Higher Specialist Diploma

Haematology

September 2024

Short Answer Questions

60 minutes

Attempt all Four Questions

Instructions to Candidates

1. Record your candidate number and HSD discipline on the front sheet of the answer booklet.

2. Record your candidate number, the question number and the page number in the spaces provided on the answer sheets.

3. Begin each new answer on a new page.

4. Each question is worth 25 marks.

1. Your current APTT reagent is no longer available according to your reagent manufacturer. Outline your approach and considerations in replacing this reagent with a new one.

2. Your manager has asked you to give a presentation to the new band 5 staff about IQC in the haematology laboratory. Briefly describe the key points and learning outcomes of your talk.

3. Your laboratory is due to become part of a regional network and will take on specialist haematology and coagulation testing for the region. Briefly discuss the challenges this will pose and how you would address them.

4. A batch of patient samples demonstrated HbA2 greater than 3.5% for every single sample. How would you investigate this finding and what action would you take?

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Essay Paper

120 minutes

Attempt 2 out of 5 Questions

Instructions to Candidates

1. Record your candidate number and HSD discipline on the front sheet of the answer booklet.

2. Record your candidate number, the question number and the page number in the spaces provided on the answer sheets.

3. Begin each new question on a new page.

4. Each question is worth 100 marks.

1. Critically discuss how you would investigate and diagnose an apparent leucoerythroblastic blood picture in a 67-year-old patient with no previous history and no clinical details. In your answer, you should refer to relevant guidelines and evaluate the contribution the laboratory makes to this particular diagnosis.

2. Critically discuss the investigation of heritable thrombophilia in a patient receiving anticoagulant therapy.

3. Critically discuss the diagnosis and treatment of TTP within the context of the haematology laboratory.

4. Critically discuss, with named examples, the haematological changes seen in infection and outline the investigations that would help confirm that the changes described are due to a reactive process.

5. Critically discuss the modes of detection and enumeration, causes and consequences of schistocytes in a peripheral blood sample

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Case Studies

120 minutes

Attempt all Case Studies

Instructions to Candidates

1. Record your candidate number and HSD discipline on the front sheet of the answer booklet.

2. Record your candidate number, the question number and the page number in the spaces provided on the answer sheets.

3. Begin **each new case study** on a new page.

4. Each question is worth 100 marks.

5. For these case study questions you are strongly advised to answer the questions as they arise during the case study to avoid later information impacting adversely on your answers to the earlier questions by presuming an "outcome".

SEEN CASE STUDY

1.

A 39-year-old male develops severe epistaxis while out shopping and rapidly becomes unconscious. An ambulance is called, and he is blue-lighted to A&E. No further details are available.

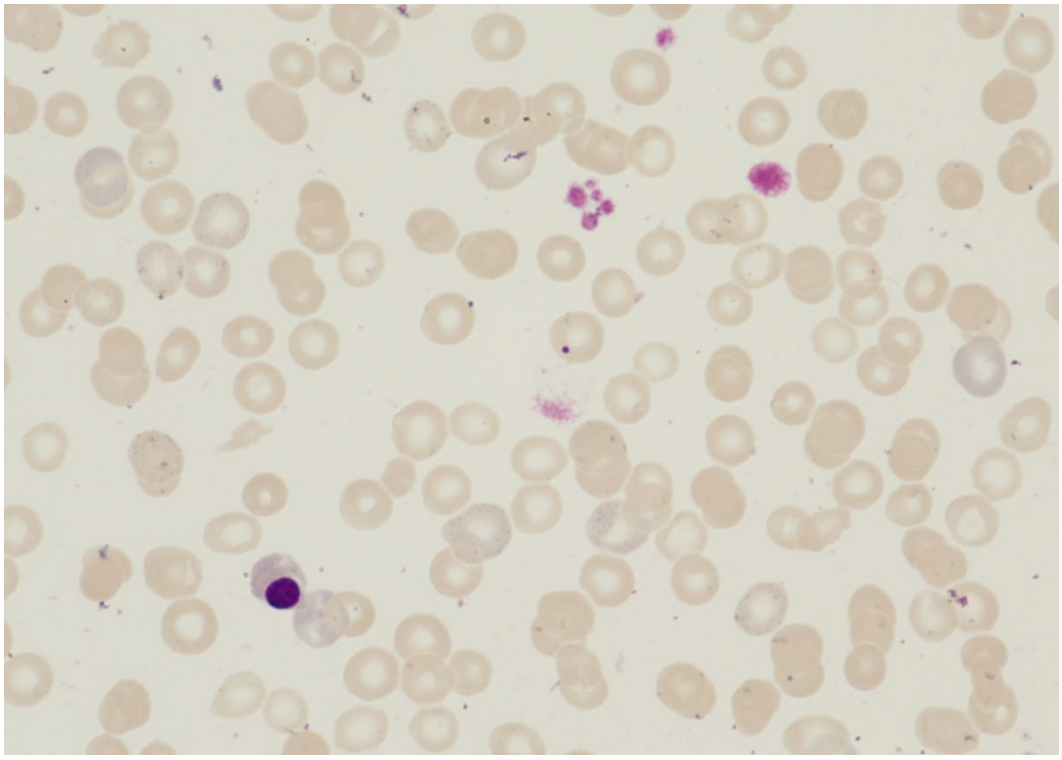
Initial blood gas analysis performed in Resus shows the haemoglobin is 64g/L. An urgent FBC and coagulation screen are sent, and the results are shown below

Parameter	**Result**	**Reference Range**	**Units**
White Blood Cell Count	9.7	4.0 - 11.5	$x10^9/L$
Haemoglobin	61	130 - 170	g/L
Platelets	17	150 - 450	$x10^9/L$
Red cell count	2.59	4.50 - 6.50	$x10^{12}/L$
Haematocrit	0.197	0.40 - 0.52	L/L
Mean Cell Volume	76.1	80 - 100	fL
Mean cell Haemoglobin	24.5	27 - 32	pg
Neutrophils	7.0	2.0 - 7.5	$x10^9/L$
Lymphocytes	2.1	1.0 - 4.0	$x10^9/L$
Monocytes	0.5	0.2 - 0.8	$x10^9/L$
Eosinophils	0.1	0.0 - 0.4	$x10^9/L$
Basophils	0.0	0.0 - 0.1	$x10^9/L$
PT	11	10 - 13	Seconds
APTT	36	30 - 40	Seconds
Fibrinogen	2.6	1.9 - 4.8	g/L

a. Discuss the results. (5 marks)

b. What further actions would you take and why? (5 marks)

c. Describe the features seen on the blood film below. (5 marks)



d. The platelet clumps are very scanty – what further actions would you take and why? (4 marks)

e. A citrated platelet count is performed, and the platelet count does not improve. What further actions would you take and why? (6 marks)

f. Explain the mechanisms behind pseudothrombocytopenia. (10 marks)

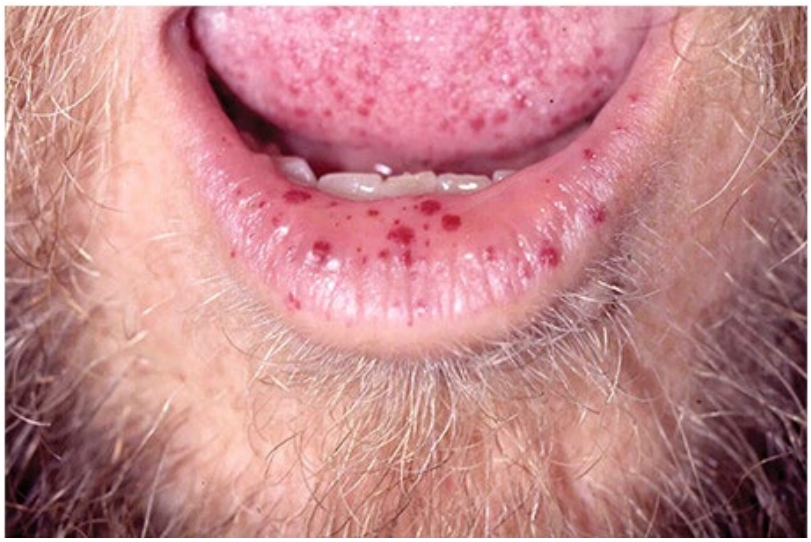
The Consultant Haematologist reviews the blood film and determines that although small platelet clumps are present the platelet count is genuinely low.

g. What is the differential diagnosis for thrombocytopenia? (10 marks)

The patient, who is now conscious, states that he has a long history of severe nosebleeds but comments that “he has never had it investigated as it’s just something that runs in the family”.

h. What is the differential diagnosis for epistaxis? (10 marks)

Extensive screening for bleeding disorders is performed and is found to be normal. Further examination of the patient reveals the following in the patient’s mouth. They are also found on the patient’s hands and fingertips.

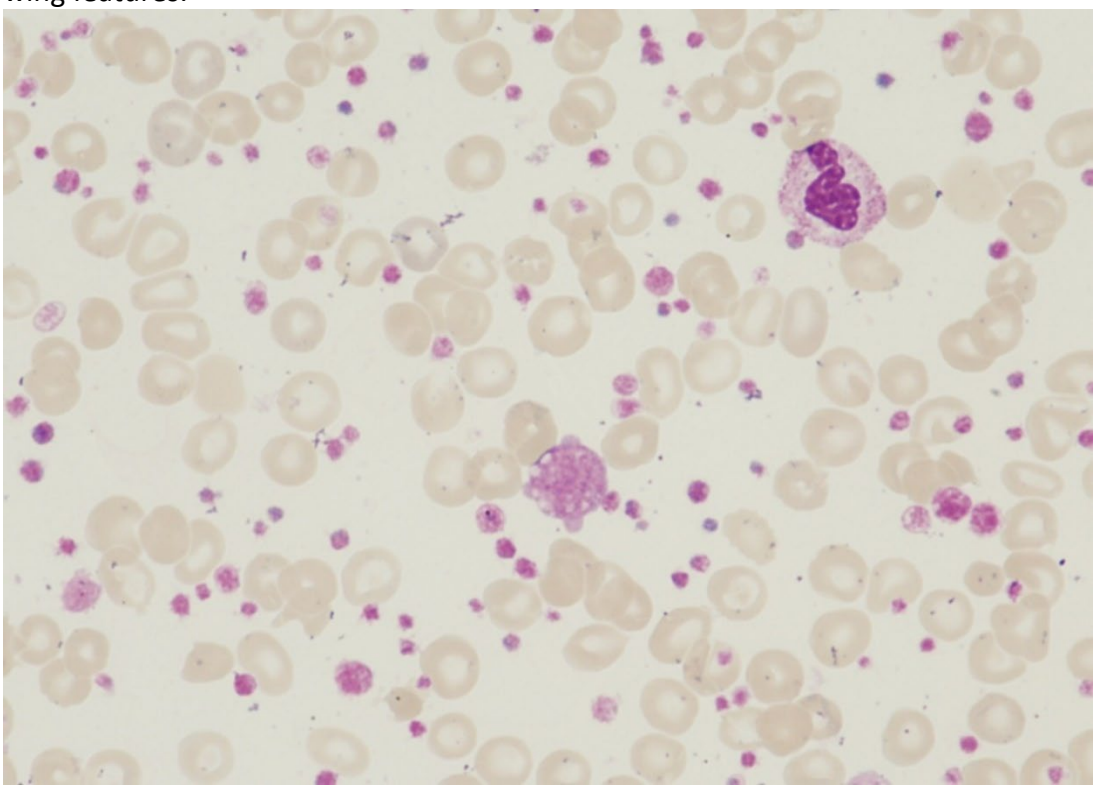


i. What are they and do they help narrow your suspected diagnosis? (5 marks)

Genetic testing is performed and shows a mutation in the *ACVRL1* gene.

j. What do these results confirm? In the absence of genetic testing what criteria need to be met to confirm the suspected diagnosis. (10 marks)

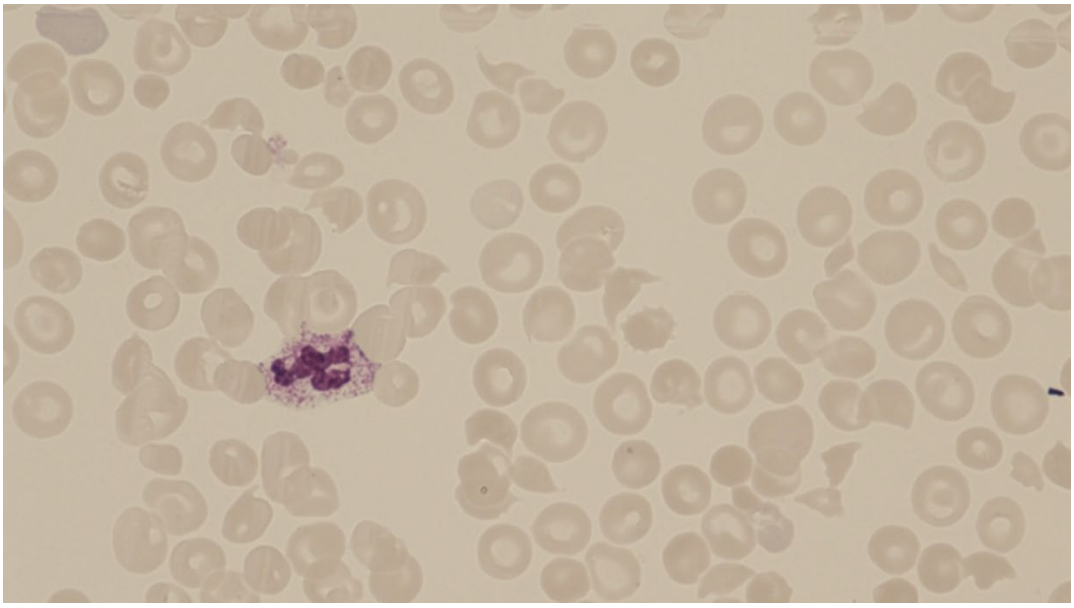
The patient continues to experience a low platelet count and it is confirmed on review of historical clinical notes now available that they have a confirmed diagnosis of ITP. The patient is started on romiplostim, and after a short period of treatment the blood film shows the following features.



k. Comment on the morphological features present and estimate the platelet count. Explain the mechanism of romiplostim and why the features are seen. (15 marks)

l. If the film image above was from a presentation film what would be your suspected diagnosis? And what genetic tests would be appropriate? (5 marks)

The romiplostim is stopped and the patient continues to deteriorate following a long stay on ITU. The blood film now shows the following features:



m. Comment on the morphology what is your suspected complication? (7 marks)

n. What is your final diagnosis for this patient? (3 marks)

UNSEEN CASE STUDIES

2.

You are on a secondment as a Biomedical Scientist working at the University of Port Harcourt teaching hospital, Nigeria. A 27-year-old male patient presented to the emergency department with generalised body pains, fever, weakness, and palpitations of four days duration. The patient is known to the hospital and has received intermittent treatment for vaso-occlusive crises from sickle cell anaemia. The patient was diagnosed with his condition at the age of two-years. The patient was in painful distress with pyrexia (temperature 38.3 °C), pallor and slight jaundice.

There was no significant peripheral lymphadenopathy. His respiratory rate was 24 breaths/minute, pulse was 120 beats/minute and regular, and his blood pressure was 120/70 mmHg. There was no abdominal tenderness, but the liver was palpably enlarged, 4 cm

below the right costal margin and had a span of 18 cm. The spleen was not palpable. Subsequent abdominal ultrasound revealed hepatomegaly and auto-splenectomy.

a. Using the patient's Full Blood Count in Table 1 and the information provided in Table 2, classify the red cells using the appropriate nomenclature. Explain the association between the **red** cell results in these tables and how the results in table 2 influence the findings in table 1. (10 marks)

Table 1: Results from the initial full blood count investigation

Parameter	**Result**	**Reference range**	**Units**
Haemoglobin	46	133 - 167	g/L
Mean Cell Volume	96	77 - 98	fL
Mean Cell Haemoglobin	29	26 - 33	pg
Mean Cell Haemoglobin Concentration	348	330 - 370	pg/L
Platelets	335	143 - 400	x 10^9/L
White Blood Cell Count	44	4.0 - 10.0	x 10^9/L
Neutrophils	4.4	2.0 - 7.0	x 10^9/L
Lymphocytes	3.08	1.0 - 3.0	x 10^9/L
Monocytes	2.5	0.2 - 1.0	x 10^9/L
Eosinophils	0.4	0.02 - 0.5	x 10^9/L
Basophils	0.02	0.02 - 0.1	x 10^9/L
Large Unstained Cells	33.6		x 10^9/L

Table 2: HPLC analysis on presentation

Haemoglobin	Concentration (%)
A	0%
S	93.7%
A2	3.1%
F	3.2%

b. Comment on the patient's **white** cell count. Explain the possible causes of these findings and, from the table only, explain the morphological features you expect to be exhibited by these white cells upon blood film examination. (15 marks)

c. Figure 1 shows the patient's initial blood film. Despite its poor quality, you review the film and comment on the key features you observe. Provide a detailed account of the blood film findings and critically review these in relation to the patient's presentation and your answer in question B. (20 marks)

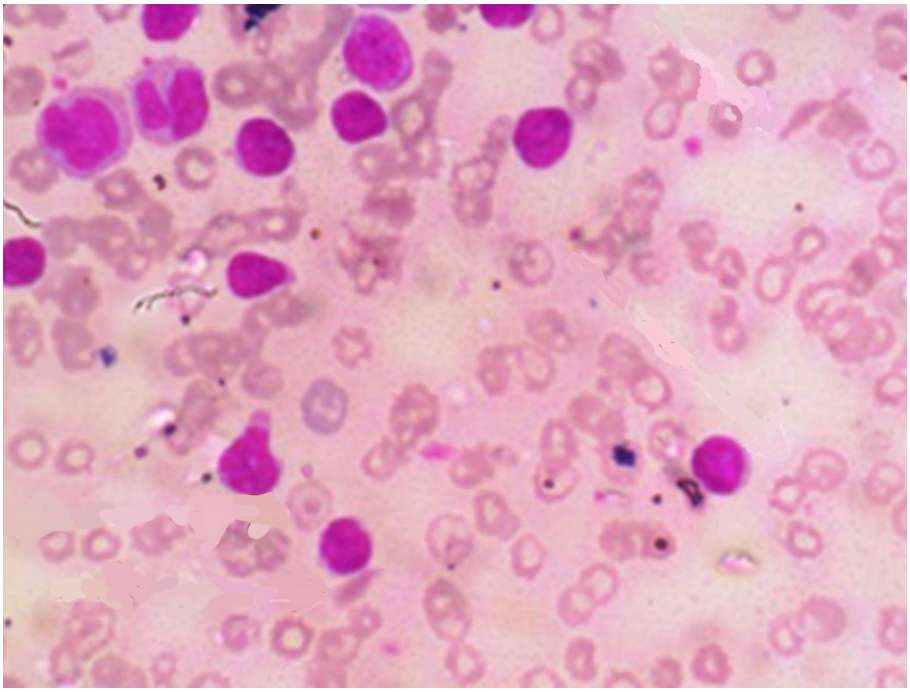


Figure 1. Peripheral blood film from initial investigations.

An ambiguous observation (not shown in Figure 1) prompted you to produce a thick and thin film from this patient's sample. An image from this thin film is shown in Figure 2.

d. Explain the differences between the preparation of thin and standard blood films and discuss the significance of the results of this analysis for this patient. From a practical perspective, explain your next course of action. (15 marks)

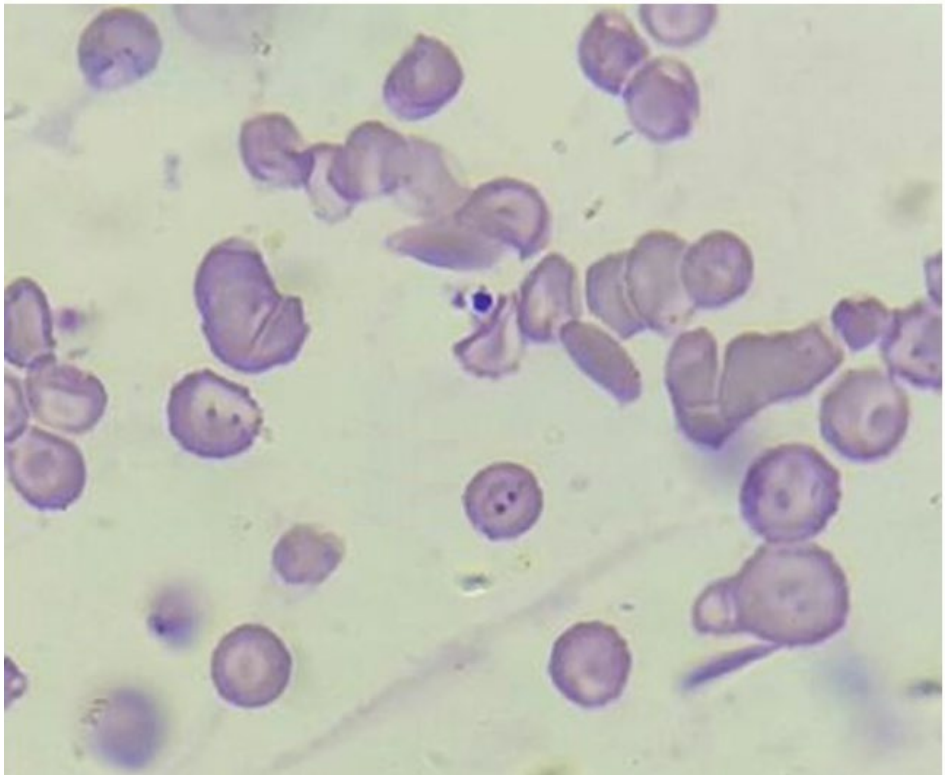


Figure 2. Supplementary thin film

Based upon the observations in figure 1, immunophenotyping was also requested. Selected results are presented in Table 3.

Table 3 Selected immunophenotyping results following blood film examination.

CD20	Positive
CD3	Negative
CD13	Negative
CD33	Negative

e. Evaluate these results and consider how these results may be reflected in the patient's clinical presentation. (20 marks)

f. Genetic analysis revealed these cells harbour 46XY, t(9;22)(q34;q11) while molecular testing demonstrated the presence of the E1a3 transcript. Critically discuss the significance of these results. (10 marks)

g. Given the information available to you, provide a definitive diagnosis for this patient and indicate the types of treatment that could be employed to manage his condition. (10 marks)

3.
A 24-year-old male is in A&E with an extensive haematoma following mild trauma. The clinician takes a clinical history and requests a full blood count and clotting screen.

a. What should the clinical history comprise? (5 marks)

Initial laboratory results are as follows:

Parameter	**Result**	**Reference range**	**Units**
WBC	5.7	4.0 - 11.0	x 10^9/L
Hb	135	130 - 160	g/L
Platelets	350	150 - 400	x 10^9/L
HCT	0.40	0.37 - 0.48	-
MCV	90	80 - 98	fL
PT	12	10 - 13	Sec
APTT	38	30 - 40	Sec
Fibrinogen	2.5	2.0 - 4.0	g/L

b. Comment on these results. (5 marks)

c. From the clinical information and these data alone, what disorder (if any) could be responsible? (5 marks)

d. The clinician asks for clotting assays – is this justified given the normal clotting screen? (5 marks)

Results are reported as follows:

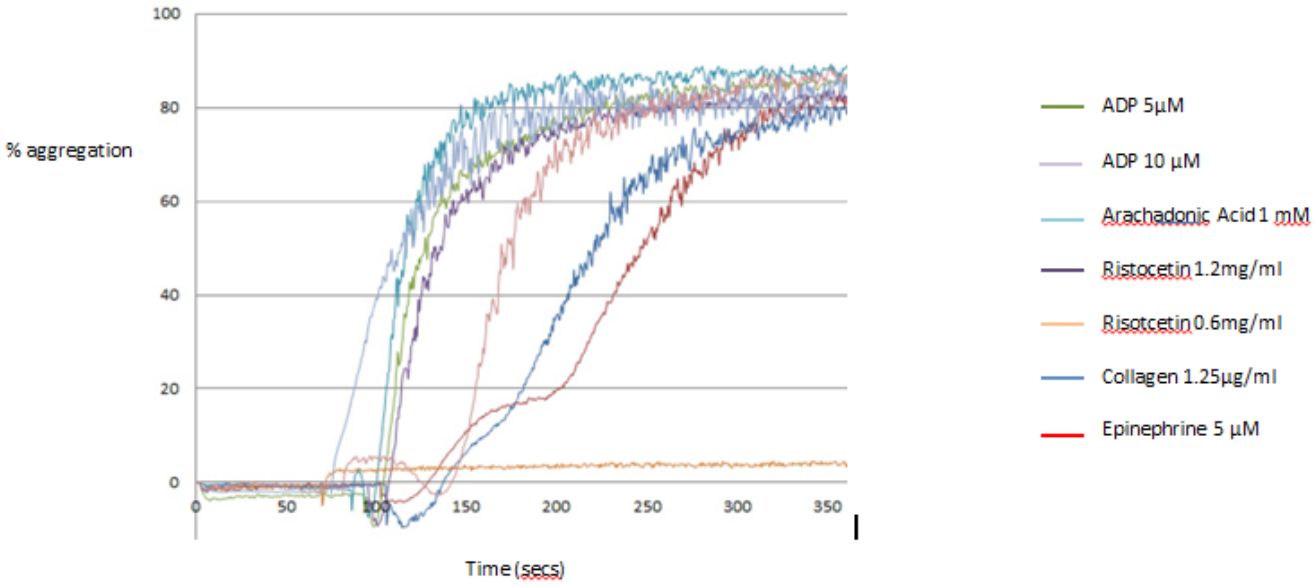
1-stage clotting assays	Result	Reference range	Units
FVIII:C	52	50 – 150	%
FIX:C	89	50 – 150	%
FXI:C	121	50 – 150	%
FXII:C	82	50 – 150	%

e. Comment on the data in this table. (10 marks)

The patient is referred to a specialist Haemophilia centre for further investigation. The haematologist takes a full history and uses the ISTH Bleeding assessment Tool (BAT).

f. Comment on the utility of this assessment tool. (5 marks)

The Haemophilia centre lab carries out platelet investigations, with the following light transmission aggregometry results:



g. Comment on these results. (10 marks)

h. How might these results have been affected if the patient had taken aspirin? (5 marks)

The haemophilia centre lab repeats the clotting screen and assays, with the following results:

Parameter	Result	Reference range	Units
PT	10	9-13	Sec
APTT	29	20 – 30	Sec
Fibrinogen	2.5	2.0 - 4.0	g/L

Assays	Result	Reference range	Units
FVIII:C	20	50 – 150	iu/dL
FIX:C	100	50 – 150	iu/dL
FXI:C	111	50 – 150	iu/dL
FXII:C	99	50 – 150	u/dL
VWF parameters			
VWF:Ag	80	50 – 150	iu/dL
VWF Activity	90	50 – 150	iu/dL
VWF:CB	80	50 – 150	iu/dL

i. Comment on these results. What conclusions and/or diagnosis can you make? (15 marks)

j. What reasons might explain the discrepant results between the two centres? (15 marks)

k. Are there any other tests you would consider to make a diagnosis in this patient? (15 marks)

A decision is made to treat this patient with FVIII concentrate if further bleeding events occur.

l. What considerations would you have with regard to monitoring the patient's treatment levels? (5 marks)