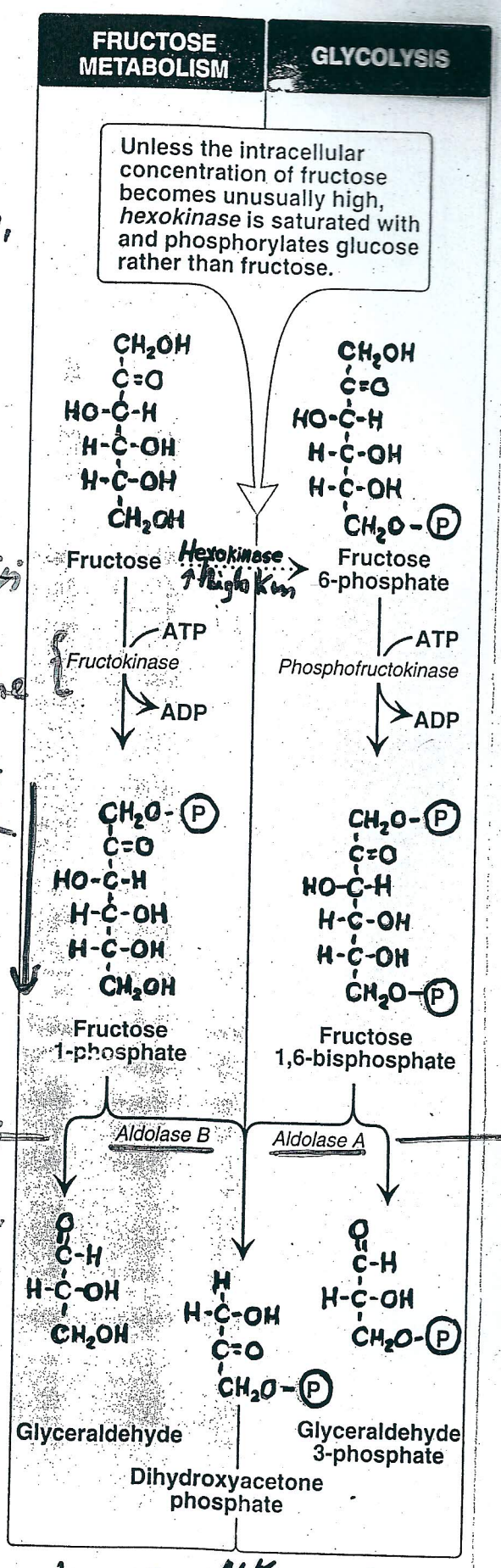
Phosphorylation of fructose and its cleavage

fructose → 10% of daily calories (~50 gr/d)

sucrose, corn syrup, fruits & honey



found in liver kidney intestine

found in liver kidney intestine

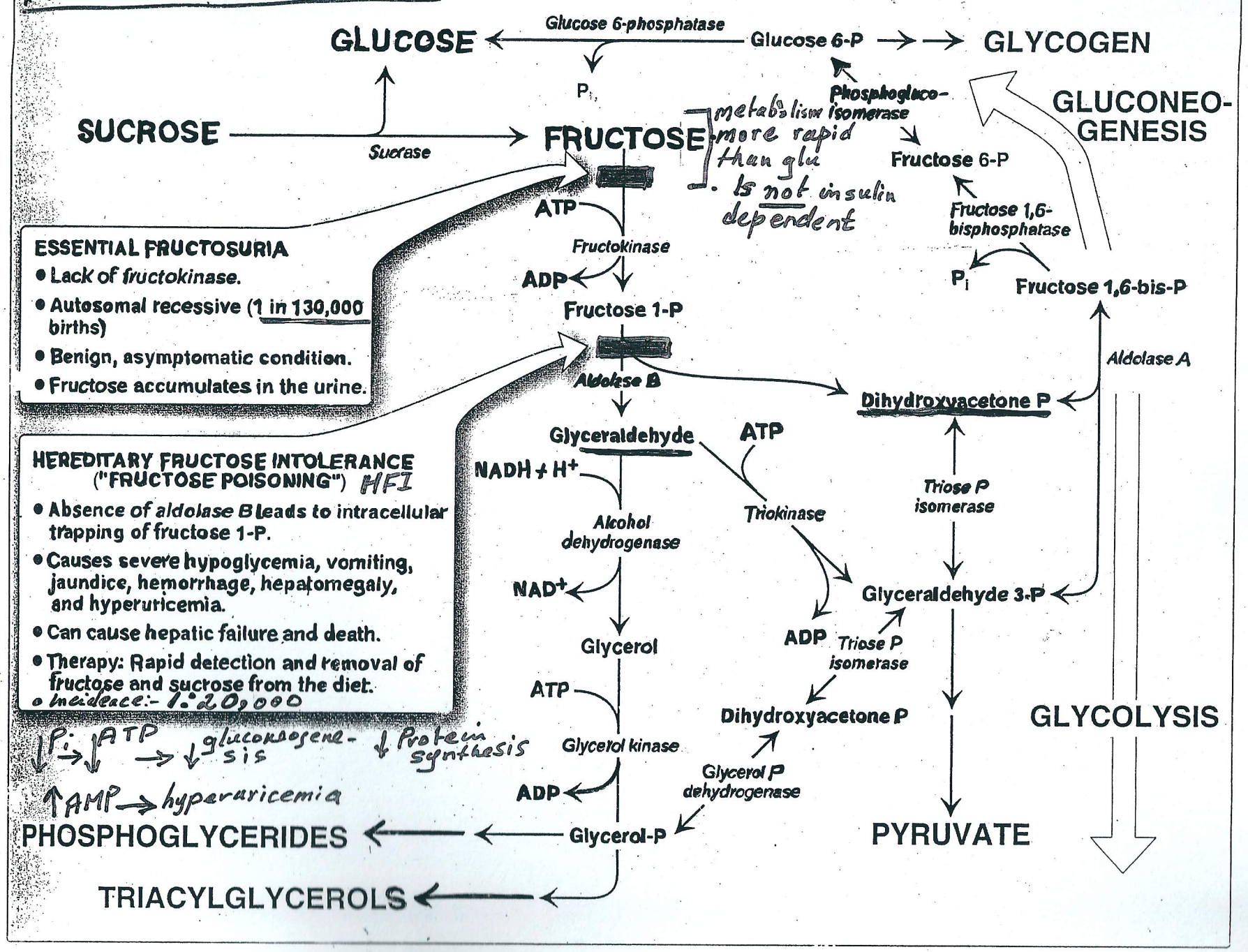
found in all tissues

Mannose (C-2 epimer) + ATP —HK→ Mannose-6-P

Most intracellular need is from fructose

PMI

FRUCTOSE METABOLISM



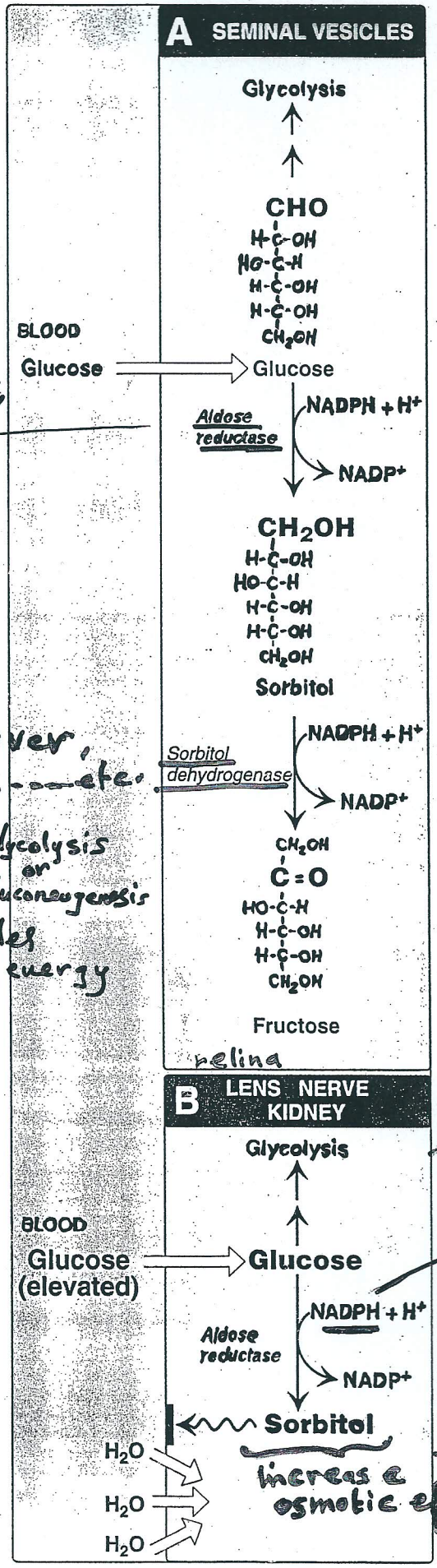
ESSENTIAL FRUCTOSURIA

- Lack of *fructokinase*.
- Autosomal recessive (1 in 130,000 births)
- Benign, asymptomatic condition.
- Fructose accumulates in the urine.

HEREDITARY FRUCTOSE INTOLERANCE ("FRUCTOSE POISONING") HFI

- Absence of *aldolase B* leads to intracellular trapping of fructose 1-P.
- Causes severe hypoglycemia, vomiting, jaundice, hemorrhage, hepatomegaly, and hyperuricemia.
- Can cause hepatic failure and death.
- Therapy: Rapid detection and removal of fructose and sucrose from the diet.
- Incidence:- 1:20,000

SORBITOL METABOLISM



Found in many tissues that include:- lense, retina, peripheral nerves, liver, Kidney, placenta, rbc, ovaries & seminal vesicles

active in liver, ovaries, seminal vesicles, ---- etc.

- Sorbitol in liver → Fru → Glycolysis or gluconeogenesis
- Sorbitol in seminal vesicles → Fru → major source of energy for sperms

Activity of sorbitol dehydrogenase low or absent

also resulting in a decrease in GSH antioxidant

Cataract
Peripheral neuropathy
nephropathy
retinopathy

increase osmotic effects

Metabolism of Galactose

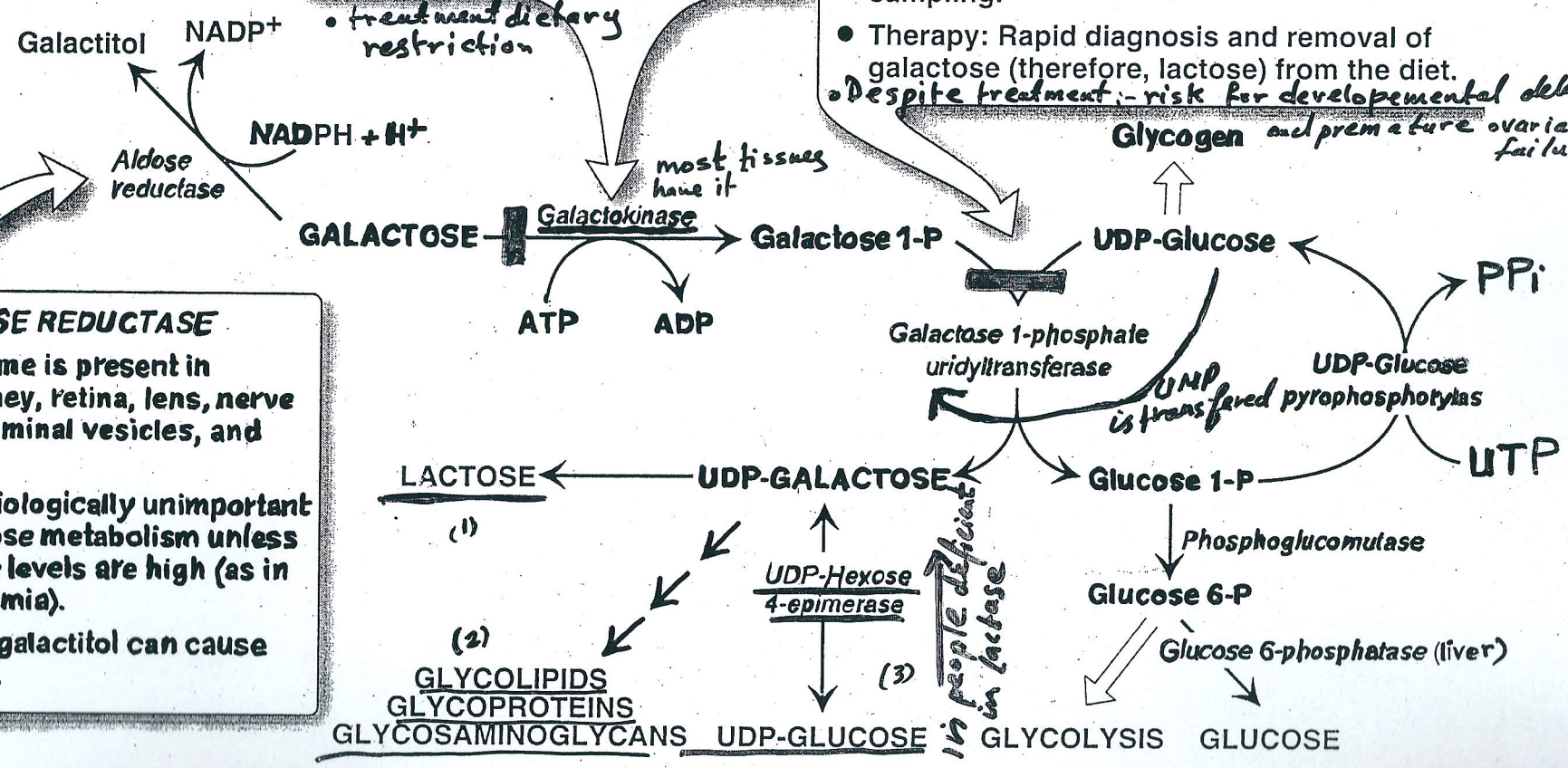
(as in fru, transport into cells is Not insulin-dependent)

GALACTOKINASE DEFICIENCY

- This causes galactosemia and galactosuria.
- It causes galactitol accumulation if galactose is present in the diet.
- rare autosomal recessive
- treatment dietary restriction

CLASSIC GALACTOSEMIA

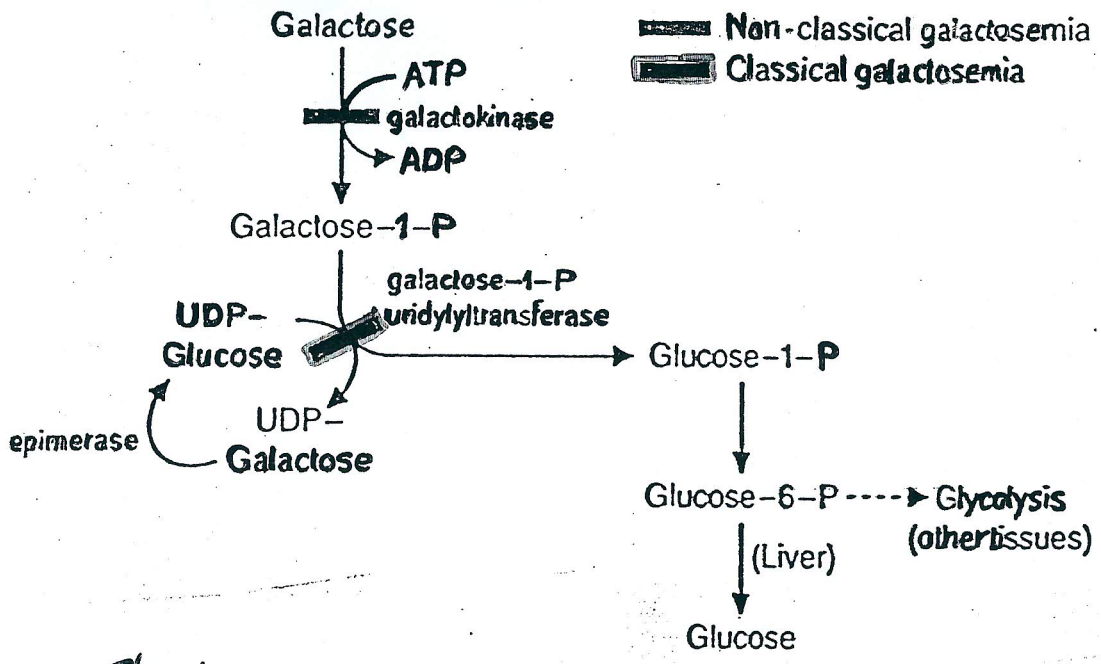
- *Uridyltransferase* deficiency.
- Autosomal recessive disorder (1 in 23,000 births).
- It causes galactosemia and galactosuria, vomiting, diarrhea, and jaundice.
- Accumulation of galactose 1-phosphate and galactitol in nerve, lens, liver, and kidney tissue causes liver damage, severe mental retardation, and cataract.
- Antenatal diagnosis is possible by chorionic villus sampling.
- Therapy: Rapid diagnosis and removal of galactose (therefore, lactose) from the diet.
- Despite treatment - risk for developmental delays and premature ovarian failure

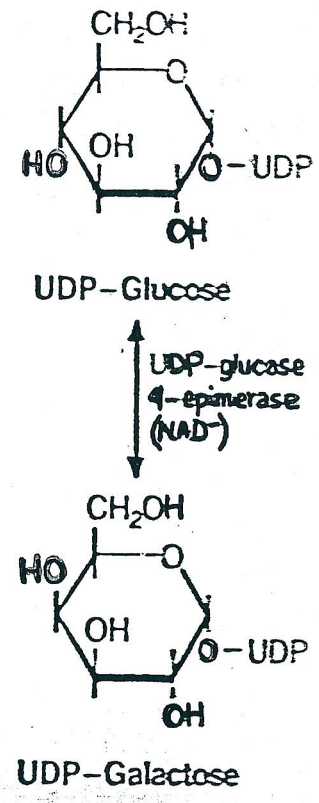


ALDOSE REDUCTASE

- The enzyme is present in liver, kidney, retina, lens, nerve tissue, seminal vesicles, and ovaries.
- It is physiologically unimportant in galactose metabolism unless galactose levels are high (as in galactosemia).
- Elevated galactitol can cause cataracts.

Galactose Metabolism:





Galactosemia :-

Gal. and Gal-1-P ↑

Gal ⟶ Galaticol ↑

- mental retardation
- Visual cataracts
- liver enlargement

Deficiency :- Transferase :- Accounts for most of cases
Galactokinase
Epimerase

- Lactose Synthesis :-

$$\text{UDP-Gal} \xrightarrow{\text{L.S.}} \text{Lactose}$$

Glu

L.S. = membrane bound $\overbrace{\text{Galactosyltransferase}}^{\text{EnzI}}$ and α-lactalbumin Complex

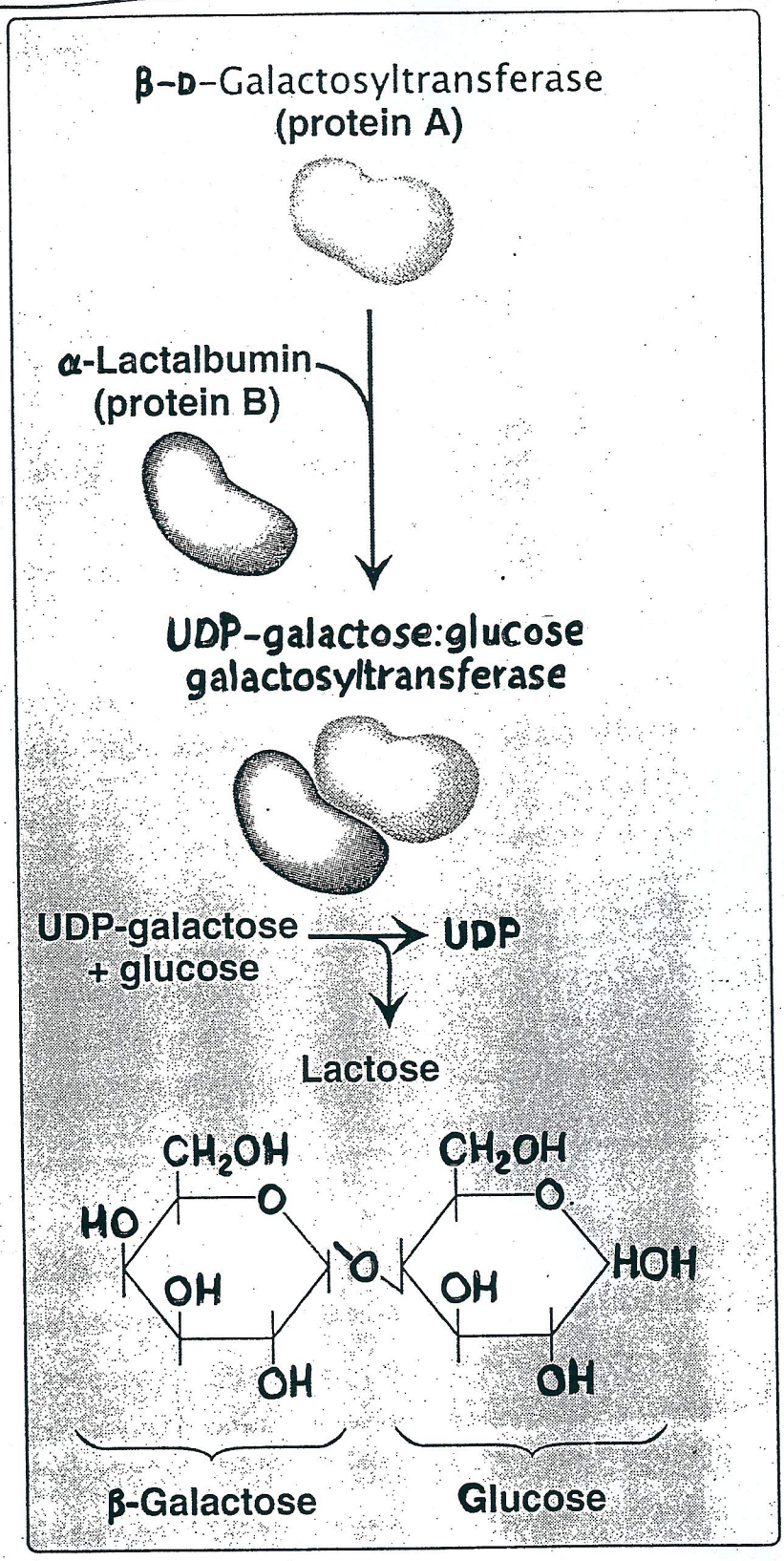
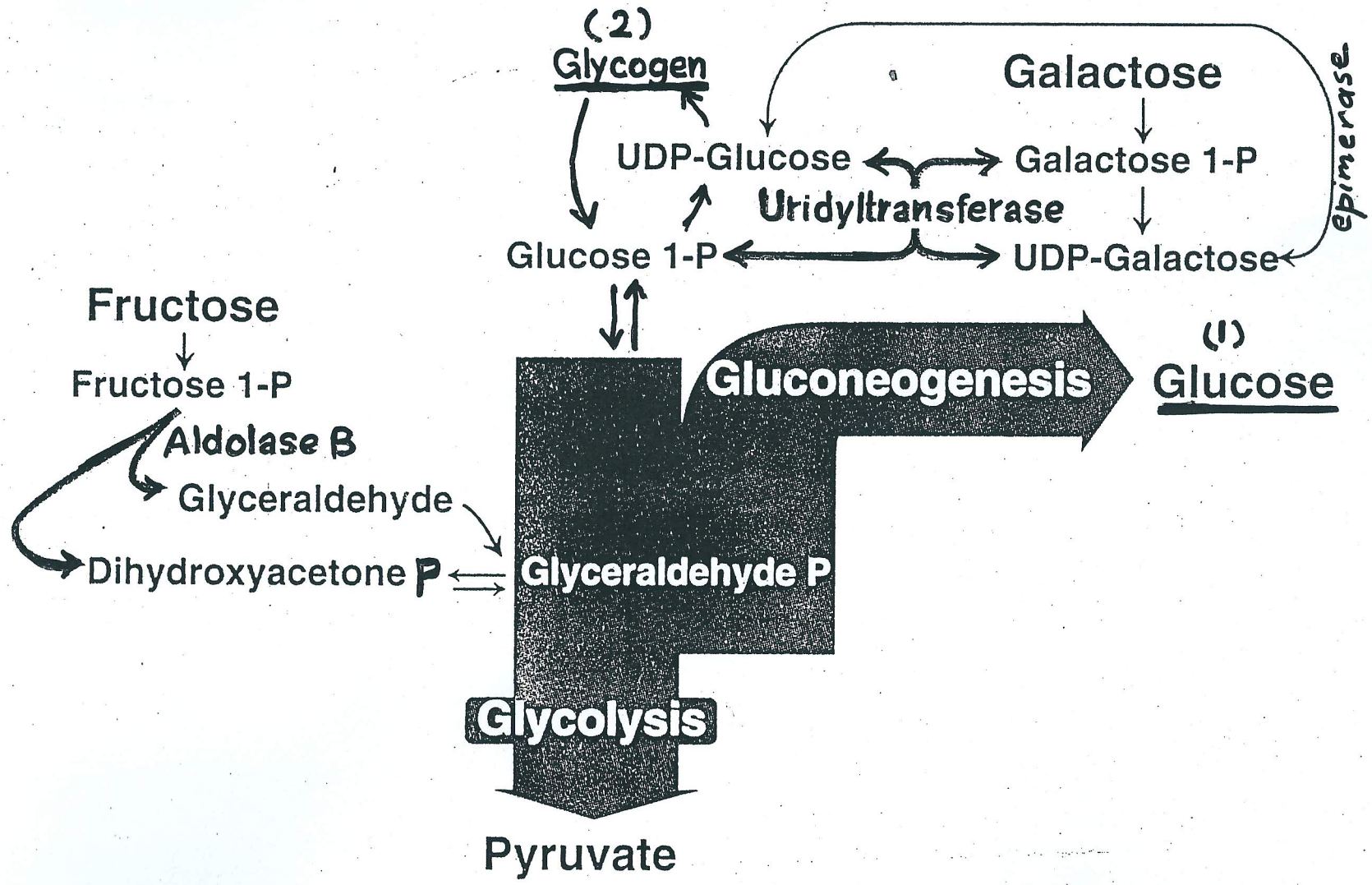
UDP-Gal + Glc-N-acetyl---- Protein

$\downarrow$ EnzI

Gal(1→4)-Glu-N-acetyl---- Protein

- α-lactalbumin Lowers the Km of Enz I from 1200 mM to 1mM

LACTOSE SYNTHESIS



Key reactions for metabolism of fructose and Galactose