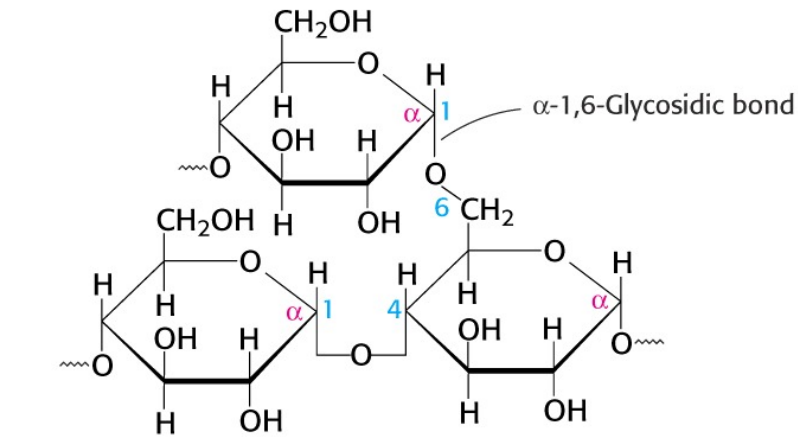
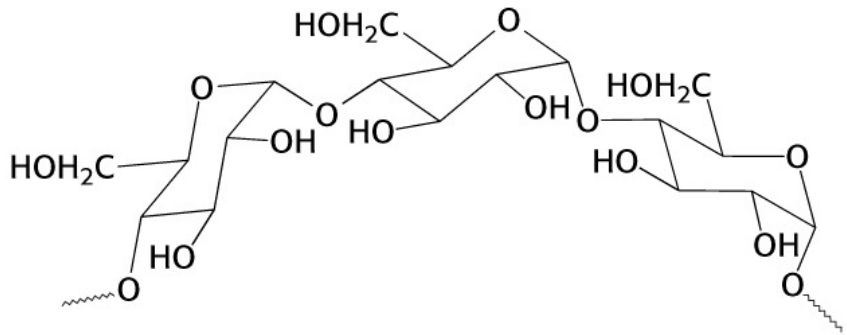
Glycogen metabolism

- Glycogen
- Glycogen breakdown
- Glycogen synthesis
- Regulation

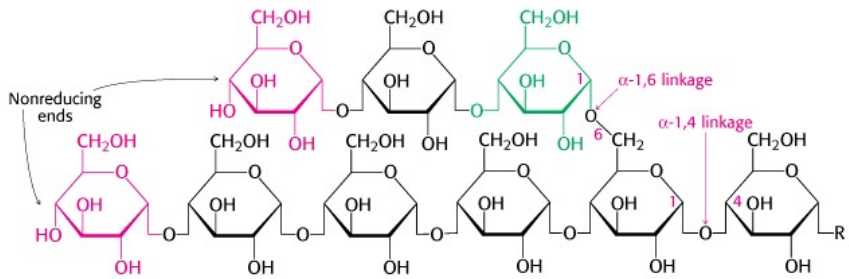


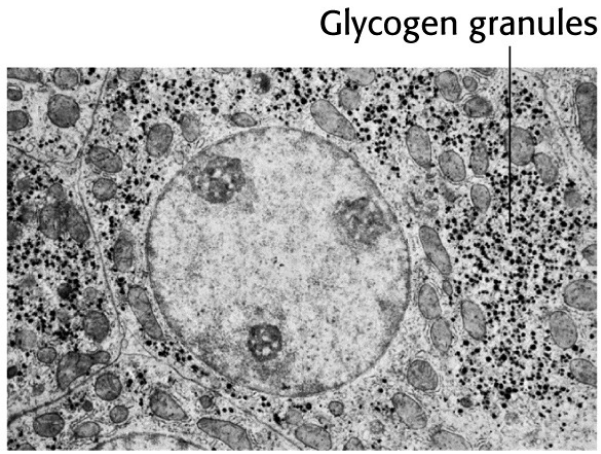


Starch and Glycogen
(α-1,4 linkages)

Glycogen

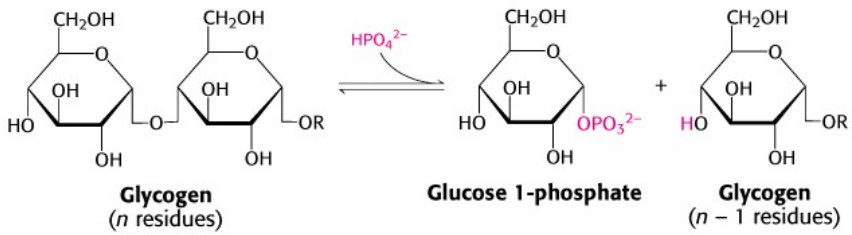
- Long chain of glucose units hooked together 1,4 linkage
- branched 1,6
- lowers osmotic potential of cell
- storage form of energy in liver and muscle



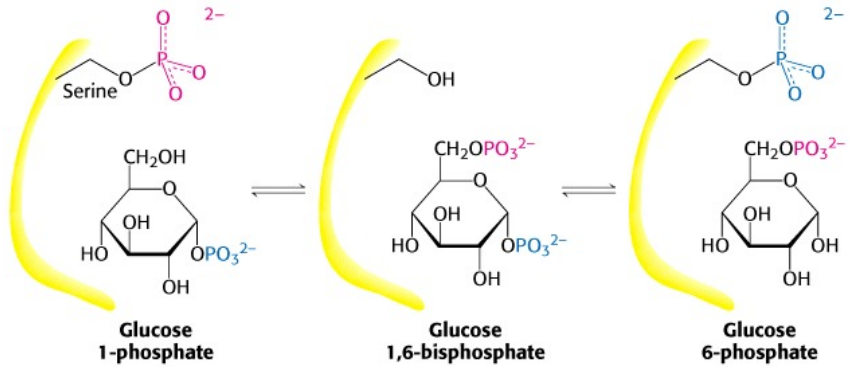


Glycogen breakdown

- glycogen phosphorylase
- inorganic phosphate, not ATP
- glucose 1-P produced
- less ATP used in glycolysis if glycogen used for energy
- phosphoglucomutase (PGM)----> glucose 6-P



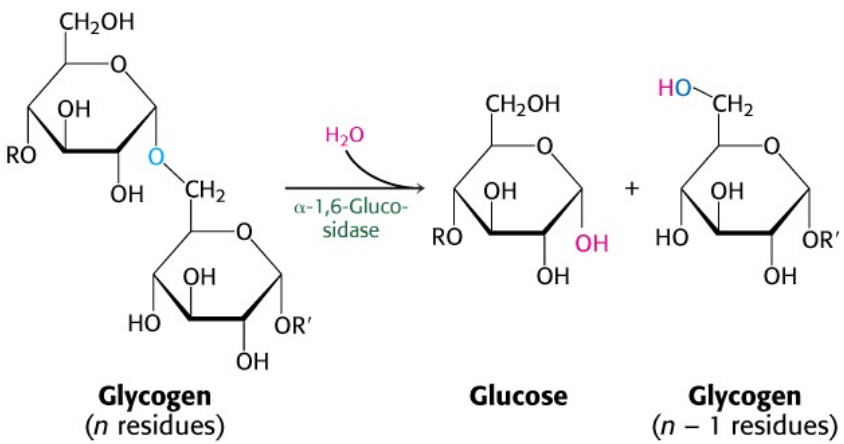
Glycogen phosphorylase

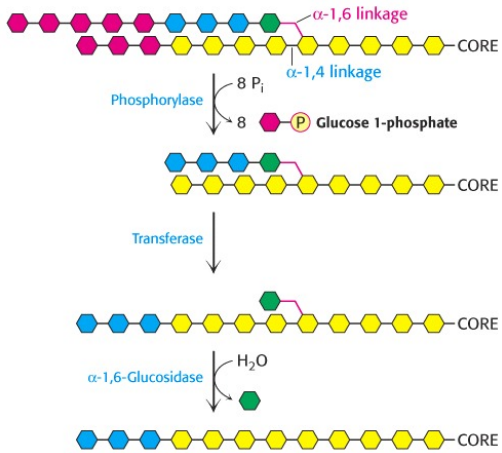


Phosphoglucomutase

Glycogen breakdown

Debranching enzyme

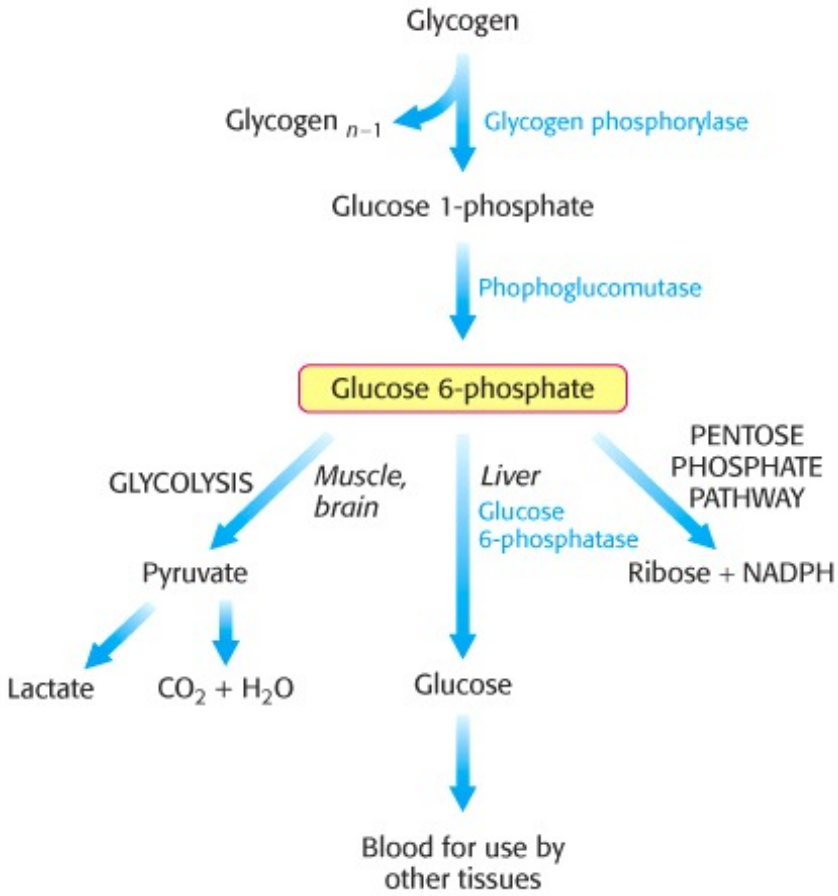




- Glucosidase --debranching enzyme
- 3 glucose residues moved
- cleaves remaining glucose for free glucose

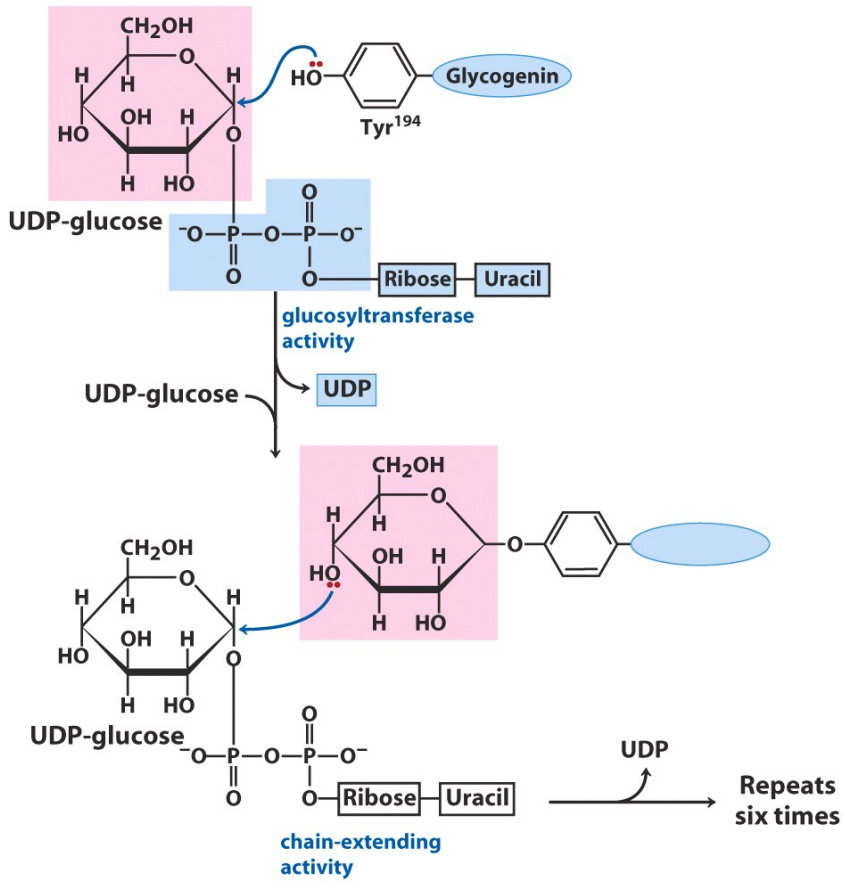
Liver Glycogen

- Glucose 6 phosphotase
- Found in Smooth Endoplasmic recticulum
- produce free glucose
- blood glucose



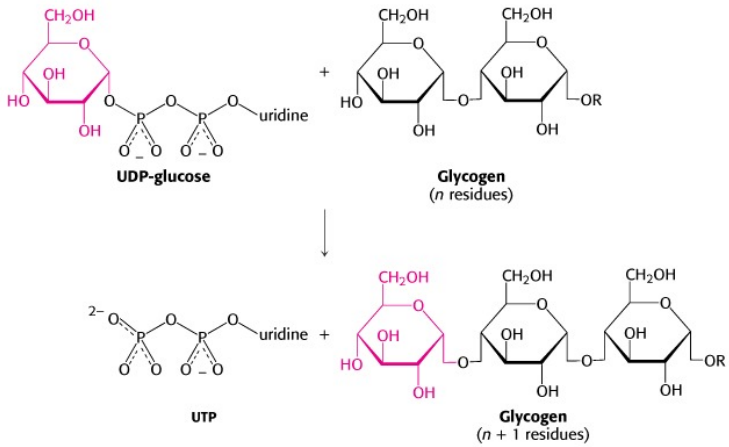
Glycogen synthesis

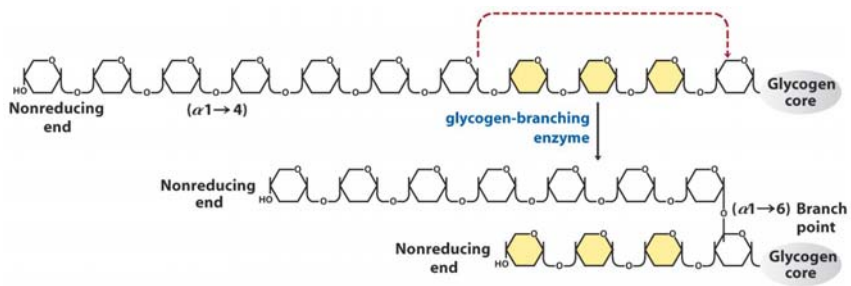
- Glycogen core started
- Protein glycogenin
- Uses primer (4 residues attached to phenolic oxygen of tyrosine



Glycogen synthesis

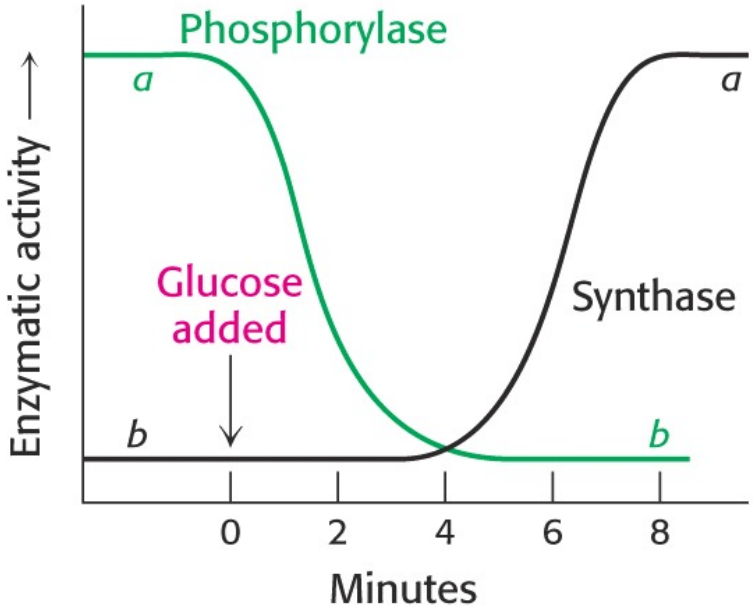
- Glycogen Synthase
- UDP-glucose used Transglycoslyase
- branching enzyme
- transfers 8-10 glucose residues

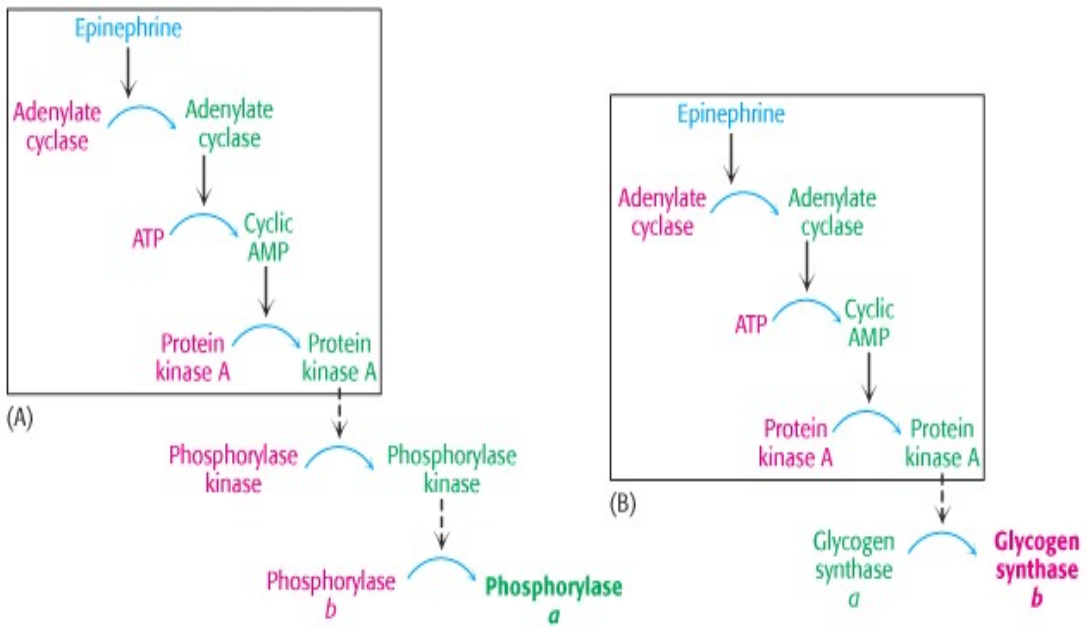




Regulation

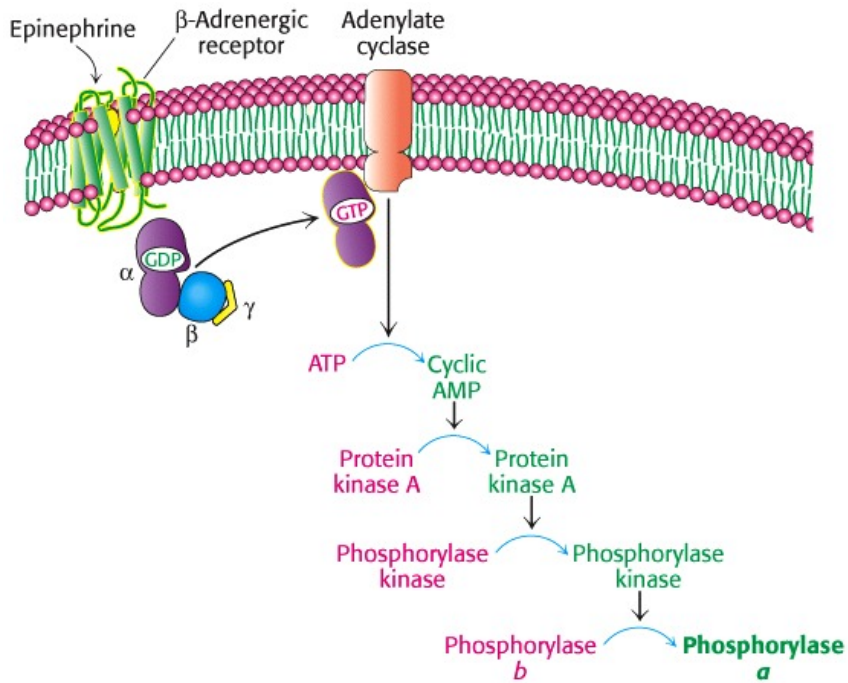
- Glycogen breakdown and synthesis are reciprocally regulated
- Controlled by hormones
- Insulin
- Glucagon
- Epinephrine
- Phosphorylation



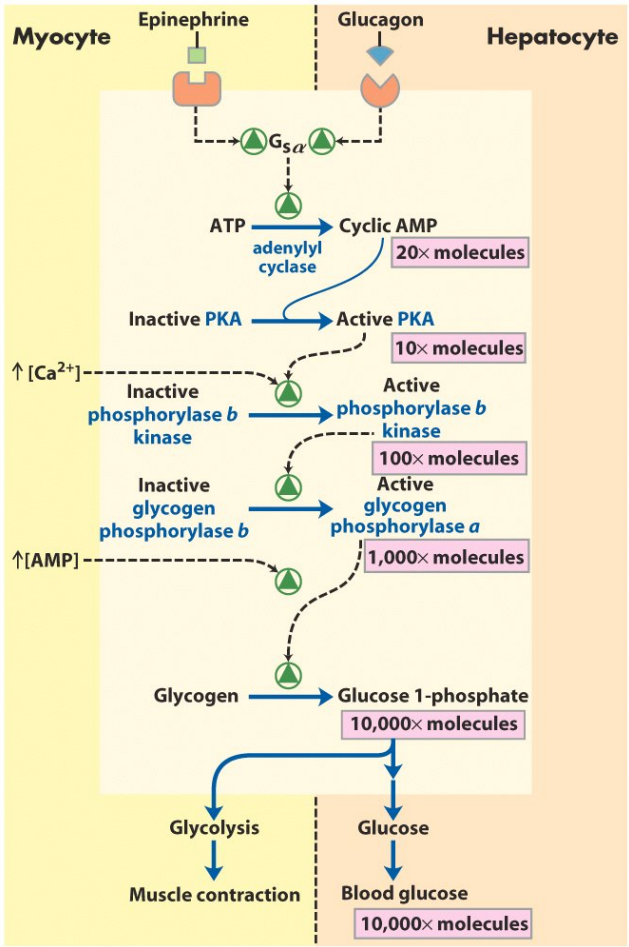


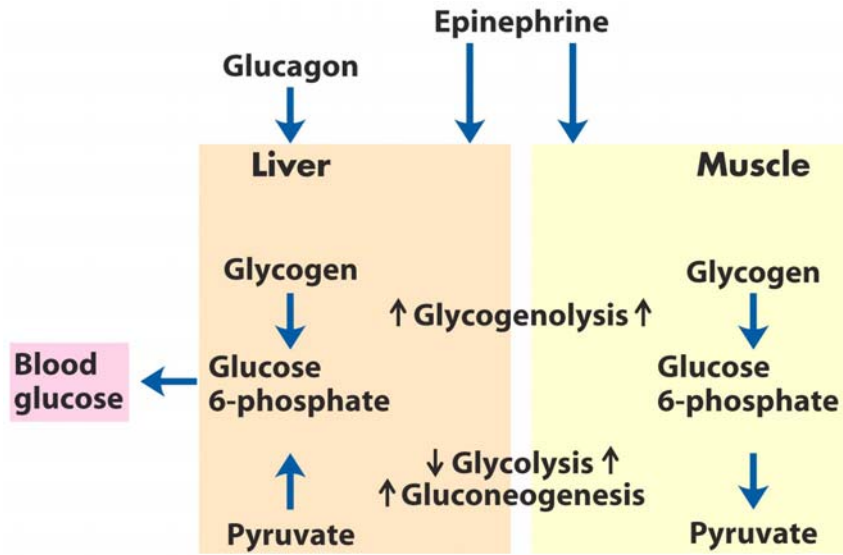
Glycogen phosphorylase

- **Epinephrine and glucagon**
- **stimulate cAMP**
- **protein kinase**
- **phosphorylase kinase**
- **phosphorylates glycogen phosphorylase**
- **glycogen breakdown**
- **phosphorylates glycogen synthase, deactivated**



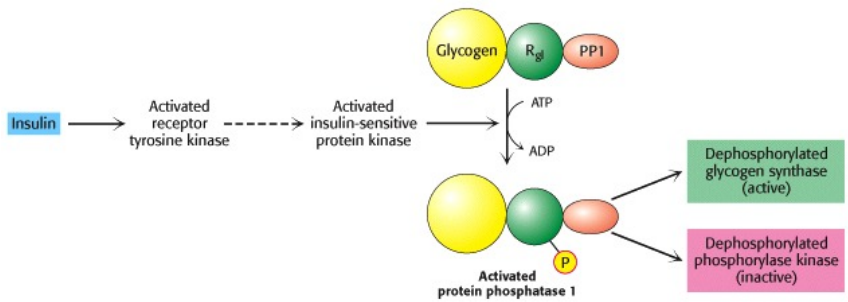
Differences in liver and muscle





Glycogen synthase

- insulin
- stimulates glycogen synthase
- dephosphorylation
- deactivates glycogen phosphorylase



Glycogen Storage Diseases

TABLE 21.1 Glycogen-storage diseases

Type	Defective enzyme	Organ affected	Glycogen in the affected organ	Clinical features
I Von Gierke disease	Glucose 6-phosphatase or transport system	Liver and kidney	Increased amount; normal structure.	Massive enlargement of the liver. Failure to thrive. Severe hypoglycemia, ketosis, hyperuricemia, hyperlipemia.
II Pompe disease	α-1,4-Glucosidase (lysosomal)	All organs	Massive increase in amount; normal structure.	Cardiorespiratory failure causes death, usually before age 2.
III Cori disease	Amylo-1,6-glucosidase (debranching enzyme)	Muscle and liver	Increased amount; short outer branches.	Like type I, but milder course.
IV Andersen disease	Branching enzyme (α-1,4 ⟶ α-1,6)	Liver and spleen	Normal amount; very long outer branches.	Progressive cirrhosis of the liver. Liver failure causes death, usually before age 2.

Note: Types I through VII are inherited as autosomal recessives. Type VIII is sex linked.

TABLE 21.1 Glycogen-storage diseases

Type	Defective enzyme	Organ affected	Glycogen in the affected organ	Clinical features
V McArdle disease	Phosphorylase	Muscle	Moderately increased amount; normal structure.	Limited ability to perform strenuous exercise because of painful muscle cramps. Otherwise patient is normal and well developed.
VI Hers disease	Phosphorylase	Liver	Increased amount.	Like type I, but milder course.
VII	Phosphofructokinase	Muscle	Increased amount; normal structure.	Like type V.
VIII	Phosphorylase kinase	Liver	Increased amount; normal structure.	Mild liver enlargement. Mild hypoglycemia.

Note: Types I through VII are inherited as autosomal recessives. Type VIII is sex linked.