

**Lewis & Clark College  
Metabolic Biochemistry  
Quiz II, Spring 2008**

*"I would rather discover a single fact, even a small one, than debate the great issues at length without discovering anything at all."*

*Galileo Galilei*

1. Familial hypercholesterolemia is associated with elevated levels of LDL cholesterol. Strategies for treating the disorder include the following: a strict dietary regimen that limits dietary cholesterol, use of drugs that competitively inhibit HMGCoA reductase, use of resins that bind the bile acids. Draw on your understanding of cholesterol metabolism and explain the molecular basis of each of these therapies.
2. One indication of the relative importance of various ATP-producing pathways is the maximum activity of key regulatory enzymes present in a tissue. The activities of several metabolic enzymes from the pectoral muscles (chest muscles used for flying) of pigeon and pheasant are presented in Table 1.

Enzyme	Pigeon	Pheasant
Hexokinase	3.0	2.3
Glycogen phosphorylase	18.0	120.0
Phosphofructokinase	24.0	143.0
Citrate synthase	100.0	15.0
Triglyceride lipase	0.07	0.01

**Table 1. Enzyme Activity in Pectoral Muscle of Pigeon and Pheasant Tissue.**

Activity for each of the enzymes is expressed as  $\mu\text{mol}$  substrate per min/g tissue.

- A. Explain why these particular enzymes were selected for comparison and discuss the relative importance of glycogen metabolism and fat metabolism for ATP generation in the muscle tissue of the two birds.
- B. The enzyme activity profile for pigeon is strikingly similar to the enzyme profile found in the pectoral muscle of migratory birds. Offer an explanation for why this particular enzyme profile is characteristically seen for birds that are long distance flyers.

3. Draw on your understanding of metabolic processes and address each of the following questions in a concise manner:
- A. Why do blood concentrations of ketone bodies rise in untreated diabetic patients?
  - B. Why is urea output increased during fasting?
  - C. Why do individuals with a hereditary deficiency of carnitine acyltransferase have muscle weakness. Why are these symptoms more severe during fasting?
  - D. What mechanism ensures against simultaneous fatty acid synthesis and oxidation in the same cell?
4. Dietary fuel molecules that are not needed for immediate generation of ATP can be metabolically converted to triglyceride. Draw on your understanding of the metabolic pathways and illustrate the means whereby excess dietary protein can be converted to triglyceride for storage. In addressing this question, you should adopt a case study approach and profile this conversion by using one of the amino acids. In your metabolic pathway, make certain that all of the carbon atoms in the triglyceride that you synthesize are derived from carbon atoms originally present in your starting amino acid. Note all key cofactors that will be required to facilitate this metabolic conversion.