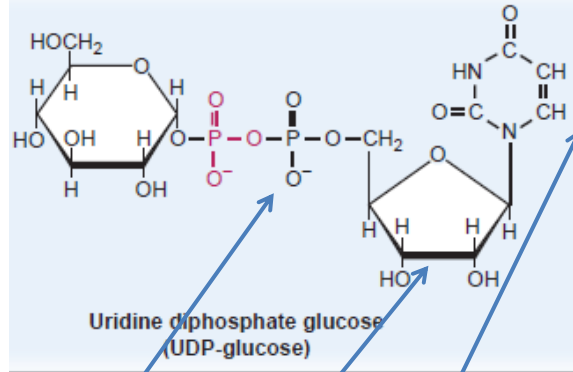
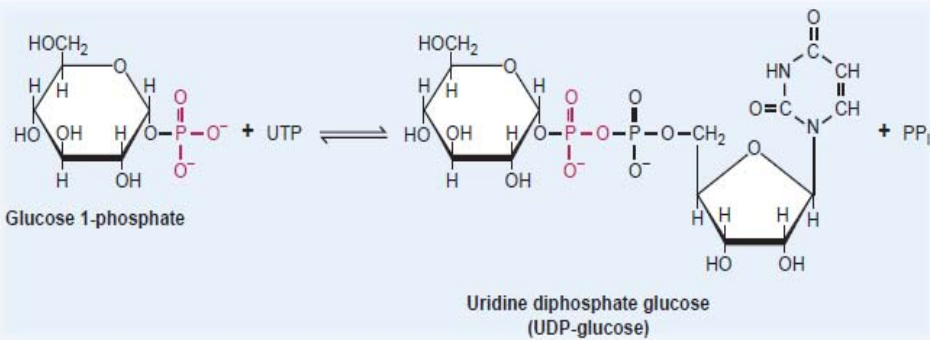
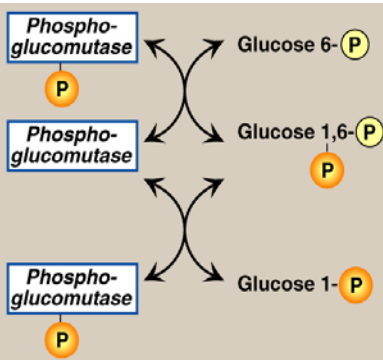


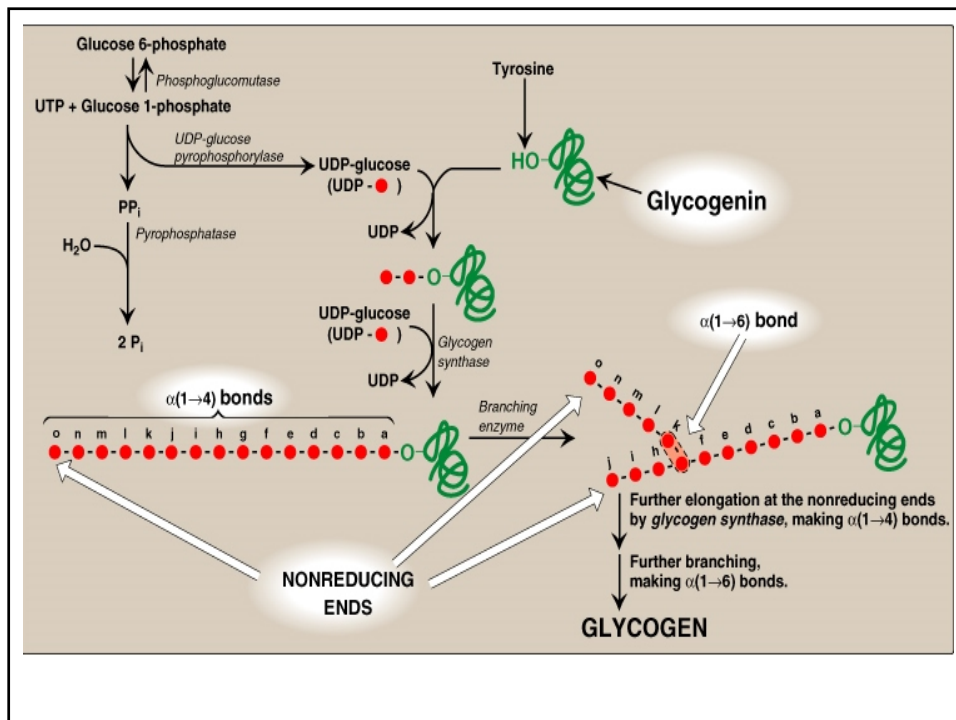
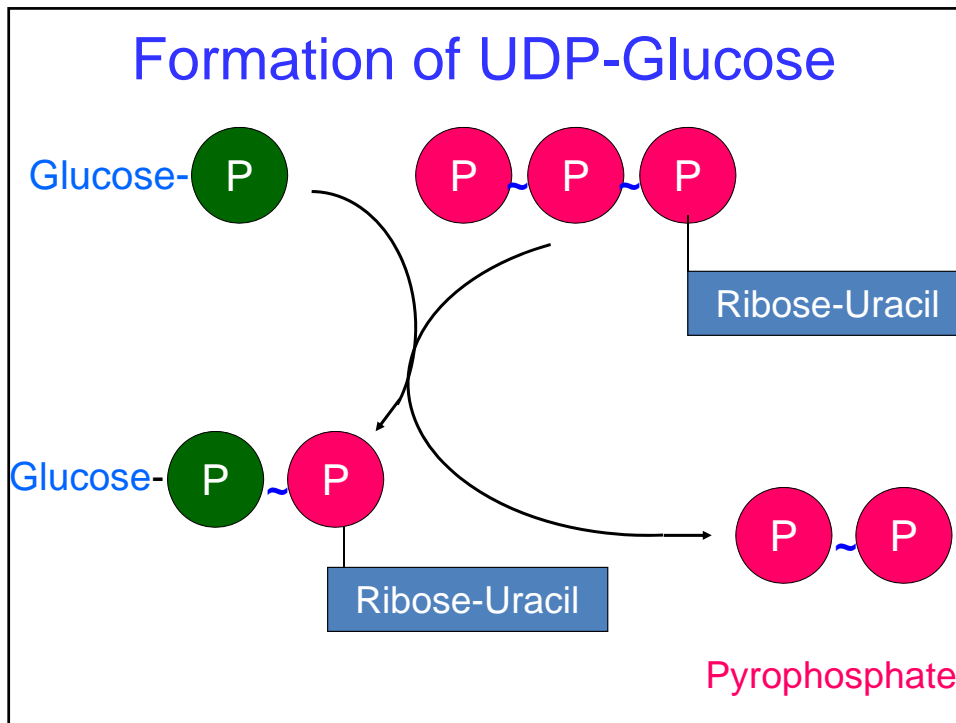
Glycogen is synthesized by adding glucose one by one
 UDP-Glucose is the active donor of glucose units



Phosphate Ribose Uracil

Formation of UDP-Glucose



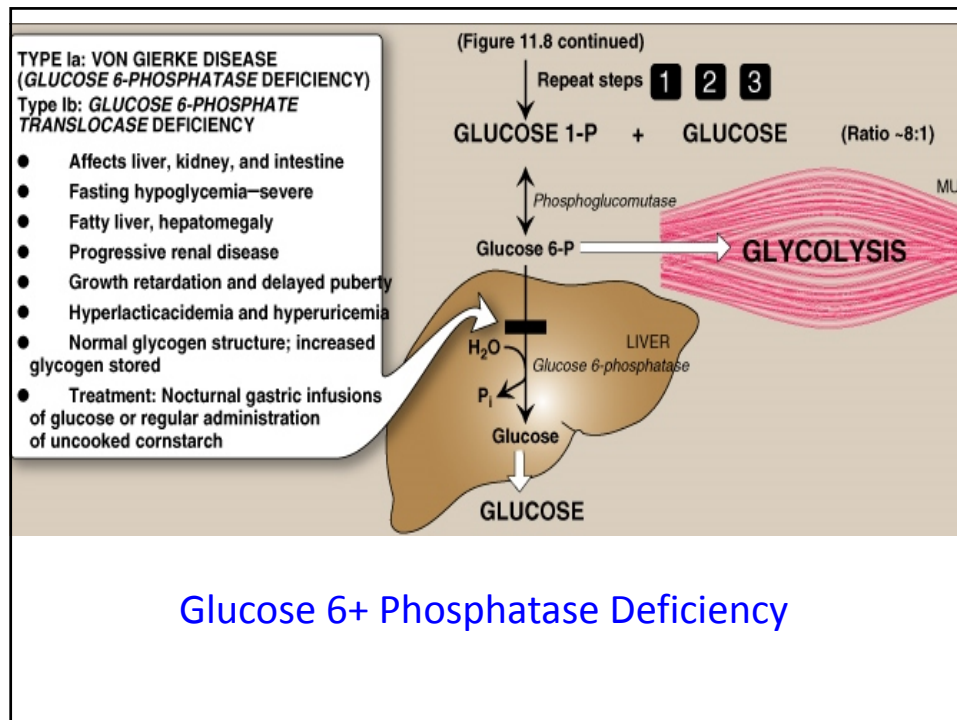


Glycogen Storage Diseases

- Genetic diseases
- Defect in an enzyme required for synthesis or degradation →
- Accumulation of excessive amount of glycogen
- In one or more tissue
- Severity: **FATAL in Infancy**..... **Mild disorder**

Glycogen Storage Diseases (examples)

- I Glucose-6-phosphatase (von Gierk's) disease
 - Liver, kidney and intestine.
 - Severe fasting hypoglycemia
 - Hepatomegaly fatty liver.
 - Normal glycogen structure.
 - Progressive renal disease.
 - Growth retardation.

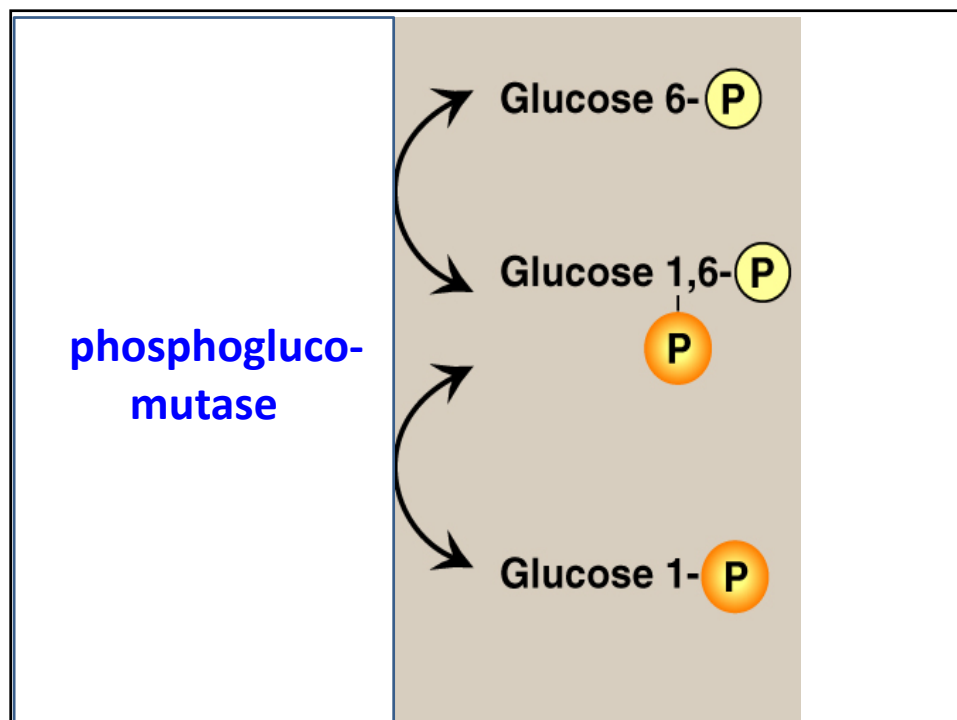


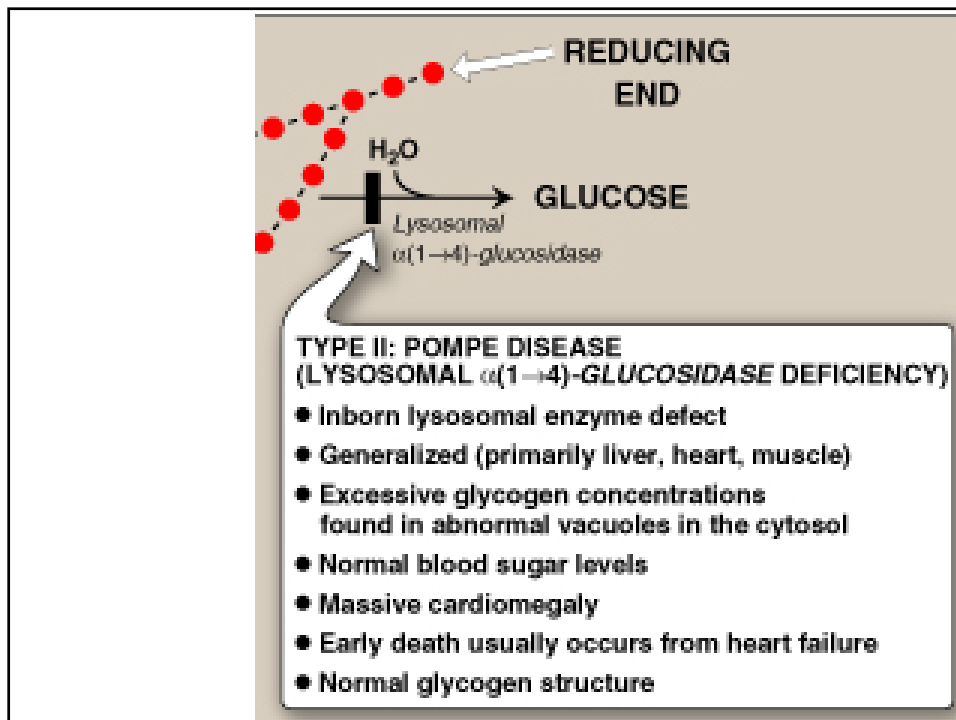
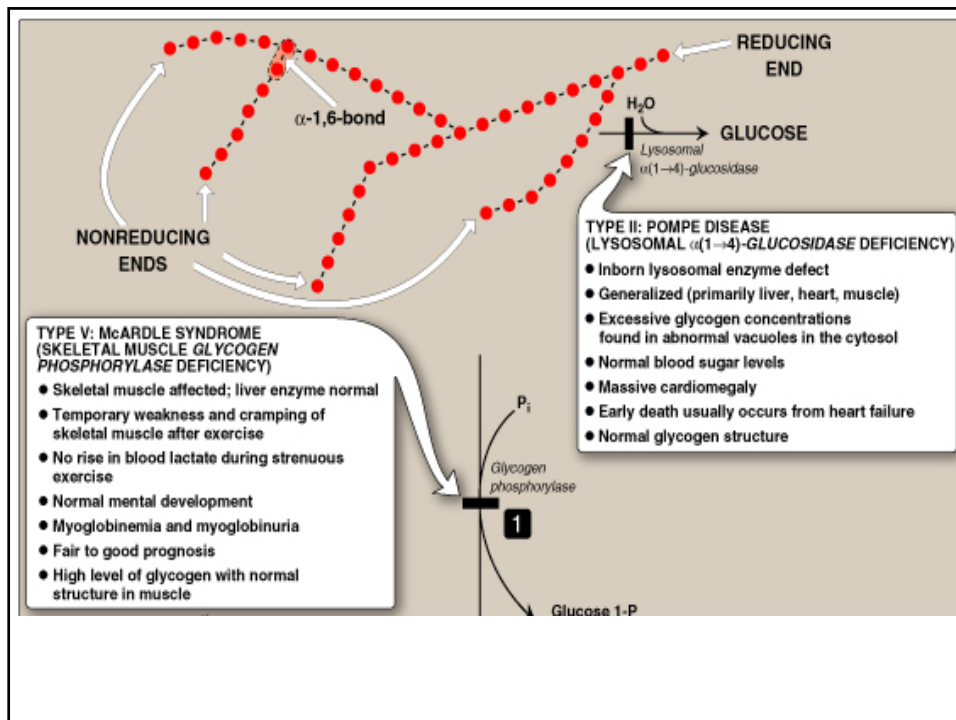
Glycogen Storage Diseases (examples)

- **V** Muscle glycogen phosphorylase (McArdle syndrome)
 - Only muscle is affected;
 - Weakness and cramping of muscle after exercise
 - no increase in [lactate] during exercise

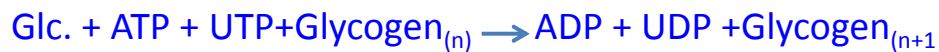
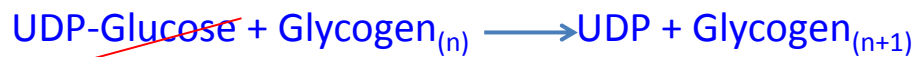
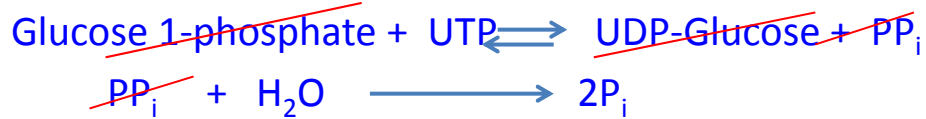
Glycogen Storage Diseases (examples)

- II Lysosomes α (1 \rightarrow 4) glucosidase \rightarrow POMP Disease
- Degradation of glycogen in the lysosomes
- \approx 3% of glycogen is degraded in the lysosomes
- Affects liver, heart and muscle
- Excessive glycogen in abnormal vacuoles in the lysosomes
- Massive cardiomegaly
- Normal blood sugar, normal glycogen structure
- Early death from heart failure.

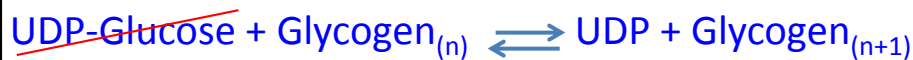
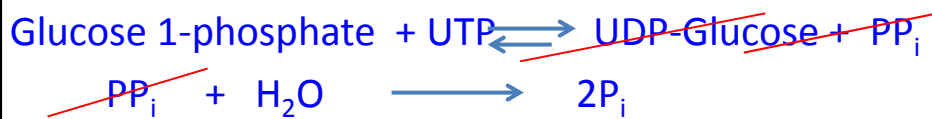




Energy needed for glycogen synthesis

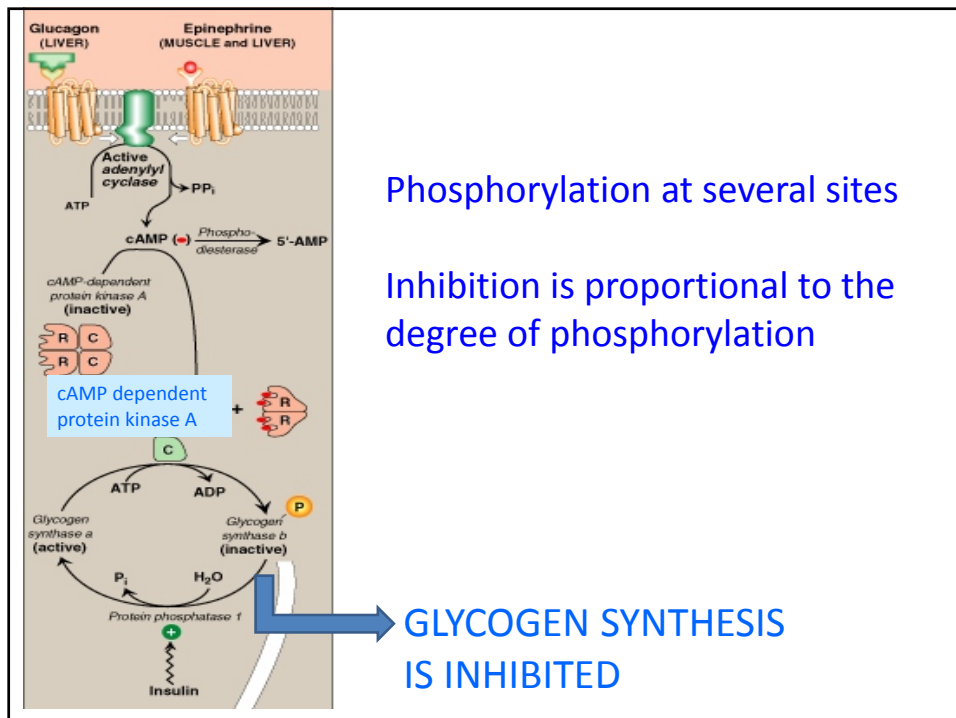
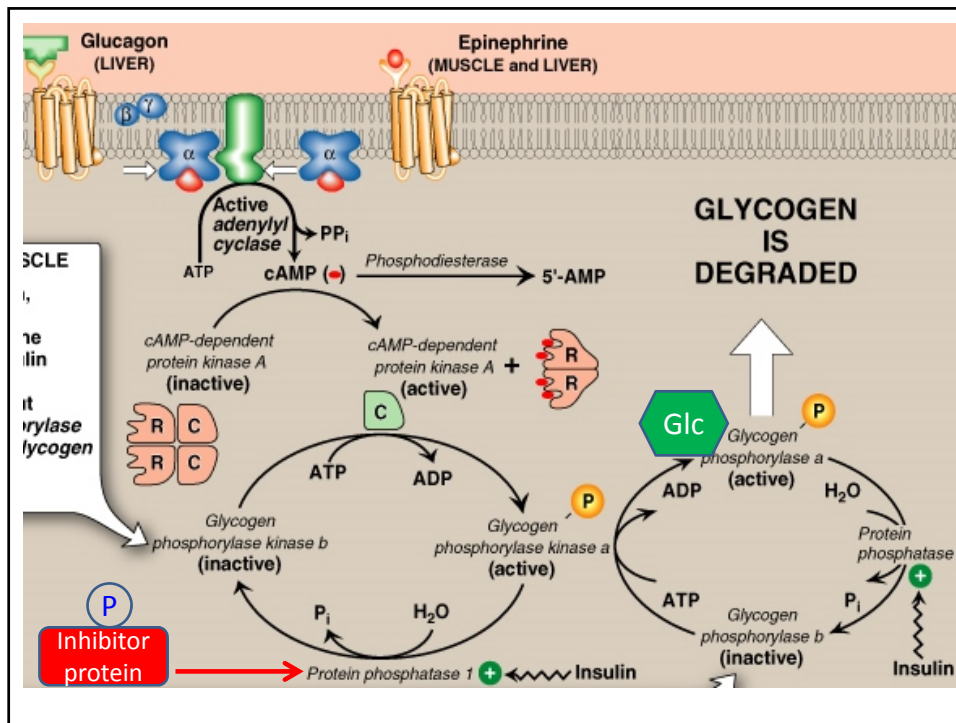


The net reaction in glycogen synthesis and degradation



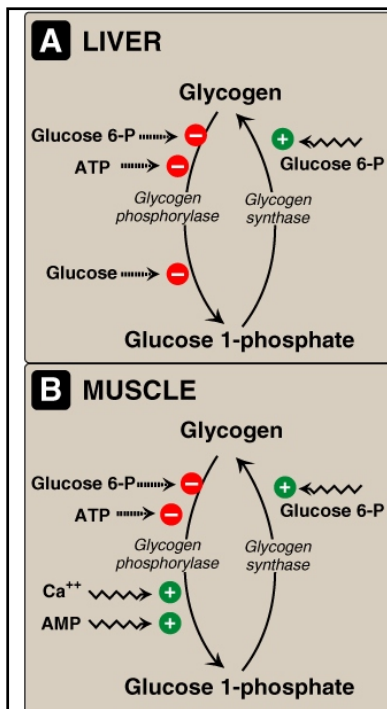
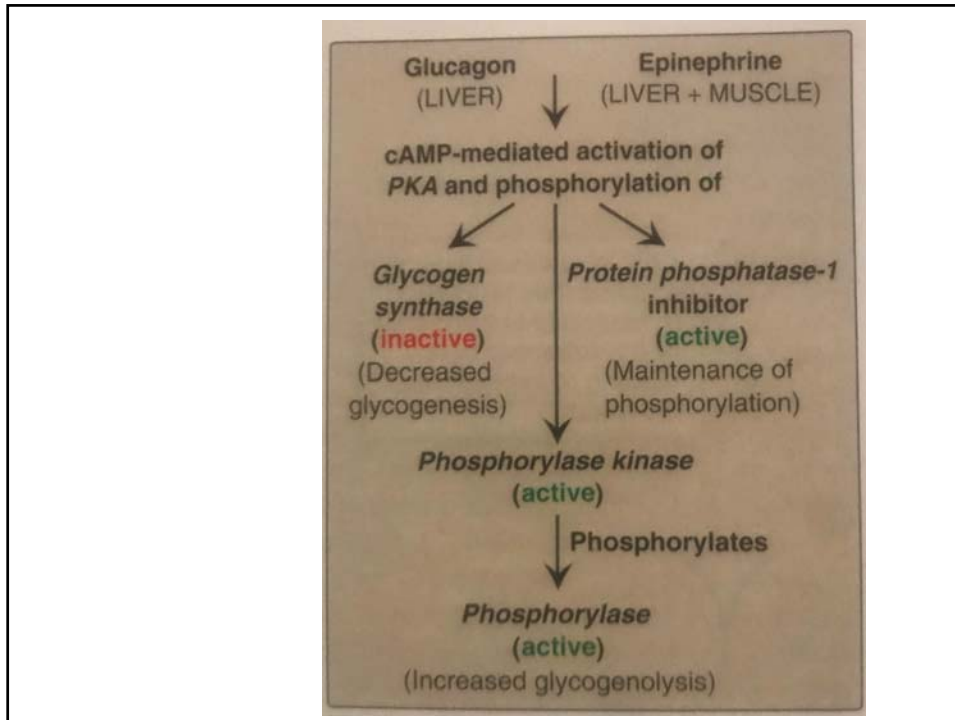
Degradation





Phosphorylation at several sites

Inhibition is proportional to the degree of phosphorylation



Allosteric Regulation
 Rapid response to cell's needs
 Available substrate and ATP → synthesis

↓↓ Glucose and ↓ ATP → Glycogenolysis

