POSTGRADUATE INSTITUTE OF MEDICINE
UNIVERSITY OF COLOMBO

POSTGRADUATE DIPLOMA IN CLINICAL HAEMATOLOGY EXAMINATION – DECEMBER 2011/JANUARY 2012

Date : 12 December 2011   **Time** 1:00 p.m. – 4:00 p.m.

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

PAPER I - Essay

1.

1.1 Discuss the importance of laboratory and clinical factors in the diagnosis of disseminated intravascular coagulation.   (70 marks)

1.2 Critically assess the available treatment options and when they should be employed.   (30 marks)

2. A 40 year old woman presented with a white cell count of 135,000/mm^3 and massive splenomegaly. Chronic myeloid leukaemia is suspected.

2.1 Discuss the molecular pathogenesis of this disease.   (30 marks)

2.2 Critically evaluate the different tests available to confirm the diagnosis, stating the sensitivity of each test.   (40 marks)

2.3 Describe the clinical and laboratory features that help to identify progression of this disease.   (30 marks)

3.

3.1 Describe the pathogenesis of disorders that arise from defects of red cell cytoskeleton (membrane).   (40 marks)

3.2 Critically evaluate the available diagnostic tests.   (60 marks)

4. Write short notes on

    4.1   Relationship between vitamin B_{12} and folate in the pathogenesis of megaloblastic anaemia.   (40 marks)

    4.2   Prognostic factors in acute lymphoblastic leukaemia.   (30 marks)

    4.3   Neonatal alloimmune thrombocytopenia.   (30 marks)

5.

    5.1   Briefly discuss the haematological changes in pregnancy (physiological).
        (50 marks)

    5.2   A woman in her 36th week of pregnancy was found to have red cell fragments in the peripheral blood with a Hb of 8.2 g/dl. Discuss the differential diagnosis. Outline the relevant investigations giving reasons.   (50 marks)

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Date : 13ᵗʰ December 2011   **Time** 9:00 a.m. – 12:00 noon.

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

PAPER II
STRUCTURED ESSAY QUESTIONS (SEQ)

1. A 55 year old female presented with dyspnoea on exertion and tiredness of two weeks duration. On examination she was pale and icteric with moderate splenomegaly. Investigations revealed Hb 5 g/dl. White cell count and platelet counts were normal. Serum indirect bilirubin 3 mg/dl. Coombs test positive with IgG and C3d.

   1.1   What further laboratory tests would you do on this patient?   (20 marks)

   1.2   Outline the management of this patient?   (30 marks)

   1.3   Briefly state the pathophysiology of this condition.   (20 marks)

   1.4   What problems would you encounter during pre transfusion testing to provide blood for this patient? How do you overcome these problems?   (30 marks)

2.

   2.1   List disorders that give rise to monoclonal gammopathy   (15 marks)

   2.2
   2.2.1. Which of the disorders you mentioned above have solid organ involvement?

   2.2.2. Which disorder has clonal neoplastic plasma cells proliferating in the bone marrow with minimal peripheral blood involvement?

   2.2.3. List the diagnostic criteria to differentiate the conditions given in 2.2.2. from a benign paraproteinaemia.   (25 marks)

   2.3   Describe the pathogenesis that gives rise to the clinical features seen in the disease that you mentioned in 2.2.2.   (40 marks)

   2.4   What criteria would you consider when staging the disease mentioned in 2.2.2?   (20 marks)

3.

    3.1   Briefly explain the role of spleen in haematopoiesis.   (30 marks)

    3.2   What are the peripheral blood and bone marrow changes in hypersplenism?
                (40 marks)

    3.3   Explain the mechanism of splenomegaly in myeloproliferative neoplasms.
                 (20 marks)

    3.4   Outline the pre operative management in a patient awaiting splenectomy.
                 (10 marks)

4.   A 40 year old female presented with sore throat and fever and was found to have a total white cell count of 1000/mm^3 with 18% neutrophils. Haemoglobin and platelet counts were normal.

    4.1   Comment on the above findings.   (10 marks)

    4.2   Give four (04) possible causes for the above findings.   (10 marks)

    4.3   Briefly describe the role of the neutrophil in infection.   (40 marks)

    4.4   In the absence of a clonal disorder, outline the management of this patient.
                 (40 marks)

5.   A 25 year old female is referred to you with the following investigations.

Hb	9.5 g/dl	
MCV	63.5 g/dl	
MCH	18.7 pg	
RBC	5.61×10^{12}/l	
Serum ferritin	166 µg/l	(normal 15 - 200µg/l)
Serum iron	84 µg/dl	(normal 37 – 145 µg/dl)
Total iron binding capacity	260 µg/dl	(normal 274 – 497 µg/dl)
Iron saturation	32%	

    5.1   Interpret the results with reasons and give the most likely diagnosis (es)
                 (20 marks)

    5.2   Hb electrophoresis at alkaline pH of this patient revealed absence of Hb A with Hb A$_2$ of 5.6% and a prominent band similar to electrophoretic mobility with sickle control sample. However the sickling test was negative.
        What is the possible diagnosis and what further investigations would you do to arrive at a diagnosis?   (30 marks)

5.3   What advice would you give to this person regarding future marriage?

(30 marks)

5.4   Briefly outline the pathophysiology of β thalassaemia major   (20 marks)

6. A 75 year old lady underwent emergency surgery for fracture neck of femur. Her pre operative haemostasis tests of PT/APTT were both in the normal reference range. However she bled from the surgical wound site during surgery.

6.1   Describe the conditions that can give rise to this situation.   (20 marks)

6.2   Discuss the value of pre operative screening tests for haemostasis.   (30 marks)

6.3   Write short notes on the role of the endothelium in haemostasis   (20 marks)

6.4   Outline the measures maintained in your laboratory to issue an accurate PT/APTT result.   (30 marks)

7. A 3 day old clinically well term newborn baby presented with jaundice. Serum bilirubin level 380 μmol/l. Hb 13.0 g/dl

7.1   List the possible causes of jaundice in this baby.   (10 marks)

7.2   Discuss the investigations which should be done to arrive at a diagnosis.

(30 marks)

7.3   What is the immediate management of this baby and what criteria would you use when deciding management options ?   (30 marks)

7.4   What selection criteria would you follow in choosing blood for this baby?

(15 marks)

7.5.   Outline the measures that can be adopted antenatally to prevent this condition?

(15 marks)