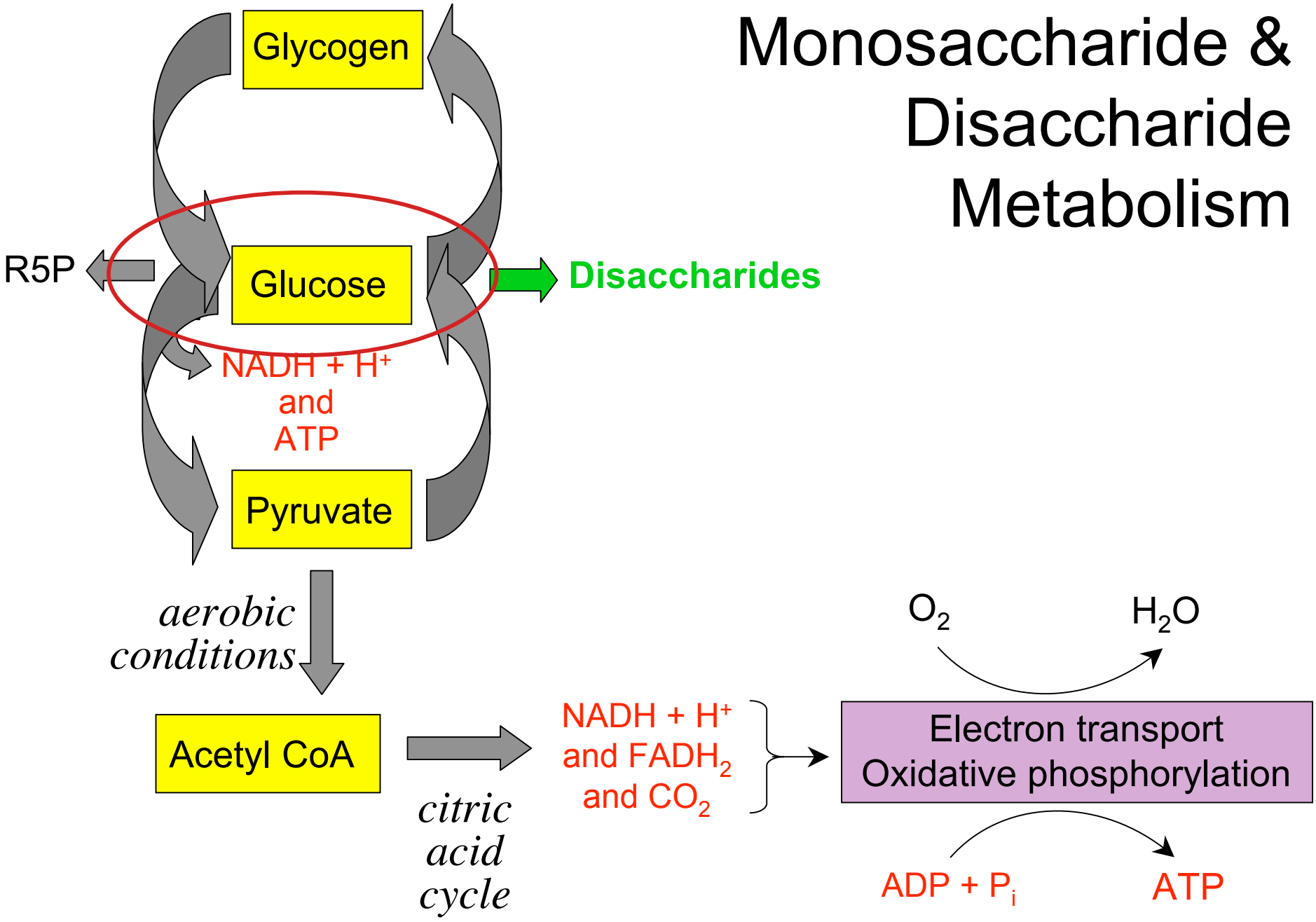
Key knowledge base & conceptual questions

- What are the two major disaccharides consumed in human diets? Know the differences in structure between these disaccharides and the monosaccharide building blocks that composed them.
- Outline how disaccharide breakdown products are shunted to glycolysis and gluconeogenesis.
- What are the two major mechanisms by which cells retain fructose and galactose for metabolism?
- Understand how aldolase B and uridyltransferase reactions are critical for fructose and galactose metabolism.
- Be able to explain different inborn errors in carbohydrate metabolism, which gene products are responsible for the disease, and cellular/clinical outcomes of the disease.
- Recognize that there are no clinical treatments for patients challenged in carbohydrate metabolism - the only recourse after diagnosis is removal of the carbohydrate from the diet

Monosaccharide & Disaccharide Metabolism



Artificial sweeteners

Sugar substitute: food additive to duplicate sugar taste but with less food energy

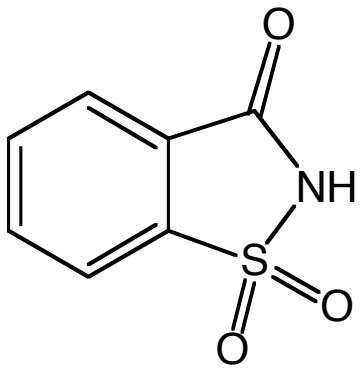
Sweetness of compounds many fold greater than glucose or sucrose

Ongoing controversy over health risks

Artificial sweeteners

Saccharin

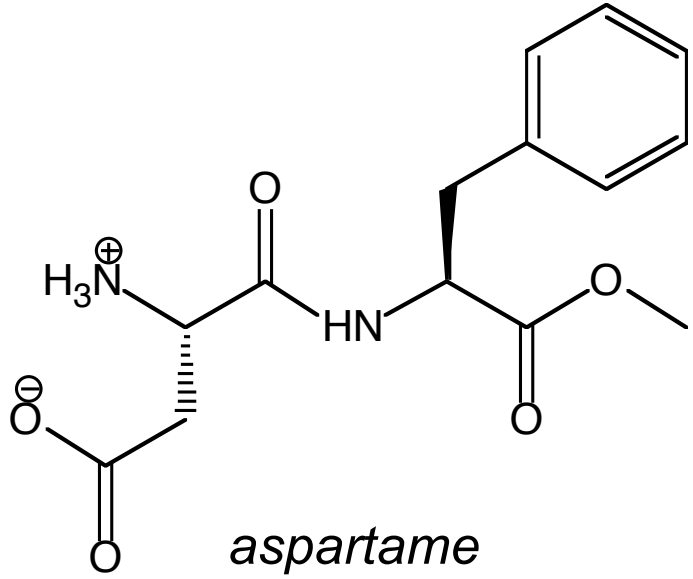
- discovered in 1879
- 300X sweeter than sucrose
- no food energy



saccharin

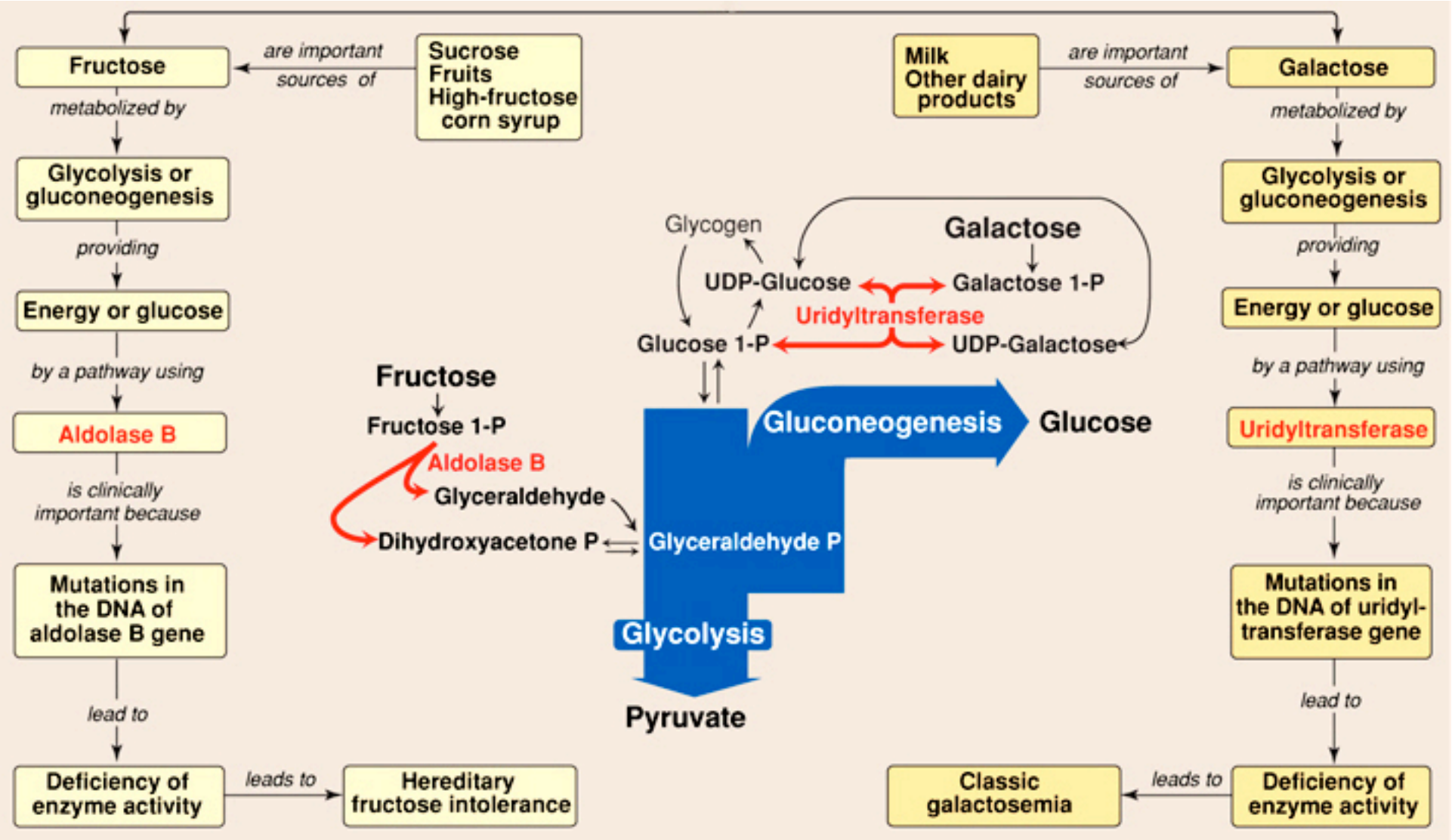
Aspartame

- Discovered in 1965
- 180X sweeter than sucrose



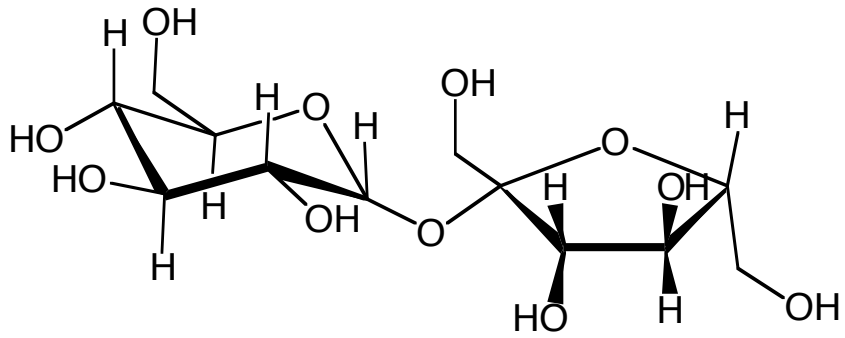
aspartame

Important Dietary Monosaccharides



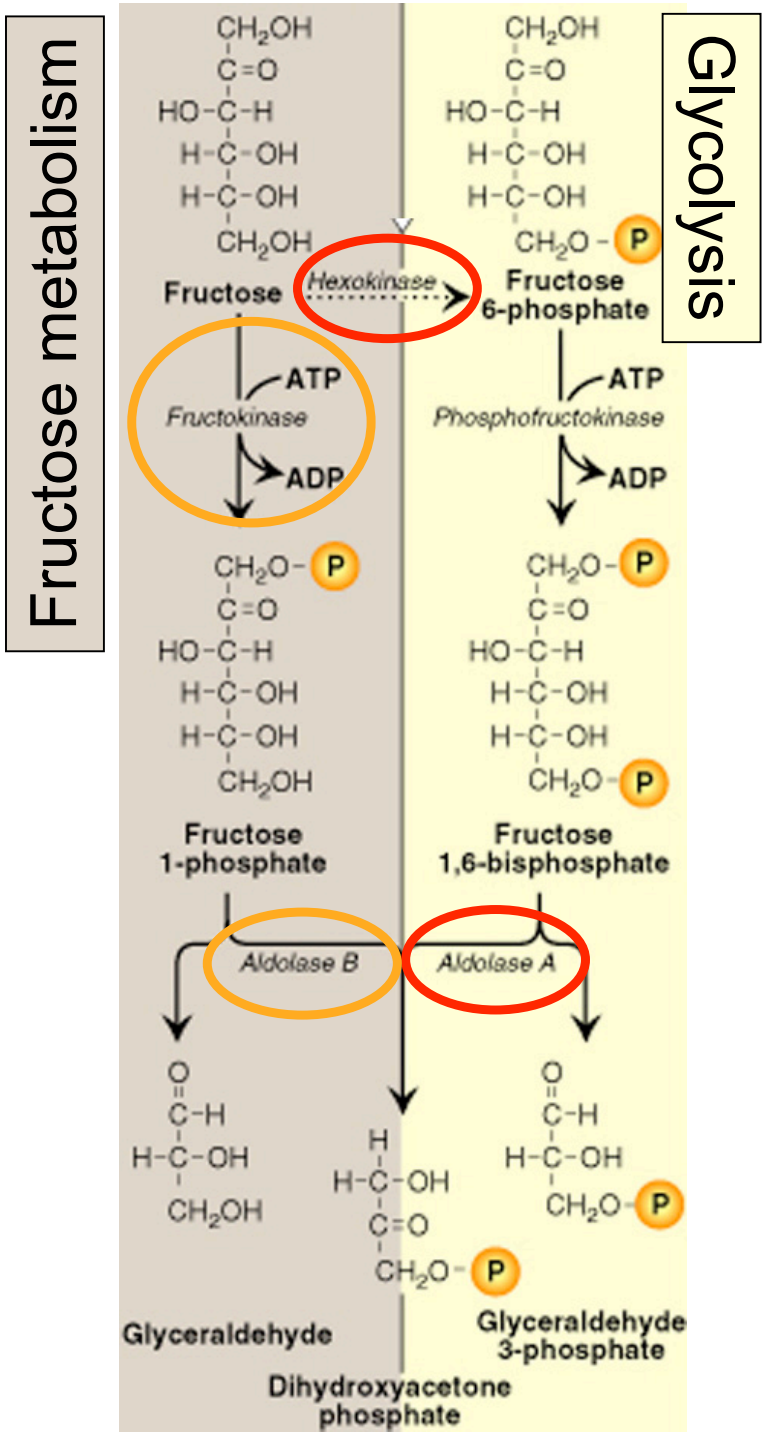
Fructose metabolism

- Typical Western diet contains 50 g fructose/day
- Monosaccharide products of sucrose are



glucose *fructose*

- Fructose found in corn syrup, fruits, and honey
- Insulin-independent

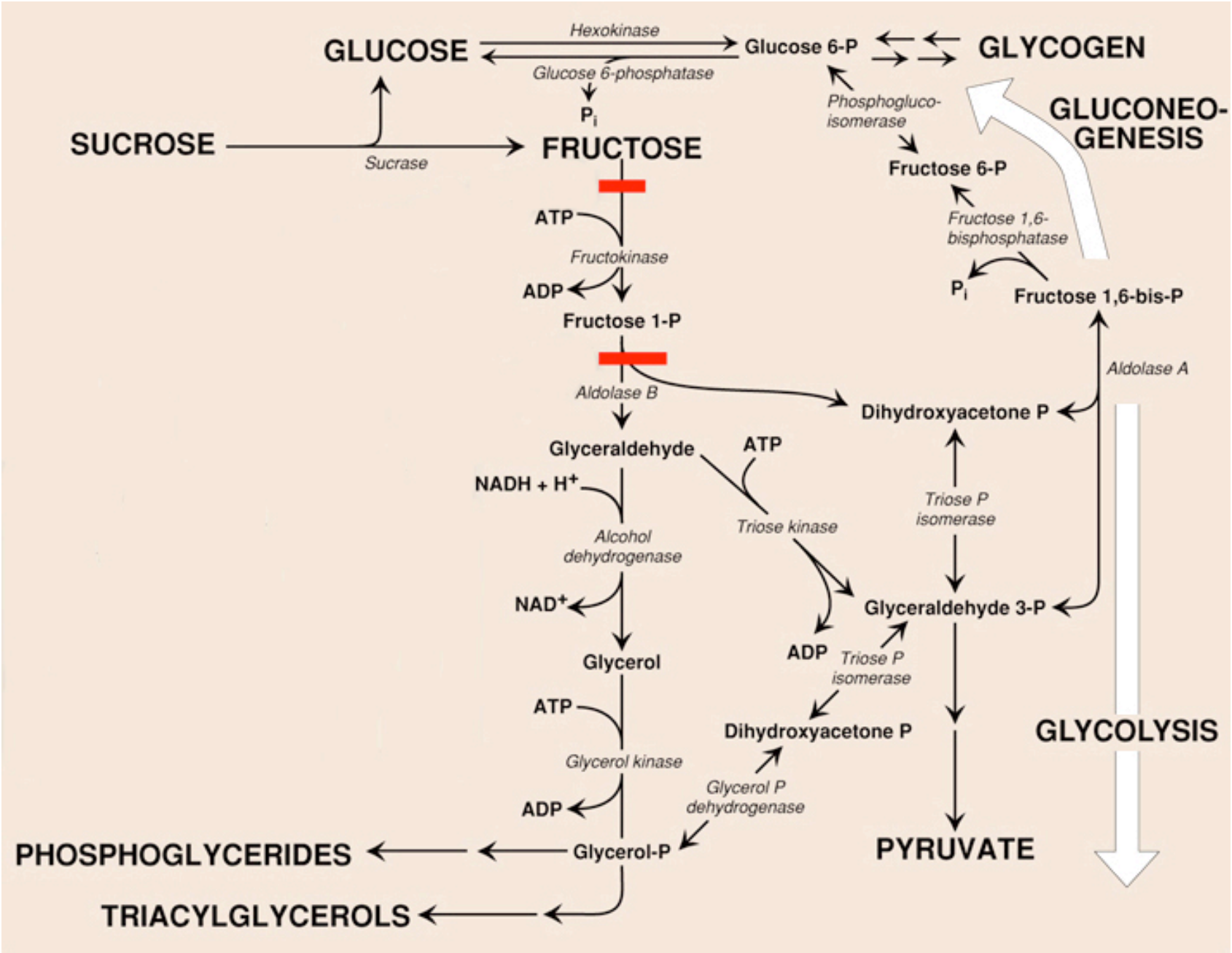


Phosphorylated products of fructose

Either hexokinase or fructokinase can covalently modify fructose.

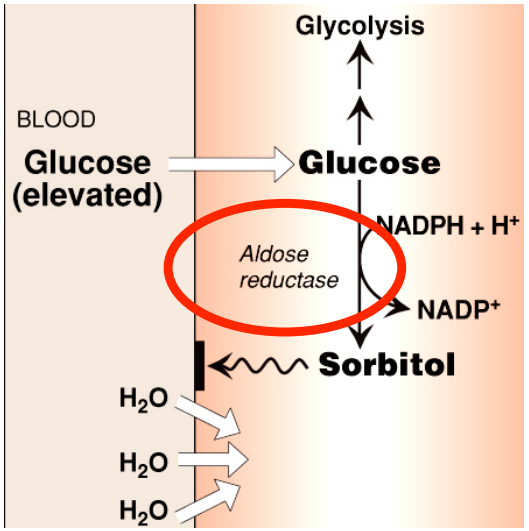
Hexokinase phosphorylates glucose rather than fructose, unless [fructose] is unusually high.

Fructokinase found in particular tissues.

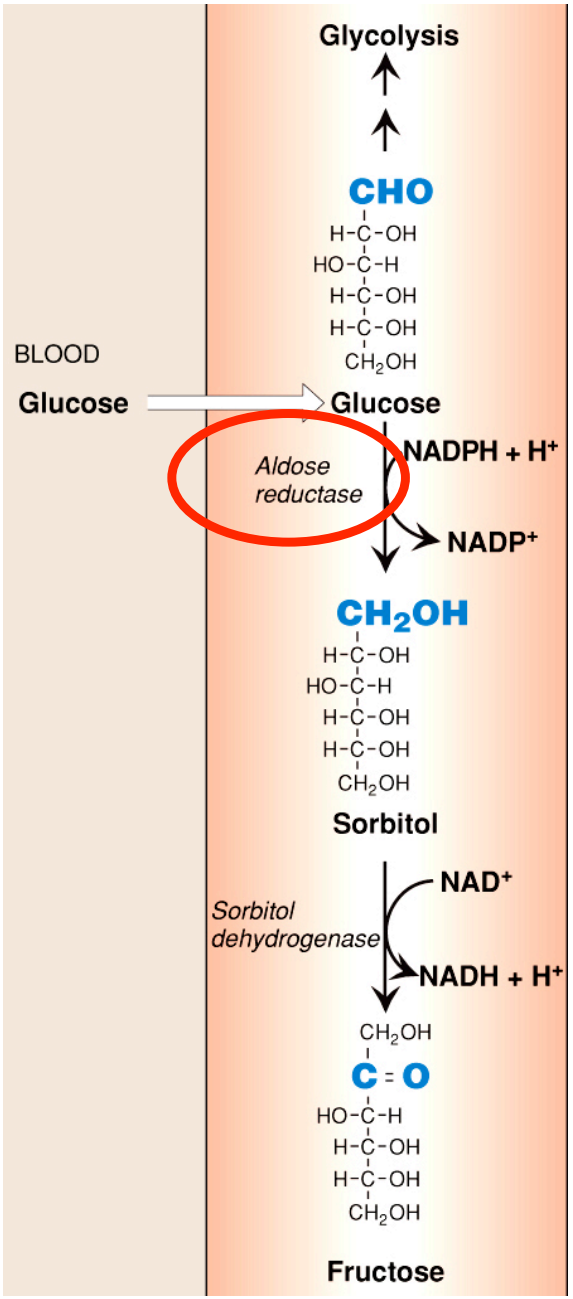


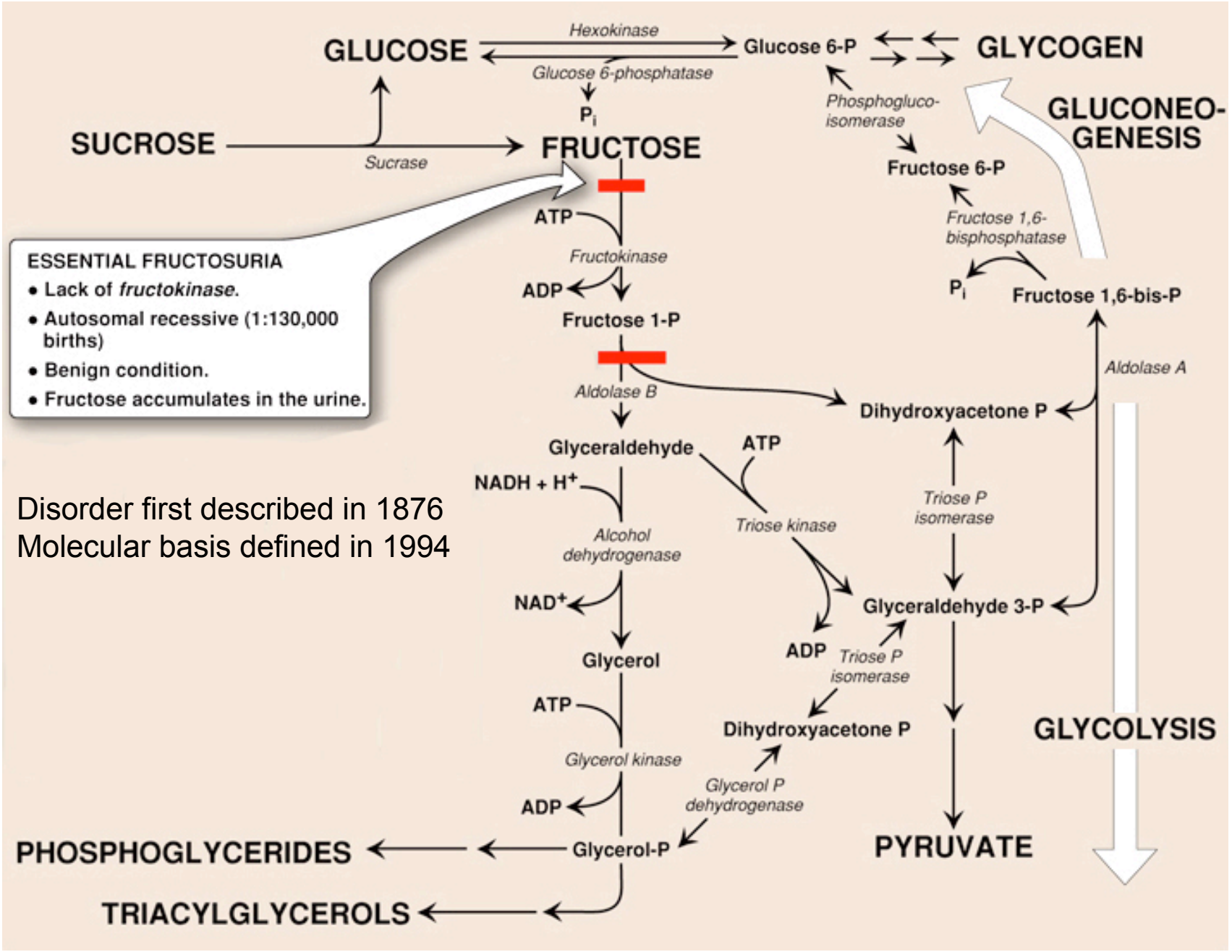
Sorbital metabolism

Lens, kidney, nerve

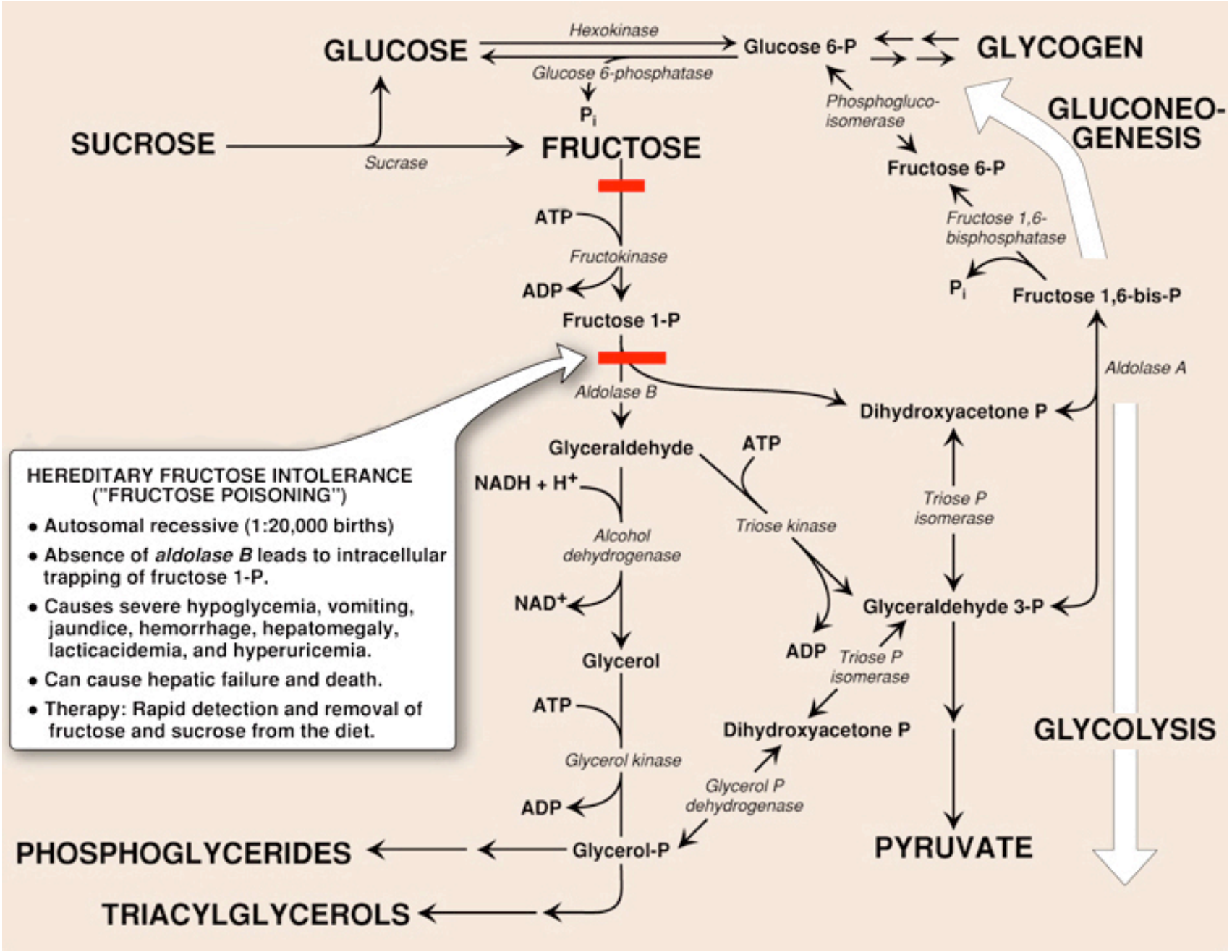


Seminal vesicles

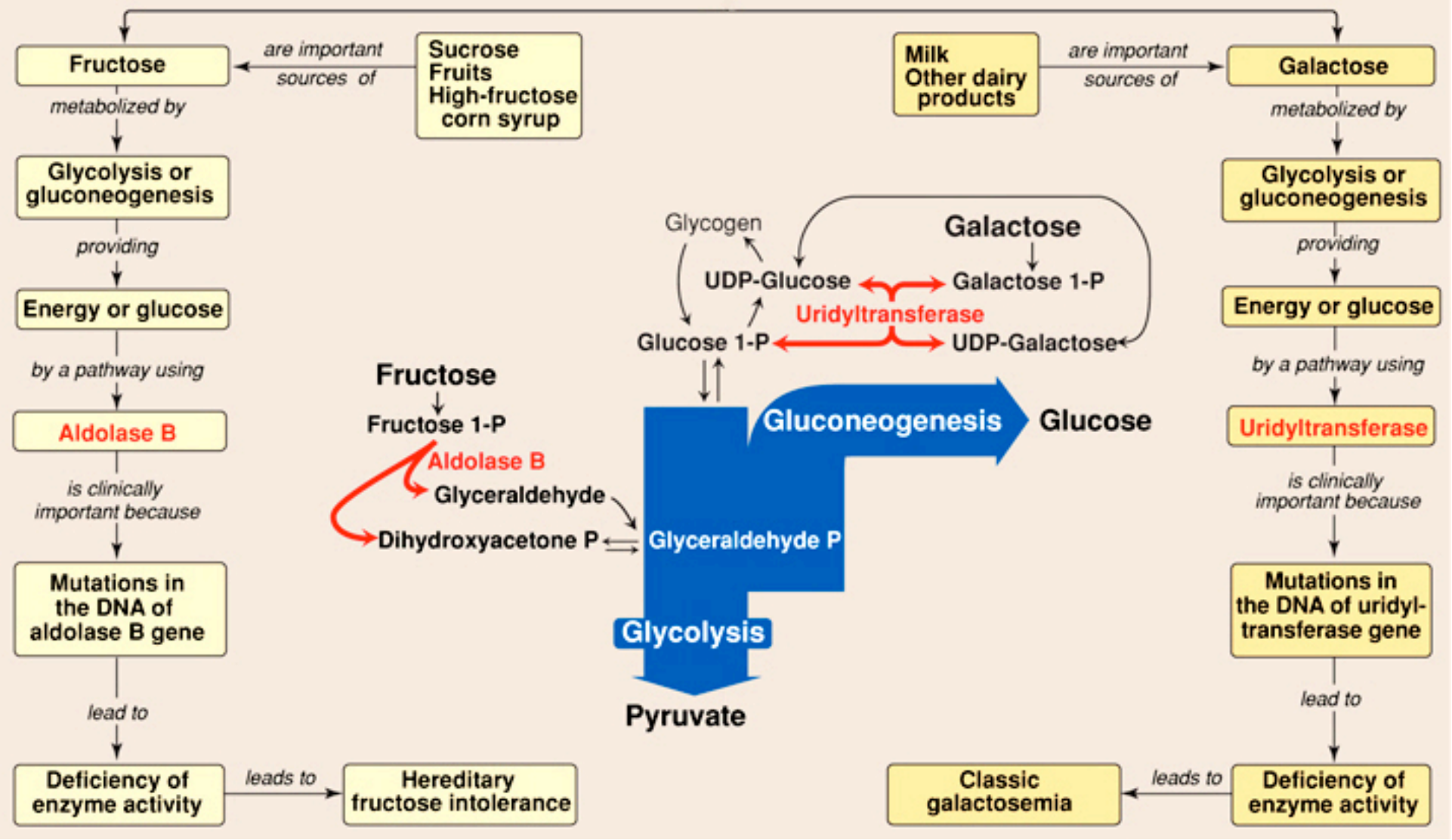




Disorder first described in 1876
Molecular basis defined in 1994

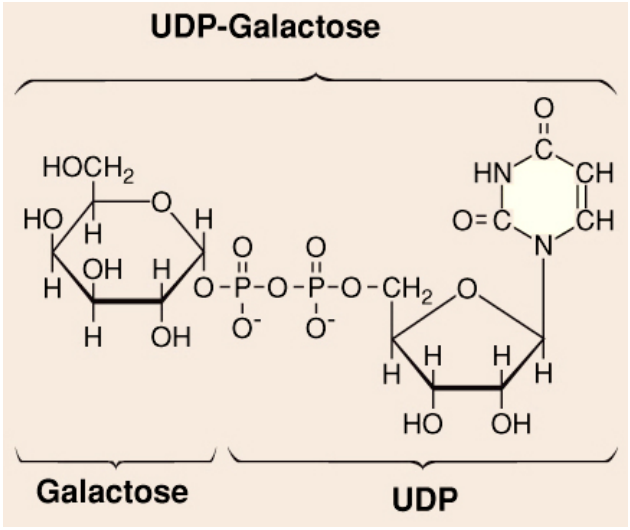


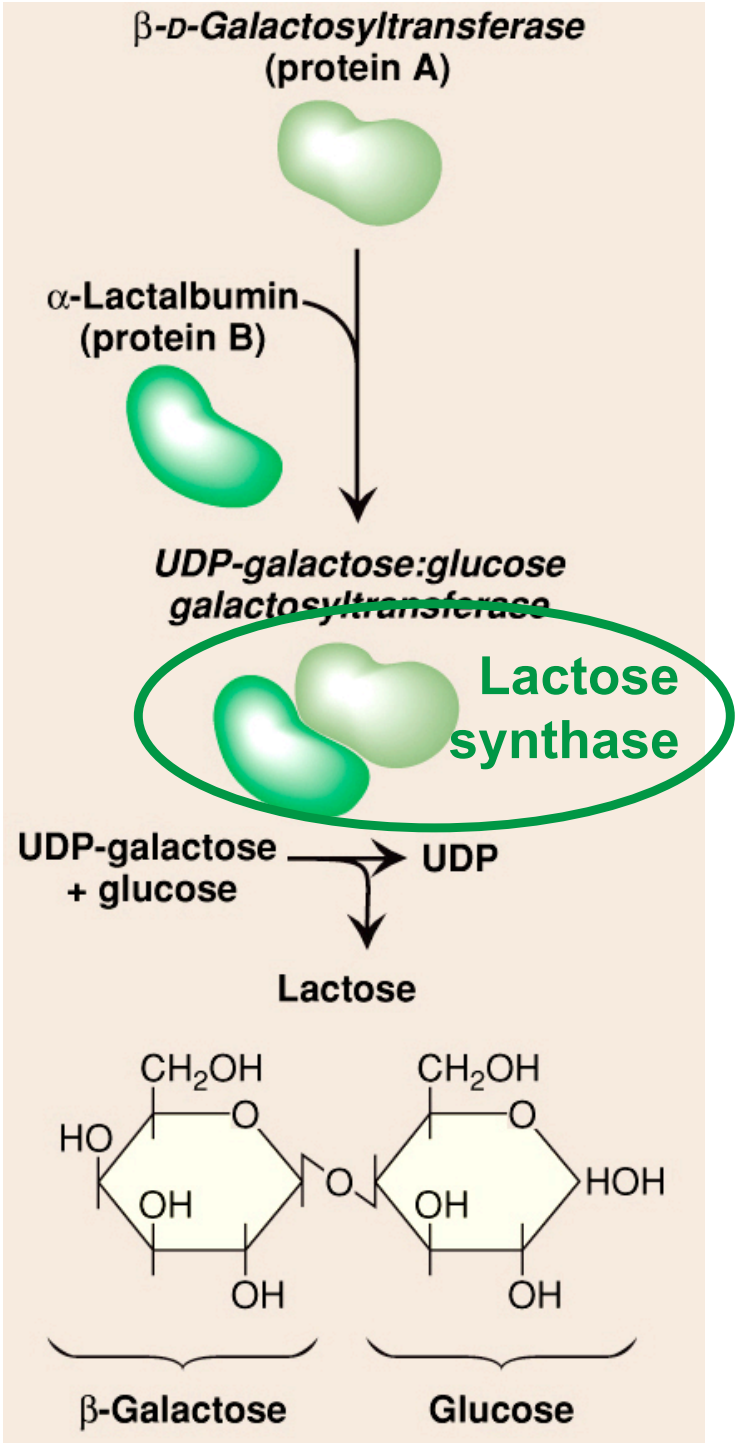
Important Dietary Monosaccharides



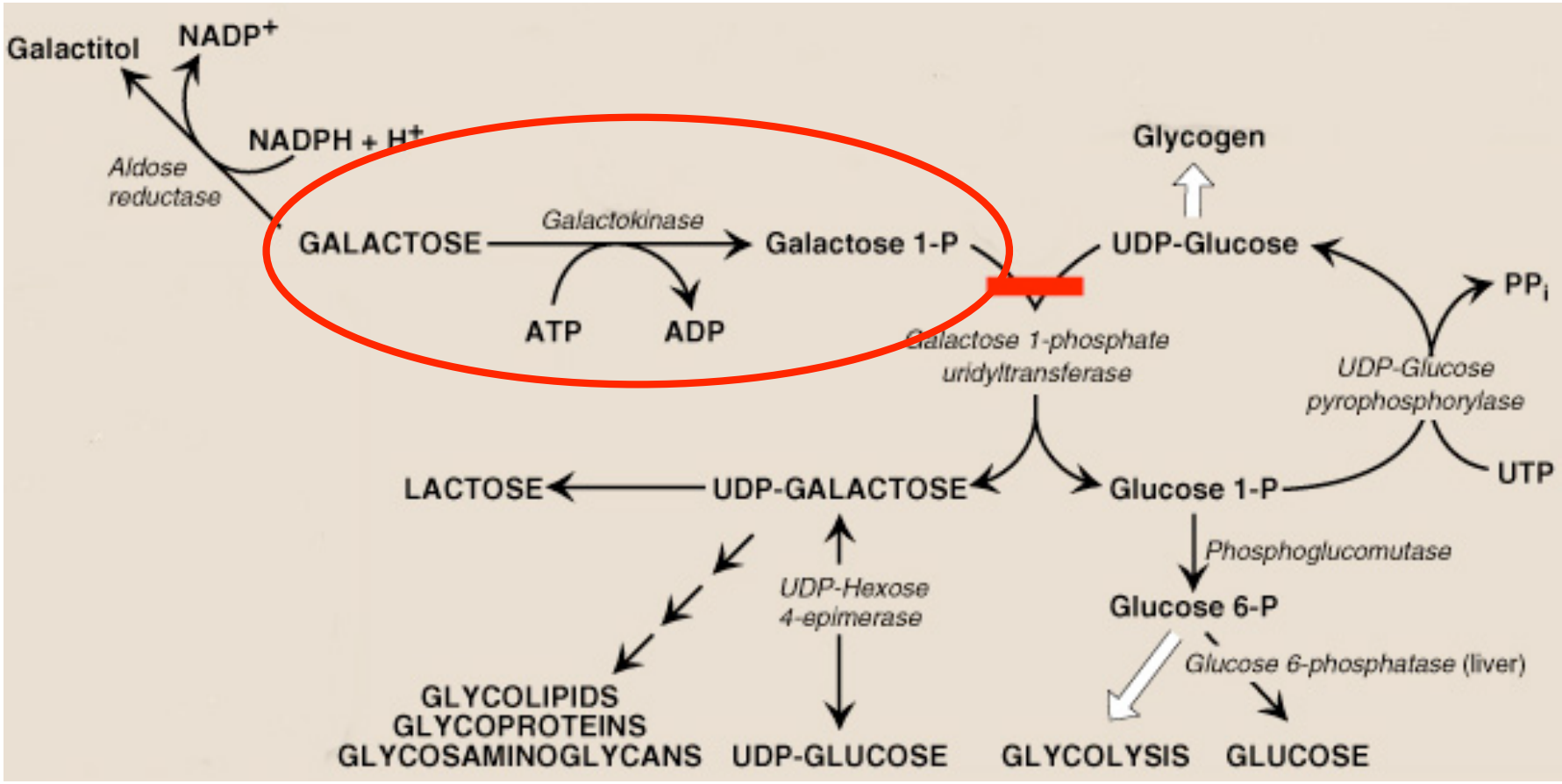
Lactose Synthesis

- Occurs in ER of human mammary glands
- Lactose synthase is a complex of two enzymes
- UDP-galactose and glucose are required substrates
- Products are UDP and lactose

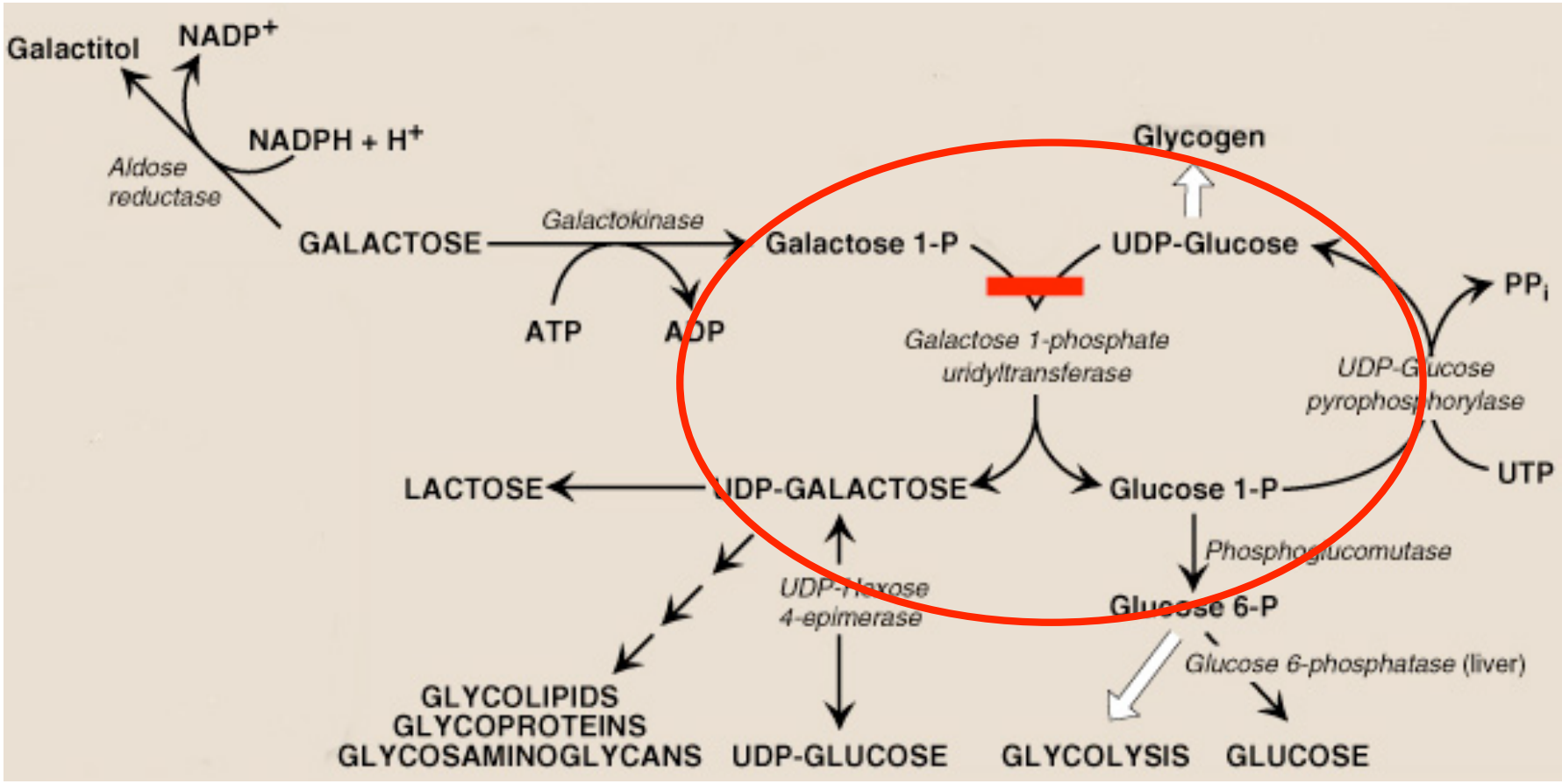




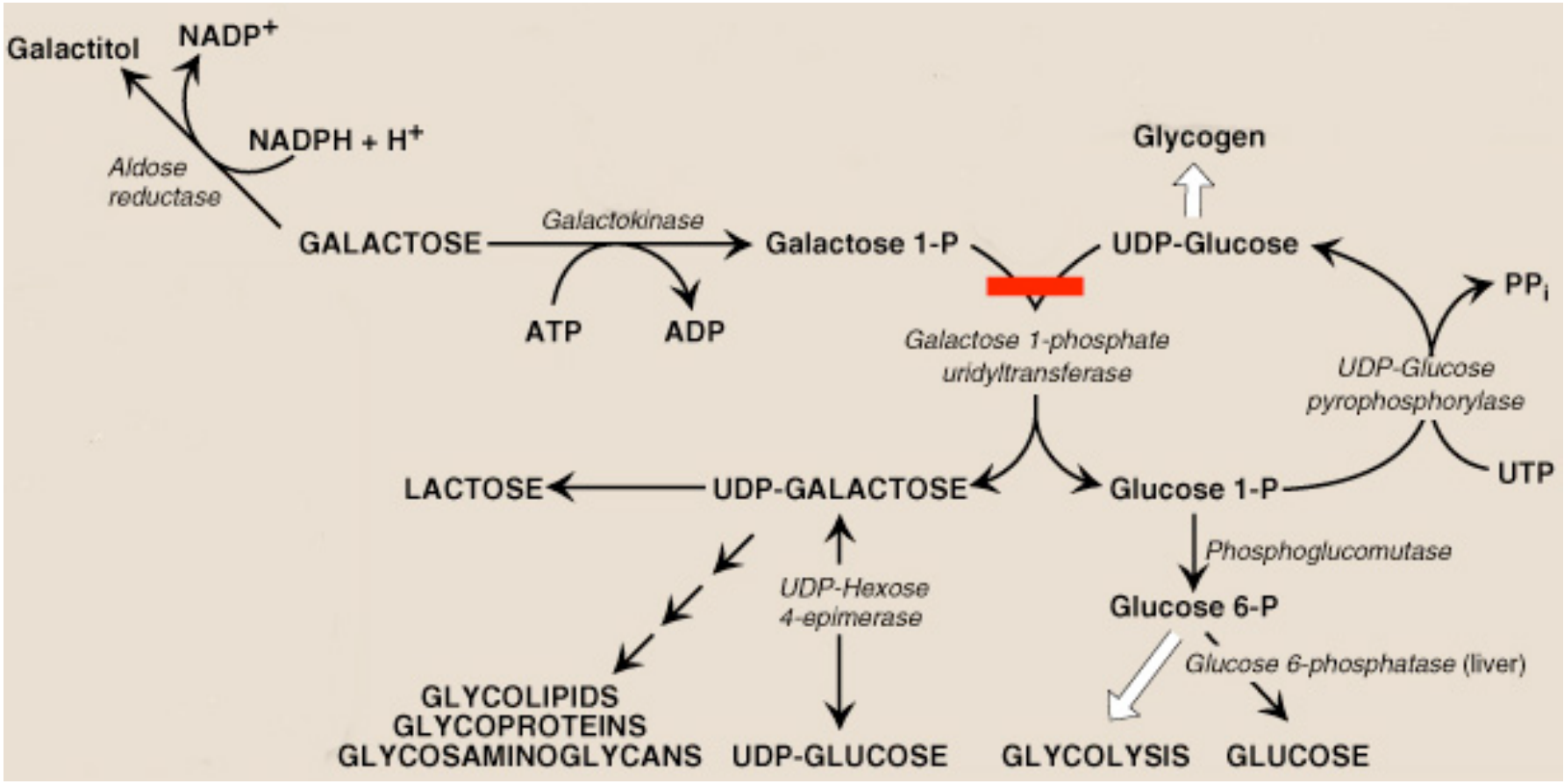
Galactose metabolism



Galactose metabolism



Galactose metabolism disorders



Classic galactosemia

Uridyltransferase deficiency
Autosomal recessive disorder (1 in 23,000 births)
Symptoms include vomiting, diarrhea, and jaundice, liver damage, severe mental retardation, and cataracts
Therapy: rapid diagnosis and removal of galactose (lactose) from diet