Boston University CH373-S20
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BU ID#

Molecular Basis for Sickle Cell Disease

Adapted from Nicholas' Story by Didem Vardar-Ulu for CH373-S20

IN-CLASS TEST

Instructions: Please recall that **we treat cheating with zero tolerance.**

This exam will be scanned and uploaded to GradeScope for grading by the course staff. In order for the exam to be properly assigned to you, please make sure **to write your name (as it appears on the BU roster) in the box to the right, and your BU ID number on the top of ALL pages.** Please write legibly, in block letters (not cursive). Make sure ALL your answers are inside the provided boxes.

Full Name (First and Last):

[ ] **Q1. The secondary structure of myoglobin and the structure of individual subunits of hemoglobin consist of the following.**

a. A tetramer with 4 alpha-helices organized around a heme group.
b. A single polypeptide chain, folded around a heme group, with 6 alpha-helices.
c. A single polypeptide chain, folded around a heme group, with 8 alpha -helices.
d. A single polypeptide chain, folded around a heme group, exhibiting an alpha-beta-alpha fold
e. None of the above.

[ ] **Q2. Upon O_2 binding in hemoglobin, the following occurs:**

a. The O_2 binds to the Fe in heme group.
b. The heme group is no longer planar.
c. Steric interactions between the heme group and amino acid residues of the polypeptide side chain cause tertiary structural changes in hemoglobin.
d. Both a and b, but not c
e. Both a and c, but not b
f. All of the above

[ ] **Q3.** In hemoglobin, the transition from T state to R state (low to high affinity) is triggered by

a. Fe binding
b. Heme binding
c. Oxygen binding
d. subunit association
e. subunit dissociation

Q4. In deoxy hemoglobin the iron in the heme is coordinated by [ ] (number) [ ] (name of the element) atoms, [ ] (number) located in the protoporphyrin IX and [ ] (number) located within the hemoglobin subunit.

[ ] **Q5. Select all the correct statements below**

a. Oxyhemoglobin has a more compact structure than deoxyhemoglobin.
b. Oxyhemoglobin structure is referred as the T state
c. Subunits reorient in the transition from deoxyhemoglobin to oxyhemoglobin
d. Oxygen binds to the Fe^{3+} state of iron
e. H-bonds are the major noncovalent interactions that stabilize the deoxyhemoglobin

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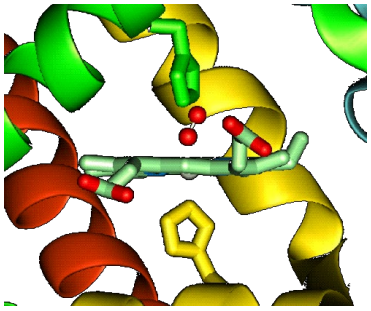
Q6. The binding site shown in the figure is taken from

Oxy- Deoxy- hemoglobin structure.



Q7. Circle proximal histidine on the figure.

Q8. In sickle cell anemia, the basis of the malfunction of the hemoglobin molecule is:

a. incorrect secondary structure
b. substitution of a single amino acid
c. faulty binding of the heme groups
d. reduced affinity for oxygen
e. insufficient iron in the diet

Q9. Sickle cells are:

a. stiff and sticky
b. stiff and flexible
c. round and sticky
d. round and flexible

Q10. Based on your molecular understanding of the SCD, which of the following conditions can trigger a pain crises in a SCD patient. Select all that apply.

a. Dehydration
b. Relaxation
c. Exercise
d. Cold exposure

Q11. The amino acid substitution of Val for Glu in Hemoglobin S results in aggregation of the protein because of ___ between molecules.

a. Covalent bonds
b. Disulfide bonds
c. Salt bridges
d. Hydrophobic effect
e. H-bonds