ACEM Primary Examination Vivas > Pathology > Haematology Organised by edvivas.com	
Title	**Page**
Anaemia 2014-2-D	1
Anaemia, Haemolytic 2010-1	2
Anaemia, Haemolytic	3
Anaemia, Iron Deficiency 2016-2-A	4
Anaemia, Iron Deficiency 2010-2	5
Anaemia, Pernicious 2006-2	6
Haemophilia A 2007-2	7
Sickle Cell Disease 2015-1-C	8
Sickle Cell Disease 2007-2	9
Thrombocytopenia 2012-2	10
Von Willebrand's Disease 2009-1	11
Von Willebrand's Disease 2007-2	12

Anaemia 2014-2-D

Stem: An 80 year old woman presents to ED following a fall secondary to an episode of melaena.			
TOPIC	QUESTIONS	KNOWLEDGE **(essential in bold)**	NOTES
Question 1 Anaemia (pp 639-665) **Subject:** Path LOA: 1	1. How are the causes of anaemia classified? Prompt if use RC morphology: How are the causes classified by mechanism? Prompt for example if not volunteered.	1. **Blood loss**: acute, chronic 2. **Increased RC destruction** Inherited genetic: H Spherocytosis, G6PD, Thal, Sickle cell Acq genetic: Parox noct hemo. Ab mediated: transfusion, drugs, Rh disease. Mech trauma: HUS, DIC, TTP, cardiac valves, runners. Infx: malaria; Toxic: envenom, clostridia, Pb. 3. **Decreased RC production** Inherited genetic: Fanconi's, thalassemia. Nutritional: B12/folate, iron. Erythropoietin deficit: renal failure, chronic dis. Immune: aplastic anaemia.	Bold main headings & 1 example of each to pass.
	2. Describe the pathogenesis of iron deficiency anaemia.	Causes: Chronic blood loss, poor diet, impaired absorption, incr reqs Iron stores used up first – **ferritin** haemosiderin. Once reserves depleted **serum iron & transferrin** decr. Erythroid activity increases, no iron in marrow macrophages. RCs become **hypochromic & microcytic.**	Bold to pass.
	3. (Please give examples of anaemias that are more common in specific ethnic groups.) Ask if there is time.	Hereditary spherocytosis: northern Europe G6PD: 10% African American, Africa, Middle East, Med Sickle cell: African descent, up to 30% Thalassemia trait: Africa, Asia, Med, India Pernicious: Scandinavian, Caucasian.	1 correct with example.

Anaemia, Haemolytic 2010-1

	antibodies persist for months	
1. Classify haemolytic anaemias	- Intravascular/extravascular Or - extrinsic/intrinsic to the RBC. Or - hereditary/acquired	One classification,
2. Describe the common features of haemolytic anaemias.	Features: - *Decreased RBC life span(< 120/7) due to premature destruction - ^ erythropoietin and erythropoiesis - Accumulation of products of Hb catabolism - reticulocytosis	• premature RBC destruction and one other feature
3. Give some important causes of intravascular haemolysis. Prompt for examples	Intravascular - Mechanical injury: cardiac valves, microangiopathic, repetitive physical trauma - Complement fixation: ABO incompatible blood transfusion - Intracellular parasites: malaria - Exogenous toxins: clostridia	• 2 of 4
If required **4. Apart from anaemia what are the results/manifestations of intravascular haemolysis?**	**- *Haemoglobinaemia** **- Haemoglobinuria** **- *Unconjugated hyperbilirubinaemia(jaundice) from catabolism of haem groups in mononuclear phagocyte system** **- Haemosiderinuria and renal haemosiderosis** **- Decreased serum haptoglobin due binding with free Hb and then cleared by monophag system.** **- Free Hb oxidized to metHb** **- Reticulocytosis**	* **Hbaemia and hyperbilirubinaemia to pass and one other** OR 3 of 7

Anaemia, Haemolytic

Question 5 Anaemia LOA: 2	**What are the causes of intravascular haemolysis?** **What are the manifestations of intravascular haemolysis?** (Prompt: In the blood? In the urine?)	-mechanical injury to cells (valves, microthrombi, other physical trauma) - complement fixation (eg transfusion reaction) -toxic injury (eg clostridia), - parasites (eg malaria) Anaemia, haemoglobinuria, haemoglobinaemia, jaundice, haemosiderinuria	**3 causes** **3 manifestations**

Anaemia, Iron Deficiency 2016-2-A

Stem: Moving onto Pathology.			
Question 3 Iron deficiency anaemia **Subject:** Pathology LOA: 1	1) What are the causes of Fe deficiency anaemia	**Chronic blood loss – GI tract**, Menorrhagia. Increased requirements – pregnancy, children. Dietary Lack –developing world, infants (prolonged breastfeeding) elderly, extreme diet Impaired absorption –celiac, gastrectomy	Bold + 3 other examples from any categories
	2) What are the symptoms of Fe deficiency anaemia	General –fatigue, weakness, dyspnoea, angina Features of cause – melaena, menorrhagia	4 for pass
	3) Are there any specific features of Fe deficient anaemia?	Koilonychia, alopecia, glossitis,pica, pharyngeal web	1 for pass

Anaemia, Iron Deficiency 2010-2

Question 4.4 Iron Deficiency Anaemia	1. What is the aetiology of Fe deficiency anaemia?	**1.1. Chronic blood loss – GIT, menorrhagia** 1.2. Increased requirement – pregnancy 1.3. Dietary deficiency – vegetarians 1.4. Impaired absorption – celiac	1. Bold + 1
	2. What are the laboratory findings in Fe deficiency anaemia?	2. 2.1. **Microcytic hypochromic anaemia** (low Hb) 2.2. Low S. Fe levels 2.3. Low S. Ferritin levels (correlates well with body iron stores) 2.4. High TIBC (high transferrin levels) 2.5. Low Transferrin saturation levels	2. Bold +3
	3. What are the clinical features of Fe deficiency anaemia?	3. 3.1. **General** - pallor, weakness, lethargy, fatigue, SOBOE, angina 3.2. Features of blood loss – GI, menorrhagia 3.3. **Specific features** – koilonychia, alopecia, glossitis, pica	3. At least 5 from 2 groups

Anaemia, Pernicious 2006-2

TOPIC: PERNICIOUS ANAEMIA _____ **NUMBER: 4** _____

OPENING QUESTION	What is the pathogenesis of pernicious anaemia?	**COMMENTS**
POINTS REQUIRED	1 Immunologically mediated (possibly autoimmune) destruction of gastric mucosa > chronic atrophic gastritis. Likely an autoreactive T-cell response > gastric mucosal injury and production of autoantibodies which may exacerbate epitheleal injury.	Must state immunologically mediated to pass
	2 Loss of parietal cells leads to reduced intrinsic factor production which in turn leads to reduced Vit B_{12} (cobalamin) absorption from the gut, resulting in macrocytic anaemia	Required to pass
	3 Also 3 types of antibodies present in many patients with P.A however their role in pathogenesis is unclear. Type 1 Ab blocks B_{12}-IF binding. Type 2 blocks B_{12}-IF complex binding to ileal receptors. Type 3 Ab recognises alpha and beta subunits of gastric proton pump. Type 3 most likely a response to gastric injury rather than causative	
PROMPTS	Which vitamin deficiency does it involve?	
SECOND QUESTION (if needed)	What are the clinical manifestations of the disease?	
POINTS REQUIRED	1 Insidious onset (due to large existing B_{12} stores) and progressive unless treated.	
	2 Moderate to severe megaloblastic anaemia. Weakness tiredness, pallor	Required to pass
	3 Leukopenia and thrombocytopenia	
	4 Mild jaundice (ineffective erythropoiesis and enhanced peripheral haemolysis	
	5 Atrophic glossitis (shiny glazed appearance)	
	6 Neurologic manifestations may include spastic paraparesis, sensory ataxia and severe paraesthesias (more commonly in lower limbs	
PROMPTS		

Haemophilia A 2007-2

TOPIC: Haemophilia A _____ **NUMBER: Q4** _____

OPENING QUESTION	What is Haemophilia A ?	**COMMENTS**
POINTS REQUIRED	A reduction in amount or activity of factor VIII.	Essential to pass
	Factor VIII is a cofactor for factor IX in the activation of factor X.	Additional information
SECOND QUESTION	X-linked recessive trait Therefore males and homozygous females 30% have no family history so probably due to a new mutation	Essential to pass Prompt: How is it inherited?
	Do patients with haemophilia A always have a family history?	
SECOND QUESTION	Why do patients with Haemophilia A bleed?	
POINTS REQUIRED	Lack of factor VIII affects the intrinsic pathway, inappropriate fibrinolysis, inadequate coagulation Normally the extrinsic pathway produces initial burst of thrombin activation, that activates the intrinsic pathway. Unable to do so in Haemophilia	Essential to pass
	Thrombin activated the intrinsic pathway via factors XI & XII	Additional information
	Thrombin activate TAFI (Thrombin activatable fibrinolysis inhibitor) which inhibits fibrinolysis	Additional information
THIRD QUESTION	**What is the association between clinical severity and Factor VIII levels?**	
	<1% severe 2-5% moderate > 6 -50% mild	Broad concept required to pass

Sickle Cell Disease 2015-1-C

Stem: Moving onto Pathology. The man has sickle cell disease.			
Question 3 Sickle Cell Disease Subject: Path LOA: 2	1. What is sickle cell disease?	**Hereditary** blood disorder **Haemoglobinopathy**	**Bold** (Prompt: is it congenital or acquired?)
	2. What are the major pathological manifestations of sickle cell disease?	• Haemolysis/Haemolytic anaemia • Microvascular occlusions (crises/Tissue ischaemia = severe pain in affected organs eg bones, lungs, liver, spleen) • Splenic enlargement, infarct and dysfunction (Increased susceptibility to infection – encapsulated organisms [eg strep pneumonia, haemophilus influenza])	**2 of 3 to pass** **2 of 5 Bold** to pass
	3. In general how are haemolytic anaemias classified?	**Inherited genetic defects** (RBC Membrane [spherocytosis], enzyme deficiencies [G6PD], haemoglobinopathies [thalassaemia, sickle cell disease]) **Antibody mediated destruction** (transfusion reactions, autoimmune) **Mechanical trauma** (Microangiopathic haemolytic anaemias [HUS, DIC, TTP], cardiac valves) **Infections of red cells** (malaria) **Toxic** (envenomation)	

Sickle Cell Disease 2007-2

Q5. Sickle cell disease	What is sickle cell disease?	Hereditary haemoglobinopathy	An abnormal haemoglobin HbS is formed because of a point mutation in the beta globin chain
		(Generally heterozygous (about 40% HbS) is asymptomatic unless severe hypoxia. Homozygous most haemoglobin is HbS – leads to alteration of the Hb when deoxygenated – sickling (morphological alteration), as well as red cell membrane changes)	Pass criteria: Must state it is an abnormal haemoglobin.
	What are the major clinical features of sickle cell disease?	1.Haemolytic anaemia (anaemia, reticulocytosis, hyperbilirubinaemia) 2.Vaso- occlusive complications/crises 3. Splenomegaly/dysfunction Prone to infections esp pneumococcus/haemophilus	Pass criteria: 2 minimum
	What are the major precipitants for a sickle cell crisis in a prone individual?	1.hypoxia 2. dehydration 3. Drop in pH	2 of 3 Optional depending on time

Thrombocytopenia 2012-2

Q5 Thrombocytopaenia LOA: 1	1. What are the causes of thrombocytopaenia?	**Decreased production of platelets** - Generalised diseases of bone marrow [Aplastic anaemia (congenital / acquired); Marrow infiltration : leukaemia/cancer] - Selective impairment of platelet production [Drug induced (alcohol, thiazides, cytotoxics); Infections (measles, HIV)] - Ineffective megakaryopoieis [Megaloblastic anaemia, Myelodysplastic syndromes ,parox noct Hburia] **Decreased platelet survival** - Immunological destruction [Autoimmune (ITP, SLE); Iso immune (post transfusion, neonatal); Drugs (quinidine, heparin, sulfa); Infections (mono, HIV, CMV)] - Non immunological destruction [DIC, TTP, giant haemangioma, micro-angiopathic haemolytic anaemia; Sequestration] - Hypersplenism; Dilutional	2 groups in bold 2 examples from each
	2. What is the pathogenesis of immune thrombocytopaenic purpura?	Triggers: Primary /Idiopathic ITP : acute / chronic Secondary : drugs ,HIV Chronic – more common – young adult women **Formation of antibodies against platelet** membrane glycoproteins (IIb-IIIa or Ib-IX); Antibodies evident 80% (plasma/platelet surface) Opsonised platelets susceptible to phagocytosis (mononuclear) Spleen probably major site of removal; 80% improve after splenectomy (site destruction + auto antibody synthesis) Acute – disease of childhood Viral illness – abrupt onset; Antiplatelet autoantibodies; Self-limiting, resolves usually within 6 months	Bold to pass

Von Willebrand's Disease 2009-1

Question 4: **von Willebrand Disease**	**What are the haematological and clinical effects of von Willebrand disease?** **Describe the types of von Willebrand Disease**	Haem: Increased bleeding time with normal platelets Increased PT time (Types 1 & 3) Decreased Ristocetin cofactor activity Clin: Spontaneous bleeding from mucous membranes Increased bleeding from wounds Menorrhagia Bleeding into joints rare except in Type 3 **1. Type 1** and **Type 3** associated with decreased circulating vWF. Type 1 most common (70%), autosomal dominant and usually mild. Type 3 autosomal recessive and severe **2. Type 2** has defective vWF, autosomal dominant, mild severity and 25% of cases.	Need 3/4 Need 2/3
Question 5:	**Describe the relationship between**	1. **Increased incidence among people with**	

Von Willebrand's Disease 2007-2

TOPIC: Von Willebrand's Disease _____ **NUMBER: Q5** _____

OPENING QUESTION	What is Von Willebrand's Disease	**COMMENTS**
POINTS REQUIRED	Bleeding disorder Compound defect in platelet function and the coagulation pathway- VWf factor VIII complex Type 1 – mild quantitative reduction in circulating vWF - commonest sub-type (70%) - autosomal dominant - relatively mild clinical syndrome Type 2 - qualitative defect in vWF - autosomal dominant - 10-15% of cases - mild to moderate bleeding syndrome Type 3 - marked quantitative reduction in circulating vWF - relatively rare - autosomal recessive - severe clinical picture	Combination platelet & coagulation abnormality required
	Clinical scenario is one of i) mucous membrane bleeding ii) excessive wound bleeding iii) menorrhagia iv)increased bleeding time v)Bleeding into joints occurs with the more severe types only.	
SECOND QUESTION	What are the effects on the clotting?	
POINTS REQUIRED	• Prolonged bleeding time and normal platelet count, possibly increased partial thromboplastin time (PTT) • Either quantitative or qualitative deficiency in VWf leading to factor VIII dysfunction • Main function is facilitation of adhesion of platelets to subendothelial collagen in haemostasis • Increases the half life of factor VIII from 2.5 to 12 hours	