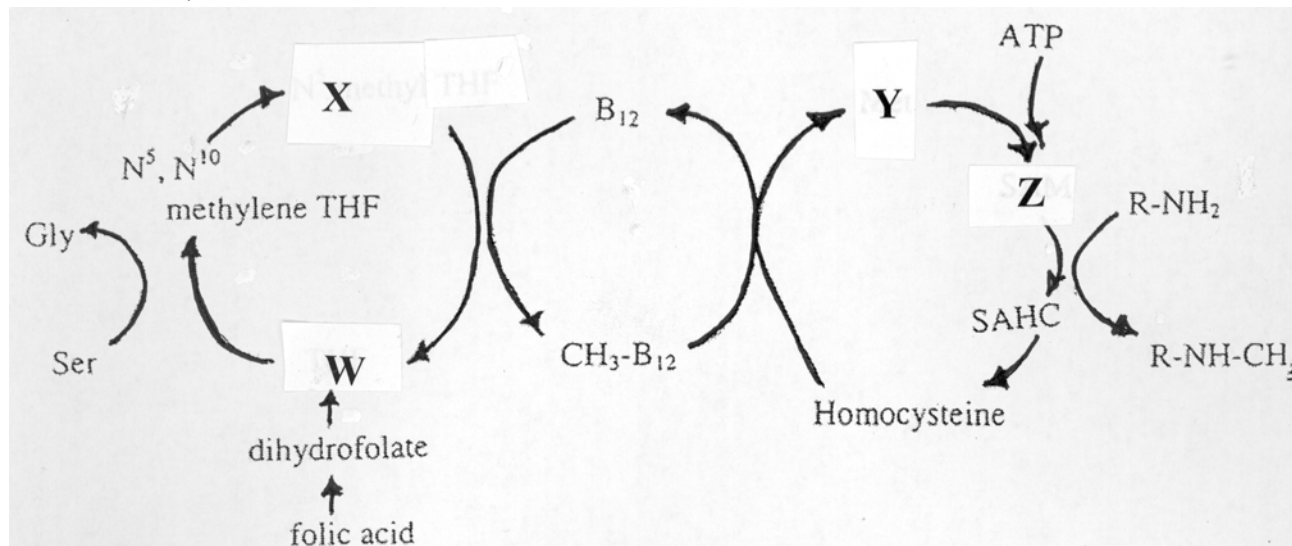


The first three questions refer to the flow chart shown below. It is suggested that you identify the compounds W, X, Y, and Z and put their names in the appropriate places before attempting to answer Questions 1-3.



1) This is “Be kind to Your Name week.” Compound W is most likely to be:

- A) dihydrofolate
- B) tetrahydrofolate
- C) N<sup>10</sup>-formyl-tetrahydrofolate
- D) N<sup>5</sup>-methyl-tetrahydrofolate
- E) N<sup>5</sup>,N<sup>10</sup>-methenyl-tetrahydrofolate

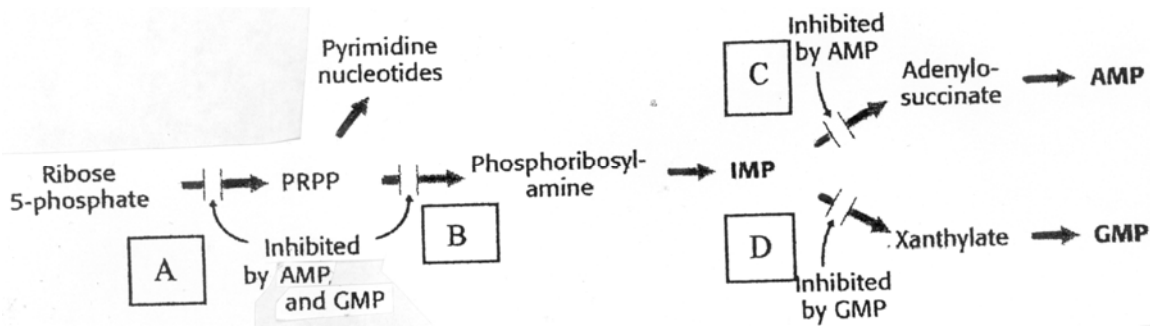
2) Compounds X, Y, and Z are most likely to be:  
(THF = tetrahydrofolate)

	<u>Compound X</u>	<u>Compound Y</u>	<u>Compound Z</u>
A)	dihydrofolate	glutamate	AMP
B)	N <sup>5</sup> -methyl-THF	methionine	S-adenosylmethionine
C)	N <sup>10</sup> -formyl-THF	cobalamin	cysteine
D)	methotrexate	S-adenosylhomocysteine	biotin
E)	THF	N <sup>5</sup> , N <sup>10</sup> -methylene-THF	methionine

3) A patient with a bladder infection is taking Bactrim, a combination drug that inhibits bacterial tetrahydrofolate synthesis. Using the above flow diagram as a guide, you would most likely find in analyzing the bacteria:  
(increased or decreased, relative to bacteria not treated with Bactrim)

	<u>Dihydrofolate reductase enzyme activity</u>	<u>Overall folate level</u>
A)	increased	increased
B)	increased	decreased
C)	decreased	decreased
D)	decreased	increased
E)	no change	no change

OK, this page still represents “Be kind to **Your Name** week.” Questions 4-6 refer to the diagram below. Choose one letter (A, B, C, or D) most appropriate for the question.



- 4) To block only AMP synthesis, you would inhibit at position: **A or B or C or D?**
- 5) To block synthesis of purine nucleotides only, you would inhibit at position: **A or B or C or D?**
- 6) To block all nucleotide synthesis, you would inhibit at position: **A or B or C or D?**
- 7) What is the difference between GDP and dGDP?
  - A) dGDP has an extra amino group attached to carbon 6 in the purine ring
  - B) dGDP has an extra phosphate attached to the 2'-carbon of the ribose
  - C) dGDP is reduced at the 2'-carbon of the ribose
  - D) dGDP is reduced at carbon 6 of the purine ring
  - E) dGDP is a purine nucleotide while GDP is a pyrimidine nucleotide

8) The enzyme and the direct product of the committed step in the *de novo* pyrimidine nucleotide synthesis pathway are:

<u>Enzyme</u>	<u>Direct Product</u>
A) carbonic anhydrase	bicarbonate
B) carbamoyl phosphate synthetase II	carbamoyl phosphate
C) carbamoyl phosphate synthetase I	orotate
D) carbamoyl phosphate synthetase II	orotate monophosphate
E) orotate phosphoribosyl transferase	uridine monophosphate

9) The mother of a 2-year-old boy is concerned about his tendency to bite himself to the point of bleeding. The boy’s fingers show scarring and several scabs and his lips are swollen and bruised. He exhibits poor coordination, poor muscle tone, and frequent jerking movements of his arms and legs. He is significantly delayed in speech. His urine is “orange” in color and appears “gritty.” Which of the following is the most likely diagnosis?

- A) Diabetes mellitus, type II
- B) Glucose 6-phosphatase deficiency
- C) Glucose 6-phosphate dehydrogenase deficiency
- D) Lesch-Nyhan syndrome
- E) Pyruvate kinase deficiency

The next **two questions** deal with the following case description.

A 72-year-old man presents with: (a) symptoms of anemia (labored breathing, fatigue, etc.); (b) dark colored stool; and (c) paresthesia (skin sensation tingling, prickling, itching). Indeed, his Complete Blood Count (CBC) showed low hematocrit, low erythrocyte count, and low hemoglobin. A peripheral blood smear shows marked anisocytosis and poikilocytosis. The mean corpuscular volume is increased above the normal range.

10) On the basis of these observations, you suspect that the patient is most likely to have:

- A) glucose 6-phosphate dehydrogenase deficiency
- B) iron deficiency anemia
- C) pernicious anemia
- D) sickle cell anemia
- E) thalassemia

11) More specifically, you suspect a deficiency in X and order administration of Y to test your hypothesis.

	<u>X</u>	<u>Y</u>
A)	vitamin B <sub>1</sub> thiamine	transketolase
B)	vitamin B <sub>3</sub> niacin	nicotinamide
C)	vitamin B <sub>12</sub> cobalamin	intrinsic factor
D)	vitamin C ascorbate	collagen
E)	vitamin H biotin	pyruvate carboxylase

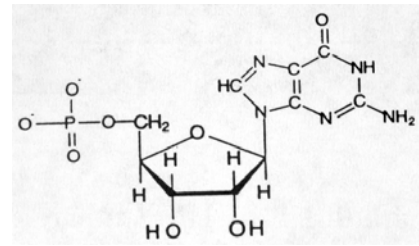
12) A 27-year-old woman presents with weakness, easy fatigability, nausea, and diarrhea but **no** neurological signs (**no** parasthesias). Her CBC and blood smear report were particularly informative: megaloblastic anemia. You run additional tests that confirm your hypothesis. You suggest that she should include green leafy vegetables in her diet and you warn her that if she becomes pregnant, her fetus might be at risk for:

- A) Beri-Beri
- B) Ehlers-Danlos syndrome
- C) Neural tube defects
- D) Osteogenesis imperfecta
- E) Scurvy

13) Methotrexate is a potent anti-cancer agent that starves dividing cells of deoxyribonucleotides through direct inhibition of which of the following enzymes?

- A) Ribonucleotide reductase
- B) Xanthine oxidase
- C) Carbamoyl phosphate synthetase II
- D) Thymidylate synthetase
- E) Dihydrofolate reductase

14) Regarding the structure shown at the right, which of the following statements is true?



- A) Inhibits overall activity of ribonucleotide reductase
- B) First pyrimidine nucleotide in *de novo* synthesis pathway
- C) Direct product of major purine salvage mechanism
- D) Product of reaction catalyzed by nucleoside phosphorylase
- E) Synthesis requires oxidation of tetrahydrofolate to dihydrofolate

15) Phosphoribosyl pyrophosphate (PRPP) is a direct, activated reactant in the regulated step of which of the following pathways?

- A) *de novo* synthesis of purine nucleotides
- B) synthesis of S-adenosylmethionine
- C) deoxyribonucleotide synthesis
- D) *de novo* synthesis of pyrimidine nucleotides
- E) thymidylate synthesis

16) What effect does the binding of CO<sub>2</sub> and H<sup>+</sup> to hemoglobin (Hb) in the peripheral tissues have on the body?

- A) the binding of CO<sub>2</sub> and H<sup>+</sup> stabilize the tense form of Hb
- B) binding of H<sup>+</sup> to Hb serves as a buffer to regulate the pH of peripheral tissues
- C) it facilitates the release of O<sub>2</sub> in the peripheral tissues
- D) all of the above
- E) none of the above

17) Problems in the glycolytic pathway can affect oxygen transport in red blood cells. How would a hexokinase deficiency alter oxygen affinity?

	<u>2,3 diphosphoglycerate (DPG)</u>	<u>oxygen affinity</u>
A)	increase	increase
B)	increase	decrease
C)	decrease	increase
D)	decrease	decrease
E)	no change	no change

18) Which of the following physiologic/pathologic conditions is most likely to result in alkalosis, provided the body cannot fully compensate?

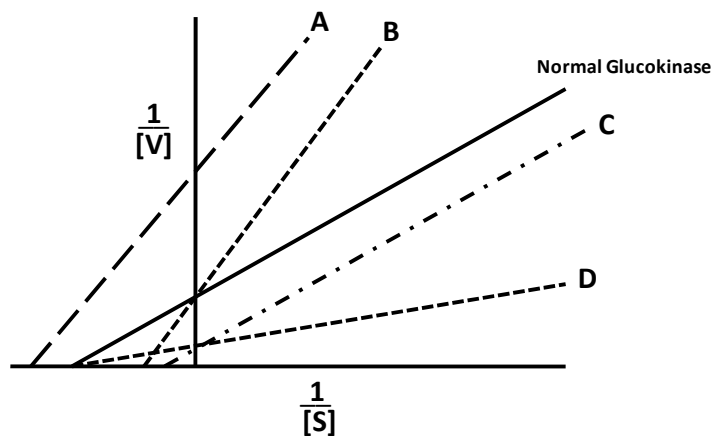
- A) Production of lactic acid by muscles during exercise.
- B) Production of ketone bodies by a patient with diabetes mellitus.
- C) Repeated vomiting of stomach contents, including HCl.
- D) Diarrhea with loss of the bicarbonate anions secreted into the intestine.
- E) An infection resulting in a fever and hypercatabolism.

- 19) Regulation of metabolic pathways includes all of the following except:
- A) Regulation of key enzymes in a pathway.
  - B) Control transcription and translation to regulate enzyme amounts.
  - C) Tissue-specific enzymes and receptors.
  - D) Reciprocal synthetic and degradative pathways utilize the exact same set of enzymes.
  - E) Compartmentation of reciprocal synthetic and degradative pathways.
- 20) Which of the following **correctly** pairs a pathway with one of its regulated enzymes?
- A) TCA cycle: fumarase
  - B) beta-oxidation of fatty acids: CAT II
  - C) glycolysis: phosphoglycerate kinase
  - D) fatty acid synthesis: acetyl CoA carboxylase
  - E) urea cycle: carbamoyl-phosphate synthetase II (CPS II)

The **next two** questions deal with the following case description.

A 6 month old boy is admitted to the clinic suffering from hyperglycemia that was first misdiagnosed as permanent neonatal diabetes. Upon further investigation, however, it appears the patient's insulin secretion system is completely intact. Lab reports suggest that the boy has a defect in the gene encoding glucokinase.

- 21) Given what you know about glucokinase, which of the following processes in the liver will **NOT** be affected by the defective enzyme?
- A) The entry of carbon into glycolysis following carbohydrate rich meal
  - B) The ability to produce NADPH through the pentose phosphate pathway
  - C) Glycogen synthesis following consumption of carbohydrate rich meal
  - D) The ability to trap all of the carbon that makes up sucrose inside the cell
  - E) The ability to generate glucose from protein rich foods
- 22) Upon careful analysis, you discover that the mutation causes the patient's glucokinase to behave exactly like hexokinase (i.e. an increased affinity for glucose (decreased  $K_m$  value) and a decrease in the  $V_{max}$ ). Which of the following lines on the Lineweaver-Burke Plot below best represents the mutated enzyme compared to normal glucokinase?



The following **three questions** deal with the scenario described below.

A slightly obese man is rushed to the emergency room by ambulance. The EMTs had noticed that the man had some newly developed diet pills on his end table next to his television remote. Upon googling the new diet pills, you realize that they are intended to increase metabolism without exercise. You quickly deduce that these pills are intended to function as a mitochondrial uncoupler.

23) If these “diet” pills really are uncouplers, which of the following would correctly describe the patient’s mitochondria in terms of oxygen consumption and ATP production?

	<u>Oxygen Consumption</u>	<u>ATP Production</u>
A)	Below normal	normal
B)	Above normal	normal
C)	Above normal	below normal
D)	Below normal	below normal
E)	Above normal	above normal

24) Your attending physician is not as biochemically inclined as you and decides to attempt to correct the metabolic imbalance in the patient by treating him with cyanide gas. What would you expect to observe in comparison to the diet pills alone?

	<u>Oxygen Consumption</u>	<u>ATP Production</u>
A)	Increased	restored
B)	Increased	unchanged
C)	Decreased	restored
D)	Decreased	unchanged
E)	unchanged	unchanged

25) Right before the patient collapsed, which of the following would you expect to see in his liver compared to a normal, non-diet pill popping couch potato?

- A) Decreased beta oxidation
- B) Increased TCA cycle activity
- C) Decreased glycolytic activity
- D) Increased glycogen synthesis
- E) Increased gluconeogenesis

26) Gluconeogenesis is not an exact reversal of glycolysis and different reactions are required to convert pyruvate to glucose. Which of the following incorrectly pairs a glycolytic enzyme with the gluconeogenic enzyme that catalyzes the “reverse” reaction?

<u>Glycolysis</u>	<u>Gluconeogenesis</u>
A) phosphofructokinase	fructose 1,6 bisphosphatase
B) pyruvate kinase	pyruvate carboxylase
C) hexokinase	glucose 6-phosphatase
D) pyruvate kinase	phosphoenolpyruvate carboxykinase
E) pyruvate kinase	pyruvate dehydrogenase

The following **6 questions** deal with the following clinical case and lab data.

A mother brings her daughter into the clinic because she is worried about her development and protruding abdomen. The young girl is clearly developmentally delayed and the mother describes frequent nose bleeds and episodes of hyperventilation, especially in the morning upon waking. You run a battery of lab tests and the results are given below.

<u>Assay</u>	<u>Patient</u>	<u>Normal</u>
Sodium (Na <sup>+</sup> )	130 meq/L	135-145
Potassium (K <sup>+</sup> )	3.9 meq/L	3.5-4.9
Calcium (Ca <sup>+2</sup> )	9.2 meq/L	8.0-10.5
Chloride (Cl <sup>-</sup> )	84 meq/L	96-110
Total CO <sub>2</sub> content	13 meq/L	20-30
pH	7.25	7.35-7.45
Anion Gap	38	14-15
Glucose	15 mg/dL	65-110
Creatinine	1.1 mg/dL	0.6-1.4
BUN	13 mg/dL	6-23
Uric Acid	9.0 mg/dL	2-7
Lactate	40 mg/dL	4.5-14.4
Pyruvate	3.6 mg/dL	0.3-0.9
Protein		
Total	17.7 g/dL	6.0-8.4
Albumin	10 g/dL	3.5-5
Globulin	7.7 g/dL	2.3-3.5
Liver biopsy		
Glycogen	10 g/100 g tissue	up to 6 g
Lipid	20 g/100 g tissue	less than 5 g
Glucose 6-phosphatase	20 U/g of liver nitrogen	170-260
Phosphorylase	20 U/g of liver nitrogen	19-25
Fructose 1,6-bisphosphatase	9 U/g of liver nitrogen	4-16

(a) pK<sub>a</sub> of bicarbonate-CO<sub>2</sub> buffer, 6.1; (b) solubility coefficient for CO<sub>2</sub> at 37°C, 0.03 mM/mmHg.

27) Based upon these lab data, what is the acid-base status of the patient?

- A) Acid-base normalcy
- B) Respiratory acidosis
- C) Respiratory alkalosis
- D) Metabolic acidosis
- E) Metabolic alkalosis

28) Given the available anion gap lab data, which of the following is closest to the concentration of HCO<sub>3</sub><sup>-</sup> in the patient?

- A) 4 mEq/L
- B) 8 mEq/L
- C) 12 mEq/L
- D) 16 mEq/L
- E) 20 mEq/L

29) Liver samples from the patient would show which of the following?

- A) Increased ketone body production
- B) Decreased gluconeogenic activity
- C) Decreased protein breakdown
- D) Decreased pentose phosphate pathway activity
- E) Increased glucose exportation

30) Which of the following statements is a possible explanation for the increased levels of lipid in the liver?

- A) Every time she eats, the liver converts the excess to fat because her glycogen stores are never depleted.
- B) As her blood glucose drops the liver cannot export the glucose, and resorts to lipid synthesis instead.
- C) The fatty acids from TAGs are being mobilized to provide precursors for glucose synthesis.
- D) A and B
- E) All of the above.

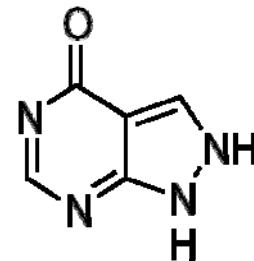
31) The increased levels of uric acid are most likely caused by which of the following?

- A) Increased uric acid production caused by excess fatty acid breakdown
- B) Decreased uric acid secretion caused by high levels of lactic acid
- C) Increased uric acid production caused by decreased glucose 6-phosphate
- D) Excess circulating glucose is broken down directly to uric acid
- E) Increased uric acid production from increased circulating ketone bodies

32) One treatment for the patients with this condition is allopurinol.

Given the structure of allopurinol (right), which of the following symptoms is the target of allopurinol and what is the rationale for its use?

- A) Hypoglycemia by inducing gluconeogenesis
- B) Lactic acidosis by inhibiting pyruvate kinase
- C) Hyperlipidemia by stimulating beta oxidation
- D) Ion imbalance by inhibiting lactate dehydrogenase
- E) Hyperuricemia by inhibiting purine nucleotide biosynthesis



Allopurinol

33) Glucose 6-phosphate is at the crossroads of many metabolic pathways, and its fate depends on the energy needs of a cell. Which of the following **incorrectly** pairs an enzyme that reacts **directly** with glucose 6-phosphate with a product of that metabolic pathway?

- A) phosphoglucoisomerase; pyruvate
- B) glucose 6 phosphatase; glucose
- C) glucose 6 phosphate dehydrogenase; ribose 5 phosphate
- D) UDP glucose pyrophosphorylase; glycogen
- E) glucose 6-phosphate dehydrogenase-NADPH



34) Which of the following statements **correctly** describes lipid delivery to the tissues?

- A) Triacylglycerols (TAGs) are endocytosed by the target tissues.
- B) VLDLs are endocytosed by the liver.
- C) LDLs require clathrin to be endocytosed by all tissues.
- D) Chylomicron remnants require hormone-sensitive lipase for endocytosis.
- E) HDLs have apoB-100 as their sole apoprotein for endocytosis.

35) Which of the following statements regarding bile salts and bile acids is **correct**?

- A) Bile salts are formed by the addition of glycine or taurine to bile acids.
- B) Bile acids and bile salts are synthesized in the gall bladder from cholesterol.
- C) The committed step in bile acid synthesis is catalyzed by HMG-Co reductase.
- D) Bile acids and bile salts are hydrophobic.
- E) The majority of bile salts are excreted.

36) Which of the following statements about VLDLs is **incorrect**?

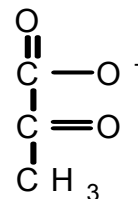
- A) VLDLs are synthesized in the liver.
- B) VLDLs have apoC-II, which is required for the delivery of triacylglycerols (TAGs).
- C) VLDLs are less dense than HDLs.
- D) VLDLs transport endogenous lipids.
- E) VLDLs are converted to IDLs and ultimately to HDLs.

37) Mr. M.'s cells can synthesize LDL receptors, but they are defective. Which of the following statements regarding his conditions would be **true**?

- A) His cells cannot carry out *de novo* synthesis of cholesterol.
- B) His doctor prescribed a drug to inhibit *de novo* synthesis of cholesterol.
- C) He is excreting too much cholesterol, so his tissues cannot make cholesterol in sufficient quantities.
- D) He has high circulating VLDLs because the lipoprotein lipase cannot cleave TAGs.
- E) He has been told to lower his dietary fiber intake, to increase his circulating LDLs.

38) Which of the following statements **correctly** describes the molecule shown below?

- A) It can spontaneously decarboxylate.
- B) A transamination reaction would convert it to alanine.
- C) A carboxylase reaction converts it to acetyl CoA.
- D) It requires NADPH to be converted to lactate.
- E) It cannot be converted back to fatty acids.



39) Which of the following statements about protein metabolism is **incorrect**?

- A) Amino acids are required for the synthesis of other nitrogen compounds.
- B) Protein degradation to provide energy does not result in loss of other functions.
- C) Essential amino acids must be obtained in the diet daily.
- D) Nitrogen is continuously lost from the body regardless of the protein intake amount.
- E) Proteins of animal origin typically have the highest biological value.

- 40) Which of the following enzymes is **not** used in the small intestine for dietary protein breakdown?
- A) Pepsin
  - B) Carboxypeptidase A
  - C) Elastase
  - D) Chymotrypsin
  - E) Trypsin
- 41) Which of the following statements **correctly** describes endogenous protein degradation?
- A) The lysosomal pathway primarily degrades proteins from endocytosis.
  - B) The attachment of ubiquitin to proteins requires UTP.
  - C) The proteasome degrades proteins within a lysosome.
  - D) Only one molecule of ubiquitin is attached to a protein to be degraded.
  - E) The lysosomal pathway requires ATP.
- 42) Which of the following statements regarding aminotransferases (a.k.a. transaminases) is **correct**?
- A) High serum levels of alanine aminotransferase (ALT) only indicate liver damage.
  - B) Aminotransferase reactions are irreversible.
  - C) Aminotransferases require pyridoxal phosphate, a co-factor derived from vitamin B<sub>6</sub>.
  - D) The co-factor of the aminotransferase is attached to the enzyme via an amide bond.
  - E) Glutamine is the most common product of aminotransferase reactions.
- 43) Which of the following enzymes are capable of fixing free ammonia (NH<sub>4</sub><sup>+</sup>)?
- A) amino acid oxidases, glutaminase, glutamate dehydrogenase
  - B) carbamoyl phosphate synthetase I (CPS I), glutaminase, glutamine synthetase
  - C) glutaminase, aminotransferases, amino acid oxidases
  - D) glutamine synthetase, carbamoyl phosphate synthetase I (CPS I), glutamate dehydrogenase
  - E) glutamate dehydrogenase, amino acid oxidases, aminotransferases
- 44) Which of the following statements about glutamate dehydrogenase is **incorrect**?
- A) In the liver, high levels of ATP inhibit glutamate dehydrogenase.
  - B) In the liver, the formation of glutamate is favored so that the NH<sub>4</sub><sup>+</sup> released goes to the urea cycle.
  - C) Glutamate dehydrogenase is capable of using NAD<sup>+</sup>/NADH and NADP<sup>+</sup>/NADPH.
  - D) Glutamate dehydrogenase is a mitochondrial enzyme.
  - E) In peripheral tissues, glutamate dehydrogenase fixes free ammonia to form glutamate.
- 45) The major purpose of the “glucose-alanine cycle” between the liver and muscle tissues allows for:
- A) The alanine taken up by the muscle to be converted back to glucose.
  - B) The liver to store the alanine as fat.
  - C) The muscle to deliver its excess nitrogen to the liver for excretion, and ultimately get back glucose, from the liver, to continue doing glycolysis.
  - D) The liver to release alanine into the blood for other tissues to use, and get back glucose so it can store it as fat.
  - E) The liver to deliver its excess nitrogen for the muscles to use in protein synthesis, and get glucose from the muscle in return.

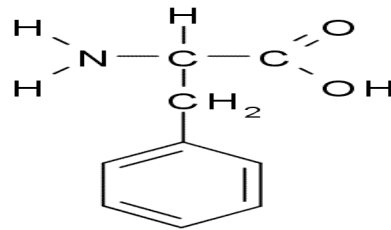
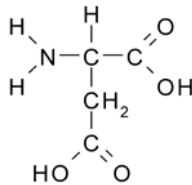
46) The urea cycle requires all of the following **except**:

- A) aspartate
- B) carbamoyl phosphate synthetase I
- C) ATP
- D) citrulline/ornithine transporter
- E) isocitrate

47) "Mixed amino acids":

- A) when degraded produce products that can be used for both glucose and ketone body synthesis.
- B) when degraded produce acetyl CoA, which can be used for glucose synthesis.
- C) are amino acids that can be used to make other amino acids.
- D) include only leucine and lysine.

48) You have just learned that the artificial sweetener aspartame can be hydrolyzed to form the following products:



People afflicted with \_\_\_\_\_ should avoid this sweetener.

- A) anemia
- B) phenylketonuria
- C) diabetes
- D) Lesch-Nyhan syndrome

49) A tissue's response to insulin includes all of the following **except**:

- A) Activation of the tyrosine kinase domain of the insulin receptor
- B) Activation of a phosphatase
- C) phosphorylation of target proteins
- D) Activation of family of proteins called insulin receptor substrates (IRS)
- E) Activation, as a general rule, of target proteins

50) Which of the following statements about glucagon and epinephrine is **correct**?

- A) The release of glucagon and epinephrine both involve a response to low blood sugar.
- B) Muscle is a target tissue for both glucagon and epinephrine.
- C) Epinephrine is derived from threonine.
- D) Both glucagon and epinephrine bind to tyrosine kinase receptors.
- E) Epinephrine stimulates glucose uptake from the blood in muscle tissue.

51) Which of the following enzymes does **not** follow the "general rule," and thus is phosphorylated and active in response to glucagon?

- A) HMG-CoA reductase
- B) glycogen phosphorylase
- C) pyruvate kinase
- D) acetyl CoA carboxylase
- E) pyruvate dehydrogenase complex

52) Which of the following statements regarding the regulation of beta-oxidation of fatty acids is **incorrect**?

- A) Beta-hydroxyacyl CoA dehydrogenase is inhibited by high NADH concentrations.
- B) Malonyl CoA is an inhibitor of beta-oxidation.
- C) Leptin stimulates beta-oxidation.
- D) Carnitine acyl transferase I (CAT I) is active.
- E) Insulin stimulates beta-oxidation of fatty acids.

53) Which of the following situations would result in an increase in ketone body synthesis?

- A) High insulin/glucagon ratio
- B) High citrate levels in the cytosol
- C) High levels of fructose-2,6-bisphosphate
- D) Low oxaloacetate levels in the mitochondria
- E) Low acetyl CoA levels in the mitochondria

54) Which of the following correctly lists, in order, the enzymes that carry out the “4 repeated steps” of fatty acid synthesis?

- A) acyl CoA dehydrogenase, enoyl CoA hydratase, beta-hydroxyacyl CoA dehydrogenase, beta-ketothiolase
- B) beta-ketoacyl ACP synthase, beta-ketoacyl ACP reductase, beta-hydroxyacyl ACP dehydratase, enoyl ACP reductase
- C) beta-ketoacyl ACP synthase, beta-ketoacyl ACP dehydrogenase, beta-hydroxyacyl ACP hydratase, enoyl ACP dehydrogenase
- D) enoyl ACP reductase, beta-hydroxyacyl ACP dehydratase, beta-ketoacyl ACP reductase, beta-ketoacyl ACP synthase
- E) beta-ketoacyl ACP synthase, beta-ketoacyl ACP reductase, beta-hydroxyacyl ACP hydratase, enoyl ACP reductase

55) Which of the following statements about the Fatty Acid Synthase complex is **incorrect**?

- A) The fatty acid synthase complex uses ATP.
- B) It is a large multi-enzyme complex.
- C) Phosphopantetheine is the cofactor of the acyl carrier protein (ACP).
- D) The removal of the carboxylic acid group from malonyl CoA drives the condensation step.
- E) The major product of the complex is palmitate (C<sub>16:0</sub>).

56) Which of the following statements regarding phosphatidate is **correct**?

- A) Phosphatidate can be used for both triacylglycerol (TAG) and phosphosphingolipid synthesis.
- B) Phosphatidate is also called ceramide.
- C) The third fatty acyl group is attached to the phosphate group of phosphatidate to form a triacylglycerol (TAG).
- D) CTP is used to activate phosphatidate to form CDP-diacylglycerol in *de novo* synthesis of phospholipids.
- E) Phosphatidate is formed by attachment of 2 fatty acyl groups to dihydroxyacetone phosphate (DHAP).

The next 4 questions refer to the case described below.

Ms. M., a 19-year old white female checks into the emergency room complaining of lethargy, vomiting, and polyuria of one-day duration. Her temperature was 97°F, blood pressure 100/50, pulse rate 120/min, respiration 40/min. The physical exam revealed Kussmaul respirations, severe dehydration, and an elevated pulse rate. Her laboratory data at admission and the normal values are shown below:

<u>Test</u>	<u>Admission</u>	<u>Normal Values</u>
pH	7.2	7.35 – 7.45
Total CO <sub>2</sub> content	6	26 – 27
pCO <sub>2</sub> (mm Hg)	8.6	35 – 45
pO <sub>2</sub> (mm Hg)	72	95 – 100
bicarbonate (mM)	4	25 – 26
sodium (meq/L)	138	136 – 145
potassium (meq/L)	5.0	3.5 – 4.5
chloride (meq/L)	102	100 – 106
fasting blood glucose (mg/dL)	300	60-100
blood urea nitrogen (mg/dL)	18	10 – 20

**Additional observations of patient's urine:** Urine was dark and contained elevated levels of sugar, acetoacetate and acetone.

(a) pK<sub>a</sub> of bicarbonate-CO<sub>2</sub> buffer, 6.1; (b) solubility coefficient for CO<sub>2</sub> at 37°C, 0.03 mM/mmHg.

57) Based on Ms. M.'s admission data, what is most likely the cause of the depletion of bicarbonate?

- A) hemoglobin
- B) glucose
- C) ketone bodies
- D) bilirubin
- E) insulin

58) Which of the following statements best explains why she has elevated glucose and ketone bodies in the urine?

- A) Ms. M. has defective glucose-6-phosphatase.
- B) Ms. M. has insulin-dependent type 1 diabetes.
- C) Ms. M. has a carnitine deficiency.
- D) Ms. M. has defective pyruvate kinase.
- E) Ms. M. has defective HMG-CoA reductase.

59) In Ms. M.'s case, at the time of her admission which of the following statements regarding her liver enzymes' phosphorylation states and activity is **incorrect**?

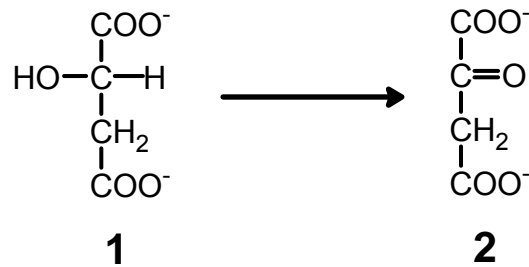
- A) The pyruvate dehydrogenase complex is phosphorylated and inactive.
- B) Glycogen phosphorylase is phosphorylated and active.
- C) Glycogen synthase is phosphorylated and inactive.
- D) Pyruvate kinase is phosphorylated and inactive.
- E) Fructose 2,6-bisphosphatase (FBP2) is phosphorylated and inactive.

60) In comparing and contrasting starvation and type 1 diabetic conditions, which of the following statements is **incorrect**?

- A) In both situations, glucagon is the hormone that is predominating.
- B) Diabetics will experience ketoacidosis, but a starving person will not.
- C) Both conditions will result in an increase in HbA<sub>1C</sub>.
- D) Both conditions will result in mobilization of fatty acids from TAG stores.
- E) Both conditions will result in protein degradation.

61) In a person with a deficiency of fructose 1,6-bisphosphatase, the predominant metabolic consequence is:

- A) failure to resynthesize glucose from lactic acid
- B) failure to split fructose bisphosphate into triose phosphates
- C) inability to degrade glycogen
- D) inability to fix CO<sub>2</sub> into organic linkages
- E) lowered yield of ATP production per mole of glucose metabolized



62) In the reaction drawn above, what has happened to molecule 1 to convert it to molecule 2?

- A) Isomerization
- B) Dehydration
- C) Reduction
- D) Phosphorylation
- E) Oxidation

63) What enzyme would carry out this reaction?

- A) Citrate synthase
- B) Malate dehydrogenase
- C) Pyruvate carboxylase
- D) Oxaloacetate reductase
- E) Pyruvate dehydrogenase

64) There are several known diseases that are caused by genetic abnormalities in the carnitine system. The clinical symptoms can range from mild, recurrent muscle cramping to severe weakness and death. Carnitine therapy has proved effective in some cases, as has the replacement of normal dietary fat by triacylglycerols containing medium chain length fatty acids. These diseases have become treatable disorders because medical students have learned the biochemistry of:

- A)  $\beta$ -oxidation of fatty acids
- B) glycolysis
- C) methylation reactions by S-adenosylmethionine
- D) pyrimidine nucleotide synthesis
- E) urea cycle

END OF EXAMINATION

Tear off this sheet and save to check your answers.

**Please remember to:**

- Write in your **form letter** in the appropriate place on the **answer sheet**.
- SIGN AND RETURN YOUR EXAMINATION** to an instructor **before leaving the exam room**.

FORM: A

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| 1. _____  | 21. _____ | 41. _____ | 61. _____ |
| 2. _____  | 22. _____ | 42. _____ | 62. _____ |
| 3. _____  | 23. _____ | 43. _____ | 63. _____ |
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