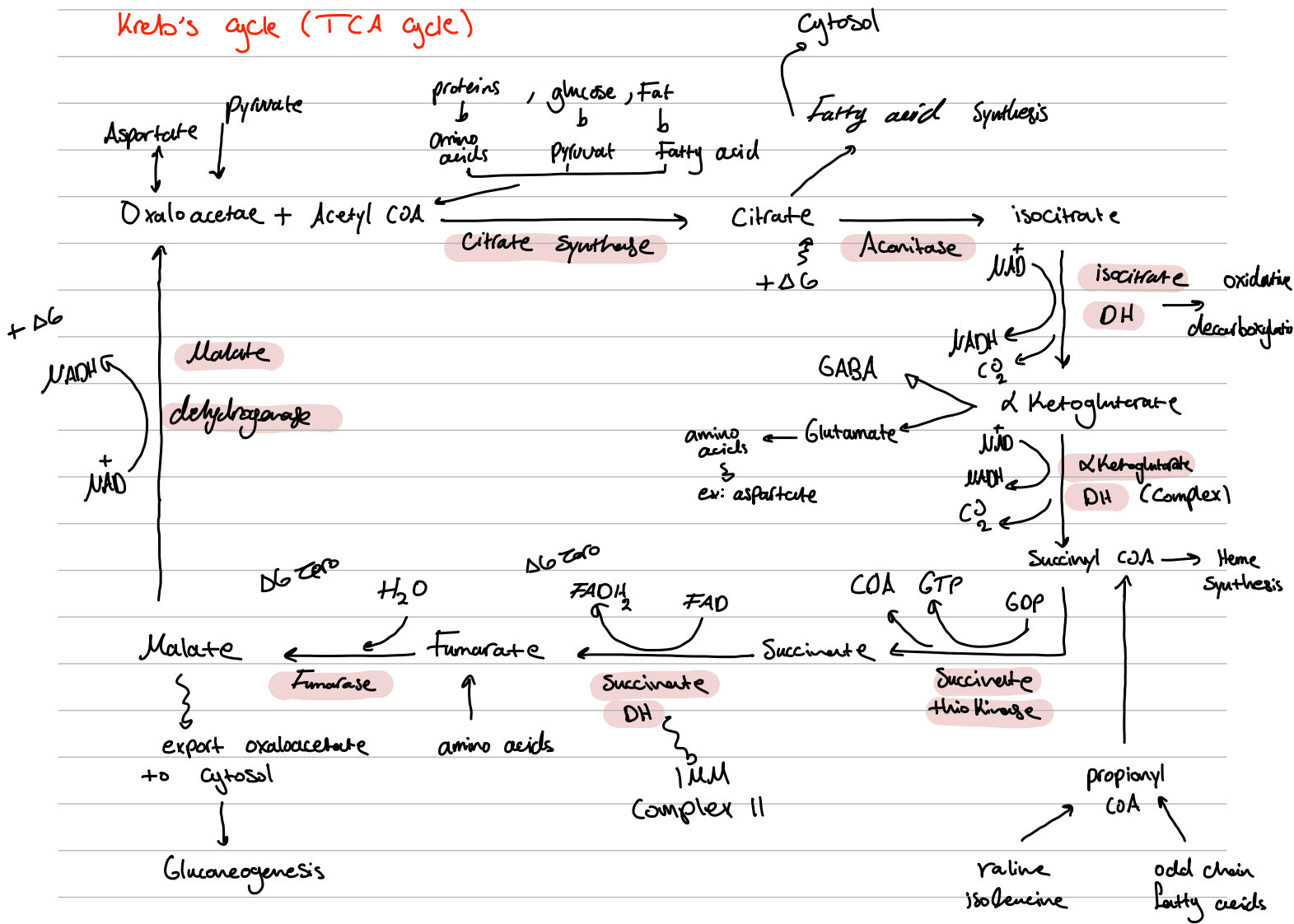
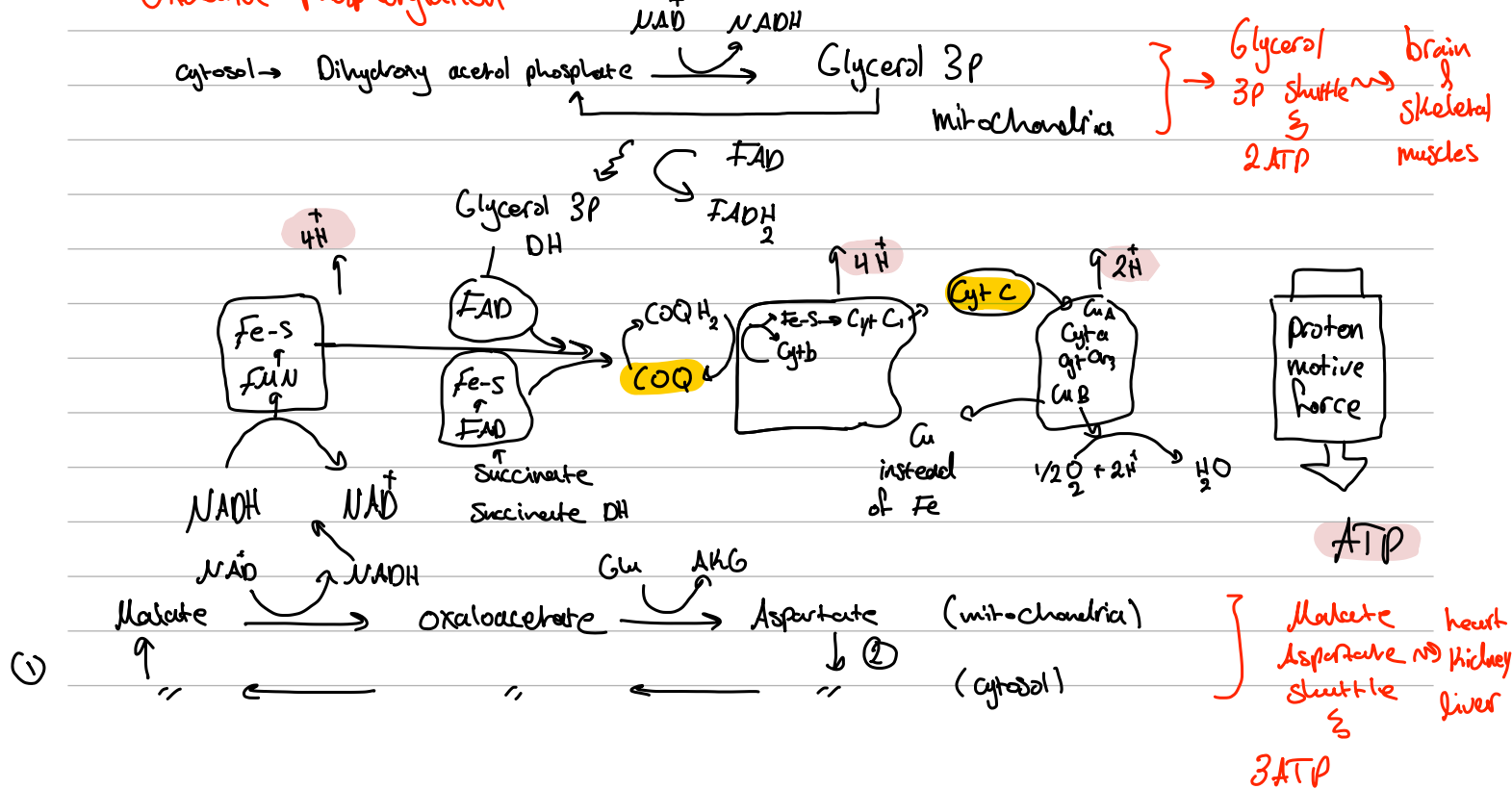


Metabolism Pathways

Krebs's cycle (TCA cycle)

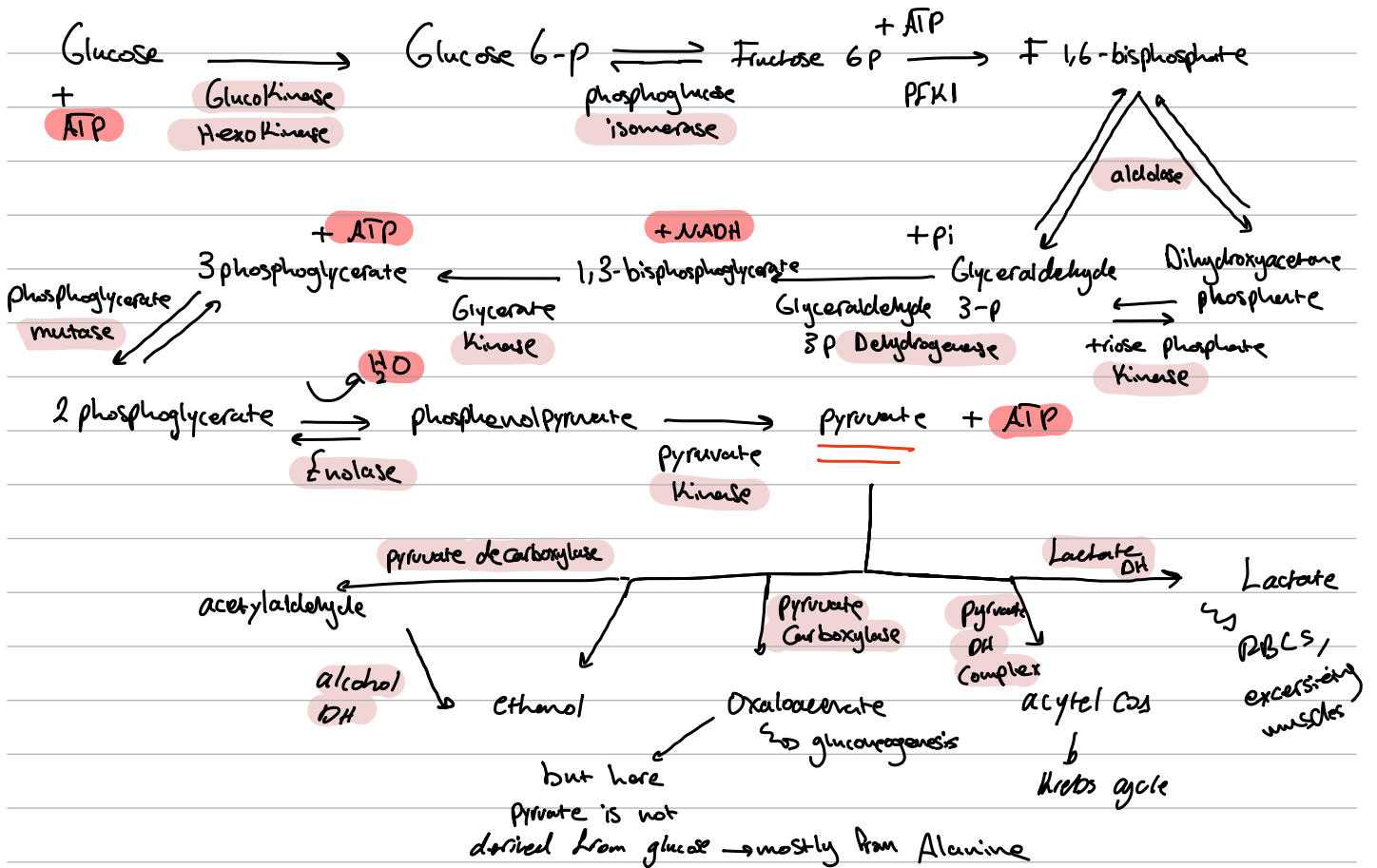


Oxidative phosphorylation

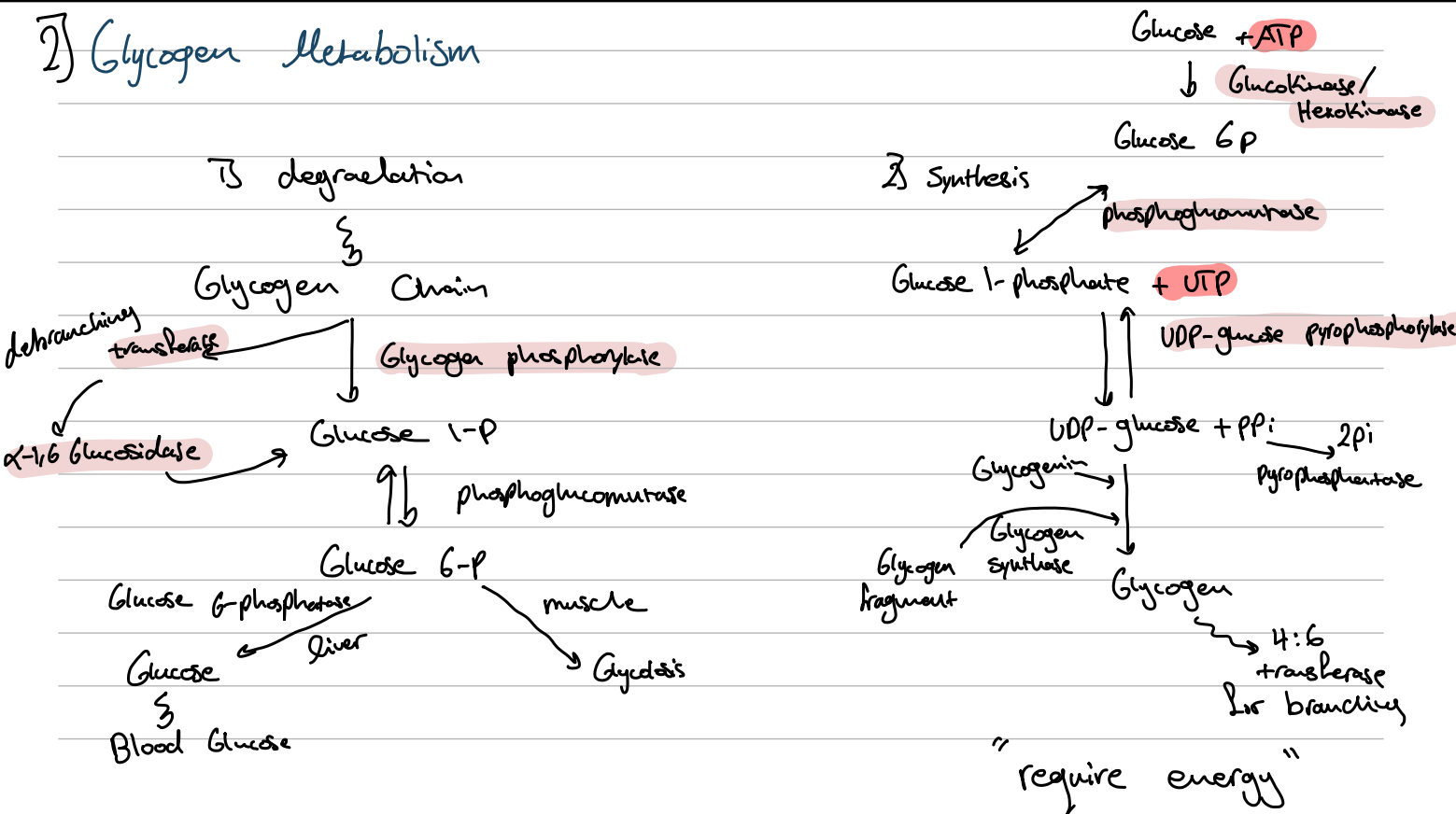


Carbohydrate Metabolism Pathway

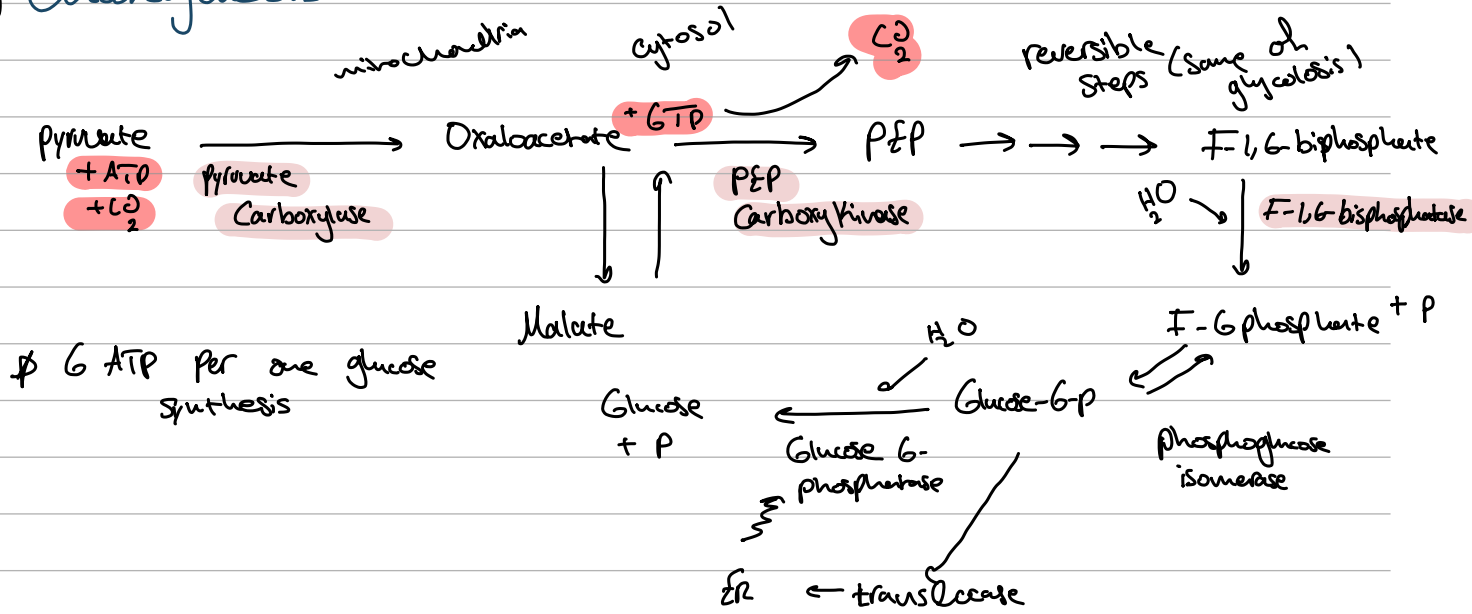
1) Glycolysis



2) Glycogen Metabolism



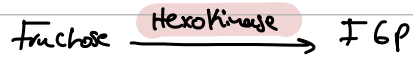
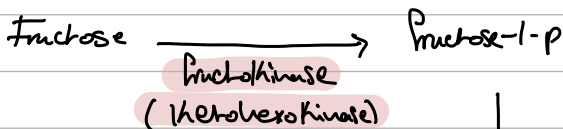
3) Gluconeogenesis



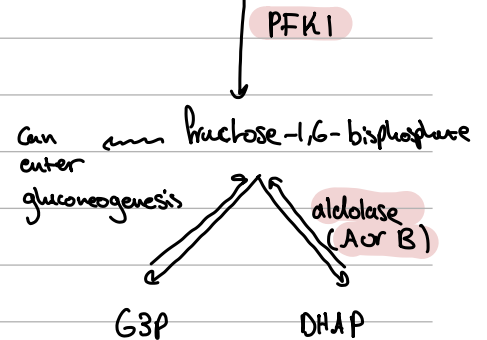
4) Fructose Metabolism (energy production) → in liver

specific pathway

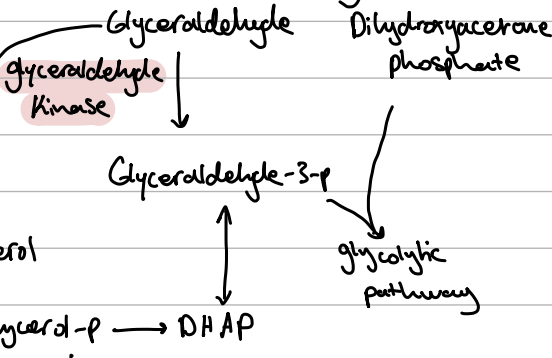
Non-specific, general pathway



aldolase B

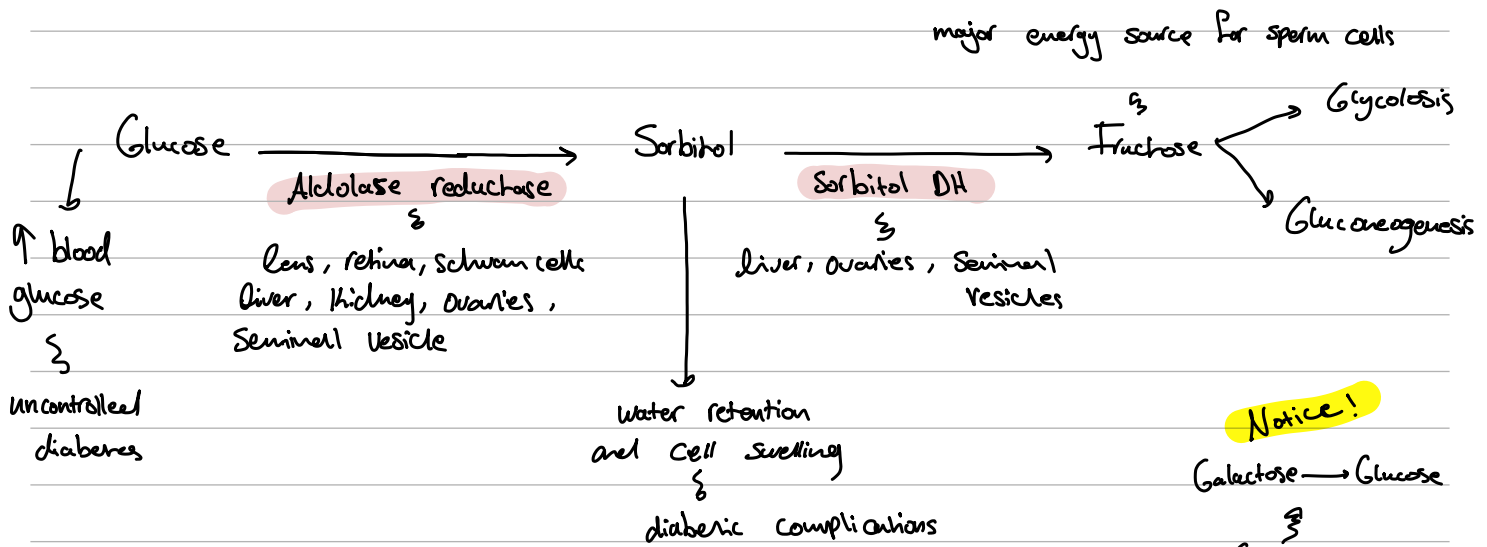


bypasses PFK 1 step (very slow) → more favorable pathway

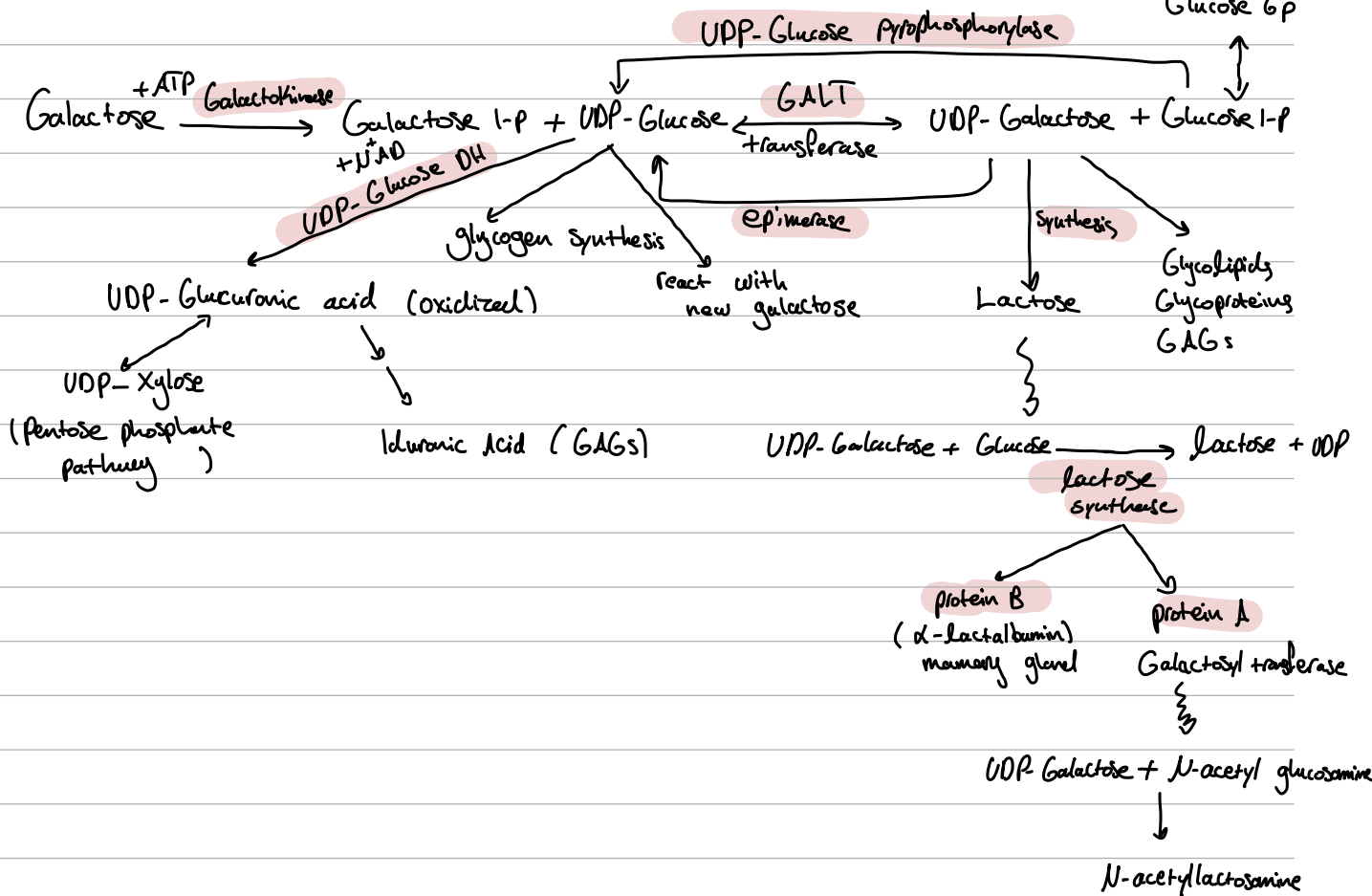


long pathway (triacylglycerols) Fats → weight gain

5) Conversion of Glucose to Fructose

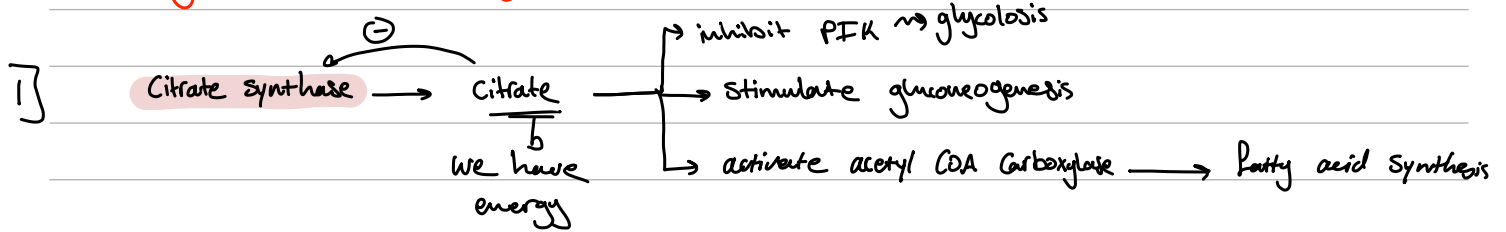


6) Galactose Metabolism



Regulation of Metabolism Pathways

Regulation of Krebs cycle

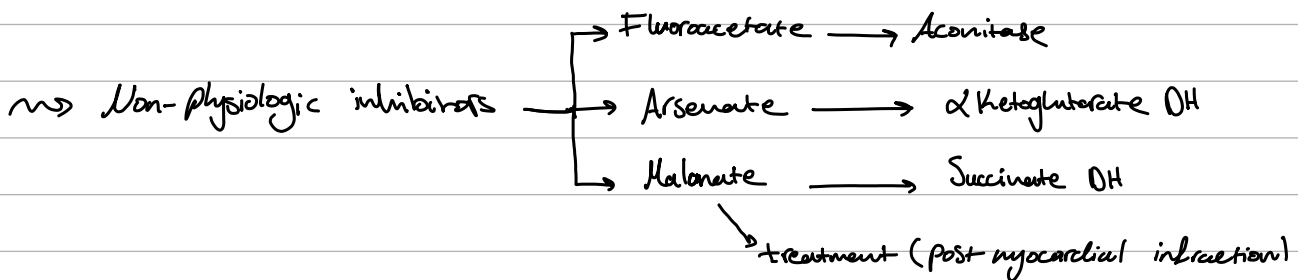


2] isocitrate DH (rate limiting) → best regulated

⊕ ADP, Ca^{+2} ⊖ ATP, NADH

shift the curve to the left → less substrate to reach high speed

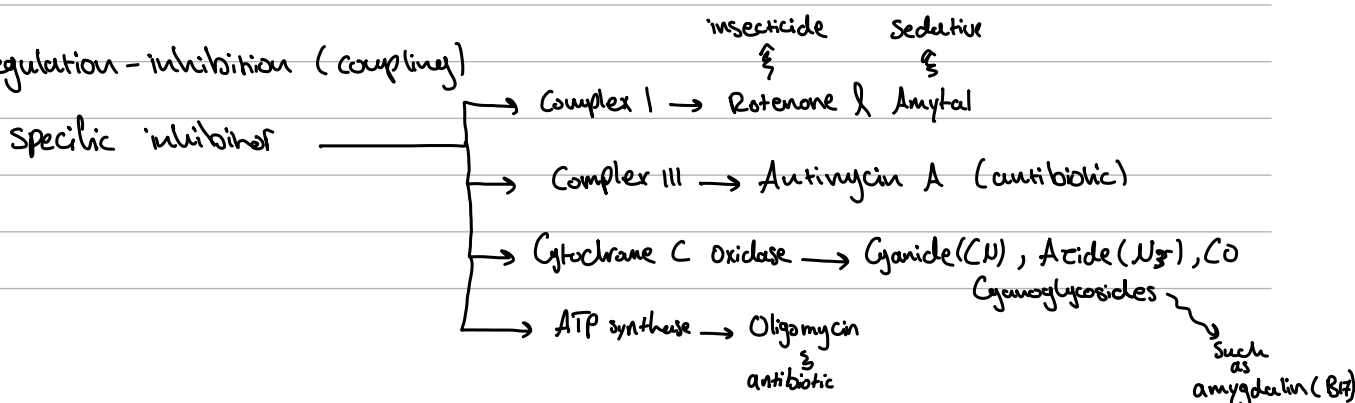
3] α Ketoglutarate DH → ⊕ Ca^{+2} ⊖ NADH, Succinyl CoA, GTP



Regulation of Oxphos

↳ respiratory control or acceptor control → ADP ⊕ Oxphos

2] Regulation - inhibition (coupling)



Fatty acid activate it

3] Regulation - Regulated Uncoupling Proteins (UCPs) → release energy of proton gradient as heat

UCP₁ (thermogenin)
UCP₂ (Most cells)
UCP₃ (skeletal muscle)
UCP₄, UCP₅ (brain)

Brain adipose tissue

↑ in infants

neck, breast
around kidneys

4] Regulation - Unregulated Chemical Uncouplers (non-physiological)

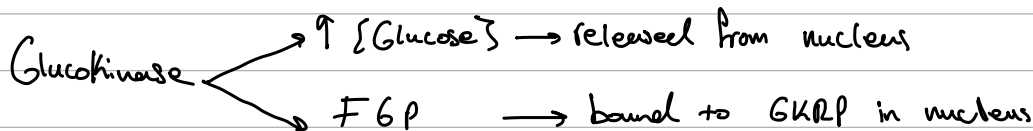
no 2,4-dinitrophenol (DNP) & other acidic aromatic compounds → ↑ O₂ consumption and oxidation

of NADH but no ATP production

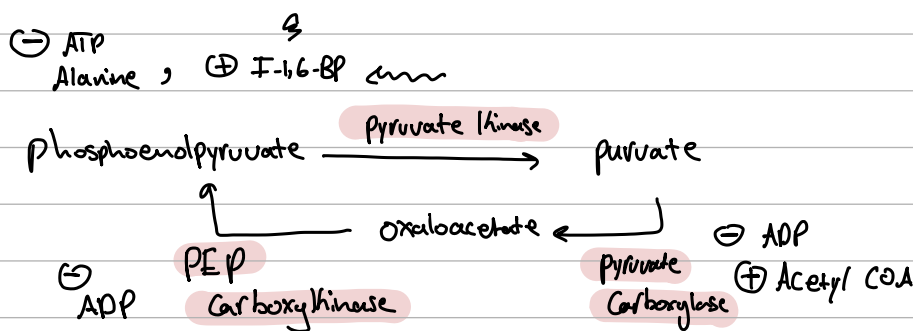
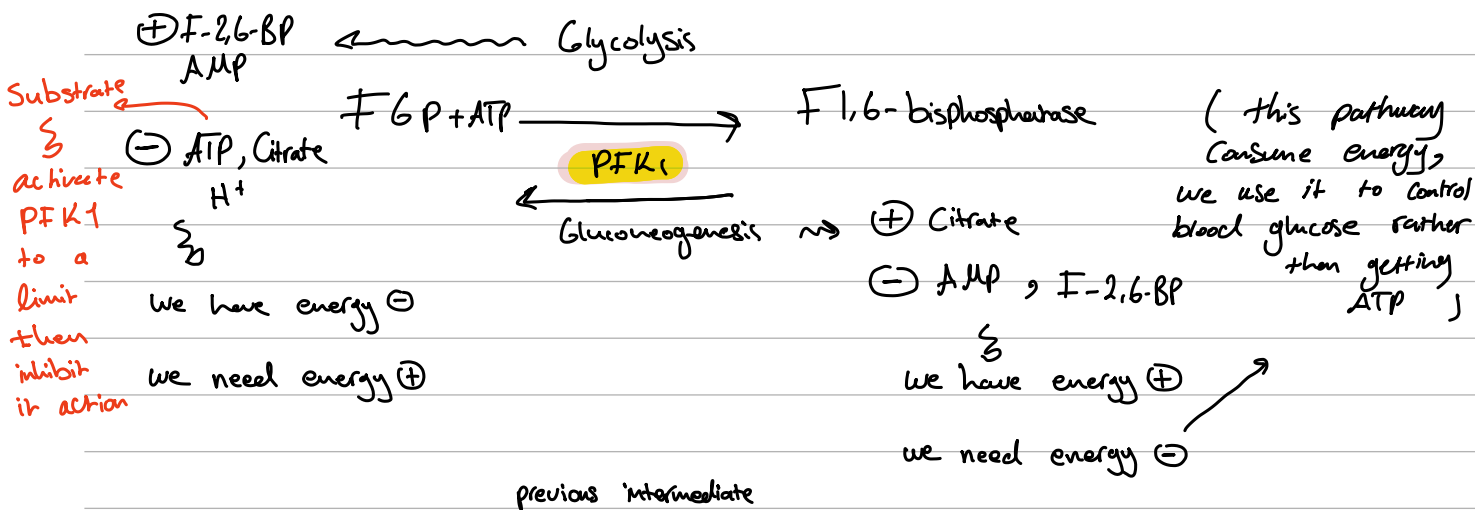
disrupt the tight coupling
of electron transport and
phosphorylation

(carries protons across IMM)

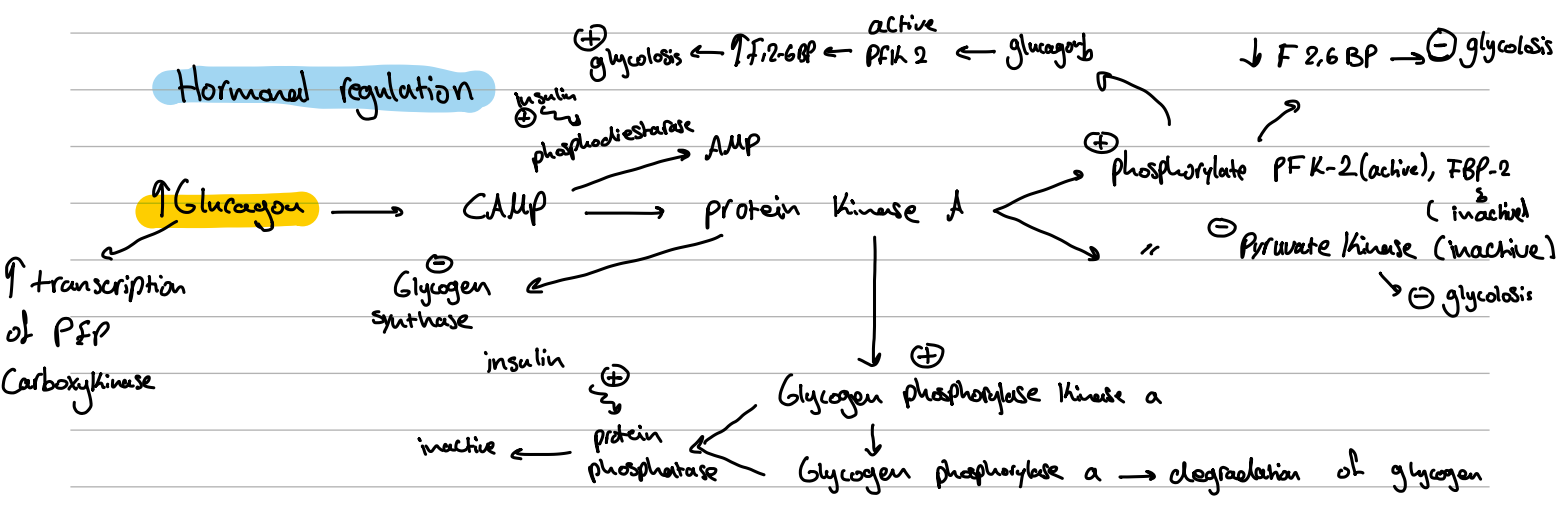
Regulation of glycolysis and gluconeogenesis



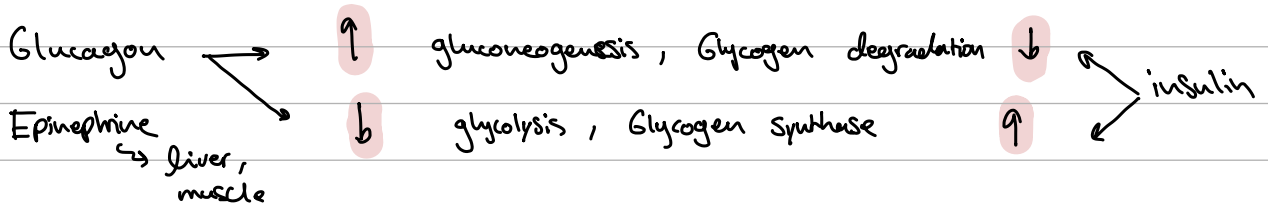
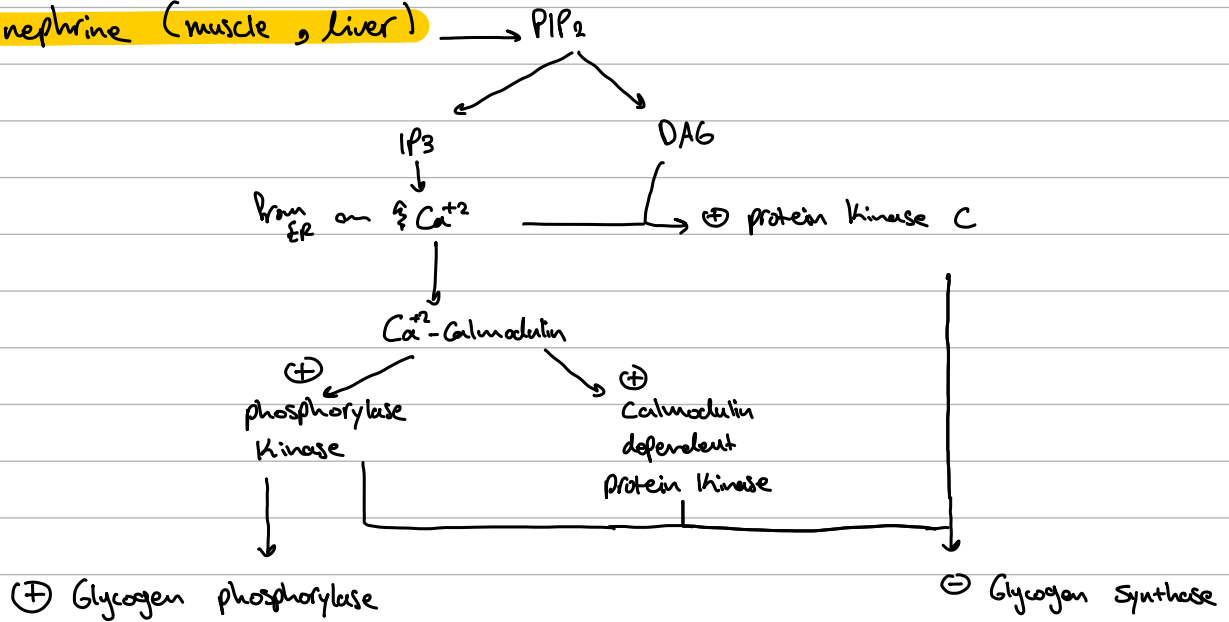
Allosteric Regulators of PFK1 \rightarrow involve in both glycolysis and gluconeogenesis



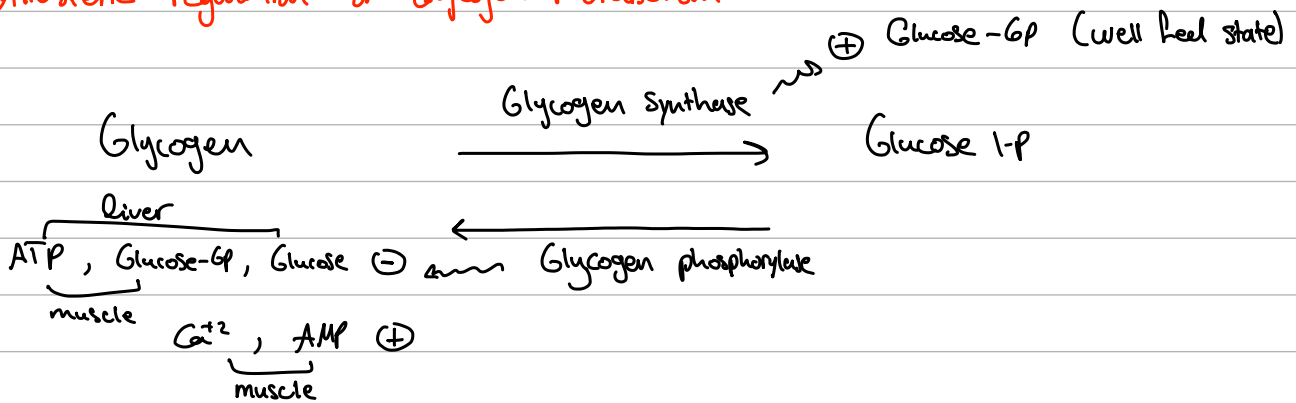
Hormonal regulation



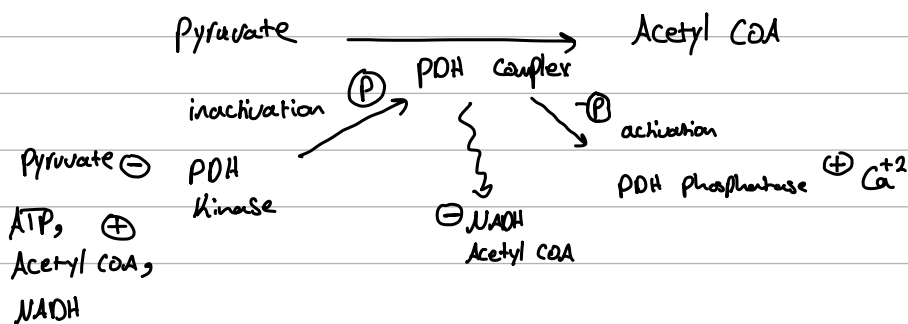
Epinephrine (muscle, liver)



Allosteric regulation of Glycogen metabolism



Regulation of PDH complex



Disorders associated with metabolism pathways

1) Oxphos Diseases (Genetic)

mutations in nDNA

mutations in mtDNA

- Complex I → LHON + Dystonia, Sporadic myopathy
- Complex III → sporadic myopathy
- Complex IV → sporadic myopathy, anemia, Encephalomyopathy

Complex I → Leigh syndrome, Leukodystrophy

Complex II → " , Paraganglioma

Complex IV → " , Leukodystrophy/tubulopathy, Cardioencephalomyopathy

Complex V
NARP, MILS, FBSU

2) Carbohydrate digestion

Sucrase-isomaltase deficiency → Gases, organic acids, other osmotically active molecules
Permeation of sucrose

Lactase deficiency → Lactose permeation by bacteria
 Carbon metabolites → bloating
 H_2 → lost → diarrhea
 H_2 can be measured in breath

3) Glycolysis

→ Pyruvate kinase deficiency → RBCs are affected → Mild to chronic hemolytic anemia

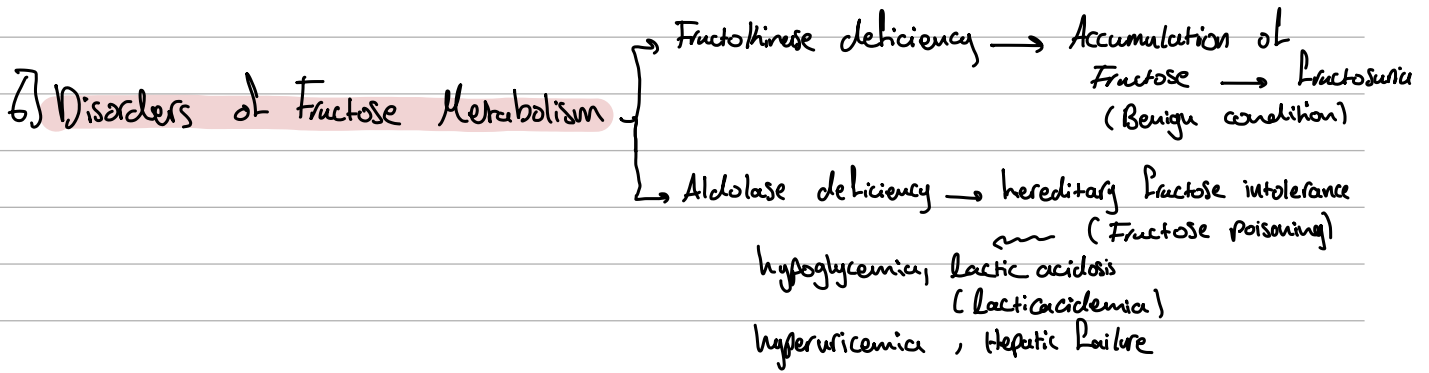
4) Pyruvate dehydrogenase deficiency

→ Affect brain → Dependent on TCA cycle
 → sensitive to acidosis

neurodegeneration, muscle spasticity, early death in neonatal onset

5) Glycogen storage diseases

- Von Gierke disease → Glucose-6-phosphatase
 liver, kidney and intestine → Lasting hypoglycemia, renal disease, Hepatomegaly, fatty liver, Growth retardation
- McArdle syndrome → Muscle glycogen phosphorylase
 ↓
 ATP shortage → weakness after exercise
- Pompe disease → Lysosomes α(1-4) glucosidase → liver, heart muscle
 ↓
 Massive cardiomegaly → early death from heart failure
 Normal blood sugar and glycogen storage



Similar consequences that in fructose intolerance

