## POSTGRADUATE INSTITUTE OF MEDICINE UNIVERSITY OF COLOMBO

## MD (PAEDIATRICS) EXAMINATION – JANUARY/FEBRUARY 2021

Date: -26th January 2021

**Time:-** 9.00 a.m. – 12.00 noon

## PAPER I (STRUCTURED ESSAY QUESTIONS)

Answer all five (05) questions.

Answer each question in a separate book.

1.

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1.1.

- 1.1.1. Outline three (03) pathophysiological mechanisms that cause brain injury due to hypoxemic ischaemia. (30 marks)
- 1.1.2. Mention the criteria that should be fulfilled in a neonate to be considered for therapeutic cooling. (25 marks)
- 1.1.3. Outline the limitations that would be encountered in establishing an island wide therapeutic cooling service in Sri Lanka. (15 marks)

1.2.

- 1.2.1. Briefly outline how you would perform a pulse oximetry test and mention the most appropriate timing of this test in a newborn. (15 marks)
- 1.2.2. What is your inference on a negative pulse oximetry result in a newborn? (15 marks)

2.

2.1. Define prediabetes.

(10 marks)

- 2.2. Mention the diagnostic criteria for Maturity Onset Diabetes of Youth (MODY). (20 marks)
- 2.3. Discuss insulin resistance .

(30 marks)

2.4. Briefly describe the acute and long-term management of Type 2 Diabetes Mellitus (T2DM) in adolescence. (40 marks)

Contd..../2-

3.

6 ,

- 3.1. Describe the clinical and laboratory features of Auto Immune Hepatitis (AIH). (35 marks)
- 3.2. Outline the pharmacological basis of the **first line medications** used in the treatment of AIH. (25 marks)
- 3.3. What is meant by overlap syndrome in AIH? (15 marks)
- 3.4. Discuss the need and the challenges of establishing a liver transplant programme in Sri Lanka. (25 marks)

4.

- 4.1. What is paediatric palliative care? (20 marks)
- 4.2. Mention four (04) broad groups of conditions identified for paediatric palliative care giving one example for each. (20 marks)
- 4.3. Outline the **pharmacological approach** of pain management in paediatric palliative care. (20 marks)
- 4.4. A 6-month-old infant diagnosed with spinal muscular atrophy type-1 is admitted with episodes of apnoea. His breathing is shallow with a respiratory rate of 12 per minute and the oxygen saturation is 88% despite high flow oxygen. You wonder whether to intubate and ventilate.

Discuss the **ethical dilemmas** in relation to mechanical ventilation that are relevant to the above clinical scenario. (40 marks)

5.

5.1.

- 5.1.1. List three (03) broad categories of ventilation perfusion mismatch, giving one (01) example each. (30 marks)
- 5.1.2. Give five (05) clinical indications to commence respiratory support in a three-month-old previously healthy baby with SARS-COV-2. (25 marks)
- 5.1.3 Mention four (04) **physiological effects** of high flow nasal cannula oxygen therapy that is beneficial in managing SARS-COV-2. (20 marks)
- 5.2. Briefly describe the proposed vaccination programme for SARS-COV-2 in Sri Lanka and it's benefit to the paediatric population. (25 marks)

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## **PAPER II – CASE HISTORIES**

Answer all five (05) questions. Answer each question in a separate book.

1. An 8-week-old girl successfully underwent hepatoportoenterostomy (Kasai) for extrahepatic biliary atresia. Liver functions normalized completely within 3 weeks. At follow-up, growth and development were normal. At the age of 6 years, she presented with an episode of fever up to 39.5°C and pain in right upper-abdominal quadrant. The pain worsened with coughing and moving. She complained of nausea and vomited once. There was no jaundice. Micturition and defecation remained normal. On admission, serum aminotransferases were normal, but elevate to three times the upper limits of normal after a few days. There was marked leucocytosis. No infectious focus could be detected, and empirical treatment was instituted with broadspectrum antibiotics. Blood cultures remained negative, liver biopsy did not present any signs of inflammation, and ultrasound was indecisive. Her fever subsided, pain disappeared, and liver function normalized on discharge after 2 weeks of treatment.

At the age of 7 years she was admitted again because of a similar episode with high fever and pain in the upper right abdomen. No clinical signs of liver involvement were present. The same therapy was given and she recovered completely. However, few weeks later she had an identical episode and was admitted for further evaluation. On physical examination, she was not drowsy, not pale or icteric and there was no hepatosplenomegaly. Liver biopsy again did not show any abnormality and repeated blood cultures were negative.

The investigation findings were as follows:

Full blood count

Hb 12 g/dL (11 - 15)WBC  $23.2 \times 10^9/L$ (4-11)N-85%, L-15% Platelet count  $259 \times 10^{9}/L$ (150 - 400)Serum creatinine 60 µmol/L (44-110)Serum Ammonia 22 mmol/L (<40)

185 mg/L	(<6)
89 IU/L	(up to 40)
92 IU/L	(up to 40)
220 IU/L	(5-27)
1136 U/L	(104-345)
2.9  mg/dL	(0.2-1.2)
1.8  mg/dL	(0.4-0.6)
80  mg/dL	
70 g/dL	(65-75)
38  g/dL	(35-50)
1.1	
No growth	
Structurally normal hear	rt and great vessels
Normal	
	89 IU/L 92 IU/L 220 IU/L 1136 U/L 2.9 mg/dL 1.8 mg/dL 80 mg/dL 70 g/dL 38 g/dL 1.1 No growth Structurally normal hear

1.1. What is the most likely diagnosis?

(30 marks)

- 1.2. List five (05) other investigations which are useful to evaluate this child. (35 marks)
- 1.3. Briefly outline the management

(35 marks)

2. A 12-year-old boy presented with a history of intermittent headache for one week and three left sided focal seizures on the day of presentation. He required a midazolam bolus for termination of seizures. He has complained of blurred vision since morning of the day of presentation. During the past 3 months he has presented with recurrent short lasting episodes of respiratory difficulty during one of which he developed a mild haemoptysis. He has also complained of non specific body aches and pains.

On examination ten hours after the last seizure, he is drowsy with a GCS of 8. He has no movements on the left side of the body. There is left sided hypertonia and hyperreflexia. He has low amplitude twitching of eyelids and limb extremities. Rest of the neurological and systemic examinations are normal.

The vital parameters show pulse rate of 120/minute, blood pressure 140/90 mmHg and a respiratory rate of 23/minute.

The initial investigations showed:

Hb	13 g/dl	(11 -15)
WBC	$15 \times 10^9/L$	(4-11)
	N - 60%, L - 2	` /
Urine full report	•	,
Red blood cells	10/hpf	
Protein	+1	
Serum sodium	143 mEq/L	(135 - 145)
Serum potassium	4.5 mEq/L	(3.5-5.5)
ESR	45 mm/1 <sup>st</sup> hour	(<20)
Random blood sugar	110 mg/dL	` ,

- 2.1. Give two (02) most likely differential diagnoses for his acute neurological presentation. (20 marks)
- 2.2. Name three (03) factors that could be contributing to the persistent low arousal status. (30 marks)

Thirty minutes later he is found to have breathing irregularities and needed intubation. ENT assistance was needed for this due to presence of tracheal stenosis. The secretions were found to be clear during intubation.

- 2.3. Name two (02) investigations you would require urgently giving reasons. (20 marks)
- 2.4. What would be the most likely underlying diagnosis? (30 marks)

3. A 2-day-old male infant presented with poor feeding and shortness of breath for one day.

He was born by normal vaginal delivery to non-consanguineous parents at term with birth weight of 2.8Kgs. Antenatal and immediate postnatal period was uneventful up to discharge. There was no fever, vomiting or seizures. He had 2-year and 5-year-old healthy siblings.

Weight on admission was 2.5Kgs and occipital frontal circumference - 35cm. Anterior fontanelle was normal. There were no dysmorphic features or cleft lip or palate. The  $SpO_2$  was 85% in room air. He was active, afebrile and had normal tone, power and reflexes. There was no pallor, jaundice, or hepatosplenomegaly. Capillary refilling time was <2 seconds. Cardiovascular system was clinically normal. There were intercostal recessions with respiratory rate of 70 breaths/minute. Auscultation revealed bilateral occasional crepitation.

Investigations done during the acute stage.

Hb	15g/dl	(12-22)
WBC	$16.0 \times 10^9/L$	(9 - 30)
	N - 50%, L -	46%, M - 4%
Platelet count	$250 \times 10^9 / L$	(150 400)
CRP	4 mg/L	(<6)
Capillary blood sugar	6 mmol/L	(5-7)
Serum sodium	148 mEq/L	(135-145)
Serum potassium	4.2 mEq/L	(4-6)
Blood culture	Pending	
Venous blood gas	Normal	
Chest x-ray with NG tube	No abnormality	

Baby was managed overnight with intravenous fluid, oxygen and intravenous antibiotics. He was started on breast feeding on the following day as the child was getting better. Immediately after establishment of breast feeding, baby deteriorated rapidly with severe respiratory distress and cyanosis.

- 3.1. Mention the most likely complete diagnosis for the above presentation. (20 marks)
- 3.2. List two (02) investigations you would request to arrive at the diagnosis. (20 marks)
- 3.3. Outline the steps in the management. (30 marks)
- 3.4. Mention two (02) long term complications child might develop if definitive treatment is not provided. (10 marks)
- 3.5. Mention four (04) other associated problems you would look for. (20 marks)

4. An 8-year-old boy presented with generalized oedema, cough and difficulty in breathing of two days duration. There was no history of fever, jaundice or urinary symptoms.

He was born vaginally at term to healthy non consanguineous parents with a birth weight of 3 kgs. Neonatal period was uneventful. Immunizations were given according to EPI schedule.

At 2 years of age, the child presented with fever, cough and severe respiratory distress and was managed at PICU for three weeks. He was ventilated for 10 days and received several courses of intravenous antibiotics.

Since then he had recurrent episodes of wheezing and cough occurring once in 2 to 4 weeks. He had been treated with several courses of oral medications including steroids, nebulization and Beclomethasone MDI. There was family history of bronchial asthma but no contact history of tuberculosis.

On examination his weight was on the 95<sup>th</sup> centile and height was on the 3<sup>rd</sup> centile. He had generalized body oedema. His SpO<sub>2</sub> was 80% on room air.

There was no cervical lymphadenopathy and BCG scar was present. Respiratory system examination revealed a respiratory rate of 40 breaths/minute and hyperinflated chest with subcostal recessions. On auscultation bilateral rhonchi were present. His pulse rate was 120 beats/minute. The heart was in dual rhythm and had no murmurs. His blood pressure was 140/100 mmHg. He had 3cm tender hepatomegaly and no ascites. His central nervous system was clinically normal.

#### Investigations

Full blood count		
Hb	11.5 g/dL	(11 - 15)
WBC	$13.4 \times 10^{9}/L$	(4-11)
	N- 62%, L- 36%,	E- 2%
Platelet count	$233 \times 10^{9}/L$	(150 - 400)
CRP	40 mg/ 🛴	
ESR	30mm/1st hour	
Chest x ray (PA)	Hyperinflated lung field	ls
ABG		
pН	7.38	(7.35 - 7.45)
$PO_2$	70 mmHg	(80 -100)
$PCO_2$	70 mmHg	(35-45)
$HCO_3^-$	33 mmol/L	(22 - 24)
UFR		
Albumin	Nil	
Cells	No	
Serum sodium	137 mmol/L	(135 - 145)
Serum Potassium	4.2 mmol/l	(3.5-5.5)
Blood Urea	21mg/dl	(15-40)
Serum Creatinine	0.4mg/dl	(0.2-0.9)

Contd..../6-

ALT 20 IU/L (<40) AST 14 IU/l (<40)

Normal

Negative Negative

Negative

Ultrasound scan abdomen Mild hepatomegaly

Bilateral normal sized kidneys

Serum Immunoglobulin

HIV Screen
Mantoux test
Mycoplasma antibodies
Sputum TB PCR

Sputum TB PCR Negative CT scan chest Generaliz

Generalized mosaic pattern (Heterogenous appearance

areas of normal and air trapping)

## Pulmonary Function assessment

	Test value (L)	Predicted value (L)	% of predicted value	Post Bronchodilator Response(BDR)	% Change post BDR
FVC	0.85	1.3	65%	0.89	4%
FEV 1	0.56	1.12	50%	0.59	5% (significant BDR -12%)
FEV1/FVC	65%			66%	

- 4.1. List three (03) other clinical signs you would look for at presentation to arrive at a diagnosis. (15 marks)
- 4.2. What is your complete diagnosis? (40 marks)
- 4.3. List two (02) other investigations you would perform in this child. (10 marks)
- 4.4. List important steps in the management. (35 marks)

5. A 4-month-old baby boy was noted to have subcutaneous nodules over abdomen measuring 3-5 mms during the routine visit for vaccination. These nodules rapidly progressed to the rest of the body over the next 2 weeks. No history of fever or any systemic symptoms.

He was the first child of healthy nonconsanguineous parents. He was born by elective lower segment caesarean section at a gestation of 38 weeks following an uneventful antenatal period. His birth weight was 3.1 kgs. His neonatal period was uneventful. He is thriving well with no previous ill health and has normal development. His immunization is up to date.

On examination, he has pallor. There is no lymphadenopathy or hepatosplenomegaly. Multiple tender skin nodules are palpable over the abdomen, chest and limbs.

His initial investigations reveals:

Hb	9.4 g/dL	(10 - 15)	
WBC	$10.2 \times 10^9/L$	(4 - 11)	
	N - 84%, L	- 8%, M - 8%	
Platelet count	$402 \times 10^9 / L$	(150 - 400)	
ESR	38 mm/1 <sup>st</sup> hour		
CRP	6 g/dL	(<6)	
Blood picture	Hypochromic mic	rocytic red cells, white cells	
	morphologically r	ormal, reactive thrombocytosis	
Ultrasound scan of the lesions - Granulomas with liquefying centres			

- 5.1. What is the diagnosis for his acute presentation? (20 marks)
- 5.2. Name three (03) tests to confirm the above diagnosis. (30 marks)
- 5.3. What is the primary source of the lesions? (10 marks)
- 5.4. What is the underlying condition that you would suspect in this child? (20 marks)
- 5.5. Enumerate three (03) investigations that you would perform to confirm your diagnosis. (20 marks)