NITROGEN METABOLISM

Past paper questions

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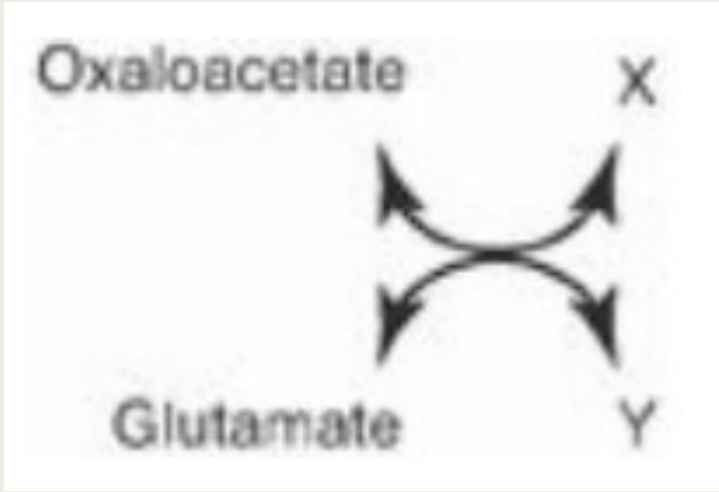
1- In the transamination reaction shown to the right, which of the following are the products X and Y?



- A. Alanine, α-ketoglutarate
- B. Asparate, α-ketoglutarate
- C. Glutamate, alanine
- D. Pyruvate, aspartate

- **Answer B**

2-Which one of the following statements about amino acids and their metabolism is correct?

- A. Free amino acids are taken into the enterocytes by a proton-linked .transport system
- B. In healthy, fed individuals, the input to the amino acid pool exceeds .the output .
- C. Liver uses ammonia to buffer protons
- D. Muscle-derived glutamine is metabolized in liver and kidney tissue to ammonia plus a gluconeogenic precursor
- E. The first step in the catabolism of most amino acids is their oxidative deamination
- F. The toxic ammonia generated from the amide nitrogen of amino acids is transported through blood as arginine

- Answer D

3- A 1-week-old infant, who was born at home in a rural area, has undetected classic phenylketonuria. Which statement about this baby and/or her treatment is correct?

- A. A diet devoid of phenylalanine should be initiated immediately.
- B. Dietary treatment will be recommended to be discontinued in adulthood.
- C. Supplementation with vitamin B6 is required.
- D. Tyrosine is an essential amino acid.

- Answer ? A

4- Which one of the following statements concerning amino acids is correct?

- A. Alanine is ketogenic.
- B. Amino acids that are catabolized to acetyl coenzyme A are glucogenic.
- C. Branched-chain amino acids are catabolized primarily in liver.
- D. Cysteine is essential for individuals consuming a diet severely limited in methionine

- Answer ? D

5- δ-Aminolaevulinic acid synthase activity:

- A. catalyses the committed step in porphyrin biosynthesis.
- B. is decreased by iron in erythrocytes.
- C. is decreased in liver in individuals treated with certain drugs such as the barbiturate phenobarbital.
- D. occurs in the cytosol.
- E. requires biotin as a coenzyme.

- Answer A

6- A patient presents with jaundice, abdominal pain, and nausea. Clinical laboratory studies give the following results:

Serum bilirubin	Urine urobilinogen	Urinary bilirubin
Increase in conjugated bilirubin	Not present	Present

What is the most likely cause of the jaundice?

- A. Decreased hepatic conjugation of bilirubin
- B. Decreased hepatic uptake of bilirubin
- C. Decreased secretion of bile into the intestine
- D. Increased hemolysis

Answer C

7- A 2-year-old child was brought to his paediatrician for evaluation of gastrointestinal problems. The parents report that the boy has been listless for the last few weeks. Lab tests reveal a microcytic, hypochromic anemia. Blood lead levels are elevated. Which of the enzymes listed below is most likely to have higher-than-normal activity in the liver of this child?

- A. δ-Aminolevulinic acid synthase
- B. Bilirubiun UDP-glucuronosyltransferase
- C. Ferrochelatase
- D. Heme oxygenase
- E. Porphobilinogen synthase

- Answer A

8- A 42-year-old male patient undergoing radiation therapy for prostate cancer develops severe pain in the metatarsal phalangeal joint of his right big toe. Monosodium urate crystals are detected by polarized light microscopy in fluid obtained from this joint by arthrocentesis. This patient's pain is directly caused by the overproduction of the end product of which of the following metabolic pathways?

- A. De novo pyrimidine biosynthesis
- B. Pyrimidine degradation
- C. De novo purine biosynthesis
- D. Purine salvage
- E. Purine degradation.

- Answer E

9- A 50-year-old man presented with painful blisters on the backs of his hands. He was a golf instructor and indicated that the blisters had erupted shortly after the golfing season began. He did not have recent exposure to common skin irritants. He had partial complex seizure disorder that had begun about 3 years earlier after a head injury. The patient had been taking phenytoin (his only medication) since the onset of the seizure disorder. He admitted to an average weekly ethanol intake of about 18 12-oz cans of beer. The patient's urine was reddish orange. Cultures obtained from skin lesions failed to grow organisms. A 24-hour urine collection showed elevated uroporphyrin (1,000 mg; normal, <27mg) The most likely diagnosis is:

- A. acute intermittent porphyria.
- B. congenital erythropoietic porphyria.
- C. erythropoietic protoporphyria.
- D. hereditary coproporphyria.
- E. porphyria cutanea tarda.

- Answer E

10- Which one of the following enzymes of nucleotide metabolism is correctly paired with its pharmacologic inhibitor?

- A. Dihydrofolate reductase—methotrexate
- B. Inosine monophosphate dehydrogenase—hydroxyurea
- C. Ribonucleotide reductase—5-fluorouracil
- D. Thymidylate synthase—allopurinol
- E. Xanthine oxidase—probenecid

- Answer ? A

11- Which substrate is common for purine, heme and creatine synthesis?

- a. Glycine
- b. Succinate
- c. Arginine
- d. Aspartate

- Answer A

12- -Which one of the following statements concerning amino acids is correct?

- a. An increase in gluconeogenesis from amino acids results in a decrease in urea formation.
- b. All essential amino acids are glycogenic.
- c. Ornithine and citrulline are found in tissue proteins.
- d. Cysteine is an essential amino acid in individuals consuming a diet severely limited in methionine.

- Answer D

13- Carbamoyl phosphate synthetase synthesized in hepatic cytosol is used for:

- a. Pyrimidine synthesis
- b. Urea cycle activator
- c. Purine synthesis
- d. Activator of IMP formation

- Answer A

14- The main acceptor of NH3 in deamination reactions:

- a. Glutamate
- b. a-ketoglutarate
- c. Glutamine
- d. Alanine

- Answer ? B

15- A patient with an inherited disorder has blue speckled discoloration of skin, was found to have kidney stones and has black urine, this disorder involves the accumulation of?

- a. Homogentisic acid (alkaptonuria)
- b. Phenylalanine
- c. Homocysteine
- d. Cystathionine

- Answer ? A

16- Which of the following is not synthesized by tyrosine?

- a. Melatonin
- b. Dopamine
- c. Melanin
- d. Epinephrine

- Answer [?] A

17- A new born who refuses feeding has been diagnosed with cystathionine-β- synthase deficiency. What is the diagnosis of his condition?

- A. Albinism
- B. Homocystinuria
- C. Maple syrup urine disease
- D. Hyperammonaemia
- E. Alkaptonuria

- Answer ? B

18- which of the following pairings is incorrect?

- A. Tyrosine - melanin
- B. Tyrosine - norepinephrine
- C. Threonine -serotonin
- D. histidine –histamine
- E. arginine and glycine - creatine

- Answer C

19- A patient who has a glutamine synthetase deficiency would have all of the following EXCEPT:

- A. Glutamate amination to glutamine is compromised
- B. Transport of ammonia from most tissues to liver is hindered
- C. Toxic levels of ammonia may accumulate in the patient’s tissues and/or blood
- D. Transport of ammonia from muscle cells to the liver is not affected
- E. Transamination of α–ketoglutarate to glutamate is downregulated

- Answer E

20- histamine is synthesized from histidine by:

- a- decarboxylation
- b- amination
- c- deamination
- d- carboxylation

- Answer ? A

21- Which of the following amino acids match with the corresponding catabolic product :

- A- (glutamate, glutamine, alanine ,arginine) > pyruvate
- B-(histidine, glutamate, arginine, proline)>alpha ketoglutarate
- C- (isoleucine, valine , tryptophan) > succinyl CoA
- D- (aspartate, phenylalanine, tyrosine) > OAA

- Answer ? B

22- Regarding heme synthesis in the liver, all statements are correct except:

- A- Uroporphyrinogen III is synthesized from porphobilinogen
- B- ALA synthase requires PRP and is located in the mitochondria
- C- ALA synthase can be induced by many drugs
- D- Synthetic pathway involves carboxylation reactions at more than one step
- E- ALA synthase is suppressed by hemin

- Answer ? D

23- Which of the following statements describes the ubiquitin mediated degradation of proteins in the cytosol?

- A- after degradation, ubiquitins are transferred extracellularly for excretion.
- B- The process is ATP independent
- C- The degradation results in free amino acids released in the cytosol
- D- this pathway is more specific to extracellular proteins
- E- One molecule of ubiquitin is attached to the protein to be degraded

- Answer B

24- which is not true regarding carbamoyl phosphate synthetase?

- A- its found in the mitochondria
- B- Its found in the cytosol
- C- Its required for pyrimidine synthesis
- D- its required for purine synthesis
- E- its required for urea synthesis

- Answer ? D

25- The following amino acids are key to the transfers of amino groups during the breakdown and synthesis of amino acids:

- A- ALA & GLN
- B- GLU & ASP
- C- GLU & ARG
- D- GLU & GLN
- E- ASP & GLN

- Answer A

26- A coenzyme derived from Vitamin B12 is needed for?

- a. Synthesis of D-Methylmalonyl CoA
- b. Formation of Guanidinoacetate
- c. Decarboxylation of Uroporphyrinogen III
- d. Propionyl CoA metabolism

- Answer D

27- If an individual has vitamin B6 deficiency, which of the following amino acids could still be synthesized and be considered non essential?

- A- Cysteine
- B- Serine
- C- Aspartate
- D- Alanine
- E- Tyrosine

- Answer --> E

28- When you are looking at the following table, which case mostly represents liver disease?

- A- III
- B- IV
- C- V
- D- I
- E- II

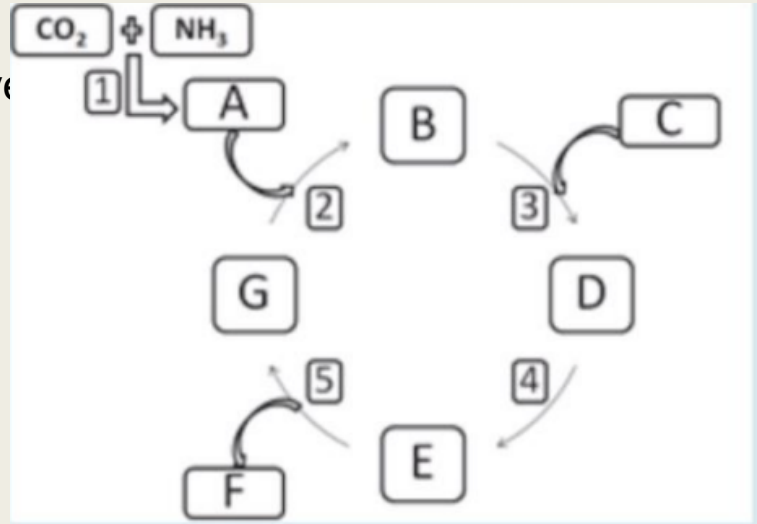
- Answer ? A

	I	II	III	IV	V
High blood levels of ALT and AST	No	No	Yes	Yes	Yes
High blood levels of bilirubin	No	Yes	Yes	Yes	No
High levels of plasma proteins	Yes	Yes	No	No	No
Edema	No	Yes	Yes	Yes	No
High levels of blood ammonia	No	Yes	Yes	No	Yes

29- considering the following diagram that represents the urea cycle, which of the following statements is incorrect?

- A- A & C represent the source of nitrogen atoms in F
- B- B & G are transported out of and into the mitochondria, respectively
- C- Step #1 is the rate limiting step
- D- Step number 5 is catalyzed almost exclusively in the live
- E- step 2 through 5 occur in the cytosol

- Answer ? E



30- All of the following regarding protein digestion are true EXCEPT:

- A- Pepsin has 2 modes of activation: acid induced and autocatalytic activation
- B- Enterokinase activates trypsinogen
- C- Stomach acidity hydrolyzes dietary proteins into long peptide chains
- D- Free amino acids and dipeptides are taken up by intestinal cells, however, by different modes of entry
- E- Trypsin is the common activator for all pancreatic zymogens

- Answer C

31- How many ATP equivalents are required to produce one molecule of urea from ammonia and aspartate?

- A- 2
- B- 4
- C- 1
- D- 3
- E- 6

- Answer ? B

32- OTC deficiency and UMP synthase deficiency can result in one of the following condition:

- A- Orotic aciduria
- B- Megaloblastic anemia
- C- Hyperphenylalaninemia
- D- Hyperammoniemia
- E- Albinism

- Answer ? A

33- one of the following is true considering nitrogen containing compounds:

- A- source of carbon atoms in creatine structure are: glycine, arginine and SAH
- B- Tyrosine hydroxylase catalyzes the rate limiting step in catecholamine synthesis
- C- In hepatocellular jaundice; stool may be pale and urinary urobilinogen is absent
- D- conjugated bilirubin is oxidized to urobilinogen then gets reduced to urobilin in the kidneys
- E- conjugated bilirubin is oxidized to urobilinogen then gets reduced to stercobilin by intestinal flora

- Answer B

34- Proteolytic enzymes must be secreted as zymogens that are activated, otherwise they would auto-digest themselves and the organs that produce them. Trypsinogen, a zymogen, is cleaved to form trypsin by a protease that is secreted by which of the following?

- A- colon
- B- liver
- C- pancreas
- D- stomach
- E- small intestine

- Answer E

35- when comparing infant I (low blood arginine, high blood ammonia) to infant II (high blood arginine, moderately high blood ammonia), infant II might have a problem in:

- A- CPS I
- B- Arginase
- C- Arginosuccinate lyase
- D- Arginosuccinate synthase
- E- OTC

- Answer B

36- The rate limiting step in prostaglandins synthesis is catalyzed by

- a) peroxidase
- b) oxygenase
- c) phospholipase A2
- d) cyclooxygenase
- e) PGG synthase

- Answer C

37- The final product of purine degradation is --------- while the final product of pyrimidine degradation is----------.

- a) Uric acid, beta alanine
- b) Urea, uric acid
- c) Uric acid, hypoxanthine
- d) Ammonia, xanthine
- e) PRPP, carbamoyl phosphate

- Answer A

38- Which of the following is NOT a precursor for de novo purine biosynthesis?

- a) N-formyl-tetrahydrofolate
- b) Aspartic Acid
- c) Glycine
- d) Arginine
- e) Glutamine

- Answer D

39- Which of the following compounds is CORRECTLY matched with the precursor amino acid:

- a) 5 Histamine, tyrosine
- b) Creatine, lysine
- c) Epinephrine, aspartate
- d) Serotonin, tryptophan
- e) Melanin, tryptophan

- Answer ? D

40- Bilirubin is transported to hepatocytes via:

- a) Facilitated diffusion
- b) Conjugation to nucleotides ion
- c) Na+/K+ pump
- d) Active transport
- e) Proton pump

- Answer A

41- Gilbert syndrome is caused by a deficiency of the following enzyme:

- a. Tyrosine hydroxylase
- b. Heme oxygenase
- c. bilirubin glucuronyl-transferase
- d. Biliverdin reductase
- e. ALA synthase

- Answer ? C

42- Which one of the following statements concerning a 1-week-old male infant with undetected classic phenylketonuria is CORRECT?

- a. Hypopigmentation is characteristic for these patients.
- b. Tyrosine is a nonessential amino acid for the infant.
- c. Therapy must begin after the first year of life.
- d. Low levels of phenylpyruvate appear in his urine.
- e. When the infant reaches adulthood, it is recommended that diet therapy be
- discontinued.

- Answer A

43- The carbamoyl phosphate that is being synthesized in the cytosol of a hepatocyte would be used to:

- a. De novo synthesize purine nucleotides.
- b. Increase amino acid degradation after a protein rich meal.
- c. Activate urea cycle.
- d. De novo synthesize pyrimidine nucleotides.
- e. Increase purine degradation and production of uric acid.

- Answer D

44- Which of the following describes heme synthesis?

- a. Fe+2 addition is the rate limiting step that requires ATP hydrolysis as an energy source.
- b. Two molecules of porphobilinogen are combined in a tetrapyrrole ring.
- c. Fe+2 is added by the cytosolic ferro chelatase enzyme.
- d. Four ALA molecules are combined to produce porphobilinogen.
- e. Gly and succinyl-CoA are precursors for heme synthesis.

- Answer E

45- Newborns jaundice can be described as follows:

- a. Liver cells function is abnormal.
- b. Glucuronyl transferase levels are lower than in a normal newborn.
- c. Conjugated bilirubin levels are higher than albumin capacity to bind them resulting in
- toxicity.
- d. Newborn exposure to blue fluorescent light produces water-insoluble photoisomers.
- e. Conjugated bilirubin levels are high in these infants.

- Answer B

46- Melatonin is made of the following amino acid:

- a. Tyr.
- b. Phe.
- c. Arg.
- d. Thr.
- e. Trp.

- Answer E

47- Dihydrofolate reductase is targeted by the following pharmacological inhibitor:

- a. Allopurinol
- b. Probenecid
- c. Methotrexate
- d. Hydroxyurea
- e. 5-fluorouracil

- Answer ? C

48- Which of the following is an essential amino acid?

- a. Cysteine
- b. Methionine
- c. Arginine
- d. Tyrosine
- e. Glutamate

- Answer B

49- A female neonate did well until approximately age 24 hours, when she became lethargic. A sepsis workup proved negative. At 56 hours, she started showing focal seizure activity. The plasma ammonia level was found to be 887 μmol/l (normal 5–35 μmol/l). Quantitative plasma amino acid levels revealed a marked elevation of citrulline but not arginosuccinate.

Which one of the following enzymic activities is most likely to be deficient in this patient?

- A. Arginase
- B. Arginosuccinate lyase
- C. Arginosuccinate synthetase
- D. Carbamoyl phosphate synthetase I
- E. Ornithine transcarbamoylase

- Answer

50- Match the deficiency of **Cystathionine β-synthase** with the corresponding clinical sign of laboratory finding in urine:

- A. Black pigmentation of cartilage
- B. Cabbage-like odor of fluids
- C. Cystine crystals in urine
- D. White hair, red eye color
- E. Increased branched-chain amino acids
- F. Increased homocysteine G. Increased methionine
- H. Increased phenylalanine

- Answer ? F

51- Match the deficiency of **Homogentisic acid oxidase** with the corresponding clinical sign of laboratory finding in urine:

- A. Black pigmentation of cartilage
- B. Cabbage-like odor of fluids
- C. Cystine crystals in urine
- D. White hair, red eye color
- E. Increased branched-chain amino acids
- F. Increased homocysteine G. Increased methionine
- H. Increased phenylalanine

- Answer ? A

52- Match the deficiency of **Tyrosinase** with the corresponding clinical sign of laboratory finding in urine:

- A. Black pigmentation of cartilage
- B. Cabbage-like odor of fluids
- C. Cystine crystals in urine
- D. White hair, red eye color
- E. Increased branched-chain amino acids
- F. Increased homocysteine G. Increased methionine
- H. Increased phenylalanine

- Answer ? D

53- A 1-year-old female patient is lethargic, weak, and anemic. Her height and weight are low for her age. Her urine contains an elevated level of orotic acid. Activity of uridine monophosphate synthase is low. Administration of which of the following is most likely to alleviate her symptoms?

- A. Adenine
- B. Guanine
- C. Hypoxanthine
- D. Thymidine
- E. Uridine

- Answer E

54- A defective glucorodyl transferase is associated with all of the following except?

- a. gilbert syndrome
- b. criggler najjjar
- c. liver cirrhosis
- d. dubin Johnson syndrome

- Answer D

Good luck *-*