

GLYCOGEN STORAGE DISEASE VI

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- glycogen is the storage form of glucose in the body primarily in liver and muscle
 - ✓ liver: stored for replenishing blood glucose during food deprivation or stress
 - ✓ muscle: stored for providing fuel during high intensity exercise in the cell where stored
- glycogen phosphorylase, the first enzyme in glycogen breakdown, has a different form in liver and muscle
- glycogen storage disease VI:
 - ✓ is a defect only of liver glycogen phosphorylase
 - ✓ patients cannot maintain blood glucose (**hypoglycemia**)
 - ✓ glycogen accumulates in the liver causing the organ to be enlarged (**hepatomegaly**)
 - ✓ accumulated glycogen pushes glucose towards other products (Figure 1)

