

POSTGRADUATE INSTITUTE OF MEDICINE
UNIVERSITY OF COLOMBO

POSTGRADUATE DIPLOMA IN CLINICAL HAEMATOLOGY EXAMINATION
JULY 2021

Date :- 14th July 2021

Time:- 1.00 p.m. – 4.00 p.m.

Answer **four (04)** questions only.
Answer each question **in a separate book**.
All questions carry equal marks.

PAPER I - ESSAY

1. Discuss how you would arrive at a diagnosis of severe acquired idiopathic aplastic anaemia in a 10-year-old boy presenting with pancytopenia.
Briefly describe the pathogenesis of this disease, and outline the management of this child. (100 marks)
2. A 56-year-old woman develops fever, chest pain and shortness of breath during a blood transfusion.
Discuss the possible causes for this presentation, pathophysiology and prevention of each cause. Outline the immediate bedside management of this patient. (100 marks)
3.
 - 3.1. Discuss how you would evaluate a patient with primary amyloidosis. (60 marks)
 - 3.2. Discuss the pathophysiology of bone disease in plasma cell myeloma. (40 marks)
4. Write notes on
 - 4.1. pathophysiology and types of lymphoma in HIV infection. (35 marks)
 - 4.2. laboratory diagnosis of acute lymphoblastic leukaemia. (35marks)
 - 4.3. tumour lysis syndrome. (30 marks)
5.
 - 5.1. Describe the pathophysiological basis for cytopenias and a leucoerythroblastic blood picture in primary myelofibrosis. (30 marks)
 - 5.2. Discuss the laboratory investigations you would perform to diagnose and prognosticate a patient suspected to have primary myelofibrosis. (70 marks)

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POSTGRADUATE DIPLOMA IN CLINICAL HAEMATOLOGY EXAMINATION
JULY 2021

Date :- 15th July 2021

Time:- 9.00 a.m. – 12.00 noon

Answer **six (06)** questions only.
Answer each question **in a separate book**.
All questions carry equal marks.

PAPER II
STRUCTURED ESSAY QUESTIONS (SEQ)

1. A 35-year-old man with haemoglobin E / β thalassaemia, has remained well, never required a transfusion and maintained a stable haemoglobin of 8.0 - 8.5 g/dL. Investigations during a routine clinic visit revealed a serum ferritin of 1500 ng/mL.
 - 1.1. Outline how you would investigate this patient further. (40 marks)
 - 1.2. Briefly discuss the pathophysiology of iron overload in major thalassaemia syndromes. (30 marks)
 - 1.3. Discuss the principles of the therapeutic use, monitoring and toxicity of the three main licensed iron chelation drugs. (30 marks)

2. A 62-year-old woman who had been previously well was admitted with a Hb value of 6.5g/dL. Auto immune haemolytic anaemia is suspected.
 - 2.1. Outline the complete diagnostic workup for this patient. (40 marks)
 - 2.2. Discuss red cell transfusion strategies for this patient. (30 marks)
 - 2.3. Briefly discuss the long term management options for this patient. (30 marks)

Contd..../2-

3. A 45-year-old man presents with cough and difficulty in breathing of two weeks duration and pruritus of five months.

His full blood count reveals

Hb	10.5 g/dL	(13.5 - 17.5)
WBC	$23.1 \times 10^9/L$	(4 - 10)
Neutrophils	$6.8 \times 10^9/L$	(2 - 7)
Lymphocytes	$3.1 \times 10^9/L$	(1- 3)
Monocytes	$0.7 \times 10^9/L$	(0.2 -1.0)
Eosinophils	$12.5 \times 10^9/L$	(0.02 - 0.5)
Basophils	$0.02 \times 10^9/L$	(0.02 - 0.1)
Platelet count	$150 \times 10^9/L$	(150 - 450)

- 3.1. Outline the relevant findings in the history and examination that you would elicit to arrive at a diagnosis. (20 marks)
- 3.2. Briefly discuss how you would investigate this man. (40 marks)
- 3.3. Outline the principles of treatment of this patient. (40 marks)
4. A 6 month old baby boy presented with the following full blood count.

Hb	6.2g/dL	(11.1 - 14.1)
RBC	$2.2 \times 10^{12}/L$	(4.1- 5.3)
MCV	108 fl	(68 - 84)
MCH	33 pg	(24 -30)
MCHC	36 g/dL	(30 -36)
WBC and platelet counts normal for age.		
Reticulocyte count	2.5%	(1.5-2.5)
Absolute reticulocyte count	$75 \times 10^9/L$	(40 -100)

His blood picture revealed oval and round macrocytes, teardrop red cells, few polychromatic cells, basophilic stippling and occasional NRBC.

The baby had no external dysmorphic clinical features.

- 4.1. Give the two (02) most likely diagnoses for the above findings. (20 marks)
- 4.2. Discuss the features you would elicit from the history and appropriate investigations you would do to arrive at a diagnosis, giving reasons. (40 marks)
- 4.3. Describe briefly the pathophysiology of the two (02) conditions you stated in 4.1. (20 marks)
- 4.4. Outline the principles of treatment for the two (02) conditions given in 4.1. (20 marks)

- 5.
- 5.1. An external quality assessment survey result for **prothrombin time** of your laboratory was reported as “unsatisfactory performance”. Write how you would investigate this problem. (35 marks)
- 5.2. Clinicians complain that the haemoglobin results of your laboratory varies in some patients. The IQC and EQA performance of the analyzers are satisfactory. Briefly state the probable causes and how you would address them. (35 marks)
- 5.3. List the causes that interfere with accurate haemoglobin A2 quantification in a thalassaemia diagnostic laboratory. (30 marks)
6. A 26-year-old pregnant woman at 30 weeks of POA is detected to have a platelet count of $80 \times 10^9/L$.
- 6.1. Discuss how you would investigate this patient to arrive at a diagnosis. (40 marks)
- At POA 34 weeks her platelet count is $20 \times 10^9/L$. She is clinically well. All other laboratory investigations are normal.
- 6.2. Explain the haematological management of this patient during pregnancy and delivery. (35 marks)
- 6.3. What advice would you give on the management of the newborn? (25 marks)
- 7.
- 7.1. Describe the pathophysiology including molecular genetics of congenital haemophilia A. (40 marks)
- 7.2. Discuss clinical applications of molecular genetic testing in haemophilia A. (30 marks)
- 7.3. State the haemostatic agents available for haemophilia A patients with inhibitors highlighting their mechanism of action. (30 marks)