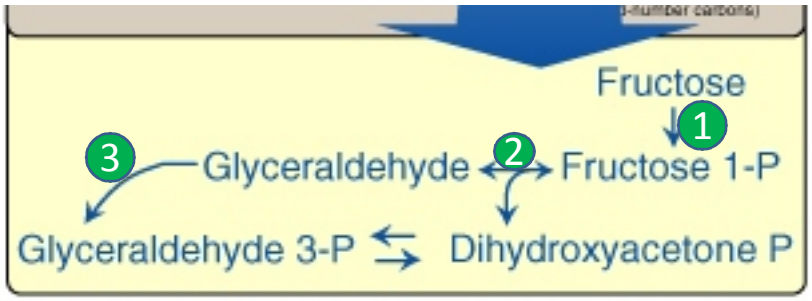
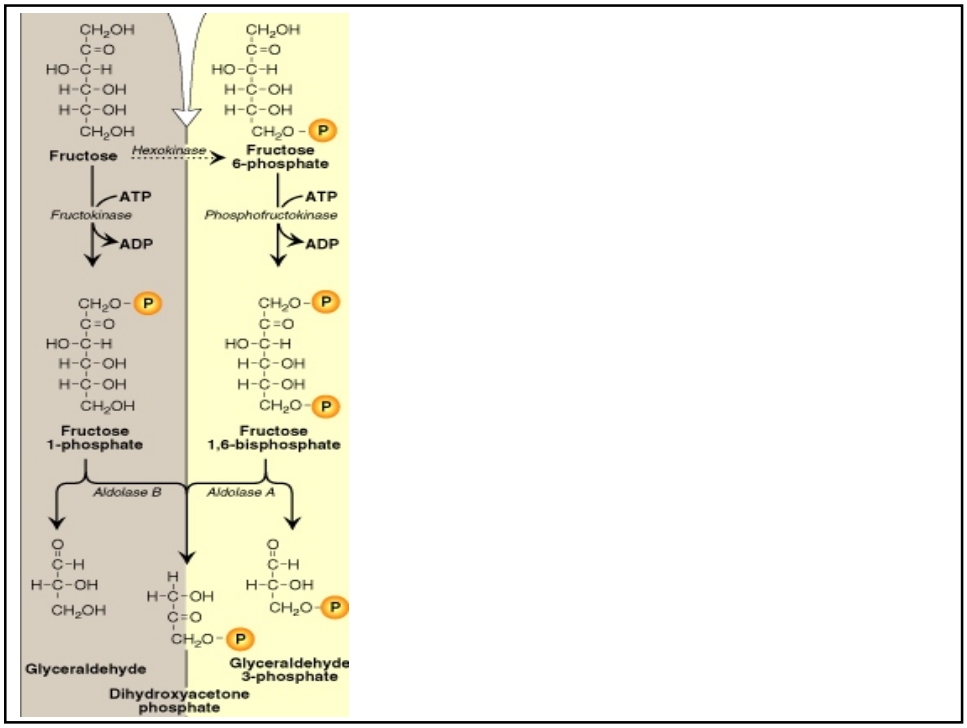
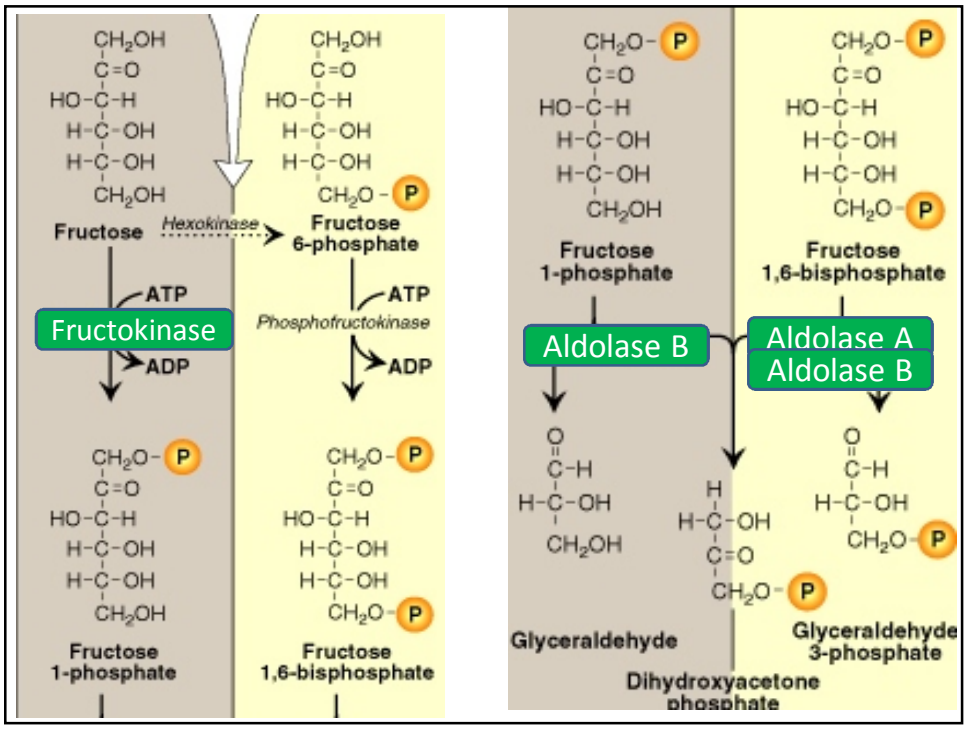
Metabolism of Monosaccharides and Disaccharides

Fructose Metabolism

- 10% of the daily calorie intake
- Sources: sucrose, Fruits, honey, high-fructose corn syrup
- Entry into cells is not insulin dependant.
- Does NOT promote the secretion of insulin




Fructose
Hexokinase
Fructose 6-phosphate
ATP
Fructokinase
ADP
ATP
Phosphofructokinase
ADP
Fructose 1-phosphate
Fructose 1,6-bisphosphate
Aldolase B
Aldolase A
Glyceraldehyde
Glyceraldehyde 3-phosphate
Dihydroxyacetone phosphate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Aldolase A
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Glyceraldehyde
Glyceraldehyde 3-phosphate
Dihydroxyacetone phosphate

Human expresses three forms of aldolase

Aldolase B

- Liver, kidney, small intestine
- Substrate

  Fruc. 1 phpsphate

  Also

  Fruc. 1,6 bisphospate

  ↓activity➔ fructose intolerance

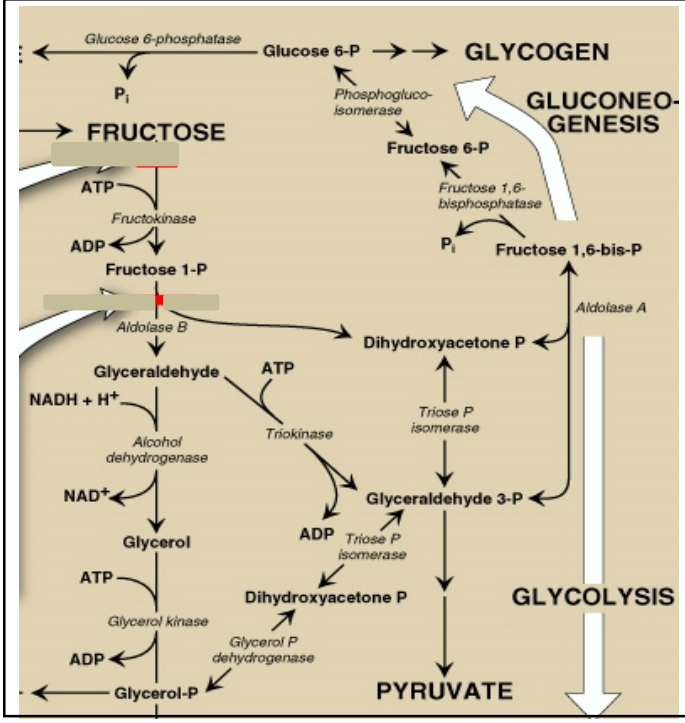
Aldolase A

- In most tissues
- Substrate

Fruc. 1,6 bisphospate

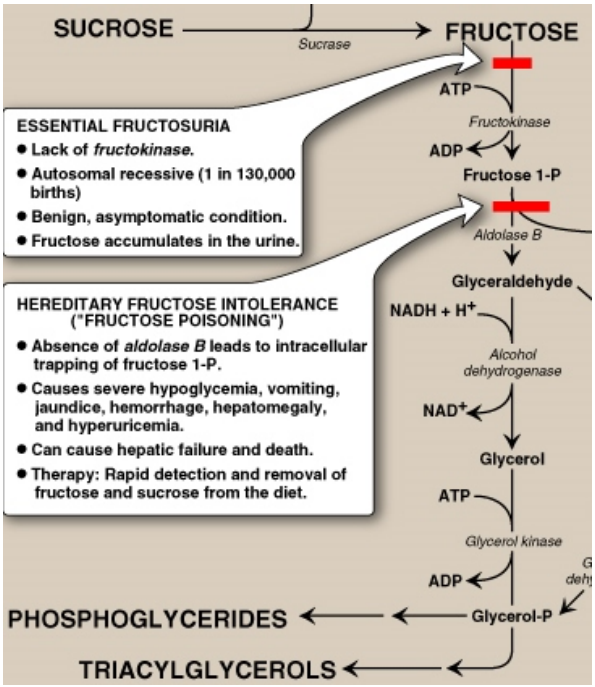
Not

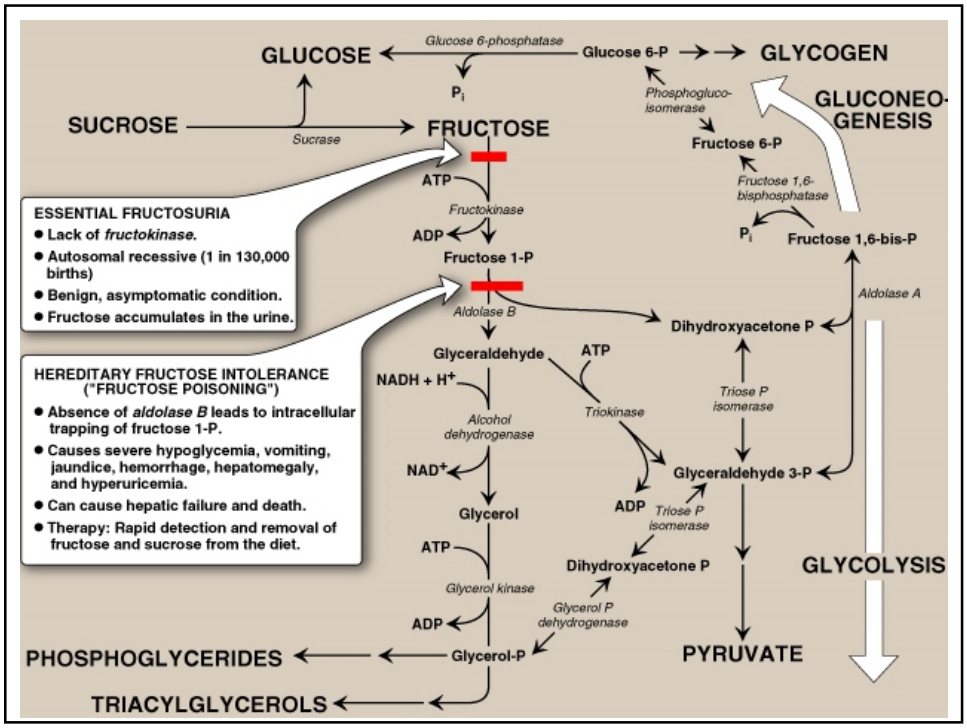
Fruc. 1 phpsphate

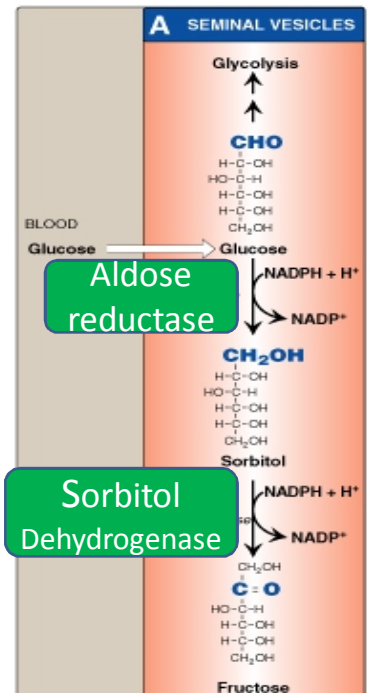


Disorders of Fructose Metabolism

- Fructokinase Deficiency ➔ essential fructosuria
  - Accumulation of fructose ➔ fructosuria
  - Benign condition
- Aldolase Deficiency ➔ hereditary fructose intolerance, (Fructose Poisoning)
  - Severe disturbance in liver and kidney metabolism
  - ↑↑↑ Fruc. 1-Phosph. ➔ drop in P_i ➔drop in ATP ➔ ↑↑ AMP ➔ ↑ degradation of AMP
  - Hypoglycemia and lacticacidemia
  - Hepatic failure





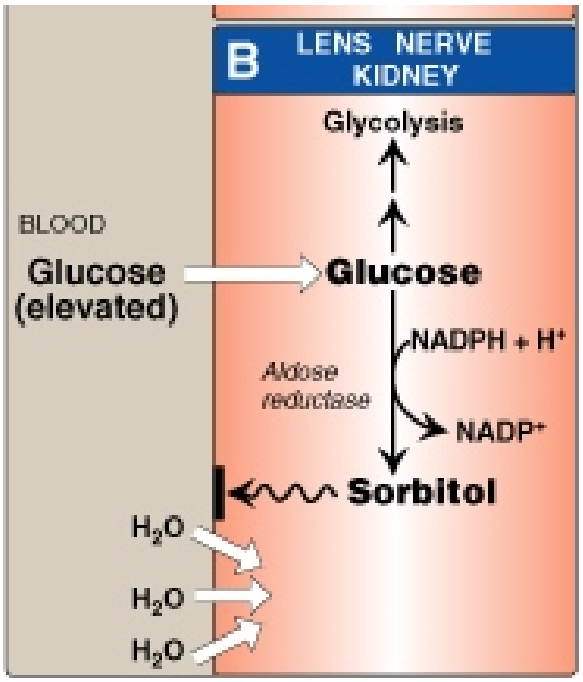


Conversion of glucose to fructose via sorbitol

Aldose Reductase:
Found in many tissues;
Lens, retina, schwan cells, liver, kidney, ovaries, and seminal viscle

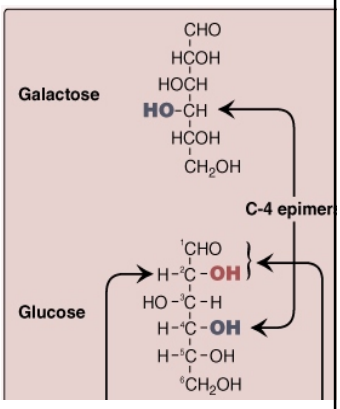
Sorbitol reductase:
Liver, ovarioes and seminal vesicles

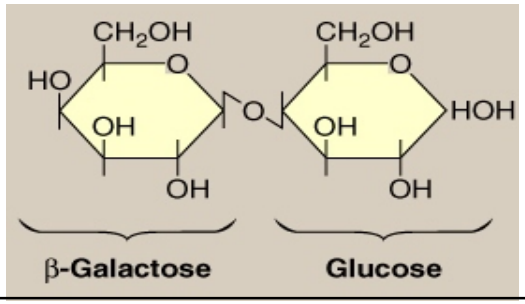
Fructose : the major energy source for sperm cells



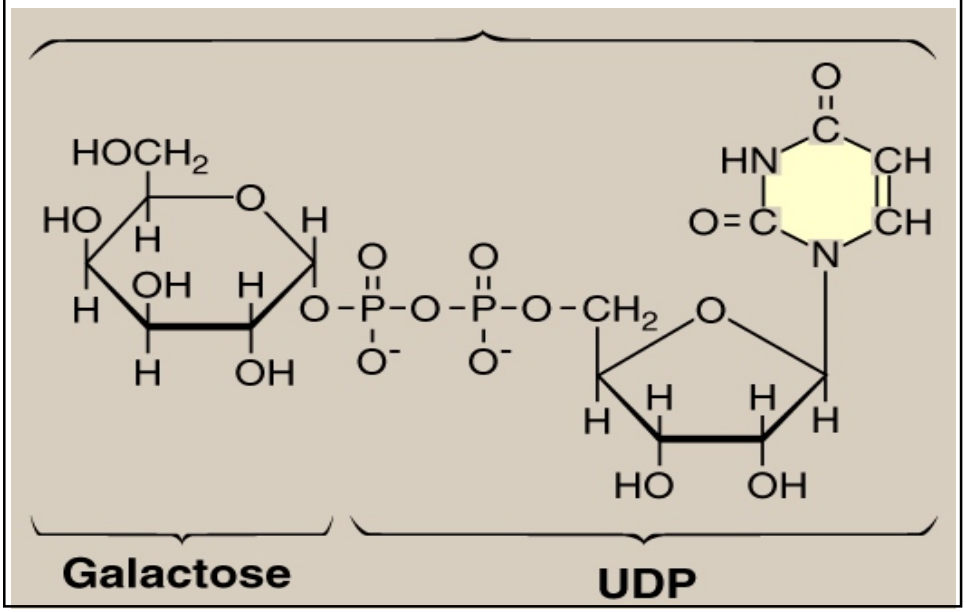
Galactose Metabolism

- Epimer of glucose
- Sources: component of lactose, glycolipids and glycoproteins
- UDP Galactose; an Intermediate in Galactose Metabolism

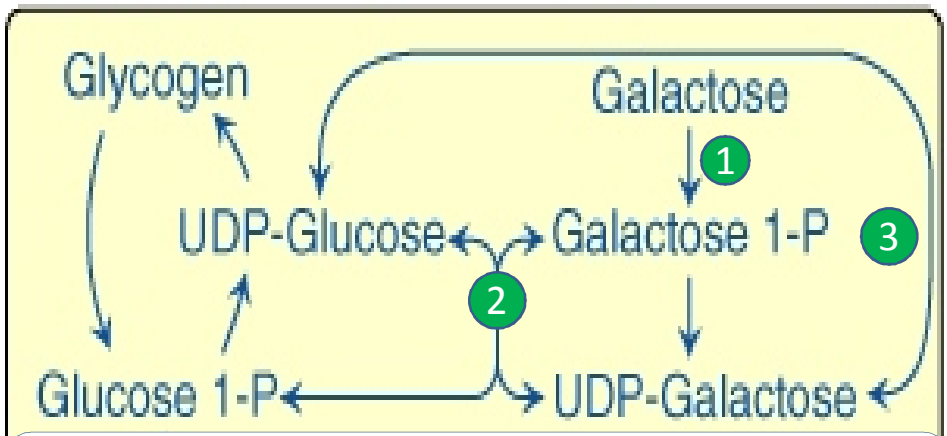


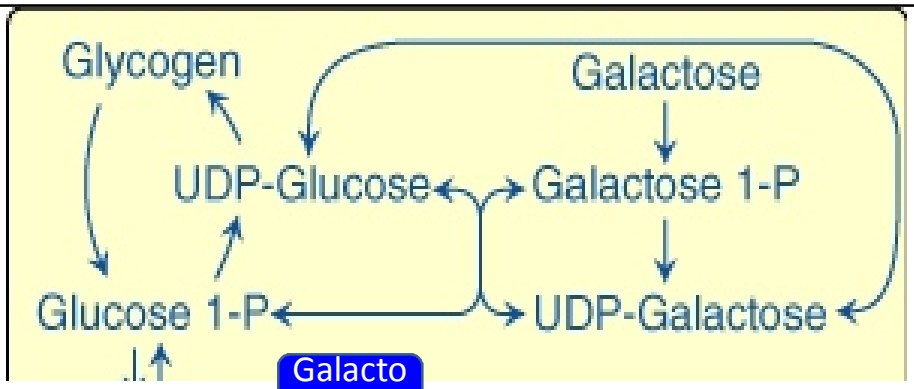


UDP Galactose; an Intermediate in Galactose Metabolism



Galactose Metabolism

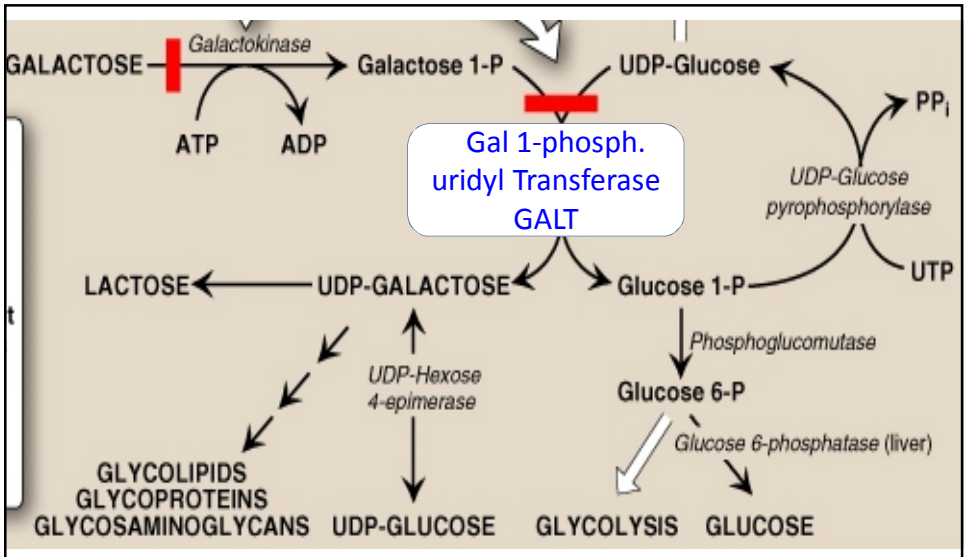




Galactose + ATP $\xrightarrow{\text{Galacto kinase}}$ Gal. 1-Phosph ADP

Gal 1-Phosph + UDP Glc. $\xrightarrow{\text{transferase}}$ UDP- Galactose + Glc1-Phosph

UDP Galactose $\xrightarrow{\text{epimerase}}$ UDP Glucose

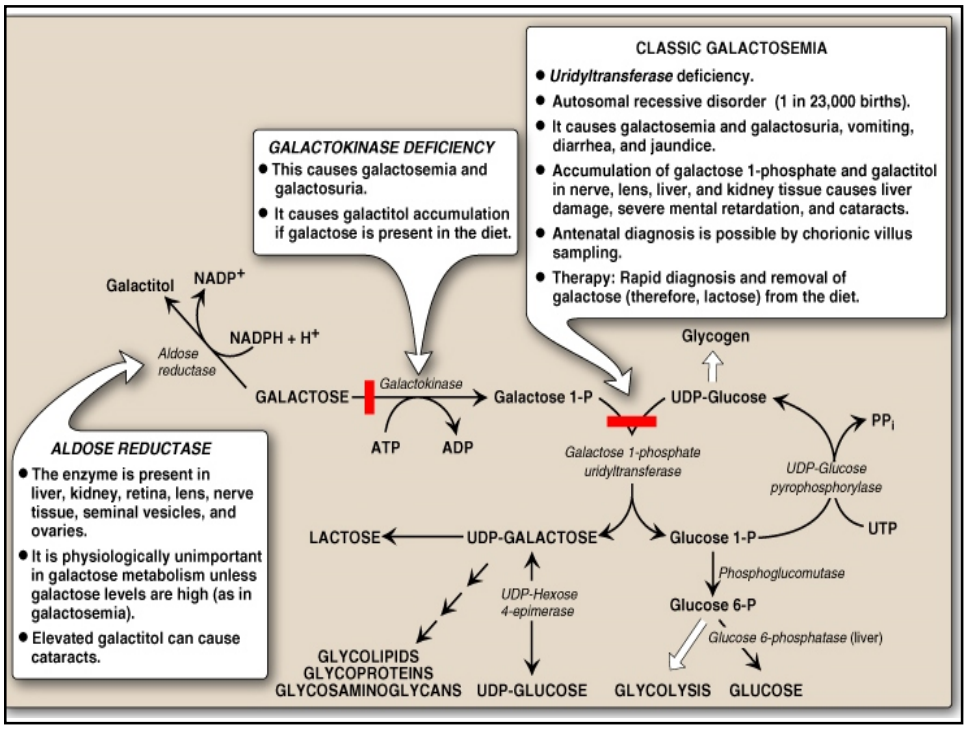


Disorders of Galactose Metabolism

- Deficiency of GALT ➔ classic Galactosemia
- Accumulation of Galactose 1-Phosphate and galactose
- Similar consequences to those in fructose intolerance

- Galactose ••••••➔ Galactitol
- Deficiency of Galactokinase

- Accumulation of Galactose ••••••➔ Galactitol



Lactose Synthesis

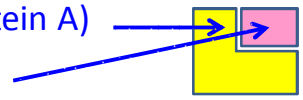
- Lactose is Galactosyl β (1→4) glucose
- Galactosyl β (1→4) glucose is found in glycolipids and glycoproteins

UDP Gal. + Glucose $\xrightarrow{\text{Lactose Synthase}}$ Lactose + UDP

- Lactose Synthase: complex of 2 proteins

Galactosyl transferase (Protein A)

α-lactalbumin (Protein B)



In glycolipids synthesis

UDP-Gal + N acetyl glucosamine $\xrightarrow{\text{Protein A}}$ N-acetyllactosamine