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Glycogen Storage Diseases

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33. What are the major glycogen storage diseases? How does the deficiencies of specific enzyme result in defects in glycogen?
    a. Glucose-6-Phosphatase Deficiency
        i. Increase in G-6-P leads to inhibition of phosphorylase, activation of synthase --> accumulation of glycogen stores
        ii. Inability to respond to glucagon --> hypoglycemia
    b. Branching Enzyme Deficiency
        i. Glycogen concentration is normal
        ii. Structure is abnormal (long chains, only one non-reducing end)
        iii. Slower breakdown of glycogen
    c. Muscle Phosphorylase Deficiency
        i. Muscle can't use glycogen for energy
    d. Liver Phophorylase
        i. Increase glycogen stores
        ii. Decreased response to glucagon
34. What is the function of UDP-glucuronate and how is it synthesized?
    a. Carries products of metabolism out to urine
        i. Bilirubin
        ii. Steroids
        iii. Drugs or toxins
    b. Synthesized from UDP-Glucose
        i. Synthesized using 2 NAD --> 2 NADH
35. What are the various functions of UDP-glucose?
    a. Glycolipids --> neurotransmitters
    b. Glycogen
    c. Galactose metabolism
    d. Oligosaccharides and glycoproteins