Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information



Hematopoiesis: Questions 1–14 3 CBC, Peripheral Smear and Bone Marrow Examination: Questions 15–40 4 Anemias: Questions 41–64 6 Hemolytic Anemias: Questions 65–84 8 Hemoglobinopathies: Questions 85–109 10

Answers to Chapter 1 14 Answers to Chapter 1 (with Explanations) 15

2 Non-Neoplastic WBC and Platelet Disorders 32 WBC and Platelet Disorders: Questions 1–15 32

Answers to Chapter 2 34 Answers to Chapter 2 (with Explanations) 35 Benign Lymph Node 38
 Benign Lymph Node: Questions 1–16 38

Answers to Chapter 3 40 Answers to Chapter 3 (with Explanations) 41

Chapter

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

# **RBC Disorders**

#### Hematopoiesis: Questions 1–14

- 1. The following are features of normal erythropoiesis except:
  - A. Immature erythropoietic cells tend to form clusters which surround macrophages
  - B. Proerythroblasts are large cells with deep blue cytoplasm with large nuclei and nucleoli
  - C. Erythropoiesis occurs predominantly in the paratrabecular areas
  - D. Reticulocytes typically have a three-day life span, of which two days are spent in the bone marrow
- 2. Which of the following is an effective marker for erythroid precursors?
  - A. CD34
  - B. CD71
  - C. CD41
  - D. CD61
- 3. Which of the following step is the rate-limiting step for heme biosynthesis?
  - A. Formation of  $\delta$  aminolevulinic acid from glycine and succinyl CoA
  - B. Formation of prophobilinogen from  $\delta$  aminolevulinic acid
  - C. Formation of heme from protoporphyrin IX
  - D. None of the above
- 4. Regarding granulopoiesis, which is true:
  - A. Secondary granules are seen only at the band and mature granulocyte stage
  - B. Blasts, promyelocytes and myelocytes all have nucleoli
  - C. Granulopoiesis in a normal marrow is seen adjacent to bone
  - D. Metamyelocytes are capable of cell division

- 5. Regarding erythropoiesis, which is true:
  - A. All normoblasts have nucleoli
  - B. The basophilic normoblast is the largest cell
  - C. Erythropoiesis occurs in islands close to the bony trabecula
  - D. The normal sequence of maturation is proerythroblast to basophilic normoblast to polychromatic normoblast to orthochromatic normoblast to reticulocyte to a mature red cell
- 6. Regarding megakaryopoiesis and megakaryocytes, which is true:
  - A. Normal development of megakaryocytes is regulated by multiple cytokines and one principal one is thrombopoietin
  - B. Clustering of megakaryocytes is a normal phenomenon
  - C. Megakaryocytes mature through endomitosis where cell division with nuclear division occurs
  - D. Megakaryocytes may cause destruction of hematopoietic cells by emperipolesis
- 7. Which of the following cell is derived from a hematopoietic stem cell?
  - A. Osteoblasts
  - B. Osteocytes
  - C. Chondrocytes
  - D. Osteoclasts
- 8. Features characteristic of erythroid precursors include all of the following except:
  - A. In a normal marrow they are found as distinctive islands
  - B. They adhere tightly to one another
  - C. Cytoplasm is deep blue in the erythroblasts
  - D. Shrinkage artifact of cytoplasm is seen in plastic-embedded sections

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

#### Section 1: Non-Neoplastic Hematology

- 9. Regarding granulopoiesis:
  - A. Both myelocytes and metamyelocytes are capable of cell division
  - B. By light microscopy, promyelocytes of the three granulocytic lineages can easily be distinguished
  - C. Myeloblasts are typically found in the paratrabecular areas and close to arterioles
  - D. Indented myeloid cells are known as bands
- 10. Regarding megakaryopoiesis, which is true:
  - A. Megakaryocytes are normally found close to bony trabeculae
  - B. Normal megakaryocytes engulf and destroy hematopoietic cells
  - C. Megakaryocytes undergo endomitosis (aka endoreduplication)
  - D. Normal megakaryocyte nuclei have multiple distinct lobes
- 11. Which agents are responsible for the stimulation of eosinophils?
  - A. IL-3
  - B. IL-5
  - C. GM-CSF
  - D. All of the above
- 12. Secondary granules of neutrophils:
  - A. Contain lactoferrin
  - B. Are also known as azurophilic granules
  - C. Contain peroxidase
  - D. Are found in immature granulocytic cells
- 13. All of the following regarding erythropoiesis are true except:
  - A. The life span of a mature erythrocyte is about four months
  - B. On average each erythroblast forms eight reticulocytes
  - C. In acute anemia, when accelerated erythropoiesis occurs, the red cells that are released into the circulation are normocytic and all bear the i antigen
  - D. The mean time from the proerythroblast stage to reticulocyte is approximately five days
- 14. Regarding megakaryopoiesis and megakaryocytes:
  - A. Megakaryocytes are approximately 10% of hematopoietic cells in a normal bone marrow

- B. Each megakaryocyte gives rise to 1,000–3,000 platelets
- C. Mature megakaryocytes contain 2N–128 N of DNA
- D. Mature megakaryocytes have little basophilic cytoplasm

#### CBC, Peripheral Smear and Bone Marrow Examination: Questions 15–40

- 15. When compared to normal adult CBC values, normal values for neonates and children differ considerably with the exception of:
  - A. RBC count
  - B. WBC count
  - C. Platelet count
  - D. Hemoglobin level
- 16. The following are true for hemoglobin measurement by automated analyzers:
  - A. The red cells are required to be lysed
  - B. The spectrophotometric method is typically used
  - C. Absorbance of light at 540 nm is measured
  - D. All of the above
- 17. Which of the following is a true statement?
  - A. In oxyhemoglobin, iron is in the ferrous state
  - B. In methemoglobin, iron is in the ferrous state
  - C. In both oxyhemoglobin and methemoglobin, iron is in the ferrous state
  - D. None of the above
- Causes of falsely elevated hemoglobin levels by the spectrophotometric method include all of the following except:
  - A. Leukocytosis
  - B. Paraproteinemia
  - C. Hyperlipidemia
  - D. Thrombocytosis
- 19. Which of the following equations is correct to calculate MCH values?
  - A. MCH= Hb/Hct
  - B. MCH=Hb/RBC count
  - C. MCH=Hb/MCV
  - D. None of the above

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

Chapter 1: RBC Disorders

- 20. One indication for warming the blood and repeating the CBC test is elevated:
  - A. Hemoglobin
  - B. Hct
  - C. MCH
  - D. MCHC
- 21. When the uncorrected WBC count is higher than the corrected WBC count, the most likely cause is:
  - A. Presence of nucleated RBCs
  - B. Presence of large platelets
  - C. High lymphocyte count
  - D. Presence of paraproteinemia
- 22. Causes of false low platelet count (pseudothrombocytopenia) include all of the following except:
  - A. EDTA-induced platelet clumps
  - B. EDTA-induced platelet satellitism
  - C. Fragmented red cells
  - D. Traumatic venepuncture
- 23. When a peripheral smear slide appears blue to the naked eye, a likely possibility is:
  - A. Underlying chronic myeloproliferative disorder
  - B. Underlying acute leukemia
  - C. Underlying hemolytic anemia
  - D. Underlying monoclonal gammopathy
- 24. A bone marrow exam reveals increased promyelocytes with paucity of more mature cells. Differential diagnosis includes:
  - A. Acute promyelocytic leukemia
  - B. Kostmann's syndrome
  - C. Sepsis in a patient with agranulocytosis
  - D. All of the above
- 25. The following are established causes of bone marrow granulomas:
  - A. Hodgkin lymphoma
  - B. Metastatic disease
  - C. Histoplasmosis
  - D. All of the above
- 26. Macrophages with "wrinkled tissue paper" appearance of the cytoplasm may be seen in all of the following conditions except:
  - A. Gaucher's disease
  - B. Niemann-Pick disease

- C. Chronic myelogenous leukemia
- D. Sickle cell disease
- 27. Characteristic features of sea blue histicoytes that may be seen in bone marrow include:
  - A. Yellow-brown macrophages with Giemsa stain
  - B. Blue-green appearance with H&E stain
  - C. They are macrophages containing ceroid
  - D. They are PAS negative
- 28. Causes of a dry tap when performing a bone marrow aspiration may be expected in:
  - A. Iron deficiency anemia
  - B. Refractory anemia with ringed sideroblasts
  - C. Hairy cell leukemia
  - D. Hereditary spherocytosis
- 29. Regarding lymphoid infiltrates in the bone marrow:
  - A. Bone marrow infiltration is seen more often in B-cell lymphomas than T-cell lymphomas
  - B. Bone marrow infiltration is seen more often in high-grade B-cell lymphomas than lowgrade B-cell lymphomas
  - C. Paratrabecular lymphoid aggregates are almost always reactive
  - D. Diffuse involvement of the bone marrow in a case of CLL imparts a favorable prognosis
- 30. Regarding iron stain on bone marrow aspirate slides, all of the following are true except:
  - A. Normal erythroblasts have one to five ironcontaining granules, distributed randomly within the cytoplasm of erythroblasts
  - B. Both intracellular and extracellular iron should be assessed
  - C. In healthy individuals, 20–50% of erythroblasts are sideroblasts
  - D. In individuals with hemochromatosis, plasma cells in bone marrow may demonstrate hemosiderin granules
- 31. Lipid granulomas of the bone marrow:
  - A. Are clinically significant
  - B. Are typically located in paratrabecular areas
  - C. Consist of fat vacuoles which are larger than marrow fat cells
  - D. May have giant cells

5

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

#### Section 1: Non-Neoplastic Hematology

- 32. Reactive or benign lymphoid aggregates may be seen in bone marrows:
  - A. In states of infection
  - B. In senior citizens
  - C. In rheumatoid arthritis
  - D. All of the above
- 33. Regarding reactive lymphoid aggregates in bone marrows, all are true except:
  - A. They are usually few in number
  - B. They are typically located in the paratrabecular areas
  - C. They consist predominantly of small mature lymphocytes
  - D. The lymphocytes exhibit pleomorphism
- 34. Bone marrow necrosis may be seen in:
  - A. Sickle cell disease
  - B. Acute leukemias
  - C. Chronic leukemias
  - D. All of the above
- 35. Regarding gelatinous transformation of the bone marrow:
  - A. There is replacement of hematopoietic elements by fat spaces
  - B. May be highlighted by Perl's stain
  - C. May be seen in individuals with anorexia nervosa
  - D. The peripheral blood will demonstrate leukocytosis
- 36. In adults, bone marrow metastasis from the following primary tumors are common except:
  - A. Prostatic carcinoma
  - B. Meningioma
  - C. Bronchogenic carcinoma
  - D. Breast cancer
- Peripheral blood findings most characteristic of metastatic tumor to the bone marrow is:
  - A. Leukoerythroblastic blood picture and presence of tear drop cells
  - B. Microcytic hypochromic red cells
  - C. Macrocytic anemia
  - D. Leukocytosis
- 38. Effect of EDTA on platelets include:
  - A. Decrease in MPV
  - B. Increase in MPV

- C. Loss of alpha granules
- D. Loss of  $\delta$  granules
- 39. Regarding reticulocytes:
  - A. Normal levels in newborns are 1%
  - B. Are raised in aplastic anemia
  - C. Contain a network of DNA in their cytoplasm
  - D. Contain ribonucleoprotein in their cytoplasm
- 40. Regarding the zeta potential of red cells:
  - A. The zeta potential of red cells is the negative charge around red cells that prevent them from aggregating
  - B. The zeta potential of red cells is the positive charge around red cells that prevent them from aggregating
  - C. Agents that increase the zeta potential will result in increased ESR
  - D. EDTA increases the zeta potential of red cells

#### Anemias: Questions 41–64

- 41. The following are true about reticulocytes except:
  - A. They are young red cells and contain ribosomal RNA
  - B. Reticulocyte count is traditionally expressed as a percentage
  - C. Reticulocyte count is low in bone marrow failure patients
  - D. Reticulocyte count is low in hemolytic anemia patients
- 42. The following are all included in the differential diagnosis of microcytic hypochromic anemia except:
  - A. HbE disease
  - B.  $\beta$  thalassemia trait
  - C. Iron deficiency
  - D. HbS disease
- 43. In a patient with β thalassemia trait, all of the following features are typically observed except:
  - A. Significantly high RDW
  - B. Mild microcytic hypochromic anemia
  - C. Target cells on the peripheral smear
  - D. Disproportionately elevated RBC count

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt <u>More Information</u>

#### Chapter 1: RBC Disorders

- 44. In a patient with iron deficiency, which of the following features are an expected finding:
  - A. Minimal anisocytosis
  - B. Abundant target cells
  - C. Reduced transferrin iron binding capacity
  - D. Elevated levels of free erythrocyte protoporphyrin (FEP)
- 45. Peripheral smear and bone marrow findings in iron deficiency anemia include:
  - A. Abundant target cells
  - B. Thrombocytopenia
  - C. Hypocellular marrow
  - D. Presence of scanty, ragged vacuolated cytoplasm in the developing erythroblasts
- 46. Which of the following is a true statement regarding sideroblastic anemia?
  - A. Sideroblastic anemia may be inherited or acquired, with the inherited form being more common
  - B. In sideroblastic anemia there occurs impaired iron utilization, and excess iron is found in the Golgi apparatus
  - C. Siderocytes and sideroblasts are abnormal and seen in sideroblastic anemia
  - D. Acquired sideroblastic anemia may be idiopathic (primary) which is part of myelodysplastic syndrome
- 47. In congenital sideroblastic anemia:
  - A. The red cells are most often macrocytic
  - B. Is a common inherited condition
  - C. In most families is transmitted as X-linked inheritance
  - D. Hemoglobin electrophoresis usually demonstrates an abnormal hemoglobin
- 48. In lead poisoning, coarse basophilic stippling is seen and this is due to:
  - A. Inhibition of the enzyme 5' pyrimidine nucleotidase
  - B. Inhibition of the enzyme ferrochelatase
  - C. Inhibition of the enzyme  $\delta$  aminolevulinic acid dehydratase
  - D. All of the above

- 49. Anemia of chronic disease is characterized by all of these except:
  - A. Low levels of serum iron
  - B. Reduced transferrin saturation
  - C. Reduced iron binding capacity
  - D. Reduced serum ferritin
- 50. Causes of macrocytic anemia include all of the following except:
  - A. Acute blood loss
  - B. Folate deficiency
  - C. B12 deficiency
  - D. Excess alcohol consumption
- 51. All of the following are established causes of B12 deficiency except:
  - A. Diet consisting of goat milk
  - B. Crohn's disease
  - C. Pernicious anemia
  - D. Fish tapeworm infestation
- 52. A 30-year-old male with long-standing history of diarrhea, has macrocytosis, Howell-Jolly bodies in RBC, target cells and small shrunken spleen. The most likely diagnosis is:
  - A. Sickle cell disease
  - B. Celiac disease
  - C. Crohn's disease
  - D. Liver disease
- 53. Bone marrow findings in megaloblastic anemia include all of the following except:
  - A. Nuclear cytoplasmic asynchrony of developing erythroid cells
  - B. Giant metamyelocytes
  - C. Hypersegmented megakaryocytes
  - D. Absent stainable iron in the bone marrow
- 54. A patient with known sickle cell disease registers with a new physician. CBC ordered by the patient's new physician demonstrates macrocytosis of the red blood cells. The most likely explanation is:
  - A. Iron deficiency
  - B. B12 deficiency
  - C. Folate deficiency
  - D. Patient is on hydroxyurea

7

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

#### Section 1: Non-Neoplastic Hematology

- 55. In pernicious anemia, all of the following are true except:
  - A. Patients may develop antiparietal cell and anti-intrinsic factor antibodies
  - B. Patients are at risk of developing gastric lymphoma
  - C. B12 levels are low
  - D. Patients may develop neurological symptoms
- 56. Iron deficiency may result in the misdiagnosis of:
  - A. Sickle cell trait
  - B. Sickle cell disease
  - C.  $\beta$  thalassemia trait
  - D.  $\beta$  thalassemia disease
- 57. In the human body, iron is found in the largest amount in:
  - A. Macrophages of the reticuloendothelial system
  - B. Hemoglobin
  - C. Myoglobin
  - D. Enzymes
- 58. The total daily loss of iron in an adult male is approximately:
  - A. 1 mg
  - B. 2 mg
  - C. 3 mg
  - D. 4 mg
- 59. Vitamin B12
  - A. Is directly required for DNA synthesis
  - B. If bound to transcobalamin I is delivered to the tissues
  - C. Is needed to convert methyl tetrahydrofolate (THF) to THF
  - D. Is absorbed in the duodenum
- 60. All of the following are causes of megaloblastoid changes in the bone marrow except:
  - A. B12 deficiency
  - B. Hydroxyurea treatment
  - C. Orotic aciduria
  - D. Gout
- 61. In a child with anemia, bone marrow examination demonstrates erythroid hyperplasia, binuclearity and multinuclearity of the erythroid precursors. Ham's acidified serum test is positive in this patient. The most likely diagnosis is:
  - A. Iron deficiency
  - B. Folate deficiency

- C. Congenital dyserythropoietic anemia, type I
- D. Congenital dyserythropoietic anemia, type II
- 62. A 50-year-old male with developed macrocytic anemia (MCV is 105 fl; red cells are round and not oval). Liver function test demonstrates elevated γ glutamyl transpeptidase (GGT) as the only abnormality. Bone marrow examination shows ring sideroblasts and erythroid multinuclearity. There is vacuolation of erythroid precursors. Cytogenetics and myelodysplastic-flourescent in situ hybridization (MDS-FISH) profile are negative. The most likely diagnosis is:
  - A. Myelodysplastic syndrome (MDS)
  - B. Megaloblastic anemia
  - C. Excess alcohol intake
  - D. Hemochromatosis
- 63. A child with a known diagnosis of hereditary spherocytosis develops "slapped cheek" appearance as well as profound anemia. Reticulocyte response to the anemia is inadequate. A bone marrow examination is most likely to demonstrate:
  - A. Hypocellular marrow with reduction of all cell lines
  - B. Erythroid hyperplasia as a response to acute hemolysis
  - C. Reduced erythropoiesis with giant erythroblasts and intranuclear inclusions
  - D. "Owl eye " appearance, due to CMV infection in erythroid precursors
- 64. Characteristic feature of Fanconi's anemia include:
  - A. Autosomal dominant mode of inheritance
  - B. Increased risk of transformation to acute myeloid leukemia
  - C. Pure red cell aplasia
  - D. Skin hyperpigmentation and nail dystrophy

#### Hemolytic Anemias: Questions 65–84

- 65. All statements regarding hereditary spherocytosis is true except:
  - A. May be diagnosed by flow cytometry
  - B. May be diagnosed by incubated osmotic fragility test

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

Chapter 1: RBC Disorders

- C. When inherited, most cases are homozygous
- D. The direct antiglobulin test (DAT) is typically negative
- 66. Regarding membrane defects of red cells, all are true except:
  - A. In hereditary elliptocytosis there is often defective spectrin dimer–dimer interaction.
  - B. In hereditary pyropoikilocytosis there is often defective spectrin dimer–dimer interaction
  - C. In making a diagnosis of hereditary stomatocytosis, >90% of red cells need to be stomatocytes
  - D. In Rh null disease, stomatocytes may be seen in the peripheral blood
- 67. Pyruvate kinase deficiency:
  - A. Is transmitted as autosomal dominant
  - B. Is the commonest enzyme deficiency in the TCA cycle
  - C. Is the commonest enzyme deficiency in the hexose monophosphate shunt
  - D. Can present as jaundice
- 68. Regarding glucose 6 phosphate dehydrogenase (G6PD) enzyme:
  - A. Normally two isotypes G6PD-A and G6PD-B – can be differentiated based on electrophoretic mobility
  - B. The A isoform is the most common type found in all populations
  - C. 90% of US black men have the G6PD-A minus isoform
  - D. The A minus isoform has the same electrophoretic mobility as the B isoform
- 69. All of the following statements regarding paroxysmal nocturnal hemoglobinuria (PNH) are true except
  - A. PNH is transmitted as X-linked recessive
  - B. PNH may be diagnosed by flow cytometry
  - C. PNH may be diagnosed by Ham's acidified serum test
  - D. Occurs due to mutation of the PIG-A gene
- 70. Which of the following observations may be seen in a positive Ham's acidified serum test?
  - A. The tube with patient's red cells and patients serum, heated to 56°C shows hemolysis

- B. The tube with patient's red cells and patient's serum, without addition of acid, shows hemolysis
- C. The tube with patient's red cells and patient's serum, with addition of acid, show hemolysis
- D. The tube with patient's red cells and patient's serum, with addition of acid and heated to 56°C shows hemolysis.
- 71. Regarding autoimmune hemolytic anemias:
  - A. In warm autoimmune hemolytic anemias (WAHA), the antibodies are often against I antigen
  - B. In warm autoimmune hemolytic anemia, hemolysis is typically intravascular
  - C. In cold hemagglutinin (CHAD) disease, antibodies are often directed against P antigen
  - D. Paroxysmal cold hemoglobinuria (PCH) is a rare condition and is sometimes seen in children following a viral infection.
- 72. Regarding the following poikilocytes, all are true except:
  - A. In individuals with HbSS disease, under low oxygen tension, tubular structures called tactoids deform the red cells and produce sickle cells
  - B. Target cells may be seen in individuals with lecithin cholesterol acyltransferase deficiency
  - C. Macro ovalocytes are commonly seen in individuals with hypothyroidism
  - D. Abundant target cells are more often seen in HbSC disease rather than in HbSS disease
- 73. Regarding the following poikilocytes all are true except:
  - A. Occasional elliptocytes may be seen in iron deficiency
  - B. Individuals with Rh null disease may exhibit stomatocytes in their peripheral blood
  - C. Echinocytes may be seen in patients with pyruvate kinase deficiency
  - D. Burr cells are also known as drepanocytes
- 74. Regarding acanthocytes
  - A. The word acanthocyte is derived from the Greek word for "urchin"
  - B. May be seen in individuals with McLeod phenotype

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

#### Section 1: Non-Neoplastic Hematology

- C. Is a feature of HbSS disease
- D. These are cells with hundreds of thorny projections
- 75. Howell-Jolly bodies are:
  - A. DNA material
  - B. Consist of iron
  - C. RNA material
  - D. An in vitro phenomenon
- 76. Pyrimidine 5 nucleotidase:
  - A. Is an enzyme in the heme biosynthetic pathway
  - B. Is responsible for the degradation of RNA in red cells
  - C. Deficiency results in target cells
  - D. Deficiency results in bite cells
- 77. All of the following are features of parasitization of RBCs with Plasmodium falciparum except:
  - A. Plasmodium parasite with two chromatin dots
  - B. Presence of marginal forms
  - C. Banana- or crescent-shaped gametocytes
  - D. Enlarged red cells
- 78. In cold hemagglutinin disease (CHAD):
  - A. The antibody is typically IgG in nature
  - B. There is an association with pneumonia due to Staph aureus
  - C. Red cell agglutination occurs in vivo
  - D. Analysis of CBC values will yield low RBC count with normal hemoglobin levels
- 79. Established causes of microangiopathic hemolysis include all except
  - A. Thrombotic thrombocytopenic purpura (TTP)
  - B. Hemolytic uremic syndrome (HUS)C. Disseminated intravascular coagulation (DIC)
  - D. Immune thrombocytopenic purpura (ITP)
- 80. Which of the following laboratory finding is expected in hereditary spherocytosis?
  - A. Increased MCHC
  - B. Low MCV and MCH
  - C. Low serum bilirubin
  - D. Low LDH
- 81. Regarding various hemolytic anemias, which statement is true:
  - A. Hereditary spherocytosis may be transmitted as autosomal recessive

- B. G6PD deficiency may be seen in females with asymmetric lyonization
- C. Myelodysplastic syndrome may result in acquired pyruvate kinase deficiency
- D. All of the above
- 82. Which of the following tests for hereditary spherocytosis has the highest sensitivity and specificity?
  - A. Osmotic fragility test
  - B. Eosin-5-maleimide (EMA) binding test
  - C. Acidified glycerol lysis test (AGLT)
  - D. DAT
- 83. Regarding cryoglobulinemia:
  - A. HCV infection is the most common cause of type I cryoglobulinemia
  - B. In an individual with multiple myeloma, type III cryoglobulinemia may be seen
  - C. High levels of complement are expected
  - D. Vasculitic skin lesions may be seen
- 84. Gaisbock syndrome refers to:
  - A. Erythrocytosis seen in polycythemia rubra vera
  - B. Erythrocytosis seen in CML
  - C. Erythrocytosis seen due to dehydration
  - D. Erythrocytosis seen in patients with renal tumors

#### Hemoglobinopathies: Questions 85–109

- 85. An example of an embryonic hemoglobin is:
  - A. Hemoglobin F
  - B. Hemoglobin A
  - C. Hemoglobin A2
  - D. Hemoglobin Gower
- The following are all hemoglobin adducts except:
   A. HbAIII
  - B. HbA1c
  - C. HbA1d
  - D. Hemoglobin CS (Constant Spring)
- 87. Regarding hemoglobin biosynthesis:
   A. The first step is formation of δ

   aminolevulinic acid from glycine and
   succinyl CoA, which takes place in the
   cytoplasm

Cambridge University Press 978-1-316-50560-1 — Hematopathology and Coagulation Amer Wahed , Jesse Manuel Jaso , Ashok Tholpady Excerpt More Information

Chapter 1: RBC Disorders

- B. Combination of heme with globin to form hemoglobin, in the cytoplasm
- C. Conversion of protoporphyrin IX to heme by the removal of iron
- D. Conversion of  $\delta$  aminolevulinic acid to porphobilinogen in the mitochondria
- 88. Regarding hemoglobinopathies, which of the following is true:
  - A. There are about 100 hemoglobinopathies and most are asymptomatic
  - B. About 5% of the population in the world are carriers of hemoglobinopathies
  - C. If an individual has HbC, then their ancestry is most likely from the Carribean
  - D. Certain hemoglobinopathies are characterized by reduced amount of normal globin chain synthesis.
- 89. Which of the following is not a sickling disorder?
  - A. HbSS disease
  - B. HbSC disease
  - C. HbSD disease
  - D. HbSE disease
- 90. HbH disease is characterized by:
  - A. Deletion of one  $\alpha$  gene
  - B. Deletion of two α genes
  - C. Deletion of three  $\alpha$  genes
  - D. Deletion of four  $\alpha$  genes
- 91. Regarding  $\alpha$  thalassemia trait:
  - A. The condition may resemble iron deficiency
  - B. Individuals have significant microcytic hypochromic anemia
  - C. If due to alpha thalassemia 1  $(-/-, \alpha, \alpha)$ , which is due to cis deletion of both  $\alpha$ genes on the same chromosome, is seen more often in the African American population
  - D. If due to  $\alpha$  thalassemia 2 (-/  $\alpha$ , -/ $\alpha$ ), which is due to trans deletion of the  $\alpha$ genes on two different chromosomes, is seen more often in Southeast Asian individuals
- 92. In α thalassemia disease:
  - A. Hb Bart's may be seen in the newborn
  - B. HbH will be evident when the patient is an adult

- C. Patients have chronic hemolytic anemia
- D. All of the above are true
- 93. An individual has microcytic hypochromic anemia (mild) with the presence of target cells on the peripheral smear. Hemoglobin electrophoresis demonstrates HbA2 (6%), HbF (3%) and HbA (91%). Iron studies are unremarkable. Patient has never received any blood transfusion. The most likely diagnosis is:
  - A. α thalassemia trait
  - B. β thalassemia trait
  - C.  $\beta$  thalassemia intermedia
  - D.  $\beta$  thalassemia major
- 94. An individual undergoes hemoglobin electrophoresis by the capillary method. HbA2 is detected at low levels,<1.5%. The following are possible causes:
  - A. α thalassemia trait
  - B. δ thalassemia
  - C. Iron deficiency
  - D. All of the above
- 95. A Greek individual undergoes hemoglobin electrophoresis for mild anemia. Results show normal HbA2 at 3.3%, HbF at 10% and the remaining hemoglobin is HbA. A possible diagnosis is:
  - A. β thalassemia trait
  - B. β thalassemia major
  - C.  $\delta \beta$  thalassemia, heterozygous state
  - D. δ thalassemia
- 96. Hemoglobin S is due to a  $\beta$  chain defect. HbS is formed when:
  - A. Glutamic acid is replaced by lysine at the 26th position of the  $\beta$  chain
  - B. Glutamic acid is replaced by lysine at the 121st position of the  $\beta$  chain
  - C. Glutamic acid is replaced by lysine at the 6th position of the  $\beta$  chain
  - D. Glutamic acid is replaced by value at the 6th position of the  $\beta$  chain
- 97. Increased levels of HbF can be seen in all of the following except:
  - A. HbSS disease
  - B. HbSS individuals on hydroxyurea