



The Royal College of **Pathologists**

Pathology: the science behind the cure

**Part 1 examination
Haematology: First paper
Tuesday 22 March 2016**

Candidates must answer ALL questions

Time allowed: Three hours

Question 1: General Haematology

A 16 year old non-European is referred for review in the haematology clinic with a history of 'thalassaemia' treated abroad with blood transfusions. Her Hb is 65g/L and her spleen is measured at 5cm below the costal margin

- a) Describe how you would assess her and the appropriate investigations that you would initiate in your first clinic.
- b) Outline what relevant review and tests would be needed to achieve a full annual review of her thalassaemia

Question 2: Transfusion medicine

What are the indications for transfusion of fresh frozen plasma and what evidence is there to support the use of this component? What are the potential adverse effects associated with FFP and how can they be prevented?

Question 3: Haemostasis and Thrombosis

You are asked to review a 28 year old female patient who is scheduled to undergo nasal polypectomy. You are informed that there is a history of von Willebrand disease in her family and she shows you a registration card from when she was 6 years old on which a VWF antigen level of 0.42 iu/ml is documented.

- a) Explain what you would do to review the diagnosis and the laboratory investigations you would perform.
- b) Discuss how the findings from question a) may affect how you advise on this patient's management.

Question 4: Haematological Oncology

A 29 year old man presents with general lethargy and petechial rash. He has no previous medical history.

His FBC shows Hb 61g/L, MCV 103 fL, WCC $0.9 \times 10^9/\text{L}$, Neutrophils $0.4 \times 10^9/\text{L}$, Platelets $11 \times 10^9/\text{L}$, Reticulocyte count $18 \times 10^9/\text{L}$.

The film shows round macrocytes, normal morphology otherwise and confirms genuine thrombocytopenia.

The bone marrow trephine reveals a profoundly hypocellular marrow with no significant dysplasia or blast cell infiltrate.

- a) Discuss any further investigations that are indicated into the diagnosis.
- b) Discuss the diagnosis and severity.
- c) Discuss the provision of blood product support for this patient.
- d) Discuss the treatment options for this patient.



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1. Discuss the clinical presentation and laboratory findings in a patient with suspected acquired haemophilia A.

Describe how you would further investigate and manage a newly diagnosed case in a 76 year old female

2. The transfusion practitioner informs you that a 32 year old woman blood group O negative has been given blood intended for another patient. How would you manage this incident and what steps would you take to prevent future errors?
3. You are contacted by your obstetric colleagues to consult on recently arrived 13 week pregnant lady with sickle cell anaemia. She has Hb of 80g/L and is confirmed Hb SS. Discuss all relevant aspects of the management of the case from this point forward to include antenatal supervision and a delivery plan.
4. A 46 year old man presented with lymphadenopathy and sweats. A PET-CT scan showed increased uptake in lymph nodes on both sides of the diaphragm, liver and spleen. A lymph node biopsy showed abnormal lymphoid infiltration around germinal centre follicles. The cells expressed B cell markers, CD5 and cyclin D1. These cells were also identified on a bone marrow aspirate and trephine



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Tuesday 24 March 2015

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Question 1: Haematological-Oncology

A 57 year old man has a diagnosis of multiple myeloma. He has received three lines of therapy over the last 6 years including a high dose melphalan conditioned autologous transplant. He is being followed up in out patients. Over a period of four weeks he develops a progressive pancytopenia. You are called by the haematology diagnostic lab to say that a senior laboratory technician has reviewed the film and this shows the presence of a small population of blast-like cells. The preliminary report from the immune-phenotype lab is that these cells are CD38 negative, B- and T- lineage marker negative but express CD 13, 33 and myeloperoxidase.

- a) What is your provisional diagnosis?
- b) Outline your approach to the further investigation and what results you might expect to find for each diagnostic test and subsequent management plan for this patient

Question 2: General Haematology

You are asked to review a 32 year old female patient with mucosal bleeding and a platelet count of $4 \times 10^9/l$ whose platelet count has failed to respond to a 20 day course of high dose steroids. The diagnosis is of ITP. Over the next 14 days her count remains in single figures with mild mucosal bleeding.

- a) Discuss the benefits versus risks of the current options for her on-going management and describe what you would explain to the patient.
- b) What counselling would you give at this stage with respect to any implications for her intention to become pregnant in the near future? Discuss issues for both the patient and baby

Question 3: Haemostasis and Thrombosis

An 84 year old patient with atrial fibrillation requires anticoagulation:

- a) Briefly outline the treatment options available and their mechanism of action
- b) Discuss the key benefits for the direct acting oral anticoagulants (DOACs) (also known as new oral anticoagulants [NOAC] or oral direct inhibitors [ODI])
- c) Discuss the potential disadvantages of the direct acting oral anticoagulants (DOAC, NOAC, ODI) in the future management of this patient

Question 4: Transfusion

An 'unknown' adult male is involved in a road accident and is brought into your hospital with massive blood loss. Explain in detail what blood and blood components should be provided in the first hour following hospital admission. If you are asked to coordinate a revision of your hospital's major haemorrhage protocol, what key elements should be included and what is the evidence for your suggestion?



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Haematology: First paper

Tuesday 23 September 2014

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Time allowed: Three hours

1. A 29 year old woman developed a deep vein thrombosis at 28 weeks in her first pregnancy.
 - a) Discuss the role of thrombophilia testing in her management
 - b) Briefly outline the advice you would give for this and future pregnancy if antithrombin deficiency is diagnosed.

2.
 - a) Describe the key clinical features that you would seek on clinical history taking and the appropriate investigation of a 35yr old female patient who has been found to have a serum ferritin of 500 ng/ml at a health screening assessment.
 - b) A 75 yr old man with myelodysplastic syndrome who has failed a trial of erythropoietin is started on a transfusion programme. Discuss the management of iron overload in this case.

3. A 49 year old man, presents with back pain to his GP. Investigations show IgG paraprotein of 68g/l immunoparesis of IgM and A and an excess of serum Lambda free light chains. In addition he had a creatinine of 220 mmol/L, adjusted Ca 2.8 mmol/L, urate 0.45 mmol/L, albumin 23 g/L, beta 2 microglobulin 1.8 mg/L, Hb 98g/L, plt $130 \times 10^9/L$, WCC $4.0 \times 10^9/L$ N $1.2 \times 10^9/L$, liver function was normal. A bone marrow aspirate showed 80% plasma cells, reduced haemopoiesis but normal morphology. Additional investigations included a skeletal survey which shows occasional lytic lesions, but no fractures. His interphase FISH results show t(11;14) with del 13q14. He has no significant previous medical history. He has 3 younger siblings. He was admitted later that day, given a single dose of pamidronate and 3 l of fluid over 24 hours and his creatinine improved to 100mmol/l, Ca to 2.2mmol/l.

His case was discussed at the local MDT 1 day ago. There are currently no trials open in your centre for this diagnosis.

You have seen him today with his wife and discussed his prognostic stage, initial management and his prognosis. You are writing a letter to his GP summarising your discussion, rationale behind the chosen therapeutic pathway as against other options and what his long term prognosis is likely to be.

4. A 50 year old man had a previous allogeneic peripheral blood stem cell transplant for poor risk CLL. He develops abnormal liver function tests and in the course of investigation of these is found to be hepatitis C antibody positive. Because of the blood components he has received in the past, transfusion transmitted infection is considered as a possible source and the Blood Service is contacted.

What steps should be taken to investigate this case?

What measures are in place to minimise the risk of transfusion-transmitted viral infection?



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Tuesday 25 March 2014

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1. a) Describe and justify your recommendations for the investigation of a 50-year-old man who presents to his GP with malaise and itch. Include a differential diagnosis at this stage:

HB 140 g/l,
WBC $11.93 \times 10^9/l$, Neut $2.34 \times 10^9/l$, Eosinophil count $3.55 \times 10^9/l$
Platelets $396 \times 10^9/l$

- b) On review 2 months later after initial investigation his blood count shows

HB 147 g/l,
WBC $36.44 \times 10^9/l$, Neut $4.53 \times 10^9/l$, Lymphocytes $2.75 \times 10^9/l$, Eosinophil count $28.2 \times 10^9/l$, Basophils $0.04 \times 10^9/l$, Monocyte $0.89 \times 10^9/l$
Platelets $299 \times 10^9/l$

Describe how you would proceed and outline the potential causes for this picture.

2. Based on loss of clinical response to factor VIII you suspect that a 4 year old boy with severe haemophilia A has developed an inhibitor.

Discuss how you would assess this in the laboratory and give a detailed plan of your future management of the case.

3. Your hospital is a very high user of platelets and has recently undertaken a clinical audit of platelet usage and wastage. Your audit results show that 25% of platelet usage was not compliant with national guidelines and 5% of platelets were ordered and not used so subsequently wasted. Explain the current recommendations for platelet transfusion and discuss how you would use these audit findings to improve practice in your hospital.
4. A 33 year old single female, who lives alone, but has a supportive family, with high risk AML (FLT-ITD positive at diagnosis) entered morphological CR following induction chemotherapy (DA 3+10). She received a second course of DA 3+8 and a repeat marrow showed continuing remission morphologically but the presence of low level MRD by immune-phenotype / FLT-3 analysis. She is blood group O+ , . CMV sero-negative. A fully matched (HLA 10/10) 23 year old male volunteer unrelated donor (blood group A+, CMV sero-positive) has been identified as her best potential donor. Stem cell transplantation in CR1 has been recommended at your MDT as her best treatment option.

Outline your discussions with this patient at her next clinic visit focussing on a description of the planned treatment, expected outcome, benefits and risks. Write your answer in the form of a letter to her GP, Dr Fox, which is also copied to the patient.