READING GUIDE

GLYCOGEN METABOLISM

1. Diagram and describe how glycogen is synthesized and degraded, including the rate-determining enzymes
2. Identify the enzyme deficiency for glycogen storage diseases, types I, II, III, and V, and contrast the clinical features and prognoses

STRUCTURE AND FUNCTION OF GLYCOGEN

- What is the structure of glycogen? (Fig 11.3)
- What is the importance of α(1->6) and α(1->4) glycosidic bonds?
- What is the function of glycogen in liver and muscle?

GLYCOGEN SYNTHESIS (GLYCOGENESIS)

- What is UDP-glucose and how is it synthesized?
- What is UDP-glucose pyrophosphorylase?
- What is glycogen synthase? (Fig 11.5)
- Why is glycogenin used? (Fig 11.5)
- How are glycogen chains elongated?
- What is phosphoglucomutase important for? (Fig 11.6)
- How are the branches of glycogen synthesized?

GLYCOGEN DEGRADATION (GLYCOGENOLYSIS)

- Glycogen chains are shortened by what enzyme? (Fig 11.7)
- There are two different enzymatic activities important for the removal of glycogen branches. What are they and how do they function (Fig 11.8)
- What is glycogen phosphorylase? What does it do? Where is it located? What is glucose 6—phosphate translocase? Why is it important? (Fig 11.8)
- Lysosomes can also degrade glycogen. What enzyme is important for this?

REGULATION OF GLYCOGEN SYNTHESIS AND DEGRADATION

- What is the difference between Glycogen Phosphorylase a and Glycogen Phosphorylase b? (Fig 11.9)
- What is the difference between Glycogen Phosphorylase kinase a and Glycogen Phosphorylase kinase b? (Fig 11.9)
- What is the difference between Glycogen Synthase a and Glycogen Synthase b? (Fig 11.10)
- How do glucagon and epinephrine regulate inhibition of glycogen synthesis? (Fig 11.10)
- How do glucagon and epinephrine regulate glycogen degradation? (Fig 11.10)
- How does Glucose-6-phosphate allosterically regulate Glycogen synthase and Glycogen phosphorylase? (Fig 11.11)
- How does calcium activate glycogen degradation? (Fig 11.12)
- How does calcium activation of liver phosphorylase kinase relate to "fight or flight" situations?

GLYCOGEN STORAGE DISEASES

- What is Glycogen Storage Disease Type I (Von Gierke disease)? (Fig 11.8)
- What is Glycogen Storage Disease Type V (McArdle syndrome)? (Fig 11.8)
- What is Glycogen Storage Disease Type II (Pompe disease)? (Fig 11.8)
- What is Glycogen Storage Disease Type III (Cori disease)? (Fig 11.8)