Subject: HLS-Biochemistry

Topic: Collected Qs from Textbooks

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1. A 27-year-old firefighter is brought to the emergency room after being exposed to smoke during a training exercise. He looks ill and has labored breathing. He is clutching his head and exhibits an altered mental status. On examination, you note that he appears red, and his pulse oximetry reads 100%. You suspect carbon monoxide toxicity. What is true of the oxygen saturation curve during carbon monoxide toxicity?

(A) The oxygen saturation curve is shifted to the left.
(B) The oxygen saturation curve is shifted to the right.
(C) The effect of carbon monoxide on hemoglobin is similar to that of having increased levels of 2,3 bisphosphoglycerate.
(D) The effect of carbon monoxide on hemoglobin is similar to that of a low pH state.
(E) The effect of carbon monoxide on hemoglobin is similar to that of an increased temperature state

2. A pediatric hematologist sees an 18-monthold patient with jaundice, splenomegaly, and hemolytic anemia. A blood smear indicates RBCs that are more rigid in appearance than normal, and a diagnosis of pyruvate kinase deficiency is made. Because pyruvate kinase catalyzes the last step in the glycolytic pathway, products before this step of the pathway will accumulate. Which one of the following products associated with the pathway will be made in abnormal amounts?

(A) Acetyl CoA
(B) Glucose
(C) 2,3-Bisphosphoglycerate
(D) OAA
(E) Pyruvate

3. A 3-year-old boy is brought to the emergency room with abdominal pain, mental status changes, and fatigue. On history, the physician finds that the patient lives in an older house and has been sucking on the paint chips that have crumbled in the windowsills, making the doctor suspicious for lead poisoning. Lead typically interferes with which of the following enzymes?

(A) Cytochrome oxidase

(B) Protoporphyrinogen oxidase

(C) UMP synthase

(D) d-ALA dehydratase

(E) Porphobilinogen deaminase

4. An 8-year-old boy sees a dermatologist because he has developed vesicles and bullae on his face and arms that appeared after a week-long trip to Florida. His father has a similar condition. A diagnosis of porphyria cutanea tarda is confirmed by finding elevated levels of porphyrins in his serum, urine, and stool. His disease is due to a deficiency of which of the following enzymes?

(A) d-ALA dehydratase

(B) Porphobilinogen deaminase

(C) Uroporphyrinogen III cosynthase

(D) Ferrochelatase

(E) Uroporphyrinogen decarboxylase

5. A postpartum woman from a rural Appalachian community recently gave birth to a baby boy with the aid of a midwife at home. She now brings the baby to the hospital because of continued bleeding and oozing from the umbilical stump. A deficiency of which vitamin might be associated to this case?

(A) Vitamin A

(B) Vitamin B

(C) Vitamin K

(D) Vitamin D

(E) Vitamin E

6. A 16-year-old African-American girl comes to the emergency room with complaints of painful muscle cramps. She states that she has sickle cell anemia and that she ran out of her pain medication. A complete blood count and smear rapidly confirm the diagnosis, and she is started on intravenous fluids and pain medications. The molecular defect underlying her disease is which of the following?

(A) A valine rather than a glutamate at position 6 of the b-globin protein

(B) A glutamate rather than a valine at position 6 of the b-globin protein

(C) A valine rather than a glutamine at position 6 of the a-globin protein

(D) A glutamine rather than a valine at position 6 of the a-globin protein

(E) Expansion of a polyglutamine repeat within the b-globin gene

7. An infant presents with neonatal jaundice. After several weeks, the jaundice becomes more exaggerated. The patient has an enzyme deficiency that inhibits conjugation of bilirubin. Which of the following reacts with bilirubin in the conjugation reaction?

(A) Vitamin C

(B) Iron

(C) Ceruloplasmin

(D) Porphyrin ring

(E) UDP-glucuronate

8. 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 $\alpha 2\delta 2$.

(D) HbA1c differs from HbA by a single, genetically determined amino acid substitution.

(E) HbA2 appears early in fetal life.

9. 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.

10. 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 saturation percentage increases.
(D) The hemoglobin tetramer binds four molecules of 2,3- bisphosphoglycerate.
(E) Oxyhemoglobin and deoxyhemoglobin have the same affinity for protons.

11. 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.

12. 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

13. Why is hemoglobin C disease a non-sickling disease?

(A) It involves no mutations

(B) The polar glutamate is replaced by polar lysine rather than by nonpolar AA

(C) HbC has a good affinity towards oxygen

(D) Hemoglobin C is a very rare disease

14. δ-Aminolevulinic acid synthase activity:
(A) catalyzes the committed step in porphyrin biosynthesis.

(B) is decreased by iron in erythrocytes.

(C) is decreased in the liver in individuals treated with certain drugs such as the barbiturate phenobarbital.
(D) occurs in the cytosol.
(E) requires tetrahydrobiopterin as a coenzyme.

15. A 50-year-old man presented with painful blisters on the backs of his hands. He was a golf instructor and indicated that the blisters had erupted shortly after the golfing season began. He did not have recent exposure to common skin irritants. He had partial complex seizure disorder that had begun ~3 years earlier after a head injury. The patient had been taking phenytoin (his only medication) since the onset of the seizure disorder. He admitted to an average weekly ethanol intake of ~18 12-oz cans of beer. The patient's urine was reddish orange. Cultures obtained from skin lesions failed to grow organisms. A 24-hour urine collection showed elevated uroporphyrin (1,000 mg; normal, <27 mg). The most likely diagnosis is:

(A) acute intermittent porphyria.

(B) congenital erythropoietic porphyria.

(C) erythropoietic protoporphyria.

(D) hereditary coproporphyria.

(E) porphyria cutanea tarda.

16. A patient presents with jaundice, abdominal pain, and nausea. Clinical laboratory results are shown below.
What is the most likely cause of the jaundice?

Plasma bilirubin	Urine urobilinogen	Urinary bilirubin
Increase in conjugated bilirubin	Not present	Present

(A)Decreased hepatic conjugation of bilirubin

(B) Decreased hepatic uptake of bilirubin

(C) Decreased secretion of bile into the intestine

(D) Increased hemolysis

17. A 2-year-old child was brought to his pediatrician for evaluation of gastrointestinal problems. The parents report that the boy has been listless for the last few weeks. Lab tests reveal a microcytic, hypochromic anemia. Blood lead levels are elevated. Which of the enzymes listed below is most likely to have higher-than-normal activity in the liver of this child?

(A) δ-Aminolevulinic acid synthase

(B) Bilirubin UDP glucuronosyltransferase

(C) Ferrochelatase

(D) Heme oxygenase

(E) Porphobilinogen synthase

For questions 18-21, choose the appropriate protein factor:

Letter	A	B	C	D	E
Factor	V	XIII	II	IX	X

18. This factor activates components of the intrinsic, extrinsic, and common pathways.

19. This factor converts the soluble clot to a hard clot.

20. This factor initiates the common pathway.

21. This factor is an accessory protein that potentiates the activity of factor Xa.

22. Which one of the following can be ruled out in a patient with thrombophilia (hypercoagulable state)?

(A) deficiency of antithrombin III
(B) A deficiency of FIX
(C) A deficiency of protein C
(D) An excess of prothrombin

23. Current guidelines for the treatment of patients with acute ischemic stroke (a stroke caused by a blood clot obstructing a vessel that supplies blood to the brain) include the recommendation that tissue plasminogen activator (TPA) be used shortly after the onset of symptoms. The basis of the recommendation for TPA is that it activates:

(A) antithrombin III.

(B) the activated protein C complex.

(C) the receptor for von Willebrand factor.

(D) the serine protease that degrades fibrin.

(E) thrombomodulin.

24. The parents of a newborn baby girl refuse to allow the baby to be giventhe injection of vitamin K that is recommended shortly after birth to prevent vitamin K deficiency bleeding, which is caused by the low levels of the vitamin in newborns. The activity of which one of the following protein factors involved in clotting would be decreased with vitamin K deficiency?

(A)FV

(B) FVII

(C) FXI

(D) FXII

(E) FXIII

25. Nephrotic syndrome is a kidney disease characterized by protein loss in the urine (≥3 g/day) that is accompanied by edema. The loss of protein results in a hypercoagulable state. Excretion of which of the following proteins would explain the thrombophilia seen in the syndrome?

(A) Antithrombin III

(B) FV

(C) FVIII

(D) Prothrombin

26. Thrombin, produced in the common pathway of clotting, has both procoagulant and anticoagulant activities. Which one of the following is an anticoagulant activity of thrombin?

(A) Activating FXIII

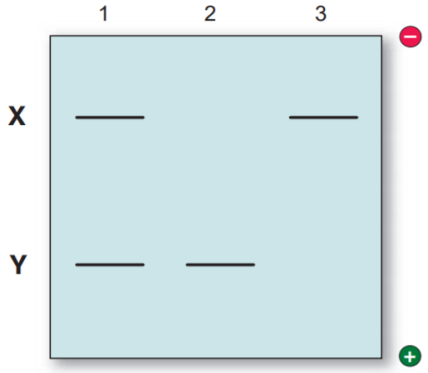
(B) Binding to thrombomodulin

(C) Increasing nitric oxide production

(D) Inhibiting FV and FVIII

(E) Inhibiting tissue factor pathway inhibitor

27. You order a hemoglobin electrophoresis on a patient suspected of having sickle cell disease. A blood sample was obtained and the red cells were isolated. Disruption of the red cells released the hemoglobin, which was run on a polyacrylamide gel. Following the electrophoresis, a Western blot was performed to locate the hemoglobin. The results of the Western blot are shown below. Which one of the following statements best represents the interpretation of the results?



(A) The band marked as X refers to the wild-type hemoglobin protein

(B) The band in lane 2 represents an individual with sickle cell disease.

(C) A carrier of sickle cell disease is represented by the band in lane 3

(D) Lane 1 represents an individual with sickle cell disease

(E) Lane 3 represents an individual with sickle cell disease

28. You see a patient on an initial visit and are struck by the bluish coloring of the skin and mucous membranes. You ask the patient about this and you are told that it is a blood problem that the patient has had for his or her entire life. The patient's father had a similar condition, but not the mother. This condition could result from which one of the following changes within the erythrocyte?

(A) An increase of 2,3-bisphosphoglycerate in the erythrocyte

(B) An E to V mutation at position 6 of the β-chain of hemoglobin

(C) Increased oxidation of heme iron to the +3 state

(D) Enhanced oxygen binding to hemoglobin

(E) A mutated hemoglobin which no longer exhibited the Bohr effect

29. Your diabetic patient has a hemoglobin A1c (HbA1c) of 8.8. HbA1c differs from unmodified hemoglobin by which one of the following?

(A) Amino acid sequence

(B) Serine acylation

(C) Valine glycosylation

(D) Intracellular location

(E) Rate of degradation

30. A critical histidine side chain in an enzyme's active site displays a pKa value of 8.2. Which of the following best describes the effect of local environment in which this histidine residue resides?

(A) A loss of quaternary structure of the hemoglobin molecule

(B) An increase in oxygen binding to hemoglobin

(C) A gain of ionic interactions, stabilizing the "T" form of hemoglobin

(D) An increase in hydrophobic interactions between deoxyhemoglobin molecules

(E) An alteration in hemoglobin secondary structure leading to loss of the "α" helix

31. Parents bring their 6-year-old son to the pediatrician due to the parents being concerned about "mental retardation." Blood work demonstrated a microcytic anemia and basophilic stippling. During the patient history, it became apparent that the boy often stayed with his grandparents, who owned a 150-year-old apartment. The boy admitted to eating paint chips that are full of lead from the radiators in the apartment. The boy's anemia is most likely the result of which one of the following?

(A) Inhibition of iron transport

(B) Reduction of heme synthesis

(C) Inhibition of the phosphatidyl inositol cycle

(D) Blockage of reticulocyte DNA synthesis

(E) Inhibition of β-globin gene expression

32. A patient, who was recently diagnosed with cystic fi brosis, displays an increased blood clotting time. This is most likely due to which of the following?

(A) Lack of proline hydroxylation

(B) Inability to catalyze transaminations

(C) Lack of dolichol and an inability to glycosylate serum proteins

(D) Inability to carboxylate glutamic acid side chains

33. A patient has had a series of blood clots, and has been placed on warfarin to reduce such incidents. Warfarin exerts its effect by blocking which of the following?

(A) Platelet biogenesis

(B) Phospholipid synthesis

(C) Zymogen synthesis

(D) Vitamin E activity

(E) Formation of γ-carboxyglutamate

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

(A) An increase in 2,3-BPG synthesis by the erythrocyte
(B) A decrease in 2,3-BPG synthesis by the erythrocyte
(C) An increase in hemoglobin synthesis by the erythrocyte
(D) A decrease in hemoglobin synthesis by the erythrocyte
(E) Decreasing the blood pH

35. Drugs are being developed that will induce the transcription of certain 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 one of the following?

(A) The α1-gene
(B) The α2-gene
(C) The γ-gene
(D) The β-gene
(E) The ζ-gene

36. A patient is a strict vegetarian and, as such, is concerned about getting sufficient iron in his diet. Which suggestion below could increase his dietary iron absorption?

(A) Never peel potatoes when preparing a potato dish.
(B) Squeeze fresh lemon juice on spinach salad.
(C) Reassure him that iron in plants is readily absorbed.
(D) Meat is the only dietary source of iron.
(E) Taking a vitamin with vitamin B12 would help iron absorption.

37. Classic hemophilia A is the absence of Factor VIII. It results in an inability to directly activate which one of the following factors?
(A) Factor II
(B) Factor IX
(C) Factor X
(D) Protein S
(E) Protein C

38. An inactivating mutation in which one of the following proenzymes would be expected to lead to thrombosis (uncontrolled blood clotting)?
(A) Factor XIII
(B) Prothrombin
(C) Protein C
(D) Factor VIII
(E) Tissue factor

39. An individual accidently ingested rat poison that interfered with vitamin K action. The activity of which one of the following clotting factors is vitamin K-dependent?
(A) Factor II
(B) Factor III
(C) Factor V
(D) Factor VIII
(E) Factor XIII

40. A patient had a stroke (a cerebrovascular accident [CVA]) caused by a clot in the carotid circulation and received "clot busters" intravenously as emergency treatment. Which clot buster below is not normally found in the human body?
(A) APC
(B) PAI-1
(C) tPA
(D) Streptokinase
(E) uPA

ANSWERS

1	A	21	A
2	C	22	B
3	D	23	D
4	E	24	B
5	C	25	A
6	A	26	B
7	E	27	E
8	A	28	C
9	A	29	C
10	C	30	C
11	B	31	B
12	C	32	D
13	B	33	E
14	A	34	A
15	E	35	C
16	C	36	B
17	A	37	C
18	C	38	C
19	B	39	A
20	E	40	D

Good Luck!!