YR 2 HEMATOLOGY UNIT EXAMINATION -- April 08, 1998.

CHOOSE THE SINGLE BEST ANSWER FOR QUESTIONS 1 - 100.

- 1. All of the following are consistent with stage I multiple myeloma EXCEPT:
 - A. Hemoglobin < 10gm/dl
 - B. Serum calcium < 12mg/fl
 - C. Serum IgG < 5gm/dl(in IgG myeloma)</pre>
 - D. Serum IgA < 3gm/dl In IgA myeloma)
 - E. Urine light chain immunoglobulin < 4gm/24h
- 2. All of the following are consistent with stage IV chronic lymphocytic leukemia EXCEPT:
 - A. Lymphocytosis in blood and in bone marrow ($\geq 15,000/ul$ and > 40%, respectively)
 - B. Enlarged lymph nodes
 - C. Enlarged spleen
 - D. Hemoglobin of 9.5gm/dl
 - E. Platelet count of 180,000/ul
- 3. Which of the following is NOT true about lymphocytes in man?
 - A. T-lymphocytes are concerned with cellular immunity and differentiate in the thymus gland.
 - B. B-lymphocyte maturation can be antigen-independent and takes place in bone marrow or antigen-dependent which takes place in secondary lymphoid organs.
 - C. T-and B-lymphocytes can be clearly distinguished from each other morphologically using a light microscope.
 - D. Natural killer (NK) cells can kill virus-infected or tumor cells without previous exposure to target cells.
 - E. Lymphocytes migrate continuously between the circulation and different lymphoid organs.

- 4. Which of the following syndromes is <u>NOT</u> usually associated with lymphopenia?
 - A. DiGeorge's Syndrome
 - B. Wiskott-Aldrich Syndrome
 - C. Richter's Syndrome
 - D. Severe Combined Immune Deficiency (SCID) Syndrome
 - E. Systemic Lupus Erythematosus
- 5. Which of the following histopathological features is most important in distinguishing Hodgkin's disease from non-Hodgkin's lymphoma?
 - A. Absence of follicular pattern
 - B. Presence of binucleated Reed-Sternberg cells
 - C. Presence of normal "reactive" hematopoietic cells
 - D. Presence of collagen fibers intermingling with tumor cells
 - E. A characteristic "starry sky" appearance on low power projection
- 6. Which of the following histologic subtypes of Hodgkin's disease has the most favorable prognosis?
 - A. Lymphocyte predominant
 - B. Nodular Sclerosing
 - C. Mixed Cellularity
 - D. Lymphocyte depleted
- 7. Which of the following histologic subtypes of non-Hodgkin's lymphoma is not considered low grade according to the international working formulation (IWF)?
 - A. Small lymphocytic lymphoma (SLL)
 - B. Follicular small-cleaved cell (FSCCL)
 - C. Follicular mixed (small and large) cell (FML)
 - D. Follicular large cell (FLCL)

- 8. All of the following are TRUE about chronic lymphocytic leukemia (CLL) EXCEPT:
 - A. It is the most common form of leukemia in Caucasians.
 - B. CLL cells are indistinguishable from normal lymphocytes by their morphologic features under light microscopy.
 - C. It is commonly associated with hypogammaglobulinemia and increased risk of infection.
 - D. It can be cured using modern combination chemotherapy regimens.
- 9. Waldenstrom's macroglobulinemia (WM) is a closely related disease to multiple myeloma. Which of the following is the least common in WM?
 - A. Monoclonal immunoglobulin spike of IgM type in the serum.
 - B. Lymphadenopathy and hepatosplenomegaly.
 - C. Skeletal involvement producing lytic lesions
 - D. Lymphoplasmacytoid features of malignant cells
- 10. All of the following infections are associated with lymphocytosis EXCEPT:
 - A. Infectious mononucleosis
 - B. Acute infectious lymphocytosis
 - C. Infectious hepatitis
 - D. Acquired Immune Deficiency Syndrome (AIDS)
- 11. Glanzman's thrombasthenia is due to:
 - A. A decrease in platelet dense granules
 - B. A decrease in platelet alpha granules
 - C. Decreased platelet dense granule ADP content
 - D. A defect in the GP IIb/IIIa complex
 - E. Decreased platelet p-selectin

- 12. A bleeding time of 20 minutes (normal <8.5 minutes) may be due to all of the following EXCEPT:
 - A. von Willebrand's disease
 - B. Bernard Soulier syndrome
 - C. Hypofibrinogenemia
 - D. Platelet dense granule deficiency
 - E. Ingestion of 650 mg aspirin by a normal individual
- 13. All of the following are manifestations of bone marrow failure EXCEPT:
 - A. Anemia
 - B. Neutropenia
 - C. Thrombocytopenia
 - D. Pancytopenia
 - E. Jaundice
- 14. A 23 year old African-American female is referred to you for evaluation. She has had multiple hospital admissions for bacterial infections. She has had a CBC every Monday for the past 4 weeks. Her absolute neutrophil counts for the last 4 weeks have been 250/ul, 900/ul, 1900/ul and 600/ul, respectively. Her hemoglobin is 13.6 gm/dl and her platelets are 212,000/ul. The best diagnosis would be:
 - A. Leukocyte adhesion deficiency
 - B. Chronic granulomatous disease
 - C. Benign neutropenia of Blacks
 - D. Cyclic neutropenia
 - E. Acute myelogenous leukemia

- 15. A 68 year old woman is allergic to aspirin. She has been taking ticlopidine since she had a transient ischemic attack 6 weeks ago. She is found to have a urinary tract infection by her physician. Her CBC shows: WBC 1600/ul (PMN 10%, lymphs 90%), hemoglobin 14.1 gm/dl, hematocrit 41%. MCV 91 fl, and platelets 237,000/ul. Her white blood cell abnormality would best be explained by:
 - A. Chronic myelogenous leukemia
 - B. Anti-neutrophil antibodies
 - C. Cyclic neutropenia
 - D. Chronic lymphocytic leukemia
 - E. Absent neutrophil precursors in the bone marrow
- 16. An 8 year old girl has a history recurrent abscesses. There is no family history of infection. Her parents are first cousins. She has not had an infection for the past 2 months. Her current WBC is 45,000/ul(90% PMN, 10% lymphs). The best diagnostic test would be:
 - A. Flow cytometry for CD 11/CD 18
 - B. Look for giant inclusion granules on peripheral smear
 - C. NBT dye reduction test
 - D. Acute leukemia phenotyping
 - E. Blood culture
- 17. All of the following may be associated with eosinophilia EXCEPT:
 - A. Hodgkin's disease
 - B. Allergic rhinitis
 - C. Hypereosinphiic syndrome
 - D. Polyarteritis nodosa
 - E. Pinworm infestation

- 18. All of the following statements about acute promyelocytic leukemia are CORRECT EXCEPT:
 - A. It is always associated with a balanced translocation between chromosomes 15 and 17
 - B. Patients usually have a preceding history of chronic anemia
 - C. Hemorrhage is a serious complication of the leukemia
 - D. The disease is treatable with a derivative of Vitamin A
 - E. More than 50% of patients can be cured with appropriate treatment
- 19. Which one of the following statements about cytogenetic changes in patients with leukemia is CORRECT?
 - A. The chromosomal abnormalities can be detected in all cells in the body
 - B. They are an important prognostic indicator of the outcome of treatment
 - C. These genetic mutations specifically suppress the growth of normal cells
 - D. Similar findings are seen in patients with acute myelogenous leukemia and acute lymphoblastic leukemia
- 20. Which of the following patients is likely to have the best outcome following treatment with chemotherapy?
 - A. A 65 year old woman with prior refractory anemia which had transformed to acute myeloid leukemia
 - B. A 20 year old male with blast crisis of chronic myelogenous leukemia
 - C. A 10 year old girl with acute monocyte leukemia [FAB M5] which presented with disseminated skin lesions
 - D. A 10 year old boy with acute lymphocytic leukemia
 - E. A 30 year old man with AML who had received successful treatment with chemotherapy for Hodgkin's disease 7 years ago.

- 21. The Philadelphia chromosome [t(9;22)] is most likely to be found in which of the following patients with leukemia?
 - A. A 45 year old man with acute lymphocytic leukemia
 - B. 82 year old woman with chronic lymphocytic leukemia
 - C. A 67 year old with refractory anemia with excess blasts
 - D. A 24 year old woman with acute myeloid leukemia, FAB M2
 - E. A 7 year old boy with acute myeloid leukemia, FAB M4
- 22. Failure to achieve effective pain control is <u>least</u> likely due to:
 - A. Fear of patient addiction by the doctor
 - B. Fear of patient addiction by the patient
 - C. Use of inadequate doses of medication
 - D. Use of inappropriately long dosing intervals
 - E. Switching to an equianalgesic dose of another medication
- 23. The diagnosis of drug addiction is **best** made on the basis of:
 - A. Frequency of demands for pain medication
 - B. Pattern of medication used during consistent outpatient follow-up
 - C. The types of pain medications requested by the patient
 - D. Patients who ask for particular medications by name
 - E. Reports of family members

24. A patient is transferred to your care with chronic pain due to lung cancer metastatic to bone. She has been managed with Demerol 200 mg orally every 6 hours for the past week. Pain control has been only "fair" with the medication not lasting the full 6 hours. She also complains of frequent twitching and having to wake up at night, in pain, to take the medication. Which of the following would be the best initial regimen for pain control? (You will of course later adjust the medication for best efficacy while following the patient.)

Equianalgesic medication table.

<u>Medication</u>	Oral mg	Parenteral mg
Marabina	30	10
Morphine	5 0	10
Oxycodone	30	NA
Codeine	200	NA
Meperidine(Demer	col) 300	75

- A. Continue Demerol but increase dose to 300mg orally every 6 hours.
- B. Switch to (325 mg acetaminophen+30mg codeine) tablets, two every four hours.
- C. Switch to intravenous demerol at 10 mg per hour continuous infusion with demerol 25 mg orally every 3 hours for breakthrough pain.
- D. Switch to oral long acting morphine (MSContin, Oromorph or Kadian) 160 mg every 12 hours with 30 mg short acting morphine every 3 hours as needed for breakthrough pain.
- E. Switch to oral long acting oxycodone (Oxycontin) 30 mg every 12 hours with 15 mg short acting every 3 hours as needed for breakthrough pain.
- 25. The best determinant if a patient has pain is the opinion of the:
 - A. Doctor
 - B. Family
 - C. Nurse
 - D. Patient
 - E. Course director

- 26. Advantages of PCA (patient controlled analgesia) include all of the following EXCEPT:
 - A. Allowing patients to control the amount of medication given
 - B. Allowing shorter medication intervals than is practical for a nurse to deliver.
 - C. Decreasing delays between the time when a patient asks for and actually gets medication
 - D. Low cost of PCA machines
- 27. All of the following are TRUE of the naturally occurring antibodies to ABH antigens EXCEPT:
 - A. Most commonly they are IgG
 - B. Are present in most people lacking the corresponding antigen
 - C. Reach peak titers at 5 to 10 years of age
 - D. Activate complement to form the membrane attack complex
- 28. The clinical significance of the D antigen is due to:
 - A. The presence of the antibodies in all patients lacking the antigen
 - B. The ability of the IgG antibodies to cross the placenta and cause hemolytic disease of the newborn
 - C. The absence of an antibody response in over 60% of the antigen negative patients exposed to the antigen
 - D. The morphological abnormalities of the D antigen negative red blood cells

- 29. If in your experience 25% of the patients undergoing a surgical procedure may receive blood transfusions, the most appropriate preoperative blood bank order is:
 - A. Type and crossmatch 2 units of packed RBC
 - B. No blood bank orders are appropriate
 - C. Type and screen
 - D. Request the blood bank to hold 4 units of 0 neg blood
- 30. A 73 year old woman is evaluated for severe anemia and neurologic abnormalities. Her hemoglobin is 5.2 g/dl. She is tired but has not other complaints. Your diagnosis is vitamin B12 deficiency. Which of the following is TRUE about RBC transfusions to this patient?
 - A. The severe anemia should be corrected by rapid transfusion of 4u of packed RBC
 - B. No transfusions are necessary if she responds well to B12 treatment
 - C. Transfusions should be given as tolerated to increase the hemoglobin to 8g/dl
 - D. Transfusions must never be given when the anemia is chronic
- 31. The expected post transfusion hemoglobin following transfusion of 3 units of packed RBC to a non-bleeding adult with a pretransfusion hemoglobin of 6g/dl is approximately:
 - A. 7g/dl
 - B. 9g/dl
 - C. 12g/dl
 - D. 13g/dl

- 32. All of the following groups of patients should be transfused with irradiated blood EXCEPT:
 - A. Severe combined immunodeficiency
 - B. Recipients of bone marrow transplant
 - C. All children less than 6 months old
 - D. Immunocompetent recipients of blood donated by first degree relatives
- 33. A positive indirect antiglobulin test is MOST indicative of:
 - A. An autoimmune process
 - B. Anti-red cell antibodies on the surface of circulating RBC
 - C. Anti-red cell antibodies in the serum
 - D. Activated complement on the surface of the RBC
 - E. Activated complement in the serum
- While in the hospital a patient receives 2 u packed RBC to correct a symptomatic anemia of 7.5g/dl Hb. The post transfusion Hb is 9.7g/dl. You see him six days later in the clinic for routine follow up. The CBC shows a Hb of 7.4g/dl. The direct Coombs test is positive. The patient is not bleeding. What is the most likely explanation?
 - A. He has a congenital hemolytic anemia causing accelerated destruction of the transfused RBC.
 - B. He has a delayed hemolytic transfusion reaction due to an anamnestic antibody response.
 - C. The hemolysis is due to a primary exposure to a RBC antigen occurring six days before.
 - D. The transfused cells were near the expiration of their storage time and survived poorly.

- 35. During surgery a patient receives one unit of blood and immediately the urine in the Foley catheter becomes red-brown and there is blood oozing in the surgical field. What are you most likely going to discover when investigating the incident?
 - A. This was the first time the surgeon ever performed the surgical procedure.
 - B. The patient had a weakly reacting antibody to a low incidence antigen.
 - C. The patient was type O and the transfused unit was type A.
 - D. The patient had a history of excessive bleeding.
 - E. The patient was type A and the transfused unit was type O.
- 36. Of the following, which is the most common adverse reaction to transfusion?
 - A. Graft versus host reaction
 - B. Transfusion related acute lung injury
 - C. Anaphylaxis
 - D. Volume overload
- 37. All of the following hematopoietic growth factors are produced primarily by marrow and other stromal elements EXCEPT:
 - A. Stem cell factor
 - B. GM-CSF
 - C. Erythropoietin
 - D. IL-11
 - E. G-CSF

38.	Which of	the	following	growth	factors	has	an	established
	therapeut	cic 1	role?					

- A. IL-5
- B. IL-7
- C. IL-9
- D. G-CSF
- E. LIF (lymphocyte infiltrating factor)
- 39. The mechanism whereby a hematopoietic growth factor generally transmits its message to the target cell involves all of the following EXCEPT:
 - A. Receptor sub-unit rearrangement
 - B. Receptor mediated endocytosis
 - C. An intermediate kinase
 - D. Phosphorylation of a signal transducing activator of transcription (STAT)
 - E. STATs modulating transcription
- 40. Based upon the relative cellular lifespan, sudden onset marrow failure will usually manifest first with:
 - A. Anemia
 - B. Thrombocytopenia
 - C. Granulocytopenia
 - D. Lymphopenia
 - E. Polycythemia

- 41. Pancytopenia presenting in childhood which is associated with skeletal abnormalities and results, in most cases, from genetic abnormalities of the DNA repair mechanisms is most consistent with a diagnosis of:
 - A. Fanconi's anemia
 - B. Congenital infection
 - C. Thrombocytopenia with absent radii (TAR) syndrome
 - D. Blackfan Diamond syndrome
 - E. Congenital dyserythropoietic anemia
- 42. Which of the following statements about paroxysmal nocturnal hemoglobinuria (PNH) is CORRECT?
 - A. Results from an abnormality of the pig-a gene which affects only the granulocyte precursors.
 - B. Is associated with an abnormal antibody detectable by the Donath Lansteiner test.
 - C. Is not associated with an increased risk of developing acute myeloid leukemia.
 - D. Frequently accompanies tertiary syphilis
 - E. Is associated with an increased frequency of vascular thrombosis.
- 43. A 20 year old African-American male has sickle cell anemia (homozygous SS). All of the following are consistent with his diagnosis EXCEPT:
 - A. Positive hemoglobin solubility test
 - B. Hemoglobin level of 8 gm%
 - C. Hemoglobin S concentration of 70% on electrophoresis
 - D. History of painful swollen hands or feet (dactylitis)
 - E. Substitution of valine for glutamine acid at position 6 of the beta globin chain.

- 44. Hemoglobin E is most commonly seen in individuals of which ancestry?
 - A. African
 - B. Southeast Asian
 - C. Northern European
 - D. Native American
 - E. Mediterranean
- 45. Regarding the alpha globin gene, which of the following is CORRECT?
 - A. The presence of 0 functional alpha globin genes is associated with hemoglobin H disease
 - B. The alpha globin gene is on the same chromosome as the beta globin gene
 - C. alpha globin gene defects are manifest at birth
 - D. Deletion of all alpha globin genes results in thalassemia major presenting at about 3 to 4 months of age
 - E. The production of alpha-globin chains is normally twice that of beta-globin chains
- 46. Which of the following is the most likely diagnosis in a child with congenital cyanosis?
 - A. Unstable hemoglobin
 - B. Hemoglobin E
 - C. High O_2 affinity hemoglobin
 - D. Hemoglobins M
 - E. Glucose-6-phosphate dehydrogenase deficiency

- 47. In homozygous beta thalassemia, all of the following are TRUE EXCEPT:
 - A. Mutations have been described in both introns as well as exons
 - B. Hematological and clinical manifestations are seen at birth
 - C. The earliest manifestation is anemia
 - D. Red cell indices are hypochromic and microcytic
 - E. Hepato-splenomegaly is frequent
- 48. Among the anemias caused by deficiency states, which is least likely to be due to poor dietary habits?
 - A. Copper deficiency
 - B. Iron deficiency
 - C. B_{12} deficiency
 - D. Folic acid deficiency
- 49. A patient is felt to have a megaloblastic anemia. Which of the following on physical exam will help distinguish B_{12} from folate deficiency?
 - A. Pale skin and "beefy" red tongue
 - B. Stocking glove parasthesias
 - C. Loss of vibratory sensation
 - D. Rales on examination of the lungs
- 50. Transfusional hemosiderosis (iron overload) is manifested by all of the following EXCEPT:
 - A. Cardiomyopathy
 - B. Diabetes mellitus
 - C. Renal failure
 - D. Growth failure
 - E. Cirrhosis of the liver

- 51. In which of the states listed below will iron absorption be the best?
 - A. Iron remains in ferric (Fe⁺³) state
 - B. Oral iron taken with tea
 - C. In patients with gastric resections
 - D. Oral iron taken with orange juice
- 52. For both vitamin B_{12} and foliate deficiency:
 - A. Dietary deficiency is the primary cause
 - B. Macroovalocytes and hypersegmented neutrophils are seen
 - C. Pregnancy decreases daily requirements
 - D. Neurologic sequella mandate early diagnosis
- 53. Most hereditary cancer predisposition syndromes are caused by mutations in:
 - A. Dominant proto-oncogenes
 - B. Growth factor genes
 - C. Cell adhesion molecule genes
 - D. Tumor suppressor genes
 - E. Nuclear matrix genes
- 54. Which approach is $\underline{\text{NOT}}$ a theoretically useful anti-cancer therapy?
 - A. Inhibition of autocrine growth factors
 - B. Inhibition of angiogenic factors
 - C. Activation of proto-oncogene function
 - D. Reconstitution of tumor suppressor gene function
 - E. Inhibition of protease activity

- 55. An 8 year old Nigerian boy presents with a large painful mass over his left jaw. A biopsy reveals Burkitt's lymphoma. What genetic abnormality will most likely be found in this boy's tumor?
 - A. Homozygous deletion of the rb gene
 - B. Translocation of chromosomes 9 and 22 with *bcr-abl* expression
 - C. Loss of heterozygosity of the NF1 gene locus
 - D. Translocation of chromosomes 8 and 14 with c-myc over expression
 - E. Amplification of the WT1 gene
- 56. Which clinical effect can be explained by tumor cell heterogeneity?
 - A. The recurrence of acute myelogenous leukemia six months after apparent complete remission
 - B. The low incidence of nausea after cis-platinum therapy in alcoholics
 - C. The development of breast cancer after mantle-zone radiation for Hodgkin's disease
 - D. The induction of melanoma by repeated exposure to UV light
 - E. Development of pulmonary fibrosis after bleomycin therapy
- 57. Which statement regarding carcinogenesis is NOT TRUE?
 - A. Initiation is an irreversible event
 - B. Progression can involve the development of drug resistance
 - C. Promotion is usually a single, genotoxic event
 - D. Most initiators are chemical or physical agents
 - E. Promotion is initially a reversible process

- 58. Type I von Willebrand disease is disease is characterized by all of the following EXCEPT:
 - A. Nose and gum bleeding
 - B. Delayed bleeding into joints, muscles, and central nervous system
 - C. Symptoms similar to those associated with thrombocytopenia
 - D. Prolonged bleeding time
 - E. Autosomal dominant inheritance
- 59. All of the following are True about Factor XII, prekallikrein, and high molecular weight kinogen(HMWK) EXCEPT:
 - A. They are frequently implicated in inherited bleeding disorders
 - B. They are apparently unnecessary in normal human hemostasis
 - C. Deficiency states may prolong the APTT
 - D. May have utility in mechanisms of inflammation, immunologic defense, and anaphylaxis
 - E. Are not screened for in the PT

60. A 45 year old male patient with sickle cell anemia develops a fever, shaking chills, and severe malaise and when examined in the Harper Hospital Urgent Care Center, he has a T of 104.3, pulse of 130, blood pressure of 80/45 mm Hg, and respiratory rate of 24/minute. Over the next several hours, the patient begins to exhibit severe bruising under the blood pressure cuff, bleeding at venepuncture sites, from his nose, and into his urine. Laboratory studies reveal the following:

		Normal range
Platelet count	14,000/ul	150,000 - 400,000/ul
Prothrombin time	15 sec	10.5 - 12.0 sec
Activated partial		
thromboplastin time	45 sec	20 - 34 sec
Thrombin time	25 sec	10 - 15 sec
Fibrinogen	60 mg/dl	150 - 345 mg/dl
Fibrin D-dimer	2 - 4ug/ml	< 0.25 ug/ml

The best interpretation of the fibrin D-dimer test in this patient's plasma is that:

- A. It suggests a high plasma fibrinogen concentration
- B. It indicates the presence of intravascular coagulation and fibrinolysis
- C. It indicates that the patient's fibrinolytic system has been inappropriately activated
- D. It demonstrates that fibrinogen cannot be converted to fibrin
- E. It implies a decrease in Factor XIII activity
- 61. An elevated erythropoietin level is least likely to be found in:
 - A. Aplastic anemia
 - B. Dyserythropoietic anemia
 - C. Iron deficiency anemia
 - D. Polycythemia vera
 - E. Secondary polycythemia

- 62. All of the following contribute to the pathogenesis of the myeloproliferative disorders EXCEPT:
 - A. Increased growth factor production
 - B. Decreased apoptosis
 - C. Decreased thrombopoietin receptors
 - D. Polyclonal fibroblast proliferation
- 63. A 63 year old woman comes to your clinic at Harper Hospital on February 28, 1998. She has had morning headaches and trouble concentrating for the past 2 months. On exam, she is plethoric. Her spleen is not palpable. Her drivers license was renewed in October 1997, and her color in the picture was normal. Her CBC shows WBC 7,000/ul, Hgb 19.3, Hematocrit 58% and platelets 225,000/ul. The test most likely to establish the diagnosis is:
 - A. Serum vitamin B12
 - B. Carbonmonoxyhemoglobin level
 - C. Leukocyte alkaline phosphatase
 - D. RBC survival study
 - E. Serum erythropoietin level
- 64. The diagnosis of essential thrombocythemia is established by which of the following tests:
 - A. Impaired 1st wave of aggregation
 - B. Prolonged bleeding time
 - C. Serum ferritin
 - D. Serum thrombopoietin level
 - E. None of the above

- 65. The development of acute leukemia may occur as part of the natural history of all of the following EXCEPT:
 - A. Polycythemia vera
 - B. Chronic myelogenous leukemia
 - C. Myelofibrosis
 - D. Myelodysplasia
 - E. Acute leukemia may develop in all of the above conditions
- 66. A 72 year old man complains of fatigue and easy bruising. His spleen is not palpable. His CBC is: WBC 2,700/ul(80% lymphs, 20% PMN), hemoglobin 7.3 gm/dl, MCV 98 fl and platelets 46,000/ul. His bone marrow is hypercellular with 18% blasts. The blasts stain with myeloperoxidase and Sudan Black B. The best diagnosis is:
 - A. Acute myelogenous leukemia
 - B. Acute lymphoblastic leukemia
 - C. Myelodysplastic syndrome
 - D. Chronic myelogenous leukemia
 - E. Thrombotic thrombocytopenic purpura
- 67. The serum LDH may be elevated in all of the following EXCEPT:
 - A. Autoimmune hemolytic anemia
 - B. Iron deficiency anemia
 - C. Pernicious anemia
 - D. Acute lymphoblastic leukemia
 - E. Reabsorption of a large retroperitoneal hematoma

- 68. "Loss of function" in the hemostatic protein, Factor VII, would likely result in:
 - A. Prolongation of the thrombin time (TT)
 - B. Prolongation of the activated partial thromboplastin time (APTT)
 - C. A clinical thrombotic disorder
 - D. A clinical bleeding disorder
 - E. None of the above
- 69. A 20 year old African-American female has been rejected as a blood donor because she is anemic. Her CBC is: WBC 7,600/ul (75% PMN, 25% lymphs) hemoglobin 11.8 gm/dl, RBC 5.3 million/ul, MCV 70 fl, RDW 14% and platelets 227,000/ul. The most likely diagnosis is:
 - A. Iron deficiency anemia
 - B. Autoimmune hemolytic anemia
 - C. Thalassemia minor
 - D. Glucose-6-phosphate dehydrogenase deficiency
 - E. Sickle cell trait
- 70. A 48 year old female has the following CBC: WBC 6,800/ul, hemoglobin 5.6 gm/dl, hematocrit 16%, platelets 325,000/ul, and reticulocytes 8%. The peripheral smear shows polychromasia. The anemia is most likely due to:
 - A. Increased RBC destruction
 - B. Decreased RBC production
 - C. Hydrops fetalis
 - D. None of the above

- 71. Intrinsic factor is necessary for the absorption of:
 - A. Folic acid
 - B. Iron
 - C. Vitamin B_6
 - D. Vitamin B_{12}
 - E. Hydrochloric acid
- 72. The lymphocytes in chronic lymphocytic anemia are most commonly of this origin:
 - A. B-cell (monoclonal)
 - B. T-cell (momoclonal)
 - C. Natural killer cell
 - D. B-cell (polyclonal)
 - E. T-cell (polyclonal)

- 73. A 28 year old female sees her physician because of fatigue. On physical exam she is jaundiced and her spleen is palpable 3 cm below the left costal margin. Her initial CBC shows: WBC 7,000/ul, hemoglobin 8.4 gm/dl, hematocrit 25%, platelets 338,000/ul. All of the following studies would be appropriate to initially evaluate the anemia EXCEPT:
 - A. Bone marrow aspiration and biopsy
 - B. Reticulocyte count
 - C. MCV
 - D. RDW
 - E. Peripheral smear

QUESTIONS 74-92 ARE TO BE ANSWERED USING THE PHOTOGRAPH PROVIDED.

- 74. A 24 year old male is found to have gallstones. His physical examination shows scleral icterus. His spleen is palpable 5 cm below the left costal margin. His initial CBC shows: WBC 6,000/ul, hemoglobin 12.2 gm/dl, hematocrit 33%. MCV 84 fl, MCHC 37 gm/dl and platelets 187,000/ul. His peripheral smear is shown in Figure 1. His anemia is most likely due to:
 - A. RBC spectrin dysfunction
 - B. RBC spectrin deficiency
 - C. A dysfunctional prosthetic heart valve
 - D. Cirrhosis
 - E. Renal failure
- 75. A previously healthy 4 year old male is brought to the emergency room by his parents. Today, two hours after eating fava beans for the first time he passed red urine and became lethargic. He is now unarousable. His CBC shows: WBC 14,000/ul, hemoglobin 3.8 gm/dl, hematocrit 11%. MCV 84 fl, platelets 298,000/ul. His peripheral smear is shown in **Figure 2**. All of the following would be TRUE EXCEPT:
 - A. Supravital staining would demonstrate Heinz bodies in the RBC
 - B. He is of Mediterranean ancestry
 - C. He has a positive direct antiglobulin (Coombs') test
 - D. His CBC was normal yesterday

- 76. A 10 year old girl is brought to the emergency room by her parents for severe nausea, vomiting and diarrhea. 14 hours earlier she had eaten a hamburger at a fast food restaurant. Her initial laboratory evaluation shows: WBC 11,200/ul(80% PMN, 20% lymphs), hemoglobin 5.2 gm/dl, hematocrit 15%, MCV 89 fl, and platelets 4,000/ul. Her creatinine is 7.6 mg/dl (normal < 1.2), PT 12.2 sec (normal < 13.1) and aPTT 28.2 sec (normal <35). Her peripheral smear is shown in Figure 3. The best diagnosis is:
 - A. Disseminated intravascular coagulation secondary to E. coli septicemia
 - B. Immune thrombocytopenic purpura
 - C. Autoimmune hemolytic anemia
 - D. Acute lymphoblastic leukemia
 - E. Hemolytic uremic syndrome

<u>DIRECTIONS</u>: Match the most appropriate diagnostic test (A-H below) with the peripheral smear numbered 77-79.

- A. Osmotic fragility test
- B. Heinz body prep
- C. Sickle cell prep
- D. Vitamin B12 level
- E. Serum Ferritin
- F. Creatinine
- G. Leukemia Phenotyping
- H. None of the above
- 77. Figure 4
- 78. Figure 5
- 79. Figure 6

DIRECTIONS:

Match the appropriate clinical case (A-E below) with the bone marrow numbered in 80-82.

- A. An 18 year old Mexican female who has been taking chloramphenical for a urinary tract infection.
- B. A 45 year old male with newly diagnosed splenomegaly. His CBC is WBC 36,000/ul (60% PMN, 15% bands, 7% metamyelocytes, 7% myelocytes, 1% promyelocytes, 4% eosinophils, 6% lymphocytes), hemoglobin 11.8 gm/dl, and platelets 524,000/ul.
- C. A 5 year old child with lymphadenopathy and splenomegaly. His CBC is WBC 25,000/ul, hemoglobin 5.4 gm/dl and platelets 13,000/ul.
- D. A 32 year old alcoholic with miliary tuberculosis.
- E. A 54 year of female with WBC 7,200/ul (PMN 80%, lymphocytes 20%), hemoglobin 7.1 gm/dl, hematocrit 21%, reticulocytes 18% and platelets 219,000. Her direct and indirect antiglobulin test are positive.
- 80. Figure 7
- 81. Figure 8
- 82. Figure 9

- 83. **Figure 10** illustrates the peripheral blood smear of a 53 year old man with generalized lymphadenopathy, mild splenomegaly and elevated WBC of one year duration. The hemoglobin is 12g/dl. What is the most likely diagnosis?
 - A. Chronic myelogenous leukemia and anemia
 - B. Acute lymphoblastic leukemia and iron deficiency
 - C. Chronic lymphocytic leukemia and elliptocytosis
 - D. Monocytosis and anemia of chronic inflammation
- 84. The peripheral blood morphology in **Figure 11** may be associated with any of the following disorders EXCEPT:
 - A. Chronic myelogenous leukemia
 - B. Essential thrombocythemia
 - C. Polycythemia vera
 - D. Refractory anemia with ring sideroblasts
- 85. The cell shown in Figure 12 is:
 - A. A leukemic myeloblast
 - B. A leukemic lymphoblast
 - C. A monocyte
 - D. An atypical lymphocyte
- 86. The peripheral blood smear in **Figure 13** is most likely seen in a patient with:
 - A. Congenital hypofibrinogenemia
 - B. A myelodysplastic syndrome
 - C. Acute appendicitis
 - D. Multiple myeloma
 - E. Decreased spectrin in the RBCs

- 87. Which statement is TRUE about the bone marrow biopsy in **Figure** 14?
 - A. May represent the end stage of several chronic myeloproliferative disorders.
 - B. It shows metastatic carcinomas
 - C. It is usually accompanied by a normal CBC
 - D. It is secondary to massive splenomegaly
- 88. **Figure 15** is a peripheral blood smear stained with cresyl violet. Based on your interpretation of the stain which statement is TRUE of the patient's anemia?
 - A. It is most likely due to bone marrow failure
 - B. The anemia is accompanied by reticulocytosis
 - C. These cells are not seen in patients with hemolytic anemia
 - D. Untreated, severe folate deficiency is the most likely cause.
- 89. Which of the following is most likely TRUE regarding the lymph node biopsy illustrated in **Figure 16**?
 - A. The lymph node is normal
 - B. The changes are typical of nodular sclerosing Hodgkin's disease.
 - C. Monoclonal proliferations with this pattern consist of T cells.
 - D. Monoclonal proliferations with this pattern consist of B cells.

- 90. All of the following are TRUE regarding both white blood cells in Figure 17 EXCEPT:
 - A. May be increased in infections
 - B. Are involved in removing bacteria
 - C. May be increased in stem cell disorders
 - D. Are immune accessory cells
- 91. This bone marrow section is stained with Prussian Blue stain. Based on the findings in **Figure 18**, the patient most likely:
 - A. Was transfused more than 100 units of RBC
 - B. Has absent iron stores
 - C. Has red blood cells with Pappenheimer's bodies
 - D. Has anemia of chronic inflammation
- 92. The PMN illustrated in **Figure 19** is most commonly associated with:
 - A. Neutrophilia with left shift
 - B. Refractory anemia with excess blasts
 - C. Vitamin B12 deficiency
 - D. Massive splenomegaly

DIRECTIONS:

Select the option (A-E below) which is most closely related to the descriptions numbered 93-97. Each alternative may be used once, more than once, or not at all.

- A. Tissue Factor
- B. Homocyst(e)ine
- C. Factor V
- D. Factor VIII
- E. von Willebrand Factor
- 93. A common mutation in this large procoagulant protein may predispose a patient to develop thrombosis.
- 94. This large protein is deficient or exhibits a loss of function in hemophilia A.
- 95. Synthesized by vascular endothelial cells and promotes platelet adhesion to damaged or activated blood vessel walls.
- 96. Accumulates under conditions of enzyme deficiency or vitamin deficiency, causing venous or arterial thrombosis.
- 97. The transport vehicle for the procoagulant protein which is missing or defective in hemophilia A.

DIRECTIONS: Select the set of coagulation studies (A-E below) which best corresponds to the diagnoses numbered $\underline{98}$ - $\underline{100}$.

Normal Range	PT 10-13.1 sec	aPTT 20-35 sec	aPTT Mixing Study < 35 sec
Α.	21.2	42.1	39.3
В.	75	24	Not indicated
C.	10.9	58.8	34.0
D.	11.1	32.3	Not indicated
Ε.	11.3	62.2	52.1

- 98. Factor VII deficiency
- 99. Hereditary factor XI deficiency
- 100. Hemophilia A with an acquired Factor VIII inhibitor