

Haematomorph Haematology FRCPATH Preparation

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Week:

General Week -Transfusion Week- Haemostasis Week- Oncology Week

Essay 01: A 62-year-old man presents with a 3-week history of worsening fatigue and shortness of breath. He also reports episodic discolouration of his fingers when exposed to cold.

Past medical history includes well-controlled hypertension. No recent travel or new medications.

On examination:

- Pale with mild scleral icterus
- Peripheral cyanosis in fingers
- No significant lymphadenopathy or hepatosplenomegaly

Investigations:

Test	Result
Hb	72 g/L
MCV	105 fL
Reticulocyte count	Elevated
LDH	Elevated
DAT	Positive for C3 only, negative for IgG

A. What is the most likely diagnosis? Explain the underlying pathophysiology. (2 marks)

B. List the common causes of this condition and key complications. (5 marks)

C. Outline your diagnostic approach, including additional investigations required. (6 marks)

D. Describe your management plan, including supportive care and disease-directed therapy. (12 marks)

Essay 02: A 25-year-old woman with known homozygous sickle cell disease (HbSS) presents with sudden onset right-sided weakness and difficulty speaking for 2 hours. She reports a preceding history of worsening headache over the past 48 hours and a recent painful vaso-occlusive crisis.

She is on folic acid and intermittent analgesia but is not on regular disease-modifying therapy.

On examination:

- Right-sided hemiparesis (power 3/5)
- Expressive aphasia
- Temperature: 37.8°C

Test	Result
Hb	78 g/L
Reticulocytes	Elevated
LDH	Elevated
CRP	Mildly elevated
CT brain	No haemorrhage
MRI brain	Acute left MCA infarct

A. What is the most likely diagnosis, and what is the underlying pathophysiology in sickle cell disease? (4 marks)

B. Outline the urgent management of this patient. (10 marks)

C. Describe strategies for secondary stroke prevention in sickle cell disease. (4 marks)

D. The patient asks about future pregnancy. Outline key considerations and risks in women with sickle cell disease planning pregnancy. (7 marks)

Essay 03: A 29-year-old woman presents with a 5-day history of severe abdominal pain, nausea, and increasing abdominal distension. She has no past medical history and is not on any medications, including oral contraceptives.

On examination:

- Mild jaundice
- Tender hepatomegaly
- Ascites present
- HR: 110 bpm, BP: 110/70 mmHg

Investigations:

Hb 135 g/L

WBC $11.5 \times 10^9/L$

Platelets $1350 \times 10^9/L$

ALT/AST Mildly elevated

Bilirubin Elevated

INR Mildly prolonged

Doppler ultrasound abdomen: Hepatic vein thrombosis

A. What is the most likely underlying diagnosis, and what are the key differential diagnoses? (4 marks)

B. Describe the diagnostic work-up required to confirm the underlying haematological disorder. (6 marks)

C. Outline the acute and long-term management of this patient. (10 marks)

D. Outline management considerations if she plans future pregnancy. (5 marks)

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Essay 01:

A 29-year-old woman is referred to haematology with progressive fatigue, recurrent oral ulcers, easy bruising, and heavy menstrual bleeding for 4 months. She also reports two recent episodes of febrile neutropenia treated with antibiotics. There is no significant past medical history.

Examination:

- Pale, with scattered petechiae over lower limbs

Initial investigations:

Test	Result	Reference Range
Hb	64 g/L	115–155
WBC	$1.2 \times 10^9/L$	4.0–11.0
Neutrophils	$0.1 \times 10^9/L$	2.0–7.5
Platelets	$8 \times 10^9/L$	150–400
Reticulocytes	0.2%	0.5–2.5
MCV	95 fL	80–96

Bone marrow aspirate:

- Markedly hypocellular marrow (<10% cellularity)

Questions

1. What are the different inherited bone marrow failure syndromes. List diagnostic investigation of each of them (05marks)
2. Discuss the first-line treatment for severe aplastic anaemia in a young patient (05 marks)

3. This patient has refractory disease after first-line immunosuppressive therapy. Discuss the management for refractory severe aplastic anaemia. (10 marks)
4. During long-term follow-up, what clonal complications should be monitored for in this patient, and how should surveillance be performed? (5 marks)

Essay 02:

A 41-year-old man is referred to haematology with progressive fatigue, abdominal pain, and two recent episodes of unprovoked venous thrombosis (hepatic vein thrombosis 18 months ago and a recent portal vein thrombosis). He also reports intermittent dysphagia.

There is no significant past medical history. He is not taking regular medications. No family history of thrombosis is reported.

Examination: Mild scleral icterus

Investigations:

Test	Result	Reference Range
Hb	78 g/L	130–170
WBC	$2.8 \times 10^9/L$	4.0–11.0
Neutrophils	$1.1 \times 10^9/L$	2.0–7.5
Platelets	$92 \times 10^9/L$	150–400
DAT	Negative	

Urinalysis:

- Haemosiderinuria present

Flow cytometry:

- Granulocyte clone: 88%
- Monocyte clone: 85%
- RBC clone: 45%

Bone marrow biopsy:

- Mild hypocellularity with reduced trilineage haematopoiesis

Questions

a. Discuss the pathophysiology of thrombosis in Paroxysmal Nocturnal Haemoglobinuria (5 marks)

- b. What are the indications for complement inhibitor therapy in PNH, and discuss appropriate first-line choice in this patient? (6 marks)
- c. Despite good control of intravascular haemolysis, the patient remains anaemic and transfusion dependent. Discuss the management options for persistent anaemia in PNH. (08 marks)
- d. Compare proximal complement inhibition (C3 inhibition) with terminal complement inhibition (C5 inhibition) in PNH, including advantages, limitations, and current clinical use. (06 marks)

Essay 03:

A 58-year-old man is referred to haematology with progressive fatigue, drenching night sweats, 8-kg unintentional weight loss over 6 months, worsening abdominal discomfort, and marked early satiety.

Past history:

- Essential thrombocythaemia diagnosed 12 years ago
- Initially treated with hydroxycarbamide and aspirin

Examination:

- Pale, cachectic appearance
- Massive splenomegaly: spleen palpable 15 cm below left costal margin

Investigations:

Test	Result	Reference Range
Hb	78 g/L	130–170
WBC	$28 \times 10^9/\text{L}$	4–11
Platelets	$110 \times 10^9/\text{L}$	150–400
Peripheral blood film Leucoerythroblastic picture		
LDH	1100 U/L	<250

Molecular studies:

- JAK2 V617F positive
- ASXL1 mutation detected
- SRSF2 mutation detected

Bone marrow trephine:

- Megakaryocytic atypia with dense clustering
- Osteosclerosis present

Questions

- a. Discuss the WHO 2022 diagnostic criteria for secondary Myelofibrosis. (5 marks)
- b. Discuss the management of the above patient. (10 marks)
- c. Discuss the role of allogeneic stem cell transplantation in this patient. (5 marks)
- d. The patient develops 12% circulating blasts 8 months later. Discuss the significance of accelerated phase myelofibrosis, its prognosis, and treatment options including bridge-to-transplant strategies. (5 marks)

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General Week -Transfusion Week- Haemostasis Week- Oncology Week

Essay 01:

A 46-year-old man is referred urgently to the haematology clinic because of persistently raised ferritin detected during investigation of abnormal liver function tests. He reports progressive fatigue, reduced libido, intermittent pain in the second and third metacarpophalangeal joints, and worsening glycaemic control over the last 2 years.

Past medical history: Type 2 diabetes mellitus, Hypertension and Non-alcoholic fatty liver disease diagnosed 5 years ago

Social history: Drinks 18–20 units alcohol/week and BMI 33 kg/m²

Family history: Elder brother died from hepatocellular carcinoma aged 52 and Mother had “liver problems”

Initial investigations: Hb 172 g/L, Platelets 180×10^9 /L, Ferritin 2,850 µg/L, Transferrin saturation 88%, ALT 96 U/L and AST 82 U/L

Questions

1. What are the possible causes of markedly elevated ferritin in this patient? (04 marks) and What additional investigations would you perform to assess severity and organ involvement? (06 marks)
2. Discuss the management of this patient. (10 marks)
3. What are the complications and late manifestations of untreated hereditary haemochromatosis? (05 marks)
4. What advice should be given regarding family screening and genetic counselling? (04 marks)

Essay 02:

A 29-year-old woman is admitted under the acute medical team with mucosal bleeding, severe headache, and new onset confusion. She is 26 weeks pregnant with her second child.

She reports Recurrent epistaxis for 2 weeks, Gingival bleeding while brushing teeth and Extensive bruising over limbs and abdomen

History: Immune thrombocytopenia (ITP) diagnosed 1 years ago and Previous splenectomy 2 years earlier due to road accident.

Current medications: Low-dose prednisolone

No hypertension, no proteinuria, and fetal movements are normal.

Examination: Drowsy but orientated and Extensive ecchymoses

Test	Result
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Hb	122 g/L
----	---------

WBC	$7.4 \times 10^9/\text{L}$
-----	----------------------------

Platelets	$1 \times 10^9/\text{L}$
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Blood film Severe thrombocytopenia, no schistocytes

CT head: Small intracranial haemorrhage without midline shift

Questions

1. What is the differential diagnosis of severe thrombocytopenia in pregnancy in this patient? (04 marks)
2. Discuss the acute management of this patient. (06 marks)
3. How would you manage her thrombocytopenia for the remainder of pregnancy and delivery planning? (10 marks)
4. What are the neonatal implications and how should the baby be monitored? (05 marks)

Essay 03:

A 24-year-old woman with is admitted to the High Dependency Unit with worsening chest pain, breathlessness, and confusion.

She reports: Severe back and limb pain for 3 days, Fever and productive cough for 24 hours. Progressive shortness of breath and Increasing drowsiness this morning

Past history:

- Recurrent vaso-occlusive crises requiring ≥ 6 admissions/year
- Silent cerebral infarct on prior MRI brain
- Chronic transfusion programme discontinued 1 year ago because of poor compliance

- Iron overload secondary to transfusions

Current medications: Hydroxycarbamide (poor adherence), Folic acid and Penicillin V

Examination: Appears toxic and distressed, Temperature 39.2°C and O₂ saturation 86% on air

Test	Result
Hb	58 g/L (baseline 88 g/L)
WBC	24 ×10 ⁹ /L
Platelets	110 ×10 ⁹ /L
LDH	Markedly elevated
CRP	220 /L

Chest imaging: Bilateral multilobar infiltrates

Hb electrophoresis: HbS 92%

Questions

1. What are the differential diagnoses for this acute deterioration? (04 marks)
2. Discuss the immediate management of this patient in detail. (06 marks)
3. What are the indications for exchange transfusion in sickle cell disease? (04 marks)
4. Discuss the role of allogeneic stem cell transplantation and gene therapy in sickle cell disease. (06 marks)
5. What pre-transplant assessments are required before curative therapy in this patient? (05 marks)

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Essay 01:

A 29-year-old woman presents with a 3-month history of progressive fatigue, exertional dyspnoea, episodic chest discomfort, generalized pruritus, and an unintentional 5-kg weight loss. Over the preceding 2 weeks, she has developed painful swelling of the left calf followed by increasing breathlessness.

She has no history of asthma, eczema, allergic rhinitis, autoimmune disease, recent travel, parasitic exposure, new medications, or illicit drug use.

On examination: Oxygen saturation: 91% on room air, Raised JVP, Mild pedal oedema and left calf swelling and tenderness

Test	Result
Hb	103 g/L
WBC	$29 \times 10^9/L$
Neutrophils	$9 \times 10^9/L$
Eosinophils	$17 \times 10^9/L$
Platelets	$185 \times 10^9/L$

Peripheral blood film: Marked mature eosinophilia. No dysplasia and No circulating blasts

Bone marrow: Hypercellular marrow with marked eosinophilic hyperplasia and Blasts 2%

Cardiac investigations: Echocardiography: Left ventricular apical thrombus with early endomyocardial fibrosis and mildly impaired left ventricular systolic function.

Imaging: CT pulmonary angiography: Bilateral segmental pulmonary emboli and Doppler ultrasound: Extensive left femoral deep vein thrombosis.

Molecular tests and Reactive causes of eosinophilia are negative.

Questions

- A. Describe the diagnostic criteria for idiopathic hypereosinophilic syndrome and outline the investigations required to exclude clonal causes of eosinophilia. (6 marks)
- B. Discuss the management of this patient, including treatment of acute life-threatening organ involvement and long-term therapy for idiopathic HES. Include management options for steroid-refractory disease. (10 marks)
- C. Two years later, she remains in remission on maintenance therapy and attends the antenatal clinic at 12 weeks' gestation with her first pregnancy. Discuss the antenatal management of this patient. (9 marks)

Essay 02:

A 30-year-old woman presents with a 4-week history of persistent high-grade fevers, drenching night sweats, profound fatigue, and an unintentional 8-kg weight loss. Over the preceding week, she has developed increasing abdominal distension and easy bruising.

She has no significant past medical history and is not taking any regular medications. There is no recent travel or history suggestive of autoimmune disease.

On examination: Temperature: 40°C, Hepatomegaly (3 cm) and Splenomegaly (8 cm below the costal margin)

Test	Result
Hb	66 g/L
WBC	$1.8 \times 10^9/L$
Ferritin	16000ug/L
Platelets	$28 \times 10^9/L$
Triglycerides	5.8 mmol/L
Fibrinogen	0.6 g/L
AST	185 U/L

Bone marrow biopsy has been arranged

Questions

- A. Describe the diagnostic criteria and discuss its underlying causes. (7 marks)
- B. Discuss the pathophysiology and investigations required to identify the underlying cause in this patient. (08 marks)

C. Outline the management of this patient, including initial treatment, and options for refractory or relapsed disease. (10 marks)

Essay 03:

A 35-year-old woman presents to the emergency department with a 5-day history of severe left flank pain, nausea, and visible haematuria. She also reports intermittent headaches, aquagenic pruritus after showering, and facial redness for the past year. She denies any recent surgery, prolonged immobilisation, oral contraceptive use, or family history of thrombosis.

On examination: Blood pressure: 152/94 mmHg, Facial plethora, Mild splenomegaly (3 cm below the left costal margin) and Left loin tenderness

Test	Result
Hb	182 g/L
Hct	0.58
WBC	$13.5 \times 10^9/\text{L}$
Platelets	$610 \times 10^9/\text{L}$
Creatinine	108 $\mu\text{mol/L}$

CT renal angiography: Acute left renal vein thrombosis extending into the inferior vena cava.

Bone marrow biopsy: Panmyelosis with pleomorphic mature megakaryocytes

Questions

A. Discuss the differential diagnosis of erythrocytosis and describe the investigations required to establish the diagnosis. (07 marks)

B. Outline the management of this patient, including treatment refractory cases. (12 marks)

C. Six years later, during clinic follow up, she complained about fatigue, early satiety and abdominal fullness. FBC shows Hb 85 mg/L WBC $28 \times 10^9/\text{L}$ Platelets $80 \times 10^9/\text{L}$. Discuss the diagnostic approach (08 marks)

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Week:

General Week -Transfusion Week- Haemostasis Week- Oncology Week

Essay 01:

A 28-year-old woman originally from Pakistan presents to the adult haemoglobinopathy clinic for review. She was diagnosed with β -thalassaemia intermedia at the age of 5 years after investigation of chronic anaemia.

She has never been regularly transfused but has required intermittent red cell transfusions during episodes of infection, pregnancy and severe anaemic symptoms.

She has recently developed worsening fatigue, reduced exercise tolerance and abdominal discomfort.

Her previous follow-up has been inconsistent due to multiple relocations.

Examination: Pale, Mild frontal bossing, Hepatomegaly: 5 cm below costal margin and Splenomegaly: 8 cm below costal margin.

Bloods: Hb 74g/l, MCV 58fl, Bilirubin 42umol and Ferritin 2850ug/l. Transferrin saturation 65%, Echo normal and ALT 72 U/L.

Questions:

- A. Outline a comprehensive management plan for this patient with β -thalassaemia intermedia. 12 marks
- B. Discuss the management of iron overload in this patient. 07marks
- C. The patient asks about curative treatment with haematopoietic stem cell transplantation (HSCT). Discuss transplantation options, donor choices and limitations. 06marks

Essay 02:

A 22-year-old man of African ancestry presents to the Emergency Department with a 24-hour history of dark-coloured urine, jaundice, profound fatigue and worsening shortness of breath.

Three days earlier, he was prescribed antibiotics by his GP for a skin infection. He is unable to recall the name of the medication. He has no previous diagnosis of anaemia or haemolytic disorder.

There is no history of recent travel, blood transfusion, or previous similar episodes.

Examination

- Temperature: 38.2°C
- Scleral icterus present
- Marked pallor
- Mild tachycardia
- Tenderness and swelling of the left calf

Investigation	Result
Haemoglobin	6.2 g/dL
MCV	104 fL
Reticulocyte count	$210 \times 10^9/L$
WCC	$14.5 \times 10^9/L$
Bilirubin (unconjugated)	78 $\mu\text{mol/L}$
LDH	1800 U/L
Haptoglobin	Undetectable

Blood film: Marked polychromasia, Bite cells, Blister cells and Occasional fragmented red cells

Doppler ultrasound: Proximal femoral vein thrombosis confirmed

Questions

- Discuss the differential diagnosis of acute haemolytic anaemia with a negative DAT in this patient. 05 marks
- Describe your approach to laboratory investigation of hemolysis in this patient. 07marks
- Outline your immediate management plan for this patient. 08marks
- What counselling should be provided regarding his diagnosis and future management? 05marks

Essay 03:

A 34-year-old woman at 32 weeks' gestation (G3P2) is referred to the obstetric haematology clinic because of persistent anaemia detected on routine antenatal screening.

She has been taking oral ferrous fumarate for the last 10 weeks but reports minimal improvement in symptoms. She describes significant gastrointestinal side effects including constipation and abdominal discomfort, resulting in intermittent compliance.

She reports: Increasing fatigue, Reduced exercise tolerance, Headaches and Palpitations on exertion

Her previous pregnancies were complicated by Postpartum haemorrhage requiring blood transfusion after her second delivery

She follows a vegetarian diet. There is no history of inflammatory bowel disease, coeliac disease, or previous gastrointestinal surgery.

Examination: Pale conjunctivae, Mild tachycardia, No signs of heart failure and Uterus appropriate for gestational age

Investigations: Hb 72g/l, Platelet 650, MCV 64fl, MCH 22pg, Ferritin 4ug/l and Transferrin saturation 4%.

Questions

- A. Discuss your management plan for this patient with iron deficiency anaemia in late pregnancy. 12 marks
- B. You are asked to counsel this patient regarding intravenous iron therapy. What information would you include in your discussion? 07 marks
- C. Outline the key information that should be included in your communication letter to the GP. 06marks

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FRCPATH Haem Part 1 Mock Exam (5th September 2026) Registration Link:

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Week:

General Week -Transfusion Week- **Haemostasis Week**- Oncology Week

Essay 01:

A 24-year-old male with severe congenital haemophilia A (baseline FVIII <1 IU/dL) presents with progressive right thigh swelling and pain following intramuscular injection given at a local clinic 5 days ago.

Current Issues

- Increasing pain and swelling despite self-administered recombinant FVIII at home
- Paraesthesia over anterior thigh

Examination

- Large tense swelling in anterior thigh
- Reduced sensation in femoral nerve distribution

Investigations

Test	Result	Reference
Hb	92 g/L	130–170
Platelets	$310 \times 10^9/L$	150–400
PT	12.5 s	11–14
APTT	110 s	25–35
FVIII activity	<1 IU/dL	50–150
FVIII inhibitor (Bethesda)	32 BU	<0.6

Ultrasound: Large intramuscular haematoma

MRI: Expanding haematoma with femoral nerve compression

Questions

1. Discuss the pathophysiology underlying FVIII inhibitor development in this patient. (05 marks)

2. Outline the immediate and definitive management of this patient's bleeding episode. (10 marks)

3. The patient is later commenced on Emicizumab prophylaxis.

Discuss:

- a. Mechanism of action and clinical benefits (5 marks)
- b. Side effects and laboratory investigation challenges (5 marks)

Essay 02:

A 68-year-old man with ischaemic heart disease, chronic kidney disease (eGFR 28 mL/min) and atrial fibrillation (previously on Apixaban) undergoes urgent CABG + mechanical aortic valve replacement.

Perioperative course

- Received high-dose unfractionated heparin intraoperatively
- Postoperatively started on IV unfractionated heparin infusion

Day 6 post-op

He develops:

- Painful, mottled left foot
- Reduced peripheral pulses
- Confusion and mild dyspnoea

Investigations

Test	Result	Reference
Hb	105 g/L	130–170
Platelets	62 ×10 ⁹ /L	150–400
(baseline pre-op)	210 ×10 ⁹ /L	—
PT/INR	2.4	—
APTT	68 s	25–35
Fibrinogen	3.8 g/L	1.5–4.5
D-dimer	Markedly elevated	—
Creatinine	210	mmol/L —

CT angiography: Acute arterial thrombosis (left tibial artery) and Pulmonary embolism

Questions

1. Discuss the differential diagnosis of thrombocytopenia and thrombosis in this patient and briefly discuss investigational approach in this case. (07 marks)

2. Outline the immediate and ongoing management of this patient. (10 marks)
 3. The patient requires redo cardiac surgery (valve-related complication) in 10 weeks.
- Discuss perioperative anticoagulation strategy in a patient with recent HIT. (08 marks)

Essay 03:

A 79-year-old woman presents with recurrent epistaxis, gingival bleeding, hematuria and prolonged bleeding after venepuncture. She is also complaining of new-onset headaches.

She has a background of hypertension, type 2 diabetes but no prior bleeding history

Examination: Multiple ecchymoses over upper limbs

Investigations

Test	Result	Reference
Hb	98 g/L	130–170
WBC	$5.8 \times 10^9/L$	4–11
Platelets	$240 \times 10^9/L$	150–400
PT	12.6 s	11–14
APTT	62 s	25–35
VWF:Ag	0.25 IU/mL	0.50–1.50
VWF activity	0.08 IU/mL	0.50–1.50
FVIII:C	0.30 IU/mL	0.50–1.50

Additional Tests

- Serum protein electrophoresis: IgM λ paraprotein 12 g/L
- Serum viscosity: mildly elevated
- Bone marrow: Lymphoplasmacytic infiltration

Questions

1. What are the features of acquired bleeding disorders 05marks
2. Discuss the mechanisms leading to bleeding in this patient. List other major causes associated with this bleeding disorder. 08marks
3. Outline the acute and long-term management of this patient. (12 marks)

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Week:

General Week -Transfusion Week- **Haemostasis Week**- Oncology Week

Essay 01:

A 34-year-old male software engineer presents to the Emergency Department with worsening central abdominal pain for 7 days, associated with nausea, early satiety, and intermittent loose stools. The pain is described as “out of proportion” to examination findings and worsens after meals. He reports an unintentional 5-kg weight loss over 2 months.

Past history: None

Examination: Mild abdominal distension with diffuse tenderness

Initial investigations: Hb 15.2 g/dL, WBC $14.8 \times 10^9/L$ and Platelets $910 \times 10^9/L$

CT Abdomen with portal venous phase:

- Extensive superior mesenteric vein thrombosis extending into jejunal tributaries
- Partial portal vein thrombosis

Questions

- a) Discuss the likely aetiology of mesenteric vein thrombosis in this patient and outline the additional investigations required. (07 marks)
- b) Describe your acute management plan for this patient (10 marks)
- c) Six weeks after admission, the patient presented again with worsening abdominal pain, rising lactate, guarding, and repeat CT demonstrates jejunal ischaemia with suspected transmural infarction. Discuss perioperative anticoagulation considerations (08 marks)

Essay 02:

A 27-year-old primigravida at 35+4 weeks gestation is referred urgently to the obstetric haematology clinic because of worsening epistaxis, easy bruising, and concern regarding delivery planning.

She was diagnosed in childhood with “possible von Willebrand disease” after recurrent mucosal bleeding and menorrhagia, but has been poorly followed up and does not know her subtype. Her mother had severe postpartum haemorrhage requiring ICU admission after childbirth.

Current pregnancy history: Two episodes of prolonged epistaxis during the third trimester

Examination: Mild ecchymoses over limbs

Laboratory investigations at 35 weeks: Hb 101 g/L, Platelets $90 \times 10^9/L$, PT normal and aPTT mildly prolonged

Haemostasis studies:

- vWF Antigen: 18 IU/dL
- vWF Activity (GP1bM): 12 IU/dL
- Factor VIII: 22 IU/dL
- Ristocetin-induced platelet aggregation (RIPA): increased sensitivity at low ristocetin concentration

Questions

a) Discuss the physiological changes in von Willebrand factor and factor VIII during normal pregnancy. Interpret the haemostatic findings in this patient and discuss the maternal and fetal risks associated with Type 2B von Willebrand disease during pregnancy and delivery. (08 marks)

b) Outline your antenatal, intrapartum, and postpartum haemostatic management plan for this patient (12 marks)

c) Three months after discharge, the patient was admitted for skin biopsy. How would you plan peri-operative haemostatic management for skin biopsy in this patient? 07marks

Essay 03:

An 18-year-old woman with known Type 3 von Willebrand disease is brought to the Emergency Department with severe right-sided epistaxis, worsening right knee and left elbow swelling, and frank haematuria following a minor fall downstairs 24 hours earlier.

She reports increasing joint pain and inability to bear weight. Her mother states that bleeding has continued despite repeated nasal packing at a local hospital. She is not on any prophylaxis.

On examination:

- Persistent active epistaxis with posterior pharyngeal blood trickling
- Large tense left elbow swelling with paraesthesia over the ulnar distribution
- Right knee markedly swollen and warm

Laboratory investigations:

- Hb 58 g/L, WBC 9.4×10^9 /L and Platelets 340×10^9 /L

Specialised haemostasis testing:

- vWF antigen: undetectable
- vWF activity: undetectable
- FVIII: <1 IU/dL

CT angiography of elbow:

- Comminuted distal humeral fracture
- Large haemarthrosis with active intra-articular bleeding
- Concern regarding impending compartment syndrome

Questions

a) Outline your emergency management of this patient during the first 24 hours (10 marks)

b) How would you manage perioperative haemostasis for emergency orthopaedic surgery in this patient? (07marks)

d) The patient develops suspected anti-vWF alloantibodies. Discuss:

- the mechanism and clinical significance of inhibitor formation in Type 3 von Willebrand disease,
- and therapeutic strategies for severe bleeding in the presence of inhibitors. (08 marks)

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Week:

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Essay 01:

A 32-year-old woman is admitted with a 5-day history of headache, fatigue and easy bruising.

On examination: Mild confusion and no focal neurological deficit

Test	Result
Hb	7.8 g/dL
Platelets	$12 \times 10^9/L$
WCC	$8.2 \times 10^9/L$
Reticulocytes	$180 \times 10^9/L$
LDH	2,900 IU/L
Bilirubin	48 $\mu\text{mol/L}$
Creatinine	195 $\mu\text{mol/L}$
PT	Normal
APTT	Normal
Blood film	Numerous schistocytes

1. What is the likely diagnosis and discuss the immediate management
10marks

2. Initially platelet count rises to $85 \times 10^9/\text{L}$ by Day 5. However, by Day 8, Platelet count falls again to $18 \times 10^9/\text{L}$ and LDH rise from 450 IU/L to 1,600 IU/L. What is the most likely explanation for the deterioration? 05marks
3. Outline the management of such refractory cases and how would you monitor the condition upon remission. 10marks

Essay 02:

An 8-year-old boy with known severe Haemophilia A presents to the Emergency Department with increasing pain and swelling of his left thigh.

He was diagnosed with severe Haemophilia A in infancy (FVIII <1 IU/dL) and has been receiving regular recombinant factor VIII prophylaxis since the age of 1 year.

His parents report that over the past 3 months he has experienced several bleeding episodes despite apparently adequate factor VIII treatment.

On examination:

- Large tender swelling of the left thigh
- Reduced knee movement

Test	Result
Hb	9.8 g/dL
Platelets	Normal
PT	Normal
APTT	Markedly prolonged
Factor VIII level (post-infusion)	<2 IU/dL
Bethesda inhibitor assay	18 BU
Ultrasound	Large intramuscular haematoma

1. What is the most likely explanation for this presentation and what investigations support the diagnosis? (4 marks)
2. How would you manage the acute bleeding episode? (8 marks)
3. Discuss the long-term management of this child. (7 marks)
4. Outline the mechanism of action, significant interactions and investigations affected by Efficizumab.(6 marks)

Essay 03:

A 35-year-old woman known to be a carrier of severe haemophilia A is referred to the joint obstetric-haematology clinic at 10 weeks' gestation during her second pregnancy.

Her father has severe haemophilia A. Her first pregnancy was complicated by a postpartum haemorrhage requiring transfusion. She has a history of menorrhagia, easy bruising, and prolonged bleeding following wisdom tooth extraction.

Current pregnancy has been uncomplicated.

Investigations at booking:

Test	Result
------	--------

Hb	112 g/L
----	---------

Platelets	$260 \times 10^9/L$
-----------	---------------------

PT	Normal
----	--------

APTT	42 sec (prolonged)
------	--------------------

FVIII	0.22 IU/mL
-------	------------

Repeat investigations at 14 weeks:

Test	Result
------	--------

FVIII	0.38 IU/mL
-------	------------

Ultrasound confirms a male fetus.

The obstetric team seeks advice regarding delivery planning.

Questions

1. List the causes of Haemophilia in female population 04marks
2. What factors influence bleeding risk in this patient and her fetus? (5 marks)
3. Discuss the antenatal, intrapartum and postpartum management of this pregnancy. (10 marks)
4. How would you manage the neonate after delivery? (6 marks)

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Week:

General Week -Transfusion Week- **Haemostasis Week**- Oncology Week

Essay 01:

A 29-year-old woman is admitted to hospital with acute left-sided weakness and expressive dysphasia. CT angiography confirms an acute right middle cerebral artery infarction. She undergoes successful thrombectomy.

Her medical history reveals: Deep vein thrombosis at age 22 following a minor ankle sprain and two first-trimester miscarriages and one fetal loss at 24 weeks despite aspirin therapy. She has never smoked and is not taking hormonal contraception. Her BMI is 35 and she is on anti-hypertensive and diabetic medications.

Investigation Result

Hb	109 g/L
Platelets	$82 \times 10^9/L$
PT	Normal
APTT	72 s (RR 25–36 s)
Fibrinogen	4.8g/L

Thrombophilia screen: Anticardiolipin IgG: >120 GPL units and Anti- $\beta 2$ glycoprotein I IgG: Strongly positive. A transthoracic echocardiogram shows small sterile vegetations on the mitral valve.

Questions

- Describe the laboratory diagnosis of antiphospholipid syndrome (APS), including the principle and limitations of screening lupus anticoagulant testing. (07 marks)
- Discuss the general management of thrombosis in antiphospholipid syndrome. (09 marks)
- Six months later, she is admitted acutely with largely deranged liver and renal function tests, respiratory distress and thrombocytopenia. Imaging

shows widespread small-vessel thrombosis. What is the likely diagnosis and its diagnostic criteria. Outline the treatment as well. (09 marks)

Essay 02:

A 19-year-old boy developed excessive bleeding following tonsillectomy under ENT. He was born to first-cousin parents of Middle Eastern origin. Since infancy, he has experienced:

- Easy bruising
- Gingival bleeding
- Recurrent epistaxis requiring nasal packing
- Prolonged bleeding after circumcision
- Menorrhagia is reported in his affected older sister
- No haemarthroses or spontaneous muscle bleeds

He has received multiple platelet transfusions during childhood. There is no history of liver disease or medication use.

On examination: slow bleeding from surgical site.

Investigation Result

Hb	68 g/L
Platelets	$68 \times 10^9/L$
MPV	16.5 fL
PT	Normal
APTT	Normal
Fibrinogen	Normal
VWF antigen	Normal

Platelet function testing:

- Normal aggregation with ADP
- Normal collagen response
- Normal adrenaline response
- Normal arachidonic acid response
- Absent response to ristocetin
- No correction after addition of normal plasma

Questions

- A. Discuss the molecular basis and the investigation required for diagnosis. (07 marks)
- B. Outline the management of the patient (10 marks)
- C. The patient is reviewed five years later before elective splenectomy for hypersplenism secondary to portal hypertension unrelated to his inherited disorder. Discuss the perioperative haemostatic management of the patient. (8 marks)

Essay 03:

A 58-year-old man is admitted with increasing breathlessness and swelling of the right leg. He was diagnosed with metastatic pancreatic adenocarcinoma four months previously with liver metastases and is receiving palliative chemotherapy.

Three months ago, he developed an incidental segmental pulmonary embolism and was commenced on therapeutic apixaban.

He now presents with:

- Progressive right leg pain and swelling
- Mild haemoptysis
- ECOG performance status 2
- Weight loss of 12 kg
- Intermittent melaena over the past two weeks

Examination reveals a swollen, tender right leg extending to the groin.

CT pulmonary angiography demonstrates new bilateral pulmonary emboli.

Compression ultrasound confirms extensive iliofemoral DVT.

Upper gastrointestinal endoscopy identifies a bleeding duodenal ulcer, which is treated endoscopically.

Questions

- A. Discuss the management of recurrent cancer-associated thrombosis in this patient. (10 marks)
- B. Discuss the role of inferior vena cava (IVC) filters in cancer-associated thrombosis. (7 marks)
- C. Two months later, he is scheduled for an urgent pancreatic bypass operation because of malignant gastric outlet obstruction. Discuss the perioperative management of anticoagulation in this patient. (8 marks)

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Week:

General Week -Transfusion Week- **Haemostasis Week**- Oncology Week

Essay 01:

A 68-year-old woman is referred to the haematology department with a 3-week history of spontaneous bruising and recurrent painful swelling of her forearms and lower legs. Over the preceding 48 hours she has developed persistent oral bleeding and haematuria.

She has never experienced abnormal bleeding previously. She underwent a hysterectomy at the age of 42 and had several dental extractions without excessive bleeding.

Her medical history includes Rheumatoid arthritis, Hypertension, Osteoporosis

Her medications include amlodipine, prednisolone 5 mg daily and alendronate. She is not taking anticoagulants, aspirin or other antiplatelet drugs.

On examination, she has extensive ecchymoses over both arms and legs and a large tender intramuscular haematoma involving the left calf. There is no splenomegaly or lymphadenopathy.

Test	Result	Reference range
------	--------	-----------------

Hb	78 g/L	115–160
----	--------	---------

Platelets	$264 \times 10^9/L$	150–400
-----------	---------------------	---------

PT	12.4 s	10.5–13.5
----	--------	-----------

APTT	86 s	25–35
------	------	-------

A repeat APTT using a different analyser remains markedly prolonged.

Questions

1. Describe the principle of APTT 03marks

1:1 mixing study is performed using patient plasma and normal pooled plasma.

Incubation	Immediate APTT	After 2 h at 37°C
Patient plasma	86 s	89 s
1:1 mixture	52 s	78 s
Normal plasma	31 s	32 s

Factor assays subsequently demonstrate:

Factor	Activity
FVIII	1.2 IU/dL
FIX	94 IU/dL
FXI	101 IU/dL
FXII	88 IU/dL

Bethesda inhibitor assay demonstrates FVIII inhibitor 48 Bethesda units/mL.

2. What is the most likely diagnosis? The patient develops progressive enlargement of the calf haematoma with a fall in Hb to 64 g/L. She requires urgent haemostatic treatment. Discuss the immediate management of the patient. 08marks
3. The patient is subsequently diagnosed with acquired haemophilia A associated with her underlying autoimmune disease. Discuss the inhibitor eradication in this patient. 06marks
4. Because of recurrent bleeding and difficulty controlling her FVIII inhibitor, the multidisciplinary team considers emicizumab. What information you need to provide the patient. 08marks

Essay 02:

A 31-year-old woman, G2P0, presents to the antenatal haematology clinic at 7 weeks' gestation after a positive pregnancy test.

She underwent mechanical aortic valve replacement with a bileaflet prosthesis 4 years ago following severe congenital aortic valve disease. She has been maintained on warfarin 6 mg daily, with a target INR of 2.5–3.0.

Before pregnancy, her INR had been relatively stable. Her most recent INR, performed 10 days ago, was 2.8.

At review, she asks whether she can simply remain on warfarin because her valve thrombosis risk concerns her more than the potential fetal risks. She is also concerned about injections.

The multidisciplinary team discusses switching her from warfarin to therapeutic-dose low-molecular-weight heparin (LMWH).

Questions

1. The patient wants to continue warfarin. Discuss the anticoagulation management with the patient during her pregnancy. 10 marks
2. Discuss the role of laboratory monitoring of anticoagulation during pregnancy. 4 marks
3. How would you manage anticoagulation from 35 weeks until delivery? 6 marks
4. Outline the postpartum anticoagulation strategy. 5 marks

Essay 03:

A 25-year-old woman presents with swelling and pain in her left leg. Doppler ultrasound confirms a proximal deep vein thrombosis (DVT).

History: no previous thrombosis, no known family history. She is not on oral contraceptives and is otherwise healthy.

She is planning pregnancy in the next year.

Questions:

- A. What laboratory investigations should be performed in this patient 06 marks
- B. The patient is found to have antithrombin deficiency. List different causes of anti-thrombin deficiency and how is antithrombin deficiency confirmed in the laboratory? 07marks
- C. Discuss the management of the patient during her pregnancy 12 marks



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Week:

General Week -Transfusion Week- Haemostasis Week- **Oncology Week**

Essay 01:

A 74-year-old woman presents with a 4-month history of progressive fatigue, drenching night sweats, unintentional weight loss (8 kg), worsening abdominal fullness, and intermittent fevers. Over the last 3 weeks, she has also developed increasing shortness of breath on exertion and new lower limb swelling.

She has a background of hypertension, atrial fibrillation (on apixaban), chronic kidney disease stage 3, and type 2 diabetes mellitus.

On examination:

- ECOG performance status: 2
- Bilateral cervical and inguinal lymphadenopathy
- Large left axillary node (4 cm)
- Massive splenomegaly (8 cm below costal margin)

Investigations

Test	Result	Reference
Hb	89 g/L	120–160
WBC	$14.5 \times 10^9/L$	4–11
Platelets	$138 \times 10^9/L$	150–400
LDH	980 U/L	<250

CT/PET scan shows:

- Extensive FDG-avid lymphadenopathy above and below diaphragm
- Bulky retroperitoneal mass (9 cm)

- Splenic and hepatic involvement
- Bone marrow uptake suspicious for involvement

Excisional lymph node biopsy shows:

- Diffuse effacement by large atypical lymphoid cells with prominent nucleoli and frequent mitoses

Bone marrow biopsy confirms involvement by lymphoma.

The diagnosis is high-grade B-cell lymphoma with MYC and BCL2 rearrangement (double-hit lymphoma), Ann Arbor stage IVB, high IPI score.

Questions

1. Discuss the diagnosis, prognostic factors, and first-line treatment of this patient. (07 marks)
2. The patient achieves partial remission after first-line therapy but progresses within 6 months of completion of treatment. Discuss the management for relapsed/refractory disease. (10 marks)
3. The patient is commenced on bispecific antibody therapy after being deemed unsuitable for CAR-T therapy.

List the important toxicities associated with bispecific antibodies and briefly discuss the recognition and management of Cytokine Release Syndrome (CRS).

Essay 02:

A 48-year-old man presents to the emergency department with a 1-week history of rapidly worsening facial swelling, breathlessness, early satiety, and severe drenching night sweats. He also reports a 2-month history of marked weight loss (10 kg), profound fatigue, and intermittent fevers.

Over the last 48 hours, he has developed worsening confusion, reduced urine output, and severe lower back pain.

He has no significant past medical history.

On examination:

- ECOG performance status: 2
- Tachycardic and mildly hypotensive
- Marked facial plethora with distended neck veins
- Large mediastinal fullness
- Massive abdominal mass palpable in the left upper quadrant

Investigations

Test	Result	Reference
Hb	92 g/L	130–180
WBC	$18 \times 10^9/\text{L}$	4–11
Platelets	$160 \times 10^9/\text{L}$	150–400
LDH	3200 U/L	135–225

CT chest/abdomen/pelvis:

- Large anterior mediastinal mass causing partial SVC compression
- Large ileocecal mass
- Bilateral renal infiltration

Bone marrow aspirate:

- Heavy infiltration by monomorphic medium-sized deeply basophilic lymphoid cells

Questions

- What are the important differential diagnoses for this presentation? (3 marks)
- Discuss the immediate and definitive management of this patient.

(12 marks)

- Within 24 hours of starting treatment, the patient develops worsening oliguria and the following blood results:

Test	Result
K ⁺	6.8 mmol/L
Phosphate	3.2 mmol/L
Calcium	1.65 mmol/L
Creatinine	420 $\mu\text{mol/L}$
Uric acid	markedly elevated

What is the most likely diagnosis? Discuss the pathophysiology and outline urgent management.

(10 marks)

Essay 03:

A 34-year-old woman, 10 weeks pregnant, presents with worsening fatigue, headaches, drenching night sweats, and increasing abdominal discomfort over the

past 3 months. She also reports recent easy bruising and intermittent visual blurring.

She has no significant past medical history, and this is her first pregnancy after several years of infertility treatment.

On examination:

- ECOG performance status: 1
- Plethoric appearance
- Massive splenomegaly (10 cm below the costal margin)
- Fundoscopy shows retinal venous engorgement

Initial Investigations

Parameter	Result	Reference Range
Hb	98 g/L	120–160
WBC	$535 \times 10^9/L$	4–11
Platelets	$680 \times 10^9/L$	150–400

Peripheral blood film:

- Marked granulocytic proliferation with all stages of myeloid maturation

Bone marrow aspirate:

- Hypercellular marrow with marked myeloid hyperplasia
- Blasts 7%

Questions

a. Discuss the diagnosis and outline the management of this patient, including the challenges posed by pregnancy. (10 marks)

b. After delivery, she is commenced on first-line TKI therapy and achieves major molecular response.

Explain Treatment-Free Remission (TFR) and discuss the eligibility criteria for attempting TFR. (07 marks)

c. Two years later, she develops rising BCR::ABL1 transcript levels despite compliance with therapy.

Discuss the causes of treatment failure, the investigations required, and management of TKI resistance. (08 marks)

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Week:

General Week -Transfusion Week- Haemostasis Week- **Oncology Week**

Essay 01:

A 72-year-old woman presents with increasing fatigue, recurrent epistaxis, blurred vision, and intermittent headaches over the past month. She also reports numbness in both feet and unintentional weight loss.

Examination: Pale appearance, Mild hepatosplenomegaly and Retinal examination show dilated tortuous veins with retinal haemorrhages

Initial laboratory results: Hb 88 g/L, WCC $6.0 \times 10^9/L$ and Platelets $140 \times 10^9/L$

Serum immunoglobulins:

- IgM: 58 g/L ↑
- IgG: 3 g/L ↓
- IgA: 0.4 g/L ↓

Serum electrophoresis:

- Monoclonal IgM- λ paraprotein: 42 g/L

PB Flow cytometry:

- CD19+, CD20+, CD22+, surface IgM λ +
- CD5 negative, CD10 negative, CD23 negative

1. What is the likely explanation for the patient's visual symptoms and headaches? (03marks)
2. List FIVE important complications associated with this condition. Discuss the management of this patient. (12 marks)
3. Six years later, the patient develops rapidly enlarging cervical lymph nodes, drenching night sweats, rising LDH, and worsening cytopenias.

PET-CT demonstrates highly FDG-avid lymphadenopathy. Outline your diagnostic and treatment approach. (10)

Essay 02:

A 33-year-old woman, gravida 1 para 0, at 24 weeks gestation presents with worsening fatigue, gum bleeding, recurrent fevers, and progressive shortness of breath over 3 weeks.

On examination: Pale and tachycardic, Multiple ecchymoses over the limbs, Gingival hypertrophy and Mild splenomegaly

Obstetric review: Fetal growth appropriate for gestational age

Initial investigations:

Test	Result
Hb	62 g/L
WBC	$68 \times 10^9/L$
Neutrophils	$1.2 \times 10^9/L$
Platelets	$18 \times 10^9/L$
PT	Prolonged
APTT	Prolonged
Fibrinogen	1.0 g/L
D-dimer	Elevated

Peripheral blood film: Circulating blasts with folded nuclei and prominent nucleoli.

Flow cytometry: CD13+, CD33+, MPO+, CD117+, HLA-DR negative

1. Describe the WHO classification of AML with recurrent genetic abnormalities. (06marks)
2. Discuss the maternal and fetal risks associated with AML and its treatment during pregnancy. (06marks)
3. Discuss the management of this patient. (10marks)
4. What supportive care measures are particularly important in this patient? (03marks)

Essay 03:

A 58-year-old man presents with a 10-month history of gradually enlarging neck and groin lumps associated with fatigue and early satiety. He reports drenching night sweats and an unintentional 6-kg weight loss over the last 3 months.

Examination: Generalized non-tender lymphadenopathy

Test	Result
------	--------

Hb	98 g/L
----	--------

WBC	$5.5 \times 10^9/L$
-----	---------------------

Platelets	$110 \times 10^9/L$
-----------	---------------------

LDH	Mildly elevated
-----	-----------------

CT-PET:

- Widespread FDG-avid lymphadenopathy above and below diaphragm
- Splenic involvement

Excisional lymph node biopsy Immunophenotype:

- CD20+, CD10+, BCL6+, BCL2+, CD5 negative and Cyclin D1 negative

1. Discuss the indications for treatment and treatment options for this patient. (07 marks)
2. Three years later, the patient develops rapidly enlarging cervical lymph nodes, worsening B symptoms, rising LDH, and a PET scan showing intensely FDG-avid nodes (SUV markedly increased). Discuss your management approach. (12 marks)
3. Briefly discuss the role of transplant and Bispecifics in this condition. (06marks)

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Week:

General Week -Transfusion Week- Haemostasis Week- **Oncology Week**

Essay 01:

A 78-year-old man is referred with progressive fatigue, drenching night sweats, and an unintentional weight loss of 10 kg over the preceding 6 months.

His medical history includes Ischaemic heart disease and chronic kidney disease stage 3

Examination reveals:

- Generalized lymphadenopathy (largest node 4 cm)
- Massive splenomegaly extending 12 cm below the left costal margin

Tests	Result
Hb	86 g/L
WBC	$38 \times 10^9/L$
Lymphocytes	$35 \times 10^9/L$
Platelets	$72 \times 10^9/L$

PET-CT: Extensive supra- and infra diaphragmatic lymphadenopathy along with Splenic involvement

Immunophenotype: CD20+ CD5+ FMC7+ Surface IgM+CD23- CD10+

Questions:

1. How would you stage and risk-stratify this patient? (6 marks)
2. Outline your initial management plan. (09 marks)
3. Sixteen months later, after initial response, he develops rapidly enlarging lymph nodes and worsening constitutional symptoms. Biopsy confirms relapsed mantle cell lymphoma with TP53 mutation. Outline your management plan (10 marks)

Essay 02:

A 31-year-old woman who is 24 weeks pregnant (G2P1) presents with a 10-day history of fatigue, gum bleeding, epistaxis, and increasing shortness of breath.

On examination:

- Multiple ecchymoses
- Tachycardia (115 bpm)
- Oxygen saturation 95% on room air

Investigations:

- Hb 71 g/L
- WBC $38 \times 10^9/L$
- Platelets $14 \times 10^9/L$
- PT prolonged
- APTT mildly prolonged
- Fibrinogen 0.8 g/L
- D-dimer markedly elevated
- Numerous abnormal promyelocytes on blood film

Questions:

1. Discuss the pathophysiology of the coagulopathy. (6 marks)
2. Outline your immediate management during the first 24 hours (10 marks)
3. On day 5 of treatment, she develops Weight gain 4 kg and Increasing oxygen requirement and WCC. What is the most likely diagnosis? Discuss pathogenesis, prevention and management. (9 marks)

Essay 03:

A 58-year-old man presents with progressive fatigue, weight loss, night sweats, and early satiety.

Examination: Splenomegaly (5 cm below costal margin)

Investigations: Hb 92 g/L, WBC $38 \times 10^9/L$, Monocytes $8.5 \times 10^9/L$ (persistent for >3 months) and Platelets $78 \times 10^9/L$

Blood film: Dysplastic neutrophils, Monocytosis with abnormal monocytes and Occasional circulating blasts

Bone marrow: Hypercellular marrow, Myeloid dysplasia and Blasts 12% (including promonocytes). Cytogenetics: normal karyotype

Questions:

- a) What is the likely diagnosis and its key diagnostic criteria 05marks
- b) Discuss the classification and poor prognostic factors 08marks
- c) Discuss the management of the above patient. Also mention the role of stem cell transplant 12marks

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Week:

General Week -Transfusion Week- Haemostasis Week- **Oncology Week**

Essay 01:

A 58-year-old Caucasian man is referred urgently to haematology after presenting with progressive fatigue, left calf swelling, chest pain and recurrent sinus infections over the past six months. He reports a 10-kg weight loss.

His history includes chronic kidney disease stage 2 and well-controlled hypertension.

On examination: Multiple tender ribs, No lymphadenopathy and tender left calf swelling.

ECOG performance status 1

Test	Result
Hb	81 g/L
WCC	$4.5 \times 10^9/L$
Platelets	$115 \times 10^9/L$
Creatinine	210 $\mu\text{mol/L}$
Calcium	3.05 mmol/L
Albumin	28 g/L
LDH	490 U/L (raised)
β 2-microglobulin	9.2 mg/L
Total protein	108 g/L

Serum protein electrophoresis demonstrates an IgA paraprotein of 52 g/L.
Bone marrow aspirate: 70% plasma cells.

1. Discuss the likely diagnosis and the prognostic significance of cytogenetic abnormalities in this condition. (04 marks)
2. Outline your initial management during the first 72 hours. (08 marks)
3. Describe the optimal first-line treatment strategy according to current UK practice (08 marks)
4. Three years later post-transplant, he develops progressive anemia and rising paraprotein. How would you manage his 1st relapse? (05 marks)

Essay 02:

A 74-year-old retired engineer is admitted with worsening fatigue, exertional dyspnoea and new onset jaundice over two weeks.

He has noticed progressive cervical swelling over the previous six months and has lost 8 kg unintentionally. He reports drenching night sweats but denies fever.

Past medical history: COPD, Permanent atrial fibrillation treated with rivaroxaban, Previous NSTEMI with coronary stent and Hypertension

Medications: Rivaroxaban, Bisoprolol and Ramipril

Performance Status: 2

Initial Investigations: Hb 61 g/L, MCV 108 fL, WCC $118 \times 10^9/L$, lymphocyte count $108 \times 10^9/L$, Platelets $42 \times 10^9/L$, Haptoglobin Undetectable, DAT Positive (IgG)

Blood film: Numerous mature lymphocytes, Smudge cells and Spherocytes

Question

- A. What is the most likely diagnosis? Discuss the immediate management of the patient. (08 marks)
- B. List the indication of treatment. Outline the 1st line treatment strategy for this condition. (08 marks)
- C. Six years later, He develops rapidly enlarging cervical nodes. LDH increases from 350 to 1700 IU/L. PET-CT demonstrates an SUV max of 18. Describe treatment options if biopsy confirms Richter transformation. (09 marks)

Essay 03:

A 20-year-old man presents with a 3-week history of worsening dyspnoea, night sweats, weight loss (8 kg), facial swelling and increasing fatigue.

On examination he has: Respiratory distress (RR 34/min), BP 95/60 mmHg, Oxygen saturation 88% on air, Raised JVP, Marked facial oedema. Large mediastinal mass on chest X-ray. The patient developed hypotension and transferred to ICU.

Initial Investigations: Hb: 84 g/L, WBC: $185 \times 10^9/L$, Platelets: $24 \times 10^9/L$, LDH: 4200 IU/L, Creatinine: 135 $\mu\text{mol/L}$

Peripheral blood: 95% blasts

Flow Cytometry: CD2+, CD7+, CD5 (dim), cyCD3+, TdT+, CD34+, CD1a-, CD8-, CD4-, CD33+, CD13+, MPO negative

Questions

- A. Discuss the emergency management during the first 24 hours. (06 marks)
- B. Discuss the differential diagnosis and diagnostic work-up. (05 marks)
- C. Discuss risk stratification and initial treatment. (06 marks)
- D. Eighteen months later, the patient develops marrow relapse with rising blasts. MRD was positive at end of consolidation. How would you manage relapsed condition?
08marks



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Week:

General Week -Transfusion Week- Haemostasis Week- **Oncology Week**

Essay 01:

A 67-year-old woman is referred to the haematology clinic with a 9-month history of progressive fatigue, exertional dyspnoea and recurrent urinary tract infections. She has also noticed increasing bruising over the last 3 months.

Her past medical history includes chronic kidney disease stage 2, hypertension and osteoarthritis. She has no history of previous chemotherapy or radiotherapy. Her regular medications are ramipril, amlodipine and paracetamol.

On examination, she is pale but haemodynamically stable. There is no palpable lymphadenopathy or hepatosplenomegaly.

Investigation	Result
Hb	76 g/L
MCV	108 fL
WBC	$2.8 \times 10^9/L$
Neutrophils	$0.9 \times 10^9/L$
Platelets	$62 \times 10^9/L$

Blood film: Macro-ovalocytes, anisopoikilocytosis, hypogranular neutrophils with occasional pseudo-Pelger-Huët cells and thrombocytopenia. No circulating blasts are identified.

Bone marrow aspirate and trephine: Hypercellular marrow with multilineage dysplasia. Blasts comprise 6% of nucleated cells. There is no evidence of metastatic malignancy or significant fibrosis.

Questions

1. Discuss your approach to the investigation of an older patient presenting with macrocytic anaemia and pancytopenia. 07 marks
2. How would you classify this patient's myeloid neoplasm and assess their prognosis? Discuss the clinical, morphological, cytogenetic and molecular factors that should be considered. 08 marks
3. discuss the management of this patient, including supportive care, disease-modifying treatment and the role of allogeneic stem cell transplantation. 10marks

Essay 02:

A 71-year-old woman presented to acute ambulatory care due to left calf swelling. She was referred to the haematology department by her GP due increasing lethargy, abdominal discomfort and recurrent episodes of dizziness over the preceding 6 months. She is still awaiting appointment.

Her past medical history includes chronic hepatitis C infection, hypertension and osteoarthritis. She has not received previous chemotherapy.

On examination, she has pallor and mild jaundice. There is no significant peripheral lymphadenopathy. The spleen is palpable 15 cm below the left costal margin.

Test	Result
Hb	7.8 g/dL
MCV	102 fL
WBC	$3.2 \times 10^9/L$
Lymphocytes	$1.4 \times 10^9/L$
Platelets	$68 \times 10^9/L$
Bilirubin	48 $\mu\text{mol/L}$
LDH	520 U/L
Haptoglobin	Undetectable
DAT	IgG positive, C3d positive

Blood film shows small mature lymphocytes, occasional villous lymphocytes and polychromasia.

Flow cytometry demonstrates a clonal B-cell population: Flow cytometry: CD19+, CD20+, CD22+, CD5-, CD10-, CD23-. CT demonstrates massive splenomegaly without significant lymphadenopathy. Bone marrow trephine shows an interstitial and intrasinusoidal infiltrate of small B lymphocytes.

Questions

1. What additional investigations would you perform to establish the diagnosis and assess prognosis? (8 marks)
2. Outline the management of this patient. (10 marks)
3. The patient achieves remission following treatment but develops symptomatic progressive disease 3 years later. Discuss the treatment options for relapsed disease. (7 marks)

Essay 03:

A 67-year-old man is referred to the regional CAR-T multidisciplinary team with relapsed diffuse large B-cell lymphoma (DLBCL).

He was initially diagnosed 18 months ago and achieved a complete metabolic response following R-CHOP. Eight months later, he developed progressive cervical and abdominal lymphadenopathy. A lymph-node biopsy confirms recurrent CD19-positive DLBCL.

He receives salvage chemotherapy with R-DHAP but develops progressive disease after two cycles.

ECOG performance status 1

Hb	94 g/L
WCC	$4.2 \times 10^9/\text{L}$
Neutrophils	$2.6 \times 10^9/\text{L}$
Platelets	$118 \times 10^9/\text{L}$
Creatinine	86 $\mu\text{mol/L}$
Bilirubin	14 $\mu\text{mol/L}$
ALT	32 U/L
LDH	680 U/L
PET-CT	Progressive FDG-avid nodal disease
MRI brain	No CNS involvement

The CAR-T MDT considers him suitable for axicabtagene ciloleucel. Leukapheresis is performed successfully.

Four weeks later, following lymphodepletion with fludarabine and cyclophosphamide, he receives axicabtagene ciloleucel.

Day +5: He develops: Temperature 39.2°C, BP 105/65 mmHg, Oxygen saturation 96% on air and No focal neurological symptoms. Blood cultures are taken and broad-spectrum antibiotics commenced. No obvious source of infection is identified.

Day +7: His fever has settled, but he becomes increasingly confused. He is unable to correctly write a sentence and has difficulty naming objects. He is awake but intermittently inattentive. There is no focal neurological deficit.

Questions

- List the indications for CAR-T therapy in the UK (05 marks)
- What factors should be considered when assessing a patient for CAR-T therapy before leukapheresis and infusion? (05 marks)
- On day +5, what are the important differential diagnoses for his fever? Outline your management if the patient has persistent fever. (07 marks)
- On day +7, what is the most likely diagnosis? How would you grade the neurological toxicity and what treatment approach would you use? (08 marks)

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Week:

General Week - **Transfusion Week** - Haemostasis Week - Oncology Week

Essay 01:

A 32-year-old woman, G3P2, attends at 10 weeks' gestation for booking.

- Blood group: O RhD-negative
- Previous obstetric history:
 - Pregnancy 1: uncomplicated, delivered a healthy D-positive baby
 - Pregnancy 2: required emergency Caesarean section for placental abruption; received 4 units RBCs

Current Investigations:

- Antibody screen: positive
- Identified antibodies:
 - Anti-D: 3 IU/mL
 - Anti-c: 5 IU/mL

Questions:

1. Discuss the role of cell fetal free DNA in pregnancy management
06marks
2. Outline the management of this pregnancy 12marks
3. A male infant is delivered at 35 weeks. He is pale and jaundiced within 2 hours. Cord blood: Hb: 85 g/L , Bilirubin: rising rapidly and DAT: strongly positive. What is the immediate management? 7marks

Essay 02:

A 34-year-old woman, G3P1, presents at 12 weeks' gestation.

Obstetric history:

- Previous Pregnancy: miscarriage at 18 weeks with unexplained fetal intracranial haemorrhage
- No maternal bleeding disorder

Current pregnancy:

- Mother: HPA-1a negative, HPA-1b positive
- Father: HPA-1a positive (homozygous)
- Maternal serum: anti-HPA-1a antibodies detected at high titre
- Blood group: O RhD-positive

Questions:

- A. Explain the mechanism of Fetal and Neonatal Alloimmune Thrombocytopenia. What factors influence severity of disease in FNAIT? 06marks
- B. Discuss the management of this pregnancy 12marks
- C. A male infant is delivered at 37 weeks by elective Caesarean section. At 4 hours of life, the baby had fits. Investigations showed platelets of $8 \times 10^9/L$. MRI brain revealed intracranial haemorrhage. Outline the management plan 07marks

Essay 03:

A 56-year-old woman with Acute Myeloid Leukaemia is admitted for chemotherapy (FLAG-Ida).

- Received multiple transfusions during admission
- One platelet pool was later identified as non-irradiated

Clinical Course (Day +12 post transfusion):

She develops:

- Persistent fever (38.5°C) despite broad-spectrum antibiotics. Blood cultures negative
- Generalised erythematous rash → progressing to desquamation
- Liver dysfunction: ALT 180 IU/L and Bilirubin $45 \mu\text{mol/L}$
- Worsening pancytopenia:

Questions:

- A. List the differential diagnoses for this presentation 05marks
- B. What tests would help confirm the diagnosis? 06marks
- C. Discuss the management of this case in detail 14marks



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Week:

General Week - **Transfusion Week** - Haemostasis Week - Oncology Week

Essay 01:

A 63-year-old man with acute myeloid leukaemia is on day 12 following induction chemotherapy. He is profoundly neutropenic and thrombocytopenic. He is transfused a pool of platelets and one unit of Red cells for a platelet count of $8 \times 10^9/L$ and Haemoglobin of 68g/L.

Ten minutes after the transfusion begins, he develops: High fever ($40.2^\circ C$), Hypotension (BP 75/45 mmHg) and Tachycardia (HR 130 bpm)

On examination: Flushed, peripherally vasodilated

Initial Investigations

Test	Result
Hb	68 g/L
WBC	$0.4 \times 10^9/L$
Platelets	$8 \times 10^9/L$
CRP	Elevated

Coagulation Early prolongation of PT/APTT

Questions

1. What is the most likely diagnosis, and what features in the case support it over other causes? (5 marks)
2. Outline the management of this patient. (12 marks)
3. Outline the specific measures used in the UK to reduce the risk of bacterial contamination in blood products, particularly platelets. (8 marks)

Essay 02:

A 89-year-old man with transfusion-dependent myelodysplastic syndrome is admitted with worsening fatigue and dizziness.

Past medical history: Chronic kidney disease (stage 4)

On admission: Hb 64 g/L

He is prescribed 2 units of red cells, each over 2 hours.

During the second unit, he develops:

- Orthopnoea
- Hypertension (BP 190/100 mmHg)

On examination:

- Bilateral basal crackles

Test Result

CXR : Features of pulmonary oedema

Questions

1. What are the differential diagnoses for this acute deterioration during transfusion? (3 marks)
2. How would you distinguish clinically and with investigations between TACO and TRALI in this case? (5 marks)
3. Outline the management of this patient. (10 marks)
4. What factors in this patient increased his risk of developing this complication? (3 marks)
5. Following recovery, what transfusion strategies would you implement to prevent recurrence? (4 marks)

Essay 03:

A 36-year-old woman with high-risk acute lymphoblastic leukaemia is scheduled for a matched unrelated donor allogeneic stem cell transplant.

Parameter	Recipient	Donor
Blood group	A RhD positive	O RhD positive
CMV status	Negative	Positive

She receives conditioning with fludarabine, cyclophosphamide, and total body irradiation, followed by peripheral blood stem cell infusion.

GVHD prophylaxis: ciclosporin + methotrexate

Day +12

- Sudden drop in Hb
- Haemoglobinuria
- Rising LDH and bilirubin

Questions

1. What type of ABO and Rh mismatch is present, and what are the expected complications in this setting? How can they be avoided (07 marks)
2. Outline the transfusion strategy for red cells, platelets, Granulocytes and plasma during early post-transplant phase (08marks)
3. Discuss the management of complication identified at Day +12? (10 marks)

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Week:

General Week - **Transfusion Week** - Haemostasis Week - Oncology Week

Essay 01:

A 68-year-old man with severe calcific aortic stenosis is scheduled for elective aortic valve replacement (AVR) in 8 weeks.

He is a Jehovah's Witness and has provided an advance directive refusing transfusion of red cells, platelets, fresh frozen plasma (FFP), and cryoprecipitate.

Past Medical History:

- Chronic kidney disease stage 3
- Iron deficiency anaemia secondary to occult gastrointestinal blood loss
- Atrial fibrillation treated with Apixaban

Test	Result
Hb	82 g/L
MCV	72 fL
Ferritin	8 µg/L
Platelets	280 ×10 ⁹ /L

The cardiothoracic surgeon requests urgent haematology advice regarding optimisation before surgery and management of potential major bleeding.

Questions

1. Which of the following products or therapies may be acceptable to some Jehovah's Witness patients? (05marks)
2. What important discussions and documentation should be completed before surgery? (05marks)
3. How would you optimise this patient's haemoglobin before surgery? (10marks)

4. The patient suffers catastrophic postoperative haemorrhage with Hb falling to 42 g/L. What emergency management options remain available despite refusal of blood transfusion? (05marks)

Essay 02:

A 29-year-old woman (Blood Group A RhD-negative) is admitted at 35 weeks' gestation with severe antepartum haemorrhage due to placenta praevia.

She has had: Two previous miscarriages, Anti-D prophylaxis at 28 weeks and No known red cell antibodies

An emergency Caesarean section is performed. During surgery she develops massive obstetric haemorrhage (estimated blood loss 5.5 L) and receives:

- 6 units RBC
- 4 units FFP
- 2 adult therapeutic doses of platelets
- 4 pools cryoprecipitate

Because of severe blood shortages and the urgency of transfusion, the first 4 units of RBCs issued are O RhD-positive emergency units.

Questions

1. What are the recognised causes of fetomaternal haemorrhage (FMH)? (05marks)
2. Explain the principles, advantages and limitations of the Kleihauer-Betke test. (06marks)
3. Discuss the management of this patient who has received a large-volume RhD-positive red cell transfusion. (10marks)
4. Despite anti-D administration, anti-D antibodies are detected 6 months later. How would you determine whether this represents passive vs immune Anti D and why is this distinction important? (04marks)

Essay 03:

A 60-year-old woman is admitted for elective hip replacement.

She has no prior bleeding history but has had two pregnancies and one transfusion 20 years ago.

During surgery she receives 2 units RBC and 1 adult therapeutic dose of platelets. Pre-op platelets: $220 \times 10^9/L$ while Day 7 post-transfusion.

She presents acutely with Extensive bruising. Platelet count $4 \times 10^9/L$

PT/APTT normal. Fibrinogen normal. Hb: 68 g/L.

1. What is the most likely diagnosis and what differential diagnoses should be considered? (4 marks)

2. Discuss the pathogenesis and laboratory investigation of this condition.
Outline the treatment options (10 marks)
3. The patient develops persistent multidrug-resistant *Klebsiella* sepsis despite optimal antimicrobial therapy. Discuss the indications, compatibility testing, administration, efficacy and risks of granulocyte transfusions. (11 marks)

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Week:

General Week - **Transfusion Week** - Haemostasis Week - Oncology Week

Essay 01:

A 28-year-old woman is brought to the Emergency Department following a high-speed road traffic accident. She was the unrestrained driver and required prolonged extrication.

On arrival

- GCS 14/15
- BP 78/46 mmHg
- Pulse 142 bpm
- Respiratory rate 30/min

Examination reveals: Open pelvic fracture and Bilateral femoral fractures

She is transferred immediately to theatre for damage control surgery. During surgery, despite activation of the Major Haemorrhage Protocol she continues to bleed diffusely. Estimated blood loss reaches approximately 8 litres over 2 hours.

She receives 10 units RBC, 4 units FFP, 1 adult therapeutic dose platelets

Laboratory Results

Hb 68 g/L

Platelets $42 \times 10^9/L$

PT 23 s

APTT 58 s

Fibrinogen 0.7g/L

TEG performed: R Time 16min (5-10min), Alpha angle 66degree (53-72) and MA 34mm (50-70).

Questions

- A. Discuss the immediate haemostatic management of this patient, including activation of the Massive Haemorrhage Protocol. (11 marks)

- B. Interpret the TEG findings and explain how they guide blood component replacement. (05 marks)
- C. Explain the causes of coagulopathy in this patient. (05 marks)
- D. List the complications of massive transfusion. (04 marks)

Essay 02:

A 76-year-old woman is reviewed in the pre-operative assessment clinic before an elective total hip replacement in 5 weeks.

Past medical history

- Heart failure (EF 38%)
- Atrial fibrillation
- Chronic kidney disease stage 4 (eGFR 24 mL/min)

Medications

- Apixaban 5 mg twice daily
- Furosemide
- Ferrous sulfate (poor adherence because of gastrointestinal side effects)

Investigations

Test	Result	Reference range
Hb	88 g/L	120–160
MCV	84 fL	80–100
Ferritin	145 µg/L	30–300
Transferrin saturation	12%	20–45
Platelets	460 ×10 ⁹ /L	150–400
INR	1.6	0.9–1.2

The orthopaedic surgeon asks for advice regarding optimisation before surgery.

Questions:

- A. Define Patient Blood Management (PBM) and describe its principles. (05 marks)
- B. Outline a comprehensive PBM strategy to optimise this patient before surgery. (10 marks)
- C. Discuss strategies to minimise peri-operative blood loss in this patient. (05 marks)
- D. Explain the role of restrictive transfusion thresholds in modern PBM and discuss circumstances in which transfusion would still be appropriate. (05 marks)

Essay 03:

A 58-year-old woman with relapsed multiple myeloma is admitted with melaena and symptomatic anaemia. She has completed six cycles of isatuximab, pomalidomide and dexamethasone. She requires urgent red cell transfusion.

Her records indicate that she experienced a severe anaphylactic reaction during a plasma transfusion five years previously. Subsequent investigations confirmed severe selective IgA deficiency with anti-IgA antibodies.

Laboratory investigations

Hb	58 g/L
WBC	$5.6 \times 10^9/L$
Platelets	$48 \times 10^9/L$
Reticulocytes	$32 \times 10^9/L$
Blood group	O RhD negative
Antibody screen	Pan-reactive (3+ all cells)
DAT	Negative
Serum IgA	<0.05 g/L

The blood transfusion laboratory reports that compatibility testing cannot identify underlying alloantibodies because of pan-reactivity.

Questions

- Explain the causes of the pan-reactive antibody screen and the likely cause in this patient. (05 marks)
- Discuss the transfusion risks associated with severe IgA deficiency. (06 marks)
- Outline your approach to providing safe red cell transfusion for this patient (08 marks)
- What pre-treatment investigations should be performed before starting anti-CD38 therapy. Explain why these investigations are important. (6 marks)

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Week:

General Week - **Transfusion Week**- Haemostasis Week- Oncology Week

Essay 01:

A 35-year-old man with HbSS sickle cell disease presents to the Emergency Department with worsening right upper quadrant pain, fever, and jaundice.

Ultrasound demonstrates acute calculous cholecystitis with multiple pigment gallstones. The surgical team recommends laparoscopic cholecystectomy within the next 24 hours.

His medical history includes: HbSS sickle cell disease, Previous acute chest syndrome requiring ICU admission 18 months ago, Recurrent vaso-occlusive crises (4 admissions/year), Pulmonary hypertension diagnosed on echocardiography and not taking hydroxycarbamide due to previous intolerance. He has history of Multiple previous blood transfusions with Anti-K and anti-Jk^a alloantibodies

Investigations: Hb 81 g/L, WCC $18 \times 10^9/L$, Platelets $520 \times 10^9/L$, ALT 110 U/L and Creatinine 128 $\mu\text{mol/L}$

Hb electrophoresis: HbS 89%, HbF 10% and HbA 1%

Blood group: B RhD Negative

Questions

- What additional investigations would you perform before deciding on transfusion and surgery? (5 marks)
- What specifications should the transfused red cells meet in this patient? (5 marks)
- Outline your peri-operative management plan to minimise sickle-related complications. (10 marks)
- What transfusion-related complications are of particular concern in this patient, and how would you minimise these risks? (5 marks)

Essay 02:

You are rotating through transfusion services in a District General Hospital as a part of your training.

You receive an urgent notification from NHS Blood and Transplant (NHSBT) that, due to a combination of reduced donor collections, manufacturing disruption and increased national demand, the UK has entered an Amber Phase Red Cell Shortage Plan. Your hospital has been instructed to reduce red cell usage immediately for the next 5 days, with only 6 O-negative units and 12 group-specific red cell units available until the next delivery.

Within the next few hours, the following patients require assessment for transfusion:

- a) A 74-year-old man with metastatic colorectal cancer has chronic iron deficiency anaemia (Hb 69 g/L) and is scheduled for palliative chemotherapy tomorrow. He is haemodynamically stable.
- b) A 68-year-old woman undergoing emergency laparotomy for perforated bowel develops major haemorrhage. Estimated blood loss is 2.8 L and she has already received 4 units of red cells.

Questions

- A. Prioritise the allocation of the available red cell units among these patients. Justify your decisions using clinical urgency, expected benefit, patient blood management principles, and national transfusion guidance. (08 marks)
- B. Discuss evidence-based strategies to reduce red cell utilisation during a national blood shortage, both at patient level and hospital level. (09 marks)
- C. Describe how you would coordinate and manage this blood shortage over the next five days. (08 marks)

Essay 03:

A 62-year-old man is admitted with severe community-acquired pneumonia. He has a background of type 2 diabetes and chronic kidney disease stage 3.

On day 2 of admission, his haemoglobin falls to 68 g/L following an episode of melaena. He receives 2 units of red cells and 1 unit of fresh frozen plasma (FFP) for suspected coagulopathy.

Thirty minutes into the FFP transfusion, he develops:

- Acute breathlessness
- Oxygen saturation falls from 96% to 82% on air
- Tachycardia and hypotension
- Fever of 38.5°C

Examination reveals bilateral crackles. Chest X-ray shows bilateral pulmonary infiltrates. Echocardiography shows normal left ventricular function and no evidence of fluid overload.

Questions:

- a) What is the most likely diagnosis? List the differential diagnoses. (05 marks)
- b) Discuss the management of this patient (12 marks)
- c) What measures have reduced the incidence of recurrence (08marks)

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Week:

General Week -Transfusion Week- Haemostasis Week- Oncology Week-
Miscellaneous Week

Essay 01:

A 46-year-old man underwent a liver transplant 4 years ago for primary sclerosing cholangitis. His immunosuppression consists of tacrolimus, mycophenolate mofetil and prednisolone.

He presents with a 6-week history of fever, night sweats, weight loss, progressive fatigue and cervical lymphadenopathy.

Investigations:

- Hb: 98 g/L

- WCC: $3.8 \times 10^9/L$
- Platelets: $112 \times 10^9/L$
- LDH: 1,280 U/L
- LFTs: mildly abnormal
- EBV DNA: 450,000 IU/mL
- CT: cervical, mediastinal and abdominal lymphadenopathy with multiple hepatic lesions
- Lymph-node biopsy: diffuse proliferation of large atypical lymphoid cells, CD20+, PAX5+, CD10-, BCL6+, MUM1+, EBER positive, high Ki-67 index

The diagnosis is monomorphic post-transplant lymphoproliferative disorder (PTLD), diffuse large B-cell lymphoma type.

1. State the main categories of PTLD according to the WHO/ICC classification. (4 marks)
2. Describe the major risk factors and pathogenesis of PTLD. (5 marks)
3. Outline the staging and baseline investigations required before treatment. (6 marks)
4. Discuss the management of this patient. (10 marks)

Essay 02:

A 64-year-old woman is referred with a 3-year history of recurrent episodes of Flushing, Severe abdominal cramps and diarrhoea, Wheeze and Dizziness and near-syncope

On two occasions, she required emergency treatment for anaphylaxis following wasp stings.

Investigations show:

- Hb: 94 g/L
- WCC: $18.5 \times 10^9/L$
- Platelets: $88 \times 10^9/L$
- LDH: elevated
- Serum tryptase: 180 $\mu g/L$

Examination reveals marked splenomegaly (8 cm below the costal margin) and hepatomegaly.

A bone marrow biopsy shows multifocal aggregates of atypical spindle-shaped mast cells. Mast cells express CD117, tryptase, CD25 and CD2. Molecular testing detects a KIT D816V mutation.

Questions

1. What is the most likely diagnosis, and how would you classify this patient according to the current classification of systemic mastocytosis? (4 marks)
2. Describe the diagnostic criteria. (6 marks)

3. What further investigations would you perform to assess the extent and severity of disease? (5 marks)

4. Discuss the management of this patient. (10 marks)

Essay 03:

A 29-year-old woman, currently 11+5 weeks pregnant, attends her booking appointment in a high-prevalence area. Her family origin is Pakistani, and her partner is of Bangladeshi origin. She reports no previous transfusions and has no known family history of a haemoglobin disorder.

Test	Result
Hb	108 g/L
MCV	65 fL
MCH	20 pg
RBC	$5.7 \times 10^9/\text{L}$
Ferritin	42 $\mu\text{g/L}$

Haemoglobin analysis by HPLC demonstrates:

- HbA: 93.5%
- HbA₂: 5.1%
- HbF: 1.4%
- No variant haemoglobin detected

Questions

A. What is the most likely maternal diagnosis? Outline the next steps in the management of this pregnancy. (6 marks)

B. The father's screening shows Hb 139 g/L, MCV 67 fL and MCH 21 pg. HbA₂ is 2.8% and HbF is 0.5%. Explain the possible causes of these findings and outline further investigations. (5 marks)

The couple later have a male baby. The newborn blood spot result at 5 days shows a pattern of F only, with no detectable HbA.

C. What are the differential diagnoses, and what are the limitations of newborn haemoglobinopathy screening? (7 marks)

The laboratory is subsequently asked to advise on haemoglobinopathy testing for the mother's sister, who received a red-cell transfusion 6 weeks previously.

D. Discuss the laboratory methods available for haemoglobinopathy diagnosis and their important limitations. (7 marks)

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