

HLS-Biochemistry

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Lecture 1

RBCs Metabolism

Medical card

Name _____

Date of b _____

Gender _____

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Date of call _____

HLS-Biochemistry **Lecture 1**

1. Glucose 6-phosphate dehydrogenase is the key regulatory enzyme in the pentose phosphate pathway, responsible for reducing glutathione. What is wrong about the disease caused by the deficiency of this enzyme?

- a. Uncontrolled diabetes mellitus is one of the causes of the hemolytic crises.
- b. May be associated with acute renal failure.
- c. It is an X-linked disorder but females may show its manifestations.
- d. All variants encoding for the enzyme result in clinical consequences.
- e. The best way for the proper diagnosis of the disease is by genetic analysis

ANSWER: D

2. Which of the following statements is true regarding the metabolic pathways in RBCs?

- a. All variants of glucose 6-phosphate dehydrogenase are accompanied by clinical consequences.
- b. The mutation $(721G \rightarrow T)$ of pyruvate kinase is a missense mutation.
- c. Phosphoglucose isomerase deficiency does not affect reduced glutathione regeneration.
- d. Phosphoglycerate kinase deficiency shows 2,3-bisphosphoglycerate accumulation.
- e. Triacylglycerols synthesis in RBCs is not affected by triose phosphate isomerase deficiency.

ANSWER: D

3. Hb inside RBCs performs the following functions EXCEPT?

- a. Allows RBCs to pass in narrow capillaries.
- b. O₂ carriage.
- c. CO₂ carriage.
- d. Acid-base buffer.
- e. Combines with glucose to form glycosylated Hb.

ANSWER: A

4. Which enzymes require zinc (Zn) as a cofactor?

- a. Superoxide dismutase (SOD), Carbonic anhydrase, Lactate dehydrogenase
- b. Hexokinase, Pyruvate kinase, Phosphofructokinase
- c. Catalase, Glutathione peroxidase, Cytochrome oxidase
- d. DNA polymerase, RNA polymerase, Helicase
- e. Adenylate cyclase, Protein kinase A, Phospholipase C

ANSWER: A

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5. In the glycolytic pathway, pyruvate is normally released from inside erythrocytes because of?

- a. The low activity of glyceraldehyde 3-phosphate dehydrogenase.
- b. The high activity of pyruvate kinase.
- c. The maintenance of hemoglobin iron in the reduced form.
- d. The absence of methemoglobin reductase enzyme.
- e. The high concentration of 2,3-bisphosphoglycerate.

ANSWER: C

6. The deficiency of pyruvate kinase is one of the causes of RBC hemolysis, but some of its manifestations can be hidden due to?

- a. Increased production of 2,3-bisphosphoglycerate.
- b. Decreased production of 1,3-diphosphoglycerate.
- c. Increased activity of Na⁺/K⁺ ATPase.
- d. Increased activity of glucose 6-phosphate dehydrogenase.
- e. Increased ATP production

ANSWER: A

7. In pyruvate kinase (PK) deficiency with chronic hemolytic anemia, which statement is ****incorrect****?

- a. Occurs in compound heterozygotes only.
- b. They affect conserved residues in structurally and functionally important domains of PK.
- c. Have a variety of clinical pictures.
- d. Usually missense mutation.
- e. Increased 2,3-BPG levels ease the anemia by lowering the oxygen-affinity of hemoglobin.

ANSWER: A

8. Identify the mismatched group of enzymes and their associated cofactors/metals in oxidative stress in RBCs.

- a. Superoxide dismutase - zinc
- b. Catalase - iron
- c. Glutathione reductase - NADPH
- d. Glutathione peroxidase - selenium
- e. Superoxide dismutase - magnesium

ANSWER: E

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9. Which statement is ****false**** about 2,3-bisphosphoglycerate (2,3-BPG)?

- a. Increased at high altitude.
- b. Eases the anemia of pyruvate kinase (PK) deficiency.
- c. Higher concentration than ATP in RBCs.
- d. Negative allosteric regulator.
- e. Increased in PK-1 deficiency.

ANSWER: E

10. ATP generated in RBCs is utilized in reactions catalyzed by?

- a. Phosphoglucose isomerase
- b. PFK-1, Hexokinases
- c. Fructose-bisphosphate aldolase

ANSWER: B

11. Zinc is a cofactor for?

- a. Carbonic anhydrase
- b. Lactate dehydrogenase
- c. Glycogen synthase

ANSWER: A

12. In the glycolytic pathway, which enzyme deficiencies affect the 2,3-bisphosphoglycerate (2,3-BPG) level?

- a. Hexokinase and pyruvate kinase
- b. Phosphoglycerate kinase and diphosphoglycerate mutase
- c. Aldolase and triose phosphate isomerase
- d. Lactate dehydrogenase and enolase
- e. Phosphofructokinase-1 and glucose-6-phosphate dehydrogenase

ANSWER: B

13. Which enzymes are involved in the nucleotide salvage pathway?

- a. AMP deaminase (AMPDA), Hypoxanthine-guanine phosphoribosyltransferase (HGPRT), Adenine phosphoribosyltransferase (ADPRT)
- b. Ribonucleotide reductase, Thymidylate synthase, Dihydrofolate reductase
- c. DNA polymerase, RNA polymerase, Helicase
- d. Adenylate kinase, Creatine kinase, Lactate dehydrogenase
- e. Carbamoyl phosphate synthetase, Aspartate transcarbamylase, Dihydroorotase

ANSWER: A

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Lecture 2

Hemoglobin synthesis

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1. The asymmetric substitution of the tetrapyrrole ring of heme starts with the activity of the following enzyme?

- a. ALA synthase.
- b. PBG synthase.
- c. Uroporphyrinogen synthase III.
- d. Coproporphyrinogen oxidase.
- e. Coproporphyrinogen decarboxylase.

ANSWER: C

2. Which of the following enzymes in the heme synthetic pathway requires a cosynthase molecule for the asymmetric substitution of the heme tetrapyrrole ring?

- a. ALA synthase.
- b. Uroporphyrinogen synthase III.
- c. PBG deaminase.
- d. Protoporphyrinogen oxidase.
- e. Uroporphyrinogen decarboxylase.

ANSWER: B

3. Different proteins are involved in iron metabolism, among them hepcidin, which is ****not**** characterized by?

- a. It is upregulated by increased iron levels to downregulate ferroportin.
- b. It is downregulated by decreased iron levels to upregulate ferroportin.
- c. Its high expression rate negatively regulates bacterial growth.
- d. Its low expression rate positively regulates bacterial growth.
- e. It directly affects iron exportation from tissues to blood.

ANSWER: E

4. In the heme synthetic pathway, one of the following sets of enzymes is responsible for starting and finalizing the asymmetrical substitutions of the four pyrrole rings of the heme molecule. Select one:

- a. Coproporphyrinogen oxidase and protoporphyrinogen oxidase.
- b. Porphobilinogen deaminase and uroporphyrinogen decarboxylase.
- c. Porphobilinogen synthase and protoporphyrinogen oxidase.
- d. ALA synthase and hydroxymethylbilane synthase.
- e. Uroporphyrinogen synthase III and coproporphyrinogen oxidase.

ANSWER: E

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5. Which one of the following sets of enzymes in the heme synthetic pathway can be inhibited by lead?

- a. ALA synthase and ALA dehydratase.
- b. PBG synthase and PBG deaminase.
- c. Uroporphyrinogen synthase III and ALA synthase.
- d. Uroporphyrinogen decarboxylase and coproporphyrinogen oxidase.
- e. Ferrochelatase and ALA dehydratase.

ANSWER: E

6. Among the following sets of proteins, one set is playing a role in cellular membranes transported iron?

- a. Hepcidin, and matriptase2.
- b. Ceruloplamin and heme carrier protein.
- c. DMT and transferrin.
- d. Duodenal cytochrome B and iron regulatory proteins.
- e. Ferritin and ferroportin.

ANSWER: B

7. Choose the ****wrong**** statement about hepcidin :

- a. Is upregulated in response to iron to induce degradation of ferroportin.
- b. Is upregulated in response to iron to induce more synthesis of ferroportin.
- c. Is synthesized by the liver.

ANSWER: B

8. Which enzyme in heme synthesis converts two of the propionyl side chains into vinyl groups?

- a. Uroporphyrinogen 3 synthase
- b. Uroporphyrinogen decarboxylase
- c. Coproporphyrinogen oxidase
- d. Protoporphyrinogen oxidase
- e. Ferrochelatase

ANSWER: C

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9. The last two enzymes that prepare the tetrapyrrole ring for iron addition in heme synthesis are?

- a. ALA synthase and ALA dehydratase
- b. Uroporphyrinogen III synthase and uroporphyrinogen decarboxylase
- c. Coproporphyrinogen oxidase and protoporphyrinogen oxidase
- d. Ferrochelatase and protoporphyrinogen oxidase
- e. PBG deaminase and coproporphyrinogen decarboxylase

ANSWER: C

10. What is true about the enzyme that converts propionyl side chains into vinyl groups in heme synthesis?

It is a mitochondrial enzyme.

11. Which of the following represents the correct sequence of proteins involved in intestinal iron absorption and transport?

- a. DcytB → DMT1 → Apo-ferritin → Ferroportin → Ceruloplasmin
- b. Ferroportin → DMT1 → Ceruloplasmin → Apo-ferritin → DcytB
- c. DMT1 → Ceruloplasmin → Ferroportin → DcytB → Apo-ferritin
- d. Apo-ferritin → DcytB → Ferroportin → DMT1 → Ceruloplasmin
- e. Ceruloplasmin → Apo-ferritin → DMT1 → DcytB → Ferroportin

ANSWER: A