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### POSTGRADUATE INSTITUTE OF MEDICINE UNIVERSITY OF COLOMBO

# **MD (PAEDIATRICS) EXAMINATION – JULY/AUGUST 2012**

**Date :-** 16<sup>th</sup> July 2012

**Time :-** 9.00 a.m. – 12.00 noon

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## PAPER I STRUCTURED ESSAY QUESTIONS

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

# Q.1.

- 1.1. Briefly describe the diagnostic criteria of Systemic Inflammatory Response Syndrome (SIRS). (15 marks)
- 1.2. Define sepsis and severe sepsis. (15 marks)
- 1.3. The spectrum of meningococcal disease varies widely. Briefly describe the recognized clinical patterns of this disease. (30 marks)

1.4. List **five (05)** morbidities which should be looked for when reviewing a child in the clinic following discharge after a severe meningococcal illness. (10 marks)

- 1.5. Briefly describe preventive health measures needed when there is a 4 year old child with meningococcal infection in the ward. (20 marks)
- 1.6. List four (04) indications for prescribing meningococcal vaccine in Sri Lanka. (10 marks)

Contd...../2-

- Q.2.
- 2.1. Briefly describe the hyperoxia test indicating its clinical use. (30 marks)
- 2.2. Explain giving examples the palliative interventions performed for cyanotic heart disease in the newborn period. (30 marks)

2.3. Outline six (06) preoperative life threatening complications of cyanotic congenital heart disease and describe in detail the pathophysiological basis for each. (40 marks)

# Q.3.

- 3.1. Mention two (02) autosomal dominant and two (02) autosomal recessive gene disorders which predispose to childhood malignancy. (10 marks)
- 3.2. Describe the clinical features of two (02) gene disorders (one autosomal dominant and one autosomal recessive) that predispose to childhood cancer which also include skin manifestations. (40 marks)
- 3.3. Discuss the growth abnormalities that occur as complications in childhood cancer survivors. (40 marks)
- 3.4. Enumerate five (05) other long term sequele of childhood cancer. (10 marks)

Contd...../3-

Q.4.

- 4.1. Mention five (05) causes of nephrocalcinosis in a 5 year old child.(20 marks)
- 4.2. Name the most sensitive investigation that detects nephrocalcinosis.

(10 marks)

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- 4.3. Mention five (05) other investigations you would perform in the above child, briefly explaining how each investigation would help in the diagnosis.(35 marks)
- 4.4. State one of the commonest causes of nephrocalcinosis in children and describe its treatment. (35 marks)

#### Q.5.

5.1. Define encephalopathy. (10 marks)

- 5.2. Give six (06) different categories of aetiological agents which cause encephalopathy in children. (30 marks)
- 5.3. Briefly describe the interventions that would minimize the incidence of encephalopathy in children aged 0 12 years. (40 marks)
- 5.4. Enumerate the principles of management of raised intracranial pressure. (20 marks)



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### PAPER II – CASE HISTORIES

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

1. A 7 year old girl was referred for admission to the paediatric ward, by school medical officer for ankle oedema.

She had been feeling unwell and lethargic for the past 3 months. There had been abdominal discomfort and she had frequently complained of pain in legs. There was no cough or shortness of breath. No urinary symptoms. She had poor appetite and loose stools about 5-6 times per day for past two weeks. Other than infrequent upper respiratory tract infections, there are no significant illnesses in the past. Her class teacher had noted that her school performance had deteriorated recently and that she had been absent several times during past one month.

She is the eldest child born to non consanguineous parents. There are two younger siblings who are healthy. Father is on treatment for chronic asthma.

Her immunization status is up to date.

Contd..../2-

On examination, she was cooperative and well oriented. Height was on the 50<sup>th</sup> centile, weight on the 10<sup>th</sup> centile. Temperature was 37.8°C. She was pale but not icteric. There were enlarged cervical lymph nodes and bilateral pitting oedema of both ankles. BCG scar was present. Cardiovascular, respiratory and nervous systems were clinically normal. Abdomen was distended, liver palpable 2cm below right costal margin and spleen not palpable. There was shifting flank dullness. Inward urine analysis was negative for proteins and glucose.

Investigations:

Haemoglobin	9 g/dl	
White Blood Cell Count	8.5 x 10 <sup>9</sup> /L	
	N - 65%, L - 2	5%, E-10%
Platelet	260 x 10 <sup>9</sup> /L	
ESR	75 mm in 1 <sup>st</sup> hour	
S. creatinine	0.5 mg/dl	(0.3-0.7)
S.sodium	130 mmol/L	(135 -140)
S.potassium	3.8 mmol/L	(3.5-4.5)
Total Protein	4.8g/dl	(6.5 - 8.0)
S. albumin	2.2 g/dl	(4.0 - 5.3)
ALT	25 IU/L	(10 - 40)
AST	30 IU/L	(10 - 48)
S. Bilirubin	12 μmol/L	(3 - 20)
Urine full report		
Protein	nil	
Sugar	nil	
Pus cells	2 - 3/field	
Chest X-Ray	Normal	

1.1. Explain the pathogenesis of oedema in this child. (20 marks)

1.2. Name the most important screening test to diagnose the aetiology of hypoprotenaemia. (20 marks)

2

Contd..../3-

- 1.3. State the three (03) most likely underlying diagnoses that you would consider in this child. (15 marks)
- 1.4. Mention two (02) investigations each to confirm each of the above diagnoses you mentioned in 1.3., giving reasons. (30 marks)

Two days after admission to ward, her oedema and ascites worsened and her urine output was found to be reduced.

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1.5. What is the most effective, immediate therapeutic measure that is indicated at this point? (15 marks)

Contd..../4-

2. A 9 year old boy was admitted with a history of limp and pain in the right hip joint. Four days ago he developed fever and vague abdominal pain while participating in a chess tournament for which he had been away from home. He was treated with two days of amoxycillin by a general practitioner and as he had some improvement he continued to play in the tournament even though he had fever and pain, on and off. The morning after he returned home he was found to have high fever and was unable to walk.

On examination he was febrile, movements at hip joint were painful and restricted. He had abdominal pain with mild right sided tenderness. He was not pale and rest of the examination was normal.

Urgent ultra sound examination of the hip revealed "normal joint space." Blood culture was performed and IV cefuroxime was commenced.

Hb	12g/dL	
WBC	$21 \times 10^{9}/L$	
	N - 80%, L - 20%.	
Platelet count	210 x 10 <sup>9</sup> /L	
ESR	60 mm 1 <sup>st</sup> hr	
C reactive protein	96 g/L (<6 g/L)	

Following day, the child was very ill with severe abdominal pain and vomiting. There was generalized abdominal tenderness with guarding. His temperature was 39°C, pulse 112/minute and blood pressure 80/60 mmHg.

2.1. State the **complete** diagnosis.

- 2.2. Outline the principles of the **immediate management** in this child. (30 marks)
- 2.3. List **four** (4) possible reasons that may have delayed the diagnosis resulting in this complication . (40 marks)

Contd...../5-

(30 marks)

3. A 12 year old girl presented to outpatients department with a history of tiredness and school refusal.

She had been well until 4 weeks ago when she complained of mild abdominal pain, loss of apetite and occasional bouts of vomiting. Her urine was tea coloured but resolved in a few days. There was no dysuria or loose stools.

She had not taken any medications recently and the family was healthy.

On examination child was febrile (39.5 °C), pale, and there were multiple petechae on her trunk. Pulse 112/min, Blood Pressure 100/70, systolic murmur heard over the precordium. Respiratory rate 24/min and lungs were clear. Liver was enlarged 3 cm below right costal margin and spleen was just palpable. Central nervous system was normal. Her height was at 75<sup>th</sup> centile and weight at 50<sup>th</sup> centile. There was no dysmorphism or any skin manifestations.

She was admitted to ward for further management.

The results of the blood investigation s are as follows.

6.7 g/dl
12x10 <sup>9</sup> /L
$1.0 \times 10^{9}$ /L N -0.2 x $10^{9}$ /L,
$L = 0.7 \times 10^9 / L$ , $E = 0.1 \times 10^9 / L$ ,
<0.5%,
report awaited
56 μmol/L
40 µmol/L
70 U/L (<40)
930 U/L (<40)
1100 U/L

Contd...../6-

- 3.1. List the **three (03) most likely diagnoses** you would consider in this child. (30 marks)
- 3.2. Describe the **investigations and expected results** that would help you to arrive at a diagnosis. (30 marks)
- 3.3. Outline your **management in the first 24 hours** (other than the diagnostic investigations you have mentioned above). (40 marks)

THE REPORT

Contd..../7-

4. A 7 year old boy was admitted to the paediatric unit with a generalized tonic-clonic seizure and continued to develop more seizures while in the ward. He has had five similar generalized tonic- clonic seizures within the last two weeks each lasting between 5 to 15 minutes. He was previously known to have a seizure disorder with learning difficulties but was off anticonvulsants for the past one year.

He was born to healthy non consanguineous parents at 32 weeks of gestation weighing 1.7 kg and had a history of hypoglycemia and meningitis during the neonatal period. At one year of age he developed his first afebrile seizure. He was initially started on sodium valproate but after one year it was changed to carbamazepine as he developed Steven-Johnson syndrome with valproate. After starting carbamazepine his seizures were largely controlled and the seizure frequency was only 2-3 per year.

At 5 1/2 years of age, there is a history of high grade continued fever lasting over two months, without response to antibiotics. During this admission he had arthritis of small and large joints, oral ulcers and a photosensitive rash. Though he had no urinary symptoms there was persistent moderate proteinuria. Blood picture showed pancytopenia without evidence of aplastic anaemia or infiltration in bone marrow. His serum was positive for both Anti Nuclear Antibodies and double stranded DNA. He was treated with prednisolone and he became clinically well. During this admission carbamazepine was changed to phenobarbitone.

Two months after changing to phenobarbitone he again developed a Stevens–Johnson syndrome like rash and phenobarbitone was discontinued. He was started on resperidone as he had some hyperactivity.

Since then (for over one year) he has been off all anti-consultants but was seizure free until the last 2 weeks. Apart from the recent seizures he has been relatively well during this period and he had even shown some improvement in school performance while on resperidone.

Contd..../8-

- 4.1. Give **two (02) possibilities** for his clinical presentation at 5 ½ years. (20 marks)
- 4.2. Briefly discuss how you would attempt to differentiate the above two conditions. (15 marks)
- 4.3. Name the **three (03)** most important investigations that would benefit his seizure management at present. (15 marks)
- 4.4. What factors would you consider when prescribing long term anticonvulsant/s at the current admission?
  Include your main concerns and considerations in this decision making process, giving specific examples. (50 marks)

Contd...../9-

8

5. Baby weighing 2.9 kg was born at term by elective caesarian section due to placenta previa. Apgar 8 and 9 at 1 and 5 minutes respectively. There was no history of consanguinity. Ambiguous genitalia was noted at birth. Baby fed well at the breast.

Examination of genitalia: Phallus of 8 mm with urethral meatus at tip. Left scrotal sac - underdeveloped with  $0.7 \times 0.4$  cm mass felt within the sac. Right side scrotal sac absent with  $0.6 \times 0.3$  cm mass felt in the right inguinal canal.

Baby was admitted to the neonatal unit.

- 5.1. What differential diagnosis would you consider at birth? (10 marks)
- 5.2. What investigations would you perform to arrive at a diagnosis? (20 marks)

Baby was found to be pale with cold peripheries on Day 3.

Investigations performed at this stage:

WBC	8.6 x 10 <sup>9</sup> /L	
C reactive protein	3.8 mg/L	(< 6)
Procalcitonin	< 3pg/ml	(< 10 pg/ml)
S.sodium	130 mmol/L	(135 - 148)
S.potassium	4.8 mmol/L	(3.6 – 5)

State the complete diagnosis at this stage. (20

(20 marks)

Name one (01) investigation which is vital in the acute management at this stage. (10 marks)

List the therapeutic interventions needed in the long term management. (40 marks)