1) A person has just eaten a large meal, complete with baked potatoes and buttered pasta. Which of the following enzymes is **NOT** necessary to get all of the carbohydrates found in this meal into portal circulation?

A) Amylase
B) Na^+/K^+ ATPase
C) Sucrase
D) Lactase
E) Fructokinase

2) A person is suffering from a mutation in a key enzyme involved in carbohydrate metabolism. You run a battery of tests and determine the patient is capable of processing glucose all the way to pyruvate at normal rates under standard conditions. During times of anaerobic stress, the patient has a decreased glycolytic capacity. Which of the following mutated enzymes is matched with the metabolite most likely to be found at abnormally high levels in this patient under anaerobic conditions?

	Mutated enzyme	**Metabolite at high concentration**
A)	Glyceraldehyde 3-P dehydrogenase	pyruvate
B)	Lactate dehydrogenase	lactate
C)	Glyceraldehyde 3-P dehydrogenase	dihydroxyacetone phosphate
D)	Lactate dehydrogenase	glyceraldehyde 3-phosphate
E)	Phosphofructokinase	pyruvate

3) A deficiency in which enzyme(s) can lead to hemolytic anemia?

A) Glucose 6-phosphate dehydrogenase
B) Phosphoenolpyruvate carboxykinase
C) Pyruvate kinase
D) Both A and C
E) None of the above

4) Wernicke-Korsakoff syndrome is characterized by psychosis and encephalopathy and is caused by severe thiamine deficiency. Which of the following enzymes would **not** be affected in a patient suffering from Wernicke-Korsakoff syndrome?

A) Pyruvate dehydrogenase
B) Transketolase
C) α-ketoglutarate dehydrogenase
D) Isocitrate dehydrogenase

5) Vitamin deficiencies may affect many metabolic pathways due to the requirement of cofactors in enzymatic reactions. The Pentose Phosphate Pathway (PPP) can be run in one of four modes, depending on the specific purpose or product(s) needed (listed below). Which of these four modes of the PPP would **NOT** be affected by thiamine deficiency?

Mode #1: NADPH + Ribose-5-P
Mode #2: NADPH only
Mode #3: NADPH + energy
Mode #4: Ribose 5-P only

A) Mode 1
B) Mode 2
C) Mode 3
D) Mode 4
E) All modes

6) An infant is suffering lethargy, vomiting, diarrhea, failure to thrive, and jaundice. Following newborn screening, the infant is diagnosed with galactosemia. Which of the following is **NOT** an enzyme that might be compromised in this patient?
   A) Galactokinase
   B) Phosphoglucomutase
   C) Galactose 1-phosphate uridyltransferase
   D) UDP-galactose epimerase (4-epimerase)

7) The local control of glycogen phosphorylase involves which of the following?
   A) Phosphorylation
   B) Glucose
   C) Biological amplification
   D) Epinephrine
   E) AMP-induced activation

8) Following a 12-hour fast, all of the following are true regarding glycogen release, **EXCEPT**:

   A) Glycogen phosphorylase will be unphosphorylated
   B) Glycogen synthase will be phosphorylated
   C) Glucagon levels will be high
   D) Adenylate cyclase will be active
   E) cAMP levels will be elevated

9) A patient with Von Gierke's disease has lipemia retinalis (milky appearance of veins and arteries of the retina) and xanthoma tuberosum disseminata (lipid filled yellowish nodules especially over elbows and knees). What is the most likely explanation for these clinical symptoms of Von Gierke's disease?

   A) Excess glycogen is being converted directly to fat
   B) Fatty acids are being released as a result of high citrate synthase activity
   C) Low circulating glucose is inhibiting the uptake of lipids into peripheral tissues
   D) Excess lipids are being released in an attempt to address energy debt caused by low blood glucose levels
   E) The excretion of lipids has been compromised by lactic acid

10) You are trying to create an artificial mitochondria with a new category II prosthetic group to replace cytochrome c. You have named this new prosthetic group substance X. Given the standard redox potentials of the two half reaction:

$$\text{Substance } X^+ + 2H^+ + 2e^- \Longleftrightarrow \text{Substance X} \qquad E_o' = 0.25$$
$$\tfrac{1}{2}O_2 + 2H^+ + 2e^- \Longleftrightarrow H_2O \qquad E_o' = 0.75$$

What is the standard free energy change of the following reaction?

$$\text{Substance X} + \tfrac{1}{2}O_2 \Longleftrightarrow \text{Substance } X^+ + H_2O$$

(Given $\Delta G^{o'} = - n \mathcal{F} \Delta E_o'$ and $\mathcal{F}$= 23 kcal/volt mol e^-)

   A) - 46 kcal/mol
   B) – 23 kcal/mol
   C) + 46 kcal/mol
   D) + 23 kcal/mol
   E) -11.5 kcal/mol

11) Which of the following correctly describes a functioning mitochondrion?

   A) Will use oxygen at a constant rate regardless of ADP levels.
   B) Will have the same P/O ratio for NADH and $FADH_2$
   C) When thermogenin (aka UCP1) is activated, ATP generation will decrease
   D) In the presence of oligomycin, the membrane potential will drop
   E) In the presence of 2,4 dinitrophenol oxygen consumption will decrease

Questions 12 through 14 apply to the following clinical case:

A person is brought into the emergency room suffering from incoordination, impaired reflexes, respiratory distress and central nervous system depression. You quickly conclude that the patient has been poisoned. Working with the local police you learn that the patient's wife is a noted biochemist. You suspect that she used her expertise to deduce she could get rid of her husband by drastically increasing his dose of Lipitor, a cholesterol lowering drug. You realize that this would lead to an almost complete loss of CoQ (also known as CoQ_{10} and ubiquinol) and ultimately lead to complete mitochondrial dysfunction.

12) Given what you know about the electron transport chain, the loss of CoQ would limit the ability of which complex or complexes to donate their reducing equivalents to the electron transport chain?

A) Complex I
B) Complex II
C) Complex III
D) A and B
E) All of the above

13) If you were able to isolate the mitochondria of the poisoned patient and measure their oxygen consumption rates before (compared to a healthy sample) and after the addition 2,4 dinitrophenol (2,4 DNP, compared to no 2,4 DNP) what would you observe?

	O_2 consumption without 2,4 DNP Compared to healthy sample	**O_2 consumption Presence of 2,4 DNP** compared to sample before 2,4 DNP addition
A)	Near normal	unchanged
B)	Near normal	increased
C)	Suppressed	unchanged
D)	Suppressed	increased
E)	Increased	unchanged

14) Given the relationship between the function of the electron transport chain and the utilization of reducing equivalents, what glycolytic enzyme would most directly be affected by mitochondrial dysfunction caused by the loss of CoQ?

A) Phosphofructokinase 1
B) Glyceraldehyde 3-phosphate dehydrogenase
C) Phosphoglycerate kinase
D) Glucokinase
E) Enolase

15) A patient is undergoing a "cleansing diet" and has not eaten in about 36 hours. He has become forgetful and seems "out of it". He is quickly diagnosed as hypoglycemic and you conclude that the patient's gluconeogenic pathway is defective. Upon further investigation, you determine the patient has high circulating levels of alanine, pyruvate, and lactate. You also realize there is little if any oxaloacetate being produced and biotin supplements seem to alleviate most of the symptoms. Which of the following enzymes is most likely defective in the patient?

A) Pyruvate carboxylase
B) Phosphoenolpyruvate carboxykinase
C) Malate dehydrogenase
D) Fructose 1,6-bisphosphatase
E) Glucose 6-phosphatase

16) Which of the following correctly characterizes the pyruvate dehydrogenase complex?

A) It uses biotin as a cofactor
B) It is positively regulated by phosphorylation
C) It is negatively regulated by AMP
D) Its product can be converted directly into glucose
E) It is critical for deciding the fate of pyruvate within the cell

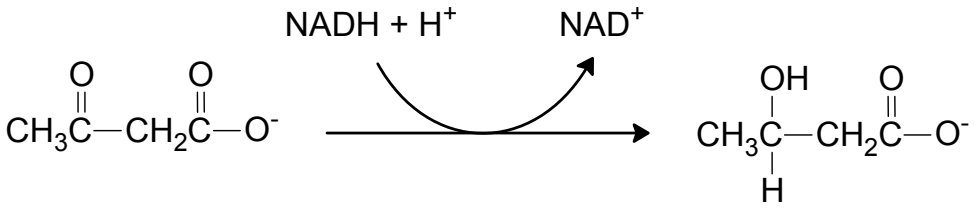
17) Which of the following correctly describes the malate-aspartate shuttle following a carbohydrate laden meal?

A) Malate will be transported into the cytoplasm
B) The production of oxaloacetate will be favored in the matrix of the mitochondria
C) Glutamate-oxaloacetate transaminase will be inactive
D) Aspartate will be transported into the matrix of the mitochondria
E) The reducing equivalents of NADH will be shuttled to the cytoplasm

18) Regulating the fate of pyruvate is essential for metabolic regulation. Which of the following correctly describes the possible fates of pyruvate within a cell?

A) Is converted to lactate in the mitochondria
B) Can be formed from acetyl-CoA via the pyruvate dehydrogenase complex running in reverse
C) Can be converted to malate via malate dehydrogenase
D) Can be converted to phosphoenolpyruvate via pyruvate kinase
E) In the presence of oxygen, donates enough carbon for one complete turn of the TCA cycle

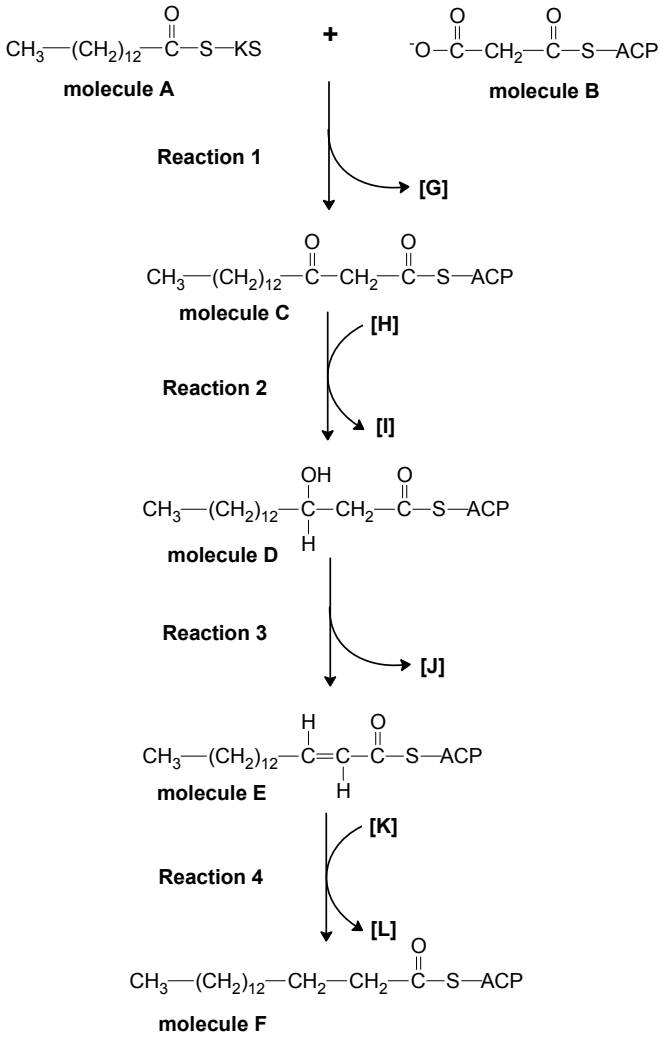
19) What is the correct enzyme name for the reaction drawn below?



A) acetoacetate dehydrogenase
B) β-hydroxybutyrate reductase
C) β-hydroxybutyrate dehydrogenase
D) acetoacetate reductase
E) β-hydroxybutyrate synthase

20) Which of the following statements regarding the ketone bodies acetoacetate and β-hydroxybutyrate is **INCORRECT**?

A) The position of the keto group on the β-carbon of acetoacetate allows it to spontaneously decarboxylate to form acetone.
B) Tissues can utilize acetoacetate and β-hydroxybutyrate as fuel sources.
C) The reduction of acetoacetate to β-hydroxybutyrate allows more ketone bodies to be available as a fuel source.
D) The reduction of acetoacetate to β-hydroxybutyrate in the liver involves the corresponding oxidation of NADPH + H^+ to $NADP^+$.
E) HMG-CoA lyase produces acetoacetate, the first ketone body formed.



The following 2 questions refer to the pathway drawn above. The letters in brackets [ ] represent additional substrates and products for the reactions indicated.

21) Which of the following statements regarding the above pathway is **CORRECT**?

A) Biotin provides the "long arm" of the complex that carries out these reactions.
B) Reaction 3 is a hydration reaction.
C) The committed step of this pathway produces the three carbon unit of molecule B.
D) The enzymes that carry out reactions 2 and 4 are dehydrogenases.
E) Reaction 2 requires NAD^+ (molecule **[H]**) and Reaction 4 requires FAD (molecule **[K]**).

22) In a liver cell, molecule F (after it is released) could be immediately used by that same liver cell for which of the following pathways:

A) Ketone body synthesis only
B) Triacylglycerol synthesis only
C) Phospholipid synthesis only
D) Triacylglycerol synthesis or phospholipid synthesis
E) Ketone body synthesis or triacylglycerol synthesis or phospholipid synthesis

23) What are the products of beta-oxidation of a C_{14} fatty acid?

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

24) What is the correct sequence of enzymes for the four repeated steps of **β- oxidation of fatty acids**?

A) carnitine acyl transferase I (CAT I), carnitine acyl transferase II (CAT II), β-hydroxyacyl-CoA, β-ketothiolase
B) acyl CoA dehydrogenase, enoyl CoA dehydratase, β-hydroxyacyl CoA dehydrogenase, β-ketothiolase
C) β-ketoacyl ACP synthase, β-ketoacyl ACP reductase, β-hydroxyacyl ACP dehydratase, enoyl ACP reductase
D) acyl CoA dehydrogenase, enoyl CoA hydratase, β-hydroxyacyl CoA dehydrogenase, β-ketothiolase
E) acyl CoA reductase, enoyl CoA hydratase, β-hydroxyacyl CoA reductase, β-ketothiolase

25) In a test tube, malonyl CoA which was ^{14}C labeled (a radioactive isotope of carbon) at carbon #3 and unlabeled acetyl CoA were mixed together with the fatty acid synthase complex. Which of the carbons of the final product, palmitate, will be ^{14}C labeled?

A) All of the carbons will be labeled
B) None of the carbons will be labeled
C) Only the methyl carbon (carbon #16) will be labeled
D) All of the even numbered carbons will be labeled, except for carbon # 16
E) Carbons #1 through #14 will all be labeled

26) Which of the following pathways is **correctly** paired with one of its regulated enzymes?

A) fatty acid synthesis: acyl-CoA synthetase
B) β-oxidation: carnitine acyl transferase I (CAT I)
C) cholesterol synthesis: HMG-CoA lyase
D) bile salt synthesis: β-ketothiolase
E) ketone body synthesis: acetyl CoA carboxylase

27) The lipid soluble vitamins

A) are all derived from cholesterol.
B) are all capable of being synthesized in the body, though not in sufficient quantities.
C) obtained in the diet are packaged into chylomicrons for delivery to tissues.
D) are readily excreted from the body.
E) include all of the B vitamins.

28) Which of the following transporters is **correctly** matched with what it transports?

A) Carnitine: fatty acids
B) LDLs: dietary triacylglycerols (TAGs)
C) Citrate: malonyl CoA
D) Albumin: ketone bodies
E) Glycerol-phosphate: protons (H^+)

29) The regulation of cholesterol synthesis involves all of the following **EXCEPT**:

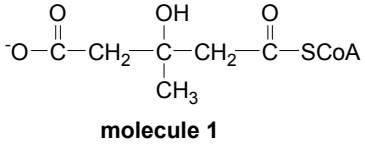
A) glucagon activates HMG-CoA reductase
B) cholesterol inhibits the synthesis of HMG-CoA reductase
C) cholesterol increases the degradation of HMG-CoA reductase
D) the dephosphorylated form of HMG-CoA reductase is active
E) cholesterol inhibits the synthesis of the LDL receptor

30) The digestion and absorption of dietary lipids involves

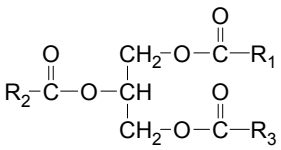
A) absorption of intact molecules of phospholipids across the intestinal mucosa.
B) hormone-sensitive lipase to cleave the dietary triacylglycerols.
C) Emulsification of large lipid droplets by bile salts.
D) lipase breakdown of triacylglycerols in the mouth.
E) complete degradation of the lipid-soluble vitamins.

31) Which of the following are all **POSITIVE** regulators of fatty acid synthesis?

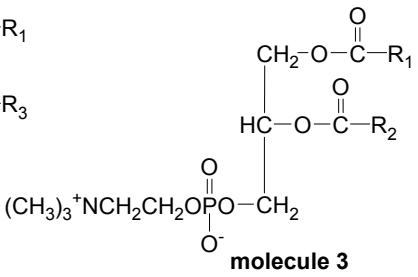
A) ATP, citrate, glucagon
B) Insulin, citrate, ATP
C) Palmitoyl CoA, AMP, insulin
D) Glucagon, epinephrine, citrate
E) Palmitoyl CoA, AMP, glucagon



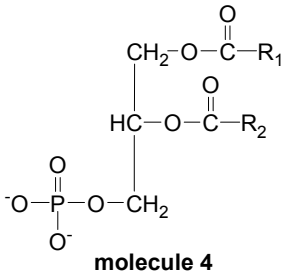
molecule 1



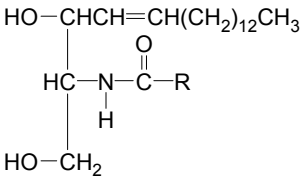
molecule 2



molecule 3



molecule 4



molecule 5

32) Which of the above molecules is the key intermediate ("branch point") in the pathways of phosphosphingolipid and glycolipid synthesis?

A) molecule 1
B) molecule 2
C) molecule 3
D) molecule 4
E) molecule 5

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

1. _____	11. _____	21. _____	31. _____
2. _____	12. _____	22. _____	32. _____
3. _____	13. _____	23. _____	
4. _____	14. _____	24. _____	
5. _____	15. _____	25. _____	
6. _____	16. _____	26. _____	
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10. _____	20. _____	30. _____	