

YR 2 HEMATOLOGY UNIT EXAMINATION -- April 07, 1999.

CHOOSE THE SINGLE BEST ANSWER FOR QUESTIONS 1 - 100.

1. A 23 year old African-American male is transferred to your hospital for replacement of his aortic valve. He has received oxacillin for 3 weeks for staphylococcal endocarditis. His current CBC is WBC 3600/ul (35% neutrophils, 65% lymphocytes), hemoglobin 14.1 gm/dl, hematocrit 42%, MCV 90 fl, RDW 13.0, and platelets 167,000/ul. The MOST useful test to explain his CBC would be:
 - A. Vitamin B12 level
 - B. Leukocyte alkaline phosphatase
 - C. Obtain his CBCs from the transferring hospital
 - D. CBC using citrate as the anticoagulant
 - E. ⁹⁹Techneium WBC survival

2. Recurrent infection with catalase producing organisms is most characteristic of individuals with:
 - A. Acute lymphoblastic leukemia
 - B. Aute myelogenous leukemia
 - C. Chediak-Higashi syndrome
 - D. Chronic granulomatous disease
 - E. Hereditary acatalasemia

3. A 55 year old man is found to have the hypereosinophilic syndrome. All the following would be compatible with this diagnosis EXCEPT:
 - A. Eosinophils > 1,500/ul for 6 months
 - B. Endomyocardial fibroelastosis
 - C. Pulmonary infiltrate on chest x-ray
 - D. Angioedema
 - E. A history of chronic allergies since childhood

4. An accelerated phase due to an Epstein-Barr virus driven lymphoproliferative disorder is part of the natural history of:
- A. Chronic myelogenous leukemia
 - B. Chronic lymphocytic leukemia
 - C. Leukocyte adhesion deficiency
 - D. Chediak-Higashi syndrome
 - E. Chronic granulomatous disease
5. A 50 year old female with rheumatoid arthritis is found to have a palpable spleen. Which of the following would be MOST compatible with a diagnosis of Felty's syndrome:
- A. Autoimmune hemolytic anemia
 - B. Immune thrombocytopenia
 - C. Neutropenia
 - D. Neutrophilia
 - E. A leukemoid reaction
6. A decrease in ATP generation is the primary defect responsible for the hemolytic anemia associated with:
- A. Hereditary pyruvate kinase deficiency
 - B. G6PD Mediterranean
 - C. Hereditary spherocytosis
 - D. Sickle cell anemia
 - E. beta-thalassemia major

7. A 38 year old African-American male (shown by the arrow) is found to have a chronic hemolytic anemia. The family tree is shown below. Dark boxes represent an abnormal gene, while open boxes represent the wild type allele.

The MOST LIKELY diagnosis for the chronic hemolytic anemia is:

- A. Sickle cell trait
 - B. Hereditary spherocytosis
 - C. G6PD A⁻
 - D. Pyruvate kinase deficiency
 - E. Hydrops fetalis
8. All of the following contribute to the shortened survival of the red blood cell in hereditary spherocytosis EXCEPT:
- A. Decreased surface area to volume ratio
 - B. Loss of membrane lipid in microvessicles
 - C. Decreased binding of the lipid bilayer to the cytoskeleton
 - D. Overexpression of the spectrin gene
 - E. Splenic trapping

9. A 65 year old women is being evaluated for a new anemia. Her direct Coombs (antiglobulin) test is negative. Her indirect Coombs (antiglobulin) test shows the presence of anti-Kell antibodies. Her red blood cells are Kell negative. She has never had a blood transfusion. The best interpretation of her laboratory testing is:
- A. Isoimmune hemolytic anemia
 - B. Alloimmune hemolytic anemia
 - C. Autoimmune hemolytic anemia
 - D. Her anemia is not immune mediated, but she has been alloimmunized during a prior pregnancy
 - E. Laboratory error
10. All of the following are manifestations of hemolytic anemia EXCEPT:
- A. Reticulocytosis
 - B. Increased LDH
 - C. Increased requirement for iron
 - D. Decreased haptoglobin
 - E. Increased unconjugated bilirubin
11. The most useful test to evaluate the etiology of thrombocytopenia is:
- A. The peripheral smear
 - B. ADP-induced platelet aggregation
 - C. Thrombopoietin level
 - D. ⁹⁹Techetium platelet survival
 - E. Platelet electron microscopy

12. Bernard-Soulier syndrome is due to:
- A. A decrease in platelet dense granule exocytosis
 - B. A decrease in platelet alpha granules
 - C. A defect in the GP 1b/IX complex
 - D. Abnormal high molecular weight von Willebrand multimers
 - E. Decreased platelet p-selectin
13. Thrombocytopenia and microangiopathic hemolytic anemia are characteristic of all of the following EXCEPT:
- A. Disseminated intravascular coagulation
 - B. Thrombotic thrombocytopenic purpura
 - C. Hemolytic uremic syndrome
 - D. Malfunctioning prosthetic heart valve
 - E. Immune thrombocytopenic purpura (ITP)
14. All of the following are associated with an absent first wave of platelet aggregation EXCEPT:
- A. Glanzman's thrombasthenia
 - B. Absent GP IIb/IIIa
 - C. Quinidine
 - D. Ticlopidine
 - E. Hypofibrinogenemia
15. The development of thrombosis in heparin associated thrombocytopenia is best explained by:
- A. Release of thrombogenic platelet microparticles
 - B. Overproduction of platelet dense granules
 - C. Direct toxic effect of heparin upon the endothelium
 - D. Induction of platelet apoptosis
 - E. Rapid clearance of heparin by antiheparin antibodies resulting in inadequate anticoagulation

16. Lymphomas may arise from all of the following EXCEPT:
- A. T lymphocytes
 - B. B lymphocytes
 - C. Follicular center cells
 - D. Myeloblasts
 - E. Natural killer cells
17. Activation of the Bcl-2 oncogene results in:
- A. Decreased apoptosis
 - B. Increased apoptosis
 - C. Easier invasion of the lymphocyte by Epstein Barr virus
 - D. Easier invasion of the lymphocyte by the human T-cell lymphotropic virus 1 (HTLV-1)
 - E. No effect on the lymphocyte
18. The Working formulation for lymphoma classification is primarily based on:
- A. Cellular morphology
 - B. Immunophenotyping
 - C. Oncogene restriction polymorphisms
 - D. Cytogenetic analysis
 - E. The whims of the committee that devised it
19. The human T-cell lymphotropic virus 1 (HTLV-1) has been MOST CLOSELY associated with the development of:
- A. Burkitt s lymphoma
 - B. Acute myelogenous leukemia
 - C. Adult T-cell leukemia/lymphoma syndrome
 - D. Follicular center cell lymphomas
 - E. The T-cell variant of Waldenstroms macroglobulinemia

20. Acute bone marrow injury or failure is LEAST LIKELY to manifest initially with which one of the following:
- A. Neutropenia
 - B. Thrombocytopenia
 - C. A combination of neutropenia and thrombocytopenia
 - D. A combination of anemia and thrombocytopenia
 - E. A combination of lymphopenia and anemia
21. A child presenting with bone marrow failure is found on history to have had removal of accessory thumbs soon after birth and on special testing to exhibit excessive chromosomal cross linking. This child is MOST LIKELY suffering from:
- A. A congenital infection with rubella
 - B. The thrombocytopenia with absent radii (TAR) syndrome
 - C. Fanconi's anemia
 - D. Erythroblastosis fetalis
 - E. Chronic myelogenous leukemia
22. Marrow infiltration in association with breast cancer may produce leukoerythroblastic changes in the peripheral blood smear which include all the follow up EXCEPT:
- A. Tear drop red blood cells
 - B. Schuffners dots in red blood cells
 - C. Nucleated red blood cells
 - D. Presence of myelocytes

23. Which of the following growth factors is produced primarily in the kidney and may also be released into the circulation in association with liver disease:
- A. G-CSF
 - B. GM-CSF
 - C. Erythropoietin
 - D. IL-3
 - E. IL-11
24. Marrow failure due to the myelodysplastic syndrome is suggested by each of the following EXCEPT:
- A. Macrocytosis
 - B. Pathological ringed sideroblasts in the marrow
 - C. Normal hemoglobin electrophoresis
 - D. A history of splenectomy and gallstones at an early age
 - E. Pseudo-Pelger Huet cells
25. All of the following infections have been documented to cause bone marrow failure EXCEPT for:
- A. Parvovirus
 - B. Hepatitis B
 - C. Cytomegalovirus
 - D. Rhabdovirus
 - E. Hepatitis C

26. Which of the following patients is most likely to present with extramedullary leukemia manifested by gingival infiltration and hypertrophy?
- A. A 65 year old woman with refractory anemia
 - B. A 20 year old male with blast crisis of chronic myelogenous leukemia
 - C. A 30 year old woman with acute monocytic leukemia [FAB M5]
 - D. A 10 year old boy with acute lymphocytic leukemia
 - E. A 30 year old man with acute myelogenous leukemia who had received successful treatment with chemotherapy for Hodgkins disease 7 years ago
27. The Philadelphia chromosome [t(9;22)] is LEAST likely to be found in which of the following patients with leukemia?
- A. A 45 year old man with acute lymphocytic leukemia
 - B. A 12 year old girl with chronic myeloid leukemia
 - C. A 30 year old female survivor of the atomic bomb in Hiroshima who presented with an elevated white blood cell count 7 years after the bombing
 - D. A 7 year old boy with acute myeloid leukemia, FAB M4
28. Which of the following individuals is MOST LIKELY to have T lineage ALL?
- A. A 77 year old woman who presents with anemia, splenomegaly and small mature lymphocytes on peripheral smear
 - B. A 20 year old male with circulating blasts containing Auer rods
 - C. An 11 year old boy with a white blood count of 120,000/u1 and a mediastinal mass on chest radiograph
 - D. A 39 year old man with pancytopenia who had previously received chemotherapy for Hodgkins Disease

29. Which of the following is most characteristic of the bone marrow in patients with myelodysplasia?
- A. Very hypocellular with erythroid hypoplasia
 - B. Hypercellular with erythroid hyperplasia
 - C. Fibrotic and difficult to aspirate
 - D. Hypercellular with 75% myeloblasts
 - E. Hypercellular with an increased myeloid:erythroid ratio
30. Which of the following patients is MOST LIKELY to have peroxidase positive blasts?
- A. A child with Burkitts leukemia/lymphoma
 - B. A child with FAB M-2 acute myelogenous leukemia (AML)
 - C. A child with B lineage acute lymphoblastic leukemia (ALL)
 - D. A child with T lineage acute lymphoblastic leukemia (ALL)
31. Visibly enlarged lymph nodes are common in individuals with:
- A. Refractory anemia
 - B. Acute progranulocytic leukemia
 - C. Acute myelogenous leukemia, FAB M2
 - D. Aplastic anemia
 - E. None of the above
32. All of the following have been associated with the development of acute leukemia EXCEPT:
- A. Radiation exposure
 - B. Living adjacent to high voltage electric wires
 - C. Exposure to benzene
 - D. Treatment with alkylating agents
 - E. Myeloproliferative disorders

33. All trans-retinoic acid has the following effect on the leukemic blasts in patients with acute promyelocytic leukemia:
- A. Induces maturation and terminal differentiation
 - B. Produces tumor lysis and worsening of the coagulopathy
 - C. Stimulates blast cell proliferation
 - D. Induces apoptosis
34. While doing a routine examination of a 10-year-old African-American girl who is new to your practice, you find her spleen to be palpable 2cm below the left costal margin. Neither the child nor her mother knows if splenic enlargement was found in the past. On further questioning, the mother reveals that the child had mild leg pains off and on which were called growing pains . The MOST LIKELY diagnosis is:
- A. Sickle cell anemia (hemoglobin S-S)
 - B. Sickle cell trait (hemoglobin A-S)
 - C. Glucose-6-phosphate dehydrogenase A⁻ heterozygosity
 - D. Hemoglobin S-C disease
 - E. Thalassemia major (homozygous beta-thalassemia)
35. Sickle cell anemia (hemoglobin S-S) has been diagnosed in a newborn infant. The mother asks you what she should expect her child s course to be. Each of the following is true EXCEPT:
- A. It is unusual to develop severe manifestations of sickle cell disease during the first 2 months of life
 - B. Dactylitis is common in childhood
 - C. There is an increased risk of developing gallstones
 - D. Painful episodes are unusual until the child becomes a teenager
 - E. Fever requires prompt medical attention because it may be a symptom of overwhelming bacterial infection

36. All of the following are life threatening complications of sickle cell disease EXCEPT:
- A. Splenic sequestration
 - B. Stroke
 - C. Pain attack
 - D. Aplastic crisis
 - E. Bacterial sepsis
37. All of the following are seen in alpha thalassemia minor EXCEPT:
- A. Normal RDW
 - B. Microcytosis
 - C. Increased hemoglobin A₂
 - D. Decreased alpha to beta globin chain synthetic ratio in reticulocytes
 - E. Detectable restriction endonuclease polymorphisms
38. The MOST frequent cause of death in beta thalassemia major is:
- A. Tissue anoxia secondary to severe anemia
 - B. Upper airway obstruction from abnormalities in facial bones from widened diploic spaces
 - C. Cardiac arrhythmias
 - D. Folic acid deficiency
 - E. Stroke
39. Of the following, which is the most common complication of sickle cell trait?
- A. Vaso-occlusive pain
 - B. Increased risk of infection
 - C. Mild hemolytic anemia
 - D. Splenic dysfunction
 - E. Hematuria

40. Total gastrectomy will lead to the development of:
- A. Iron deficiency because non heme iron won't be reduced
 - B. Vitamin B12 deficiency
 - C. Folic Acid deficiency
 - D. Hemochromatosis
 - E. Small bowel lymphoma
41. World-wide, the MOST COMMON cause of anemia is:
- A. Folate deficiency
 - B. Pernicious anemia
 - C. Iron deficiency
 - D. Sideroblastic anemia
 - E. G6PD Mediterranean
42. All of the following individuals generally need extra iron added to their diet EXCEPT:
- A. Pregnant women
 - B. Adolescent girls
 - C. Men with beta thalassemia minor
 - D. 6 month old children
 - E. Men with chronic bleeding due to esophageal varices
43. Transfusional hemosiderosis (iron overload) is associated with all of the following EXCEPT:
- A. Cardiomyopathy
 - B. Diabetes mellitus
 - C. Renal failure
 - D. Growth failure
 - E. Cirrhosis of the liver

44. A patient is felt to have a megaloblastic anemia. Which of the following on physical exam will help distinguish B12 from folate deficiency?
- A. "Beefy" red tongue
 - B. Stocking glove parasthesias
 - C. Loss of vibratory sensation
 - D. Rales on examination of the lungs
 - E. Spooning of the fingernails
45. A 60 year old male patient has nodular sclerosing Hodgkin's Disease (HD) diagnosed on biopsy of an enlarged left supra-clavicular lymph node. Staging work up is positive for lymph nodes in the right hilum and left axilla. CAT Scans show no other sites of disease. Bone marrow biopsy showed the presence of HD. His disease would be MOST accurately stage as:
- A. Stage I
 - B. Stage II
 - C. Stage III
 - D. Stage IV
 - E. Stage V
46. All of the following are consistent with Stage IV chronic lymphocytic leukemia (Rai Staging) EXCEPT:
- A. Lymphocytosis
 - B. Splenomegaly
 - C. Hemoglobin of 9.5 gm/dl
 - D. Platelet count of 120,000/ul

47. All of the following are manifestations of multiple myeloma EXCEPT:
- A. Retinal hemorrhage secondary to hyperviscosity
 - B. Anemia
 - C. Hypercalcemia
 - D. Lytic bone lesions
 - E. Lymphadenopathy
48. Hairy cell leukemia is characterized by all of the following EXCEPT:
- A. Splenomegaly
 - B. Pancytopenia
 - C. Male predominance
 - D. Tartrate-resistant acid phosphase (TRAP) positive lymphocytes
 - E. 80% chance of cure with chemotherapy
49. Tumor lysis Syndrome can be seen in chemotherapy-sensitive rapidly growing tumors undergoing treatment. It is characterized by all of the following laboratory features EXCEPT:
- A. Hyperuricemia
 - B. Hyperkalemia
 - C. Hyperphosphatemia
 - D. Hypercalcemia
 - E. Increased BUN

50. The presence of "B" symptoms has prognostic significance for patients with Hodgkin's disease and non-Hodgkin's lymphoma. Which of the following is NOT considered a "B" symptom in such patients?
- A. Fever $>38^{\circ}\text{C}$ ($>100.4^{\circ}\text{F}$)
 - B. Night sweats
 - C. Pruritus (itching) involving at least 1/3 of body skin
 - D. Weight loss of 10% or more of body weight
51. Which of the following is NOT a clonal disorder?
- A. Chronic myelogenous leukemia
 - B. Acute myelogenous leukemia
 - C. Essential thrombocythemia
 - D. Polycythemia vera
 - E. All of the above are clonal disorders
52. Mutations in the erythropoietin receptor (EpoR) can cause:
- A. Familial erythrocytosis
 - B. Polycythemia vera
 - C. Chronic myelogenous leukemia
 - D. Acute myelogenous leukemia
 - E. Iron deficiency anemia
53. Which of the following is NOT associated with polycythemia vera?
- A. Enlarged spleen
 - B. Leukocytosis
 - C. Thrombocytosis
 - D. Red blood cell lifespan of 140 days
 - E. Elevated red cell mass

54. Which is the most specific test to establish the diagnosis of chronic myelogenous leukemia?
- A. Analysis for the Philadelphia chromosome
 - B. BCR-Abl translocation analysis
 - C. Leukocyte alkaline phosphatase
 - D. Absolute neutrophil count
 - E. Sokal score or "new" score
55. Essential thrombocythemia:
- A. Is a diagnosis of exclusion
 - B. Is often discovered serendipitously on a routine cbc
 - C. Can cause pathological bleeding
 - D. Can cause pathological thrombosis
 - E. All of the above
56. All of the following components of the hemostatic system are synthesized in the liver EXCEPT:
- A. Factor V
 - B. Factor VII
 - C. Factor VIII
 - D. von Willebrand factor
 - E. Plasminogen
57. Antithrombin III deficiency is likely to produce clinical signs and symptoms most similar to those of:
- A. Prothrombin deficiency
 - B. Factor VIII deficiency
 - C. Protein C deficiency
 - D. von Willebrand disease
 - E. Factor XIII deficiency

58. All of the following are common causes for vitamin K deficiency in hospitalized patients EXCEPT:
- A. Diminished dietary intake
 - B. Antibiotic therapy may eliminate gut bacteria that synthesize Vitamin K
 - C. Patients with biliary tract disease may have difficulty absorbing Vitamin K
 - D. Because it is water soluble, dietary Vitamin K may be lost from the gut in diarrheal diseases
 - E. Humans have very low Vitamin K stores
59. Which of the following statements concerning hyperhomocyst(e)inemia is MOST ACCURATE?
- A. Elevations of homocyst(e)ine of less than twice normal are of little concern
 - B. The disorder arises from enzyme deficiencies in the metabolic conversion of isoleucine
 - C. The disorder may be worsened by a dietary deficiency of folic acid
 - D. Accumulation of homocyst(e)ine increases the concentrations of prothrombin and factor X
 - E. Hyperhomocyst(e)inemia produces clinical bleeding
60. Transgenic animal studies are important to understanding the hemostatic system because:
- A. It is likely that hemostatic system components play a role in ontogeny and organogenesis
 - B. Homozygous deficiency states of some hemostatic components may be lethal in early fetal life resulting in apparent infertility
 - C. The impact of mixed genetic defects may become predictable from multiple "knockout" or "knock-in" experiments
 - D. Unexpected consequences of transgenic experiments may permit the detection of illnesses not currently identified as genetically determined
 - E. All of the above are true

DIRECTIONS: Match the acute leukemia numbered 61 - 62 with the most common cytogenetic abnormality (A-E below).

- A. t(8:14)
- B. Inv 16
- C. Trisomy 8
- D. t(8;21)
- E. t(15;17)

- 61. Acute promyelocytic leukemia
- 62. Burkitt's leukemia/lymphoma (FAB L3)

DIRECTIONS: Match the descriptions numbered 63 - 66 with the diseases A - E below. Each answer may be used once, more than once or not at all.

- A. High grade lymphoma commonly involving extranodal sites
- B. High grade lymphoma primarily involving the lymph nodes
- C. Low grade lymphoma
- D. Lymphocyte predominant Hodgkin's disease
- E. Cutaneous T-cell lymphoma
- F. Waldenstrom's macroglobulinemia

- 63. Lymphoma in HIV infection
- 64. Same cell of origin as chronic lymphocytic leukemia
- 65. Reed Sternberg cell
- 66. Sezary cell

67. Which of the following is suggestive of a reactive neutrophilia:
- A. Neutrophil counts >100,000/ul
 - B. Accompanying basophilia
 - C. The presence of toxic granulations and Dohle bodies
 - D. Pseudo Pelger-Huet abnormalities
 - E. Ineffective myelopoiesis
68. The plasma of a normal person who has blood type AB Rh positive would be expected to contain:
- A. Anti-A, anti-B, and anti-Rh antibodies
 - B. Anti-A and anti-B antibodies
 - C. Anti-Rh antibodies
 - D. No anti-ABO or anti-Rh antibodies
 - E. Findings are too variable to predict
69. All of the following are manifestations of hemolytic disease in utero (before birth) EXCEPT:
- A. Hyperbilirubinemia
 - B. Anemia
 - C. Extramedullary hematopoiesis
 - D. Circulating IgG antibodies of maternal origin
 - E. Hepatosplenomegaly
70. Indications for anti-Rh immunoglobulin to prevent Rh sensitization include:
- A. Rh negative woman who delivered an Rh positive child
 - B. Pregnant, Rh negative woman with anti-Rh antibodies in her serum
 - C. Type A, Rh negative woman who delivered a type O Rh negative baby
 - D. Following a miscarriage regardless of the Rh of the woman

DIRECTIONS: For questions 71 - 72, match the following clinical situations with the most appropriate blood product (A-E below). Each alternative may be used once, more than once or not at all.

- A. Packed red blood cells
- B. Platelet concentrate
- C. Fresh frozen plasma
- D. Cryoprecipitate
- E. No transfusion required

71. A patient with disseminated intravascular coagulation secondary to bacterial sepsis who has a fibrinogen of 90 mg/dl (normal 150 - 250 mg/dl) who is having life threatening bleeding
72. A patient with a platelet count of 6,000/ul due to thrombotic thrombocytopenic purpura. The patient is covered with petechiae.

DIRECTIONS: For questions 73 - 74, match the following clinical presentations or pathophysiology with the most likely transfusion reaction or complication (A-G below). Each answer may be used once, more than once or not at all.

- A. Acute intravascular hemolysis
- B. Delayed hemolytic reaction
- C. Febrile reaction
- D. Allergic reaction
- E. Graft versus host reaction
- F. Circulatory overload
- G. Transfusion related acute lung injury (TRALI)

73. While receiving a unit of red blood cells, the patient became febrile (38° C), had back and flank pain, and became hypotensive. A posttransfusion urine showed hemoglobinuria.
74. A 43 year old woman who had five children received three units of red blood cells during surgery without complications. Her hemoglobin is 10 gm/dl on discharge. Five days later she develops jaundice. Her hemoglobin is 7.3 gm/dl and the total bilirubin is 3.3 mg/dl (unconjugated bilirubin 3 mg/dl).

75. An elevated d-dimer is most indicative of:
- A. Hemolytic anemia
 - B. Fibrin polymerization
 - C. Fibrinolysis
 - D. Fibrin polymerization followed by fibrinolysis
 - E. Fibrinolysis followed by fibrin polymerization
76. Which of the following is most suggestive of von Willebrand's disease compared to Hemophilia A:
- A. Normal bleeding time
 - B. Decreased factor VIII activity
 - C. Normal von Willebrand antigen
 - D. Recurrent bleeding into joints
 - E. Sons inheriting the disease from their fathers
77. A prolonged prothrombin time (PT) and prolonged activated partial thromboplastin time (aPTT) is compatible with a diagnosis of:
- A. Afibrinogenemia
 - B. Factor X deficiency
 - C. Long standing vitamin K deficiency
 - D. Disseminated intravascular coagulation
 - E. All of the above
78. Which of the following tests would be expected with the Factor V Leiden defect:
- A. Decreased factor V procoagulant activity (factor V activity assay)
 - B. Abnormal activated protein C resistance test
 - C. Prolonged prothrombin time (PT)
 - D. Prolonged activated partial thromboplastin time (aPTT)
 - E. Prolonged thrombin time

79. A patient has a preoperative prothrombin time (PT) and activated partial thromboplastin time (aPTT) performed. The PT is 19.2 seconds (normal < 13.1 seconds) and the aPTT is 26 seconds (normal 22.0 (inhibitor test) is 12.6 seconds. The best diagnosis is:
- A. Factor VIII deficiency
 - B. Factor II deficiency
 - C. Factor VII deficiency
 - D. Factor VIII inhibitor
 - E. Factor VII inhibitor
80. The MOST important criteria to differentiate acute myelogenous leukemia from a myelodysplastic syndrome is:
- A. Extent of dysplastic changes in the orthochromatophilic normoblasts
 - B. Presence or absence of blasts in the peripheral blood
 - C. Leukocyte alkaline phosphatase score
 - D. Cytogenetic analysis
 - E. Percentage of blasts in the bone marrow
81. A 72 year old man comes to the emergency room for back pain. He is found to have a hemoglobin of 7.8 gm/dl. His peripheral smear is shown in **Figure 1**. Which of the following statements is MOST LIKELY true:
- A. Serum IgM is decreased
 - B. The bone marrow is aplastic
 - C. He has a palpable liver and spleen
 - D. He has palpable lymph nodes
 - E. The cells are myeloperoxidase positive

82. You are asked to review a lymph node biopsy. The biopsy is shown in **Figure 2**. Which of the following is true:
- A. This disease is most commonly seen in the 10-20 year age group
 - B. This disease is associated with a t(8;14) translocation
 - C. The oncogene is a BCR-cAbl hybrid
 - D. The disease is of B-cell origin
 - E. The disease is of T-cell origin
83. A 60 year man has a routine physical exam. He is found to have an elevated white blood count, a hemoglobin of 11.8 gm/dl and a normal platelet count. His peripheral smear is shown in **Figure 3**. Which of the following is true:
- A. He is more likely Asian than Caucasian
 - B. The most likely phenotype is CD3 and CD4 positive
 - C. The most likely phenotype is CD15 and CD 30 positive
 - D. The most likely phenotype is IgM kappa positive
 - E. A lymph node biopsy would show follicular lymphoma
84. Which of the following would be the LEAST LIKELY presenting symptom for the bone marrow biopsy shown in **Figure 4**:
- A. Pruritus
 - B. Pneumonia
 - C. Petechiae
 - D. Shortness of breath
 - E. Epistaxis (nose bleed)

85. An African-American couple seek genetic counseling prior to having children. Their hemoglobin electrophoreses are shown in **Figure 5**. The chance of their first child having sickle cell anemia is best approximated as:
- A. 0%
 - B. 25%
 - C. 50%
 - D. 100%
 - E. It depends upon the sex of the child
86. A 24 year old female is referred to you for evaluation of anemia. Her mother has hereditary spherocytosis. The patient's hemoglobin is 11.0 gm/dl, MCHC 36.6 gm/dl and 8% reticulocytes. An osmotic fragility test is shown in **Figure 6**. Of curves A, B and C, which is the curve MOST LIKELY to represent the patient?
- A. Curve A
 - B. Curve B
 - C. Curve C
87. A 28 year old woman has increasing fatigue. On exam, she has pallor. Her spleen and liver are not palpable. Her CBC shows: WBC 6,700/ul, hemoglobin 5.2 gm/dl, hematocrit 15%, MCV 107 fl, platelets 310,000/ul and reticulocytes 12%. Her peripheral smear is shown in **Figure 7**. The mechanism of her anemia is MOST LIKELY:
- A. Decreased production
 - B. Increased destruction
 - C. Hemolysis
 - D. Splenic sequestration

88. A 45 year old man sees his physician for fatigue. His spleen is palpable 3 cm below the left costal margin. His white blood count is 68,000/ul and his hemoglobin is 9.7 gm/dl. His peripheral smear is shown in **Figure 8**. Which of the following is the MOST LIKELY diagnosis:

- A. Acute monoblastic leukemia
- B. Acute lymphoblastic leukemia
- C. Chronic lymphocytic leukemia
- D. Chronic myelogenous leukemia
- E. Leukemic hiatus

DIRECTIONS: Match the most appropriate description (A-E below) with the peripheral smears numbered 89-91.

- A. Nuclear fragment
- B. Iron deposition
- C. Ribosomes and RNA
- D. Insoluble oxyhemoglobin
- E. Insoluble deoxyhemoglobin
- F. Malarial parasite

- 89. Figure 9
- 90. Figure 10
- 91. Figure 11

DIRECTIONS: Match the most appropriate description (A-E below) with the peripheral smears numbered 92-93.

- A. Decreased surface area to volume ratio
- B. Substitution of valine for glutamic acid
- C. Horizontal spectrin defect
- D. Excess red cell membrane
- E. Remodeling by the spleen

- 92. Figure 12
- 93. Figure 13

94. A 26 year old female comes to the emergency room complaining of red urine and the worst headache of her life. Her sclerae are icteric and she is jaundiced. Her CBC is: WBC 9,800 (neutrophils 75%, lymphocytes 25%), hemoglobin 7.1gm/dl, hematocrit 21%, MCV 110 fl, platelets 3,000/ul, reticulocytes 20%. The creatinine is 2.2 mg/dl (normal < 1.5). Her prothrombin time (PT) is 11.1 seconds (normal < 13.1) and activated prothrombin time is 29.6 seconds (normal < 38.1 seconds). Her peripheral smear is shown in **Figure 14**. Her symptoms are MOST LIKELY due to:
- A. Leukostasis
 - B. Hyaline thrombi
 - C. Anti-platelet antibodies
 - D. IgM antibodies
 - E. Disseminated intravascular coagulation
95. The cells in **Figure 15** are most commonly associated with an abnormality of the:
- A. Retinoic acid receptor gene
 - B. C-Myc oncogene
 - C. Breakpoint cluster region
 - D. Abelson oncogene (c-abl)
 - E. IL-3 gene
96. A toddler is brought to the emergency room after hitting the left side of his head on a coffee table while learning to walk. He is shown in **Figure 16** (ignore the dermatitis on his cheeks and chin). All of the following would be appropriate tests to initially evaluate him EXCEPT:
- A. CBC
 - B. Bleeding time
 - C. Bone marrow aspiration and biopsy
 - D. Prothrombin time (PT)
 - E. Activated partial thromboplastin time (aPTT)

97. The bone marrow aspirate in **Figure 17** would most likely be due to a deficiency of which growth factor:
- A. G-CSF
 - B. GM-CSF
 - C. IL-3
 - D. IL-1
 - E. Erythropoietin
98. A 34 year old woman is found to have a hemoglobin of 6.4 gm/dl. Her peripheral smear is shown in **Figure 18**. She would be expected to have all of the following EXCEPT:
- A. Increased relative distribution width (RDW)
 - B. Decreased serum ferritin
 - C. Increased serum transferrin
 - D. Increased serum ferritin
 - E. Microcytosis
99. A 73 year old women has a syncopal episode. In the emergency room her CBC is: WBC 3,100/ul (neutrophils 30%, lymphocytes 70%), hemoglobin 5.4 gm/dl, MCV 120 fl, and platelets 97,000/ul. Her peripheral smear is shown in **Figure 19**. A Schilling test is ordered by an overzealous intern. Excretion in Part I is 1% and in Part II is 10% (normal >8%). The best diagnosis is:
- A. B₁₂ deficiency secondary to fish tapeworm
 - B. B₁₂ deficiency secondary to pancreatic insufficiency
 - C. Pernicious anemia
 - D. Folic acid deficiency
 - E. Lead intoxication

100. A woman sees her physician for fever and night sweats for the past 2 months. Her hemoglobin is 10.2 gm/dl. Her bone marrow biopsy is shown in **Figure 20**. The best diagnosis is:
- A. Aplastic anemia
 - B. Acute Myelogenous leukemia
 - C. Chronic lymphocytic leukemia
 - D. Pernicious anemia
 - E. None of the above