The Royal College of Pathologists
Pathology: the science behind the cure

Part 1 examination
Haematology First paper
Tuesday 24 September 2019
Candidates must answer ALL questions
Time allowed: Three hours

Question 1 - Transfusion

A 54-year-old male with Hb SS with recurrent crisis and history of acute chest syndrome several years and pulmonary hypertension, has been admitted following a mechanical fall. X-ray findings confirm fracture right neck of femur. He needs urgent orthopaedic surgery but is reluctant to agree to have blood transfusion as he is concerned about the safety of blood and his wife is a Jehovah's Witness. He is blood group B RhD negative. His Hb S was 84% when last checked 4 months ago and Hb at admission was 90 g/L.

a) Discuss how you will manage his concerns and what you need to do to optimise him for surgery. Discuss potential complications in this patient including those related to treatment. (20 marks)

b) Describe the specification of blood components, volume required for exchange transfusion and targets for treatment. (5 marks)

Question 2 - Haemato-oncology

A previously fit and healthy 62 year old man presents with fatigue and back pain and is found to have a Hb of 82g/l and Creatinine of 350umol/l. A diagnosis of multiple myeloma is suspected:

a) Discuss the initial investigations and urgent acute management that is required. (10 marks)

b) What are the recommended options for imaging in newly diagnosed patients with myeloma? (4 marks)

c) Discuss the options for therapy for this man and the factors that would guide your decisions. (6 marks)

d) 28 months later, he then presents with rapid onset pain and weakness in his left leg associated with urinary incontinence. Discuss the likely diagnosis and initial plans for management in this situation. (5 marks)

Question 3 - Haemostasis

A 74-year-old man underwent a complicated femoral-popliteal bypass as is started on unfractionated heparin post-operatively.

a) What clinical situations is unfractionated heparin used in preference to low molecular weight heparins. (8 marks)

b) List three methods for monitoring the effect of unfractioned heparin and describe one of these methods in detail discussing the advantages and disadvantages of it (9 marks)

c) The patient needs to go back to theatre in the next few hours as there are worries over the patency of the graft, describe how the effect of unfractionated heparin can be reversed and compare this to the reversal of low molecular weight heparin.
(8 marks)

Question 4 - General Haematology

A 35-year-old man referred urgently with Hb 210g/L. The patient had been attending a gym and had an episode of lightheadedness.

a) What is your advice to the GP and management plan? (10 marks)

4 weeks later he is in Haematology clinic with a negative Jak2 result.

b) What further investigations? Discuss ongoing management (15 marks)

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Part 1 examination
Haematology First paper
Tuesday 26 March 2019
Candidates must answer ALL questions
Time allowed: Three hours

Question 1 – General Haematology

a) Design a guideline for GP's for patients with mild eosinophilia with a referral pathway? (40%)

b) A 36 year old woman with normal Hb Eosinophilia of $10x10^9$g/L, normal platelets is referred urgently to clinic. Outline your approach to investigation and management. (60%)

Question 2 – Haemostasis and Thrombosis

A 74 year old man with hypertension attends hospital for a routine cholecystectomy. He is found to be in atrial fibrillation.

a) Describe how you would assess whether he would benefit from anticoagulation and what treatment options are available giving the advantages/disadvantages of each. (40%)

He has mitral valve disease and the decision is made to start him on warfarin.

b) Describe how the PT is performed and the INR derived. (30%)

c) Detail how you would start the patient on warfarin and how you determine the quality of anticoagulation. (30%)

Question 3 – Haem-oncology

A 60 year old man who was previously fit and well presents to his GP with a three month history of neck swelling accompanied by fatigue, lethargy and loss of 10kg of weight. On examination he had lymphadenopathy in all cervical stations, the largest being 3cm in diameter. A CT scan showed disease in the axillae, mediastinum and retroperitoneum, as well as 18 cm splenomegaly. He was referred to an ENT surgeon for a biopsy. The pathology is presented at your MDT and reveals a diagnosis of mantle cell NHL.

a) Describe the pathological and immunophenotypical features you would expect to see. Discuss your investigation and management plan for this patient. How would you explain this to the patient? (50%)

b) 36 months later at a routine clinic appointment he reports recurrence of his presenting symptoms and has clinical evidence of relapse with widespread lymphadenopathy and hepatosplenomegaly. What are the options for further systemic anticancer treatment now and which is your preferred treatment? Briefly describe the mode of action of any treatment you might use and any side effects that you would look out for. (35%)

c) 12 months later he relapses again with no B symptoms and palpable left axillary nodes only. How would you treat him now? (15%)

Question 4 – Transfusion

You are contacted by the hospital transfusion laboratory at 2 am to say that the transplant team are about to perform a heart transplant on a 44 year old man, but he has a positive direct antiglobulin test (DAT) and the red cell antibody panel is pan-reactive.

a) What are the possible causes of the positive DAT and what laboratory investigations will help you decide on the cause? (50%)

b) What is the differential diagnosis for this patient? (30%)

c) How would you manage the provision of red cell support? (20%)

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Part 1 examination

Haematology: First paper

Tuesday 25 September 2018

*Candidates must answer **all** questions. Each question is worth a total of 25 marks.*

Time allowed: 3 hours

1. Haemato-oncology.

A 22 year old woman, presented with a 2 week history of bruising on her arms and legs and a 3 day history of intermittent epistaxis. Examination revealed large bruises of varying ages on her limbs, and a number of oral mucosal haemorrhages. There were no other abnormal findings.
Full blood count shows:

Haemoglobin 96 g/L (NR: 120-160)

White cell count 135 x 10^9/L (NR: 4-10)

Platelets 20 x 10^9/L (NR 140-400)

Blood film showed a large number of abnormal, immature myeloid cells, many with folded nuclei. The cells contained multiple granules and infrequent Auer rods.

Fluorescence in situ hybridisation demonstrated the presence of a PML-RARA rearrangement

Part 1

What is the diagnosis? What immediate additional tests are required? Describe your initial management of this patient. What complications of treatment would you advise the patient about? **(12 marks)**

Part 2

5 days after the start of treatment, the patient complained of increasing breathlessness, and was noted to have gained 5 kg in weight since presentation. Chest XRay showed patchy bilateral opacification. What is the differential diagnosis? How would you manage each possible cause of these findings? **(8 marks)**

Part 3

The patient achieved a complete remission after her initial therapy. 18 months after finishing her initial treatment, there was molecular evidence of relapse. Blood count showed
Hb 112 g/L, white cell count 1.3 x 10^9/L, platelets 60 x 10^9/L.
How would you manage her now? **(5 marks)**

2. Haemostasis and thrombosis

You are asked to see in clinic a 29 year old woman at the request of the obstetric department. She has a history of 3 consecutive first trimester miscarriages and an anatomical cause has been excluded. Investigations have shown a normal haemoglobin, white count and platelet count and PT and fibrinogen, but the APTT is prolonged 15 seconds beyond the upper limit of normal.

a) Discuss in general the differential diagnosis of an isolated prolonged APTT and the history and laboratory investigations you would perform to clarify the cause.
(15 marks)

b) Describe the clinical and laboratory features and the criteria for diagnosis of anti-phospholipid syndrome. **(10 marks)**

3. General Haematology

A 35yr old woman is referred by her GP with a month history of tiredness and easy bruising. Full blood count is as follows

Hb 60 g/L (NR:	120-160)
White Cell Count	2.0 $X10^9$/L (NR: 4-10)
Neutrophil Count	0.03 x 10^9/L (NR: 1.5-7)
Platelets	12 X 10^9/L (NR: 140-400)
Reticulocytes	9 x 10^9/L (NR: 25-85)

The blood film confirms that the counts are genuine. There is no cellular atypia or blast cells in the film. The count is repeated 2 days later and the findings are almost exactly the same. The bone marrow sample shows gross hypocellularity and mild dyserythropoeisis only

Outline your further investigation.
Based on the current criteria state the diagnosis precisely.
Describe the approach to transfusion in this case.
Discuss the management plan for the disorder with justification for choices
(25 marks)

4. Transfusion

What measures are taken by UK blood services to reduce the risk of an adverse transfusion - related event? Explain the rationale for these measures considering all the steps from recruitment of donors through to the dispatch of blood from the transfusion laboratory
(25 marks)

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Part 1 examination

Haematology: First paper

Tuesday 20 March 2018

*Candidates must answer **all** questions. Each question is worth a total of 25 marks.*

Time allowed: 3 hours

Question 1: General Haematology

You are asked by a surgical colleague for your opinion on a 23yr old lady who has been admitted with abdominal pain, but was noted to have Haemoglobin 90g/L, elevated LDH of 600U/L and low haptoglobin.

a) Write a differential diagnosis as a list. Outline your investigation plan with justification for each test that you suggest. (12.5 marks)

b) A diagnosis of Paroxysmal Nocturnal Haemoglobinuria (PNH) is established - outline your management plan of this lady. (12.5 marks)

Question 2: Transfusion Medicine

A 33 year old woman is admitted with symptoms of abdominal pain and confusion.

Full blood count shows Haemoglobin 90g/L, white blood cell count 11 x 10^9/L and platelet count 6 x 10^9/L. Red cell fragments are noted on blood film. Lactate Dehydrogenase (LDH) is > 1200U/L. Blood group is A RhD negative.

A provisional diagnosis of Thrombotic Thrombocytopenic Purpura (TTP) is made and the patient is referred for plasma exchange following placement of central line.

a) Discuss the management of blood product support for this patient. (12.5 marks)
b) Prepare an information leaflet for patients requiring plasma exchange (12.5 marks)

…/questions continue over

Question 3: Haematological Oncology

A 60 year old man presents with drowsiness, nosebleeds, headaches and blurred vision. On examination he has retinal haemorrhages, 4 fingerbreadths of splenomegaly and extensive 1-2 cm lymphadenopathy. Blood count shows Haemoglobin 78 g/L, platelets 76 x10^9/L, and white blood cell count 1.5 x 10^9/L. His serum total protein is elevated, and total IgM is raised at 28 g/L with a paraprotein demonstrated on serum electrophoresis. A bone marrow trephine biopsy reveals dense infiltration with lymphoplasmacytoid lymphocytes.

a) What is the diagnosis? What further investigations are required to identify the cause of his presenting symptoms and clarify the underlying problem? Indicate the appropriate initial management of the patient (7 marks)

b) With appropriate initial management, his symptoms improve significantly. What options for systemic anti-cancer treatment would you offer him to treat the underlying condition? Describe what aims of treatment and potential adverse effects you would discuss with the patient in order to gain written informed consent for the proposed treatment (9 marks)

c) He responds well to first line chemotherapy and achieves a remission. 12 months later he represents with renewed nosebleeds and a significant rise in the IgM paraprotein. What is the appropriate management at this stage? Are there any molecular genetic tests that might guide your choice of therapy? (9 marks)

Question 4: Haemostasis and Thrombosis

You are asked for advice on a 70 year old woman with a platelet count of 45 x 10^9/L who had undergone coronary artery bypass graft surgery 6 days previously. Full blood count and coagulation screen (prothrombin time and activated partial thromboplastin time) are otherwise normal. The blood film confirms the thrombocytopenia is genuine with no other abnormalities seen and review indicates that the preoperative platelet count was normal.

a) Discuss the differential diagnosis and any further history you will obtain to help establish the diagnosis (7 marks)

b) Outline the pathophysiological basis of heparin induced thrombocytopenia (HIT). Discuss the laboratory tests used to investigate suspected cases of HIT and how the findings of these may be interpreted in its diagnosis (12 marks)

c) Outline the anticoagulation options available for the treatment of HIT including the duration of treatment and describe any factors that may influence your choice of treatment. (6 marks)

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Haematology

First Paper

Tuesday 26th September 2017

Candidates must answer ALL questions

Time allowed: 3 hours

Question 1: Haemostasis and thrombosis

A 28 year old woman presents for the first time to antenatal clinic at approximately 34 weeks gestation. Her haemoglobin, white cell count, and differential are normal but platelet count is 58 x 10^9/l. The blood film confirms genuine thrombocytopenia. You are asked to see her by the obstetrics team.

a) Detail the history, examination and investigation you would perform to establish the differential diagnosis.
b) Outline the differential diagnosis in an otherwise completely well patient
c) Outline the management of the mother and baby for a diagnosis in this case of ITP

Question 2: Haemato-oncology

A 64 year old man describes 6 months of fatigue, loss of appetite, sweating and weight loss. On examination he has 3 finger breadths of splenomegaly and palpable liver edge.

His FBC is: Haemoglobin 110 g/L, white cell count 20 X 10^9/L (neutrophils 15.3 basophils 0.3), platelets 110 x 10^9/L. Blood film examination reveals a leucoerythroblastic picture with numerous tear drop poikilocytes. Blasts account for <1% of circulating white blood cells and there are giant platelets.

a) What is the most likely diagnosis and the differential diagnosis? Outline the investigations you would perform and the results you would expect to confirm the diagnosis.
b) Outline the natural history of this condition, and how you would predict the clinical course in this patient.
c) Discuss the management options for this patient.
d) Summarise in short note form the information you would wish to convey to this patient during your consultation.

Question 3: Transfusion medicine

A blood sample sent to the hospital blood bank for crossmatching is found to have a positive direct antiglobulin test (DAT).

What are the possible causes of the positive DAT, what further features in the history and laboratory investigations would be appropriate and how would you manage the provision of red cell support?

Question 4: General Haematology

A GP seeks advice on a 23 year old patient with (haemoglobin) Hb SS, estimated 10 weeks pregnant. She is para 0 + 0.

She wants to know if she is going to have an affected child & what antenatal care she should have.

a) Outline the detail of the investigations that you would recommend the GP to send to the laboratory prior to the patient being seen in 1 weeks time in clinic

b) List the relevant tests for her partner and the rationale for these

c) Draw up an outline of how you would approach care in pregnancy with reference to potential complications

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Part 1 examination

Haematology: First paper

Tuesday 21 March 2017

Candidates must answer ALL QUESTIONS

Time allowed: Three hours

Question 1: Haemostasis and thrombosis

You are asked to see a 35 year old female in clinic who requires tonsillectomy. She has been referred from pre-op assessment clinic having reported “bleeding” following a previous dental extraction.

Discuss features of the clinical history and examination that would be relevant. Which investigations you would subsequently request – please explain why you would perform these investigations and any caveats in the interpretation of results.

Question 2: Transfusion

Describe the strategies used to keep the blood supply free from potential viral infections, using 2 example viruses (include one virus recognised as an issue in the past 3 years) to illustrate your answer. What practices, other than strategies to minimise the risk of viral contamination of a product, maximise clinical safe and effective blood transfusion for patients?

Question 3: Haemato-oncology

A 26 year old lady presents to her General Practitioner with a 4 month history of worsening wheeze, associated with lumbar back pain, drenching night sweats and 6 kg weight loss. Her full blood count is as follows: Haemoglobin 102 g/L (MCV 69) platelets 470, white cell count 16 (neutrophils 11.9, lymphocytes 0.3, eosinophils 2.0). She has a 3cm left axillary lymph node palpable and 2 fb splenomegaly palpable below the costal margin. A CXR reveals a wide mediastinum and pulmonary infiltrates. Biopsy of the axillary node reveals a nodular appearance with extensive bands of fibrosis. There are scattered large cells that are CD 15 and 30+ve and are –ve for CD 20 and 45. There is positive staining for LMP-1. There is a background scattered positivity for both B and T cell markers.

Discuss the diagnosis based on these results.

Outline your management plan, including further investigations and choice of treatment.

Describe what you would tell the patient and her family at the first consultation.

Question 4: General Haematology

a) A 50 year old man is referred with a ferritin of 800mcg/L. Outline your evaluation of this patient to include a clear relevant clinical history and description of investigations to clarify your diagnosis.

b) Write an outline for a nurse led haemochromatosis clinic of patients with an established diagnosis including procedure, monitoring parameters, referral criteria for medical review & key performance indicators.