Aim AIIMS

Biochemistry

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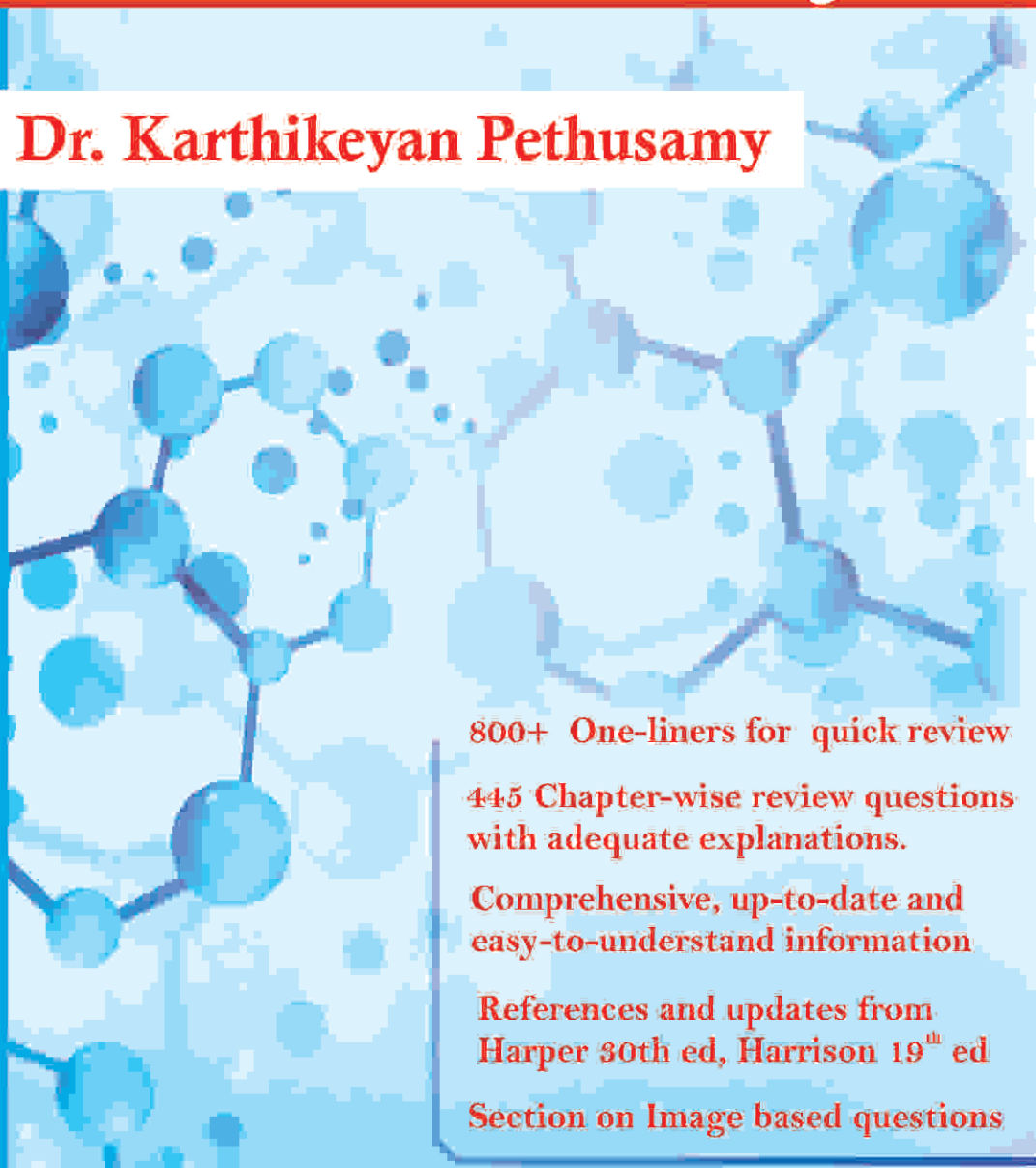


- 800+ One-liners for quick review
- 445 Chapter-wise review questions with adequate explanations.
- Comprehensive, up-to-date and easy-to-understand information
- References and updates from Harper 30th ed, Harrison 19th ed
- Section on Image based questions

ALTIS VORTEX

Metabolism of Amino Acids

1. Which of the following is the storage form of proteins in human body?
a. Collagen b. Titin
c. Both d. None

2. To which of the following amino acids of target protein ubiquitin is covalently added?
a. Glycine b. Valine c. Serine d. Lysine

3. What is the nature of bond between ubiquitin and the amino acid of the target protein?
a. Glycosidic bond b. Hydrogen bond
c. Isopeptide bond d. Hydrophobic interaction

4. What is the minimum number of ubiquitin molecules that must be added to a protein for its degradation in a proteasome?
a. 1 b. 3 c. 4 d. 6

5. Which of the following is NOT true about ubiquitin-proteasomal system?
a. Intracellular, short-lived regulatory proteins are degraded
b. Non-α peptide bond is formed by ubiquitin
c. Amino terminal Asp or Arg accelerate ubiquitination
d. ATP independent

6. Which of the following organ is the major source of alanine for liver in between meals?
a. Muscle b. Brain
c. Kidney d. Adipocyte

7. In postprandial state, which of the following amino acid is extracted predominantly by muscle, having been spared by the liver?
a. Alanine b. Valine
c. Glutamate d. Glutamine

8. Which is the only amino acid that undergoes significant amount of oxidative deamination in the hepatic mitochondira?
a. Lysine b. Alanine
c. Threonine d. Glutamate

Ans. 1. (d) 2. (d) 3. (c) 4. (c) 5. (d) 6. (a) 7. (b) 8. (d)

9. Ammonia is detoxified in brain into _____

a. Urea b. Glutamine
c. GABA d. Glutamate

10. In transaminases, to which of the following amino acids is PLP covalently attached?

a. Lysine b. Alanine
c. Threonine d. Glutamate

11. Which of the following is an allosteric activator of hepatic glutamate dehydrogenase?

a. ATP b. ADP c. GTP d. NADH

12. All of the following are true statements about urea cycle, EXCEPT

a. Operates in both cytoplasm & mitochondria
b. 4 mol of ATPs are required for synthesis of 1 mol of urea
c. There is no net loss or gain of ornithine
d. Ornithine transcarbamoylase gene is present in X-chromosome

13. During co-translational synthesis of selenocysteine, $tRNA^{Sec}$ is initially charged with

a. Selenocysteine b. Serine
c. Cysteine d. Methionine

14. Glycine is formed from the transamination of

a. Alanine b. Aspartate
c. Glutamate d. Glyoxylate

15. In ureotelic animals, carbamoyl group are transferred to

a. Urea b. Uric acid
c. Ornithine d. Creatine

16. Which of the following enzyme is not used by the urea cycle?

a. CPS-I b. CPS-II
c. Arginase d. Argininosuccinase

17. Hydrolysis occurs at which step of urea cycle

a. Cleavage of arginine
b. Formation of Argininosuccinate
c. Formation of citrulline
d. Formation of fumarate

18. All of the following amino acids are converted to Succinyl-CoA EXCEPT

a. Methionine b. Isoleucine
c. Valine d. Leucine

Ans. 9. (b) 10. (a) 11. (b) 12. (b) 13. (b) 14. (d) 15. (c) 16. (b) 17. (a) 18. (d)

19. Taurine is produced from

a. Arginine b. Leucine
c. Valine d. Cysteine

20. Cysteine can be synthesised from

a. Methionine b. Arginine
c. Histidine d. Lysine

21. Tryptophan loading test is done to detect the deficiency of

a. Folate b. PLP c. TPP d. Biotin

22. Tetrahydrobiopterin is required for the metabolism of

a. Arginine b. Lysine
c. Phenylalanine d. Serine

23. All of the following are derived from tyrosine, EXCEPT

a. Norepinephrine b. Melatonin
c. Thyroxine d. Dopamine

24. Which of the following compounds is accumulated in Maple Syrup urine disease?

a. Homogentisate
b. Homocysteine
c. Branched chain α-keto acids
d. Methyl Malonyl-CoA

25. Homocystinuria is due to abnormal metabolism of

a. Methionine b. Valine
c. Cysteine d. Leucine

26. Enzyme defective in Tyrosinemia Type I is

a. Fumaryl acetoacetate hydrolase
b. Tyrosine amino transferase
c. Para hydroxyl phenyl pyruvate hydroxylase
d. Para hydroxyl phenyl pyruvic acid oxidase

27. Richner-Hanhart syndrome is due to defect in

a. Homogentisate oxidase
b. Partial defect in HGPRTase
c. Fumarylacetoacetate hydrolase
d. Tyrosine aminotransferase

28. In Hartnup's disease, the absorption of which of the following is defective?

a. Tryptophan b. Tyrosine
c. Xanthurenic acid d. Homogentisic acid

Ans. 19. (d) 20. (a) 21. (b) 22. (c) 23. (b) 24. (c) 25. (a) 26. (a) 27. (d) 28. (a)

29. Guthrie's bacterial inhibition test for PKU detects
a. Phenyl alanine b. Phenyl pyruvate
c. Phenyl lactate d. All of the above

30. $FeCl_3$ screening test for PKU detects
a. Phenyl alanine b. Phenyl pyruvate
c. Phenyl acetate d. Phenyl lactate

31. Mousy odor of urine in phenylketonuria is due to
a. Phenyl alanine
b. Phenyl pyruvate
c. Phenyl acetate
d. Phenyl lactate

32. In Phenylketonuria, the first line therapy is:
a. Replacement of the defective enzyme
b. Replacement of the deficient product
c. Limiting the substrate for deficient enzyme
d. Giving the missing amino acid by diet

33. Carbon skeleton of which of the following amino acids cannot produce intermediates of TCA cycle?
a. Histidine b. Threonine
c. Proline d. Leucine

34. Which of the following is the substrate for rate-limiting enzyme of polyamine biosynthesis?
a. Histidine b. Cadaverine
c. Ornithine d. Anandamide

35. Carbon skeleton of all of the following amino acids enter the TCA cycle at the level of α-keto glutarate, EXCEPT
a. Proline b. Methionine
c. Arginine d. Histidine

36. Nitisinone can be used in
a. Alkaptonuria
b. Hereditary tyrosinemia type I
c. Both
d. None

37. Which of the following group of amino acids are absorbed in the enterocytes through Meister cycle?
a. Basic amino acids
b. Dibasic amino acids
c. Dicarboxylic amino acids
d. Neutral amino acids

Ans. 29. (d) 30. (b) 31. (c) 32. (c) 33. (d) 34. (c) 35. (b) 36. (c) 37. (d)

38. A boy was brought with complaints of red, scaly rash and mild cerebellar ataxia by his mother with the history of same symptoms in her older daughter. It was clear that the boy did not have the usual dietary-deficiency form of pellagra. The boy was found to excrete excessive free amino acids in the urine. Which of the following amino acid transporter could be defective in this boy?

a. Dibasic aminoacid transporter
b. Dicarboxylic amino acid transporter
c. Imino acid transporter
d. Neutral amino acid transporter

39. 5-hydroxyindoleacetate is a catabolic product of

a. Phenylalanine b. Tyrosine
c. Tryptophan d. Histidine

40. Which of the following chemical reaction is involved in the production of GABA from glutamate?

a. Deamination
b. Decarboxylation
c. Oxidative decarboxylation
d. Transamination

41. 3-phosphoglycerate is the precursor molecule in the synthesis of

a. Aspartate b. Proline c. Serine d. Threonine

42. All of the following are characteristics of phenylalanine hydroxylase, EXCEPT

a. Mixed function oxidase
b. Tetrahydrobiopterin is a cofactor
c. NADPH provides the reducing power
d. Vitamin C is a cofactor

43. All of the following are required for lysyl hydroxylase reaction, EXCEPT

a. Alpha keto glutarate b. Copper
c. Iron d. Vitamin C

44. All of the following are cofactors for Nitric Oxide Synthase reaction, EXCEPT

a. NAD^+ b. FMN
c. Heme d. Tetrahydrobiopterin

45. Aspartame is made up of

a. Aspartic acid and methionine
b. Asparagine and methionine
c. Asparagine and phenylalanine
d. Aspartate and phenylalanine

Ans. 38. (d) 39. (c) 40. (b) 41. (c) 42. (d) 43. (b) 44. (a) 45. (d)

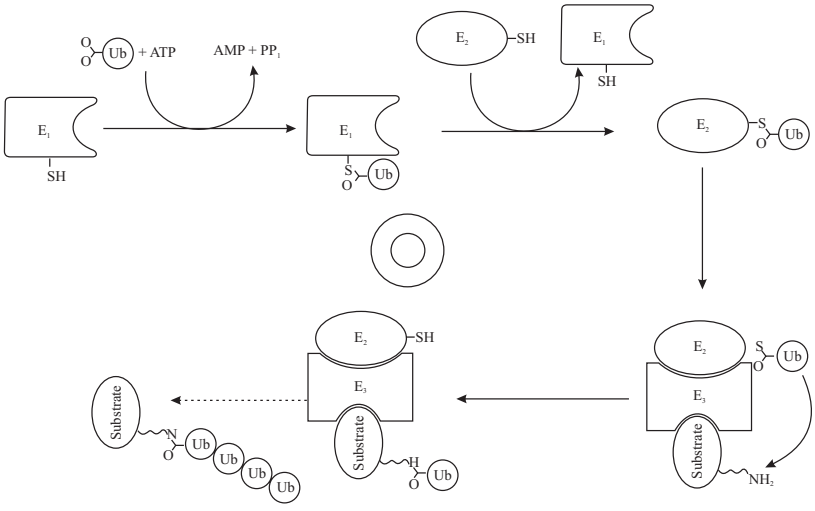
Explanations

1. (d) None ***(Ref. Harper 30th Ed. p. 544)***

Starch is the storage form of carbohydrate in plants. Glycogen is the storage form of carbohydrate in animals. Triacylglycerol is the storage form of lipids. But there is no storage form of proteins in humans. Excess dietary proteins are degraded.

"There is little or no evidence that athletes and body builders require large amounts of protein; simply consuming more of a normal diet providing about 14% of energy from protein will provide more than enough protein for increased muscle protein synthesis—the main requirement is for an increased energy intake to permit increased protein synthesis."

2. (d) Lysine ***(Ref. Harper 30th Ed. p. 288; Fig 28-2)***



PEST (Proline, Glutamate, Serine & Threonine) sequences target some proteins for rapid degradation. That doesn't mean that ubiquitin is covalently added to these sequences. You can clearly see the epsilon amino group of lysine in the substrate to which ubiquitin is added in the figure above.

Cover page of 30th edition of Harper denotes the Ubiquitin-Proteasomal system. Ubiquitin is a small protein having many roles.

- It marks the protein for degradation in proteasomes.
- Monoubiquitylation of histone proteins is associated with gene activation, repression, and heterochromatic gene silencing.

3. (c) Isopeptide bond ***(Ref. Harper 30th Ed. p. 336; Q.7)***

Peptide bond is formed between α amino group and α carboxylic group. Isopeptide bond forms between non-α groups. e.g. Glutathione - γ carboxyl group of glutamate is linked to α amino group of cysteine.

Carboxy terminal amino acid of ubiquitin is glycine. This binds to ϵ amino group of target protein through isopeptide bond. One ubiquitin molecule is attached to another ubiquitin molecule through isopeptide bond.

4. (c) 4 ***(Ref. Harper 30th Ed. p. 620)***

Addition of a single ubiquitin molecule does not target the protein for degradation. A minimum of **FOUR** ubiquitin molecules must be attached for targeting into proteasome.

5. (d) ATP independent ***(Ref. Harper 30th Ed. p. 288; Fig 28-2)***

Ubiquitin-proteasomal system	Lysosomal Protein degradation
ATP dependent	ATP-independent
Degrades intracellular, short-lived, regulatory proteins	Degrades extracellular, long-lived structural proteins as well as intracellular proteins
Proteolysis is achieved by 20S core subunit of proteasome	Proteolysis is done by acid hydrolases like cathepsins.

6. (a) Muscle ***(Ref. Harper 30th Ed. p. 289)***

"Maintenance of steady-state concentrations of circulating plasma amino acids between meals depends on the net balance between release from endogenous protein stores and utilization by various tissues. Muscle generates over half of the total body pool of free amino acids"

Muscle releases alanine. This serves two purposes, i.e. removal of ammonia and providing carbon skeleton for synthesis of glucose. "The rate of hepatic gluconeogenesis from alanine is far higher than from all other amino acids."

7. (b) Valine ***(Ref. Harper 30th Ed. p. 289; Figure 28–7)***

"Following a protein-rich meal, the splanchnic tissues release amino acids while the peripheral muscles extract amino acids, in both instances predominantly branched-chain amino acids. Branched-chain amino acids (Valine, Leucine, Isoleucine) thus serve a special role in nitrogen metabolism. In the fasting state, they provide the brain with an energy source, and postprandially they are extracted predominantly by muscle, having been spared by the liver."

8. (d) Glutamate ***(Ref. Harper 30th Ed. p. 291)***

Amino group from all the amino acids are accepted by α-ketoglutarate through transamination since glutamate is the only amino acid that undergoes significant amount of oxidative deamination in liver mitochondira. This oxidative deamination generates NH_3 which is used for urea cycle in the same location i.e. liver mitochondria.

9. (b) Glutamine

Excess ammonia is toxic to brain. Multiple reasons are there:

Excess NH_3 shifts the equilibrium of glutamate dehydrogenase reaction towards formation of glutamate. This depletes α-Ketoglutarate and impairs TCA cycle in neurons.

So, NH_3 must be quickly detoxified. Urea can be produced only in liver. Thus, ammonia has to be transported from brain in a non-toxic form other than urea. Transamination of amino acids produce glutamate. This glutamate accepts another NH_3 to produce glutamine. In this way, two NH_3 can be detoxified into one molecule.

Glutamine synthetase deficiency in neonates results in severe brain damage, multiorgan failure, and death.

10. (a) Lysine ***(Ref. Harper 30th Ed. p. 291)***

- PLP is the prosthetic group found in catalytic site of all aminotransferases.
- Carbonyl group of the PLP is bound as a Schiff base (imine) to the ε-amino group of a lysine residue of the enzyme.

11. (b) ADP ***(Ref. Harper 30th Ed. p. 291)***

High-Yield Points about Glutamate dehydrogenase (GDH):

- Predominantly found in hepatic mitochondria.
- Can use either NAD^+ or $NADP^+$as cofactor
- GDH catalysed reaction is reversible.
- Allosterically inhibited by ATP, GTP, and NADH, and is activated by ADP.

12. (b) 4 mol of ATPs are required for synthesis of 1 mole of urea ***(Ref. Harper 30th Ed. p. 292)***

Synthesis of 1 mol of urea requires 3 mol of ATP, 1 mol each of ammonium ion and of aspartate.

13. (b) Serine ***(Ref. Harper 30th Ed. p. 286)***

Selenocysteine is synthesised co-translationally.

Selenocysteine insertion sequence is a stem loop structure present in the untranslated region of mRNA. It directs the protein synthesising machinery to synthesise selenocysteine from $tRNA^{sec}$ carrying serine. Serine is converted to selenocysteine and inserted.

14. (d) Glyoxylate ***(Ref. Harper 30th Ed. p. 283)***

- Glyoxylate is formed from the metabolism of hydroxyproline and deamination of glycine.
- Transamination of glyoxylate produces glycine.
- Failure to catabolize glyoxylate formed results in 1° hyperoxaluria

15. (c) Ornithine ***(Ref. Harper 30th Ed. p. 293)***

This is the other way of asking, "In Urea cycle, carbamoyl phosphate combines with……"

16. (b) CPS-II

Argininosuccinase is the other name of the urea cycle enzyme argininosuccinate lyase.

CPS II is the rate limiting enzyme of pyrimidine synthesis.

	CPS I	CPS II
Subcellular location	Mitochondria	Cytoplasm
Pathway involved	Urea cycle	Pyrimidine biosynthesis
Source of NH_3	Free NH_3 liberated from glutamate dehydrogenase reaction	Glutamine
Regulated by N-Acetyl Glutamate	Yes	No

17. (a) Cleavage of arginine ***(Ref. Harper 30th Ed. p. 293; Fig.28-16)***

This question test your ability to differentiate lyase from hydrolase. Both are involved in cleavage of covalent bonds. Lyase doesn't need water.

- Argininosuccinase is a lyase
- Arginase is a hydrolase

18. (d) Leucine ***(Ref. Harper 30th Ed. p. 298; Figure 29–1)***

This Figure 29–1 in Harper is very important. Many questions are being asked from this concept of fate of carbon skeleton of amino acids.

Leucine is purely ketogenic.

19. (d) Cysteine ***(Ref. Harper 30th Ed. p. 314)***

Taurine is a sulfonic acid derived from cysteine.

Cholyl-CoA +Taurine → Taurocholate (primary bile salt)

20. (a) Methionine ***(Ref. Harper 30th Ed. p. 309; Figure 29–19)***

- Cysteine is a dietarily non-essential amino acid as it can be synthesised from methionine.
- Conversion of methionine to cysteine is affected in homocystinuria and cysteine becomes dietarily essential in these patients.

21. (b) PLP ***(Ref. Harper 30th Ed. p. 307; Fig 29-16)***

Loading test	Urinary metabolite	Vitamin deficiency
Histidine loading test	FIGLU	Folate
Tryptophan loading test	Xanthurenic acid	PLP
-	Methylmalonic acid	B_{12}

22. (c) Phenylalanine

Tetrahydrobiopterin (THB) is a nucleotide cofactor derived from Guanosine triphosphate by 3 enzymatic reactions. The first one, GTP cyclohydrolase I is the rate limiting.

THB is a cofactor for

- Aromatic amino acid (Tryptophan, Phenylalanine and Tyrosine) hydroxylases
- Nitric acid synthase
- Serotonin synthesis

Some types of phenylketonuria are due to defect in the synthesis or recycling of THB.

DOPA-responsive dystonia (DRD) or the Segawa variant (DYT5) is a dominantly inherited form of childhood-onset dystonia caused by a mutation in the gene GTP cyclohydrolase-I. (Ref. Harrison 19th Ed., p.2620)

23. (b) Melatonin ***(Ref. Harper 30th Ed. p. 316)***

Melatonin is derived from tryptophan.

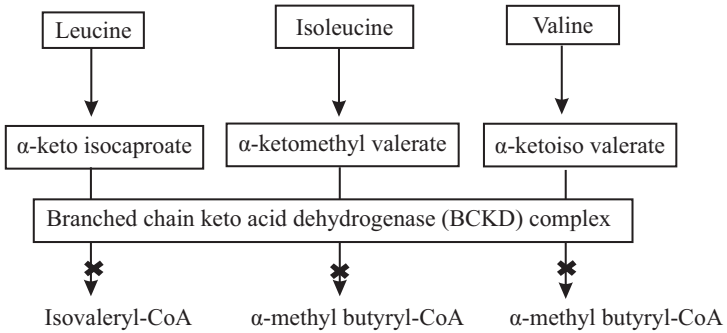
Special products derived from amino acids

Special Product	Constituents
Creatine	Glycine, Arginine, Methionine (Entire Glycine + Formamide group of Arginine + CH_3group of SAM)

Glutathione		γ - Glutamyl Cysteinyl Glycine (Peptide bond between glutamate and cysteine is an Isopeptide bond)
Sarcosine		N - methyl glycine
β-Alanyl Dipeptides	Carnosine	β – Alanyl histidine
	Anserine	N – methyl carnosine
Homocarnosine		GABA + Histidine
Tryptophan Derivatives	Serotonin	5-hydroxy tryptamine
	Melatonin	N-acetyl-5-methoxy tryptamine
	Niacin	60 mg tryptophan = 1 mg Niacin

24. (c) Branched chain α-keto acids ***(Ref. Harper 30th Ed. p. 309)***

Maple Syrup Urine Disease (MSUD) is an autosomal recessive disease due to the deficiency of mitochondrial multienzyme complex **B**ranched **C**hain **K**etoacid **D**ehydrogenase (BCKD). This enzyme requires thiamine as a cofactor.



Clinical presentation: Disease manifests in the first week of life with vomiting, hypoglycemia & **neurological impairment**. Characteristic maple syrup/**burnt sugar odor** *(not taste)* of urine & sweat is due to branched chain keto acids.

Diagnostic findings:

- ↑ Branched chain ketoacids and branched chain amino acids in blood & urine is confirmatory.
- Presence of alloisoleucine (stereoisomeric metabolite of isoleucine) is characteristic.
- Enzyme defect can be demonstrated in cultured fibroblasts.

Treatment:

- **Restriction of branched chain amino acids** low enough to prevent mental retardation but sufficient enough to allow normal growth
- **High-dose thiamine** in thiamine-responsive MSUD

25. (a) Methionine ***(Ref. Harper 30th Ed. p. 309; Fig. 29-19)***

Cysteine is produced from Methionine. Defect in this leads to homocystinuria and cysteine becomes a dietarily essential amino acid in this condition.

26. (a) Fumaryl acetoacetate hydrolase ***(Ref. Harper 30th Ed. p. 300; Table 29-2)***

Hereditary tyrosinemia type I (Hepatorenal tyrosinemia) is due to deficiency of the enzyme fumaryl acetoacetate hydrolase (fumarylacetoacetase).

Clinical features: Liver failure, cirrhosis, rickets, failure to thrive, peripheral neuropathy, "boiled cabbage" odor

27. (d) Tyrosine aminotransferase

Richner-Hanhart syndrome is the other name of Tyrosinemia type II (oculo-cutaneous tyrosinemia) which is due to deficiency of Tyrosine transaminase (Tyrosine aminotransferase).

Clinical features: Palmoplantar keratosis, painful corneal erosions with photophobia, intellectual disability

Extra Edge: Tyrosinemia type III is due to deficiency of 4-hydroxyphenylpyruvate dioxygenase.

28. (a) Tryptophan ***(Ref. Harper 30th Ed. p. 557)***

Hartnup disease is an autosomal recessive disease with defective absorption of tryptophan and other neutral amino acids in intestinal & renal tubular epithelial cells due to mutation in the transporter → excretion of tryptophan in urine → ↓ synthesis of niacin → Pellagra.

Treatment: High protein diet & Niacin

29. (d) All of the above

Biochemical basis of Guthrie test:

- Growth of *Bacillus subtilis* (ATCC 6051) in agar media is inhibited by beta-2-thienylalanine.
- Addition of phenylalanine, phenyl pyruvic acid and phenyl lactic acid overcome the inhibition.
- If the blood from normal infant is added, there will be no growth as there is no phenylalanine to relieve the inhibition.
- When the blood from phenylketonuric infant is added, bacteria will proliferate since unmetabolized phenylalanine and derivatives are abundant in blood.

30. (b) Phenyl pyruvate

To 5 ml of fresh urine sample, 3-4 drops of ferric chloride solution is added. Phenylpyruvate gives green/blue colour.

31. (c) Phenyl acetate

32. (c) Limiting the substrate for deficient enzyme

- Restriction of phenylalanine low enough to prevent mental retardation but sufficient enough to allow normal growth is the first-line therapy.

33. (d) Leucine ***(Ref. Harper 30th Ed. p. 298; Fig. 29-1)***

Leucine produces acetyl-CoA and acetoacetate which are ketogenic and are not the intermediates of TCA cycle.

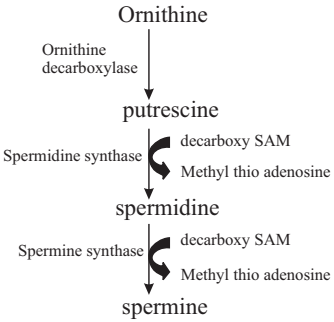
34. (c) Ornithine ***(Ref. Harper 30th Ed. p. 314)***

If there is arginine instead of ornithine, you can select arginine as it is the precursor of ornithine.

Polyamines are polycations synthesised from non-protein amino acid ornithine.

Putrescine (**di**amine), spermidine (**tri**amine) & spermine (**tetra**mine) are the major polyamines.

Rate limiting enzyme: **Ornithine decarboxylase** which is regulated by *ubiquitin independent proteasomal degradation*.



Role of polyamines:
Presence of regularly spaced multiple positive charges is the reason behind the functional capability of polyamines.

- Role in DNA stabilization, Signal transduction, Cell cycle regulation, Chromatin modulation, Cell migration.
- Used as a growth factor in cell culture technique

Polyamine synthesis inhibitor:

Difluoromethylornithine (DFMO), suicidal inhibitor of ornithine decarboxylase enzyme is used in,

- Sleeping sickness – injection form
- Facial hirsutism – topical application

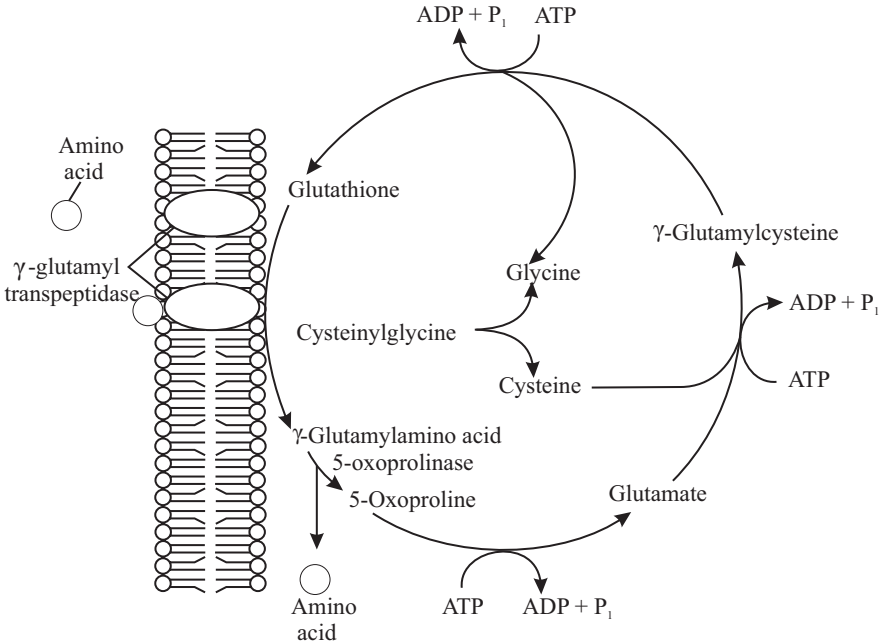
35. (b) Methionine ***(Ref. Harper 30th Ed. p. 298; Fig. 29-1)***

VIM (Valine, Isoleucine, Methionine) enter TCA cycle at the level of Succinyl-CoA.

36. (c) Both ***(Ref. Harrison 19th Ed. p. 434e-5)***

Nitisinone, also known as NTBC inhibits 4-Hydroxyphenylpyruvate dioxygenase. It prevents the formation of maleylacetoacetate and fumarylacetoacetate, which have the potential to be converted to toxic succinyl acetone. It also prevents the formation of homogentisic acid.

37. (d) Neutral amino acids



- Absorption of neutral amino acids is effected by the gamma glutamyl cycle a.k.a meister cycle in the enterocytes.
- Tripeptide glutathione (GSH) (gamma glutamyl cysteinyl glycine) is essential for Meister cycle. It reacts with the amino acid to form gamma glutamyl amino acid. This is catalysed by gamma glutamyl transferase (GGT)

- The glutamyl amino acid is then cleaved to give the free amino acid. The net result is the transfer of an amino acid across the membrane. The transport of one molecule of amino acid and regeneration of GSH requires 3 molecules of ATP.

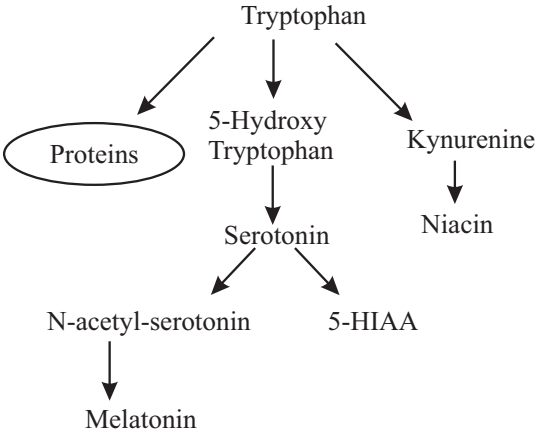
38. (d) Neutral amino acid transporter ***(Ref. Harper 30th Ed. p. 557)***

See the answer of Q.28

39. (c) Tryptophan ***(Ref. Harper 30th Ed. p. 316)***

If you know indole group is present in tryptophan, you will answer this easily.

- Serotonin is a derivative of tryptophan.
- Degradation of serotonin by monoamine oxygenase produce 5-hydroxyindoleacetic acid (5-HIAA), urinary excretion of which is increased in carcinoid syndrome.



40. (b) Decarboxylation ***(Ref. Harper 30th Ed. p. 315)***

Decarboxylation of amino acids produces biogenic amines. Pyridoxal phosphate serves as a cofactor.

Amino acid	Biogenic amine
Glutamate	Gamma-amino butyric acid (GABA)
Histidine	Histamine
Tryptophan	Tryptamine → 5HT (serotonin)
Tyrosine	Tyramine
DOPA	Dopamine → Norepinephrine & Epinephrine

41. (c) Serine ***(Ref. Harper 30th Ed. p. 284)***

42. (d) Vitamin C is a cofactor ***(Ref. Harper 30th Ed. p. 285; Fig. 27-12)***

- Not all hydroxylase enzymes require Vitamin C.
- Not all carboxylase enzymes require Vitamin K.

43. (b) Copper ***(Ref. Harper 30th Ed. p. 628)***

- Lysyl oxidase requires copper.
- Lysyl and prolyl hydroxylase require Ascorbate, Fe^{2+}, and α-ketoglutarate

44. (a) NAD+ ***(Ref. Harper 30th Ed. p. 314; Fig. 30-2)***

Nitic Oxide Synthase is unique in the way that it requires the following five cofactors:

1. NADPH
2. FAD
3. FMN
4. Heme
5. Tetrahydrobiopterin (BH_4)

45. (d) Aspartate and phenylalanine

Aspartame is a synthetic dipeptide artificial sweetener made up of methyl ester of the aspartic acid & phenylalanine.

- Used in cold drinks
- Contraindicated in phenylketonurics and children.