GLYCOGEN- METABOLISM

Glycogen is a polysaccharide that is the principal storage form of glucose in animal/human cells.

Glycogen metabolism is synthesis and breakdown of glycogen. It is a very large, branched polymer of glucose residues that can be broken down to yield glucose molecules when energy is needed. The pathway for the synthesis and degradation of glycogen is not reversible.

Glycogen

GLYCOGENESIS

Definition: The synthesis of glycogen from glucose is called glycogenesis

Location: Cytosol of Liver and Muscle cell

Requirements: ATP (Adenosine triphosphate), UTP (Uridine Triphosphate), Glucose

Steps:

1. Synthesis of UDP glucose
2. Requirement of primer to initiate glycogenesis
3. Glycogen synthesis by glycogen synthase
4. Formation of branches in glycogen

GLYCOGENOLYSIS

Definition: The breakdown or degradation of stored glycogen into glucose is called glycogenolysis. Glycogen is degraded by breaking α 1, 4-bonds and α 1, 6-bonds glycosidic bond.

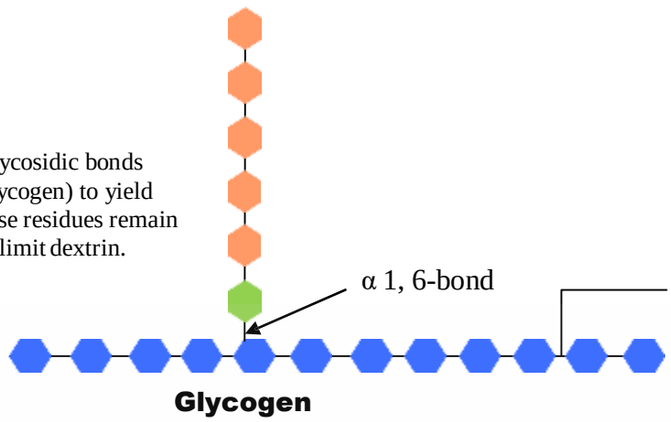
Location: Cytosol of Liver and Muscle cell

Steps:

1. Action of glycogen phophorylase
2. Action of debranching enzyme
3. Formation of glucose-6-phosphate and glucose

GLYCOGENOLYSIS

Breakdown of glycogen into glucose



Glycogen

1. Phosphorylasis
Glycogen phosphorylase cleave α 1, 4 glycosidic bonds sequentially (from non reducing end of glycogen) to yield glucose 1 phosphate, continues until 4 glucose residues remain on the either side of branching to form limit dextrin.

Pi — Glycogen Phosphorylase — 1

Glucose-1-Phosphate

2. Debranching
Glycosyl tranferase removes fragments of 3-4 glucose of branch chain and transfer them to another chain

3. Cleavage of α 1, 6-bond by α 1-6 glucosidase activity. Release of a free glucose

Limit dextrin

4. Remaining molecule of glycogen is again cleaved by phosphorylase and debranching enzyme to repeat the reaction 1 to 3.

Debranching enzyme (Tranferase activity) — 2

Debranching enzyme (α 1-6 glucosidase activity) — 3

Free Glucose

5. Isomerization
By phosphoglumutase

Further action of Phosphorylase — 4

Glucose-1-Phosphate

6. Release of phosphate group from glucose 6 phosphate by glucose 6 phophatase in liver and kidney to form glucose

Phosphoglucomutas — 5

Glucose-6-Phosphate → Glycolysis

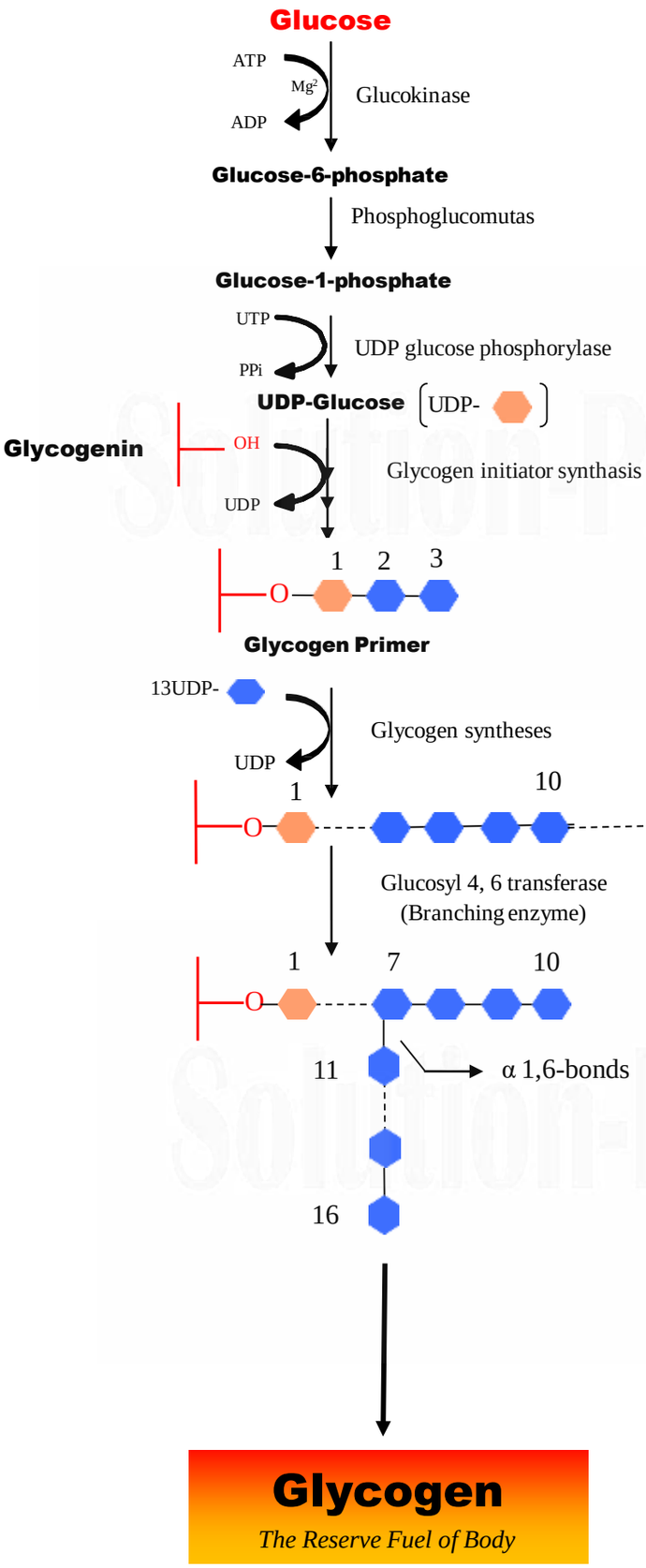
Glucose-6-Phosphatase (In Liver and Kidney only) — 6

Glucose

'The Power of Body'

GLYCOGENESIS

Storage of glucose into glycogen to serve you later (when your body really needs it)



Phosphorylation

Isomerisation

UDP- Uridine Diphosphate Glucose Synthesis

Glyocgenin protein accept glucose from UDPG to from glycogen primer to initiate glycogen synthesis

Glycogen synthase
Transfer Glucose from UDPG to the non reducing end of glycogen formation to form α 1, 4 glycosidic linkages

Formation of branches in glycogen by glucosyl 4, 6 transferase

Further elongation of glycogen by glycogen synthase forming α 1, 4-bonds

Branching by glucosyl 4, 6 transferase forming α 1, 6-bonds

GLYCOGEN STORAGE DISESASE

Metabolic defect in the glycogen synthesis and degradation are collectively known as Glycogen storage disease (GSD). In these condition, deposition of normal or abnormal type of glycogen in one or more tissue.

Glycogen Storage Diseases

Type	Name	Enzyme defect	Organ(s) involved	Characteristic features
0	Glycogen synthase	—	Liver	Hypoglycemia, ketonemia, early death.
I	von Gierke's disease (type I glycogenosis)	Glucose 6-phosphatase	Liver, kidney and intestine	Glycogen accumulates in hepatocytes and renal cells, enlarged liver and kidney, fasting hypoglycemia, lactic acidemia; hyperlipidemia; ketosis; gouty arthritis.
II	Pompe's disease	Lysosomal α-1,4 glucosidase (acid maltase)	All organs	Glycogen accumulates in lysosomes in almost all the tissues; heart is mostly involved; enlarged liver and heart, nervous system is also affected; death occurs at an early age due to heart failure.
III	Cori's disease (limit dextrinosis, Forbe's disease)	Amylo α-1,6-glucosidase (debranching enzyme)	Liver, muscle, heart, leucocytes	Branched chain glycogen accumulates; liver enlarged; clinical manifestations are similar but milder compared to von Gierke's disease.
IV	Anderson's disease (amylopectinosis)	Glucosyl 4-6 transferase (branching enzyme)	Most tissues	A rare disease, glycogen with only few branches accumulate; cirrhosis of liver, impairment in liver function.
V	McArdle's disease (type V glycogenosis)	Muscle glycogen phosphorylase	Skeletal muscle	Muscle glycogen stores very high, not available during exercise; subjects cannot perform strenous exercise; suffer from muscle cramps; blood lactate and pyruvate do not increase after exercise; muscles may get damaged due to inadequate energy supply.
VI	Her's disease	Liver glycogen phosphorylase	Liver	Liver enlarged; liver glycogen cannot form glucose; mild hypoglycemia and ketosis seen.
VII	Tarui's disease	Phosphofructokinase	Skeletal muscle, erythrocytes	Muscle cramps due to exercise; blood lactate not elevated; hemolysis occurs.

Rare glycogen disorders VIII, IX, X and XI have been identified. They are due to defects in the enzymes concerned with activating and deactivating liver phosphorylase.