



Benha University
Faculty of medicine
Medical Biochemistry department

second semester second year exam
Time allowed 1.5 hours
25/5/2013



MODEL ANSWERS

Course code: 0704

Important Instructions For All Students:Please Read Carefully

Allocated time	(1.5 hours including MCQ)
Total allocated marks	(75marks)
Number of papers	(7 papers including MCQ)
Number of questions	(7 questions including MCQ)
Attention	All questions are to be answered

Question number	Mark	Signature
Q I	/14	
Q II	/17	
Q III	/8	
Q IV	/10	
Q V	/4	
Q VI	/12	
Q VII	/10	
Total marks	/75	

Code number

Total mark

/75

Total marks:

Signature:

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Code number

Student's full name:.....

Student's roll name:.....

I*-Question number 1:

(14marks)

A-case 1

(4marks)

A 3 Years old child with light brown color hair and white color skin suffers of mental retardation .His parents took him to the hospital seeking for medical advice .The laboratory investigation showed elevated phenyl alanine , phenyl lactic , phenyl acetic in the urine of the patient .Serum amino acid analysis showed elevated phenylalanine and decreased tyrosine.

i-What is the possible diagnosis of this case? (0.5 mark)

Phenyl-ketonuria

ii-Name the enzyme defected in this case. (one mark)

Phenylalanine hydroxylase enzyme

iii- Explain on biochemical bases why the color of the hair is light brown color hair and the skin color is white. (1.5 marks)

Phenylalanine hydroxylase enzyme deficiency leads to decreased conversion of phenylalanine to tyrosine leading to decreased formation of melanin which is the pigment present in skin and hair

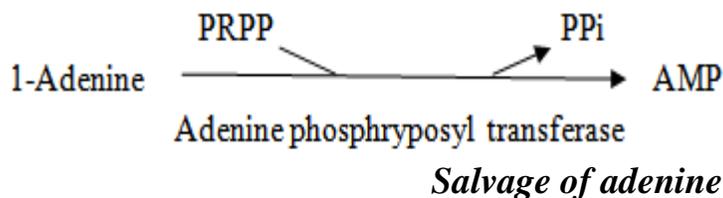
iv-Explain the treatment for this case? (one mark)

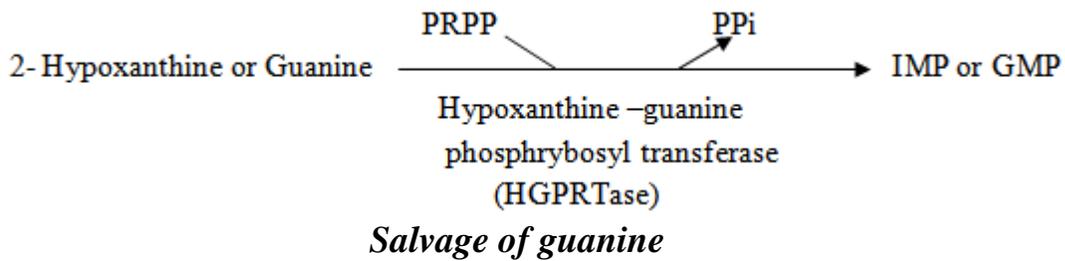
Treatment: must be started early in the first days of life by giving diet very low in phenyl alanine and rich in tyrosine.

B- Describe the salvage pathway of purine nucleotides biosynthesis and the disorders affecting it? (10marks)

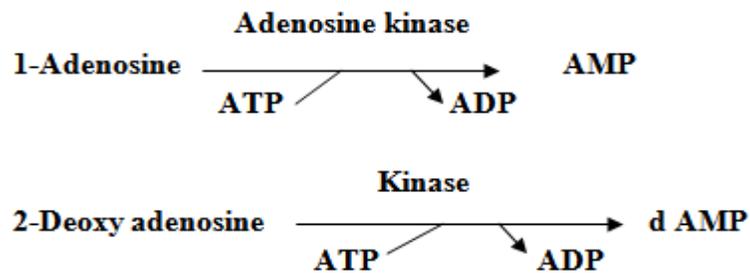
The significance of salvage system is to supply purine nucleotides to certain tissue or cell where the de novo synthesis is not active e.g. brain, red cells and lymphocytes. There are two systems for purine salvage.

A- Salvage of free purines: (5marks each 2.5 marks)





B- Salvage of purine nucleosides (3 marks each 1.5 marks)



Lesch-Nyhan syndrome: Congenital deficiency of HGPRTase enzyme leading to increase PRPP so de novo synthesis of purines increase much more than tissue needs so uric acid formation increase and it is associated with mental retardation. **(2 marks)**

II*Question number 2- Enumerate: **(17 marks)**

A-Functions of amide group of glutamine (four only) **(4 marks)**

- 1-** The amide group of glutamine has important functions as follows: (4 each one mark)
- a- Conversion of aspartate to asparagine.
 - b- Formation of N₃ & N₉ of purines.
 - c- Formation of carbamoyl phosphate in pyrimidines synthesis N₃.
 - d- Conversion of UTP to CTP.
 - e- Synthesis of amino sugars.
 - f- Conversion of XMP to GMP.
 - g- Conversion of nicotinic acid to nicotinamide which is important for NAD⁺ & NADP⁺ formation.

B-Causes of deficiency of vitamin D:**(4marks each one mark)**

1. Deficiency of vitamin D can occur in people who are not exposed to sunlight properly.
2. Nutritional deficiency of calcium or phosphate may also produce similar clinical picture.
3. Malabsorption of vitamin (obstructive jaundice and steatorrhea).
4. Abnormality of vitamin D activation due to Liver and renal diseases.

C-Importance and functions of vitamin K:**(3marks each one mark)**

- a. Vitamin K is necessary for coagulation. Factors dependent on vitamin K are Factor II (pro- thrombin); Factor VII ; Factor IX and Factor X.
- b. All these factors are synthesised by the liver as inactive zymogens. They undergo post-trans-lational modification; gamma carboxylation of glutamic acid residues. These are the binding sites for calcium ions. The gamma carboxy glutamic acid (GCG) synthesis requires vitamin K as a co-factor .
- c. Vitamin K dependent gamma carboxylation is also necessary for the functional activity of structural proteins of kidney, lung, spleen and Osteocalcin which is a unique protein synthesised by osteoblasts and seen only in bone. It binds tightly to hydroxy apatite crystals of bone; this binding is dependent on the degree of gamma carboxylation

D-Causes of proteinuria:**(3marks each one mark)****1-Physiological proteinuria:**

- Following severe muscular exercise.
- After high protein meal.
- Orthostatic or postural due to temporary impairment in renal circulation.

2-Pathological proteinuria: may be

- **Prerenal:** in case of heart failure and hypertension.
- **Renal:** due to increased permeability of renal glomeruli as a result of kidney diseases e.g.glomerulonephritis ,nephritic syndrome and poisoning of renal tubules by heavy metals like mercury and arsenic.
- **Post renal:** due to inflammation of the urinary tract.

3-Bence Jones proteinuria :

These are special types of globulines of low molecular weight ,they are found in serum and excreted in urine in case of myloid leukemia and multiple myeloma .They are precipitated at 45 -60°C but redissolve on boiling and reprecipitate on cooling.

E-Non protein nitrogenous compounds(NPN) in blood & mention the normal level of each one: (3marks)

- 1- Urea: (10-50 mg/dl) (0.5 mark)
- 2- Free amino acids: (3-6 mg/dl). (0.5 mark)
- 3- Uric acid: 3-6 mg/dl (in females), 4-7 mg/dl (in males). (0.5 mark)
- 4- Creatine (0.2 -0.9 mg/dl) and Creatinine (0.6 -1.4 mg/dl) (one mark)
- 5- Ammonia: (Less than 0.05 mg/dl) (0.5 mark)

III*-Question number 3: (8marks)

A-Complete the following: (2marks each 0.5 mark)

1-Serotonin is catabolized by ...monoamine oxidase...(MAO)....to5-hydroxy indole acetic acid (5-HIAA)...

which is increased
in...argentaaffinoma.....

2-catecholamines are catabolized to...vanillyl mandelic acid (VMA).....

B- Name the enzyme defects in the following diseases: (6 marks each 0.5 mark)

Diseases	Enzyme defects
Neonatal tyrosinemia	P-hydroxyphenylpyruvate hydroxylase
Alkaptonuria	Homogentisate oxidase
Cystathioninuria	Cystathioninase
Hyperargininemia	Arginase
Citrullinemia	<u>Argininosuccinate synthetase</u>
TypeI hyperammonemia	Carbamoyl phosphate synthetase I

IV- Question number 4: Explain on biochemical basis: (10 marks)

A-Different types of Anemia can occur in vitamin C deficiency: (3 marks each one mark)

- 1. Microcytic hypochromic anemia:** The reasons for this type of anemia may be due to:
 - a. Loss of blood by hemorrhage.
 - b. Decreased iron absorption.
- 2. Macrocytic hyperchromic anemia:** The reason for this type of anemia is decreased activation of folic acid to tetrahydro folic acid.
- 3. Accumulation of met-hemoglobin leading to functional anemia.**

B--Deficiency of vitamin B6 produce convulsions: (2marks)

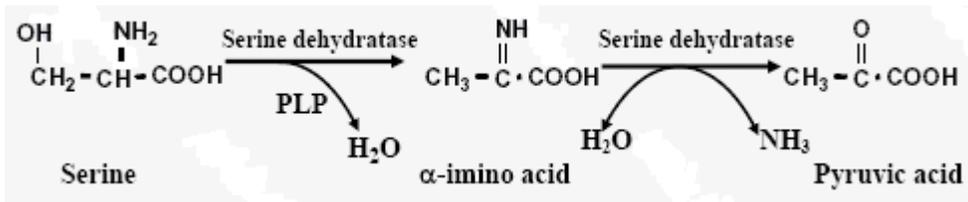
Deficiency of vitamin B6 leads to decreased formation of GABA from glutamic acid by decarboxylase which need pyridoxal phosphate coenzyme from vit B6 .GABA is a chemical inhibitory neurotransmitter

C-Methionine amino acid is a lipotropic factor: (2marks)

- 1- It forms ethanolamine and choline which helps in the formation of phospholipids and so help is synthesis of VLDL which helps mobilization of fat from liver.
- 2- It is an essential amino acid needed for synthesis of apoprotein needed for VLDL.

D-Serine is a glucogenic amino acid (explain the reaction with formulae): (3marks)

Serine by serine dehydratase gives glucose



Conversion of serine to pyruvic acid

V- Question number 5-Differentiate between colostrum and mature milk: (4 marks)

Colostrums:

It is a yellowish fluid secreted by the mammary gland during the first week of lactation. It differs from mature milk in the following point:

- 1- **Proteins:** They are higher in colostrums (5.5% in human colostrums) than mature milk .It is rich in immunoglobulin which gives the babies immunity.
- 2- **Minerals:** They are higher than mature milk (0.35% in human colostrums).
- 3- **Vitamins:** They are higher than in mature milk.
- 4- **Fats:** They are lower than in mature milk (2.5% in human colostrums),but with higher content of fat soluble vitamins and carotenes, and this is due to its yellowish color.
- 5- **Lactose:** it is lower than in mature milk (3.5 % in human colostrums), making is less sweet and more suitable for the newly born infant.

Mature milk:

Human milk contains; water (87.7%), lactose (7.4%), lipids (3.5%), proteins (1.2%), minerals (0.2%) and vitamins. The latter is rich in Vit A & B, poor in Vit C & K and contains fair amounts of the other vitamins .

VI- Question number 6 : (12marks)

A-Define and explain causes of creatinuria? (5marks)

Creatinuria: (one mark)

It is increase excretion of creatine in urine under normal physiological or pathological states.

Causes of physiological creatinuria: (one mrk)

- 1- In young children (decreased muscle mass).
- 2- In females during pregnancy and early postpartum period.

Causes of pathological creatinuria: (3 mark)

In all condition of muscle wasting which include:

- 1- Starvation.
- 2- Diabetes mellitus.

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- 3- Fevers.
 - 4- Hyperthyroidism.
 - 5- Hypogonadism.
 - 6- Vit E deficiency.
 - 7- Degenerative muscle diseases (myopathies).

B-List and explain metabolic errors of glycine amino acid: (3marks)

1- Primary hyperoxaluria : (2 marks)

- Congenital disease caused by decreased metabolism of glyoxalic acid either by:
 - a- Oxidative decarboxylation to formic acid, or
 - b- Transamination to glycine.
- Glyoxalic acid is accumulated and is oxidized to oxalic acid
- There is excretion of large amount of oxalate in urine.
- Oxalate reacts with calcium forming insoluble Ca, oxalate stone in urinary tract.
- Large stones if bilateral may cause renal failure.

2- **Hyperglycinemia:** There is increased glycine level in blood, caused by deficiency of glycine cleavage system. (0.5 mark)

3- **Glycinuria:** There is increased excretion of glycine in urine, caused by congenital decreased renal tubular reabsorption of glycine. (0.5 mark)

C-Define Xenobiotics & Detoxification –list the compounds that are detoxified in our body?

(4marks)

Detoxification; is the biochemical process whereby the toxic substances become less harmful, more water soluble and easily excreted from the body. **(one mark)**

Xenobiotics; are compounds which may be ingested or taken as drugs or compounds produced in the body by bacterial mechanism. **(one mark)**

The compounds that are detoxified include: (2 marks)

1. Compounds accidentally ingested like preservatives and food additives.
2. Drugs taken for therapeutic purposes.
3. Compounds produced in the body which are to be eliminated e.g. Steroids and bilirubin.
4. Compounds produced by bacterial metabolism e.g. Amines produced by decarboxylation of amino acids:
 - i. Tyrosine → Tyramine
 - ii. Tryptophan → Tryptamine.



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7-Choose the correct answers : (10 marks each 1 mark)

1-Transmethylation of guanido acetic acid gives:

- A**
(A) Creatine
(B) Creatinine
(C) Choline
(D) n-methyl nicotinamide

2- NH₃ is removed from brain mainly by:D

- (A) Creatinine formation
(B) Uric acid production
(C) Urea formation
(D) Glutamine formation

3- Pellagra occurs in population dependent on:C

- (A) Wheat (B) Rice
(C) Maize (D) Milk

4- The precursor of CoA is:D

- (A) Riboflavin (B) Pyridoxamine
(C) Thiamin **(D) Pantothenate**

5- Lesch-Nyhan syndrome, the sex linked recessive disorder is due to the lack of the enzyme:A

- (A) Hypoxanthine-guanine phosphoribosyl transferase**
(B) Xanthine oxidase
(C) Adenine phosphoribosyl transferase
(D) Adenosine deaminase

6- The major catabolic product of pyrimidines in human is:A

- (A) β -Alanine** (B) Urea
(C) Uric acid (D) Guanine

7- The amino acid which detoxicated benzoic acid to form hippuric acid is:A

- (A) Glycine** (B) Alanine
(C) Serine (D) Glutamic acid

8- Calcitriol is:D

- (A) 1-hydroxy cholecalciferol
(B) 25-hydroxy cholecalciferol
(C) 24, 25-dihydroxy cholecalciferol
(D) 1, 25-dihydroxy cholecalciferol

9- Milk is a good source of all of the following Except:B

- (A) Essential amino acids
(B) Vitamin C
(C) Galactose
(D) Calcium and phosphorous

10- Bence Jones proteins may be excreted in urine of patients suffering from:C

- (A) Tuberculosis (B) Diabetes mellitus
(C) Multiple myeloma (D) Hyperthyroidism

NB:

Oral exam will be held on Saturday 25th & Sunday 26th of May 2013 in medical biochemistry department.

Student numbers 1- 200 will attend the oral exam at 11:30 am on Saturday 25th of May 2013

Student numbers 200-(till the end) will attend the oral exam at 10 am on Sunday 26th of May 2013

Good luck

