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## POSTGRADUATE INSTITUTE OF MEDICINE UNIVERSITY OF COLOMBO

## MD (PAEDIATRICS) EXAMINATION – JANUARY/FEBRUARY 2017

**Date :-** 24<sup>th</sup> January 2017

**Time :-** 9.00 a.m. – 12.00 noon

### PAPER I (STRUCTURED ESSAY QUESTIONS)

Answer all five questions.

Answer each question in a separate book.

#### **Q.1.**

- 1.1.
  - 1.1.1. Name the species of Shigella that are known to cause bacillary dysentery. (10 marks)
  - 1.1.2. List five (05) extra intestinal manifestations of Shigella infection. (20 marks)
  - 1.1.3. Outline the steps in the initial management of an 11 year old boy with bacillary dysentery, who presents with hypotension and a capillary refill time of 3 seconds. (30 marks)
- 1.2.
- 1.2.1. Describe the mechanism of action of oral rehydration solution. (25 marks)
- 1.2.2. Mention the osmolarity and the composition of oral rehydration solution. (15 marks)

#### Q.2.

2.1. Explain the term "genetic counselling".

(20 marks)

- 2.2. Briefly describe how you would prepare yourself for a genetic counselling session. (40 marks)
- 2.3. Outline the specific categories of medical therapies used in the management of genetic disorders. (20 marks)
- 2.4. Name two (02) types of non-traditional inheritance and give one example for each type. (20 marks)

#### Q.3.

3.1. State the aetiological classification of vesicoureteral reflux (VUR).

(20 marks)

3.2. Explain the relative advantages and disadvantages of different imaging tests in diagnosing VUR. (30 marks)

3.3. Outline the clinical implications of VUR

(20 marks)

3.4. Briefly discuss the management of a child with a dilating grade of VUR.

(30 marks)

#### Q.4.

4.1. Describe the patho-physiology of sickle cell disease (HbSS). (30 marks)

4.2. Describe briefly the important complications known to occur in five (05) different organs of the body due to sickle cell disease (HbSS). (50 marks)

4.3. List the treatment strategies of sickle cell disease (HbSS). (20 marks)

#### Q.5.

5.1. Describe briefly the following terms in relation to allergic diseases.

5.1.1. Allergic March

(15marks)

5.1.2. Hygiene hypothesis

(15marks)

5.1.3. Oral tolerance

(15 marks)

- 5.2. A 1 year old child is brought by the mother who claims that the child is allergic to eggs.
  - 5.2.1. State five (05) features in the history that would help in making a clinical diagnosis in this child. (25 mark)
  - 5.2.2. State three (03) confirmatory tests you would perform to confirm egg allergy. (15 marks)
  - 5.2.3. Outline the principles of management of a child with confirmed egg allergy. (15marks)

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#### **PAPER II – CASE HISTORIES**

Answer all five questions.

Answer each question in a separate book.

1. A baby boy was born by normal vaginal delivery at 37 weeks gestation to a 32 year old mother in her second pregnancy. The pregnancy had been complicated by gestational diabetes, needing insulin. The latest HbA1C was 8.5. The baby cried at birth and the Apgar at one minute and five minutes was 10.

The baby had fed well and the capillary blood sugar estimations were normal. However, at 8 hours of age, the baby was found to be lethargic and tachypnoeic.

Examination revealed a lethargic baby, with occasional grunting and the respiratory rate of 72/minute. Heart rate was 160/minute and capillary refill time was 4 seconds. No significant murmurs were heard. There was no hepatomegaly.

The oxygen saturation in the right upper limb was 80% and the left lower limb 70%. Chest X-ray revealed a white-out of the lung fields.

The investigations are as follows:

Capillary blood sugar was 112 mg/dl.

Arterial blood gas analysis revealed:

рН	7.21	(7.35-7.45)
$PO_2$	40 mmHg	(50-70)
PaCO <sub>2</sub>	50 mmHg	(35-45)
$HCO_3$	13 meq/L	(20-24)
BE	-15	(+/-5)
WBC	$18 \times 10^9$ /L N-80%	
PCV	51%	
Platelet	$108 \times 10^9 / L$	(250 - 450)
CRP	0.5 mg/L	(<6)

- 1.1. List five (05) differential diagnoses for the above presentation.

  (40 marks)
- 1.2. Describe the management of this neonate in the next 6 hours. (40 marks)
- 1.3. Enumerate five (05) important investigations that would be helpful in the management. (20 marks)

2. A 10 year old girl was referred by the school medical officer for shortness of breath while walking, for 2 months. She was in the school netball team, but had been unable to perform well in the recent past. She attributed this to her knee joint pain and tiredness. Moreover, her school performance has also deteriorated.

The family physician, who examined the child could not find anything abnormal, and prescribed flunerazine for one month. For the last 2 weeks she had refused to go to school due to increasing tiredness.

She is the 3<sup>rd</sup> child of non-consanguineous parents and from a poor socio-economic background. However, her 2 sisters are studying in the same school and performing well academically.

On examination, she was afebrile, ill looking, and pale. She was found to have puffiness of face with oedema of legs. The heart was in dual rhythm and with no audible murmurs. The abdomen was distended with 2 cm tender hepatomegaly and ascites. The air entry was equal bilaterally.

Multipara monitoring revealed a heart rate of 170/minute, a respiratory rate of 44/minute and sinus tachycardia with low voltage complexes. The blood pressure was 84/60mmHg.

The chest X-ray showed a globular heart with cardiomegaly and obliteration of both costophrenic angles.

Haemoglobin	8.8g/dl	(12-15)
WBC/DC	$5.5.0 \times 10^9/L$	N - 57%, L - 40%, E - 03%
Platelet count	$100 \times 10^9 / L$	(150-450)
Serum Creatinine	96 μmol/L	(20-80)
UFR		
Protein	++	
Pus cells	2-3/hpf	
RBC	15-20 /hpf	
Urine culture	No growth	
CRP	0.8mg/L	(<0.8)
Blood culture	Sterile	
Albumin	4.6 g/dl	(3.5-5.5)

- 2.1. Give three (03) possible causes for the above presentation. (30 marks)
- 2.2. Mention the immediate intervention needed. (20 marks)
- 2.3. Name five (05) investigations which would help you to arrive at a diagnosis. (20 marks)
- 2.4. Outline the principles of management of the most likely diagnosis. (30 marks)

3. A 13.8 year-old girl presented with a history of headache for 2 months duration. Brain imaging was performed. She was admitted to neurosurgical unit for surgical intervention. However, she was referred to paediatrician for a comprehensive evaluation.

She is the 3<sup>rd</sup> child of nonconsanguineous healthy parents, from a poor socioeconomic background. She was born at term with a birth weight of 2.9kg and her postnatal period was uneventful.

Her immunization and development were age appropriate. She studied up to grade 6. However, she had dropped out because of declining school performance. She has been apparently healthy until this presentation and was not on any medications.

Recently, she developed a headache which was progressively increasing in severity. She had no history of visual problems or vomiting. According to mother, she has been a quiet child and a poor eater. She had a single episode of vaginal bleeding at the age of 12 years. There is no family history of delayed puberty.

On examination, her weight was 34 kg (weight age was 10.5 years) and the height is 115cm (height age is 6 years). The mid parental height was 162cm (just above 25<sup>th</sup> centile). She was pale but not icteric. There were no signs of micronutrient deficiencies. She did not have a goiter. She was noted to have a depressed nasal bridge and puffiness of the face. Her hair was thin and brittle and the skin was dry and coarse. The Tanner staging revealed B-2, P-1. Her blood pressure was 90/60 mmHg and the pulse rate was 64/min with a regular rhythm. There were no heart murmurs.

## Investigations:

FBC- Hb 8g/dl

Renal and liver functions were normal.

Serum cholesterol 7.1 mmol/L (4.1 - 5.2) Triglycerides 3.2. mmol/L (<2.3)

MRI report 1.5 cm x 1.8 cm homogenous

sellar-suprasellar lesion.

USS of the pelvis Bilateral enlarged multicystic ovaries

with a small uterus

Hormonal profile		
Prolactin	50 ng/mL	(4.5-23)
LH	0.9 mU/mL	(2.4 - 12.6)
FSH	8.6 mU/mL	(3.5 - 12.5)
Oestradiol	0.21 nmol/L	(0.20.8)
Cortisol (8am)	220 nmol/L	(230 - 750)
T3	0.3  ng/mL	(0.8 - 2)
Free T4	$0.4  \mu \text{g/dL}$	(1.5 - 2.9)
TSH	800 µIU/L	(0.27 - 4.2)

- 3.1. What is the most likely (complete) diagnosis? (20 marks)
- 3.2. List two (02) other possible conditions for this clinical presentation. (20marks)
- 3.3. Mention two (02) other relevant investigations you would perform. (10 marks)
- 3.4. Outline the management of this child. (30 marks)
- 3.5. With appropriate management, describe briefly the expected overall outcome of her problems. (20 marks)

4. A 12 year old girl was referred to the paediatric clinic for evaluation of fatigue of 3 months.

She was a member of the hockey team at school. She broke into tears when she said that she had had to give up hockey one month ago due to unusual tiredness she experienced with practices. The general practitioner had performed several blood tests which were normal. He had prescribed a multivitamin and asked her to rest at home. Her mother says that she had been very cheerful and was always smiling. However, this had changed recently. Her fatigue became worse despite the treatment given. She also experienced double vision when she did some reading in the night.

She has had an unremarkable past history. She was the second child of non consanguineous parents. Except for asthma, in her elder brother, other family members were healthy.

On examination, she did not have skin rashes or lymphadenopathy. She was found to have normal intelligence. Her face was expressionless. The double vision that she complained about was intermittent and had no particular pattern. It was difficult for her to stand up from the squatting position. Deep tendon reflexes and sensory examination were normal.

The investigations are as follows:

Haemoglobin	11.3 g/dl	(12-15)
WBC	$9.8 \times 10^9 / L$ N- 55%	6, L-40% E-5%
Platelet count	$285 \times 10^{9}/L$	
RBC	$3.9 \times 10^{9}/L$	
MCV	75fl	(78-90)
MCH	20pg	(27-30)
MCHC	30g/dl	(32-35)
ESR	20mm/hour	, ,
C-Reactive protein	4mg/dl	(<6)
Creatine phosphokinase	200	(<6) (70-367)
Anti nuclear antibodies	Negative	
Rheumatoid factor	Negative	
Thyroid function tests	Normal	

4.1. Mention the most likely diagnosis.

(20 marks)

4.2. Give one (01) other possible diagnosis.

(10 marks)

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4.3. List three (03) other clinical features that would support the diagnosis mentioned in 4.1. (30 marks)

4.4. Suggest four (04) other useful investigations with expected findings in the diagnosis mentioned in 4.1. (40 marks)

5. A 12 year old boy presented to the gastroenterology clinic for routine follow up. He complained of a chronic cough of 3 months. He had been treated by two general practitioners with oral antihistamines and antibiotics before clinic visit. Although it started as a mild nocturnal cough, now it is more "moist", and noted both during day and night. His mother stressed that the cough was getting progressively worse and there were audible wheezy noises in his chest especially when he sleeps.

He was diagnosed to have severe Chrons disease at the age of 4 years. The boy had received several courses of both intravenous and oral steroids to control his colitis. He was commenced on oral mesalazine as his maintenance drug but changed over to azathioprine (50 mg daily) as the disease could not be controlled with mesalazine. Last year, he had developed a perianal fistula and was had been treated with monthly intravenous infliximab (anti-TNF antibody) for 6 months. His current medications include azathioprine and a calcium supplement. Chrons disease had been well controlled in the preceding 6 months. At present, he has a normal appetite although he feels miserable at times.

On examination, he looks unwell. His temperature is 99.8° F. His height and weight are both below 3<sup>rd</sup> centile. There is no lymphadenopathy. Respiratory rate is 36 cycles per minute. Air entry to the right lung base is reduced. The abdominal examination revealed no organomegaly and no active lesions were found in the perianal area.

His investigations are as follows:

Haemoglobin WBC/DC Platelet count ESR C reactive protein Serum sodium Serum potassium Blood urea	6.2 g/dL 3.9 x10 <sup>9</sup> /L N - 30%, L 114 x10 <sup>9</sup> /L 60 mm/1 <sup>st</sup> hour 12 mg/dL 138 mEq/L 4 mEq/L 5.2 mmol/L	(11-16) - 70% (150-400) (<20) (<6) (135-145) (3.5-6.0) (4 - 6)
ALT	26 IU/L	(4 - 6)
AST Chest X-ray	Asymmetrical widening of the mediastinum with collapse of the right ower lobe.	

5.1. Mention the most likely diagnosis. (20 marks)

5.2. Give one (01) other possible diagnosis. (10 marks)

5.3. Discuss the investigations you would request to arrive at a diagnosis. (20arks)

5.4. Outline the management of this child. (50 marks)