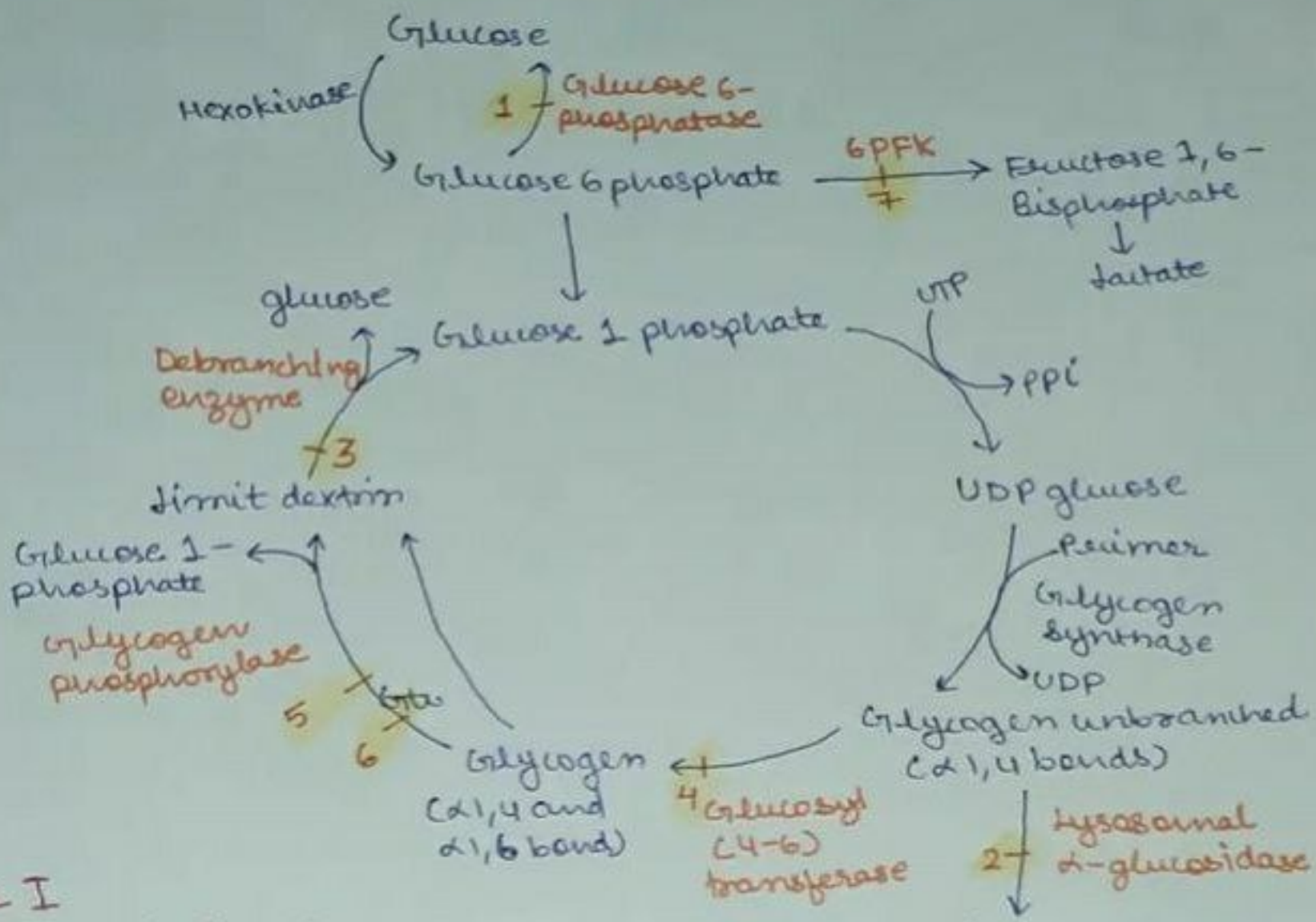
# GLYCOGEN STORAGE DISEASE




## Type I
### Von Gierke's disease

- Enzyme : Glucose 6 phosphatase
- Organ : liver, kidney, intestine
- Feature : Glycogen accumulates in hepatocytes and renal cells,
  - enlarged liver and kidney,
  - fasting hypoglycemia
  - lactic acidemia
  - hyperlipidemia
  - ketosis
  - gouty arthritis.

## Type II
### Pompe's disease

- Enzyme : lysosomal α1,4 glucosidase (acid maltase)
- Organ : All organs
- Features : heart mostly affected.
  - enlarged liver and heart,
  - death occurs at an early age due to heart failure.

## Type VII
### Tarui's disease

- Enzyme : Phosphofructokinase
- organ : Skeletal Muscles, RBC.
- Features : Muscle cramps - blood lactate
  - Hemolysis donot elevated.

## Type III
### Cori's disease (limit dextrinosis, Forbe's disease)

- Enzyme : Amylo α-1,6 glucosidase (debranching enzyme)
- organ : liver, muscle, heart, WBC
- Features : Branched chain glycogen accumulated, liver enlarged.

## Type IV
### Anderson's disease (amylopectinosis)

- Enzyme : Glucosyl 4-6 transferase (branching enzyme)
- Organ : Most tissues
- Features : A rare disease, glycogen with few branches accumulated.
  - cirrhosis of liver.
  - impairment of lives function.

Type V
McArdle's disease
- Enzyme : Muscle glycogen phosphorylase
- Organ : Skeletal Muscle
- Features : Muscle glycogen stores very high.
  - not available during exercise.
  - Muscle cramps.
  - blood lactate and pyruvate do not increases after exercise.
  - Muscles may get damaged due to inadequate energy supply.

Type VI
Her's disease
- Enzyme : liver glycogen phosphorylase.
- organ : liver
- Features : liver enlarged.
  - liver glycogen cannot form glucose.
  - Mild hypoglycemia, ketosis
  - not very serious disease.