Questions

1) A 12-month-old girl presents for evaluation of extreme irritability without fever. She came to Texas shortly after birth as a Somalian refugee, and seemed to feed and grow well with a slight falloff in weight recently. Physical examination reveals a slightly enlarged liver, a palpable spleen, and tenderness over her extremities. A peripheral smear shows sickle-shaped red blood cells and subsequent studies confirm a diagnosis of sickle cell anemia. When told this is a genetic disorder, the parents are skeptical because she had been so healthy early in life. Which of the following explanations concerning hemoglobin chain structure best explains her lack of symptoms the first few months after birth?

a. The (α1 -α2)(β1 -β2) subunit structure shifts to (α1 -β1)-(α2 -β2) at puberty.

b. The α1 -α2 -α3 -α4 subunit structure switches to (α1 -β1)-(α2 -β2) during pregnancy.

c. β-β-β-α Subunit structure switches to (α1 -β1)-(α2 -β2) during infancy.

d. The (β1 -β2 -β3 -α1) subunit structure switches to (α1 -β1)-(α2 -β2) during pregnancy.

e. The (α1 -γ1)-(α2 -γ2) subunit structure switches to (α1 -β1)-(α2 -β2) at 2 to 6 months.

2) A 17-year-old African American adolescent is evaluated for anemia and is found to have increased indirect bilirubin in serum and plasma free hemoglobin suggestive of hemolysis (lysis of red blood cells). Enzyme assays reveal deficiency of bisphosphoglycerate mutase with low concentrations of 2,3-bisphosphoglycerate (BPG) in her red blood cells. The teenager shows greater respiratory distress than expected for the degree of anemia. Which of the following statements best explains the consequences of this enzyme deficiency?

a. BPG shifts the curve to the right, so less BPG will cause greater release of oxygen to tissues.

b. BPG shifts the curve to the right, so less BPG will cause lesser release of oxygen to tissues.

c. BPG shifts the curve to the left, so less BPG will cause greater release of oxygen to tissues.

d. BPG shifts the curve to the left, so less BPG will cause lesser release of oxygen to tissues.

e. BPG binds oxygen directly and has no effect on the curve.

3) An 18-year-old African American female is evaluated for her first prenatal visit and is found to have chronic anemia, a slightly enlarged spleen, increased reticulocyte count, and mild elevation of indirect-reacting (unconjugated) bilirubin in serum. Incubation of the female's red cells with glucose yields decreased amounts of ATP as compared to controls, even in the presence of oxygen. The female's anemia is explained by the fact that ATP is produced by which of the following pathways?

a. Glycogen breakdown.

b. Glycolysis.

c. Oxidative phosphorylation.

d. Pentose phosphate cycle.

e. Lactate conversion to glucose (Cori cycle).

4) Increased resistance to malaria is seen in persons with hemoglobin AS, where A is the normal allele and S is the allele for sickle hemoglobin. Which of the following terms applies to this situation?

a. Founder effect.

b. Heterozygote advantage.

c. Genetic lethal.

d. Fitness.

e. Natural selection.

5) A female presents with fatigue, pallor, and pale conjunctival blood vessels. She gives a recent history of metrorrhagia (heavy menstrual periods). Which of the following laboratory findings is most likely?

a. High serum haptoglobin.

b. High serum iron.

c. High numbers of transferrin receptors.

d. High saturation of transferrin.

e. High serum ferritin.

6) A 19-year-old man is referred to an oncologist after his primary care physician detects a soft tissue mass along the distal femur. The oncologist diagnoses the patient with

osteosarcoma and places him on a chemotherapeutic regimen that includes a dihydrofolate reductase inhibitor. A week later, he returns to the clinic with nausea, vomiting, and diarrhea. Which of the following is the best course of intervention?

(A) Administer erythropoietin.

(B) Administer filgrastim.

(C) Folic acid supplementation.

(D) Iron supplementation.

(E) Vitamin B12 supplementation.

7) An 8-year-old boy has a history of chronic and severe hemolytic anemia, hepatosplenomegaly, and maxillary overgrowth. He has received blood transfusions since early infancy but has not received a transfusion in >4 months. Hemoglobin (Hb) electrophoresis shows marked elevation of HbF, increased HbA2, and absence of HbA1. Which of the following diagnoses is most consistent with this patient's electrophoresis?

(A) a-Thalassemia minor.

(B) b-Thalassemia major.

(C) b-Thalassemia minor.

(D) Glucose 6-phosphate dehydrogenase deficiency.

(E) HbH disease histocompatibility complex class II.

(F) RBCs containing Hb Bart.

8) Which biochemical component of the erythrocyte cell surface is primarily responsible for determining blood type (eg, the A-B-O system).

a. Fatty acid.

b. Carbohydrate.

c. Nucleic acid.

d. Protein.

e. Cholesterol.

9) 18) What cell in circulating blood is the precursor to microglia and most antigen-presenting cells?

a. Eosinophil.

b. Basophil.

c. Lymphocyte.
d. Monocyte.
e. Mast cell.

10) What is the approximate life span of a circulating erythrocyte?
a. 8 days.
b. 20 days.
c. 5 weeks.
d. 4 months.
e. 1 year.

11) Which cell type has cytoplasmic granules that contain heparin and histamine?
a. Eosinophils.
b. Basophils.
c. Lymphocytes.
d. Monocytes.
e. Neutrophils.

12) A differential cell count of a blood smear from a patient with a parasitic infection is likely to reveal an increase in the circulating numbers of which cell type?
a. Neutrophils.
b. Lymphocytes.
c. Monocytes.
d. Basophils.
e. Eosinophils.

13) Which of the following blood cells differentiate outside of the bone marrow?
a. Neutrophils.
b. Basophils.
c. Eosinophils.
d. T lymphocytes.
e. Megakaryocytes.

14) Examination of a normal peripheral blood smear reveals a cell more than twice the diameter of an erythrocyte with a kidney-shaped nucleus. There cells are < 10% of the total leukocytes. Which of the following cell types is being described?

a. Monocyte.

b. Basophil.

c. Eosinophil.

d. Neutrophil.

e. Lymphocyte.

15) A 43-year-old anatomy professor is working in her garden, pruning rose bushes without gloves, when a thorn deeply penetrates her forefinger. The next day the area has become infected. She removes the tip of the thorn, but there is still pus remaining at the wound site. Which of the following cells function in the formation of pus?

a. Cells with spherical nuclei and scant cytoplasm.

b. Biconcave cells with no nuclei.

c. Cells with bilobed nuclei and many acidophilic cytoplasmic granules.

d. Very small, cell-like elements with no nuclei but many granules.

e. Cells with polymorphic, multiply lobed nuclei.

16) A 35-year-old woman's physician orders laboratory blood tests. Her fresh blood is drawn and centrifuged in the presence of heparin as an anticoagulant to obtain a hematocrit. From top to bottom, the fractions resulting from centrifugation are which of the following?

a. Serum, packed erythrocytes, and leukocytes.

b. Leukocytes, erythrocytes, and serum proteins.

c. Plasma, buffy coat, and packed erythrocytes.

d. Fibrinogen, platelets, buffy coat, and erythrocytes.

e. Albumin, plasma lipoproteins, and erythrocytes.

17) A hematologist diagnoses a 34-year-old woman with idiopathic thromobocytic purpura (ITP). Which of the following symptoms/ characteristics would one expect in this patient?

a. Normal blood count.

b. Hypercoagulation.
c. Decreased clotting time.
d. Abnormal bruising.
e. Light menstrual periods.

18) Which of the following can be used to describe megakaryocytes?
a. Multinucleated.
b. Formed by fusion of haploid cells.
c. Precursors to bone marrow macrophages.
d. A minor but normal formed element found in the circulation.
e. Possess dynamic cell projections from which one type of formed element is released.

19) Which one of the following statements concerning the hemoglobins is correct?
A. HbA is the most abundant hemoglobin in normal adults.
B. Fetal blood has a lower affinity for oxygen than does adult blood because HbF has an increased affinity for 2,3-bisphosphoglycerate.
C. The globin chain composition of HbF is α2δ2.
D. HbA1c differs from HbA by a single, genetically determined amino acid substitution.
E. HbA2 appears early in fetal life.

20) Which one of the following statements concerning the ability of acidosis to precipitate a crisis in sickle cell anemia is correct?
A. Acidosis decreases the solubility of HbS.
B. Acidosis increases the oxygen affinity of hemoglobin.
C. Acidosis favors the conversion of hemoglobin from the taut to the relaxed conformation.
D. Acidosis shifts the oxygen-dissociation curve to the left.
E. Acidosis decreases the ability of 2,3-bisphosphoglycerate to bind to hemoglobin.

21) Which one of the following statements concerning the binding of oxygen by hemoglobin is correct?
A. The Bohr effect results in a lower oxygen affinity at higher pH values.

B. Carbon dioxide increases the oxygen affinity of hemoglobin by binding to the C-terminal groups of the polypeptide chains.

C. The oxygen affinity of hemoglobin increases as the percentage saturation increases.

D. The hemoglobin tetramer binds four molecules of 2,3- bisphosphoglycerate.

E. Oxyhemoglobin and deoxyhemoglobin have the same affinity for protons.

22) β-Lysine 82 in HbA is important for the binding of 2,3-bisphosphoglycerate. In Hb Helsinki, this amino acid has been replaced by methionine. Which of the following should be true concerning Hb Helsinki?

A. It should be stabilized in the taut, rather than the relaxed, form.

B. It should have increased oxygen affinity and, consequently, decreased oxygen delivery to tissues.

C. Its oxygen-dissociation curve should be shifted to the right relative to HbA.

D. It results in anemia.

23) A 67-year-old man presented to the emergency department with a 1-week history of angina and shortness of breath. He complained that his face and extremities had taken on a blue color. His medical history included chronic stable angina treated with isosorbide dinitrate and nitroglycerin. Blood obtained for analysis was brown. Which one of the following is the most likely diagnosis?

A. Carboxyhemoglobinemia.

B. Hemoglobin SC disease.

C. Methemoglobinemia.

D. Sickle cell anemia.

E. β-Thalassemia.

24) A compensatory mechanism to allow adequate oxygen delivery to the tissues at high altitudes, where oxygen concentrations are low, is which of the following?

A. An increase in 2,3-bisphosphoglycerate synthesis by the red cell.

B. A decrease in 2,3-bisphosphoglycerate synthesis by the red cell.

C. An increase in hemoglobin synthesis by the red cell.

D. A decrease in hemoglobin synthesis by the red cell.

E. Decreasing the blood pH.

25) Drugs are being developed that will induce the transcription of globin genes, which are normally silent in patients affected with sickle cell disease. A good target gene for such therapy in this disease would be which of the following?

A. The a1-gene.

B. The a2-gene.

C. The gamma-gene.

D.The b –gene.

E. The zeta-gene.

26) A mature blood cell that lacks a nucleus is which of the following?

A. Lymphocyte.

B. Basophil.

C. Eosinophil.

D. Platelet.

E. Neutrophil.

27)) A family has two children, one with a mild case of thalassemia and a second with a severe case of thalassemia, requiring frequent blood transfusions as part of the treatment plan. One parent is of Mediterranean descent, the other is of Asian descent. Neither parent exhibits clinical signs of thalassemia. Both children express 20% of the expected level of b-globin; the more severely affected child expresses normal levels of a-globin, whereas the less severely affected child expresses only 50% of the normal levels of a-globin. Why is the child who has a deficiency in a-globin expression less severely affected?

A. Thalassemia is caused by a mutation in the a-gene, and the more severely affected child expresses more of it.

B. The less severely affected child must be synthesizing the zeta-gene to make up for the deficiency in a-chain synthesis.

C. The more severely affected child also has HPFH.

D. The more severely affected child produces more inactive globin tetramers than the less severely affected child.

E. Thalassemia is caused by an iron deficiency, and when the child is synthesizing normal levels of a-globin, there is insufficient iron to populate all of the heme molecules synthesized.

28) The edema observed in patients with noncalorie protein malnutrition is caused by which of the following?

A. Loss of muscle mass.

B. Ingestion of excess carbohydrates.

C. Increased fluid uptake.

D. Reduced protein synthesis in the liver.

E. Increased production of ketone bodies.

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Do not hesitate to ask me if you have any questions.

GOOD LUCK