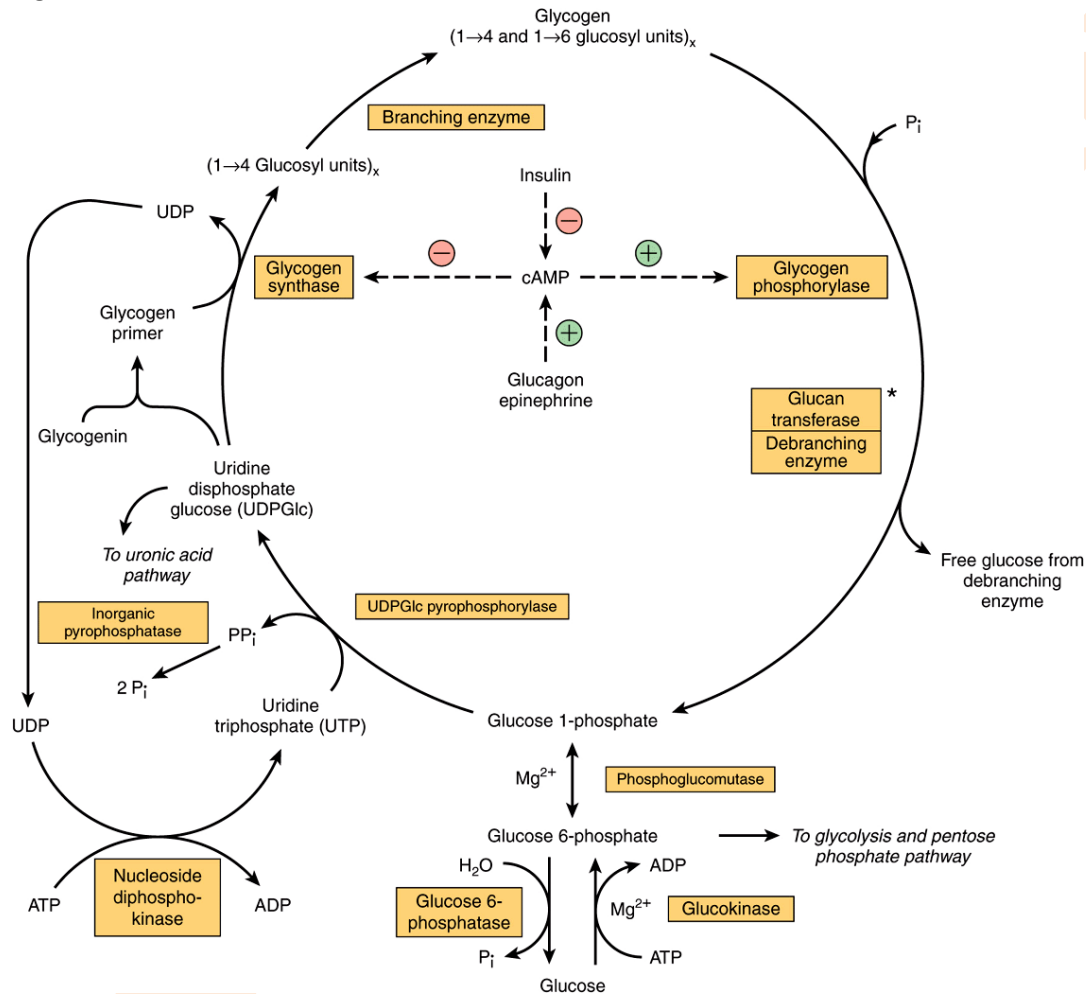


GLYCOGEN METABOLISM

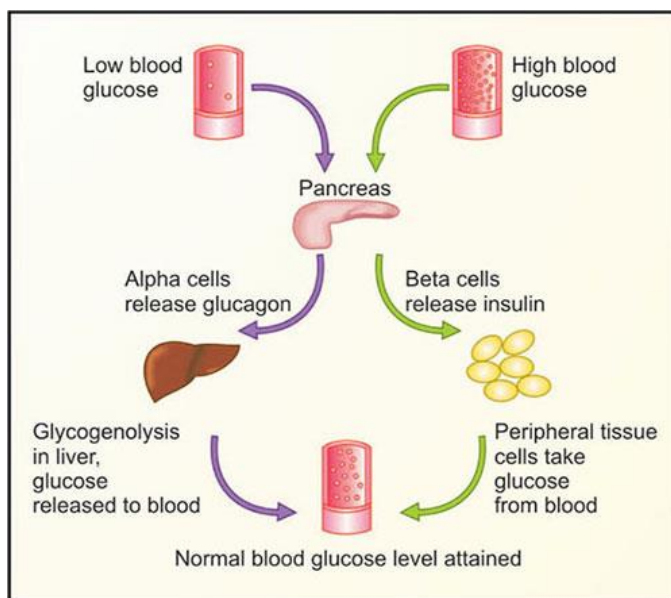
❖ Glycogen metabolism

- **Glycogen** is the major storage form of glucose mainly in the muscle and liver. Also most of the cells store minute amounts of glycogen. Glycogen is composed of glucosyl units which are linked by alpha-1,4 glycosidic bonds. The formation and utilization of glycogen in the body with the help of enzymatic system is called glycogen metabolism.



❖ Regulation of blood glucose

- Inside the pancreas, the hormone insulin is **made in the beta cells, which are part of the Islets of Langerhans**. These islets also have alpha cells, which make glucagon,
- Glucagon helps blood glucose levels rise back up in multiple ways, including:
 - Glucagon triggers your liver to convert stored glucose (glycogen) into a usable form and then release it into your bloodstream. This process is called glycogenolysis.
 - Glucagon can also prevent your liver from taking in and storing glucose so that more glucose stays in your blood.
 - Glucagon helps your body make glucose from other sources, such as amino acids.



Hormones involved in glucose homeostasis		
Hormone	Principal actions	
insulin	increases	cellular glucose uptake glycogen synthesis protein synthesis fatty acid and triglyceride synthesis
	decreases	gluconeogenesis glycogenolysis ketogenesis lipolysis proteolysis
glucagon	increases	glycogenolysis gluconeogenesis ketogenesis lipolysis
adrenaline (epinephrine)	increases	glycogenolysis lipolysis
growth hormone	increases	glycogenolysis lipolysis
cortisol	increases	gluconeogenesis glycogen synthesis proteolysis
	decreases	tissue glucose utilization

❖ Carbohydrate metabolism disease

i) Diabetes

Deficiency	Disorder	Clinical Feature
Galactose-1-phosphate uridyl transferase	Galactosemia	Swollen and inflamed liver; jaundice; weight loss; hypoglycemia; kidney failure; ovarian failure in girls; stunted physical and mental growth; cataracts in eyes
Fructose-1-phosphate-aldolase	Fructosemia	Accumulation of fructose in liver, kidney, and intestine; dislike for sweets and fruits. Fructose intake results in abdominal pain, vomiting, and hypoglycemia. If untreated, can cause liver and kidney damage.
Hepatic fructokinase	Fructosuria	Increased fructose in blood and urine. Rare and harmless.
L-xylulose	Pentosuria	Increased L-xylulose in urine. Harmless. Occurs almost exclusively in Ashkenazim Jews of Polish-Russian origin.

ii)

iii) Glycogen storage disease

- Glycogen storage disease (GSD) is a rare condition that changes the way the body uses and stores glycogen, a form of sugar. It is passed down from parents to children (inherited).

Type	Defective enzyme	Organ affected	Glycogen in the affected organ	Clinical features
I Von Gierke	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	α -1,4-Glucosidase (lysosomal)	All organs	Massive increase in amount; normal structure.	Cardiorespiratory failure causes death, usually before age 2.
III Cori	Amylo-1,6-glucosidase (debranching enzyme)	Muscle and liver	Increased amount; short outer branches.	Like type I, but milder course.
IV Andersen	Branching enzyme (α -1,4 \rightarrow 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.
V McArdle	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	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.

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