POSTGRADUATE INSTITUTE OF MEDICINE
UNIVERSITY OF COLOMBO

MD (PAEDIATRICS) EXAMINATION
JANUARY / FEBRUARY 2012

Date : 24th January 2012 Time : 9.00a.m. – 12.00 noon

PAPER 1
STRUCTURED ESSAY QUESTIONS

Answer all Five Questions
Answer each question in a separate book

Q.1.

1.1 Write on bilirubin metabolism. (25 marks)

1.2.

(a) Name five [05] hereditary disorders of bilirybil metabolism and indicate their patterns of inheritance. (15 marks)

(b) Mention the basic underlying defect/s in each of the conditions given in 1.2.(a) (15 marks)

1.3 Mention the treatment modalities available globally for the condition mentioned in 1.2.(a) which presents with severe unconjugated hyperbilirubinaemia in the neonatal period. (30 marks)

1.4. Outline the underlying mechanisms of action of phototherapy in managing indirect hyperbilirubinaemia. (15 marks)

Q.2.

2.1 Enumerate four [04] anthropometric methods and four radiological methods for determining adiposity in children (25 marks)

2.2 Discuss their advantages and disadvantages in clinical practice. (25 marks)

2.3 Discuss the important concepts in the dietary management of Type 1 diabetes mellitus in a 7 year old who is treated with twice a day Mixtard insulin. (50 marks)

Q.3.

3.1. Name five [05] ultrasonographic parameters that assess placental function during the third trimester (20 marks)

3.2. What is the "gold standard" for assessment of utero-placental function during labour ? (10 marks)

3.3. Describe the pathophysiology of Hypoxic Ischaemic Encephalopathy. (30 marks)

3.4. Discuss the principles of management of Hypoxic Ischaemic Encephalopathy. (30 marks)

3.5. Name two [02] investigations that could predict a poor outcome in Hypoxic Ischaemic Encephalopathy. (10 marks)

Q.4.

4.1. Describe the physiology of calcium homeostasis. (30 marks)

4.2.

(a) Outline five [05] patho-physiological mechanisms through which hypercalcaemia occurs in paediatric practice. (25 marks)

(b) List five [05] clinical manifestations of hypercalcaemia. (25 marks)

4.3. Outline the initial mamagement of a child with hypercalcaemia. (20 marks)

Q.5.

5.1. Describe the pathogenesis of migraine. (20 marks)

5.2. Briefly describes the different types of migraine. (25 marks)

5.3. Outline the management of migraine. (30 marks)

5.4. Name two [02] clinical entities that are recognized precursors of migraine. (10 marks)

5.5 Outline the clinical features and management of the conditions mentioned in 5.4. (15 marks)

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Date: 25th January 2012 Time: 9.00a.m.-12.00 noon

PAPER 11 – CASE HISTORIES

Answer all five questions
Answer each question in a separate book.

1. A 7 year old boy is admitted with malaise and lethargy of 3 weeks duration. On examination his height is below the 3rd centile and weight is at 10th centile. He is afebrile and pale with no jaundice, no lymphadenopathy or bone tenderness. There is hepatomegaly of 5 cm, splenomegaly of 7 cm, right sided optic atropy and impaired hearing on left side. Rest of the clinicl examination is unremarkable.

Investigations :

Hb	7g/dl
WBC	$2.3X10^9/1$ N 25%, L 60%, M 10%, E 5%
Platelets	$93 X 10^9/1$
Blood Film	pancytopenia with myeloid precursors and nucleated red cells. No blast cells
ESR	10mm, in the first hour
Ultrasound Scan of abdomen	confirmed hepatospenomegaly, no other masses

1.1. What is the most likely diagnosis ? (20 marks)

1.2. Mention two [02] investigations that would confirm the diagnosis and give the expected findings. (30 marks)

1.3. Mention two [02] other complications of this disease. (20 marks)

1.4. Name two [02] drugs that are useful in the treatment. (30 marks)

2 A 3½ year old boy was brought to the Emergency department by his grandmother with whom he had been spending the day whilst his parents were away. He was apparently well and active when the parents handed him over in the morning, but at noon when she tried to feed him she noted that the child was breathless. He complained of abdominal pain, vomited several times and then became progressively lethargic and drowsy.

Apart from frequent mild upper respiratory tract infections, he has been a healthy child. He is the only child born to non consanguineous parents who are teachers. His birth weight was 3 kg. His grandfather is on treatment for dementia, ischaemic heart disease and cerbrovascular accident and is totally dependent on his wife for care.

On examination he was unconscious and febrile. (39.5^0C). His respiratory rate was 45/min with clear lungs on auscultation. His pulse rate was 125/min, regular with good volume, Blood pressure was 80/50. Heart was in dual rhythm with no murmurs. Abdominal examination was unremarkable and there were no focal neurological signs.

2.1. Mention two [02] possible conditions you would suspect prior to performing any investigations. (15 marks)

2.2 What additional information would you elicit from the history and examination to arrive at a definite diagnosis ? (20 marks)

Subsequently the following investigation results became available.

Hb	13g/dl	
WBC/DC	8.2 x 10^9/L , N-60%, l-35%, E-3%, M-2%	
UFR		
pH	5	
Alb	Nil	
Reducing substances	++	
Cells	1-2/hpf.	
Blood urea	50mg/dl	(15-40 mg/dl)
Serum creatinine	0.85 mg/dl	(0.2-0.9mg/dl)

Serum electrolytes		
Sodium	128mmol/l	(135-145mmol/l)
Potassium	2.8mmol/I	(3.5-5.5mmol/l)
Random Blood Sugar	8mmol/l	

2.3 What is the diagnosis ? (15 marks)

2.4 Give three [03] investigations that you would now perform. (20 marks)

2.5 Briefly outline the principles of management of this child. (30 marks)

3. A 7 year old child presented with productive cough, difficulty in breathing and low grade fever of 3 weeks duration. He has had two episodes of mild haemoptysis. He has also had some weight loss.

He was born to 2nd degree consanguineous parents. The birth weight was 3 kg. Neonatal period was uneventful.

He gives a history of chronic rhinorrhoea and recurrent otitis media since the age of 10 months. He had undergone myringotomy with pressure equalizing tube placement. Audiometry had shown conductive hearing loss.

At the age of 4 years he was admitted for a lower respiratory tract infection with wheezing and was treated with IV antibiotics and bronchodilators. Since he had a persistent cough, he was also started on steroid prophylactic inhaler therapy and followed up at the paediatric clinic. He was treated with oral antibiotics whenever he had a mild respiratory tract infection.

There is no history of diarrhoea, abdominal symptoms, skin rashes or bleeding manifestations. Immunizations were given according to EPI schedule. Both his parents are healthy and he has a healthy 10 year old sister.

On examination weight was on the 3rd centile and the weight for height was below 2SD (WHO). Finger clubbing was present. There was no cervical lymphadenopathy and BCG scar was present. Skin and mucous membranes were normal. Respiratory systems examination revealed respiratory rate of 30/min with sub-costal recessions. On auscultation bilateral coarse basal crepitations were present.

Examination of cardiovascular system, abdomen and nervous system were clinically normal.

Investigations

FBC		
	Hb	10g/dl
	WBC	14 x 10^9/L
		N-63%, L-33%, E- 4%
ESR		76mm/1st hour
CXR		prominent bronchovascular markings with some Inflammatory shadows

Sputum for culture	growth of streptococcus pneumonia and moraxella catarrhalis
Mantoux test	negative
Pulmonary function assessment	
FVC	2.84 L (72% of predicted)
FEVI	2.05 L (64% of predicted)
FEVI/FVC	72%
HRCT of the chest	signet ring appearance seen bronchi with beaded contour seen in some segments of the lungs

Serum electrolytes, liver function tests, renal function tests – normal

3.1 What is the diagnosis ? (10 marks)

3.2. Mention four [04] underlying causes which could have led to this condition. (20 marks)

3.3. List five [05] investigations you would perform to identify the underlying cause giving the expected results for each. (40 marks)

3.4. Outline the management of this child. (30 marks)

4. A 4 day old baby was brought with a history of jaundice of 1 day duration. He was born by a normal vaginal delivery at term. Birth weight was 3.0Kg., Apgar score was 8 at one minute and 10 at five minutes and the baby was given to the mother to breast feed soon after birth. The perinatal period was uneventful and mother and baby were discharged on day two. He had been feeding well at home but the mother noticed that the baby was icteric on the 3rd day after birth. Since the jaundice was gradually worsening, the baby was brought to hospital.

The mother's blood group was O positive. She was healthy with no history of post partum problems.

This was the second child of a non consanguineous family. There was a similar history in the elder boy who also had a similar neonatal history. He had been treated with phototherapy and had received SCBU care for one week. He had required two blood transfusions at the age of 3 and 5 years and is now an apparently healthy 10 year old school boy.

On examination, the baby was afebrile and active but icteric. There was no obvious pallor. The liver was palpable (1cm) and the spleen was just palpable below the costal margin.

Investigations :

Serum bilirubin	422µmol/L (total) Indirect fraction – 402 µmol/L
FBC:	
Heamoglobin	14.0g/dl
WBC count	9 x $10^{9/}$L Neutrophils – 60% Lymphocytes – 40%

4.1. State three [03] likely causes for this baby's jaundice. (20 marks)

4.2. Enumerate what further investigations you would plan in this baby to arrive at a diagnosis and give the expected results. (30 marks)

4.3. Compare and contrast the long term complications and therapeutic options in two of the conditions mentioned in 4.1. (50 marks)

5. A 10 year old girl has had several episodes of abdominal pain associated with vomiting for the last two months. The pain was episodic and she was admitted to a paediatric ward for observation and evaluation. On admission she had marked vomiting and developed a generalized tonic clonic seizure.

Mother revealed that there was loss of appetite and loss of weight over the last two months. The child had felt faintish and weak and returned from school on several occasions.

She was born at term. There were no antenatal of postnatal complications. Immunization and development are age appropriate. There were no other significant illnesses in the past.

Parents are non consanguineous. Mother is taking thyroxine for the last 8 years. Father and the older sister are healthy.

On examination her weight was below the 3rd centile and height was at the 10th centile. She was ill , thin and wasted with a dark complexion. There were no rashes. There was significant dehydration and pallor. Bilateral cervical and axillary lymphadenopathy were noted. BCG scar was present. Abdomen was soft and non tender. Liver was just palpable and the spleen was palpable 2 cm. below the costal margin. Cardiovascular, respiratory and nervous systems are normal.

Investigations :

WBC	4.8 x $10^{9/L}$	
	N 55%, L 45% , E 5%	
Platelet count	180 x 10^{9}/L	
Hb	6 g/dl	
Serum sodium	118 meq/L	(135 – 145)
Serum potassium	5.7meq/L	(3.5 – 4.6)
Blood urea	33 mg/dl	(7 – 18)
Random blood sugar	52 mg/dl	(60 – 100)
ALT	126 iu/L	(10 – 40)
AST	180 iu/L	(9 – 48)
Mantoux test	negative	
Reticulocyte count	2.5%	

Coombs test	positive
Blood picture	No abnormal white cells few spherocytes
Bone marrow biopsy	normal active marrow. No evidence of Infiltration
Lymph node biopsy	reactive changes only
USS abdomen	mesenteric adenitis and splenomegaly

5.1. Give two [02] likely causes for the seizure. (10 marks)

5.2. Give the most likely complete diagnosis. (30 marks)

5.3. Give five [05] further investigations that are relevant and reasons for their selection. (30 marks)

5.4 Outline the management in the first 24 hours. (30 marks)