

MEDICAL BIOCHEMISTRY 531 - ACKERMAN
TEACHING-LEARNING EXAMINATION (FALL 2004)

November 30, 2004

100 questions, 2 hour time period (1.2 minutes/question)

(NOTE: Final Exam has 80 questions @ 1.2 minute/question = 1 hr 36 min. MPL \approx 53%)

(a list of answers is provided on the last two pages of this TLE - NO PEEKING!!!)

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Notes: *The TLE questions are clearly not in the order of coverage in the lectures. However, the final exam questions will be in the general order of coverage in the lectures. So, questions from the first 24 lectures will (more or less) come first in the final exam. Also, the final exam includes more questions on interorgan/intertissue relationships during different physiologic states or diseases than appear on this TLE.*

1. The **most serious** form of galactosemia in humans is generally due to a deficiency of:
 - (A) galactokinase, leading to a build-up of galactose-1-phosphate and galactose
 - (B) galactose-1-phosphate uridylyltransferase, leading to a build-up of free galactose and galactose-1-phosphate
 - (C) galactosidase, leading to a build-up of free galactose and galactitol in the blood
 - (D) lactase, leading to a build-up of galactose in the blood from an inability to digest lactose in milk.
 - (E) UDP-galactose epimerase, leading to a build-up of UDP-galactose.

2. All of the following metabolic disorders are accompanied by metabolic acidosis EXCEPT:
 - (A) maple syrup urine disease
 - (B) methylmalonic acidemia
 - (C) ornithine transcarbamoylase deficiency
 - (D) pyruvate dehydrogenase deficiency
 - (E) type I glycogen storage disease

3. Removal of the chief metabolite of cholesterol from the body is used in one of the treatments of hypercholesterolemia. This therapy is directed towards the elimination of:
 - (A) neutral sterols
 - (B) cortisol
 - (C) progesterone
 - (D) bile salts
 - (E) carbon dioxide

4. Infants with lethargy, hypotonia, metabolic acidosis without ketosis, and very high levels of urinary β -hydroxy- β -methylglutarate in the urine suffer from:
 - (A) HMG-CoA lyase deficiency
 - (B) β -ketothiolase deficiency
 - (C) maple syrup urine disease
 - (D) medium-chain acyl-CoA dehydrogenase deficiency
 - (E) propionyl-CoA carboxylase deficiency

5. A 12 year old male patient is admitted to the hospital with a red scaly rash and mild cerebellar ataxia. His mother thinks he is suffering from pellagra, because similar symptoms in her daughter had been diagnosed as a dietary deficiency of niacin. The boy does not have the usual dietary deficiency form of pellagra, but large amounts of aromatic and neutral free amino acids are found in his urine. When an older sister also has an attack of ataxia, her urine is also found to contain excessive amounts of these amino acids as well. Two other siblings are also found to have aminoaciduria, but four others are normal. This patient most likely has _____ disease in which inadequate absorption of _____ leads to a niacin-deficiency and negative nitrogen balance?
- (A) Hartnup's; tryptophan
 - (B) Tyrosinemia; tryptophan
 - (C) Alcaptonuria; linoleic acid
 - (D) Classical PKU; phenylalanine
 - (E) Both A and D are correct answers
6. Failure to digest dietary lipids leads to steatorrhea. All of the following substances are required for digestion of dietary lipid EXCEPT:
- (A) bile pigments
 - (B) bile salts
 - (C) colipase
 - (D) pancreatic lipase
 - (E) pancreatic phospholipase
7. The elevated blood glucose seen in diabetes mellitus is a consequence of:
- (A) a failure of glucose to enter liver cells
 - (B) increased glycolytic activity in liver
 - (C) increased activity of gluconeogenic enzymes due to enzyme induction by glucagon
 - (D) increased liver glycogenesis as a result of augmented formation of glycogen synthase in response to glucagon
 - (E) conversion of fatty acids to glucose
8. A deficiency of which one of the following enzymes results in hypermethioninemia and homocystinuria?
- (A) Methionine aminopeptidase
 - (B) S-Adenosylhomocysteine hydrolase
 - (C) S-Adenosylmethionine hydrolase
 - (D) Cystathionine lyase
 - (E) Cystathionine synthase

9. Allopurinol, an inhibitor of xanthine oxidase, is used to treat gout. Which one of the following enzymes is rate limiting for de novo purine nucleotide synthesis, and thus could also be a target for development of drugs for the treatment of gout?
- (A) Carbamoyl-phosphate synthetase II
 - (B) Hypoxanthine-guanine phosphoribosyl transferase (HGPRT)
 - (C) Phosphoribosylpyrophosphate-glutamine amidotransferase (PRPP amidotransferase)
 - (D) Xanthine dehydrogenase
 - (E) Carbamoyl-phosphate synthetase I
10. A deficiency of which one of the following enzymes leads to diarrhea and failure to thrive in newborns owing to inadequate cleavage of trypsinogen to trypsin in the intestinal lumen?
- (A) Carboxypeptidase A
 - (B) Enterokinase (enteropeptidase)
 - (C) Chymotrypsinogenase
 - (D) Leucine aminopeptidase
 - (E) Pepsin
11. Cyanide (CN^-), which is one of the most rapidly acting poisons in humans, blocks oxidative phosphorylation by:
- (A) dissipating the proton gradient
 - (B) binding to the ferrous iron in myoglobin
 - (C) inhibiting ATP synthase
 - (D) lowering the oxygen-carrying capacity of the blood
 - (E) binding to the ferric iron of cytochrome a_1 in cytochrome oxidase
12. Enzyme deficiencies result in the inability to convert substrates into products. The accumulation of precursors and their aberrant metabolism can lead to various maladies. Enzymes perform their catalytic function by:
- (A) decreasing the energy of activation (E_a) of a reaction
 - (B) decreasing the standard free energy change (ΔG°) of a reaction
 - (C) decreasing the entropy change (ΔS) of a reaction
 - (D) increasing the equilibrium constant (K_m) of a reaction
 - (E) increasing the total kinetic energy ($1/2 mv^2$) of the reactants
13. All of the following are true statements regarding a deficiency of bile salts EXCEPT:
- (A) the deficiency impairs lipid digestion and absorption
 - (B) the deficiency impairs the absorption of vitamins A, D, E, and K
 - (C) the deficiency leads to steatorrhea (increased lipid in the stool)

- (D) the deficiency promotes cholelithiasis (gallstones)
 - (E) the deficiency leads to hypercholesterolemia
14. All of the following are correct statements regarding gluconeogenesis and glycolysis EXCEPT:
- (A) both gluconeogenesis and glycolysis require ATP as a reactant in some steps
 - (B) both gluconeogenesis and glycolysis are affected by changes in fructose 2,6-bisphosphate concentration
 - (C) both gluconeogenesis and glycolysis proceed with the liberation of free energy
 - (D) both gluconeogenesis and glycolysis result in the net loss of ATP
 - (E) both gluconeogenesis and glycolysis involve the interconversion of two triose phosphates and fructose 1,6-bisphosphate

15. Rotenone is used in several commercial rat poisons, and this substance inhibits the functioning of Complex I of electron transport. This substance inhibits mitochondrial electron transport and prevents the translocation of hydrogen ions:
- (A) from the intermembrane space to the mitochondrial matrix
 - (B) from the cytosol to the intermembrane space
 - (C) from the mitochondrial matrix to the intermembrane space
 - (D) from the intermembrane space to the cytosol
 - (E) from the intermembrane space to the outer mitochondrial membrane
16. With the knowledge that citrate is an intermediate in de novo fatty acid synthesis from glucose, which of the following conclusions can be deduced?
- (A) The primary regulatory step of the Krebs cycle must be downstream from the citrate synthase reaction
 - (B) The citrate synthase reaction is reversible in order to release acetyl-CoA into the cytosol
 - (C) The first committed step of the Krebs cycle is catalyzed by aconitase
 - (D) The citrate synthase reaction is markedly inhibited when the energy charge is high
 - (E) The reversible α -ketoglutarate dehydrogenase reaction permits four-carbon metabolites to be converted to acetyl-CoA
17. The final product of glycolysis in human cells under anaerobic conditions, or in cells lacking mitochondria, is:
- (A) carbon dioxide
 - (B) ethanol
 - (C) lactate
 - (D) phosphoenolpyruvate
 - (E) pyruvate
18. The elevation of blood ketone bodies seen in hyperketotic hypoglycemia results from a deficiency in which of the following enzymes?
- (A) β -Ketothiolase
 - (B) Hydroxymethylglutaryl-CoA synthase
 - (C) Hydroxymethylglutaryl-CoA lyase
 - (D) Hydroxymethylglutaryl-CoA reductase
 - (E) Succinyl-CoA:acetoacetate CoA-transferase
19. What is the first purine nucleotide formed during de novo purine synthesis?
- (A) Carbamoyl phosphate

- (B) Orotidine monophosphate
- (C) Inosine monophosphate
- (D) 5-Phosphoribosyl-1-pyrophosphate (PRPP)
- (E) 5-Phosphoribosylamine

20. Which one of the following is a true statement regarding non-competitive inhibitors?
- (A) They are structural analogues of the substrate
 - (B) They include captopril, an angiotensin converting enzyme (ACE) inhibitor, that resembles the physiological substrate
 - (C) They lead to the same y-intercept on Lineweaver-Burk plots as does the noninhibited enzyme
 - (D) Their action can be overcome by increasing the concentration of the varied substrate
 - (E) They decrease the apparent V_{\max} of an enzyme
21. An inborn error of metabolism that is associated with lactic acidosis, secondary carnitine deficiency, and excretion of medium chain dicarboxylic acids is due to a deficiency of which one of the following enzymes?
- (A) Medium-chain fatty acyl-CoA dehydrogenase
 - (B) Succinyl-CoA: acetoacetate CoA-transferase
 - (C) Carnitine acyltransferase I
 - (D) β -Ketothiolase
 - (E) β -Hydroxyacyl-CoA dehydrogenase
22. Which one of the following shuttles can move reducing equivalents both INTO and OUT OF the mitochondrion?
- (A) The glycerol phosphate shuttle
 - (B) The malate-aspartate shuttle
 - (C) The ATP/ADP shuttle
 - (D) The dinitrophenol shuttle
 - (E) The carnitine/acylcarnitine shuttle
23. Protoporphyrin IX is derived from eight molecules of succinyl-CoA and eight molecules of:
- (A) alanine
 - (B) valine
 - (C) glutamate
 - (D) histidine
 - (E) glycine
24. Myoglobin, a monomeric protein, exhibits all of the following types of structure EXCEPT
- (A) primary
 - (B) secondary

- (C) tertiary
 - (D) quaternary
 - (E) alpha helix
25. Muscle lactate dehydrogenase deficiency is accompanied by fatigue and myoglobinuria. This lactate dehydrogenase (LDH) deficiency is characterized by all of the following properties **EXCEPT**:
- (A) there is a decrease in activity of M subunits of LDH found in muscle
 - (B) there is little or no change in blood pyruvate levels following exercise
 - (C) there is little or no change in blood lactate levels following exercise
 - (D) there is no change in cardiac pyruvate/lactate production because H subunit expression of lactate dehydrogenase is normal
26. Dinitrophenol (DNP) uncouples electron transport and oxidative phosphorylation in which one of the following ways?
- (A) By dissipating the proton gradient between the mitochondrial matrix and the intermembrane space
 - (B) By inhibiting the translocation of protons across the mitochondrial membrane
 - (C) By blocking the reduction of oxygen to water
 - (D) By inhibiting the oxidation of ubiquinol (coenzyme QH₂)
 - (E) By blocking the transfer of electrons from cytosol to the mitochondrial matrix
27. Aspirin is an irreversible inhibitor of:
- (A) cyclooxygenase via acetylation of the enzyme
 - (B) lipoyxygenase via acetylation of the enzyme
 - (C) phospholipase A₁ via blockage of arachidonate cleavage from the SN1 position of phospholipids
 - (D) phospholipase A₂ via blockage of arachidonate cleavage from the SN2 position of phospholipids
 - (E) phospholipase C via acetylation of the enzyme
28. Which one of the following mechanisms explains how ADP regulates the rate of electron transport during oxidative phosphorylation?

- (A) Proton translocation across the inner mitochondrial membrane is decreased when ATP synthase lacks bound ADP and Pi, secondarily retarding electron transport
 - (B) The ATP/ADP antiport system is not functional with low mitochondrial [ADP]
 - (C) The transmembrane proton gradient is dissipated with low [ADP]
 - (D) Low [ADP] accelerates Krebs cycle reaction rates, thereby providing more NADH to activate electron transport
 - (E) AMP concentrations are increased as ADP concentrations fall
29. Both the plasma and urine of patients with Maple Syrup urine disease contain:
- (A) elevated levels of valine
 - (B) elevated levels of phenylketones
 - (C) low levels of leucine
 - (D) low levels of tyrosine
 - (E) normal levels of isoleucine
30. The conversion of monocarboxylic acids to dicarboxylic acids (e.g., dodecanedioic acid), which is prevalent in medium-chain fatty acyl-CoA dehydrogenase deficiency (MCAD), is called:
- (A) α -oxidation
 - (B) β -oxidation
 - (C) δ -oxidation
 - (D) ω -oxidation
 - (E) the final common pathway of metabolism
31. The major regulatory step of glycolysis in liver can be bypassed in the catabolism of which one of the following sugars?
- (A) Fructose
 - (B) Galactose
 - (C) Glucose
 - (D) Mannose
 - (E) Trehalose
32. Which one of the following compounds is **NOT** an allosteric regulator of phosphofructokinase activity?
- (A) AMP
 - (B) ATP
 - (C) Citrate
 - (D) Fructose 6-phosphate
 - (E) Fructose 2, 6-bisphosphate

33. A deficiency of any one of the following amino acids leads to a negative nitrogen balance in humans **EXCEPT**:
- (A) methionine
 - (B) isoleucine
 - (C) leucine
 - (D) ornithine
 - (E) phenylalanine
34. Reversing the effects of the glucagon-induced metabolic cascade in the liver involves all of the following processes **EXCEPT**:
- (A) cessation of adenylyl cyclase stimulation
 - (B) conversion of cyclic AMP to 5'-AMP
 - (C) re-formation of the inactive RC form of protein kinase A
 - (D) dephosphorylation of phosphatase-1 inhibitor, phosphorylase a, phosphorylase kinase, glycogen synthase, fructose-6-phosphate-2-kinase/phosphatase, and pyruvate kinase
 - (E) increased dephosphorylation of fructose 1,6-bis-phosphate to form fructose 6-phosphate

35. The rate-limiting reaction in prostaglandin biosynthesis is catalyzed by:
- (A) phospholipase A₁
 - (B) phospholipase A₂
 - (C) phospholipase C
 - (D) phospholipase D
 - (E) 5-lipoxygenase
36. A deficiency of which one of the following enzymes is associated with markedly reduced levels of cholesterol esters in **plasma** lipoproteins?
- (A) Acid lipase
 - (B) Acyl-CoA: cholesterol acyltransferase (ACAT)
 - (C) Hepatic lipase
 - (D) Lecithin: cholesterol acyltransferase (LCAT)
 - (E) Lipoprotein lipase
37. A new hexokinase has been developed that gets rid of the nuisance of the inhibition by glucose 6-phosphate. But the owner of this enzyme is obese. To help with this problem you could:
- (A) genetically remove the hexokinase such that glucose could no longer go down the pentose-phosphate pathway.
 - (B) remove the ATP inhibition of phosphofructokinase. Now glucose will not be sent down the pentose-phosphate pathway.
 - (C) remove the malonyl CoA inhibition of fatty acid oxidation. Now the fatty acids would be burned instead of going to triacylglycerols.
 - (D) eliminate the glucagon receptor so that all of the calories would be burned up.
 - (E) B and C might just possibly help this individual.
38. A liver hepatocyte goes berserk and destroys (eliminates) all of its available malate. In this cell:
- (A) the glycolytic pathway would be partially inhibited.
 - (B) the urea cycle would not be affected by this action.
 - (C) there would now be a great excess of NADPH produced.
 - (D) the concentration of aspartate in the cell would increase.
39. You are trying to build up the blood glucose levels of one of your patients. The diet you prescribe is rich in L-tryptophan. The next thing you do is:
- (A) resign your position since L-tryptophan is purely ketogenic.
 - (B) inhibit the GTPase activity of the alpha subunit of the G-proteins such that

- gluconeogenesis is promoted.
- (C) increase the amount of insulin, since your patient is in an anabolic mode.
 - (D) release the controls of the PFK1 - fructose 1,6 bisphosphate phosphatase so that the system will be more efficient in gluconeogenesis.

40. In the Pentose-Phosphate Shunt:
- (A) one of the two decarboxylations is performed by glucose 6-phosphate dehydrogenase.
 - (B) lactonase breaks a lactone ring after the action of glucose 6-phosphate dehydrogenase.
 - (C) transketolase transfers three carbon units.
 - (D) transaldolase requires thiamine as an essential cofactor.
 - (E) none of the above answers are correct.
41. Which of the following is **NOT** CORRECT?
- A UDP derivative is used in the:
- (A) synthesis of glycogen.
 - (B) conversion of glucose into galactose.
 - (C) formation of the branches in glycogen.
 - (D) activation of glucuronic acid.
 - (E) synthesis of deoxythymidylic acid.
42. In the degradation of glycogen:
- (A) phosphorylase requires the presence of cofactor pyridoxal phosphate.
 - (B) phosphorylase kinase converts phosphorylase from the (a) form to the (b) form.
 - (C) glucagon action leads to the phosphorylation of the phosphoprotein phosphatase inhibitor.
 - (D) phosphorylase converts the glucose at the end of the chain directly into glucose 6-phosphate.
 - (E) the process goes on only in the liver since this is the only tissue that gives us blood glucose.
 - (F) A and C are both correct
43. Acetyl CoA carboxylase:
- (A) is one of those few carboxylation enzymes that do not use biotin.
 - (B) is inhibited by citrate.
 - (C) is activated by phosphorylation.
 - (D) is inhibited by long chain fatty acids.
 - (E) is made less active through the actions of insulin.
44. In the cytosol:
- (A) malate is used as a source of NADH.

- (B) fatty acid synthesis uses malonyl CoA.
- (C) unsaturation (double bonds) can only be introduced from the methyl end up to the #9 carbon.
- (D) the peroxisomes are used to increase the length of unsaturated fatty acids.
- (E) we do not have the machinery to add a hydroxyl group to the alpha carbon of a fatty acid.

45. Which of the following is **NOT** CORRECT?

The incorporation of substantial amounts of ethanol in the diet would promote the formation of:

- (A) 6-phosphogluconate.
- (B) cholesterol.
- (C) uric acid.
- (D) OMP (orotidine monophosphate) from orotic acid.
- (E) 2,3-bis phosphoglycerate (2,3-diphosphoglycerate).

46. To control the production of the prostaglandins one uses:

- (A) aspirin to inhibit lipoxygenase.
- (B) hydrocortisone to inhibit phospholipase A₂.
- (C) carbon monoxide to inhibit cyclooxygenase.
- (D) ibuprofen to inhibit phosphoprotein phosphatase.
- (E) theophylline to inhibit phosphodiesterase

47. In cell membranes:

- (A) the enzyme that carries phospholipids from one side of the membrane to the other is called a “flippase”.
- (B) the integral proteins are absolutely essential to the existence of the membrane.
- (C) the phospholipids facing inside of the cell are less highly charged than those facing out.
- (D) the phospholipids have long organic tails, they move across the surface of the membrane very slowly when compared with the integral proteins.
- (E) the phosphatidyl inositol anchors are used with proteins tied to the outside of the cell.
- (F) A and E are both correct answers

48. Which of the following is **NOT** TRUE?

Membranes allow the passage across their lipid bilayer of:

- (A) steroids
- (B) carbon monoxide
- (C) water
- (D) glutamine
- (E) nitric oxide

49. Your patient is excreting large amounts of citrulline. You restrict his nitrogen intake. You then diagnose a deficiency of:
- (A) arginase and so you prescribe an arginine supplement.
 - (B) ornithine transcarbamylase and ask for a mitochondrial transplant.
 - (C) argininosuccinate synthase and add arginine to the diet.
 - (D) carbamoyl phosphate synthase II and nothing can be done.
 - (E) N-acetyl glutamate and suggest eating more arginine.

50. Which of the following does **NOT** require a “One Carbon” cofactor for its metabolism?
- (A) tryptophan
 - (B) histidine.
 - (C) creatine.
 - (D) serine.
 - (E) All of the above require a “one carbon” donor
51. A novel mutation produces hemoglobin 423. In this polymer, 2,3 bis phosphoglycerate combines with **both** the deoxy and the oxy form of hemoglobin. This hemoglobin:
- (A) should be more difficult to oxygenate than the normal hemoglobin.
 - (B) would unload oxygen easily and would very easily pick up oxygen.
 - (C) would be more sensitive to pH changes than the normal variety.
 - (D) should be more easily converted to methemoglobin than is usual .
52. Which of the following is **NOT CORRECT**?
Phosphoribosyl pyrophosphate (PRPP) is:
- (A) used in the synthesis of phosphoribosylamine.
 - (B) used in the 2-deoxy form when added to thymine.
 - (C) used in the recovery of the pyrimidine bases U and T.
 - (D) used to convert orotic acid into orotidine monophosphate (OMP).
 - (E) formed by the pentose phosphate pathway.
53. The kidney cortex:
- (A) uses fatty acids but does not require oxygen in this pathway.
 - (B) uses glutamine or ketone bodies as a fuel source.
 - (C) uses glucose as its major fuel.
 - (D) converts fatty acids into ketone bodies for the use of the medulla.
 - (E) uses glucose produced by the kidney medulla
54. A marathon runner intending to impress his/her fiance sprints past the reviewing stand half way through the race. As a result of this impulsive action:
- (A) the Cori cycle runs backward since the muscle now really needs glucose.
 - (B) the amount of fatty acids metabolized increases markedly. The resulting ketone bodies hurt and the runner regrets his/her behavior.
 - (C) the respiratory quotient briefly increases and then returns to the original level.
 - (D) the sudden increase in the alanine cycle means that the kidney will have to form more urea.
 - (E) the sudden demand for oxygen causes a brief blackout and the runner stumbles and falls.

55. A new enzyme inhibitor - developed exclusively for this exam - blocks the action of glucose 6-phosphate dehydrogenase. This inhibitor would affect:
- (A) the inhibition of phosphofructokinase-1 by ATP.
 - (B) glyceraldehyde 3-phosphate dehydrogenase.
 - (C) enolase.
 - (D) pyruvate kinase.
 - (E) both B and D are correct answers

56. Which of the following is **NOT** TRUE?

Ethanol in the diet:

- (A) provides an abundance of NADH and is a good source of acetyl CoA.
- (B) limits the use of glucose in providing carbon skeletons for the synthesis of triacyl glycerol.
- (C) is metabolized to acetaldehyde and this active acetaldehyde inhibits phosphofructokinase-1.
- (D) increases the use of the pentose-phosphate pathway.

57. Which of the following is **NOT** a substrate for gluconeogenesis?

- (A) lysine
- (B) propionate
- (C) alanine
- (D) succinate
- (E) tryptophan

58. Which of the following **DOES NOT** correctly tie together the items in columns A and B?

A	B
(A) Mitochondrial malate dehydrogenase.	Source of NADPH
(B) Thiamine	Transketolase
(C) Glucose 6-phosphate dehydrogenase	Inhibition by NADPH
(D) Ribose	Possible product of pentose-phosphate shunt

59. Which of the following is **NOT** CORRECT?

Fructose can be:

- (A) used to synthesize palmitate in the liver
- (B) converted into threonine.
- (C) made into galactose.
- (D) metabolized in the muscle without any help from insulin.
- (E) metabolized in either the glycolytic or the pentose-phosphate pathway.

60. Glucagon helps in the:

- (A) conversion of glucose-1 phosphate into glycogen.
- (B) formation of the polyamines from ornithine.
- (C) pathway that turns acetyl CoA into cholesterol.

- (D) synthesis of triacylglycerol from glycerol and fatty acyl CoA substrates.
- (E) synthesis of glucose from alanine.

61. In the metabolism of glycogen:
- (A) branching enzyme produces the α 1-6 linkages.
 - (B) the synthetic reaction requires glucose in the form of CDP-glucose.
 - (C) the first degradation product after glycogen is glucose 6-phosphate.
 - (D) branching enzyme deficiency produces glycogen with short branches.
 - (E) the synthesis takes place in the cytosol and the degradation takes place in the mitochondria.
62. There is an old saying that “fat burns in the flame of carbohydrate.” One possible reason for this could be that:
- (A) glucose makes dihydroxyacetone phosphate which is then converted into glycerol phosphate. This is needed in lipid metabolism.
 - (B) carbohydrates produce oxaloacetate which is used in the citric acid cycle.
 - (C) carbohydrates produce acetyl CoA which is then burned.
 - (D) when conditions are appropriate to burn fats, then carbohydrates are also burned.
 - (E) carbohydrates produce citrate, which is necessary for fatty acid oxidation.
63. In the presence of the phosphoprotein phosphatase inhibitor in its phosphorylated form:
- (A) glycogen synthase becomes activated.
 - (B) hormone sensitive lipase becomes less active.
 - (C) β -hydroxymethyl glutaryl CoA reductase is increased in activity
 - (D) ornithine decarboxylase is increased in activity.
 - (E) phosphorylase kinase remains in the activated form.
64. In response to a low insulin to glucagon ratio, hormone-sensitive lipase:
- (A) becomes attached to the capillary membranes by a carbohydrate polymer composed of glycosoaminoglycans prior to activation and cleavage of VLDL.
 - (B) hydrolyzes the phospholipids of VLDL and chylomicrons to produce fatty acids.
 - (C) hydrolyzes intracellular triacylglycerols in adipose tissue to generate fatty acids for muscle and glycerol for gluconeogenesis.
 - (D) is first activated by apolipoprotein ApoCII before it can efficiently cleave VLDL produced by the liver or chylomicrons from fat in the diet.
65. Which of the following is **NOT** used in fatty acid oxidation?
- (A) Desaturation.
 - (B) Decarboxylation, but only for α -oxidation of branched chain fatty acids
 - (C) Oxidation of a beta hydroxyl group to a beta ketone group.
 - (D) A cleavage of a beta keto acyl CoA with another Coenzyme A molecule.
 - (E) All of the above are used in the oxidation of fatty acids.
66. Cholesterol:

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| (B) phosphofructokinase-1 | fructose 1,6-bisphosphate |
| (C) pyruvate kinase | phosphoenolpyruvate |
| (D) pyruvate dehydrogenase | ATP |
| (E) pyruvate carboxylase | acetyl CoA |

71. A new (hypothetical) virus emerges that encodes an enzyme that destroys fructose 2,6-bisphosphate in liver hepatocytes. Infection with this virus will likely:

- (A) increase the effects of insulin
- (B) lead to an uninhibited phosphofructokinase-1 (PFK-1)
- (C) increase the effects of glucagon
- (D) block gluconeogenesis
- (E) immediately kill anyone who becomes infected.

72. Consider the following two series of reactions in glycogen metabolism:

- 1) Glycogen \rightleftharpoons Glucose-1-phosphate \rightleftharpoons Glucose 6-phosphate
- 2) Glucose-1-phosphate \rightleftharpoons UDP-glucose \rightleftharpoons Glycogen

In a resting liver cell, this cycle is controlled by the allosteric effector:

- (A) glucose-6-phosphate
 - (B) AMP
 - (C) fructose 1,6-bisphosphate
 - (D) cyclic AMP
 - (E) all of the above are correct
73. Which of the following **DOES NOT** link the lipid with the correct definition, components or description?

Lipid	Definition or Components
(A) phospholipid	complex lipid
(B) ceramide	sphingosine plus a fatty acid
(C) plasmalogen	glycerol and an ether linked vinyl chain
(D) lecithin	sphingosine plus choline phosphate
(E) prostaglandin	arachidonate-derived

74. Which of the following is **NOT** true?

On their way from adipose tissue to the final state of oxidation, fatty acids are found as different derivatives. In this process, the fatty acids are:

- (A) stored in the adipose tissue as glycerol esters
 - (B) transferred from the adipose tissue to the other tissues (muscle, etc) as the glycerol esters
 - (C) transported in the blood as free or albumin-bound fatty acid
 - (D) taken across the mitochondrial membrane as the carnitine ester
 - (E) metabolized in the mitochondrial matrix as a coenzyme A thiolester
75. Which of the following is **NOT** true?

In the mammalian system of fatty acid synthesis:

- (A) the pantothenic acid side chain is attached to the acyl carrier protein
- (B) enzyme activities are carried on two large polypeptide chains
- (C) the cofactor used to reduce the ketones to alcohols and the double bonds to a saturated chain is NADPH

- (D) malonyl CoA is used for all of the carbon atoms except those provided by the primer
- (E) the primer is acetyl CoA
- (F) all statements are true as near as I can tell!

76. Cholesterol:
- (A) must be built into the membrane at the time that the structure is being put together
 - (B) is removed from the membrane inside of the cell as the membrane is taken into the cell in a recycling process
 - (C) is converted into the cholesterol ester through the direct action of the lipoprotein lipase
 - (D) is transported to the tissues in a form that is covalently bound to the lipoprotein. It is released as this protein cholesterol ester is digested in the lysosome
 - (E) is excreted only by the liver and this is in the form of the bile acids or bile salts
 - (F) B and E are both correct answers.
77. In amino acid metabolism:
- (A) cysteine is degraded only by the reactions of the citric acid cycle
 - (B) arginine is degraded directly to aspartic acid and then into a citric acid cycle intermediate
 - (C) tyrosine is converted into compounds that are used to synthesize both glucose and the ketone bodies
 - (D) proline is acted on by enzymes to glycolytic pathway
 - (E) methionine is the only purely ketogenic non-essential amino acid
78. Which of the following is **NOT TRUE** with regard to the indicated amino acids?
- (A) valine, isoleucine and leucine are transaminated to three alpha keto acids
 - (B) amino acids that have branched chains all use a reaction similar to that of pyruvate dehydrogenase and so generate NADH during their oxidation.
 - (C) valine and isoleucine make use of FAD in forming a double bond
 - (D) valine and isoleucine require the use of vitamin B₁₂ (cobalamin) on their way to form glucose
 - (E) leucine and lysine differ from the others in being both ketogenic and glucogenic
79. In the purine biosynthetic pathway:
- (A) one of the nitrogen atoms comes from glutamine while all of the rest come from other nitrogen donors
 - (B) two of the nitrogen atoms in guanosine come from aspartic acid
 - (C) glycine is used to provide the carbons that join together the six and five-membered rings
 - (D) the ribose phosphate is added after the purine ring is made. In this respect, the synthesis of the purines and of the pyrimidines is similar
 - (E) GTP is the nucleotide used to provide the energy required for synthesis
80. With regard to the urea cycle in mitochondria:
- (A) fatty acid oxidation is stimulated in order to generate allosteric signals used in the control of gluconeogenesis and the urea cycle

- (B) our systems are unable to make use of the reduced cofactors generated in amino acid oxidation. This energy is therefore used in gluconeogenesis
- (C) the synthesis of argininosuccinate in the urea cycle is controlled by the K_m relationships with its two substrates, citrulline and aspartic acid
- (D) the transport of ornithine coming into the mitochondria is balanced (except for the charge difference) by the citrulline which is leaving this organelle
- (E) one of the nitrogen atoms in urea is added by the addition of argininosuccinate to the urea precursor
- (F) Answers A, C and D are each correct statements

81. In the gut, liver, and adipose tissue systems, plasma:
- (A) cholesterol can be taken from the tissues to the liver where it is burned to CO₂ and water
 - (B) glycerol is released by the adipose tissue and used by the liver
 - (C) carries the ketone bodies released by the adipose tissue for use by the liver or the gut
 - (D) maintains a balance of amino acids. All tissues use the same relative amounts of these acids such that the distribution in the plasma does not change
 - (E) contains the lipoproteins released by the adipose tissue and returns the free fatty acids to the adipose tissue for conversion into more lipoproteins
82. Tissues in the body are specialized such that they prefer to use certain fuels for energy. In this regard, which of the following **DOES NOT** use glucose as a major fuel?
- (A) lung
 - (B) fast twitch muscle fiber
 - (C) kidney medulla
 - (D) adipose tissue
 - (E) kidney cortex
83. The kidney contributes to normal acid-base pH balance by:
- (A) secretion of ammonia
 - (B) reabsorption of bicarbonate
 - (C) Increased ketogenesis
 - (D) Decreased CO₂ uptake
 - (E) Both A and B contribute
84. In the hepatic metabolism of ethanol, which of the following is **INCORRECT**?
- (A) hepatic oxidation of lactate is impaired.
 - (B) gluconeogenesis is impaired.
 - (C) the NADH/NAD⁺ ratio increases.
 - (D) fatty acid oxidation is impaired.
 - (E) acetaldehyde dehydrogenase changes the NADPH/NADP⁺ ratio.
85. Metabolism of which of the following will yield a net synthesis of glucose in the liver or kidney?
- (A) acetoacetic acid
 - (B) leucine
 - (C) palmitic Acid
 - (D) acetic Acid
 - (E) glutamate
86. Which of the following fatty acids would yield the **HIGHEST** level of gluconeogenic precursors?

- (A) palmitic acid (16:0)
- (B) heptadecanoic acid (17:0)
- (C) stearic acid (18:0)
- (D) phytanic acid (C-20 polyisoprenoid)
- (E) arachidonic acid (20:4)

87. Which of the following intermediates in the oxidation of odd-chain length fatty acids is likely to appear in the urine in patients with a vitamin B₁₂ deficiency?
- (A) formic acid
 - (B) methylmalonate
 - (C) pentanoic acid
 - (D) propionate
 - (E) succinate
88. Which of the following statements about ketogenesis is **INCORRECT**?
- (A) ketone bodies appear in the urine during both fasting and starvation.
 - (B) ketone bodies are produced during periods when the liver is glucogenic.
 - (C) ketone bodies are a major source of energy in the liver during fasting and starvation.
 - (D) ketones have more caloric value as an energy source per gram than glucose.
 - (E) ketogenesis is a mechanism for regeneration of CoA in the mitochondrion.
89. A three year old patient with mild mental retardation was found to have cloudiness of the lens, indicative of cataract formation. When abnormally high blood concentrations of a sugar alcohol was detected, the child was immediately placed on a milk-free diet. The enzyme most likely to be defective in this child is:
- (A) hexokinase
 - (B) galactose-1-phosphate uridyl transferase
 - (C) galactokinase
 - (D) lactase
 - (E) B and C are both possibilities

Problems 90-93 refer to the following case:

A 1.5 month old male infant was admitted to the hospital because of vomiting and failure to gain weight. During hospitalization, he became drowsy, lethargic, and finally comatose with convulsions. He improved after receiving intravenous glucose and came out of the coma. A few days later, his plasma ammonium ion concentration was found to be 180 $\mu\text{mol/L}$ (normal, 11-50 $\mu\text{mol/L}$).

90. The abnormally high level of ammonium ions is consistent with a genetic defect in:
- (A) synthesis of carbamoyl phosphate.
 - (B) formation of N-acetylglutamate.
 - (C) synthesis of glutamine from glutamate.
 - (D) synthesis of citrulline.
 - (E) any of the above.
91. High blood ammonium ion concentrations in some patients like this can be lowered by treatment with arginine. The reason for this is that:

- (A) the defective enzyme prevents arginine biosynthesis.
 (B) a defect in the utilization of arginine in protein biosynthesis requires excess arginine, especially in developing infants.
 (C) the enzyme arginase is defective.
 (D) arginine increases the effective concentration of an acceptor for a carbamoylation reaction in the liver.
 (E) arginine is an essential amino acid for infants.
92. With informed consent of the parents, the infant was challenged with a high protein load. After the high protein diet, one might expect that some ketone bodies would form. Ketone bodies can be obtained from carbon atoms of which of the following amino acids?
- (A) arginine, isoleucine, valine.
 (B) arginine and valine.
 (C) isoleucine and leucine.
 (D) leucine only.
 (E) none of the above amino acids would be ketogenic in this patient.
93. The patient's protein intake was maintained at a low level. However, one must be careful that the diet contains sufficient amounts of which essential amino acids?
- (A) isoleucine, phenylalanine, threonine
 (B) isoleucine and threonine
 (C) phenylalanine and tyrosine
 (D) tyrosine only
 (E) ornithine and citrulline
94. What is the pKa of an acid which has a pH of 6.8 when its base:acid ratio is 1:20?
- A. pKa = 5.5
 B. pKa = 6.3
 C. pKa = 8.1
 D. pKa = 7.5
 E. pKa = 8.0
95. A patient comes into the emergency room feeling faint. He says that he is an insulin-dependent diabetic. His blood gases have been determined and you see his value for $[\text{HCO}_3^-] = 18$ milliequivalents per liter and for $\text{P}_a\text{CO}_2 = 38$ mm Hg. Based on your calculations this patient has
- A. A blood pH of 7.3 and is just slightly acidotic. He is in no real danger.
 B. A blood pH of 5.78 and by all rights should be dead!
 C. A blood pH of 6.42 and is extremely acidotic. He should be treated immediately.
 D. A blood pH of 7.95 and is suffering from alkalosis. He should be treated immediately.

- E. You cannot determine the blood pH from these values.
96. Choose the **INCORRECT** statement concerning the modification of amino acids:
- A. Ser, Thr and Tyr are all reversibly phosphorylated and these modifications can control enzyme activities and signal transduction cascades.
 - B. Asn is frequently modified by large N-glycosidically linked oligosaccharide structures that frequently play a role in protein folding, stability and function.
 - C. Ascorbic acid (Vitamin C) is critical for the activity of the enzymes that hydroxylate Arg and Lys residues in collagen.
 - D. The covalent addition of fatty acids, prenyl groups and glycosylphosphatidylinositol (GPI) anchors help proteins to associate with membranes.
 - E. The nonenzymatic glucosylation of hemoglobin amino groups generates the isoform HbA1c which is used to determine the long term maintenance of diabetics' blood glucose levels.

97. Which type of non-covalent bond is the primary contributor to the secondary structure of proteins?
- A. Peptide bond
 - B. Hydrophobic interactions
 - C. Hydrogen bond
 - D. Van der Waals forces
 - E. Disulfide bond
98. You have found that there are elevated levels of a specific enzyme in the serum of patients in the early stages of emphysema. It happens that this enzyme exists in two isoforms, A and B, that have the same molecular weights but differ slightly in their amino acid sequences. It seems that the B isoform predominates in the lung whereas the A isoform is found in most other tissues. You need to find a way to distinguish these two isoforms so you can distinguish lung tissue damage from other tissue damage. Which protein separation technique would you try first?
- A. Ultracentrifugation
 - B. Gel filtration
 - C. Nondenaturing or native gel electrophoresis
 - D. SDS-polyacrylamide gel electrophoresis (SDS-PAGE)
 - E. Western immunoblot analysis
99. As catalysts of biochemical reactions, enzymes:
- A. Alter the equilibrium of the reaction.
 - B. Alter the energy of activation of the reaction.
 - C. Alter the Gibbs free energy of the reaction.
 - D. Alter the enthalpy of the reaction.
 - E. Alter the entropy of the reaction.
100. You have identified a 4 step biosynthetic pathway for an unusual amino acid required for protein synthesis in a parasite that is deadly to humans. One way to treat infected patients is to block the pathway by inhibiting a critical enzyme of the pathway. You must work quickly! Which enzyme would you choose to study first and why?
- A. You would choose the last enzyme in the pathway because it is right before product formation.
 - B. You would choose the first enzyme in a pathway because it is likely to be under feedback control.
 - C. You would choose an enzyme that has the longest half-life because it works longer before being degraded.
 - D. You would choose the enzyme that works at equilibrium because it works more steadily.

- E. It is likely that all the enzymes are under some sort of control so it wouldn't really matter which one you started to study first.

That's all folks!!!

Good luck on the final exam!

...Answers on the next two pages...

MEDICAL BIOCHEMISTRY 531 - ACKERMAN
TEACHING-LEARNING EXAMINATION FALL 2003

THE ANSWERS REVEALED!

- | | | |
|----------------------|----------------------|----------------------|
| 1. <u> B </u> | 21. <u> A </u> | 41. <u> C </u> |
| 2. <u> C </u> | 22. <u> B </u> | 42. <u> F </u> |
| 3. <u> D </u> | 23. <u> E </u> | 43. <u> D </u> |
| 4. <u> A </u> | 24. <u> D </u> | 44. <u> B </u> |
| 5. <u> A </u> | 25. <u> B </u> | 45. <u> E </u> |
| 6. <u> A </u> | 26. <u> A </u> | 46. <u> B </u> |
| 7. <u> C </u> | 27. <u> A </u> | 47. <u> F </u> |
| 8. <u> E </u> | 28. <u> A </u> | 48. <u> D </u> |
| 9. <u> C </u> | 29. <u> A </u> | 49. <u> C </u> |
| 10. <u> B </u> | 30. <u> D </u> | 50. <u> A </u> |
| 11. <u> E </u> | 31. <u> A </u> | 51. <u> A </u> |
| 12. <u> A </u> | 32. <u> D </u> | 52. <u> B </u> |
| 13. <u> D </u> | 33. <u> D </u> | 53. <u> B </u> |
| 14. <u> D </u> | 34. <u> E </u> | 54. <u> C </u> |
| 15. <u> C </u> | 35. <u> B </u> | 55. <u> B </u> |
| 16. <u> A </u> | 36. <u> D </u> | 56. <u> C </u> |
| 17. <u> C </u> | 37. <u> E </u> | 57. <u> A </u> |
| 18. <u> E </u> | 38. <u> A </u> | 58. <u> A </u> |
| 19. <u> C </u> | 39. <u> B </u> | 59. <u> B </u> |
| 20. <u> E </u> | 40. <u> B </u> | 60. <u> E </u> |

- | | | | |
|-----|------------------|------|------------------|
| 61. | <u> A </u> | 86. | <u> D </u> |
| 62. | <u> B </u> | 87. | <u> B </u> |
| 63. | <u> E </u> | 88. | <u> C </u> |
| 64. | <u> C </u> | 89. | <u> E </u> |
| 65. | <u> E </u> | 90. | <u> E </u> |
| 66. | <u> A </u> | 91. | <u> D </u> |
| 67. | <u> C </u> | 92. | <u> C </u> |
| 68. | <u> B </u> | 93. | <u> A </u> |
| 69. | <u> C </u> | 94. | <u> C </u> |
| 70. | <u> B </u> | 95. | <u> A </u> |
| 71. | <u> C </u> | 96. | <u> C </u> |
| 72. | <u> A </u> | 97. | <u> C </u> |
| 73. | <u> D </u> | 98. | <u> C </u> |
| 74. | <u> B </u> | 99. | <u> B </u> |
| 75. | <u> B </u> | 100. | <u> B </u> |
| 76. | <u> F </u> | | |
| 77. | <u> F </u> | | |
| 78. | <u> C </u> | | |
| 79. | <u> E </u> | | |
| 80. | <u> C </u> | | |
| 81. | <u> D </u> | | |
| 82. | <u> E </u> | | |
| 83. | <u> E </u> | | |
| 84. | <u> E </u> | | |

85. E