- 4) Abdominal examination: it is generally negative.
- 5) Simple investigations: urine, stool, blood count and abdominal x-ray are all normal.