Exam Procedures:

STEP 1 - NAME (*Print clearly*) _____ _____
(first) *(last)*

STEP 2 – Fill in your answer sheet with a #2 scoring pencil, as follows:

- ☑ Your Student PID Number (excluding "A")
- ☑ Your last name and first name
- ☑ Course ID in "subject" …… **this is BMB 514 Exam #2**
- ☑ Date ……**10/07/13**
- ☑ Exam form in "period" …..**this is form A**
- ☑ By signing this coversheet for this exam, the student certifies that he/she has adhered to the policies of academic honesty in the performance of this exam.

_____
Signature

STEP 3 - Read these instructions:

- ☑ Page 2 of this exam contains information that may be useful to you: (a) abbreviations for the amino acids; (b) pKa values of functional groups; and (c) table of logarithms.
- ☑ Make sure your exam has **32** questions.
- ☑ Read each question very carefully. Choose the single, best answer and mark this answer on your answer sheet. No points will be added for correct answers which appear on the exam page but not on the answer sheet.
- ☑ The proctors have the authority/responsibility to assign any student a different seat at any time, without implication and without explanation, before or during the examination, as they deem necessary. Accomplish any relocation quietly and without discussion.
- ☑ We will not answer questions of clarification. However, if you think there is an error on your exam, summon an exam proctor.
- ☑ When you finish, place all exam materials (except the tear sheet) into the manila envelope. When you leave the exam room, please turn in your envelope to the proctors. Once you exit the auditorium, please leave the area. Hallway conversations disturb those still taking the exam.
- ☑ There will be answer keys to this exam posted on the course website by 5:00 p.m. the day of the exam. You may wish to copy your responses from your answer sheet onto the answer grid on the LAST page of this exam so that you can check your results. You can tear off the last page and take it with you.
- ☑ We will close the exam promptly at **9:10 a.m.** At the announcement of the examination end time, the examination and scantron and images (if provided as part of the examination) must immediately be placed into the manila envelope provided.

STEP 4 – Wait until instructed to proceed with the exam!

INFORMATION THAT MAY BE USEFUL FOR THE EXAM

Abbreviations for Amino Acids			
Amino Acid	3-Letter Abbreviation	Amino Acid	3-Letter Abbreviation
Alanine	Ala	Leucine	Leu
Arginine	Arg	Lysine	Lys
Asparagine	Asn	Methionine	Met
Aspartic Acid	Asp	Phenylalanine	Phe
Cysteine	Cys	Proline	Pro
Glutamine	Gln	Serine	Ser
Glutamic Acid	Glu	Threonine	Thr
Glycine	Gly	Tryptophan	Trp
Histidine	His	Tyrosine	Tyr
Isoleucine	Ile	Valine	Val

Ionizable Group	pKa
α-COOH of any aa	2
β-COOH of Asp	4
γ-COOH of Glu	4
imidazole of His	6
SH of Cys	8
α-NH_2 of any aa	9
phenolic OH of Tyr	10
ϵ-NH_2 of Lys	10
guanidino of Arg	12

Tables of Logarithmic Relationships

Number	Decimal									
	.0	.1	.2	.3	.4	.5	.6	.7	.8	.9
1.	.00	.04	.08	.11	.15	.18	.20	.23	.26	.28
2.	.30	.32	.34	.36	.38	.40	.41	.43	.45	.46
3.	.48	.49	.51	.52	.53	.54	.56	.57	.58	.59
4.	.60	.61	.62	.63	.64	.65	.66	.67	.68	.69
5.	.70	.71	.72	.72	.73	.74	.75	.76	.76	.77
6.	.78	.79	.79	.80	.81	.81	.82	.83	.83	.84
7.	.85	.85	.86	.86	.87	.88	.88	.89	.89	.90
8.	.90	.91	.91	.92	.92	.93	.93	.94	.94	.95
9.	.95	.96	.96	.97	.97	.98	.98	.99	.99	1.00
10.	1.00					logs				

E.g. log 3.5 = 0.54

Reminder: How to form logs of multiples .....

log 35 = log (3.5×10^1) = (log 3.5 + log 10^1) = (0.54 + 1) = 1.54

log 350 = log (3.5×10^2) = (log 3.5 + log 10^2) = (0.54 + 2) = 2.54

log 0.35 = log (3.5×10^{-1}) = (log 3.5 + log 10^{-1})= (0.54 - 1) = - 0.46

The following three questions deal with the following case:

Prior to beginning medical school, a student received a clean bill of health during his routine yearly physical, including lab reports showing all tests within normal range. The stress of his medical school schedule is beginning to set in and because of his upcoming medical school biochemistry 514 exam, the student has not eaten in 36 hours.

1) Following completion of his BMB 514 exam, he decides to have a couple of drinks to celebrate. Prior to consuming enough alcohol to become truly inebriated, the student begins to shake, sweat, and display signs of being confused. You hypothesize that your classmate is suffering from alcohol-induced hypoglycemia. Which of the following best explains why alcohol would induce hypoglycemia under these conditions?

   A. Alcohol metabolism produces NADH that inhibits gluconeogenesis
   B. Alcohol metabolism produces acetyl-CoA that inhibits pyruvate dehydrogenase
   C. Alcohol allosterically inhibits pyruvate carboxylase
   D. Alcohol metabolism inhibits the production of fructose 2,6-bisphosphate
   E. Alcohol metabolism produces citrate that inhibits glucose 6-phosphatase

2) Approximately one hour after consuming a meal consisting of pasta, meat loaf and an ice cream sundae, which of the following pathways would most likely be active in the patient's liver.

   A. Ketone body synthesis
   B. β-oxidation
   C. Gluconeogenesis
   D. Glycogenolysis
   E. Cholesterol synthesis

3) If you were able to analyze his blood following the completion of his meal of pasta, meat loaf, and an ice cream sundae, which of the following would you expect to find?

   A. High glucagon levels
   B. Free fatty acids
   C. Chylomicrons
   D. Ketone bodies
   E. Hyperglycemia

4) Highly metabolic cells that are not dividing (i.e. no nucleic acids are needed) require high concentrations of NADPH for synthetic reactions. Which of the following contains a pair of enzymes that are both required for the production of NADPH without also producing excess ribose 5-phosphate?

   A. Phosphoglucose isomerase and glucose 6-phosphate dehydrogenase
   B. Pyruvate kinase and phosphoglycerate kinase
   C. Glucose 6-phosphate dehydrogenase and phosphoglycerate kinase
   D. Glucose 6-phosphate dehydrogenase and glyceraldehyde 3-phosphate dehydrogenase
   E. Tranketolase and phosphoglycerate kinase

5) Coenzyme Q is best described by which of the following?

   A. Is only capable of carrying protons
   B. It feeds electrons directly to complex IV
   C. HMG-CoA reductase can impact its synthesis
   D. Is a water soluble, electron transport chain intermediate
   E. Transfers electrons from complex I to Complex II

6) A patient (7 years old) is brought into the clinic suffering from ataxia. Upon serum analysis the only abnormalities that were observed were elevated lactate, alanine, and pyruvate. The patient was partially stabilized by the treatment of thiamine. What do you think is wrong with the patient?

A. Pyruvate kinase deficiency
B. Lactate dehydrogenase deficiency
C. Pyruvate carboxylase deficiency
D. Pyruvate dehydrogenase deficiency
E. Phosphofructokinase deficiency

7) A patient is brought into the emergency room near death. You have determined that he has been exposed to a mitochondrial poison. His life depends on determining to what poison he has been exposed. You quickly determine that his mitochondria are not making ATP nor utilizing O_2. Addition of 2,4-DNP does not restore oxygen consumption and there appears to be higher than normal concentrations of reduced cytochrome c. What poison most likely caused the patient's conditions?

A. Amytal
B. Rotenone
C. Carbon monoxide
D. Antimycin
E. Oligomycin

8) Classic galactosemia is caused by a defect in galactose 1-phosphate uridyltransferase. Which of the following describes the impact of a deficiency in galactose 1-phosphate uridyltransferase?

A. Inability to metabolize table sugar
B. Build up of glucose 6-phosphate in the cells
C. Tissue damage caused by phosphate sequestration
D. Build up of UDP-galactose
E. Tissue damage caused by inability to release glycogen stores

9) During starvation, ketone bodies are produced. Which of the following correctly describes ketone bodies or their metabolism?

A. There are two ketone bodies found in humans, 3-hydroxybutyrate and acetone
B. Ketone bodies are used by the liver for energy during times of starvation
C. Ketone synthesis shares an intermediate with cholesterol biosynthesis
D. Ketone bodies must be bound to protein to safely move through circulation
E. Ketone body synthesis can be induced by a high carbohydrate diet

The following 3 questions deal with the following case study

A 38 year old woman comes into the clinic complaining of feeling tired and physically "drained" for the last 5 days. She has yellow sclerae and has informed you that her urine is very dark. She has made no foreign trips and mentions the only thing she did different prior to this bout of fatigue, is she consumed two meals of broad beans, including fava beans. In both meals, she estimates she consumed approximately one pound of beans. Her lab reports are detailed in the chart below:

TEST	RESULT	Normal
Hemaglobin	55 g/l	115-151 g/l
Reticulocyte count	2.1%	0.5 - 1.5%
Vitamin B12	475 ng/l	125-600 ng/l
RBC Folate	597 ug/l	215-650 ug/l
Bilirubin	48 umol/l	<17 umol/l
Lactate dehydrogenase	2197 U/l	50-450 U/l
Glucose 6-phosphate dehydrogenase	7.2 U/g Hb	4.6-13.5 U/g Hb

10) Given the relationship between the appearance of symptoms and the two meals of fava beans, you suspect she is deficient in erythrocyte glucose 6-phosphate dehydrogenase (G6PDH) activity. What role does G6PDH play in erythrocytes that render them sensitive to oxidant-induced lysis in G6PDH deficient patients?

A. The NADPH it produces is necessary to maintain reduced glutathione.
B. The lactone it produces is a necessary building block for cell wall maintenance
C. The NADPH it produces is used to synthesize the steroids necessary for cell wall biosynthesis
D. The lactone it produces can be used to inhibit phospholipases
E. The NADPH it produces is necessary for salvage of partially degraded phospholipids

11) According to the laboratory data, the patient is suffering from anemia (i.e decreased hemoglobin). Given the patient's decreased oxygen carrying capacity, her muscle cells might become anoxic, even under non-strenuous activity. Under these conditions (i.e. anoxic muscle cells) what would you expect the state of glycogen metabolism to be in her muscle cells?

A. High levels of glucose 6-phosphate would stimulate glycogen phosphorylase
B. High levels of Ca^{2+} would stimulate glycogen synthase
C. Low levels of ATP would activate glycogen synthase
D. High levels of AMP would activate glycogen phosphorylase
E. High levels glucose 1-phosphate would inhibit glycogen synthase

12) If a muscle biopsy could be performed under these anoxic conditions, what would you expect to find, in terms of lactate production and electron transport chain (ETC) activity compared to a muscle cell that is under normoxic (Normal O_2 levels) conditions ?

	Lactate production	ETC activity
A.	Increased	Increased
B.	Increased	Decreased
C.	Decreased	Increased
D.	Normal	Normal
E.	Decreased	Decreased

The following three questions deal with the following case:

A three year old boy is brought into the clinic suffering from cardiorespiratory distress following an infection. This is the fifth time the boy was admitted under similar circumstances. He had a brother who died of an undiagnosed liver problem after falling into a coma. His other two siblings are well. Upon admission, the patient's lab reports were:

TEST	RESULT	Normal
Serum Glucose	15 mg/dl	60-100 mg/dl
alanine aminotransferase (ALT)	179 I.U./l	10-45 I.U./l
aspatate aminotransferase (AST)	337 I.U./l	6-36 I.U./l
pH	7.37	7.35-7.44
serum ketone levels		< 1 mg/dl

Previously the boy was admitted to the hospital in a coma and suffered a cardiac arrest. He was extremely hypoglycemic (plasma glucose = 15 mg/dl) and showed hepatomegaly. Electrocardiogram, brain scan and chromosomal analysis all proved unremarkable. Muscle and liver biopsies contained large amounts of neutral lipids. A carnitine deficiency was suspected and a 32 hour fasting study was performed with the following result:

Fasting test	At start	24 hrs later	Normal
Glucose	91 mg/dl	66 mg/dl	60-100 mg/dl
Triglycerides	66 mg/dl	122 mg/dl	< 150 mg/dl

13) Loss of carnitine will most directly affect which of the following processes?

A. Fatty acid translocation into the matrix of the mitochondria
B. Fatty acid packaging into chylomicrons
C. Transport of free fatty acids from adipocytes to the liver
D. Salvage pathway for phospholipids
E. Phospholipase induced release of free fatty acids

14) On the way up to the floor, someone spilled coffee on the lab report so that you are unable to determine if the patient is undergoing ketosis. Being the good biochemists, you can deduce what it will say. Is the patient positive for serum ketone levels, and why or why not?

A. The ketone levels will be high to meet energy demands since glucose is low
B. The ketone levels will be high because of the high levels of fatty acids in the liver will be converted to ketones.
C. The ketone levels will be low because fatty acids cannot get into the mitochondria to be converted to ketones.
D. The ketone levels will be low because the hypoglycemia will decrease the energy needed for their synthesis.
E. The ketone levels will be low because the triglycerides will continue to build up and inhibit ketone synthesis.

15) Why is the patient unable to maintain glucose levels during the 32 hour fast and ultimately became hypoglycemic?

A. The high circulating triglycerides inhibit glucose 6-phosphatase
B. Gluconeogenesis is inhibited by the lack of energy caused by the inability to use fatty acids for ATP production.
C. The fat deposits in the liver will inhibit lactate dehydrogenase
D. The borderline acidemia will inhibit the conversion of malate to oxaloacetate in the mitochondria
E. Pyruvate carboxylase will be inhibited by the high levels of acetyl CoA in the matrix

16) Which of the following correctly describes the regulation of β-oxidation?

A. During starvation conditions β-ketothiolase will be inactivated
B. High levels of NADH will activate β-oxidation
C. Carnitine acyl transferase I (CATI) is positively regulated by malonyl CoA
D. Acetyl CoA will activate β-oxidation through stimulation of β-ketothiolase
E. Glucagon and epinephrine will stimulate β-oxidation through lipase activation

17) Surfactants use dipalmitoylphosphatidyl choline (DPPC) as the primary phospholipid. Which of the following correctly describes the de novo synthesis of DPPC?

A. It is an energy independent process
B. It takes place entirely in the cytosol
C. It uses phosphatidic acid as a intermediate
D. It requires the removal of the phosphate group to complete the synthesis
E. It can be inhibited by statins

18) The glycolipids are

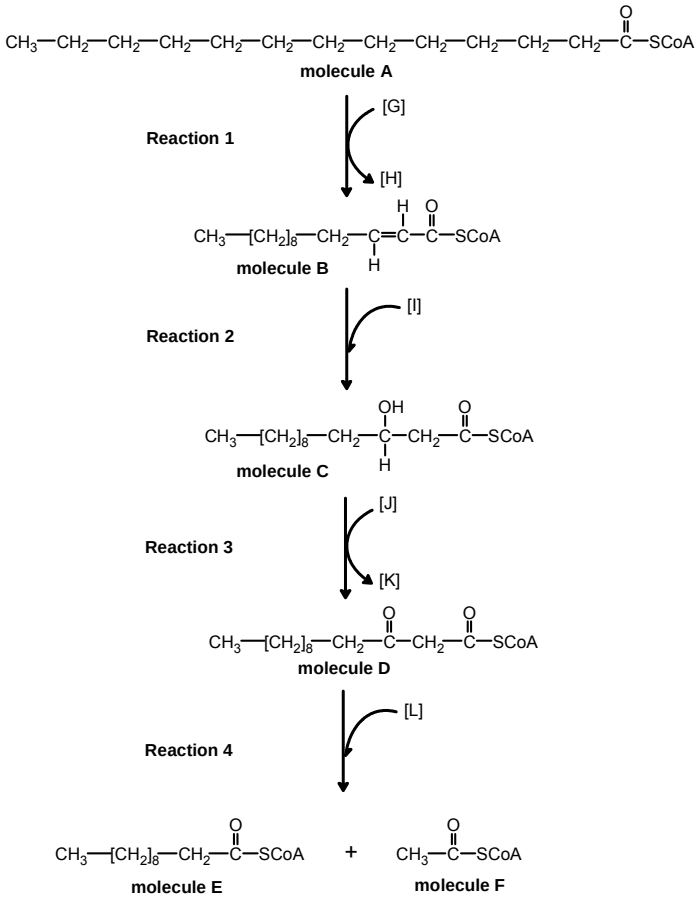
A. lipids which function mainly as a storage form of energy.
B. membrane lipids containing a phosphate-alcohol headgroup.
C. lipids that are synthesized using phosphatidate as the key intermediate of the pathway.
D. degraded to form diacylglycerol as a second messenger.
E. lipids in which the backbone is initially synthesized from palmitoyl CoA and serine.

19) You decide to try the Atkins diet. During the first stage of this diet, you are not allowed to eat any carbohydrates. Which of the following pathways would increase in your liver under these conditions?

A. Glycolysis and fatty acid synthesis
B. ketone body synthesis and glycolysis
C. gluconeogenesis and fatty acid synthesis
D. glycolysis and beta-oxidation
E. β-oxidation, ketone body synthesis, and gluconeogenesis

20) The pyruvate dehydrogenase (PDH) complex is tightly regulated by hormones and cellular metabolites. Which of the following are responsible for **ACTIVATING** the kinase involved in regulating the PDH complex?

A. Acetyl CoA and Insulin
B. ADP and glucagon
C. Acetyl CoA and pyruvate
D. NADH and Insulin
E. Acetyl CoA and NADH



The following four questions refer to the pathway drawn above.

21) Which of the following statements describing the above pathway is correct?

A. The enzymes that carry out all 4 reactions are organized into a large complex.
B. Reaction #1 can be inhibited by statins.
C. The 4 reactions, respectively, are dehydrogenation, hydration, dehydrogenation, and cleavage.
D. Citrate is the carrier of molecule A into the mitochondrial matrix.
E. The pathway oxidizes NADPH to $NADP^+$ in reactions 1 and 3.

22) Repeating reactions 1 through 4 will ultimately yield:

A. 8 acetyl CoA, 7 $FADH_2$, 7 ($NADH + H^+$)
B. 1 propionyl CoA, 6 acetyl CoA, 6 $FADH_2$, 6 ($NADH + H^+$)
C. 6 acetyl CoA, 6 $FADH_2$, 6 ($NADH + H^+$)
D. 1 propionyl CoA, 5 acetyl CoA, 5 $FADH_2$, 5 ($NADH + H^+$)
E. Palmitate

23) Deficiencies in one of the enzymes that catalyzes reaction #1 will lead to which of the following phenotypes?

A. Starvation induced hypoglycemia
B. Inability to synthesize phospholipids
C. Overproduction of ketone bodies
D. Decreased triacylglyceride synthesis due to decreased substrates
E. Normal since there are a total of four enzymes that perform this reaction

24) Which of the molecules is a negative regulator of reaction #4?

A. Molecule C
B. Molecule D
C. Molecule E
D. Molecule F
E. Molecule L

25) Which of the following statements about bile acids and bile salts is **CORRECT**?

A. They function as detergents because they are amphipathic.
B. Conjugation of the bile salts with taurine or glycine produces bile acids.
C. The addition of taurine or glycine makes these molecules more insoluble in water.
D. Bile acids and bile salts are synthesized in the gall bladder.
E. The majority of bile acids and bile salts are excreted from the body daily.

26) Which of the following correctly describes the regulation of cholesterol metabolism?

A. HMG-CoA reductase can be inhibited by insulin
B. Cholesterol can positively regulate its own synthesis via inhibition of HMG-CoA reductase transcription
C. Cholic acid can activate HMG-CoA via inhibition of protease responsible for HMG-CoA reductase proteolysis
D. Cholesterol can activate LDL receptor activity, thus increasing cholesterol uptake into cells
E. Cholesterol allosterically inhibits HMG-CoA reductase via product inhibition

27) Following consumption of a high calorie meal, which of the following correctly describes the state of the malate/citrate/pyruvate shuttle?

A. Acetyl-CoA will be moving from the cytosol to the mitochondria
B. It uses malate dehydrogenase, the enzyme involved in the TCA cycle and gluconeogenesis
C. It uses a isozyme of citrate synthase to produce acetyl Co in the cytosol
D. Acetyl CoA is cotransported with malate across the inner mitochondrial membrane
E. The process produces large amounts of NADH in the cytosol

28) Which of the following correctly describes lecithin:cholesterol acyltransferase (LCAT) function?

A. Involved in the synthesis of phospholipids
B. Is responsible for cholesterol esterase synthesis in LDLs
C. Can only use phosphotidylcholine as an acyl substrate
D. Makes cholesterol more water soluble
E. Involved in the mitochondrial translocation of fatty acids

29) Which of the following statements about high density lipoproteins (HDLs) is correct?

A. HDLs transport dietary triacylglycerols (TAG).
B. HDLs are the largest of the lipoprotein particles.
C. HDLs are synthesized by the intestinal mucosal cells.
D. HDLs can transport cholesterol from the tissues to the liver.
E. HDLs cannot be endocytosed.

30) Which of the following correctly describes the process of fatty acid synthesis

A. Uses malonyl CoA as a source of new carbon units being added to the growing fatty acid
B. Is an energy independent process
C. Occurs in the matrix, using the same four enzymes involved in β-oxidation
D. Will be activated when glucagon levels are high
E. Double bonds can be introduced at any carbon in the chain

31) A 3 day old male infant has poor feeding, decreased muscle tone, and several seizures per day. Significant laboratory investigations include elevated levels of lactate in the cerebrospinal fluid, massive elevations of fumarate, and decreased levels of malate. You suspect he MOST likely has a deficiency of:

A. citrate synthase
B. succinate thiokinase
C. pyruvate dehydrogenase
D. fumarase
E. α-ketoglutarate dehydrogenase

32) Which statement BEST describes the clinical course of Leigh disease?

A. Infants are born with feeding difficulties and poor muscle tone, but gradually improve
B. Affected infants can be expected to have manifestations at birth
C. Age of onset is variable, and manifestations get worse over time
D. Manifestations are usually severe, but can be effectively treated
E. Muscle and eye are generally always affected, but brain is spared

BMB 514 SS 2013 Exam #2
FORM: A

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