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Student's Biochemistry

Important Solved Questions with Clinical Cases (All India)

Veena Shripad Hatolkar

JAYPEE

Contents

1. Chemistry and Metabolism of Carbohydrates 1
2. Chemistry and Metabolism of Lipids 7
3. Chemistry of Proteins and Metabolism of Amino Acid 12
4. Chemistry and Metabolism of Hemoglobin 21
5. Biological Oxidation 25
6. Chemistry and Metabolism of Nucleic Acids 28
7. Replication, Transcription, Translation, and DNA Recombinant Technology 32
8. Enzymes 37
9. Mineral Metabolism 40
10. Vitamins 44
11. Starvation 48
12. Energy Metabolism and Nutrition 49
13. Free Radical and Antioxidants 50
14. Xenobiotics/Detoxification 52
15. Organ Function Tests 54
16. Immunology and Oncology, Oncogenesis 56
17. Extracellular Matrix 57
18. Water, Electrolyte, and Acid-Base Balance 59
19. Case Studies in Biochemistry 62

3 CHAPTER

Chemistry of Proteins and Metabolism of Amino Acid

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BRIEF ANSWER QUESTIONS (2 MARKS)

1. What are essential amino acids ? Enumerate them.

Ans: The amino acids which cannot be synthesized in body and therefore need to be supplied through diet are called essential amino acids.
For example: (1) Methionine, (2) Arginine, (3) Threonine, (4) Tryptophan, (5) Valine, (6) Isoleucine, (7) Leucine, (8) Phenylalanine, (9) Histidine, (10) Lysine.
Arginine and histidine are semi-essential as they cannot be synthesized by children but partly can be synthesized by adults.

2. Which is the 21st amino acid ? Name the enzymes which contain it.

Ans: Selenocysteine is the 21st amino acid. It is found at active site of certain enzymes like glutathione peroxidase, glycine reductase, 5' deiodinase, thioredoxin reductase.

3. What is a zwitterion and isoelectric pH?

Ans: Amino acid rarely exist in neutral form with free carboxylic (-COOH) and free amino group (NH_2). In strong acidic medium the amino acid is positively charged.
In strong alkaline medium, the amino acid acquires negative charge.
Each amino acid has a characteristic pH at which it carries both positive and negative charge and exists as zwitterion or dipolar ion.
This characteristic pH is known as isoelectric pH of that amino acid (e.g., leucine, pI -6)

4. Enumerate the biologically important peptides with example.

Ans: Tripeptide: Glutathione, thyrotropin releasing hormone
Pentapeptide: Enkephalins
Octapeptide: Angiotensin II
Nonapeptide: Oxytocin,vasopressin (ADH), bradykinin
Decapeptide: Angiotensin I

5. Name the amino acids present in glutathione and give its functions. (any two)

Ans: Glutathione consists of glycine, glutamic acid and cysteine.
Functions of glutathione:
1. Maintains RBC membrane structure and integrity and protects hemoglobin from getting oxidised by agents such H_2O_2.
2. Involved in detoxication process.

3. Involved in transfer of amino acid in intestine and kidney tubules via γ-glutamyl cycle or Meister's cycle.
4. Serves as a redox buffer by maintaining equilibrium between its oxidised and reduced state within a cell.

6. Name the bonds responsible for protein structure.

Ans: Covalent bonds, e.g., peptide bond, disulfide bond
Non-covalent bonds, eg., hydrogen bond, hydrophobic bond, electrostatic/ionic bond, Van der waal's interactions.

7. Nutritional classification of proteins.

Ans: Based on nutritional point of view proteins are classified into three categories:

1. **Complete protein:** Have all ten essential amino acids in required proportion by human body to promote growth, e.g., egg albumin, milk casein.
2. **Partially incomplete:** These proteins partially lack one or more essential amino acids, e.g., wheat and rice proteins (lack lys, thr).
3. **Incomplete proteins:** These completely lack one or more essential amino acids, e.g., gelatin (lack trp), zein.

8. Define transamination and write its importance.

Ans: The transfers of amino group from an amino acid to keto acid is known as transamination catalysed by transaminases/aminotransferases and PLP as coenzyme. Its importance is:
A. Synthesis of nonessential amino acid
B. Transfer of amino group.

9. Diagrammatic presentation of amino acid metabolism.

Ans:

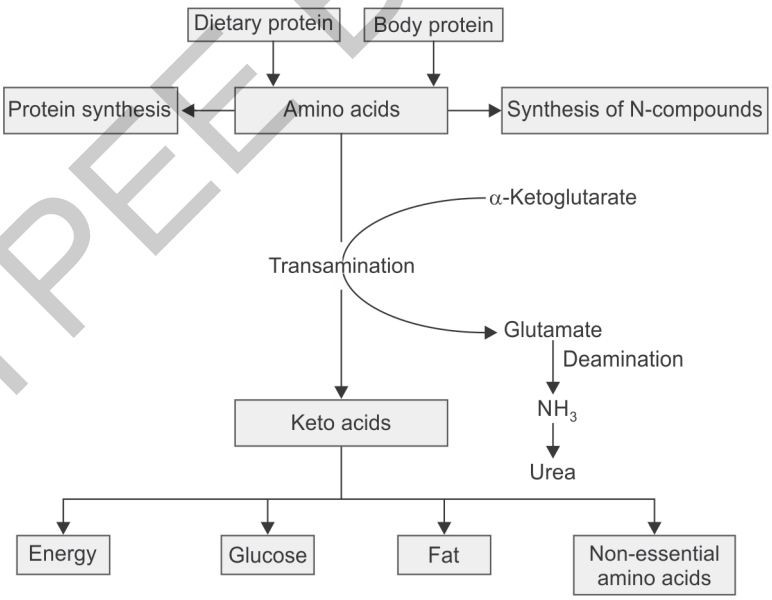

Fig. 1: Overview of amino acid metabolism.

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10. Diagrammatic presentation of body amino acid pool.

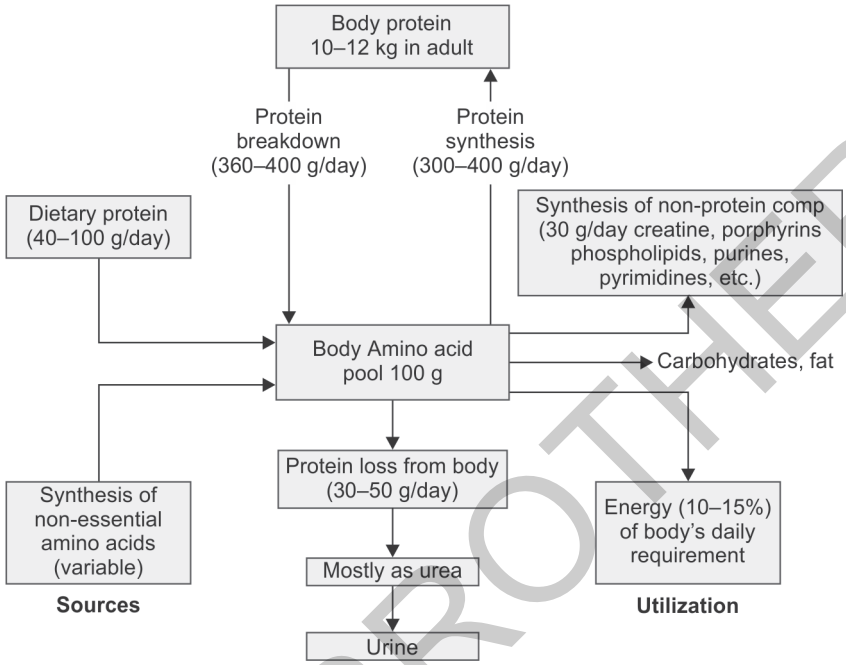

Fig. 2: Body amino acid pool.

11. What is transdeamination?

Ans: Transamination is followed by oxidative deamination is known as transdeamination

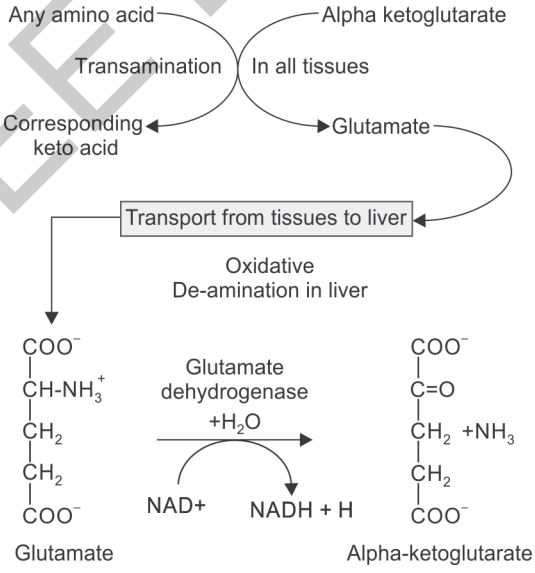

Fig. 3: Transdeamination.

12. Write sources of ammonia in body.

Ans: 1. From transamination and deamination of amino acids
2. From amino groups of purines and pyrimidines

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3. Biogenic amines
4. By action of intestinal bacteria on urea

13. What are the functions of ammonia?

Ans: Ammonia is not just a waste product of nitrogen metabolism. It is involved in synthesis of many imp compounds in body.

1. Non-essential amino acids
2. Purines and pyrimidines
3. Amino sugars, asparagine, etc.
4. Involved in acid-base balance of body as ammonium ions (NH_4^+).

14. Enumerate the metabolic defects in urea cycle with the enzymes involved.

Ans:

	Defect	**Enzymes involved**
1.	Hyperammonemia type I	Carbamoyl phosphate synthase I
2.	Hyperammonemia type II	Ornithine transcarbamylase
3.	Citrullinemia	Argininosuccinate synthase
4.	Argininosuccinic aciduria	Argininosuccinase
5.	Hyperargininemia	Arginase

15. Enumerate the specialized products synthesized from glycine.

Ans:

1. Glutathione
2. Heme
3. Creatine
4. C4, C5 and N7 of purine ring
5. Glycocholic acid and chenodeoxycholic acid

16. What is sparing action of tyrosine on phenylalanine?

Ans:

- Phenylalanine and tyrosine are aromatic amino acid.
- Phenylalanine is essential and tyrosine is nonessential.
- Phenylalanine is converted to tyrosine in body.
- So ingestion of tyrosine in diet can reduce the dietary requirement of phenylalanine. This is referred as sparing action of tyrosine on phenylalanine.

17. Give reaction for synthesis of tyrosine from phenylalanine and name the related disorder.

Ans: Disorder is phenylketonuria due to deficiency of phenylalanine hydroxylase.
Reaction:

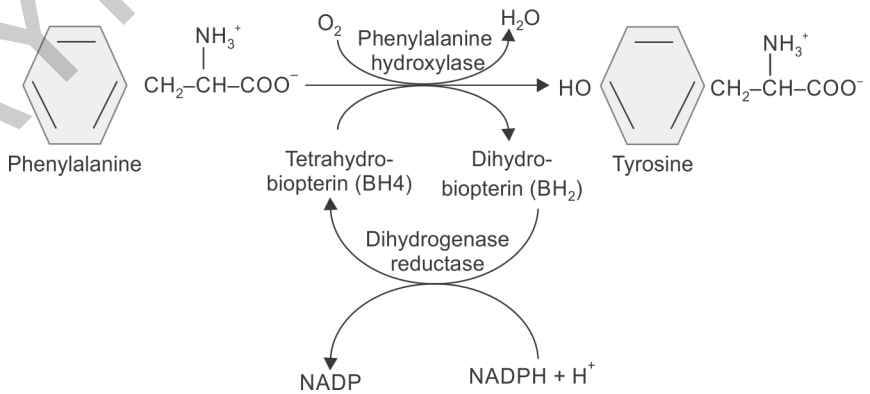

Fig. 4: Tyrosine synthesis.

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18. Enumerate the biologically important compounds formed from tyrosine.

Ans: 1. Melanin
2. Dopamine
3. Norepinephrine
4. Thyroxine

19. Outline the metabolism of phenylalanine in phenylketonuria (PKU).

Ans:

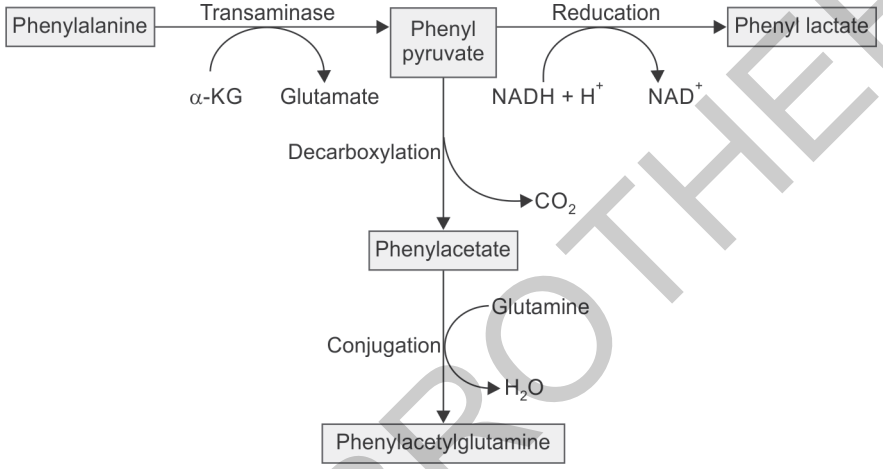

Fig. 5: Phenylalanine metabolism in PKU.

All these metabolites are excreted in urine in high concentration in phenylketonuria.

20. Name the pathways involved in metabolism of tryptophan and the important products derived from these pathways.

Ans: Kynurenine pathway → NAD^+, $NADP^+$
Serotonin pathway → Serotonin and melatonin

21. Define transmethylation. Give two examples.

Ans: Transfer of methyl group from active methionine to an acceptor is known as transmethylation. Enzyme is methyl transferase.

Methyl group accepter	*Methylated product*
Guanidinoacetate	Creatine
Nor-epinephrine	Epinephrine
Epinephrine	Metanephrine
Ethanolamine	Choline
Serine	Choline

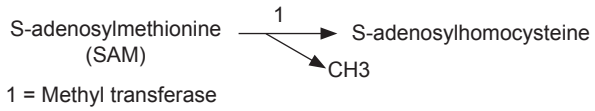

Fig. 6: Transmethylation.

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22. Name the sulfur containing amino acids and the disorders associated with their metabolism.

Ans: *Sulfur containing amino acid:* Methionine, cysteine and cystine.
Disorders associated are: Cystinuria, homocystinuria, cystinosis.

23. Enumerate the coenzymes of tetrahydrofolate involved in one carbon metabolism.

Ans: N^{10} formyl THF
N^5N^{10} Methenyl THF
N^5 N^{10} Methylene THF
N^5 Methyl THF

24. Name the branched chain amino acids and the disorders related to them.

Ans: *Branched chain amino acids are:* Valine, leucine, isoleucine.
Disorders associated are: Maple syrup urine disease, intermittent branched chain ketonuria, isovaleric acidemia, hypervalinemia.

25. Name the amino acid involved in synthesis of nitric oxide. Give two functions of nitric oxide.

Ans: Arginine is involved in synthesis of nitric oxide.
Functions of nitric oxide:
- ❖ Acts as vasodilator involved in regulation of blood pressure
- ❖ Involved in penile erection.

26. Name the amino acid involved in carnitine synthesis and write its biomedical role.

Ans: Methionine and lysine are involved in synthesis of carnitine.
Carnitine plays key role in fatty acid oxidation for transport of acyl-CoA across mitochondrial membrane in the form of acyl carnitine.

27. Name the specialized products formed from glutamate.

Ans: 1. Glutathione
2. N-acetylglutamate
3. γ-aminobutyric acid (GABA)
4. γ-carboxyglutamate in clotting factors 2,7,9,10

28. What are zymogens? Give two examples of it.

Ans: They are proenzymes, inactive at the time of secretion but will be activated in GIT, e.g., trypsinogen, chymotrypsinogen pepsinogen, proelastase.

29. What is an endopeptidase? Give examples of it.

Ans: It is an enzyme which acts on peptide bonds inside the protein molecule during digestion process, e.g., trypsin, pepsin.

30. What is an exopeptidase? Give examples of it.

Ans: It is an enzyme which acts on the terminal end (amino or carboxyl) of a protein liberating amino acids sequentially one at a time, e.g., carboxypeptidase, aminopeptidase.

31. Name the precursors involved in urea synthesis.

Ans: Carbon atom derived from CO_2.
First nitrogen atom derived from ammonia and the other from aspartic acid.

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32. What are the two carbamoyl phosphate synthetase?

Ans:

CPS I	CPS II
1. Urea synthesis	1. Pyrimidine synthesis
2. Present in mitochondria	2. In cytosol

33. Write the characteristic features and enzyme defective in homocystinuria.

Ans: Characteristic feature: Mental retardation, subluxation of lens, thrombosis.
Enzyme defective: β-cystathionine synthase.

34. Name the conditions in which pellagra is manifested and write its clinical features.

Ans: Characteristic feature of pellagra: Diarrhea, dermatitis, dementia, death (if not treated) condition in which pellagra is manifested.

1. Niacin deficiency
2. Pyridoxal deficiency
3. Tryptophan deficiency/Hartnup's disease
4. Carcinoid syndrome

35. Name the aminoaciduria associated with mental retardation and their enzyme defects.

Ans:

Aminoaciduria	**Defective enzyme**
1. Phenylketonuria	Phenylalanine hydroxylase
2. Maple syrup urine disease	α-keto acid dehydrogenase
3. Homocystinuria	Cystathionine β-synthase

36. Write salient features and diagnostic test for phenylketonuria.

Ans: Clinical feature:

1. Mental retardation
2. Seizures
3. Tremors
4. Growth failure and delayed milestones

Diagnostic tests:

1. Increased blood phenylalanine levels by Guthrie bacterial bioassay test.
2. Ferric chloride test on urine for detection of phenylpyruvate
3. Prenatal diagnosis by cultured amniotic cells
4. Use of DNA probes

37. Name any two clinically important biogenic amines with function.

Ans:

Biogenic amine	**Amino acid**	**Biological importance**
1. Histamine	Histidine	Vasodilator, BP ↓ ↑ HCl secretion and pepsin Anaphylaxis
2. γ-aminobutyric acid	Glutamic acid	Inhibitory neurotransmitter
3. Taurine	Cysteine	Bile acid constituent
4. Tryptamine	Tryptophan	Vasoconstrictor
5. Ethanolamine	Serine	Choline formation

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38. Show schematically biosynthesis of creatine.

Ans:

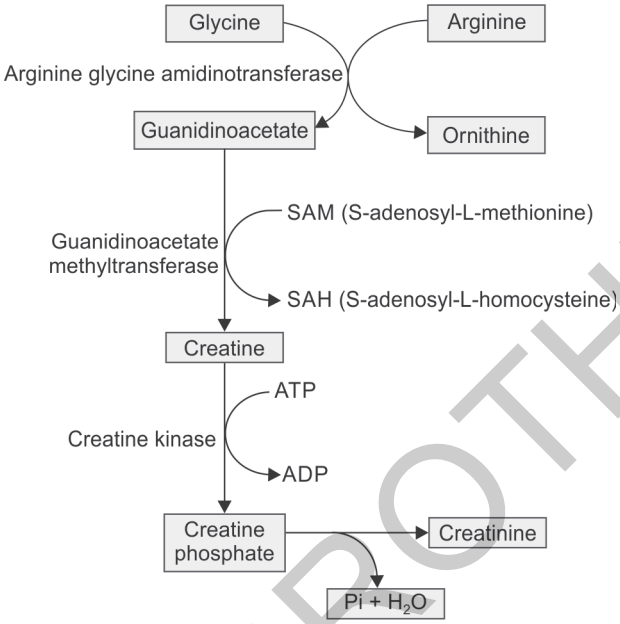

Fig. 7: Biosynthesis of creatine.

39. What is Lohman reaction?

Ans: Creatine-P is the reservoirs of energy in muscles. When muscles contract energy is derived from breakdown of ATP to ADP+Pi. ATP most be reformed quickly to supply the energy.

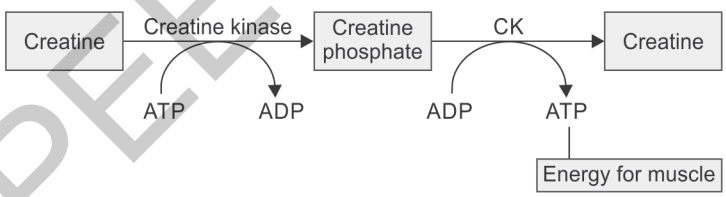

Fig. 8: Lohman reaction.

The high energy phosphate of creatine-P is transferred to ADP and ATP is formed. This reaction is called Lohman reaction and it takes place during muscle activity.

40. Enumerate any four factors affecting nitrogen balance.

Ans: Following are the factors affecting nitrogen balance:

- Growth
- Hormone
- Pregnancy
- Acute and chronic illness
- Protein deficiency

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SHORT ANSWER QUESTIONS (5 MARKS)

1. Essential and non-essential and semi-essential amino acids with examples.
2. Classification of amino acids with examples.
3. Functional classification of proteins.
4. Isoelectric pH and zwitterion.
5. Nutritional classification of proteins.
6. Biologically important peptides.
7. Primary structure of protein.
8. Secondary structure of protein.
9. Tertiary structure of protein.
10. Denaturation of protein.
11. Digestion and absorption of proteins.
12. Transamination and its importance.
13. Deamination.
14. Transmethylation reaction and its importance.
15. Toxicity of ammonia in brain.
16. Amino acid pool.
17. Nitrogen balance.
18. Functions of plasma protein.
19. Separation technique for plasma protein or electrophoresis.
20. Phenylketonuria.
21. Alkaptonuria.
22. Cystinuria.
23. Albinism.
24. Maple syrup urine disease.
25. Hartnup's disease.
26. Urea cycle.
27. Important compound derived from glycine.
28. Important compound derived from tryptophan.
29. Biogenic amines and its importance.
30. Serotonin.

LONG ANSWER QUESTIONS (10 MARKS)

1. Define and classify proteins with suitable examples.
2. Describe structural organization of protein at different levels with example.
3. Describe digestion and absorption of proteins and different stages of metabolism of amino acids
4. Describe formation and transport of ammonia. Write reactions of urea cycle. Add a note on inborn errors of urea cycle.
5. Describe glycine metabolism in detail and its role in biosynthesis of specialized products. Add a note on inborn errors of it.
6. Describe phenylalanine and tyrosine metabolism in detail. Add a note on inborn errors associated with their metabolism.
7. Describe metabolism of sulfur containing amino acids. Add a note on inborn errors associated with the pathway.

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