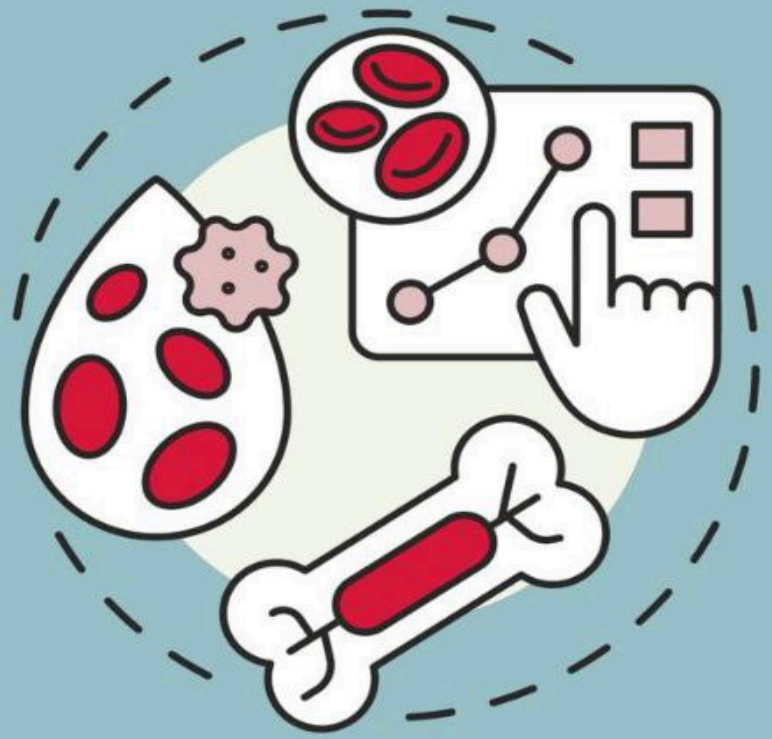
Edited past paper



By Farah Yousef

Hematology and oncology Test bank

Collected by :Ibrahim Elhaj,
Doctor (20 and 18)

1)A 17 year old male presents to emergency department complaining of headache, dizziness, and red urine that started **1 day after eating fava beans**. He looks jaundiced and sweaty. A diagnosis of glucose-6-phosphate dehydrogenase (G6PD) deficiency is suspected .Which of the following statements about this condition is **FALSE:**

a. In addition to fava beans, haemolysis can be triggered by certain medications which should be avoided.
b. Plasma exchange is the treatment of choice in adults with severe cases
c. Presence of bite cells on blood film is characteristic
d. This condition makes red blood cells susceptible to oxidative stress resulting in acute episodes of hemolysis
e. The disease is caused by point mutations in G6PD gene on X-chromosome

ANSWER:B

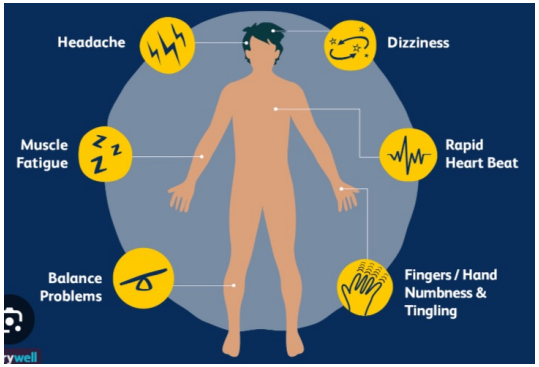
Therapy

- Avoid precipitating factors.
- Blood transfusion in severe hemolysis.
- Maintenance of good urine output during hemolytic episodes
- Folic acid.
- Exchange transfusion in newborn

2)A 58 year old male is reviewed after complaining of gradual onset of odd behaviour with psychotic symptoms.He has irritability and paresthesia in hands and feet. Examination reveals an imbalanced gait and loss of vibration sensation. A diagnosis of subacute combined degeneration of the spinal cord is suspected. **The underlying diagnosis is likely:**

a. Myelodysplastic syndrome
b. Autoimmune haemolysis
c. Vitamin b12 deficiency
d. Aplastic anaemia
e. iron deficiency

ANSWER:C

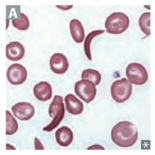


3)All of the following are associated with sickle cell disease **except**:

a. AML
b. Priapism
c. Stroke
d. Acute chest syndrome
e. Anemia

ANSWER:A

Sickle cell anemia

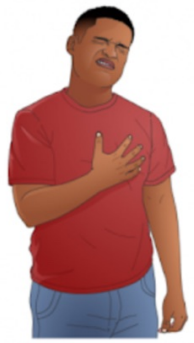


Point mutation in β-globin gene → single amino acid substitution (glutamic acid → valine) alters hydrophobic region on β-globin chain → aggregation of hemoglobin. Causes extravascular and intravascular hemolysis.
Pathogenesis: low O_2, high altitude, or acidosis precipitates sickling (deoxygenated HbS polymerizes) → vaso-occlusive disease.
Newborns are initially asymptomatic because of ↑ HbF and ↓ HbS.
Heterozygotes (sickle cell trait) have resistance to malaria.
Sickle cells are crescent-shaped RBCs A.
"Crew cut" on skull x-ray due to marrow expansion from ↑ erythropoiesis (also seen in thalassemias).

Complications:
- Aplastic crisis (transient arrest of erythropoiesis due to parvovirus B19).
- Autosplenectomy (Howell-Jolly bodies) → ↑ risk of infection by encapsulated organisms (eg, *Salmonella* osteomyelitis).
- Splenic infarct/sequestration crisis.
- Painful vaso-occlusive crises: dactylitis (painful swelling of hands/feet), priapism, acute chest syndrome (respiratory distress, new pulmonary infiltrates on CXR, common cause of death), avascular necrosis, stroke.
- Sickling in renal medulla (↓ P_{O_2}) → renal papillary necrosis → hematuria (also seen in sickle cell trait).

Hb electrophoresis: ↓↓ HbA, ↑ HbF, ↑↑ HbS.
Treatment: hydroxyurea (↑ HbF), hydration.

Complications related to sickle cell anemia include:
- Hand-foot syndrome.
- Splenic crises.
- Infections.
- Acute chest syndrome.
- Pulmonary hypertension.
- Delayed growth and puberty.
- Stroke.
- Eye problems.
- Priapism.
- Gallstones.
- Leg ulcers.
- Multiple organ failure.



This section reviews each of these complications.

4) A 60 year old male presents with dizziness and fatigue of 3 weeks duration. He also has shortness of breath upon exertion and headaches, He has constipation and has lost 5 kg over the last two months. His physical examination reveals pallor and tachycardia. Blood tests show **anemia** with **low MCV** (mean corpuscular volume) and high RDW (red cell distribution width) .Which of the following statements is **FALSE**:

a. Iron deficiency without anemia does not need treatment
b. A cause of iron deficiency should always be pursued
c. A detailed history about diet and iron intake should be carried out
d. Blood film is expected to show microcytic hypochromic red blood cells.
e. Ferritin and serum iron should be checked

ANSWER:A

IDA always need treatment

Iron deficiency	↓ iron due to chronic bleeding (eg, GI loss, heavy menstrual bleeding), malnutrition, absorption disorders, GI surgery (eg, gastrectomy), or ↑ demand (eg, pregnancy) → ↓ final step in heme synthesis. Labs: ↓ iron, ↑ TIBC, ↓ ferritin, ↑ free erythrocyte protoporphyrin, ↑ RDW, ↓ RI. Microcytosis and hypochromasia (↑ central pallor) A. Symptoms: fatigue, conjunctival pallor B, restless leg syndrome, pica (persistent craving and compulsive eating of nonfood substances), spoon nails (koilonychia). May manifest as glossitis, cheilosis, Plummer-Vinson syndrome (triad of iron deficiency anemia, esophageal webs, and dysphagia).

5) 43-year-old man presents to his physician with fatigue. The patient says he Is concerned about his fatigue because he has a strong **family history of cancer.** He thinks that if his parents did not drink, smoke, and eat such poor diets they would have lived longer lives. He says that because of all this, he never drinks alcohol or smokes tobacco. He has also followed a **strict vegan diet for 10 years**. He says that all of his meals are high in leafy green vegetables. Laboratory tests show a Hb level of 9 and MCV of 112. macrocytes appeared on a peripheral blood smear. **Most appropriate next step:**

a. Check Vitamin B 12 levels in serum
b. Check Folate level
c. Check Homocysteine level
d. Check Serum gastrin levels
e. Urine vitamin B12 level

ANSWER:A

he is a vegetarian for 10 years ,and the only source for V.B12 is meat.

6) Which of following can be used to cure transfusion-dependent beta-thalassemia:

a. splenectomy
b. blood transfusions
c. deferasirox
d. plasmapheresis
e. Bone marrow transplantation

ANSWE:E

Treatment/ Prevention of B thal major

- Blood Transfusion
- Iron chelation: deferroxamine (parenteral)
- ?splenectomy
- Allo-BMT
- Supportive
- **Prevention**

فحصيه قبل الزواج

Oral deferasirox

7) One of the following statements about glucose-6-phosphate dehydrogenase (G6PD) is **FALSE**:

a. Is an X-linked recessive disease
b. Splenectomy is the treatment of choice in case of severe attacks
c. Is the most common enzyme deficiency affecting humans.
d. In this disease, red blood cells become more susceptible to oxidative stresses
e. Most patients are asymptomatic with episodes of intravascular hemolysis and consequent anemia

ANSWER: B

G6PD deficiency	X-linked recessive. G6PD defect → ↓ NADPH → ↓ reduced glutathione → ↑ RBC susceptibility to oxidative stress (eg, sulfa drugs, antimalarials, fava beans) → hemolysis. Causes extravascular and intravascular hemolysis.	Back pain, hemoglobinuria a few days after oxidant stress. Labs: ↓ G6PD activity (may be falsely normal during acute hemolysis), blood smear shows RBCs with Heinz bodies and bite cells. "Stress makes me eat bites of fava beans with Heinz ketchup."

Therapy

- Avoid precipitating factors.
- Blood transfusion in severe hemolysis.
- Maintenance of good urine output during hemolytic episodes
- Folic acid.
- Exchange transfusion in newborn

8) Which of the following is **correct** about G6PD deficiency:

a. X-linked dominant
b. infections are a common trigger of hemolysis
c. less common in persons of African descent
d. anti-malarials reduce oxidative stress, and thus prevents hemolysis from taking place
e. Plasma exchange is the treatment of choice in adults with severe cases

ANSWER: B

G6PD deficiency	X-linked recessive. G6PD defect → ↓ NADPH → ↓ reduced glutathione → ↑ RBC susceptibility to oxidative stress (eg, sulfa drugs, antimalarials, **fava beans**) → hemolysis. Causes extravascular and intravascular hemolysis.	Back pain, hemoglobinuria a few days after oxidant **stress**. Labs: ↓ G6PD activity (may be falsely normal during acute hemolysis), blood smear shows RBCs with **Heinz** bodies and **bite** cells. "**Stress** makes me eat **bites** of **fava beans** with **Heinz** ketchup."

9)19 yr old male presented with “anemia syndrome”, fever and easy bruising. No splenomegaly and no abnormal cells on peripheral smear. Hb 6 g/dl, WBC 1500 : Neutrophils 10%, Lymphocytes 80%, others 10%. Retics© 0,001%. MCV 105fl, platelet count 20k. Bone marrow biopsy has shown hypocellular Bone marrow composed mainly of fat and stromal cells. All of the following is true **except**:

a. Recurrent infections may take place in this patient.
b. The pathogenesis of this condition revolves around bone marrow failure due to hematopoietic stem cell deficiency
c. Treatment in case of severe cases and young age in the presence of a sibling donor include bone marrow transplant.
d. most common presenting symptom of patient’s condition is bleeding.
e. It is believed that B lymphocytes are primarily causal in the bone marrow failure.

ANSWER: E (pancytopenia)

APLASTIC ANEMIA

- Aplastic anemia is a severe, life threatening syndrome in which production of erythrocytes, WBCs, and platelets has failed.
- Aplastic anemia may occur in all age groups and both genders.
- The disease is characterized by peripheral pancytopenia and accompanied by a hypocellular bone marrow.

APLASTIC ANEMIA

– The primary defect is a reduction in or depletion of hematopoietic precursor stem cells with decreased production of all cell lines
  - This may be due to quantitative or qualitative damage to the pluripotential stem cell.
  - In rare instances it is the result of abnormal hormonal stimulation of stem cell proliferation
  - or the result of a defective bone marrow microenvironment
  - or from cellular or humoral immunosuppression of hematopoiesis.

Causes of Bone Marrow Failure

Acquired	Inherited
-Idiopathic	-Fanconi anemia
-PNH	-Diamond-Blackfan Anemia
Secondary	-Other rare conditions
-Drugs	
-- radiation	
-Viruses	

10) All of the following commonly cause hemolysis in G6PD patients **except:**

a. Sulfonamides
b. erythromycin
c. Primaquine
d. Infections
e. Methylene blue

ANSWER: B

- Drugs that have been linked to G6PD
- Primaquine
- Sulphonamide antibiotics
  Sulphones e.g. dapsone used against leprosy
- Other sulphur-containing drugs: glibenclamide (an anti-diabetic drug
  Nitrofurantoin
- Vitamin K analogues
- Several others
- Henna can cause a hemolytic crisis in G6PD deficient infants

11) Which of the following is curative for hemochromatosis of chronic dialysis in beta thalassemia major

a. B.M transplant
b. Defirasorax
c. Splenectomy
d. Transfusion
e. Deferoxamine

ANSWER: A

Treatment/ Prevention of B thal major

- Blood Transfusion
- Iron chelation: deferroxamine (parenteral)
- ?splenectomy
- Allo-BMT
- Supportive
- Prevention

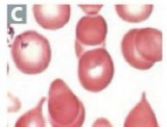
Oral deferasirox

12) a case describing pt with bone marrow findings of fibrosis and **teardrop** cells:

a. Idiopathic myelofibrosis
b. G6PD Deficiency
c. Sideroblastic Anemia
d. Hereditary Elliptocytosis
e. Lead poisoning

ANSWER:A

Dacrocytes ("teardrop cells")



Bone marrow infiltration (eg, myelofibrosis), thalassemias.

RBC "sheds a tear" because it's mechanically squeezed out of its home in the bone marrow.

13) Patient with thalasaemia was started on chelation therapy, which of the following is **true** :

a. Chelation therapy cannot be started before age of 7 years
b. Chelation therapy is given with vit c bacause it increases iron secretion
c. Before giving therapy, if organ damage was established it cannot be reversed when iron chelation therapy is started
d. Deferoxamine is first line of treatment
e. Oral deferasirox is the standard of care.

ANSWER: E

Treatment/ Prevention of B thal major

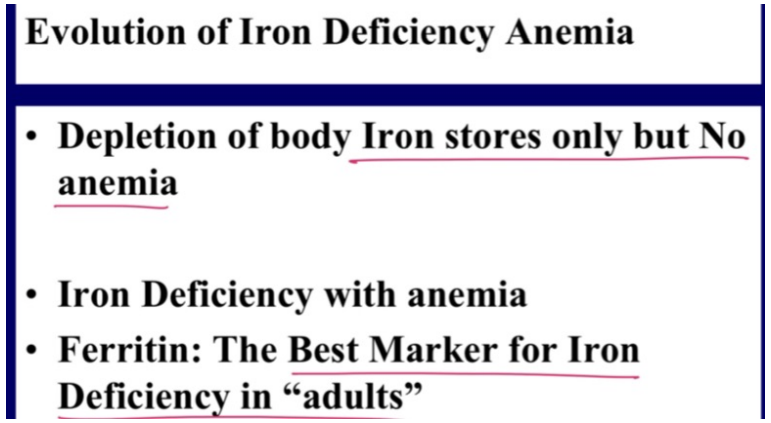
- Blood Transfusion
- Iron chelation: deferroxamine (parenteral)
- ?splenectomy
- Allo-BMT
- Supportive
- Prevention

Oral deferasirox

14) Which of the following laboratory investigations has the highest specificity and sensitivity in the diagnosis of iron deficiency anemia?

a. Serum ferritin level
b. Serum iron level
c. Serum TIBC
d. Serum MCV
e. Bone marrow biopsy

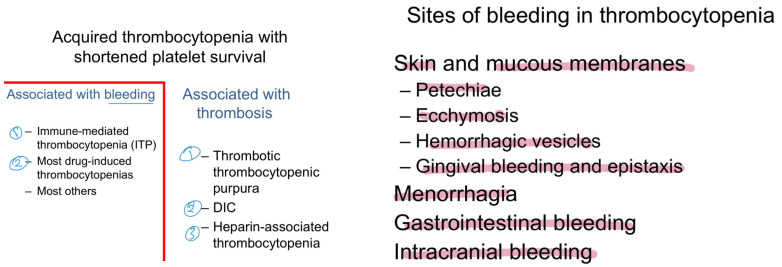
ANDWER:A



15) A 20-year-old female college student presented to the student's clinic with the new onset of bruising and epistaxis. Physical examination reveals ecchymoses on her extremities and petechiae on her ankles. Her spleen was not palpable and there was no lymph node enlargement. She has been otherwise well with no infective or B symptoms and she takes no medications. Blood tests: Hemoglobin 13 gm/dl (12-15.5), WBCs 6x109/l (4-10) , Platelets: 15 x9/l (150-450). PT: 13/13 INR 1.0. PTT 32/32. Blood film: Thrombocytopenia with large platelets. Normal red and white blood cells. LDH 375 u/l (240-480). Albumin 4.2 g/dl (3.5-5.2).
The most likely diagnosis is:

a. Immune thrombocytopenia
b. Pseudothrombocytopenia
c. Disseminated intravascular coagulation
d. Vitamin B12 deficiency
e. Thrombotic thrombocytopenia purpura

ANSWER: A



Sites of bleeding in thrombocytopenia

Skin and mucous membranes
– Petechiae
– Ecchymosis
– Hemorrhagic vesicles
– Gingival bleeding and epistaxis
Menorrhagia
Gastrointestinal bleeding
Intracranial bleeding

16) An 18 year old female was admitted with pallor, abdominal pain and long-standing history gum bleeding. She has been complaining of mucosal bleeding ever since she remembers. Her periods have always been heavy lasting more than 1 wk. She was admitted before and received bld TX for bleeding. She has summer epistaxis and bad bleeding gums. Her parents are 1st degree relatives. Her coagulation profile has shown prolonged BT, normal PT, PTT, TT and Plt, and absent clot retraction. The most likely diagnosis in this case is:

a. vWD
b. Hemophilia A
c. ITP
d. Glanzmann's thrombasthenia
e. TTP

ANSWER:D

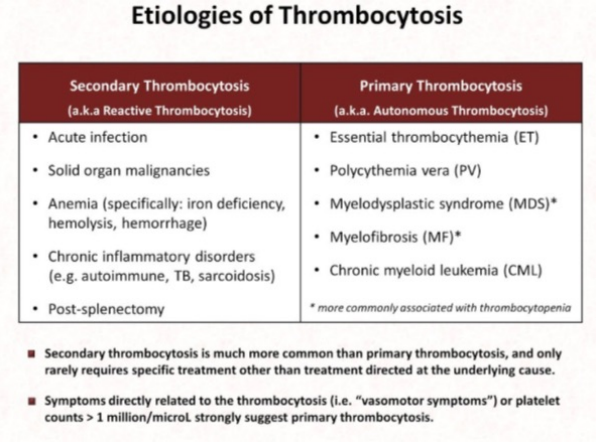
GT Laboratory/ Diagnostic tests

- Normal platelet count and morphology
- Prolonged bleeding time
- Absent or impaired clot retraction
- Absent or reduced plt fibrinogen
- No aggregation with physiological aggregating agents
- Absent or reduced GPIIb-IIIa
- Treatment is supportive

17) All of the following are associated with thrombocytosis **except**:

a. Post-splenectomy
b. Chronic stage of CML
c. POLYCYTHEMIA VERA.
d. Pernicious anemia
e. Iron deficiency

ANSWER:D



Etiologies of Thrombocytosis

Secondary Thrombocytosis (a.k.a Reactive Thrombocytosis)	Primary Thrombocytosis (a.k.a. Autonomous Thrombocytosis)
• Acute infection	• Essential thrombocythemia (ET)
• Solid organ malignancies	• Polycythemia vera (PV)
• Anemia (specifically: iron deficiency, hemolysis, hemorrhage)	• Myelodysplastic syndrome (MDS)*
• Chronic inflammatory disorders (e.g. autoimmune, TB, sarcoidosis)	• Myelofibrosis (MF)*
• Post-splenectomy	• Chronic myeloid leukemia (CML)
	** more commonly associated with thrombocytopenia*

- Secondary thrombocytosis is much more common than primary thrombocytosis, and only rarely requires specific treatment other than treatment directed at the underlying cause.
- Symptoms directly related to the thrombocytosis (i.e. "vasomotor symptoms") or platelet counts > 1 million/microL strongly suggest primary thrombocytosis.

18) One of the following bleeding patterns is a feature of coagulatory factor defect rather than a platelet defect?

a. Gum bleeding
b. Small superficial bruises
c. Bleeding into Joints
d. Epistaxis
e. Petechial rash

ANSWER: C

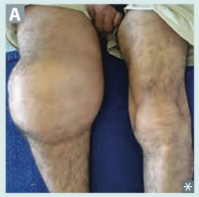
Clinical Features of Bleeding Disorders

	Platelet disorders	Coagulation disorders
Site of bleeding tissues	Skin (epistaxis, gum, vaginal, GI tract)	Deep in soft Mucous membranes, joints, muscles)
Petechiae	Yes	No
Ecchymoses ("bruises")	Small, superficial	Large, deep
Hemarthrosis / muscle bleeding	Extremely rare	Common
Bleeding after cuts & scratches	Yes	No
Bleeding after surgery or trauma	Immediate, usually mild	Delayed (1-2 often severe

19)A 19 year old male is seen in clinic with repeated attacks of large joint painful swelling especially in his knees for several years. His maternal uncle has similar condition. Examination reveals swollen hot right knee with effusion and limitation of movement. Blood tests: Prothrombin time (PT): 14/14 sec. Partial thromboplastin time (PTT): 80/31 seconds. Mixing study of PTT: 32/31 platelets 268 x109/l FactorVII: 41%.**The most likely diagnosis is:**

a. Glanzmann's thrombasthenia
b. Haemophilia B
c. Haemophilia A
d. Von Willebrand disease (VWD)
e. Disseminated intravascular coagulation (DIC)

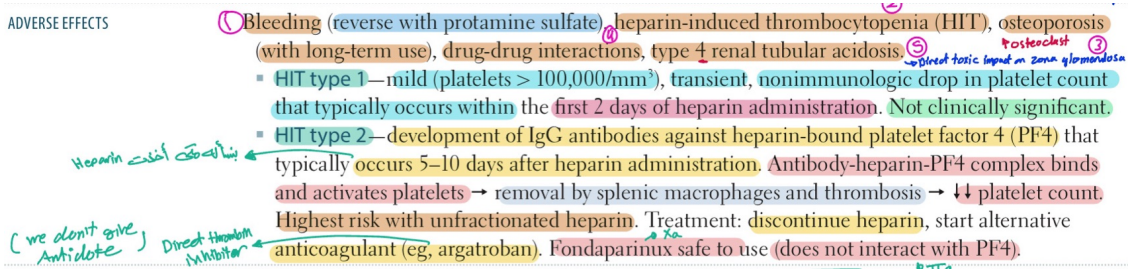
ANSWER:C

DISORDER	PT	PTT	MECHANISM AND COMMENTS
Hemophilia A, B, or C 	—	↑	Intrinsic pathway coagulation defect (↑ PTT). ▪ A: deficiency of factor VIII; X-linked recessive. Pronounce "hemophilia Ate (eight)." ▪ B: deficiency of factor IX; X-linked recessive. ▪ C: deficiency of factor XI; autosomal recessive. Hemorrhage in hemophilia—hemarthroses (bleeding into joints, eg, knee A), easy bruising, bleeding after trauma or surgery (eg, dental procedures). Treatment: desmopressin, factor VIII concentrate, emicizumab (A); factor IX concentrate (B); factor XI concentrate (C).
Vitamin K deficiency	↑	↑	General coagulation defect. Bleeding time normal. ↓ activity of factors II, VII, IX, X, protein C, protein S.

20) What is the most appropriate step in the management of a patient with heparin-induced thrombocytopenia and thrombosis?

A. Continue heparin and administer warfarin
B. Discontinue heparin and administer argatroban
C. Discontinue the heparin substitute with warfarin
D. Continue heparin and add lepirudin
E. Continue heparin and monitor closely

ANSWER:B



ADVERSE EFFECTS

Bleeding (reverse with protamine sulfate), heparin-induced thrombocytopenia (HIT), osteoporosis (with long-term use), drug-drug interactions, type 4 renal tubular acidosis.

- HIT type 1—mild (platelets > 100,000/mm³), transient, nonimmunologic drop in platelet count that typically occurs within the first 2 days of heparin administration. Not clinically significant.
- HIT type 2—development of IgG antibodies against heparin-bound platelet factor 4 (PF4) that typically occurs 5–10 days after heparin administration. Antibody-heparin-PF4 complex binds and activates platelets → removal by splenic macrophages and thrombosis → ↓↓ platelet count. Highest risk with unfractionated heparin. Treatment: discontinue heparin, start alternative anticoagulant (eg, argatroban). Fondaparinux safe to use (does not interact with PF4).

21) All of the following help to distinguish qualitative platelet disorder **except**:

a. platelet size
b. clot retraction
c. platelet aggragation
d. bone marrow cytogenatics
e. platelet flow cytometry

ANSWER:A

22)23-year-old woman with systemic lupus erythematosus diagnosed 2 years ago presents to the clinic because of tenderness in her left calf that Is worse with flexion of the foot. After an initial work-up, her rheumatologist suspects that she may have antiphospholipid antibody (APA) syndrome. Which of the following laboratory findings is **most likely in this patient?**

a. Decreased partial thromboplastin time, corrected by mixing with fresh-frozen plasma
b. Decreased partial thromboplastin time, not corrected by mixing with fresh-frozen plasma
c. Prolonged partial thromboplastin time, corrected by mixing with fresh-frozen plasma
d. Prolonged partial thromboplastin time, not corrected by mixing with fresh-frozen plasma
e. Decreased Prothrombin Time, corrected by mixing with fresh-frozen plasma

ANSWER:D

Antiphospholipid antibodies (aPL antibodies) [1][4]

Reserve testing for patients with characteristic clinical features to avoid false positives.

- **Lupus anticoagulant (LA)**: antibodies against certain phospholipids in cellular membranes; detection involves a three-step procedure
  1. Screening for phospholipid-dependent coagulation with either:
     - Prolonged aPTT
     - Prolonged dilute Russell viper venom time (dRVVT)
  2. Mixing study: The patient's plasma is mixed with normal plasma (which contains clotting factors).
     - aPTT or dRVVT normalize: Presence of lupus anticoagulant ruled out; prolonged aPTT may be due to a lack of clotting factors.
     - aPTT or dRVVT remain prolonged: Lupus anticoagulant may be present.
  3. Confirmation of phospholipid dependence: Phospholipid is added.
     - aPTT or dRVVT normalize: Presence of lupus anticoagulants is confirmed.
     - aPTT or dRVVT remain prolonged: Consider a factor deficiency.
- **Anticardiolipin antibodies (IgG and IgM)**: antibodies against cardiolipin, a phospholipid in cellular membranes
- **Anti-β2-glycoprotein antibodies (IgG and IgM)**: antibodies directed against the cardiolipin binding factor β2 glycoprotein I that have prothrombotic effects [2]

23)A patient with gingival bleeding and increased bleeding time and **normal PT** and PTT and normal platelet count, parents are cousins, Based on the scenario above, the diagnosis is :

a. Glanzmann's thrombasthenia
b. VWD
c. Hemophilia
d. ITP
e. TTP

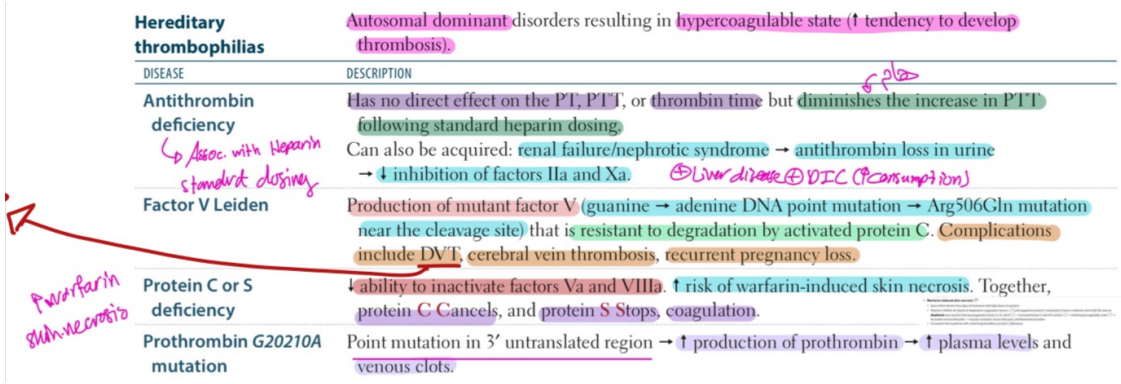
ANSWER:A

DISORDER	PC	BT	NOTES
Bernard-Soulier syndrome	–/↓	↑	Autosomal recessive defect in adhesion. ↓ GpIb → ↓ platelet-to-vWF adhesion. Labs: ↓ platelet aggregation, Big platelets.
Glanzmann thrombasthenia	–	↑	Autosomal recessive defect in aggregation. ↓ GpIIb/IIIa (↓ integrin $\alpha_{IIb}\beta_3$) → ↓ platelet-to-platelet aggregation and defective platelet plug formation. Labs: blood smear shows no platelet clumping.

24) Upon arriving home following a 14-hour flight from Japan, a 58-year-old white woman notices significant swelling and pain in her right calf. She arrives at her apartment exhausted and falls asleep. In the middle of the night she wakes up severely short of breath.
This woman most likely has what disorder?

a. Antithrombin III defidency
b. Factor V leiden
c. Protein C deficiency
d. Protein S deficiency
e. Prothrombin gene mutation

ANSWER:B

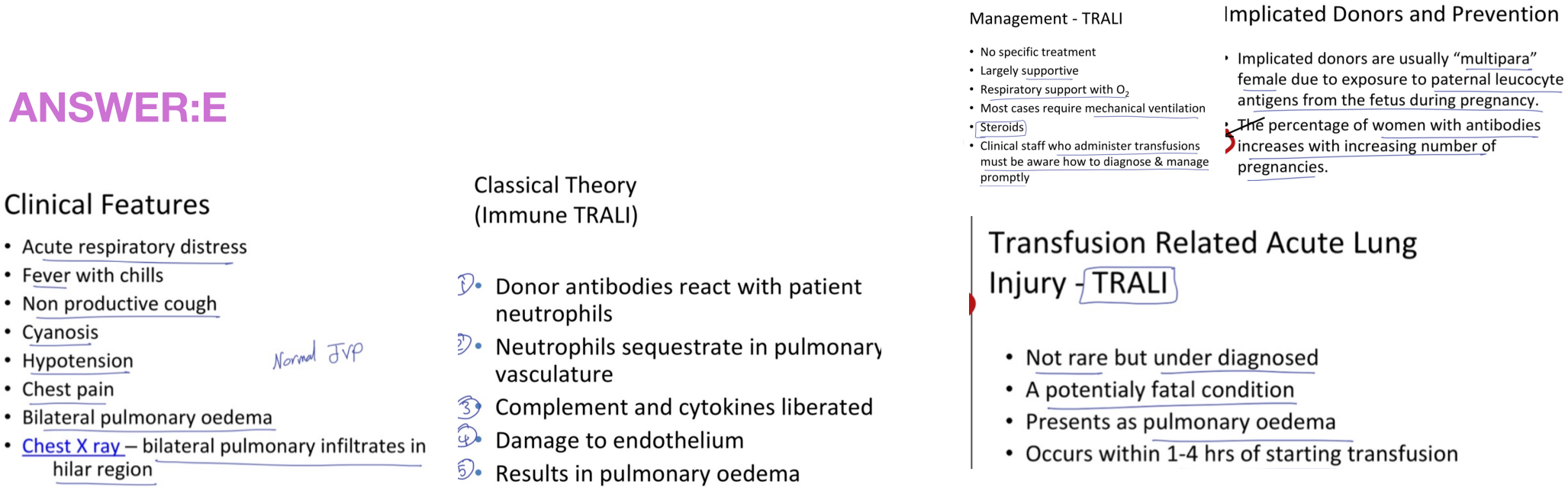


Hereditary thrombophilias	Autosomal dominant disorders resulting in hypercoagulable state (↑ tendency to develop thrombosis).
DISEASE	DESCRIPTION
Antithrombin deficiency	Has no direct effect on the PT, PTT, or thrombin time but diminishes the increase in PTT following standard heparin dosing. Can also be acquired: renal failure/nephrotic syndrome → antithrombin loss in urine → ↓ inhibition of factors IIa and Xa.
Factor V Leiden	Production of mutant factor V (guanine → adenine DNA point mutation → Arg506Gln mutation near the cleavage site) that is resistant to degradation by activated protein C. Complications include DVT, cerebral vein thrombosis, recurrent pregnancy loss.
Protein C or S deficiency	↓ ability to inactivate factors Va and VIIIa. ↑ risk of warfarin-induced skin necrosis. Together, protein C Cancels, and protein S Stops, coagulation.
Prothrombin *G20210A* mutation	Point mutation in 3′ untranslated region → ↑ production of prothrombin → ↑ plasma levels and venous clots.

25) All of the following are true about Transfusion Related Acute Lung Injury, except:

a. takes place within 6 hours of transfusion
b. Implicated donors are usually “multipara” female
c. donor anti-leukocyte antibodies are the cause of this transfusion reaction
d. causes noncardiogenic pulmonary edema with bilateral pulmonary infiltrates
e. a minority of cases require mechanical ventilation

ANSWER:E



26) female told that she has heterozygous factor V leiden ... what would you tell her

a. start of LMWH for lifelong
b. start on heparin and warfarin then stop heparin
c. do nothing
d. do lower limb doppler and treat accordingly
e. Order a CT Scan of the chest

ANSWER:C

How is heterozygous Factor V Leiden treated?

How is factor V Leiden (FVL) treated? The factor V Leiden mutation itself doesn't have any specific treatment. But when a person is diagnosed with an acute deep vein thrombosis (DVT) or pulmonary embolism (PE), treatment with anticoagulants (blood thinners) will be necessary and should be started as soon as possible. ٢٠٢٢/١٠/٠٤

27) One of the following is a mutation thrombophilia that Causes VTE:

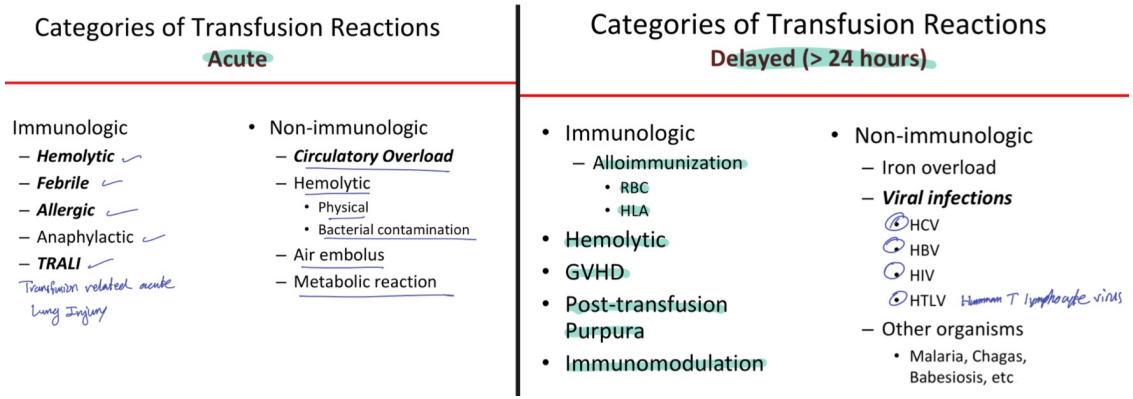
a. Protein C def
b. Protein A def
c. Anticardiolipin
d. Lupus anticoagulant
e. Factor V leiden

ANSWER: E

28) not an acute complication for blood transfusion:

a. Rh hemolysis
b. Anaphylactic transfusion reaction
c. TRALI
d. Metabolic reaction
e. Circulatory overload

ANSWER:A



Categories of Transfusion Reactions

Acute

Immunologic
- ***Hemolytic***
- ***Febrile***
- ***Allergic***
- Anaphylactic
- ***TRALI***

Transfusion related acute Lung Injury

- Non-immunologic
  - ***Circulatory Overload***
  - Hemolytic
    - Physical
    - Bacterial contamination
  - Air embolus
  - Metabolic reaction

Categories of Transfusion Reactions

Delayed (> 24 hours)

- Immunologic
  - Alloimmunization
    - RBC
    - HLA
- Hemolytic
- GVHD
- Post-transfusion Purpura
- Immunomodulation

- Non-immunologic
  - Iron overload
  - ***Viral infections***
    - HCV
    - HBV
    - HIV
    - HTLV Human T lymphocyte virus
  - Other organisms
    - Malaria, Chagas, Babesiosis, etc

29) Blood transfusion risks include all of the following except:

a. Iron overload
b. Secondary hemochromatosis
c. hypokalemia
d. Acute hemolytic transfusion reaction
e. Post-transfusion purpura

ANSWER:C

The risks of blood transfusions include:

- An allergic reaction. This can be mild or severe. ...
- Fever. ...
- Destruction of red blood cells by the body (hemolytic reaction). .
- Too much blood in the body (transfusion overload). ...
- Too much iron in the body (iron overload). ...
- Viruses being transmitted. ...
- Graft-versus-host disease.

30) which of the following is the definitive test to distinguish hemophilia A from B:

a. PTT
b. factor assay
c. Bleeding test
d. X linked
e. PT

ANSWER:B

Factor Assays. Factor assays may be part of the testing process to determine the cause of an unexpected, prolonged PT or aPTT. This test is performed after mixing studies have been performed and a deficiency of one or more coagulation factors is suspected.

Haemophilia occurs in two sub-types: haemophilia A and haemophilia B. In haemophilia A, there is a lack or total absence of coagulation factor VIII. In haemophilia B, there is a serious shortage or total absence of coagulation factor IX.

31)37 yr old lady was admitted with high fever, seizure and confusion for 3 days.P/E shown. Temp 40.5,BP 80/50, Pulse: 122 regular, low volume. Bleeding from needle puncture sites and bruising. Hb 9g/dl, retcs 6%, bilirubin 5 (d1), WBC 19k, Plt 25k, LDH 1400, PT 14/12s, PTT 35/32s, TT 13/11s, Creatinine 2.3. Bld film shown.Fibrinogen. 140mg/dl. All of the following regarding this case is correct except :

a. If untreated, mortality may exceed 90%
b. Initial Treatment involves plasma exchange daily until recovery
c. On MRI, leukoencephalopathy and brain infarcts may be detected
d. VWF cleaving protease levels are severely reduced
e. On blood film, spherocytes are expected to be seen.

ANSWER:E

Thrombotic microangiopathies	Disorders overlap significantly in symptomatology. May resemble DIC, but do not exhibit lab findings of a consumptive coagulopathy (eg, ↑ PT, ↑ PTT, ↓ fibrinogen), as etiology does not involve widespread clotting factor activation.	
TTP	**Thrombotic thrombocytopenic purpura**	**Hemolytic-uremic syndrome**
EPIDEMIOLOGY	Typically females	Typically children
PATHOPHYSIOLOGY	Inhibition or deficiency of ADAMTS13 (a vWF metalloprotease) → ↓ degradation of vWF multimers → ↑ large vWF multimers → ↑ platelet adhesion and aggregation (microthrombi formation)	Predominately caused by Shiga toxin–producing *Escherichia coli* (STEC) infection (serotype O157:H7), which causes profound endothelial dysfunction.
PRESENTATION	Triad of thrombocytopenia (↓ platelets), microangiopathic hemolytic anemia (↓ Hb, schistocytes, ↑ LDH), acute kidney injury (↑ Cr)	
DIFFERENTIATING SYMPTOMS	Triad + fever + neurologic symptoms	Triad + bloody diarrhea
LABS	Normal PT and PTT helps distinguish TTP and HUS (coagulation pathway is not activated) from DIC (coagulation pathway is activated)	
TREATMENT	Plasma exchange, glucocorticoids, rituximab	Supportive care

32) Wrong about glanzman thrombasthenia:

a. normal platelet count
b. There's a defect in platelet plug formation.
c. abnormal clot retraction test
d. del ch (17)
e. abnormal flowcytometry due to lack of expression of CD19 + CD38

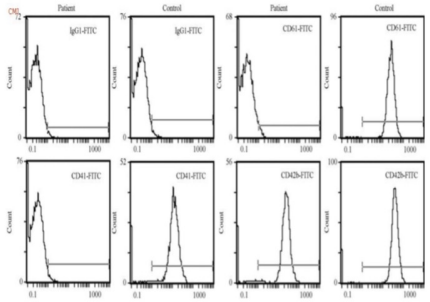
ANSWER: E

GT Laboratory/ Diagnostic tests

- Normal platelet count and morphology
- Prolonged bleeding time
- Absent or impaired clot retraction
- Absent or reduced plt fibrinogen
- No aggregation with physiological aggregating agents
- Absent or reduced GPIIb-IIIa
- Treatment is supportive

Absent CD41 61

Flowcytometry of platlets with GT



33)A patient presented to the ER with a sudden onset of dyspnea and unilateral calf swelling. On exam, respiratory and heart rates were increased. Which of the following would **least** likely be present in this patient:

a. acute respiratory acidosis
b. pitting edema
c. pleuritic chest pain
d. Normal chest X-ray.
e. increased pulmonic sound (P2)

ANSWER:A

- **Common features of PE** [5]
  - Acute onset of symptoms
  - **Dyspnea** (> 75% of cases) [5]
  - Tachycardia and **tachypnea** (up to 50% of cases) [5]
  - **Sudden pleuritic chest pain** (~ 20% of cases) [5]
  - Cough and **hemoptysis**
  - Associated features of DVT: e.g., unilaterally painful leg swelling [6]
- **Less common features of PE** [5]
  - Decreased breath sounds
  - Dullness to percussion
  - Split S2
  - Low-grade fever
  - Rarely, upper abdominal pain [7][8]
- **Features of massive PE**: (e.g., due to a saddle thrombus)
  - Presyncope or syncope
  - Jugular venous distension and Kussmaul sign
  - Hypotension and obstructive shock
  - Circulatory collapse
- **Features of intraoperative PE**: all features typically have an abrupt onset [9][10]
  - Decrease in $EtCO_2$ (early sign)
  - Decrease in SpO_2
  - Hypotension
  - Tachycardia

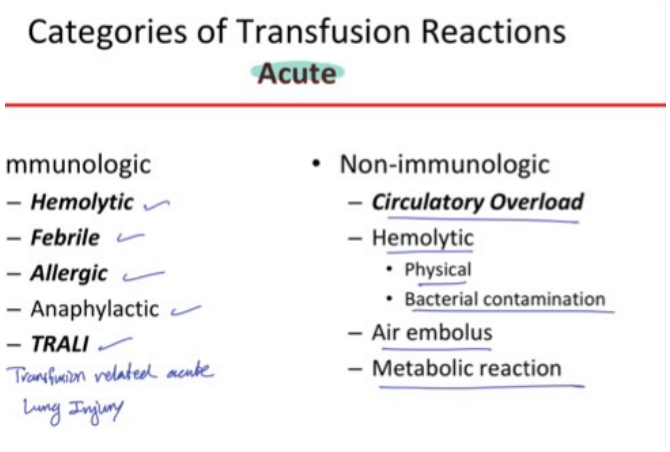
Why does pulmonary embolism cause respiratory alkalosis?

Thus, most patients with PE present with a lower than normal arterial PCO2 and respiratory alkalosis because of an increased total minute ventilation. Limited data suggest that the increased total minute ventilation occurs because of reflex stimulation of irritant and juxta capillary sensors in the lung. Dec 2, 2003

34) All of the following are **acute** complications of blood transfusion **except**:

a. iron overload
b. TRALI
c. WBC reaction
d. ABO incombitability hemolysis
e. Anaphylactic transfusion

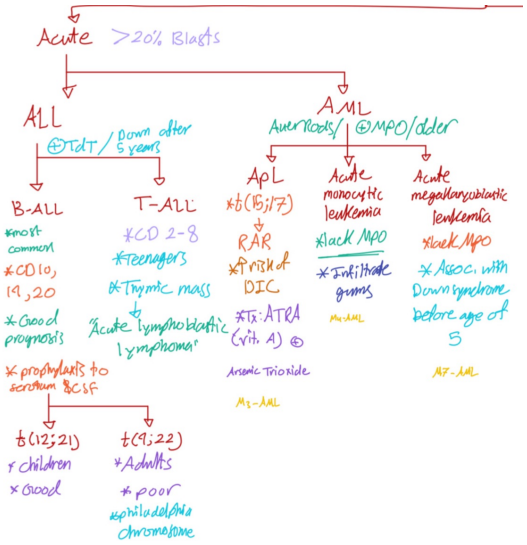
ANSWER:A



35)Which of the following goes more with ALL than AML?

a. TdT and CD10
b. Auer rods
c. a subtype of ALL is more commonly associated with DIC
d. t(15;17)
e. ATRA syndrome

ANSWER:A



36) All of the following are features of Symptomatic myeloma distinguishing it from smoldering myeloma except:

a. bone lytic lesion
b. recurrent bacterial infections
c. renal insufficiency
d. bone marrow plasma cells 15%
e. normocytic anemia

ANSWER:D

Clinical features

common tetrad of multiple myeloma is ***CRAB***

- **C = Calcium (elevated)**
- **R = Renal failure**
- **A = Anemia**
- **B = Bone lesions**

Related organ or tissue impairment

B – Lytic bone lesions – visible on x-ray in 85% of patients. **Hint** – Osteoclasts activated, not osteoblasts.
C – Hypercalcemia (Ca > 11 mg/dL)
A – Anemia (Hb < 10)
V – Hyperviscosity – especially common in the rare IgM secreting myeloma
I – Bacterial infections (>2)
A – Amyloidosis
R – Renal (Crt > 1.96 mg/dL): **HINT** – occurs 50% of the time because most often the light chains are toxic to the tubules.

These are the end organ manifestations of myeloma

Active Multiple Myeloma
Both criteria must be met:
1- Clonal bone marrow plasma cells 10% or biopsy-proven bony or extramedullary plasmacytoma
2- Any one or more of the CRAB features

Smouldering MM
Both criteria must be met:
Serum monoclonal protein (IgG or IgA) 3 g/dL, or urinary monoclonal protein 500 mg per 24 h and/or clonal bone marrow plasma cells 10%–60%
Absence of myeloma defining events or amyloidosis Mu

37) Patient with multiple myeloma, what makes him stage 3?

a. β2 microglobulin above 5.5 mg/dl
b. serum albumin > 3.5 g/dl
c. serum β2 microglobulin between 3.5-5.5 mg/dl
d. BM biopsy revealing 8% abnormal plasma cells
e. Elevated monoclonal immunoglobulin spike

ANSWER:A

Prognostic Factors

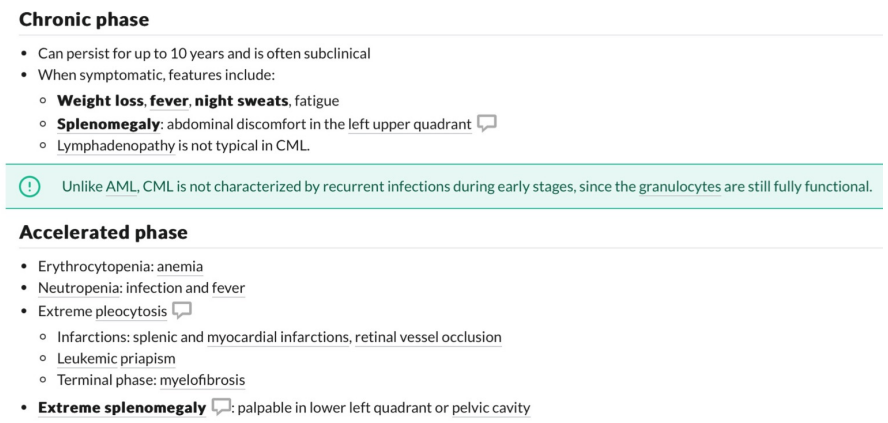
International staging system
I (good prognosis)
Serum albumin > 3.5 g/dl
Serum β2 microglobulin < 3.5 mg/dl
II
Not I or III
III
β2 microglobulin: >5.5 mg/dl

albumin < 3.5
β_2 -microglobulin: 3.5-5.5

38)A 63-year-old woman has a 6-month history of increasing fatigue, weakness, and anorexia. Physical examination reveals splenomegaly and pale conjunctiva, and laboratory studies show leukocytosis and anemia. Karyotyping of suspicious cells reveals a t(9;22) translocation Which of the following is the most likely diagnosis ?

a. CML
b. CLL
c. ALL
d. Multiple myeloma
e. Hodgkin lymphoma

ANSWER:A



Chronic phase

- Can persist for up to 10 years and is often subclinical
- When symptomatic, features include:
  - **Weight loss**, **fever**, **night sweats**, fatigue
  - **Splenomegaly**: abdominal discomfort in the left upper quadrant
  - Lymphadenopathy is not typical in CML.

Unlike AML, CML is not characterized by recurrent infections during early stages, since the granulocytes are still fully functional.

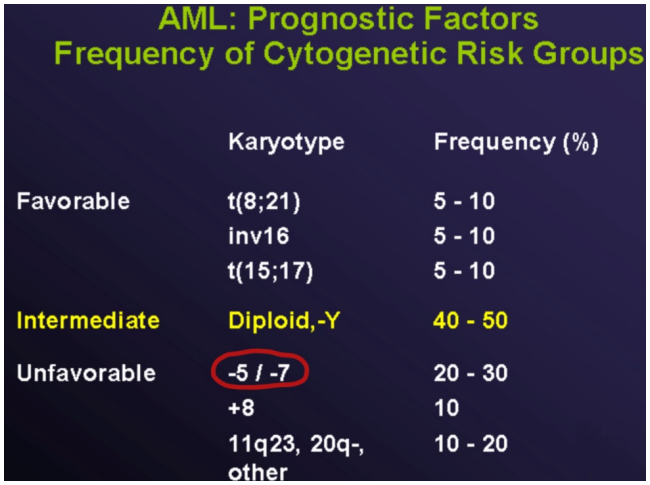
Accelerated phase

- Erythrocytopenia: anemia
- Neutropenia: infection and fever
- Extreme pleocytosis
  - Infarctions: splenic and myocardial infarctions, retinal vessel occlusion
  - Leukemic priapism
  - Terminal phase: myelofibrosis
- **Extreme splenomegaly**: palpable in lower left quadrant or pelvic cavity

39)Poor prognosis in AML?

a. NMP1 mutaion
b. t (15;17)
c. inv(16)
d. t (8;21)
e. chromosome 7 deletion

ANSWER:E



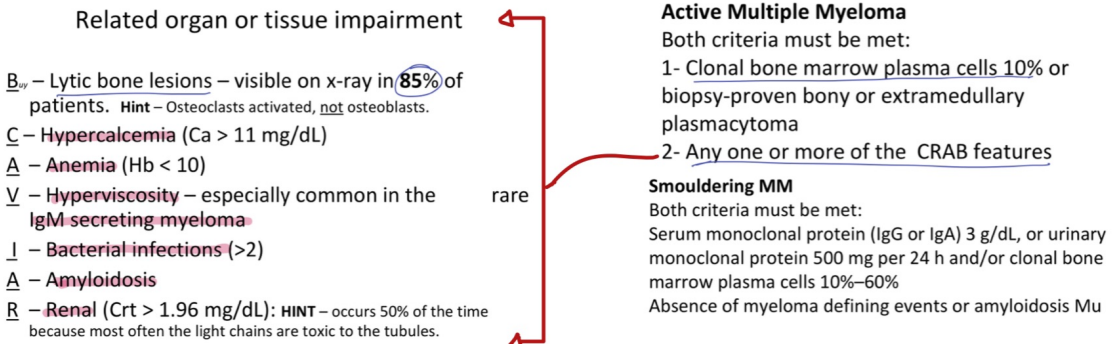
AML: Prognostic Factors
Frequency of Cytogenetic Risk Groups

	Karyotype	Frequency (%)
Favorable	t(8;21)	5 - 10
	inv16	5 - 10
	t(15;17)	5 - 10
Intermediate	Diploid,-Y	40 - 50
Unfavorable	-5 / -7	20 - 30
	+8	10
	11q23, 20q-, other	10 - 20

40) One of the following findings in a patient with multiple myeloma is **NOT** a distinctive feature of symptomatic disease (related organ or tissue damage):

a. Serum calcium of 12 mg/dl (8.6-10.2)
b. Creatinine of 2.4 mg/dl (0.6-1.2)
c. Haemoglobin of 9 gm/dl (13.5-16.5)
d. Multiple lytic bony lesions
e. Bone marrow plasma cells of 15%

ANSWER:E



41)A 72 year old male is seen with a new diagnosis of Chronic Lymphocytic Leukaemia. Diagnosis was made by flow cytometry of peripheral blood and after reviewing his full blood count. Which of the following is NOT an expected finding?

a. Lymphadenopathy
b. Bone lytic lesions
c. Reduced immunoglobulins
d. An absolute lymphocytic count of more than 5x 109/1
e. Splenomegaly

ANSWER: B

CLL Clinical Presentation

- Lymphocytosis
  - Morphologically mature
  - Immunologically immature
  - Accumulation in PB, BM and lymphatic tissues
- Enlarged Lymph nodes
- Splenomegaly
- Hypogammaglobulinaemia

Laboratory studies [2][6][5][1]

Obtain a CBC, peripheral blood smear, and flow cytometry for all patients.

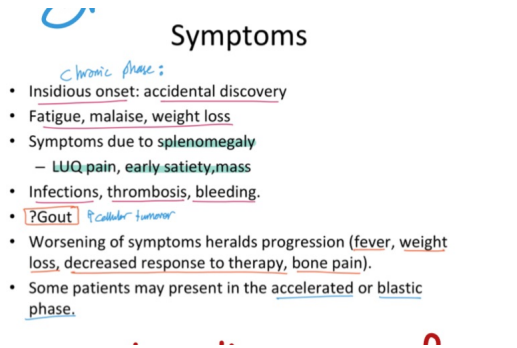
- **Diagnostic criteria**
  - Persistent (≥ 3 months) **lymphocytosis** (≥ 5000 cells/mm^3) [6][1]
  - CLL immunophenotype confirmed by flow cytometry, including:
    - Expression of **CD5**, **CD19**, CD20, CD23 [2]
    - Light chain restriction (kappa or lambda)
- **Additional findings**: may further support the diagnosis
  - Cytopenias
    - Anemia (usually normochromic, normocytic): may be caused by AIHA [5][7]
    - Thrombocytopenia
    - Granulocytopenia
  - Peripheral blood smear may show:
    - High percentage of **small mature lymphocytes**
    - **Smudge cells (Gumprecht shadows)**: mature lymphocytes that rupture easily; appear as artifacts on a blood smear [8]

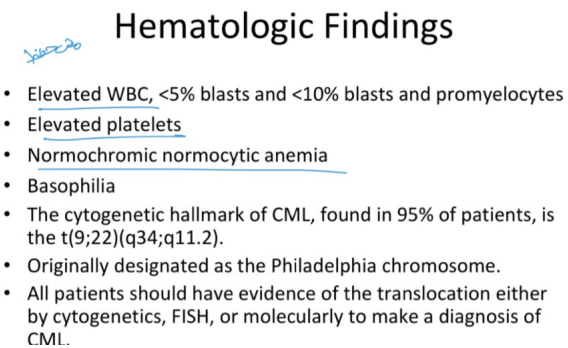
42)A 62-year-old male with a white blood count of 120 x109/l is seen in the clinic. He has been feeling weak and excessively fatigues recently. He has no infective or bleeding symptoms and looks well overall while being interviewed in the clinic. Physical exam is notable for splenomegaly that is felt 3 cm below costal margin. Blood tests: Hemoglobin 14 gm/dl(12-15.5), WBCs 120x109/1 (4-10) , Platelets: 800 x9/1 (150-450). Blood film: Leukocytosis with granulocytes showing different stages of maturation (from myeloblasts to neutrophils). Basophilia is noted. The most appropriate test to send to establish the correct diagnosis is:

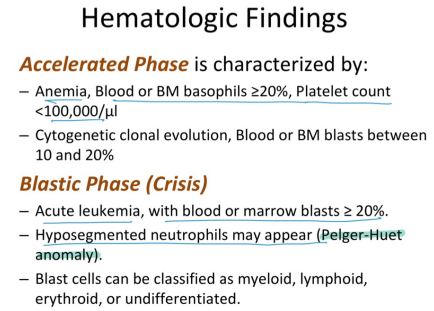
a. Protein electrophoresis
b. Cytogenetics for Philadelphia chromosome: t(9;22)
c. Bone marrow aspiration and biopsy
d. Spleen biopsy
e.Flowcytometry (immunophenotyping)

ANSWER:B

The diagnosis is CML







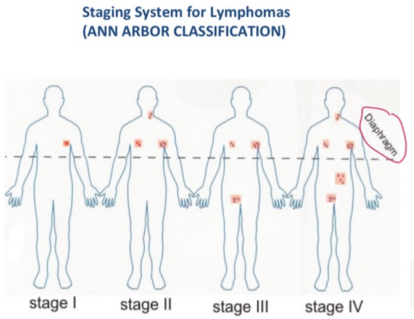
43)A patient diagnosed with Hodgkin lymphoma after further investigations was found to have LN involvement of cervical and inguinal groups, with no involvement of the spleen, liver, bone marrow or mediastinum. The patient complains of sweating, weight loss and fever. According to Ann Arbor staging system, the patient is in stage:

a. IIA
b. IIIA
c. IIIIA
d. IIB
e. IIIB

ANSWER:E

Staging: Ann Arbor

I. 1 lymph node region or structure
II. >1 lymph node region or structure, same side of diaphragm
III. Both sides of diaphragm
IV. Extranodal sites diffuse, beyond "E" designation



Stage I	Involves 1 ganglionic chain (I) or localized involvement of 1 extralymphatic organ (IE)
Stage II	Involves 2 or more ganglionic chains ipsilateral to the diaphragm (II) or involves 1 extralymphatic site and 1 or more ganglionic chains ipsilateral to the diaphragm (IIE)
Stage III	Involves various ganglionic chains on both sides of the diaphragm (III), possibly associated with extralymphatic sites (IIIE), the spleen (IIIS), or both (IIIES)
Stage IV	Involves various extralymphatic sites with or without ganglionic involvement or 1 isolated site associated with distal ganglions
Type A	Without systemic symptoms
Type B	With systemic symptoms

44)all of the following are indicated to stage a patient with NHL **except**:

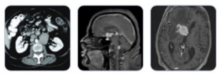
a. brain CT
b. bone marrow biopsy
c. chest, abd, pelvic CT
d. LDH
e. bone scan

ANSWER: D

Staging and classification

Staging [15]

- **Imaging**
  - Indicated in all patients for staging and to assess response to therapy
  - Choice of imaging modality depends on the suspected subtype of NHL (uptake of FDG varies between subtypes)
    - FDG-avid NHLs (most subtypes): PET-CT
    - Non-FDG-avid NHLs: CT whole body with contrast
- **Bone marrow aspiration and biopsy**: indicated in most newly diagnosed patients with NHL
- **Assessment of CNS involvement**
  - Indications
    - Patients considered high-risk for CNS involvement [21]
    - Primary CNS lymphoma
    - Patients with neurological signs and symptoms
    - Patients with HIV
  - Recommended modalities include:
    - Imaging (MRI or CT brain)
    - Lumbar puncture with CSF assessment (cytology; detection of **EBV** DNA)



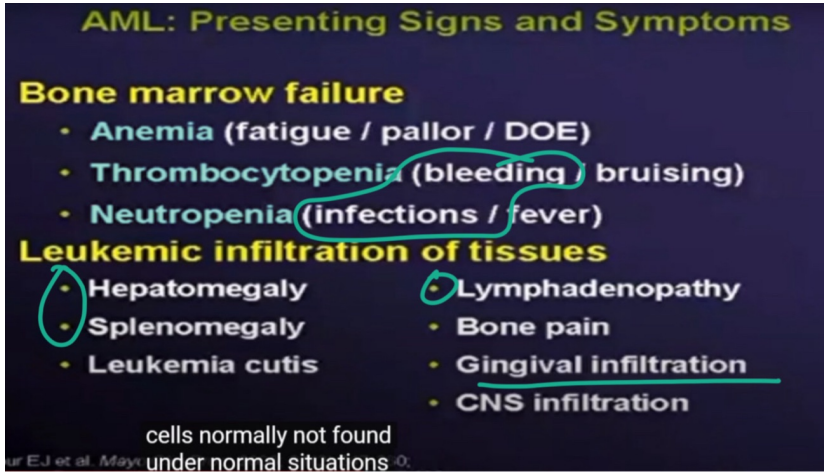
Prognostic factors in non-Hodgkin's lymphoma

- ***Adverse factors:***
- ***Age > 60 years***
- ***Stage III or IV, i.e. advanced disease***
- ***High serum lactate dehydrogenase level***
- ***Performance status (ECOG 2 or more)***
- ***More than one extranodal site involved***

45) One of the following is NOT an expected finding in the marrow of a patient with newly diagnosed AML:

a. Decreased erythropoiasis
b. Hyper-cellular marrow
c. 60% blast cells
d. Auer rod cells
e. 15% plasma cell

ANSWER:E



46) A 22-year-old college student presents to the clinic complaining of fever, fatigue, and sore throat that have not improved for the last 2 weeks. Physical examination reveals painful lymphadenopathy. Further investigations were done and patient was found to have EBV infection. Most commonly, the route of transmission of EBV in this case is:

a. Feco-oral (ingestion)
b. Aerosol
c. vector borne
d. close contact (Oral secretions)
e. Blood transfusion

ANSWER:D

- **Pathogen**: **Epstein-Barr virus (EBV)**, also called human herpesvirus 4 (HHV-4)
- **Transmission**: Infectious mononucleosis spreads via bodily secretions, especially saliva. Therefore, it is also called **kissing disease**.

47)which of the following is associated with an increase in RBCs, WBCs and platelets:

a. Polycythemia Rubra Vera
b. Essential thrombocythmia
c. Myelofibrosis
d. CML
e. Glanzmann Thrombasthenia

ANSWER:A

Polycythemia vera (PV)

1. What is PV?
   - Increase in RBC, WBC and platelets but main presentation is based on too much RBC.

	RBCs	WBCs	PLATELETS	PHILADELPHIA CHROMOSOME	*JAK2* MUTATIONS
Polycythemia vera	↑	↑	↑	⊖	⊕
Essential thrombocytosis	–	–	↑	⊖	⊕ (30–50%)
Myelofibrosis	↓	Variable	Variable	⊖	⊕ (30–50%)
CML	↓	↑	↑	⊕	⊖

48) A 25 year old male patient was reffered with the diagnosis of acute myeloid leukemia. He was found to have Hb 8gm/dl, WBC 80000/ul, platelets count 17000/ul. Blood film showed 80% blasts with bilobed appearance. BM was heavily infiltrated by blasts with abundant cytoplasmic granules and aur rods. Granules were positive for myeloperoxidase, but negative for butyrate esterase. Blasts were CD33 and CD13 positive. Cytogenetics studies were done. Which of the following cytogenetic abnormalities is **most likely in this patient:**

a. T(15;17}
b. T(8;16)
c. T(8:21)
d. T(1;22)
e. T(9;22)

ANSWER:A

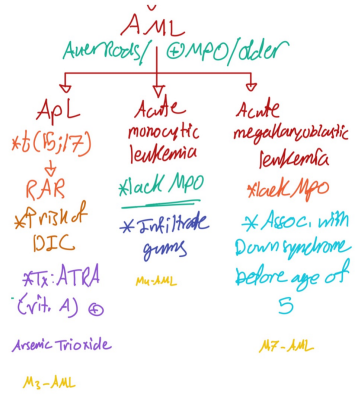
Promyelocytic leukemia M3

1. Associated with t (15;17) involving the retinoic acid receptor (RAR) gene .
2. Good prognosis category.
3. Commonly associated with **DIC.**

- Prominent Auer rods. pathognomic for AML

Treatment aml/m3

- **Tretinoin (all tran retinoic acid)** is an oral drug that induces the differentiation of leukemic cells bearing the **t(15;17)**; it is not effective in other forms of AML.
- APL is responsive to **cytarabine** and **daunorubicin**, but about **10%** of patients treated with these drugs die from **DIC** induced by the release of granule components by dying tumor cells.



49) A 65 year old healthy male was incidentally found to have on his labs: WBC of 70K with lymphocyte predominance, platelets of 250k, Hb 12. He does not have any lymph node enlargement or splenomegaly or hepatomegaly. Cytoanalysis of lymphocytes showed 5% of cd38 (ZAP) positive cells. The rest were cd 19 and cd 20 positive. Smudge cells were seen. Which of the following is **true** about this case:

a. The patient will survive for years
b. The patient will develop generalized lymphadenopathy and splenomegaly in a year
c. The Patient should receive initial therapy with fludarabine
d. The patient has an advanced stage disease
e. P53 loss/mutation is an expected genetic abnormality in this specific patient.

ANSWER:A

Mutation status of IgHV genes

- Unmutated:
  - Pregerminal centre cell
  - Rapid progression
- Mutated:
  - Postgerminal centre cells
  - Slow progression
- Surrogate markers
  - ZAP 70 and CD38

CLL

Staging: Rai and Binet staging systems for CLL

Clinical staging systems for CLL

	Stage			
Value	Rai	Binet		Median survival
Lymphocytosis (>15,000/mm³)	0	-		150 months (12.5 years)
Lymphocytosis plus nodal involvement	I	A	<3 node groups	101-108 months (8.5-9 years)
Lymphocytosis plus organomegaly	II	B	>3 node groups	60-71 months (5-6 years)
Anemia (RBCs)	III Hgb <11 g/dL	C	Hgb <10 g/dL	19-24 months (1.5-2 years)
Lymphocytosis plus thrombocytopenia (platelets)	IV PLT <100,000/mm³		PLT <100,000/mm³	

50) patient with recurrent viral, fungal, & protozoal infection. This patient most likely has a defect In:

a. B cell
b. T cell
c. macrophage
d. neutrophil
e. immunoglobulin

ANSWER:B

51) A 54 year old male complains of abdominal discomfort , weight loss, sweating and headache. P/E: showed splenomegaly. Platelets 800k.WBC 120K neutrophils 80% and basophils 2%.Uric acid elevated.Hb 13. What is the diagnosis?

A. ALL
B. CLL
C. CML in chronic phase
D. Richter transformation

ANSWER: C

Differential diagnosis between leukemoid reaction and myeloproliferative neoplasms

Disease	CBC and peripheral blood smear	LAP score	Genetics/cause
Polycythemia vera	• **Erythrocytosis** • Thrombocytosis • Leukocytosis	• ↑	• ***JAK2* mutation** in 95% of cases
Primary myelofibrosis	• Progressive **pancytopenia** • **Dacryocytes**	• ↑	• *JAK2* mutation in up to 60% of cases
Essential thrombocythemia	• Isolated **thrombocytosis**	• ↑	• *JAK2* mutation in 50% of cases
Chronic myeloid leukemia	• **Extreme leukocytosis** • **Basophilia** • Neutrophils may have the Pseudo-Pelger-Huet anomaly	• ↓	• **Philadelphia chromosome**
Leukemoid reaction [13]	• **Leukocytosis** • Fewer granulocytic precursors, basophils, and eosinophils compared to CML • Neutrophils have toxic granulation, toxic vacuolation, and Dohle bodies	• ↑	• No mutation • Typically secondary to infections or drugs (e.g., steroids) • Associated with certain solid tumors (e.g., lung and kidney cancer)

Chronic myeloid leukemia (CML) is a type of myeloproliferative neoplasm involving hematopoietic stem cells that results in overexpression of cells of myeloid lineage, especially granulocytes. It is caused by a reciprocal translocation between chromosomes 9 and 22, resulting in the formation of the Philadelphia chromosome, which contains the *BCR-ABL1* fusion gene. The *BCR-ABL1* fusion gene encodes a hybrid tyrosine kinase with increased enzymatic activity that leads to unregulated proliferation of hematopoietic stem cells. These cells subsequently differentiate into mature myeloid cells, resulting in CML. CML has three distinct phases: the chronic phase (CP-CML), the accelerated phase (AP-CML), and the blast phase (BP-CML, also referred to as blast crisis. Patients with CP-CML may be asymptomatic or present with nonspecific symptoms (e.g., fever, weight loss, night sweats) and splenomegaly. AP-CML is characterized by complications secondary to the suppression of other hematologic cell lines (e.g., thrombocytopenia, anemia, recurrent infections), and the clinical picture of BP-CML is similar to that of acute leukemia. Important diagnostic features in BP-CML are severe leukocytosis (with leukocyte counts as high as 500,000/mm^3), basophilia, and massive splenomegaly. The most effective treatment for CML is targeted therapy with tyrosine kinase inhibitors (TKIs). This class of drug has revolutionized the efficacy of treatment and greatly improved the prognosis of patients with CML.

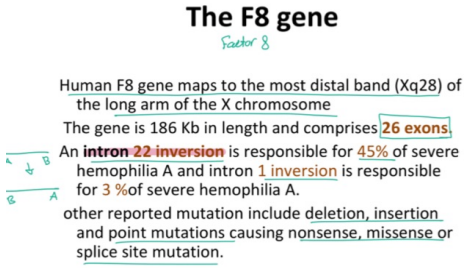
52) a pt with chronic low back pain of 6 months duration, his prostate is enlarged and his prostate specific antigen is elevated, he's having pancytopenia, his bone marrow biopsy revealed abnormal dysplastic cells,
what's the cause of his **pancytopenia:**

A) infiltration of the bone marrow by metastatic prostate cancer
B) myeloid plastic syndrome

ANSWER: A

53)A mutation that indicates severe hemophilia?

ANSWER: inversion 22



54)True about hemophilia:

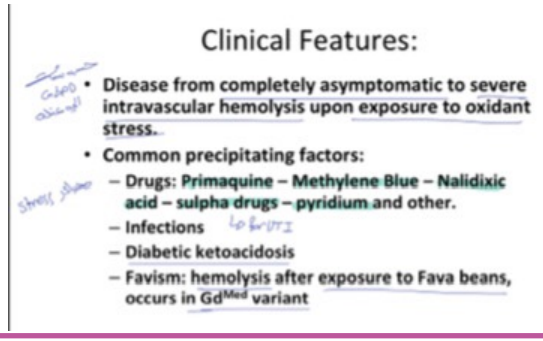
ANSWER: factor 8 level correlates with severity.

55)Most common cause of severe hemophilia A in Jordan is?

ANSWER: Intron 22 inversion

56)a jordanian soldier was given anti malarial prophylaxis, then he developed symptoms of anemia, on blood smear was found to have heinz bodies. what is the most likely dx :

ANSWER:G6PD deficiency



Clinical Features:

- Disease from completely asymptomatic to severe intravascular hemolysis upon exposure to oxidant stress.
- Common precipitating factors:
  - Drugs: Primaquine – Methylene Blue – Nalidixic acid – sulpha drugs – pyridium and other.
  - Infections
  - Diabetic ketoacidosis
  - Favism: hemolysis after exposure to Fava beans, occurs in Gd^{Med} variant

57) which of the following is the characteristic cell seen in the peripheral blood in patients with autoimmune hemolytic anemia warm antibodies:

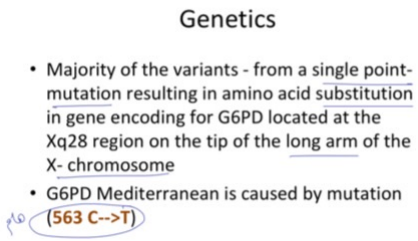
ANSWER:densely hemoglobinated spherocytes

58)Pt with pneumonia and anemia, reticulocytes 8%, Which is wrong:

ANSWER:LDH is usually within normal

59)G6PD Mediterranean is associated with:

ANSWER:mutation (563 C—>T)


Genetics

- Majority of the variants - from a single point-mutation resulting in amino acid substitution in gene encoding for G6PD located at the Xq28 region on the tip of the long arm of the X- chromosome
- G6PD Mediterranean is caused by mutation (563 C-->T)

60)True in DIC:

ANSWER:increased PT

61)patient post.op, develop drug induced ischemic signs in his hands, what do u expect to find in labs?

ANSWER: decrease >50% in platlet count

62) seen in HIT?

ANSWER:skin necrosis at site of injection

63)wrong in TIP case:

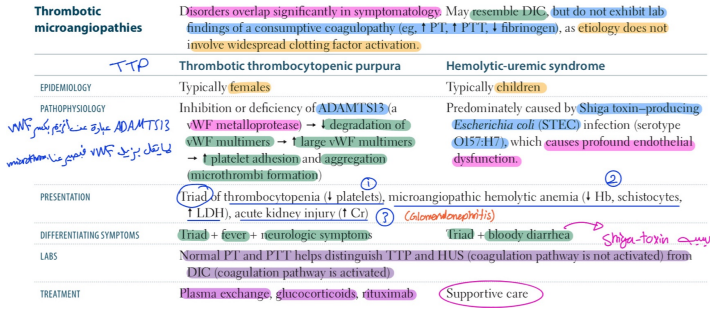
ANSWER: give platelets.

Approach to the Treatment of ITP

Initial treatment	Glucocorticoids IVIG
Curative therapy	Glucocorticoids Splenectomy Rituximab
Rescue therapy	High dose glucocorticoids IVIG
Chronic therapy	Many agents **Thrombopoietin receptor agonists**

64) A female patient with headache bleeding from mucosal surface was found to have low platelet count. Hb is decreased with schistocytes on blood smear.Her temperature is 38.5. WBC are normal,PT and PTT are normal. What's the most probable explanation:

ANSWER:autoantibodies to plasma protease ADAMTS13



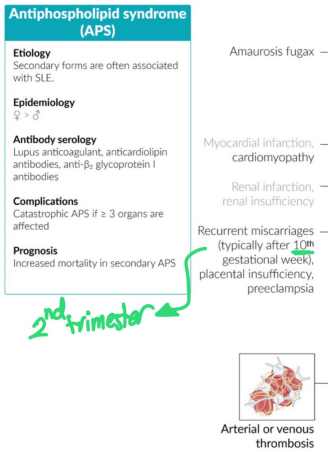
Thrombotic microangiopathies	Disorders overlap significantly in symptomatology. May resemble DIC, but do not exhibit lab findings of a consumptive coagulopathy (eg, ↑ PT, ↑ PTT, ↓ fibrinogen), as etiology does not involve widespread clotting factor activation.	
	Thrombotic thrombocytopenic purpura	Hemolytic-uremic syndrome
EPIDEMIOLOGY	Typically females	Typically children
PATHOPHYSIOLOGY	Inhibition or deficiency of ADAMTS13 (a vWF metalloprotease) → ↓ degradation of vWF multimers → ↑ large vWF multimers → ↑ platelet adhesion and aggregation (microthrombi formation)	Predominately caused by Shiga toxin–producing *Escherichia coli* (STEC) infection (serotype O157:H7), which causes profound endothelial dysfunction.
PRESENTATION	Triad of thrombocytopenia (↓ platelets), microangiopathic hemolytic anemia (↓ Hb, schistocytes, ↑ LDH), acute kidney injury (↑ Cr)	
DIFFERENTIATING SYMPTOMS	Triad + fever + neurologic symptoms	Triad + bloody diarrhea
LABS	Normal PT and PTT helps distinguish TTP and HUS (coagulation pathway is not activated) from DIC (coagulation pathway is activated)	
TREATMENT	Plasma exchange, glucocorticoids, rituximab	Supportive care

64) Which of the following doesn't occur in blood transfusion?

ANSWER:hypokalemia

65)patient with recurrent 2nd tri. pregnancy loss, Which of the following is least likely going to be changed:

ANSWER: PT



66) Worst lymphoma:

ANSWER: Burkitts lymphoma

67) lymphoma with best prognosis?

Answer: Follicular lymphoma

68)AML (M3) treatment:

ANSWER:all-trans retinoic acid

69)Multiple myeloma stage 3 treatment :

ANSWER: Chemotherapy and Bone marrow transplant

Treatment of Multiple Myeloma

Standard Chemotherapy +steroids

- Dexa and Thalidomide
- Dexa and Bortezomib(Velcade)/ Lenalidomide
- Melphalan and prednisone
- High Dose Chemotherapy

with Bone marrow transplant (auto)

70)CLL case, treatment of autoimmune hemolytic anemia:

ANSWER:Steroids

71) CML case: wrong about it?

ANSWER:positive globulin test.

72)What is the genetic abnormality in CML?

ANSWER:t(9.22)

73)Hb 14.5 , WBC 56000 , plts 960,000 , 80% neutrophils, 3% basophils In this case, the diagnosis most likely is :

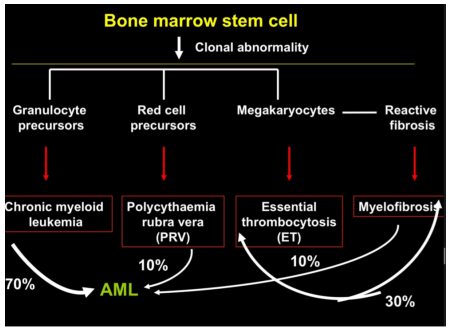
ANSWER:CML

74)Not used for Dx of NHL

ANSWER:B2 microglobulin

75) wrong about polycythemia vera?

ANSWER:50% transform into AML



76)AML with myelomonocytes, what do you see in cytogenetics:

ANSWER:inv 16

M4: Myelomonocytic leukemia

- With inverted 16 and associated eosinophilia this is a good prognostic category
- Associated with leukemia cutis.
- CNS disease may occur.

77)A patient who was on warfarin developed an episode of melena, his INR was 7. 9, hemoglobin was 9. 3. In addition to omitting warfarin,what is the next step:

Ans: give vitamin K.

78)Long case with image for Aure bodies:

ANSWER:: acute leukemia

79)long case scenario (heme-arthrosis, painful, tenderness knee joint + swelling and redness) what is special; markedly elevated PTT,PT normal,WBC normal,Platelet normal,decreased Hb(forgot the number),what to do next:

A. Factors X,V,II
B. Factors VII,IX,XI,XII
C. Mixing study

ANSWER: C

80)Long case scenario (pregnant lady ; fatigue, SOB, MCV= 60, low MCH, HbF <1% (normal level),HbA2 4. 5%(normal 1. 5–3. 5%) ,what is the diagnosis:

A. Iron deficiency anemia
B. Beta thalassemia trait
C. Sickle cell anemia

ANSWER:B

81)which one is Not considered a variable in MDS classification according to IPSS:

A. Percentage of blasts
B. Creatinine
C. Anemia
D. Thrombocytopenia
E. Karyotype

ANSWER: B

82)Wrong about IDA:

A. High reticulocyte count
B. High TIBC
C. Low ferritin
D. Low serum iron
E. Low MCV

ANSWER: A

83)21 year old female with purpura on chest and back and low platelet count with hx of recent URI (There is a similar case in the slides)

A. ITP
B. HSP

ANSWER:: A

84)Patient with fever and drenching sweats,red steenberg cells,has enlarged lymph nodes on both sides of the diaphragm and hypodense liver lesion,what is her ann arbor stage:

A. II
B. III A
C. III B
D. IV A
E. IV B

ANSWER: E

85)A 30 year old woman was brought to the ER by her husband and was disoriented to surrounding,decreased Hb,normal WBCs,decreased platelets,Cr 4,LDH 859,haptoglobin low,bilirubin 3,peripheral blood smear
is shown,what is the diagnosis:

ANSWER:TTP

86)A 55 year old male patient suffers from itching after taking hot shower baths,all of the following are cause to his presentation except:

A. Polycythemia rubra vera
B. Hemochromatosis
C. Renal cell carcinoma
D. Dehydration

ANSWER: B

87)which is not a risk factor of IDA?

A. vegan
B. old age
C. multiparity
D. menorrhagia

ANSWER:B

88)Defect in TTP ? ADAMTS13 deficiency

89) Poor prognosis in AML ? chr 7 deletion

90) A case of CLL, what is the prognosis? – The patient will live for a long time without treatment (he's in the chronic phase)

91) A case of CML, what to do? – Do FISH looking for BCR-ABL translocation

92) Most common cause of severe hemophilia A in Jordan is? – Intron 22 inversion

93) Most common presentation of acute hemolytic reaction in a young adult is? – Back pain, red urine, and headache

94) True about TRALI? occurs in the first 6 hours following transfusion

95) A case with spherocytes, high LDH and other features of hemolytic anemia, most likely Dx? Hemolytic anemia, autoimmune

96) A case of bleeding gums and heavy menstruation, PT, PTT, plt count are all normal, Dx? Glanzmann Thrombasthenia

97) Treatment to cure transfusion-dependent beta-thalassemia – Bone marrow transplantation

11- Patient with multiple myeloma, what makes him stage 3? – beta2-microglobulin above 5. 5

Extra questions

A 22-year-old man presents to the emergency department with mucosal bleeding and epistaxis. Laboratory tests show an increased bleeding time and an Increased partial thromboplastin time. His mother was anemic throughout her life and required several blood transfusions after a minor operation.

Which of the following is the most likely diagnosis?

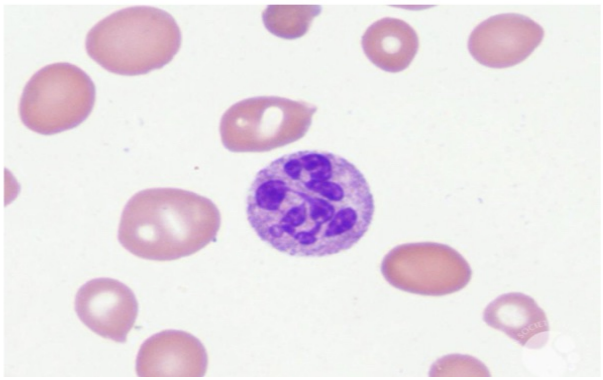
- **A.** Bernard-Soulier syndrome
- **B.** Glanzmann disease
- **C.** Hemophilia A
- **D.** Vitamin K deficiency
- **E.** Von Willebrand disease

ANSWER:E

A 43-year-old man presents to his physician with fatigue. The patient says he Is concerned about his fatigue because he has a strong family history of cancer. He thinks that if his parents did not drink, smoke, and eat such poor diets they would have lived longer lives. He says that because of all this, he never drinks alcohol or smokes tobacco. He has also followed a strict vegan diet for 10 years. He says that all of his meals are high in leafy green vegetables. Laboratory tests show a hematocrit of 35% with a normal RBC distribution width. Results of a peripheral blood smear are shown In the Image.

Which of the following laboratory tests will most definitively determine the likely cause of his abnormal blood smear?

- **A.** Folate level
- **B.** Homocysteine level
- **C.** Methylmalonic acid level
- **D.** Serum gastrin levels
- **E.** Urine vitamin B_{12} level



ANSWER:C

A 70-year-old woman presents to her physician with shortness of breath. On further questioning, she admits to feelings of lethargy, weakness, and occasional bone pain. Her serum calcium levels are found to be elevated, and a CBC reveals an abnormally low hemoglobin and hematocrit. Urinalysis reveals Increased protein levels, and urine electrophoresis demonstrates an M spike in the y-globulin fraction.

Which of the following would be found in a peripheral blood smear from this patient?

- **A.** Broken and destroyed lymphocytes
- **B.** Increased levels of circulating promyelocytes and myelocytes
- **C.** Microcytosis and hypochromasia
- **D.** Myeloblasts with Auer rods
- **E.** Rouleaux formation

ANSWER:E

A 9-year-old boy with Down syndrome presents to his pediatrician with fever, fatigue, pain in both arms and legs, and a headache that started about 2 weeks ago and rapidly became worse in the last few days. Physical examination demonstrates generalized lymphadenopathy and petechiae on his trunk. Laboratory studies show leukocytosis and profound anemia and thrombocytopenia

Which of the following is the most likely diagnosis?

- **A.** Acute infective endocarditis
- **B.** Acute lymphocytic leukemia
- **C.** Chronic myelogenous leukemia
- **D.** Multiple myeloma
- **E.** *Streptococcus pyogenes*-induced toxic shock syndrome

ANSWER:B

A 76-year-old man presents to the emergency department with a 3-day history of constipation. He reports fatigue and changes in bowel habits over the period of a year, including dark stools. On physical examination the patient's abdomen is remarkably distended. Barium enema reveals an apple- core lesion of the rectosigmoid junction, and biopsy demonstrates adenocarcinoma. Laboratory studies show:

WBC count: 10,200/mm
Hemoglobin: 8.8 g/ dL
Hematocrit: 26.%
Platelet count: 231,000/ mm3
The patient's low hematocrit likely reveals what type of anemia?

The patient's low hematocrit likely reveals what type of anemia?

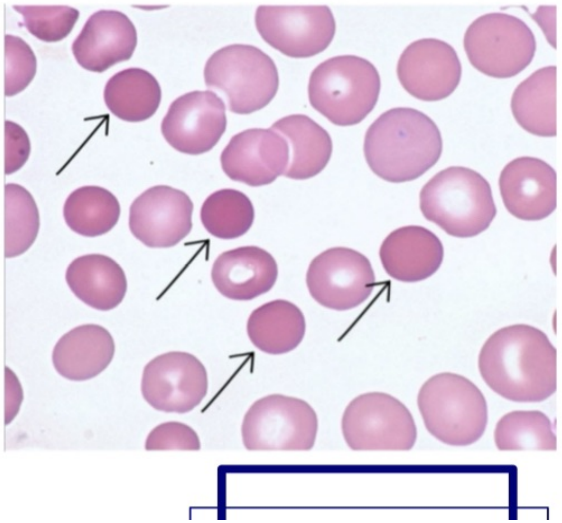
- **A.** Aplastic anemia
- **B.** Hemolytic anemia
- **C.** Megaloblastic anemia
- **D.** Microcytic anemia
- **E.** Sideroblastic anemia

ANSWER:D

A 10-month-old infant is taken to the doctor for persistent jaundice and easy fatigability. The mother is worried because the baby's father had his spleen removed as a child because "something was wrong with his blood." On physical exam, the child is mildly tachycardic with splenomegaly. A peripheral blood smear is obtained, and shows the presence of abnormal red cells, as seen in this image.

Which of the following would be expected in this patient's blood?

- **A.** Decreased mean corpuscular hemoglobin concentration
- **B.** Hypersegmented polymorphonuclear cells
- **C.** Increased mean corpuscular hemoglobin concentration
- **D.** Increased total iron-binding capacity
- **E.** Positive direct Coombs test
- **F.** Rouleaux formation of RBCs



ANSWER:C

A 30-year-old woman presents to the emergency department because of new-onset dyspnea and pleuritic chest pain. Physical examination Is notable for tachypnea, tachycardia, and tenderness to deep palpation in her right leg. Arterial blood gas studies show an increased alveolar-to-arterial gradient. Laboratory studies show an increased D-dimer level.

Which of the following is the most common condition predisposing this patient to her current presentation?

- **A.** A genetic defect that causes the prothrombin level to increase
- **B.** Autosomal dominant condition preventing protein C from degrading factor V
- **C.** Genetic absence of antithrombin III
- **D.** Genetically abnormal fibrinogen causing clots
- **E.** Protein deficiency resulting in increased factor Va and VIIIa

ANSWER:B

. A 7-year-old girl is brought to the emergency department because of a 7-day history of bloody diarrhea. She has had generalized fatigue for the past 2 days. Her temperature is 37°C (98.6°F), pulse is 112/min, respirations are 14/min, and blood pressure is 105/65 mm Hg. Examination shows pallor and scleral icterus. The liver is not enlarged or tender. Laboratory studies show:

Hemoglobin	6 g/dL
Mean corpuscular volume	80 μm^3
Leukocyte count	18,000/mm^3
Reticulocyte count	12%
Platelet count	50,000/mm^3

Which of the following is the most likely explanation for these findings?

A) Acute lymphoblastic leukemia
B) Bone marrow suppression
C) Glucose 6-phosphate dehydrogenase deficiency
D) Hemoglobin SS disease
E) Hemolytic-uremic syndrome
F) Iron deficiency

Extra Best next step:
Look for schistocytes in blood film

ANSWER:E