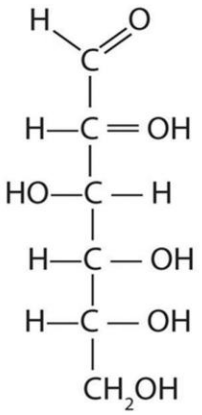
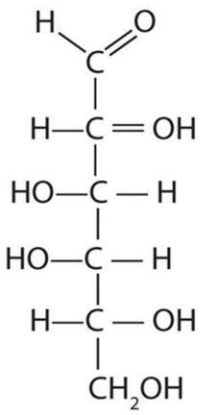
Other source of carbohydrate

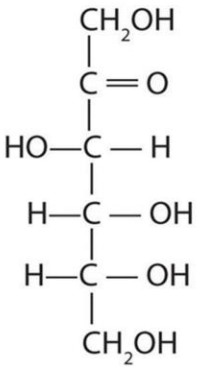
DR. Bariaa Ali



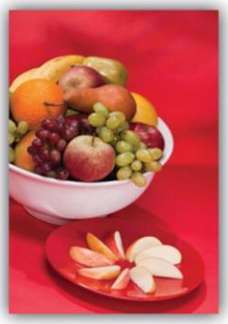
D-(+)-glucose



D-(+)-galactose



D-(–)-fructose



Sources of Sugars

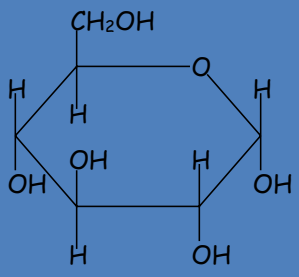
-**Glucose:** lactose (dairy products) and sucrose (table sugar)

-**Fructose:** fruits and sucrose

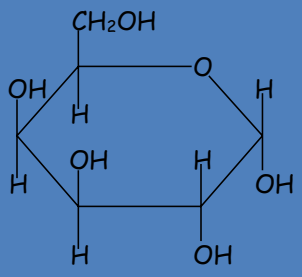
-**Galactose:** lactose

-**Mannose:** polysaccharides and glycoproteins

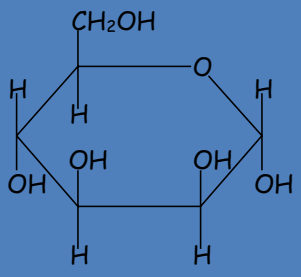
Common Hexoses



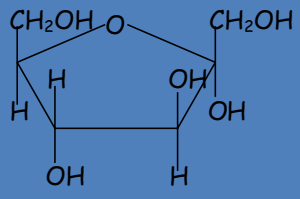
a-D-Glucose



a-D-Galactose

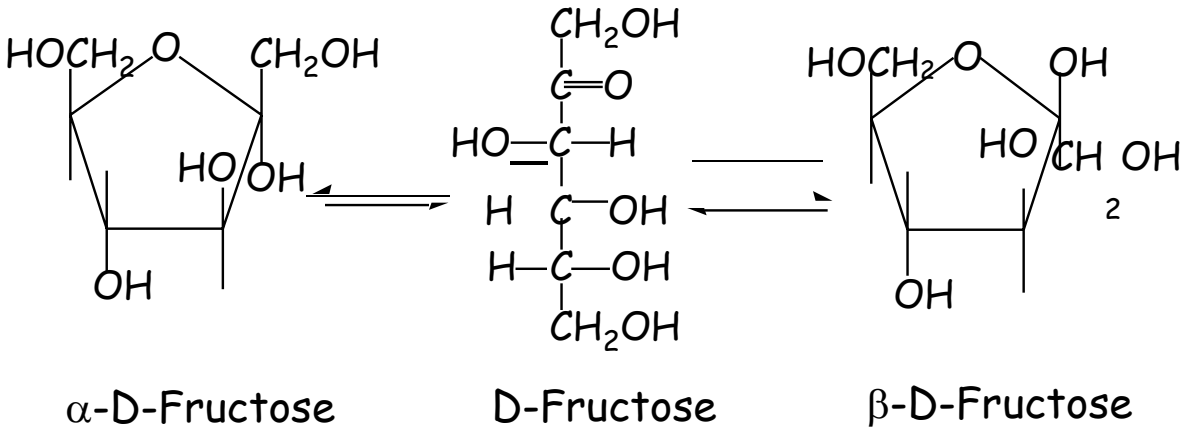


a-D-Mannose



a-D-Fructose

Mutarotation of Fructose



α-D-Fructose D-Fructose β-D-Fructose

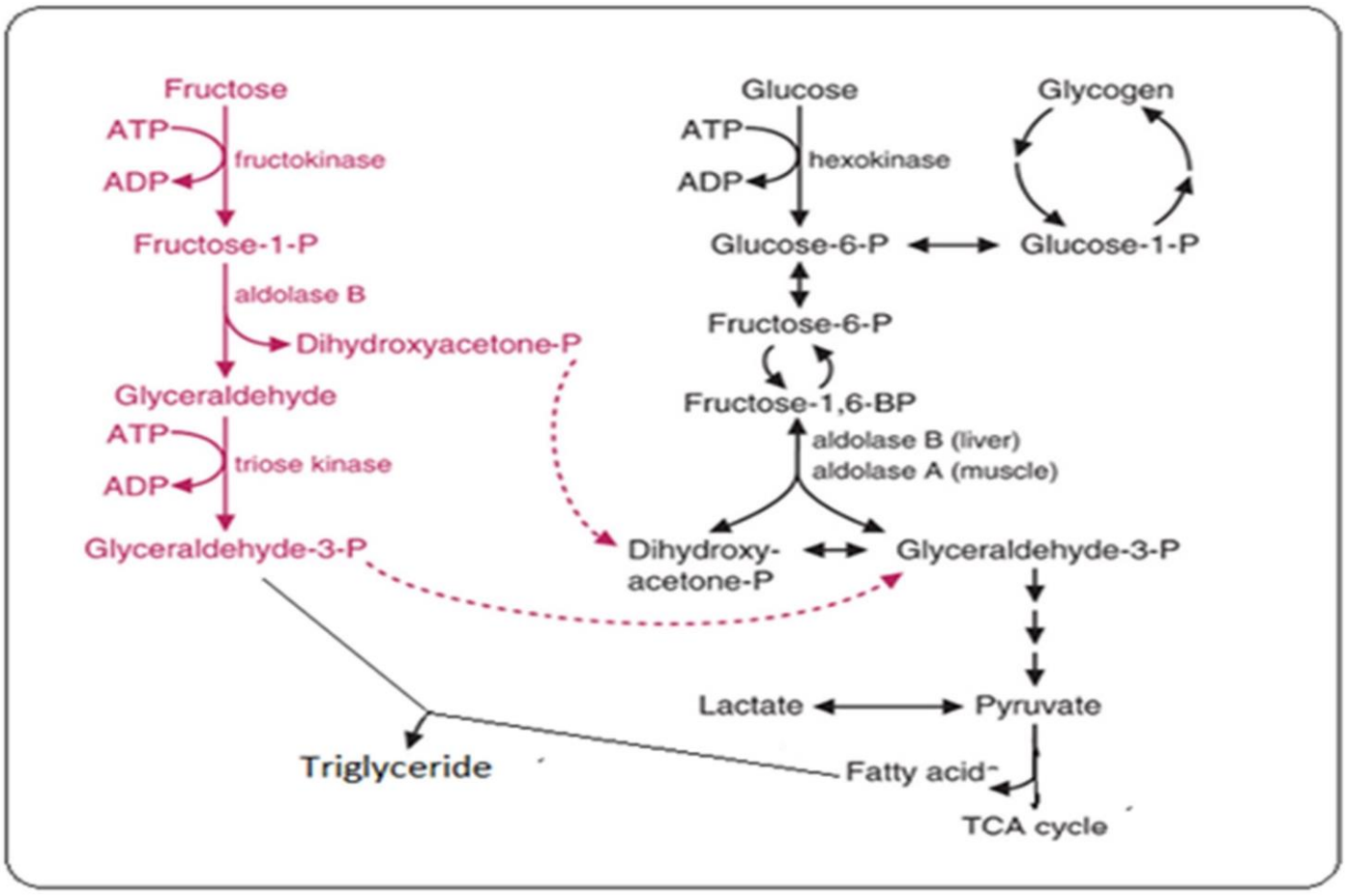
Fructose Metabolism

Fate of fructose:

Although glucose is the most abundant monosaccharide derived from the diet, fructose is also usually obtained in significant quantities. The major dietary source of fructose is the disaccharide sucrose in table sugar and fruit, or can be oxidized to pyruvate and reduced to lactate,.

, but it is also present as the ..The monosaccharide in corn syrup, which is used as a sweetenermetabolism of fructose at this point yields intermediates in gluconeogenic pathway leading to glycogen synthesis

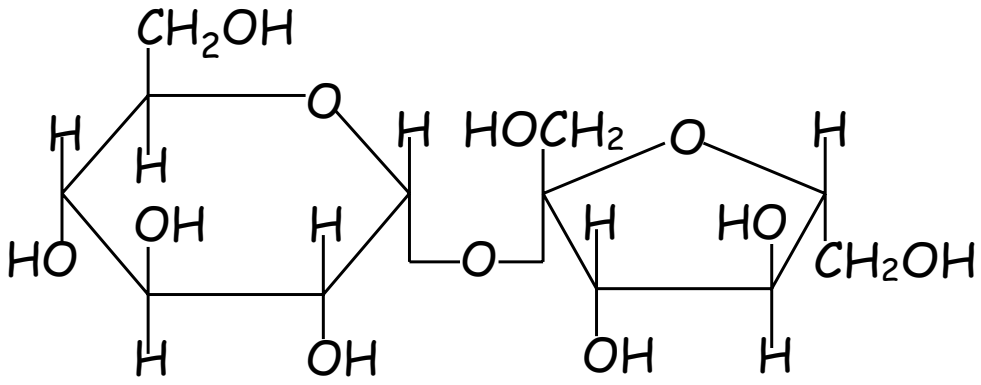
or be decarboxylated to acetyl CoA in the mitochondria and directed toward the synthesis of free fatty acid, resulting finally in triglyceride synthesis



Fate of Fructose

Sucrose

(Table Sugar)



O-α-D-Glucopyranosyl-(1—> 2)-β-D-Fructofuranoside

All Tissues

Sucrose ⟶ α-D-glucose + β-D-fructose ⟶ α-D-fructose

a-Glucosidase
(Invertase)

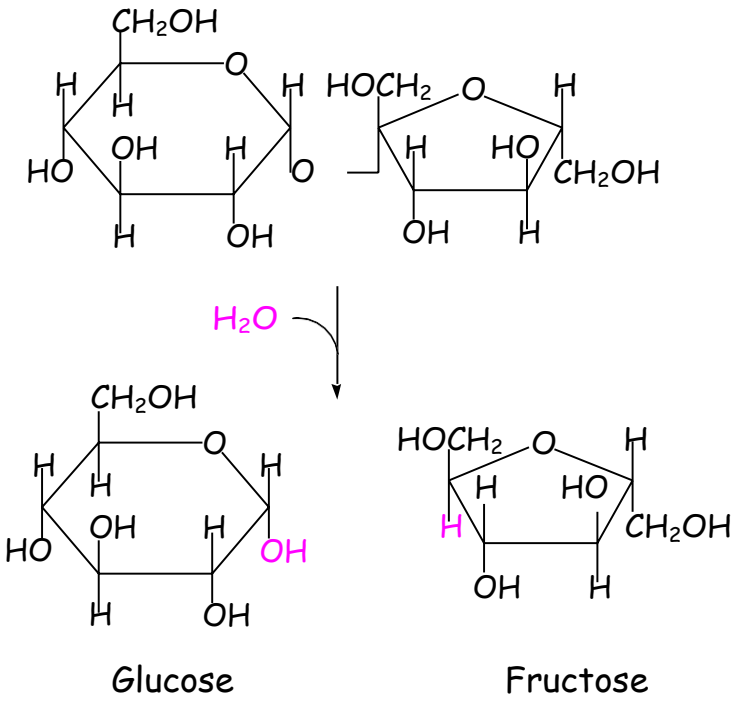
Mutarotation

↓

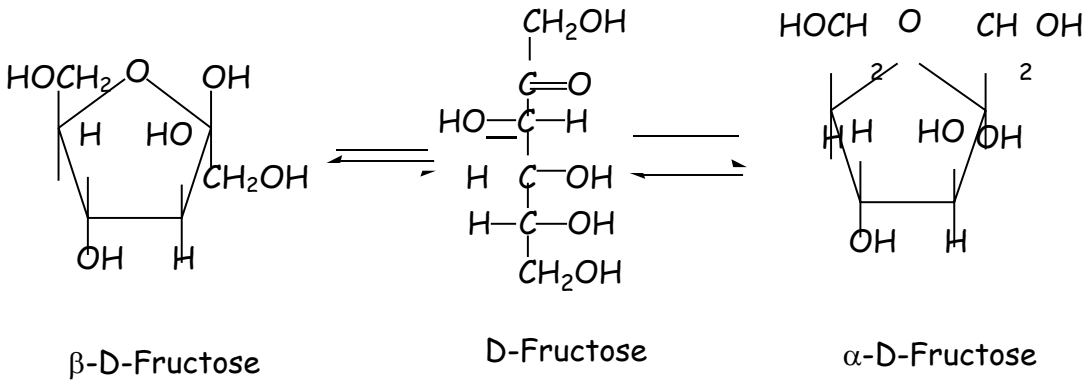
Glycolysis

Cleavage of Sucrose

(β-glucosidase or invertase)



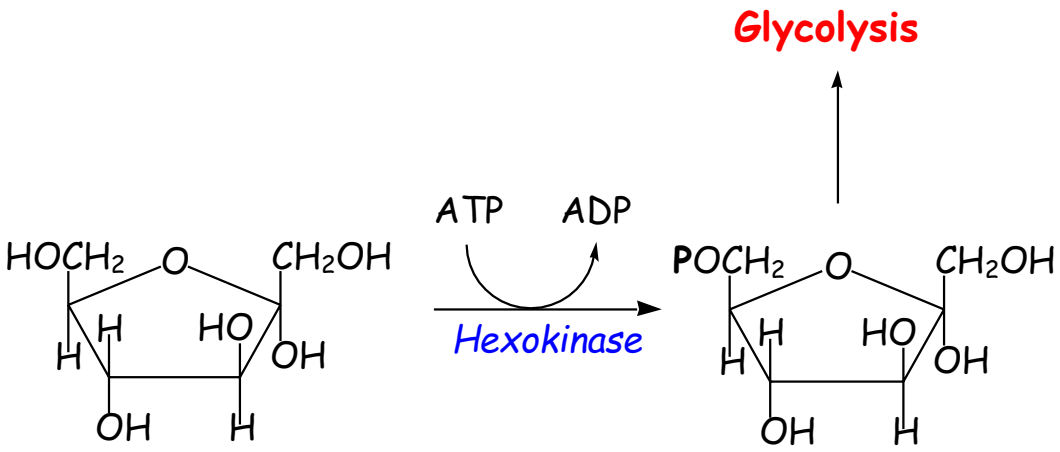
Mutarotation of Fructose



Muscle Metabolism of Fructose

(Anaerobic Glycolysis)

Large Amounts of Hexokinase

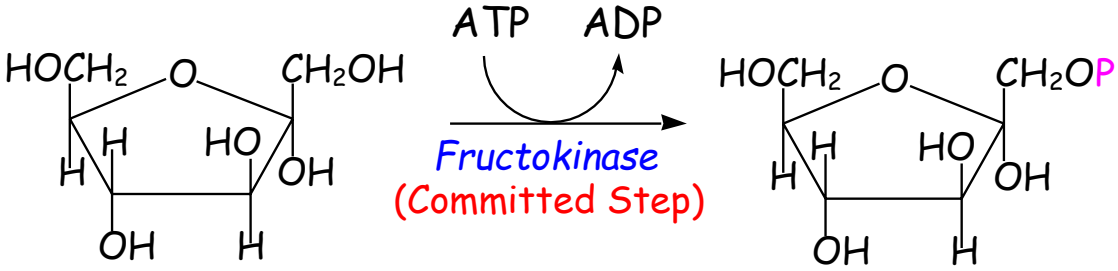


α-D-Fructose

Fructose-6-P

Liver Metabolism of Fructose I

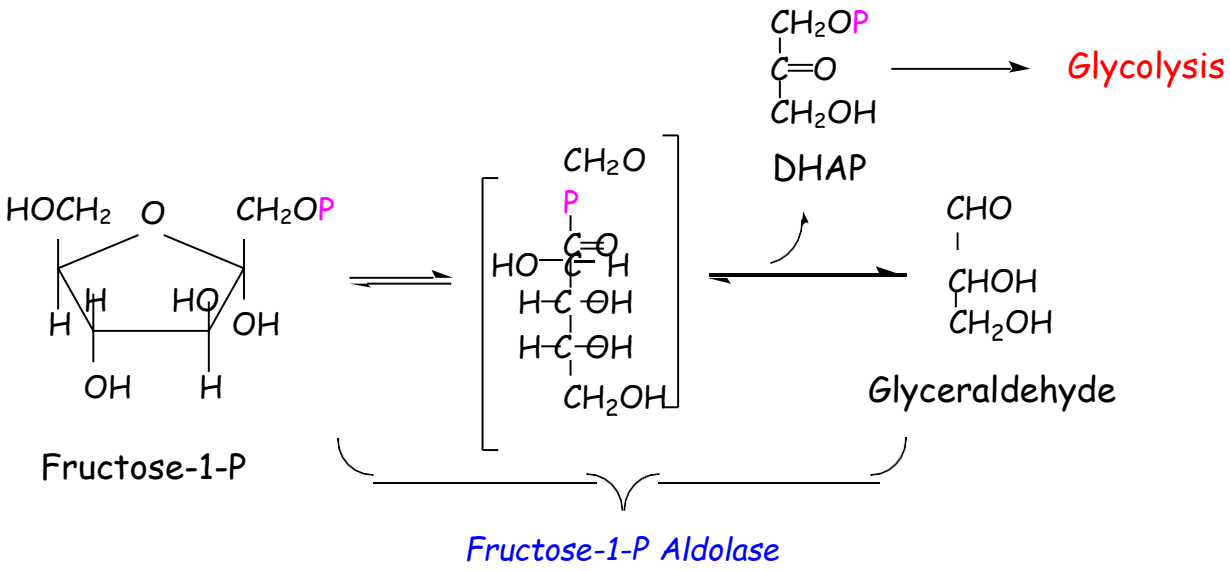
(Little Hexokinase)



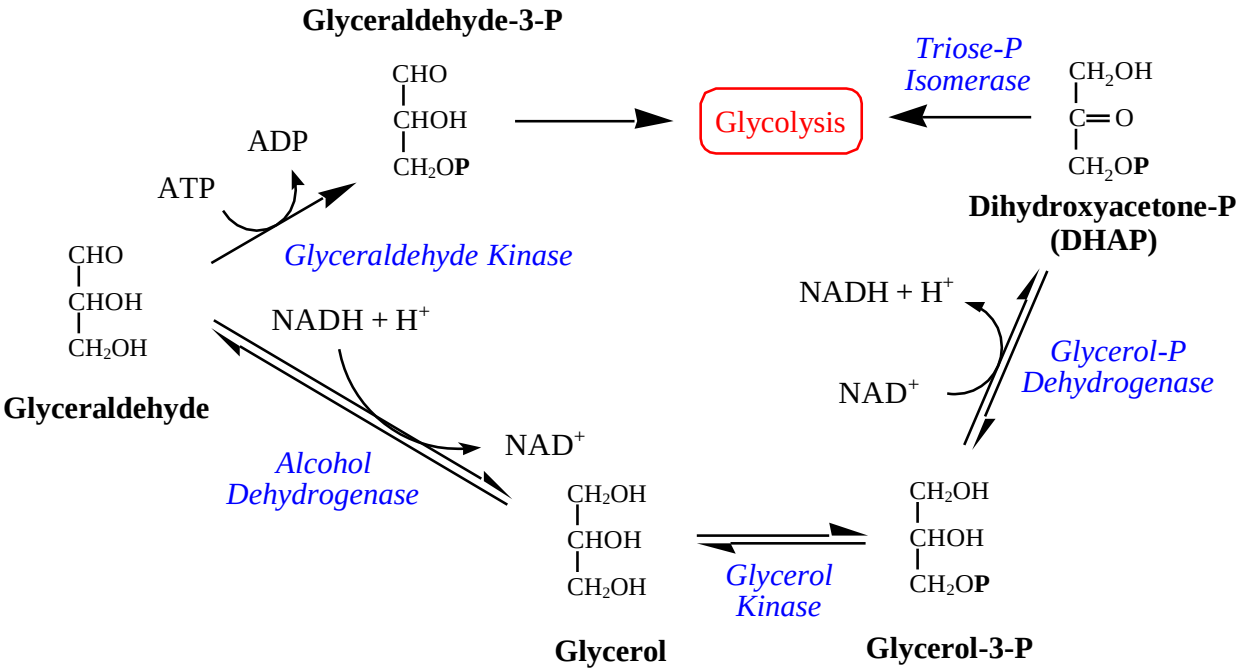
α-D-Fructose

Fructose-1-P

Liver Metabolism of Fructose II



Liver Metabolism of Fructose III



Complexity of Liver Metabolism

Breakdown of Many Metabolites

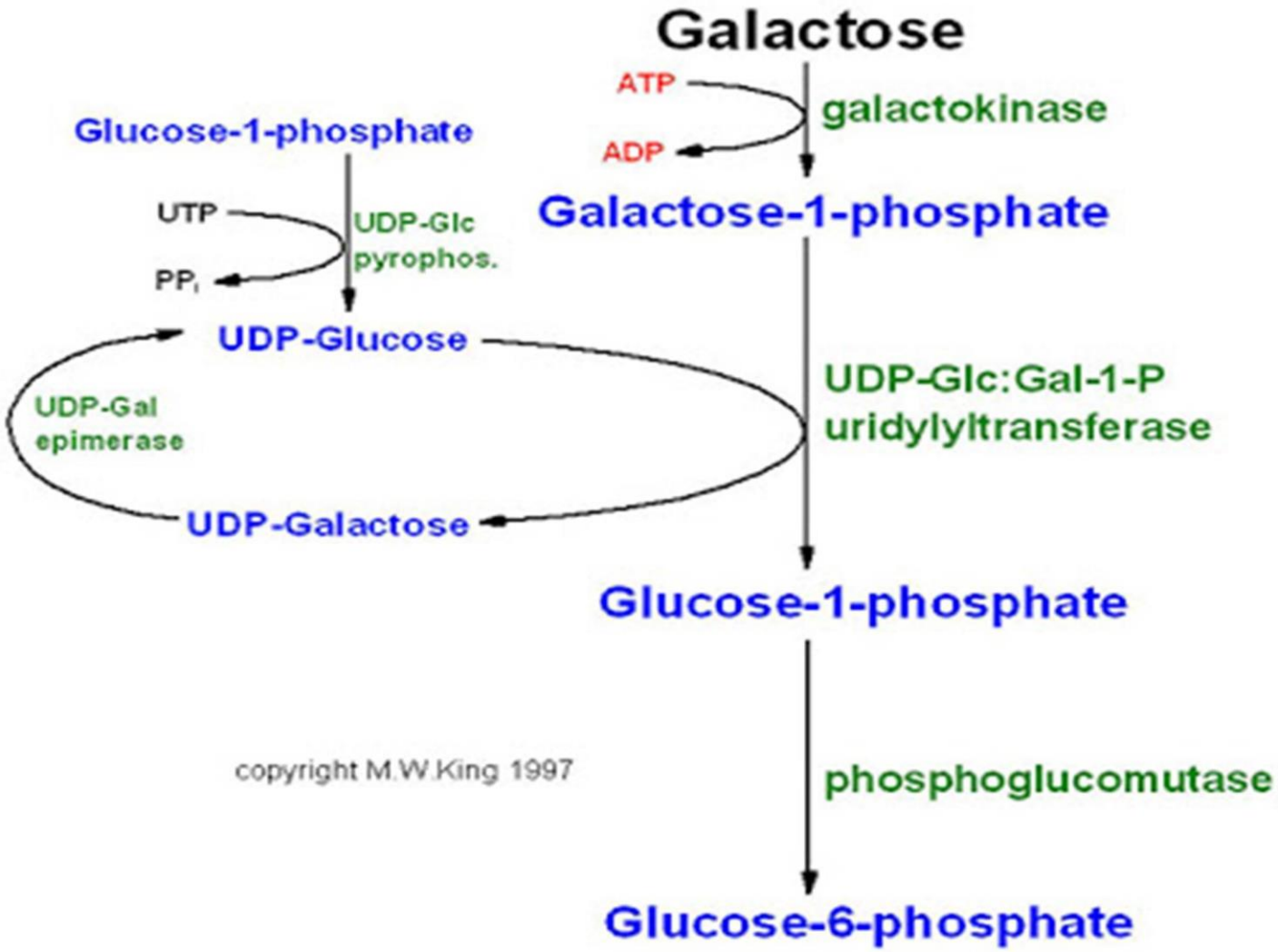
Fructose Intolerance

***Too Much Fructose**

- *-Fructose-1-P Aldolase* is rate-limiting
- Depletion of P_{i1}
- Reduction in [ATP]
- Increase in glycolysis
- Accumulation of lactate (acid) in blood

-Fructose-1-P Aldolase **Deficiency (Genetic Disease)**

Galactose Metabolism

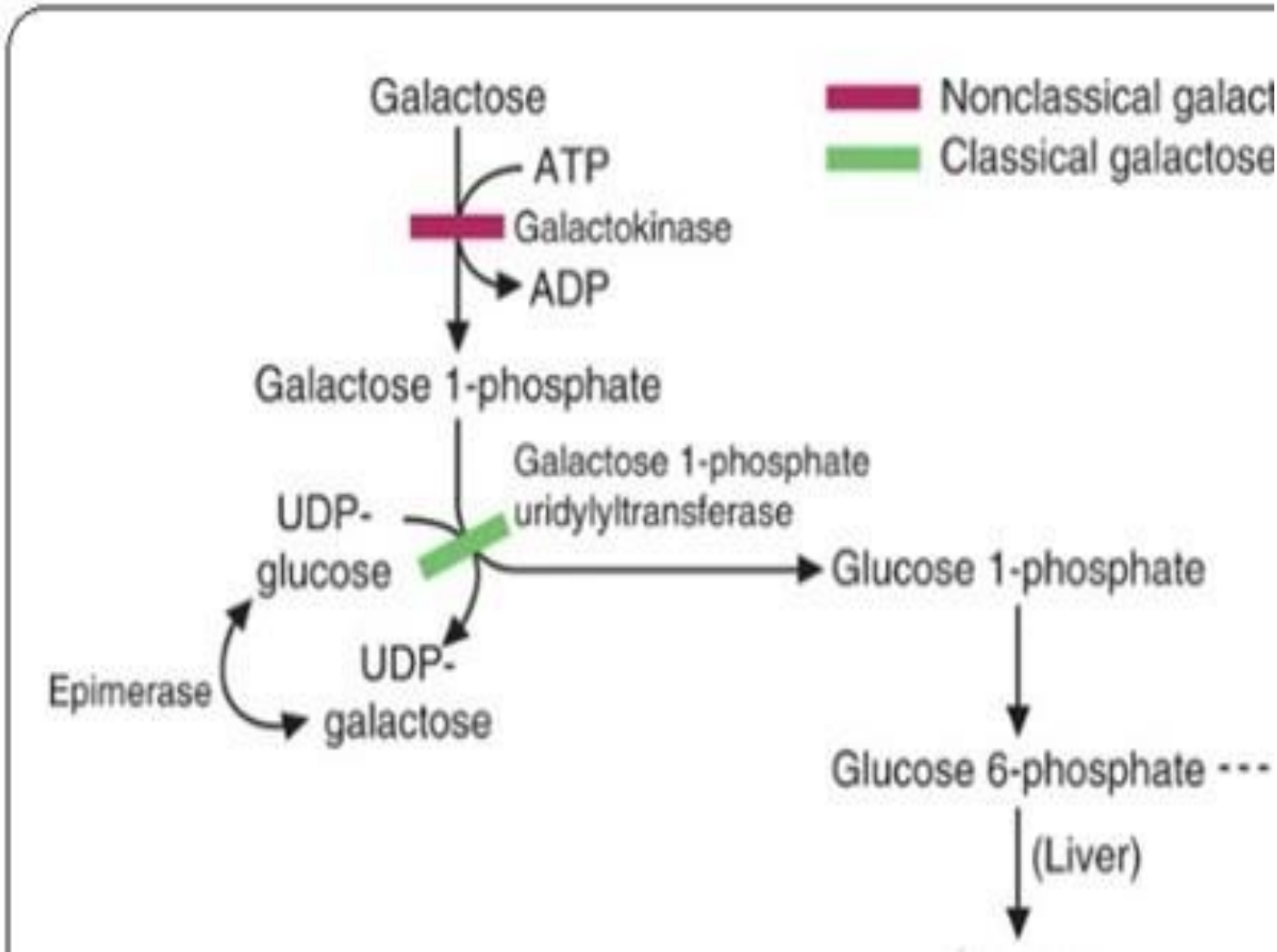


Galactose is classified as a monosaccharide, an aldose, a hexose, and is a reducing sugar.

- Galactosemia - Genetic Enzyme Deficiency:

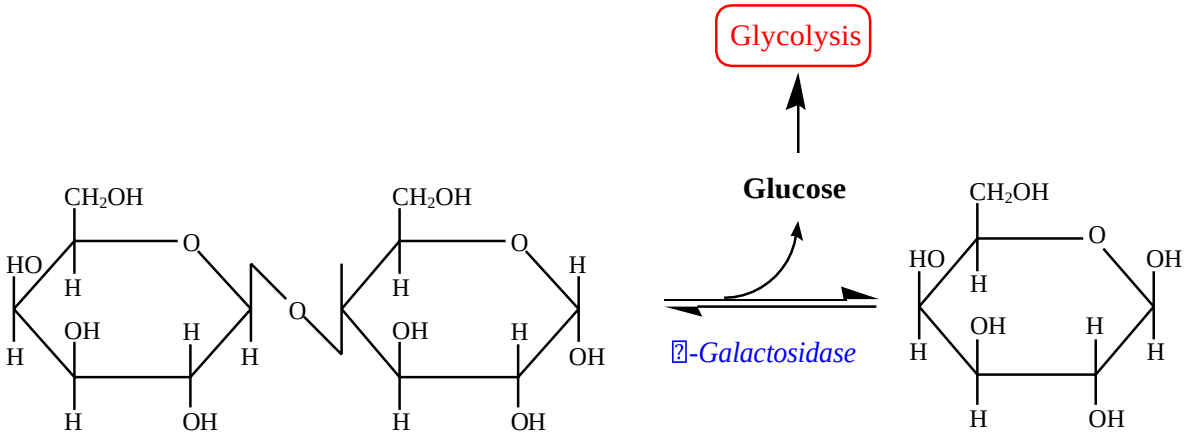
One baby out of every 18,000 is born with a genetic defect of not being able to utilize galactose. Since galactose is in milk as part of lactose, it will build up in the blood and urine. Undiagnosed it may lead to mental retardation, failure to grow, formation of cataracts, and in sever cases death by liver damage. The disorder is caused by a deficiency in one or more enzymes required to metabolize galactose.

The treatment for the disorder is to use a formula based upon the sugar sucrose rather than milk with lactose. The galactose free diet is critical only in infancy, since with maturation another enzyme is developed that can metabolize galactos



Lactose Metabolism

(Dairy Products)

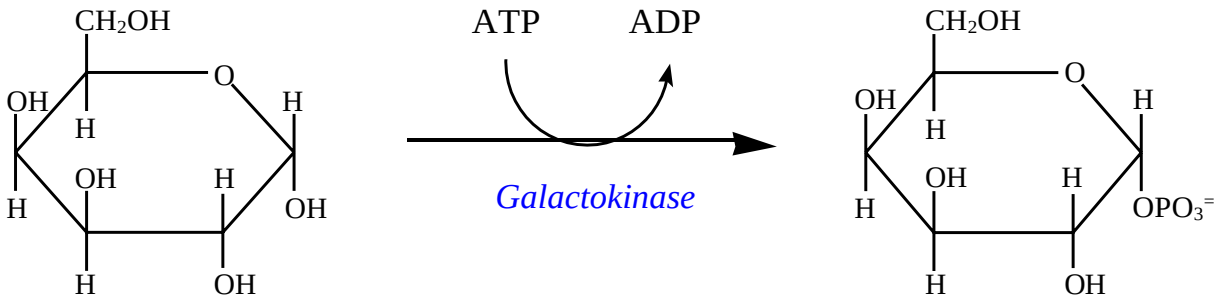


Lactose **β-D-Galactose**

Mutarotation of β-D-Galactose

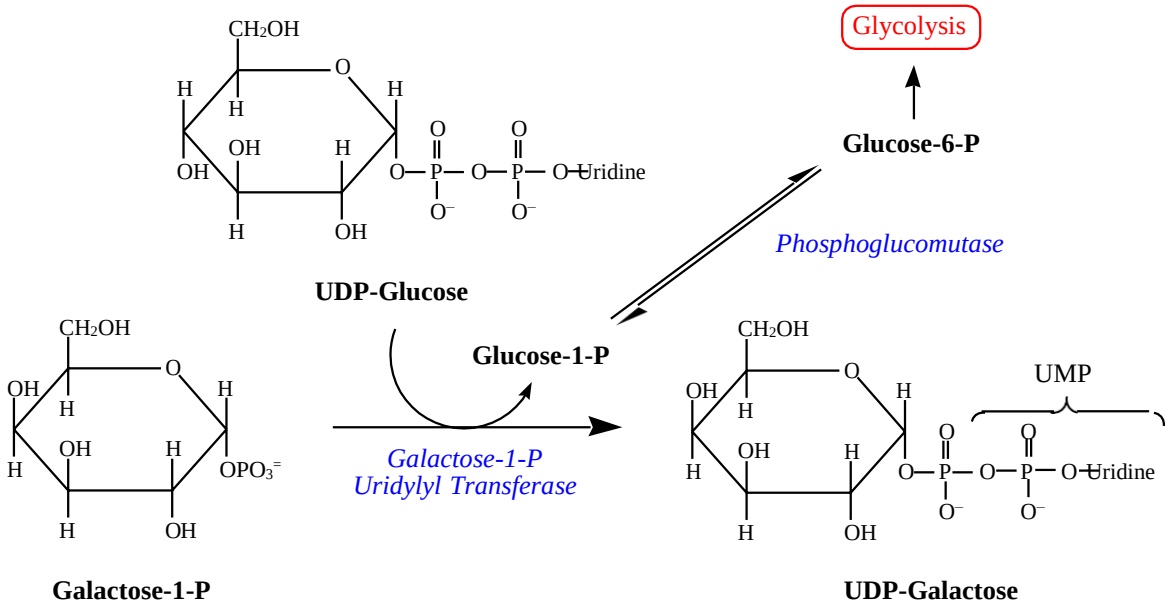
Glycolytic Enzymes are specific and do not recognize galactose!

Phosphorylation of Galactose

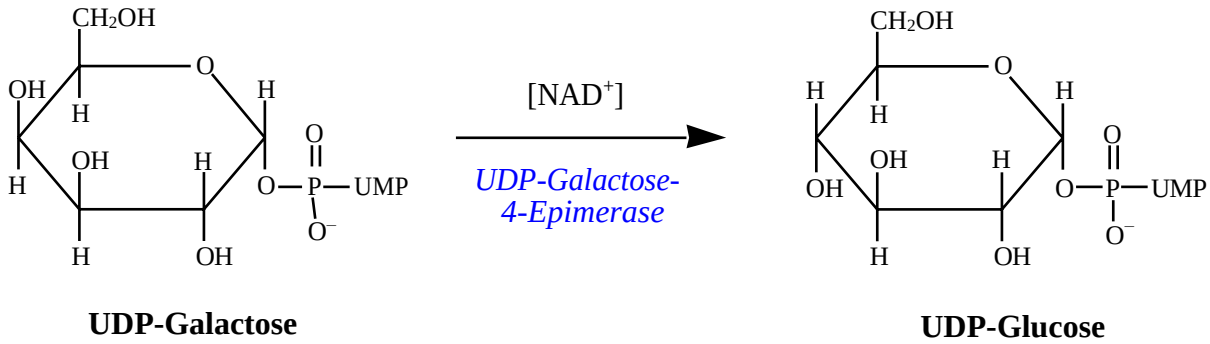


Galactose **Galactose-1-P**

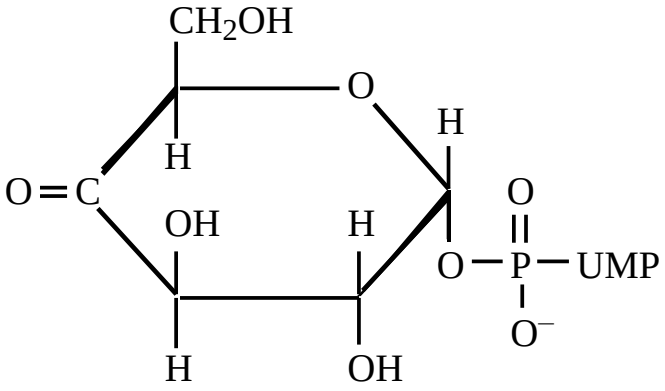
Activation of Galactose



Epimerization of UDP-Galactose



Mechanism of Epimerization



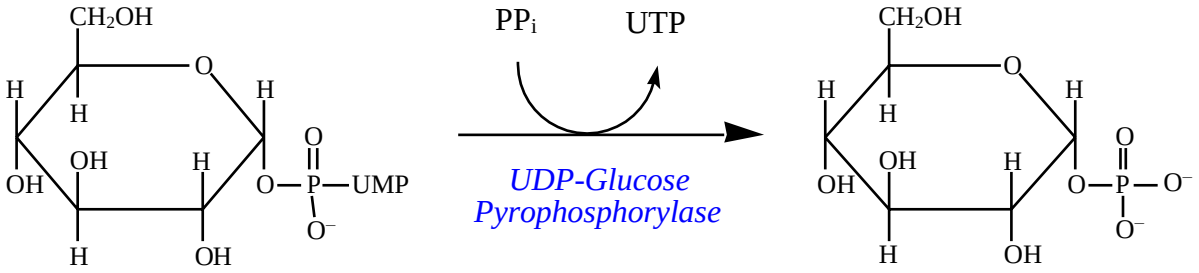
Why UDP-Galactose?

Glycoproteins

Glycolipids

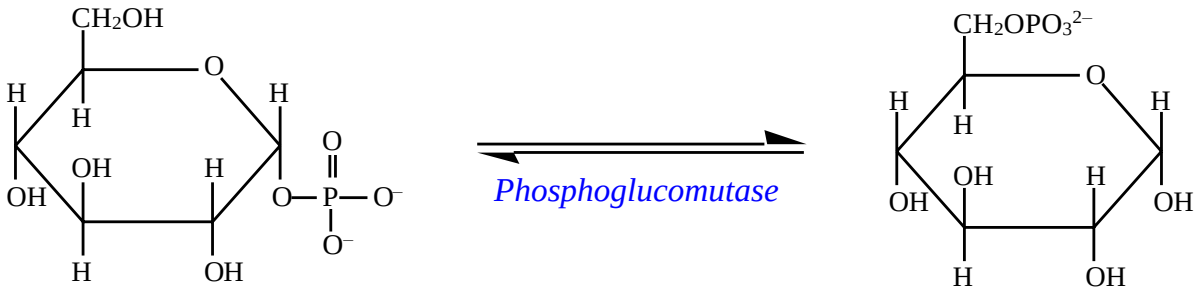
(Require UDP-Galactose)

Formation of Glucose-1-P



UDP-Glucose **Glucose-1-P**

Formation of Glucose-6-P



Glucose-1-P **Glucose-6-P**

Glucose-6-P ——> Glycolysis

Galactosemia

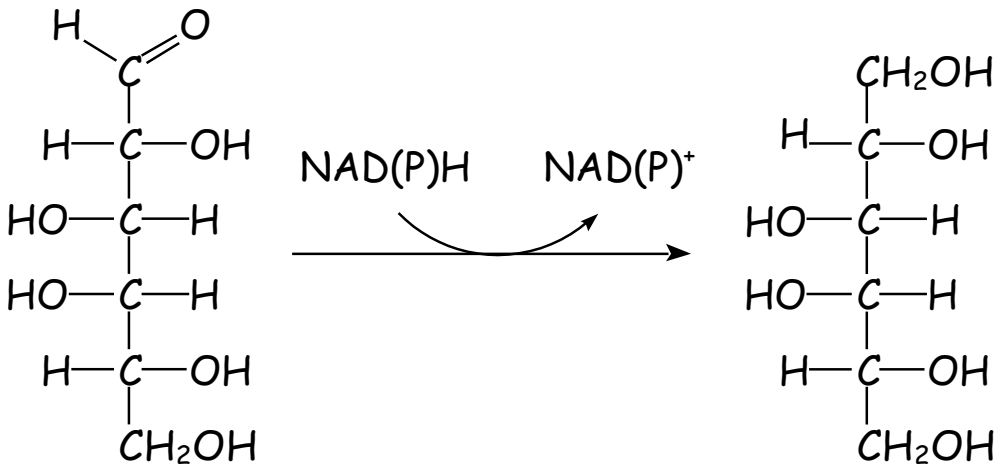
(Mental Retardation and Death)

Treatment

Galactose-free diet

(reversal of all symptoms except mental retardation)

Cataracts

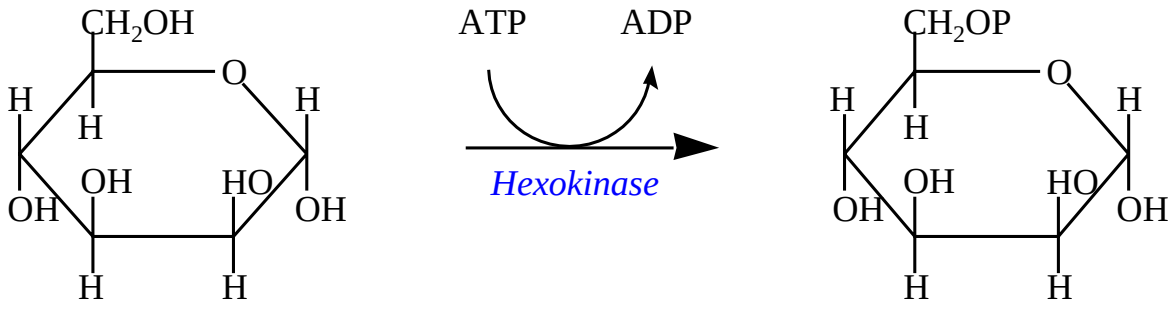


Galactose

Galactitol

Mannose Metabolism

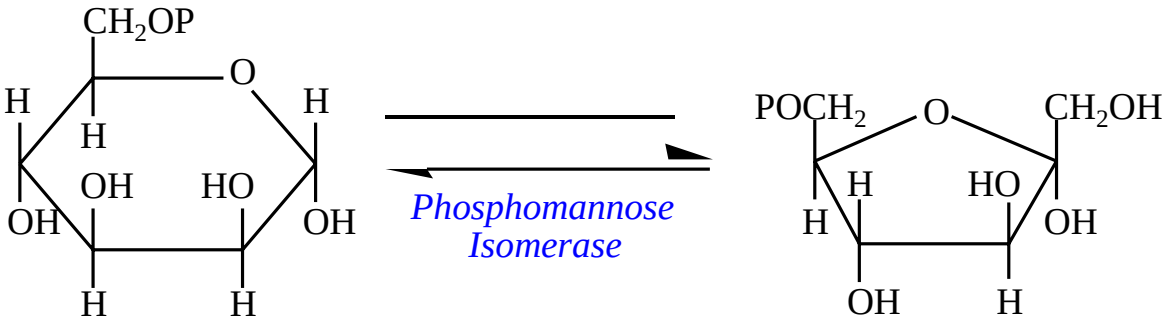
Phosphorylation of Mannose



-D-Mannose

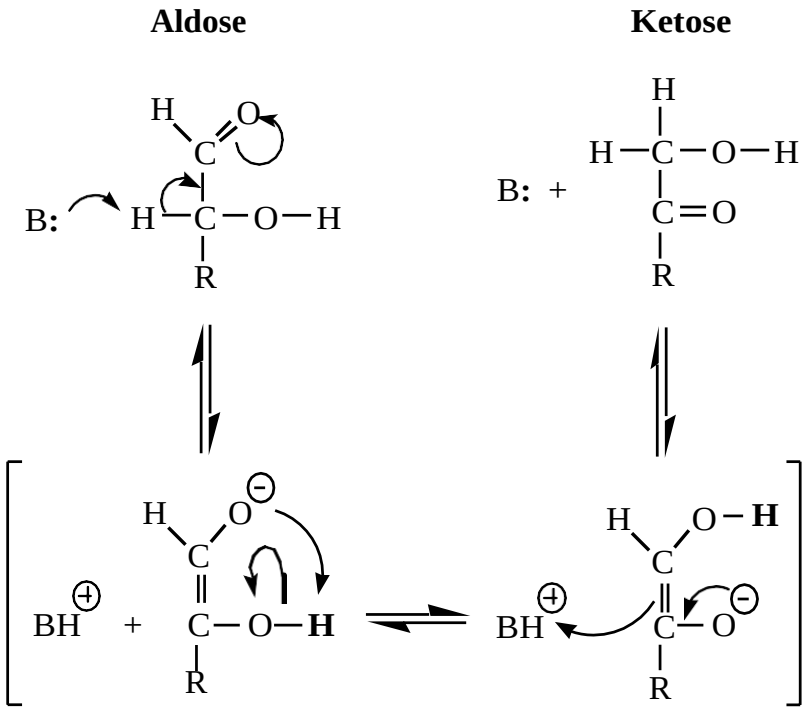
Mannose-6-P

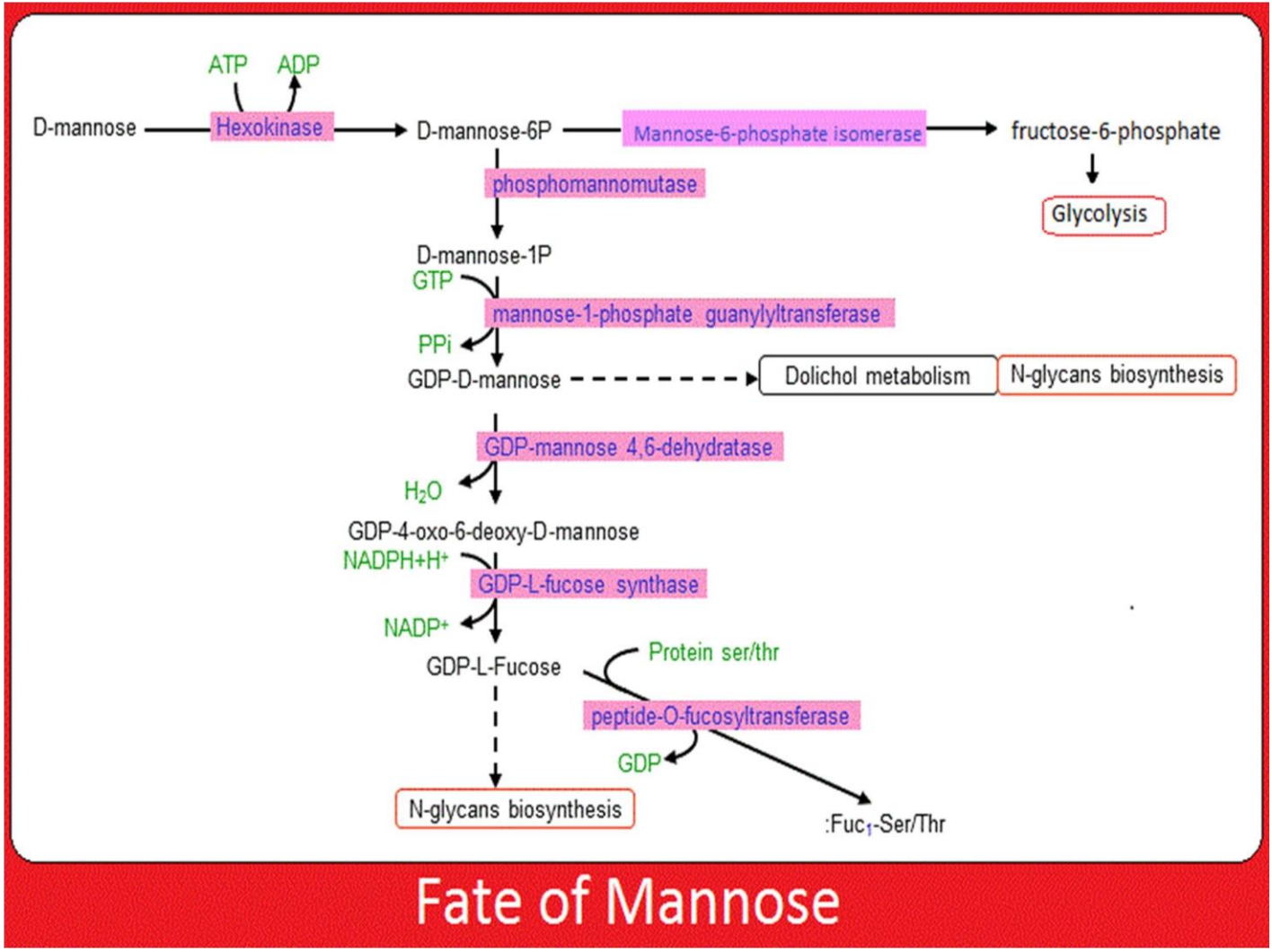
Isomerization of Mannose-6-P



Mannose-6-P **Fructose-6-P**

Mechanism of Isomerization



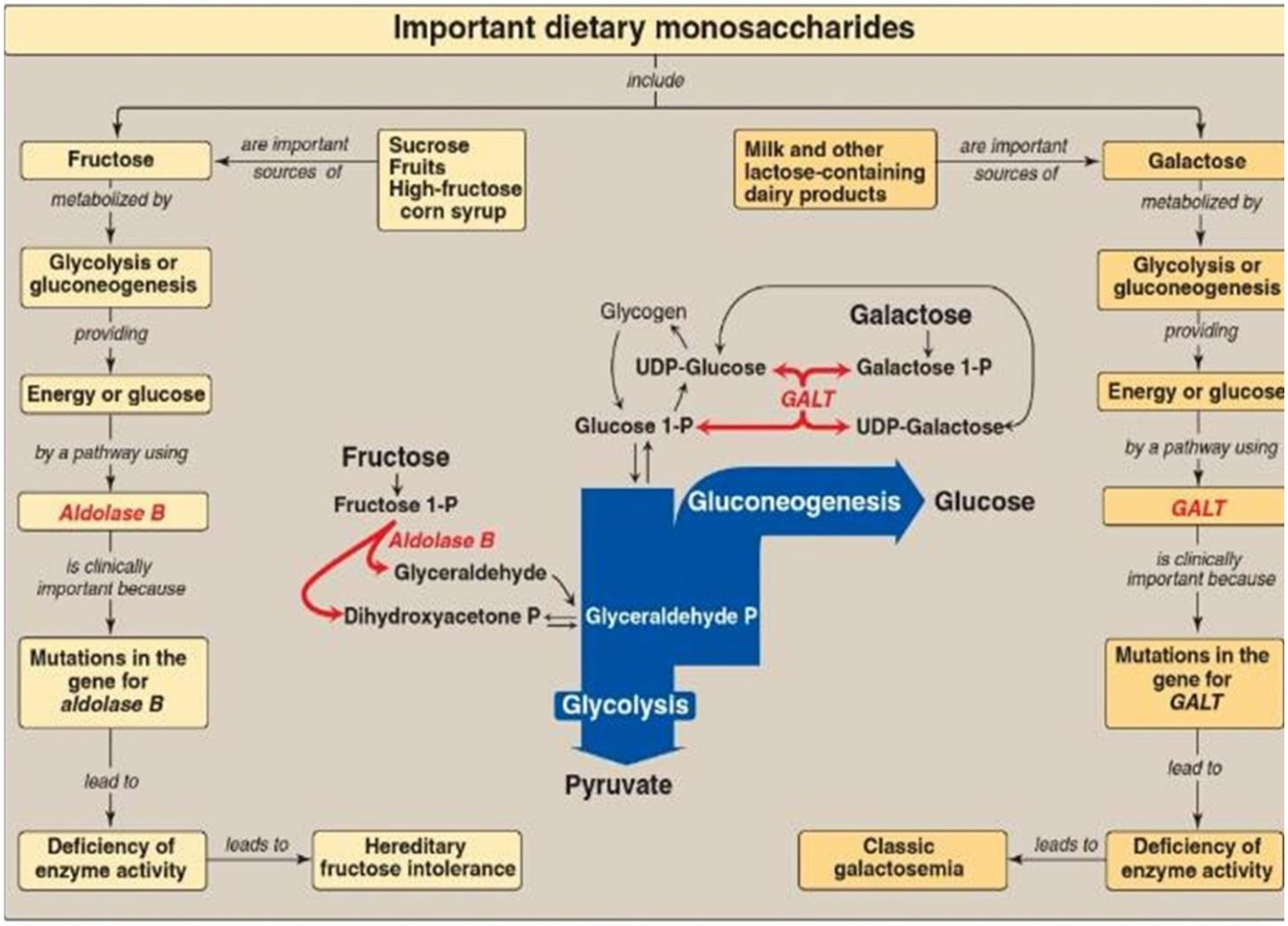


Fate of Mannose

The digestion of many polysaccharides and glycoproteins yields mannose which is phosphorylated by hexokinase to generate mannose-6-phosphate. Some of the foods that contain D-mannose are cranberries, black currants, red currants and gooseberries. Others are tomatoes, apples, peaches, oranges, blueberries and palm kernel oil.

Mannose is not well metabolized in humans and it does not significantly enter most carbohydrate metabolism pathways when taken orally. Indeed it appears that 90% of mannose ingested is excreted unconverted into the urine within 30 – 60 minutes, with 99% of the remainder being excreted within the following 8 hours.

What little mannose is metabolized appears to be phosphorylated and either converted to GDP mannose and sent on to N-glycan biosynthesis or it is converted to fructose-6-phosphate, by the enzyme phosphomannose isomerase, and then enters the glycolytic pathway.


Important dietary monosaccharides
include
Fructose
are important sources of
Sucrose
Fruits
High-fructose corn syrup
Milk and other lactose-containing dairy products
are important sources of
Galactose
metabolized by
Glycolysis or gluconeogenesis
providing
Energy or glucose
by a pathway using
Aldolase B
is clinically important because
Mutations in the gene for aldolase B
lead to
Deficiency of enzyme activity
leads to
Hereditary fructose intolerance
Glycogen
Galactose
UDP-Glucose
Galactose 1-P
GALT
Glucose 1-P
UDP-Galactose
Fructose
Fructose 1-P
Aldolase B
Glyceraldehyde
Dihydroxyacetone P
Glyceraldehyde P
Gluconeogenesis
Glucose
Glycolysis
Pyruvate
metabolized by
Glycolysis or gluconeogenesis
providing
Energy or glucose
by a pathway using
GALT
is clinically important because
Mutations in the gene for GALT
lead to
Classic galactosemia
leads to
Deficiency of enzyme activity