



MedEd

Lecture No.- 03

FARRE

1ST PROF 2025

Biochemistry

Topic- Carbohydrate Metabolism

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TOPICS

to be covered

DKA
DM
Lactase ↓
Sucrase ↓

Galactosemia

HFI

GSD - VG

Pompos
McArdle

G6PDH

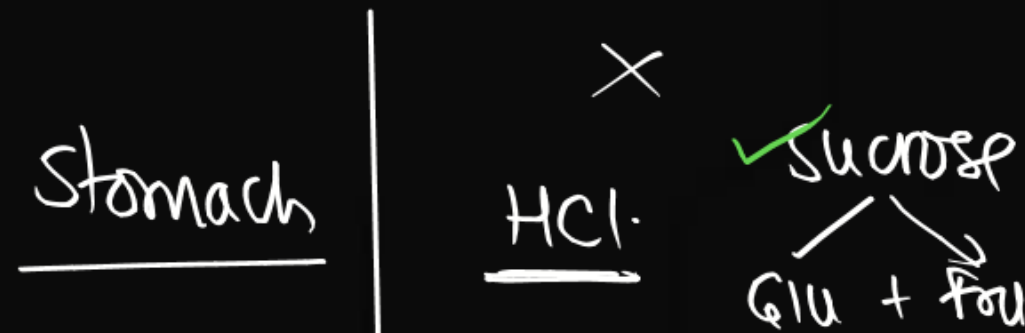
- 1 Carbohydrate digestion and Absorption
- 2 Glycolysis and R1 cycle
- 3 Gluconeogenesis from lactate, Glycerol, Propionyl CoA, Amino acids
- 4 Glycogen metabolism and Regulation and Disorders
- 5 HMP shunt and G6PD deficiency
- 6 Galactose and Fructose metabolism

Digestion & Absorption

Starch, Glycogen
Lactose, Glu, Fru, cellulose

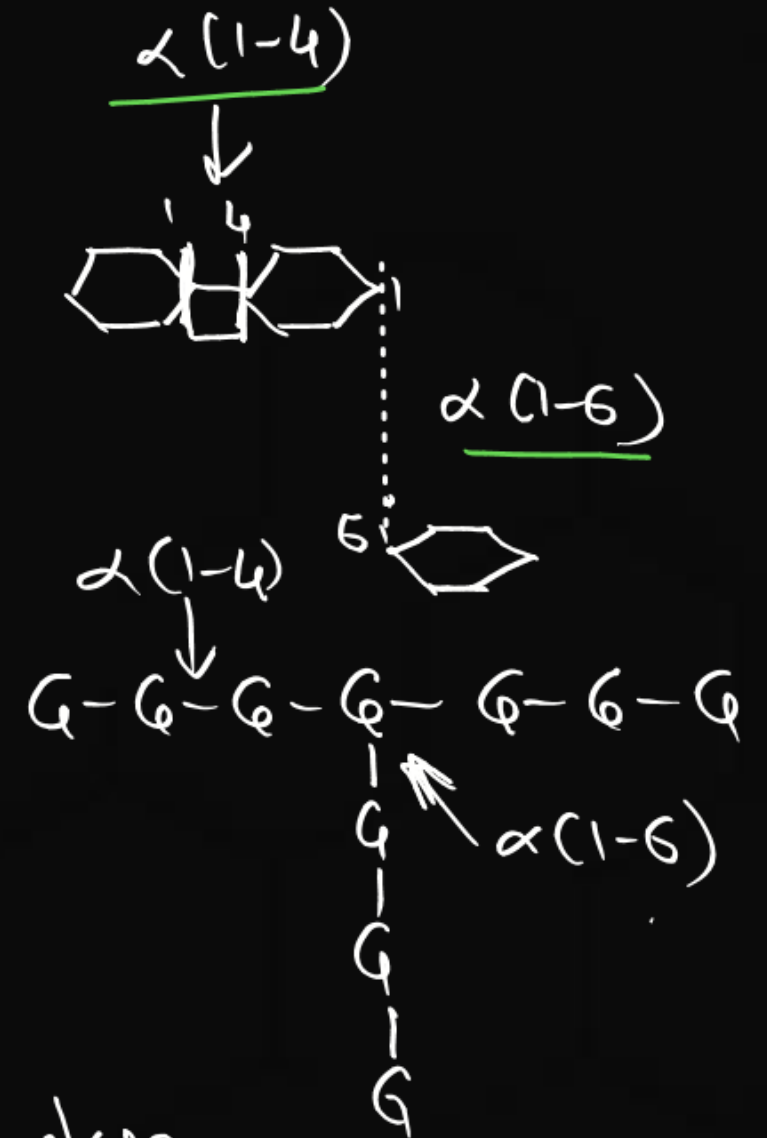


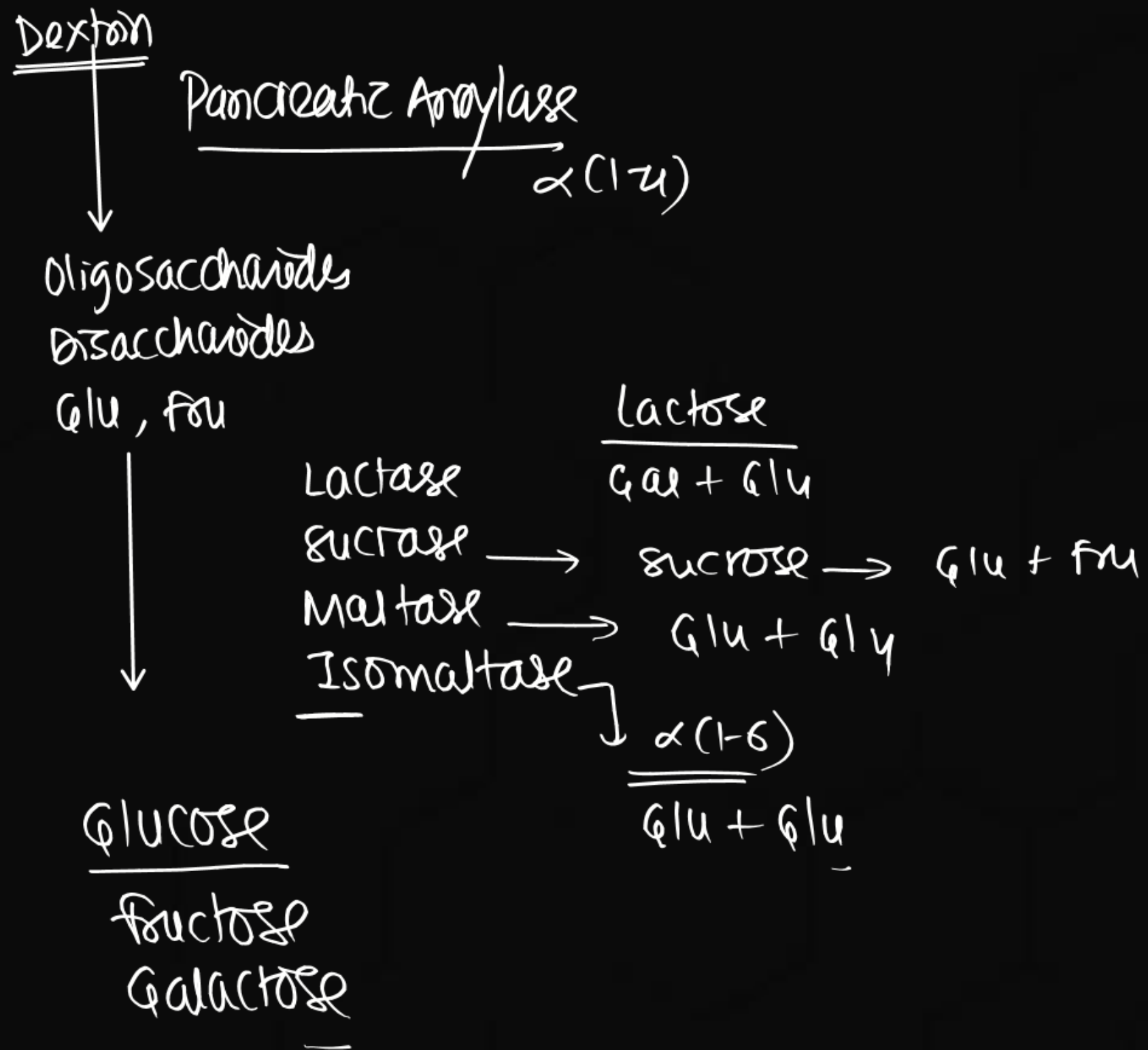
Oligosaccharide



Chyme
duodenum ↓

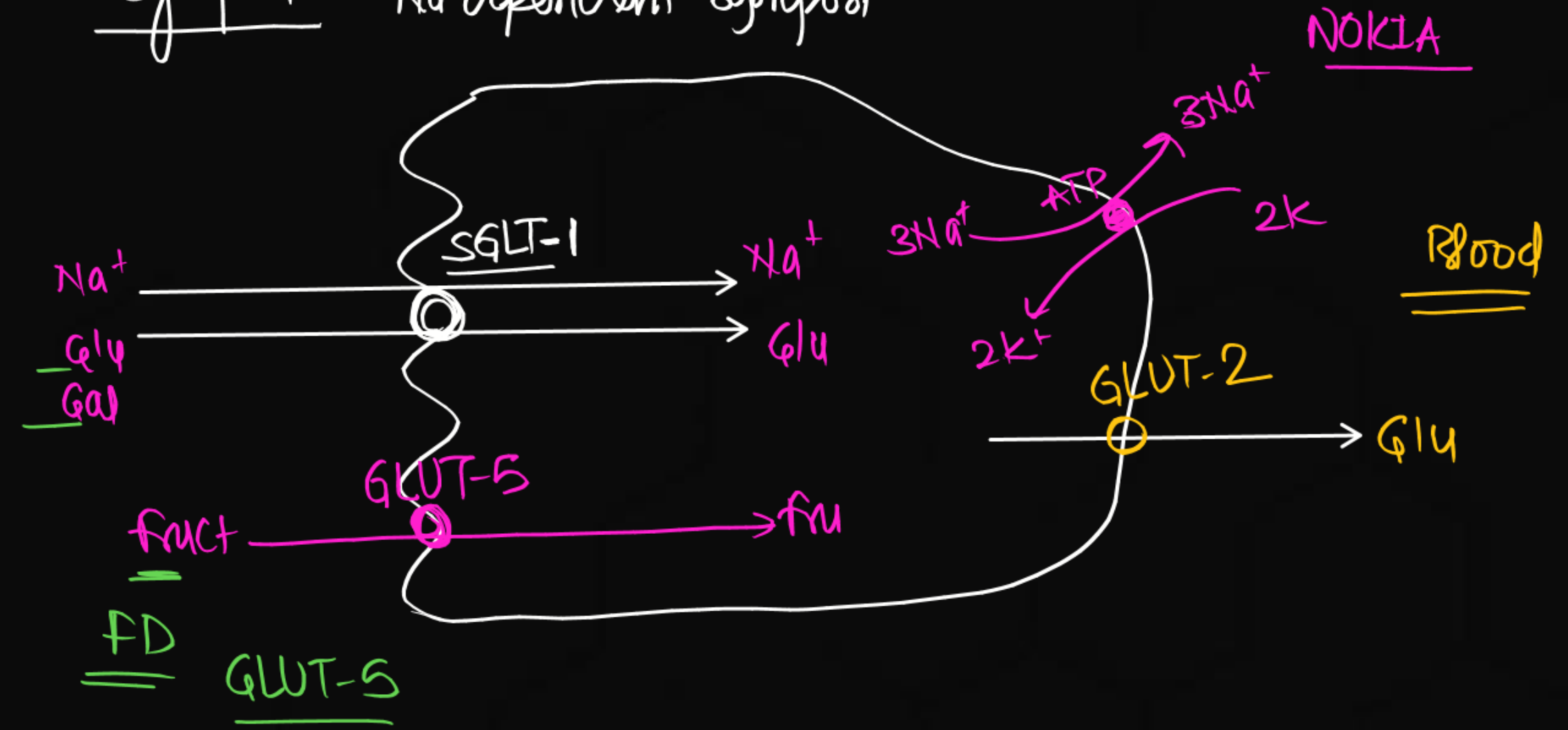
CCK → Pancreatic Amylase
Secretion → causes release of HCO_3^-





Absorption

Symport - Na dependent symport



GLUT → FD

* GLUT 1 → Kidney, Placenta, RBCs, Brain, Colon

✓ 2 → B/L intestinal cell, B/L renal tubular cell, β -cell, Liver
Regulation

3 → Neurons, Kidney, Placenta

VIP
MCA

4 → sk. muscle, Heart, Adipose tissue → Insulin dependent

5 → Intestine, testis → Abs. of fructose

Glycolysis

Glucose



Pyruvate



Lactate

1. Energy investment

2. Energy generation phase

Embden Meyerhof pathway

All cells

Aerobic/Anaerobic



✓ ③ Irreversible

② Substrate level phosphate

③ Inhibitor

① Reversible kinase reaction

Aerobic Anaerobic

RL

characteristics
steps

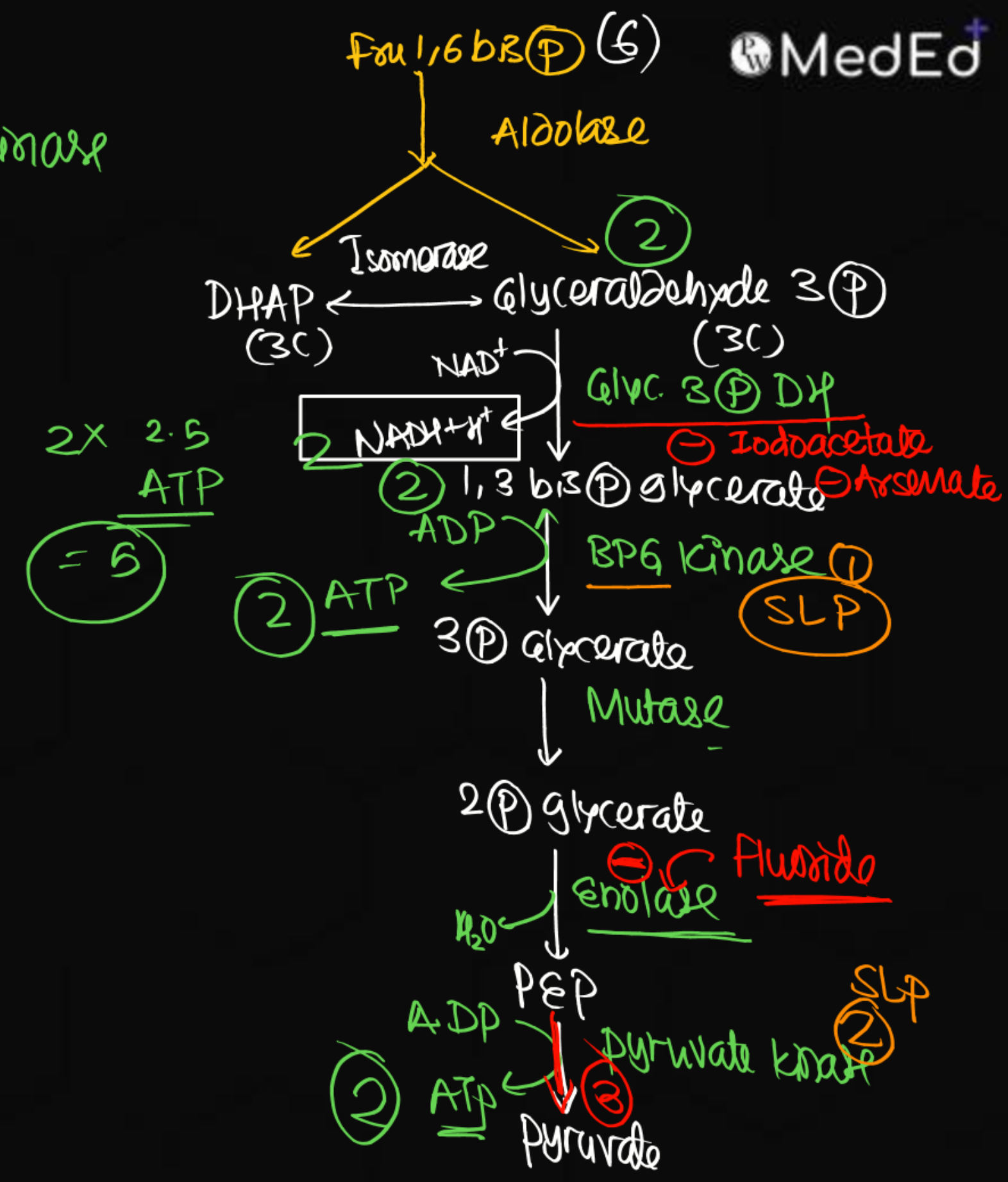
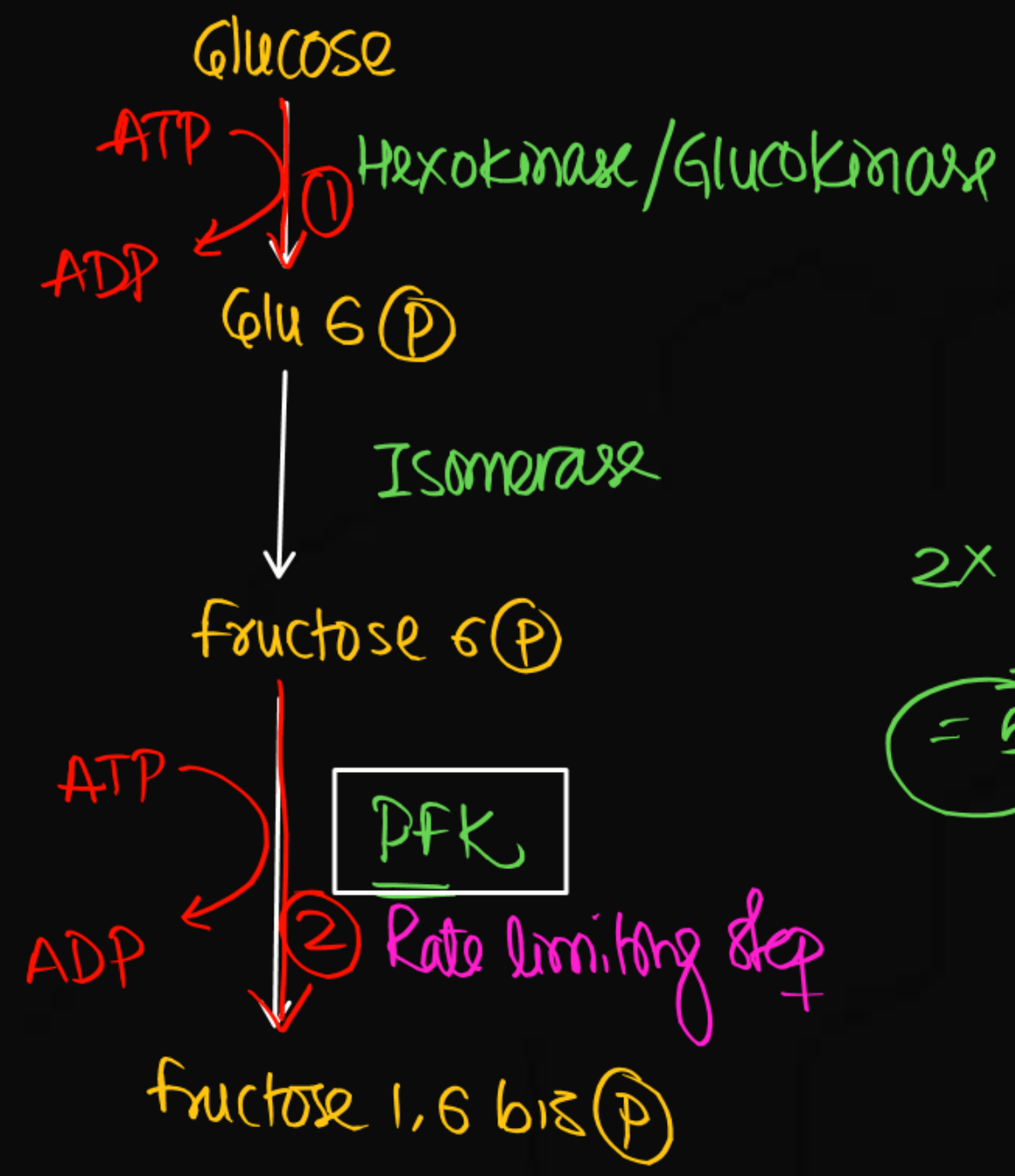
energetics

Regulation - ①

C/S

②

③



Energetics

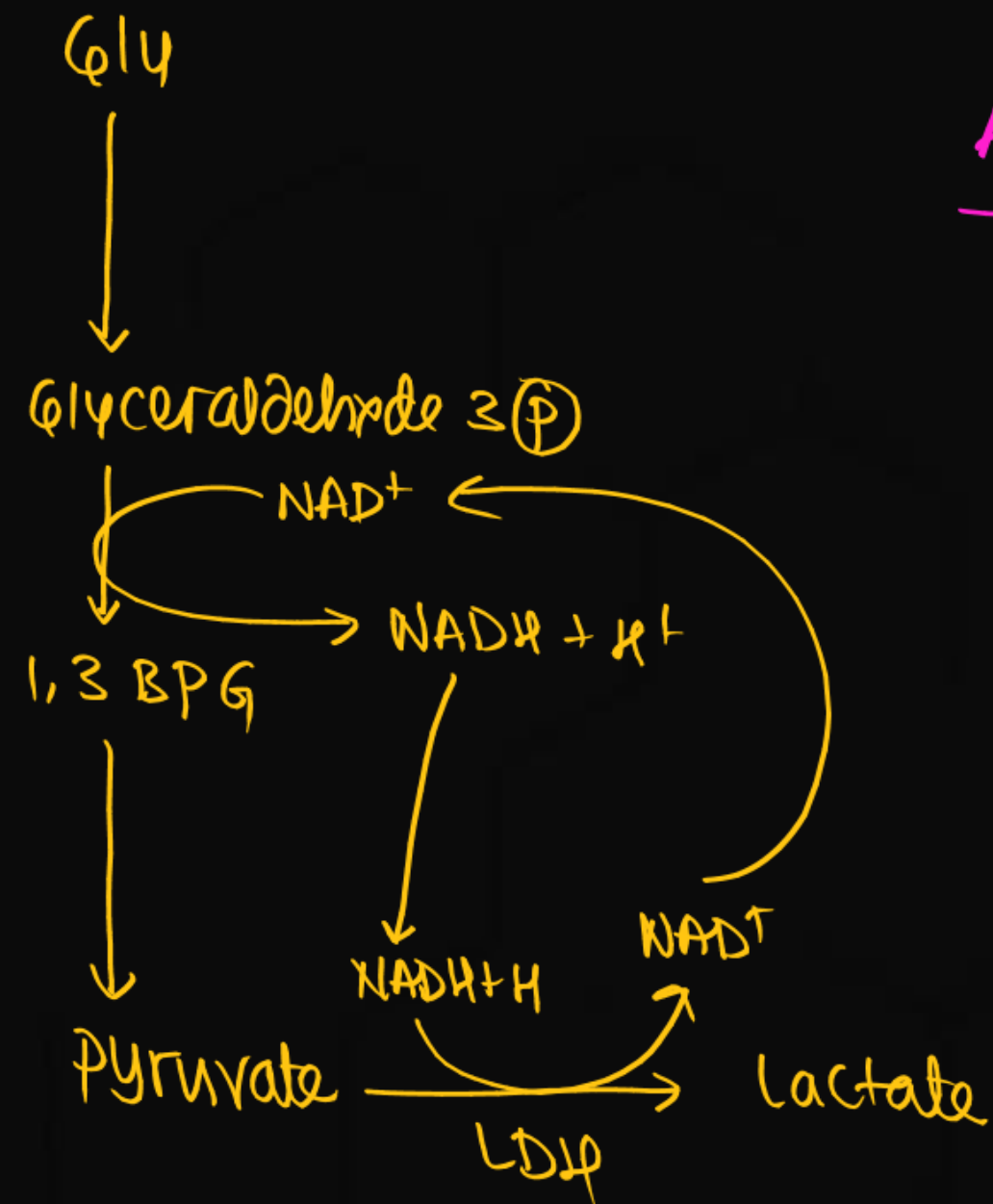
Aerobic



9 ATP

- 2 ATP

7

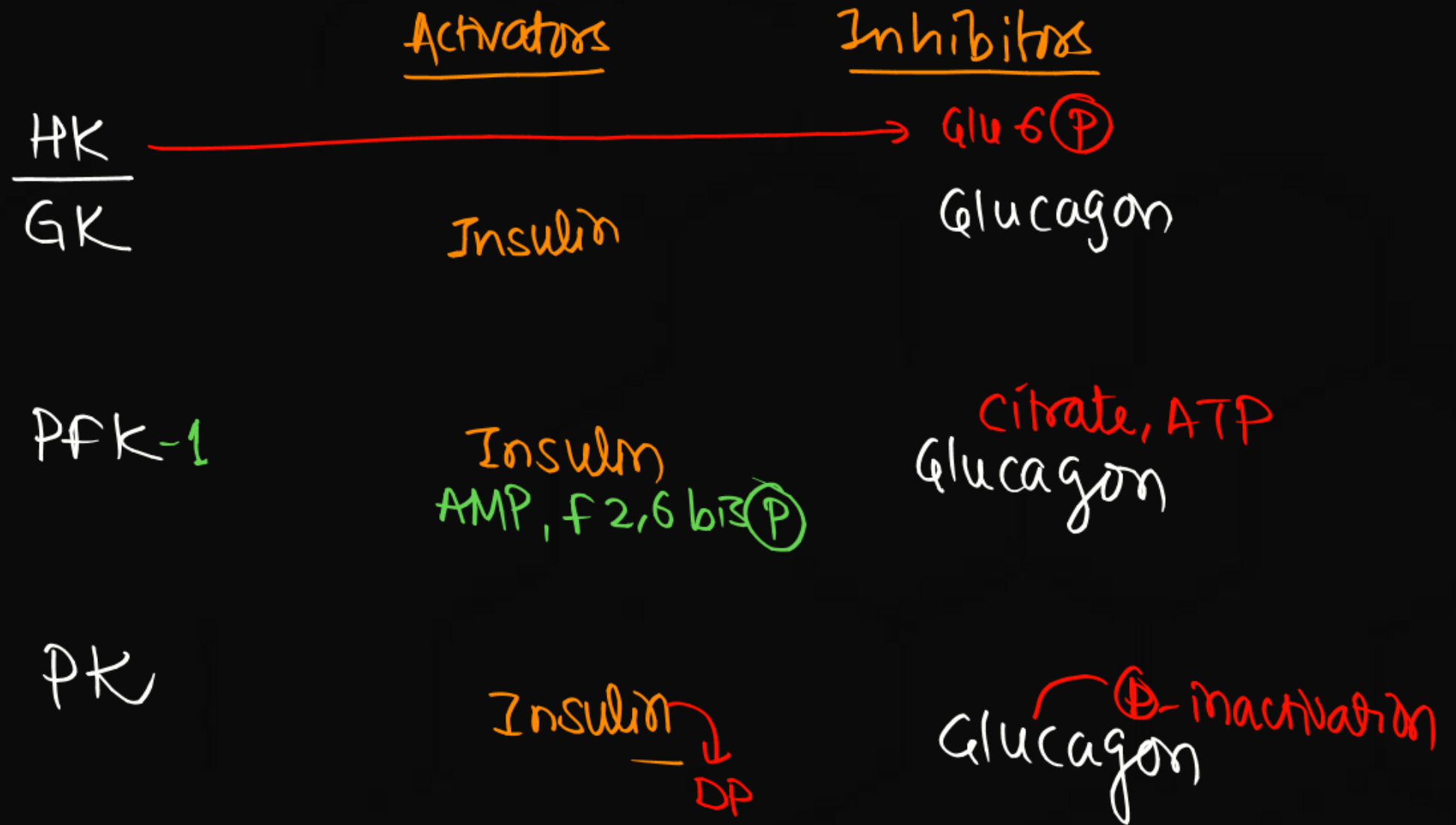


Anaerobic glycolysis

$$\begin{array}{r}
 -2 \text{ ATP} \\
 \times \text{ NADH} + \text{H}^+ \\
 \hline
 4 \text{ ATP} \\
 \hline
 4 - 2 \\
 \hline
 \textcircled{2} \text{ ATP}
 \end{array}$$

Regulation

③ Irreversible Reaction



C/S - fluoride bulb

PFK deficiency

Tarui's ds



- exercise intolerance
- mild hemolysis

1. Hormonal Regulation

Insulin & Glucagon

2. Covalent mod. in Enzy

Ⓟ & ⓈⓅ

3. Allosteric activator

Rapoport leubering cycle

SAQ

RBC's

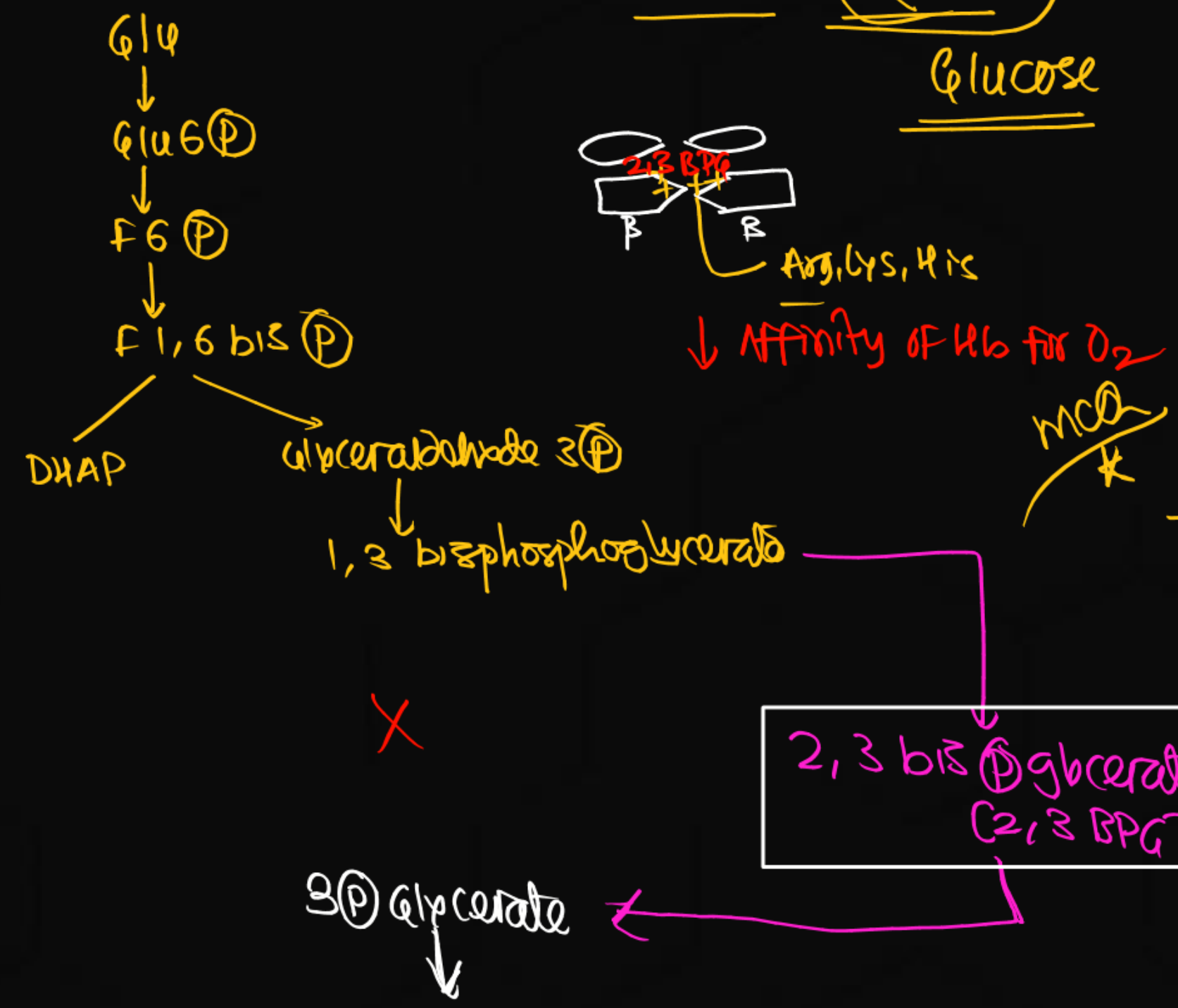
Glucose

What is produced

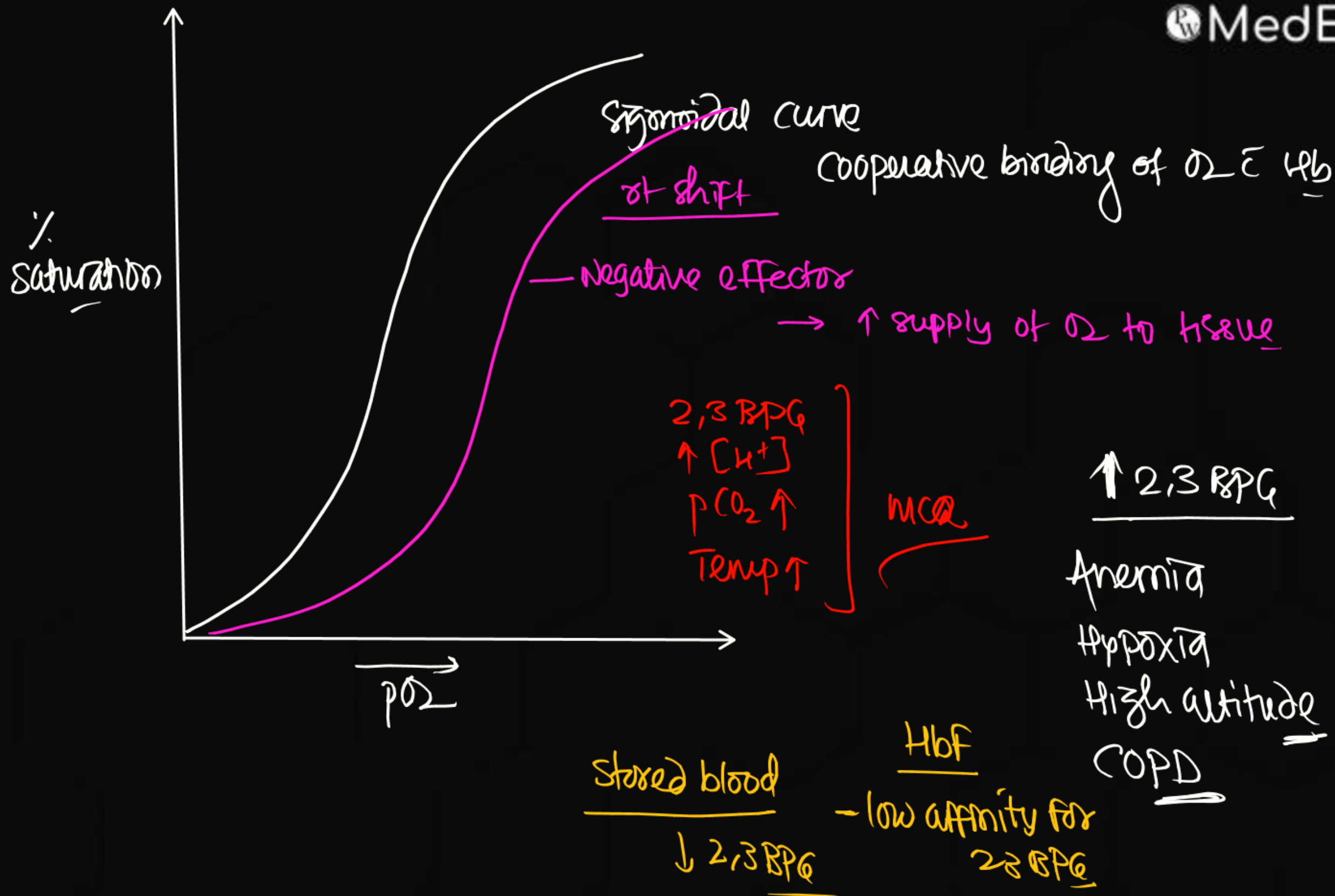
Significance

ATP

st shift



2,3 BPG
 ↑ [H⁺]
 pCO₂ ↑
 Temp ↑



Gluconeogenesis

④ LAQ

Non carbohydrate precursors (substrate)

Lactate

Glycerol

propionyl coA

Glucogenic AA

Glucose

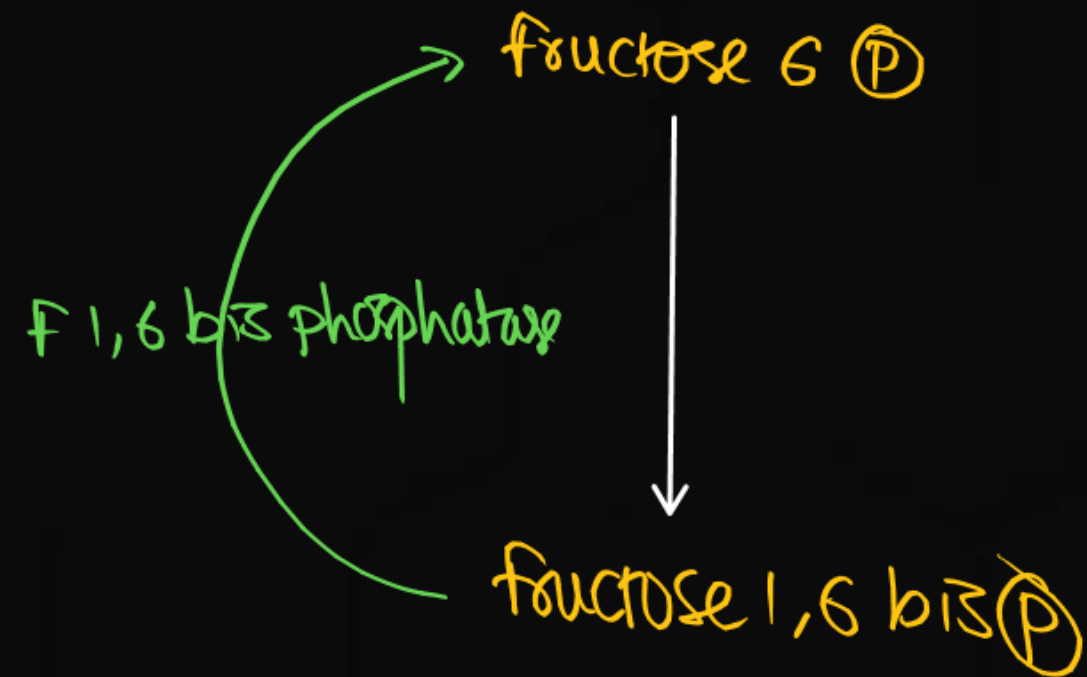
12-18 → 48 Ps
Start

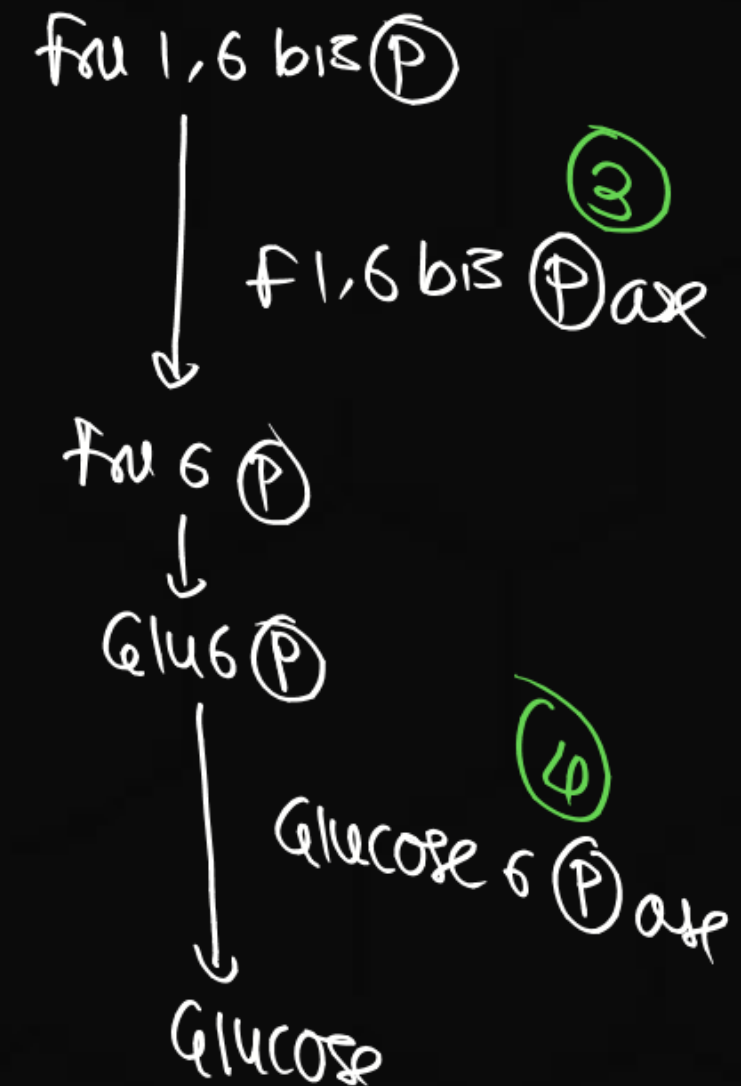
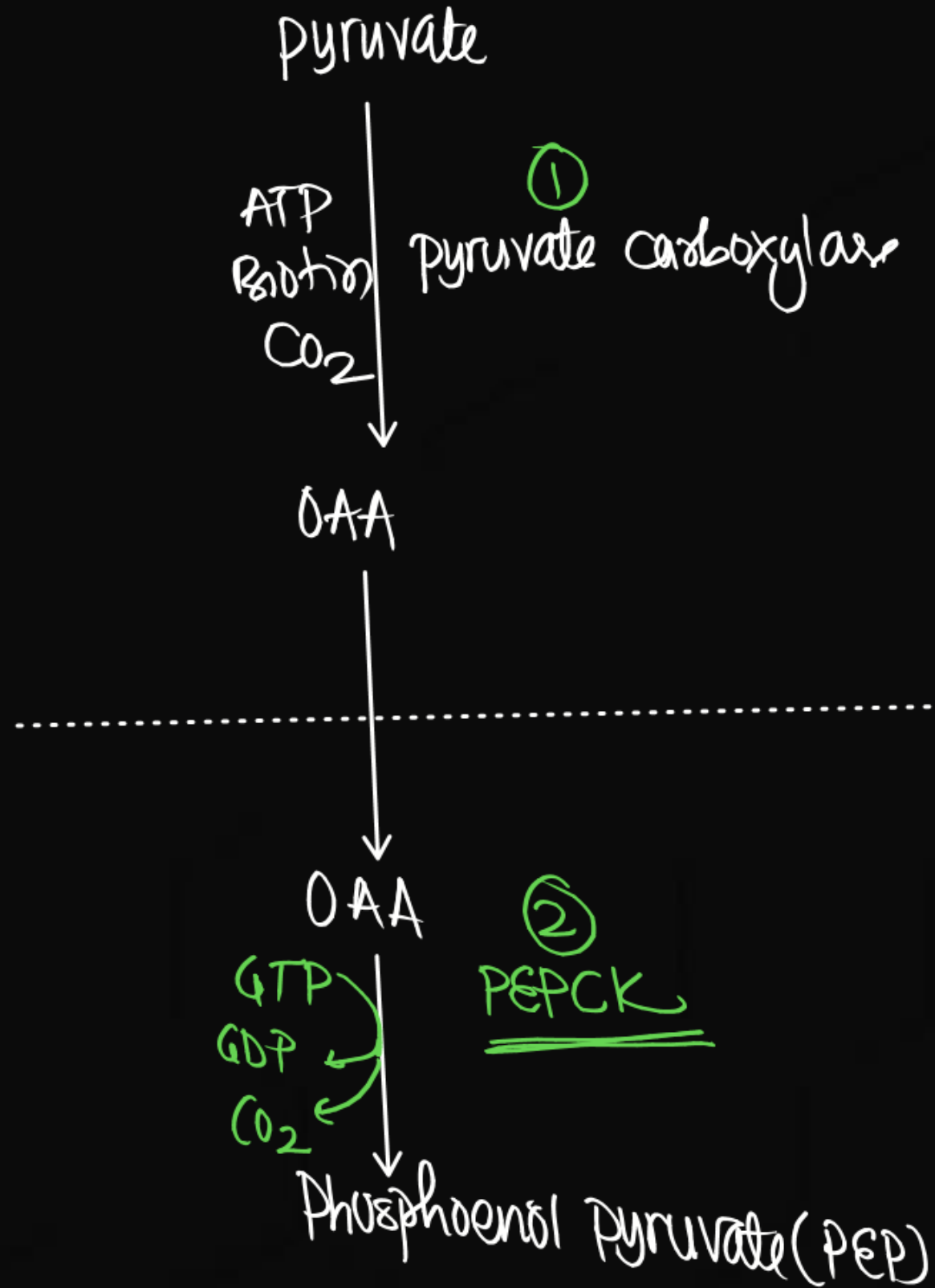
MedEd+

* which of the following will not give u glucose.

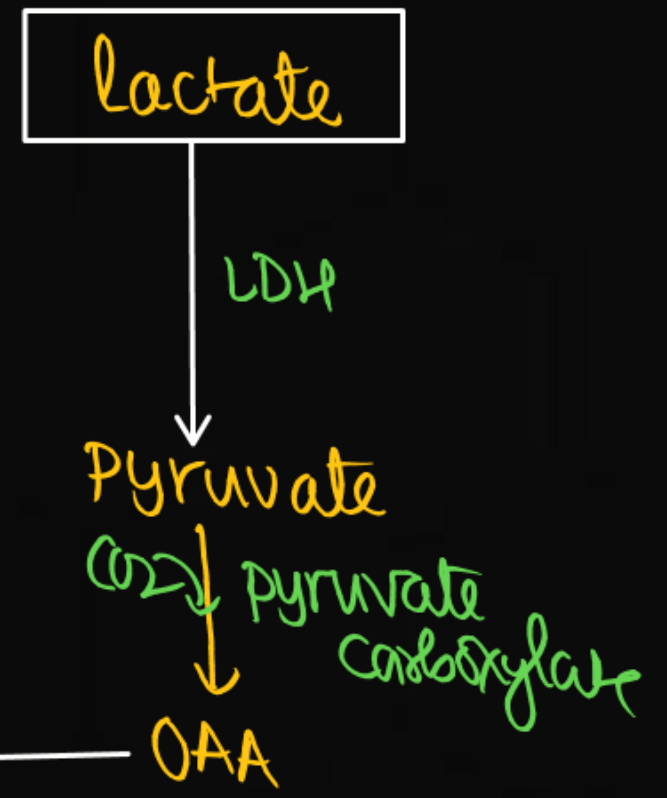
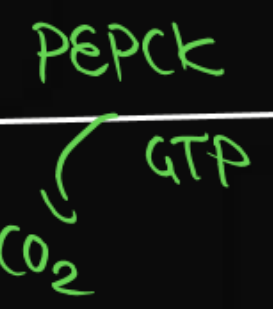
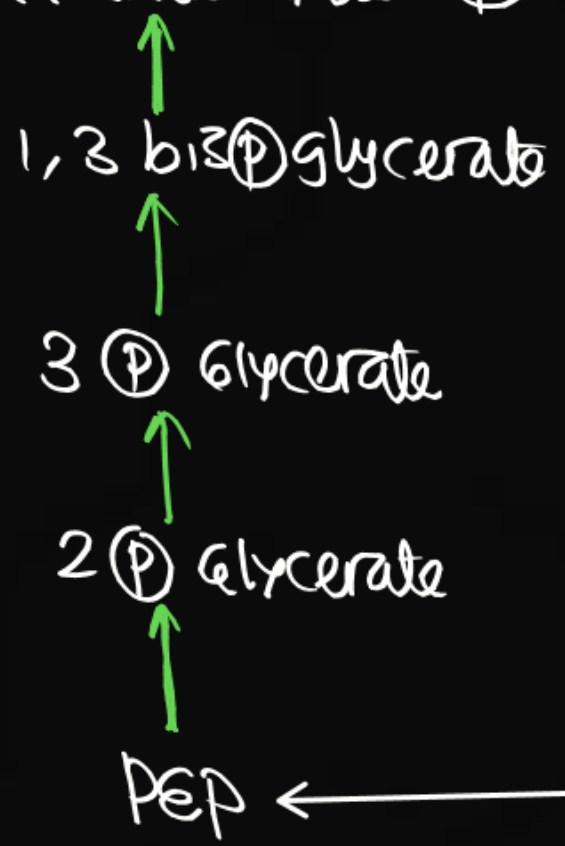
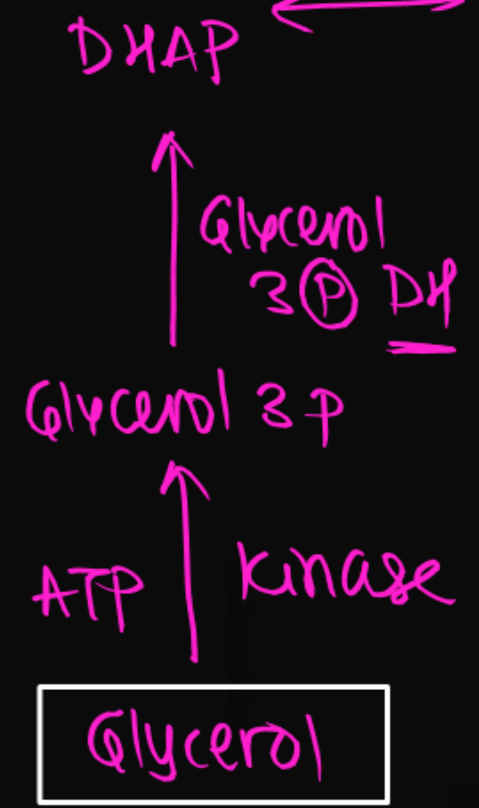
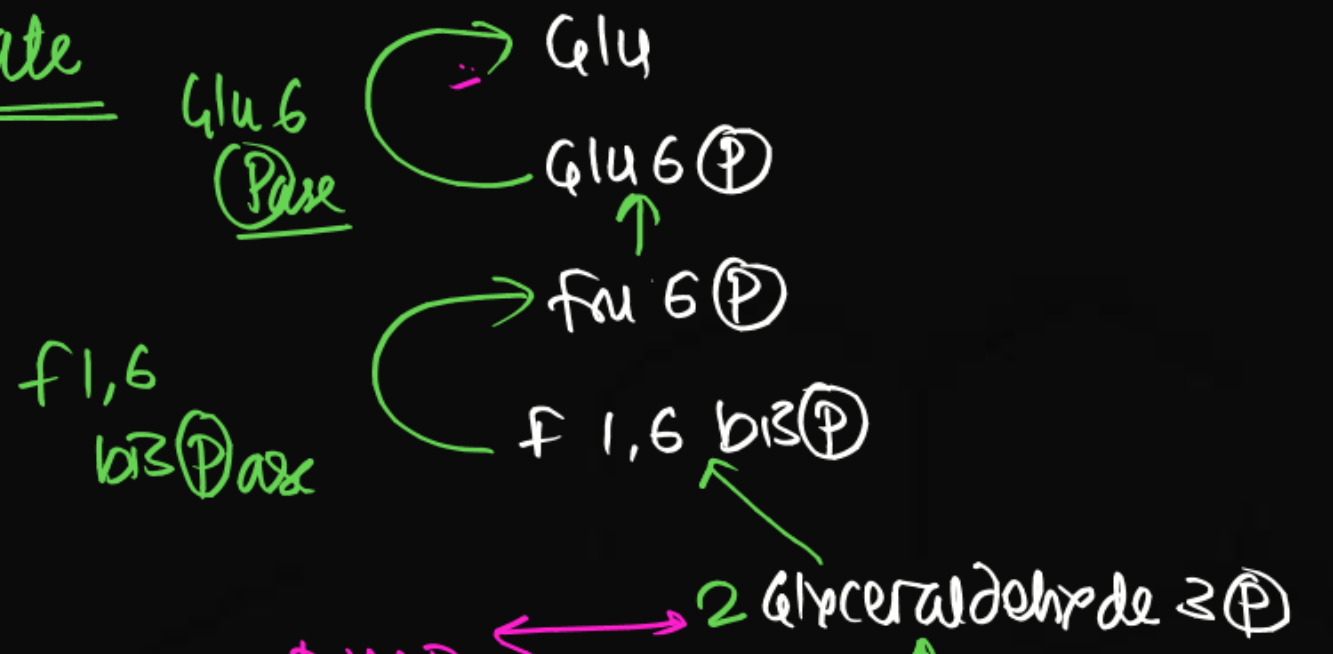
- Even chain FA
- Acetyl coA

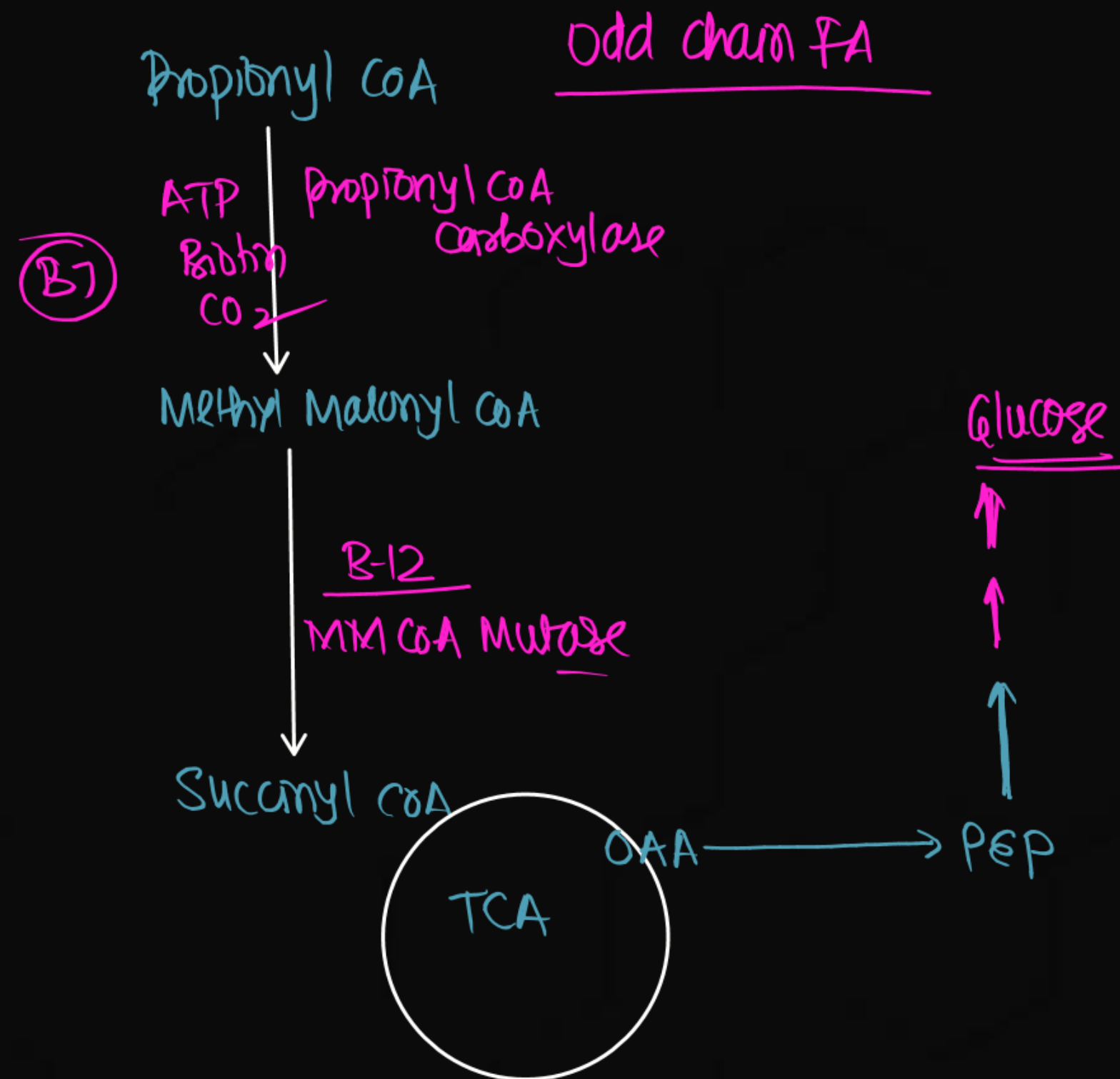
Irreversible steps of Glycolysis

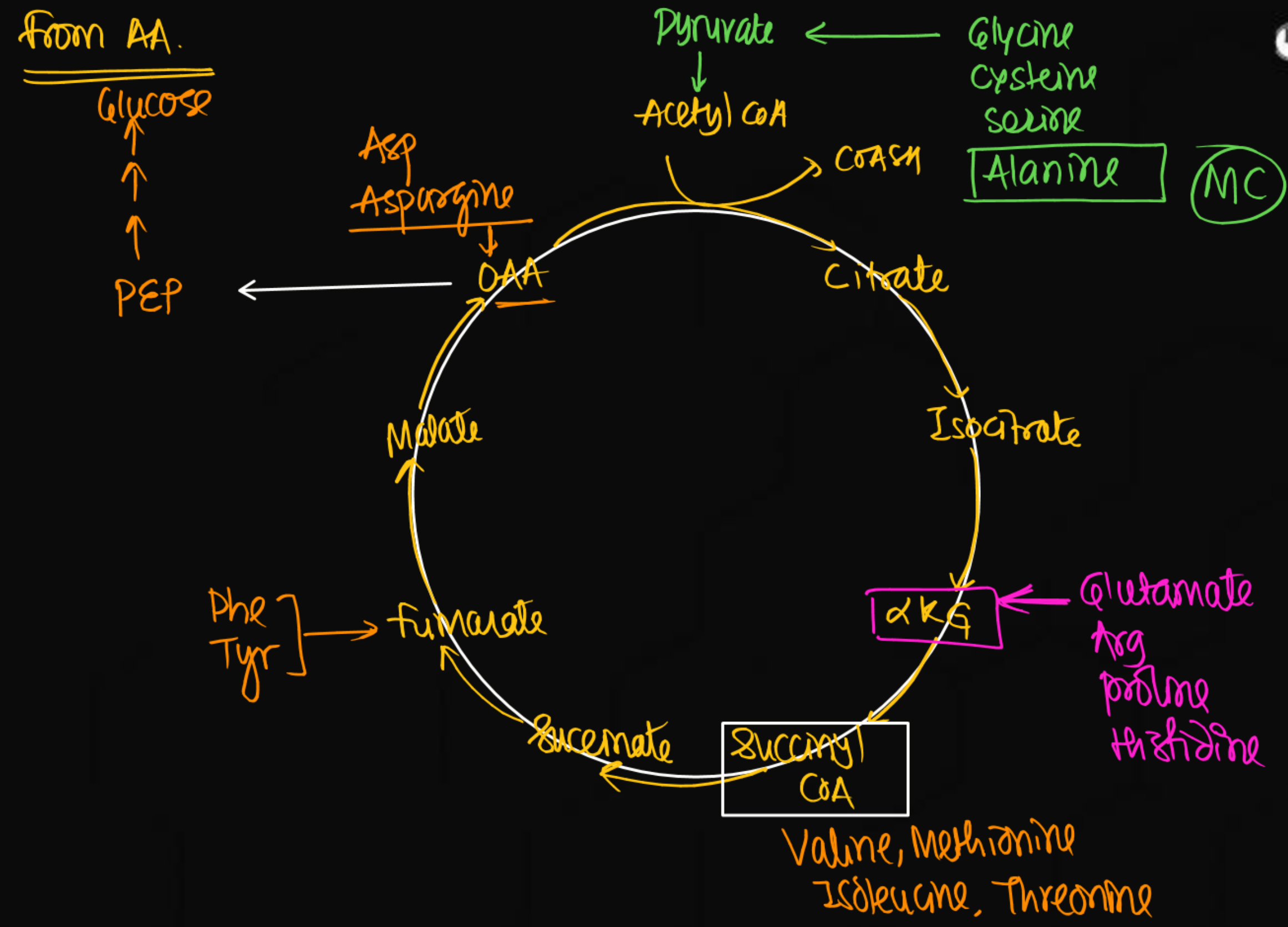


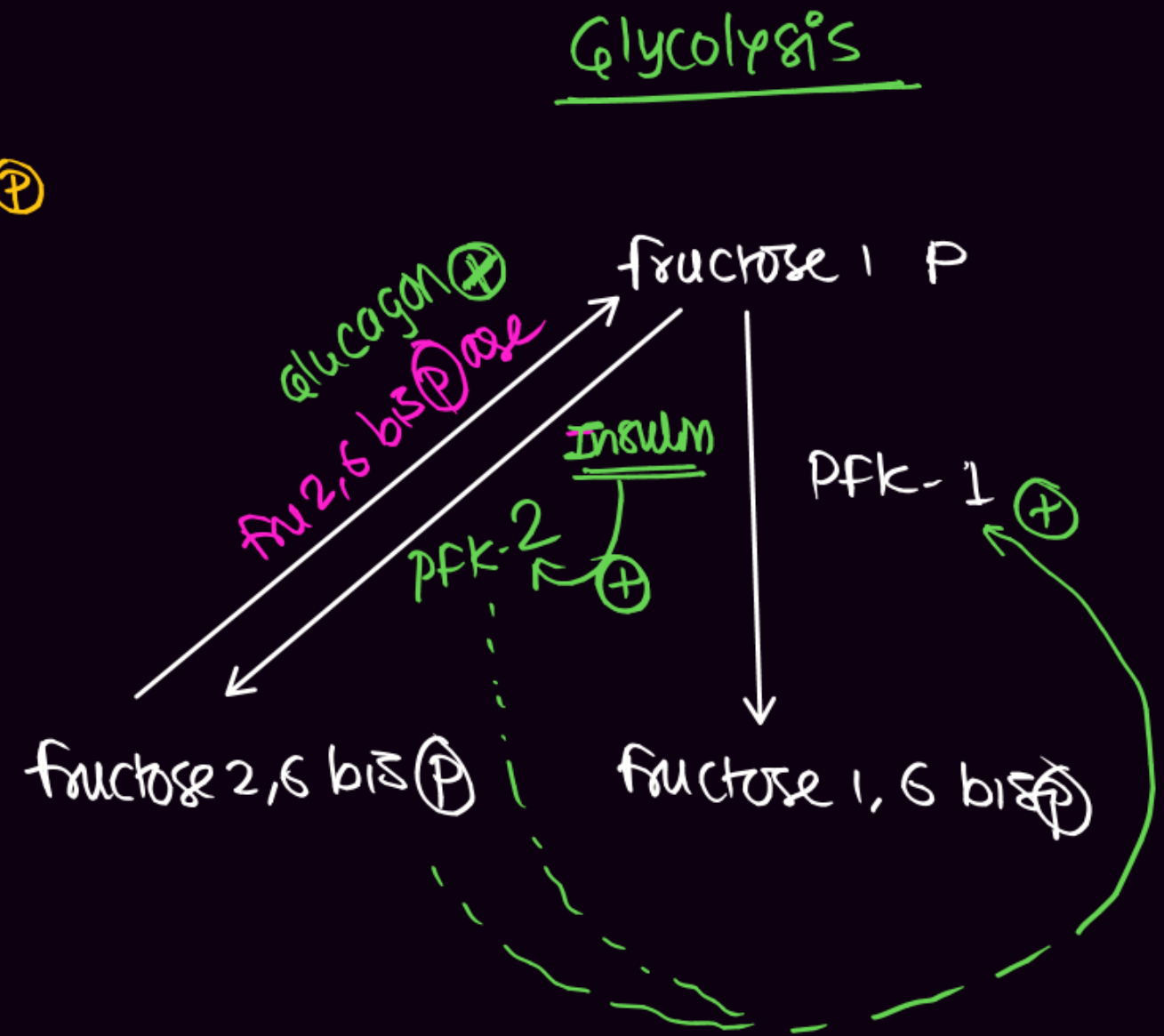
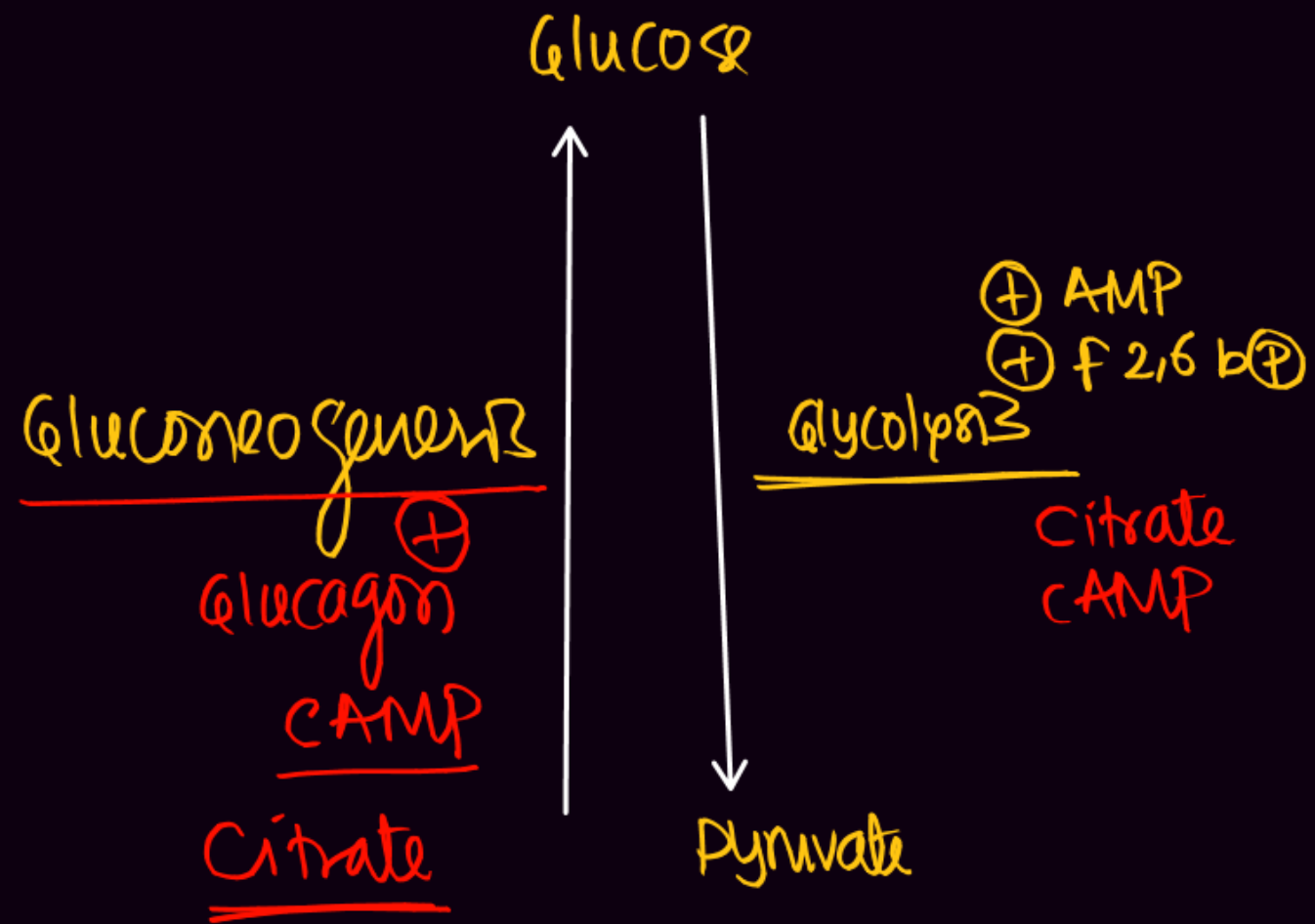


Lactate

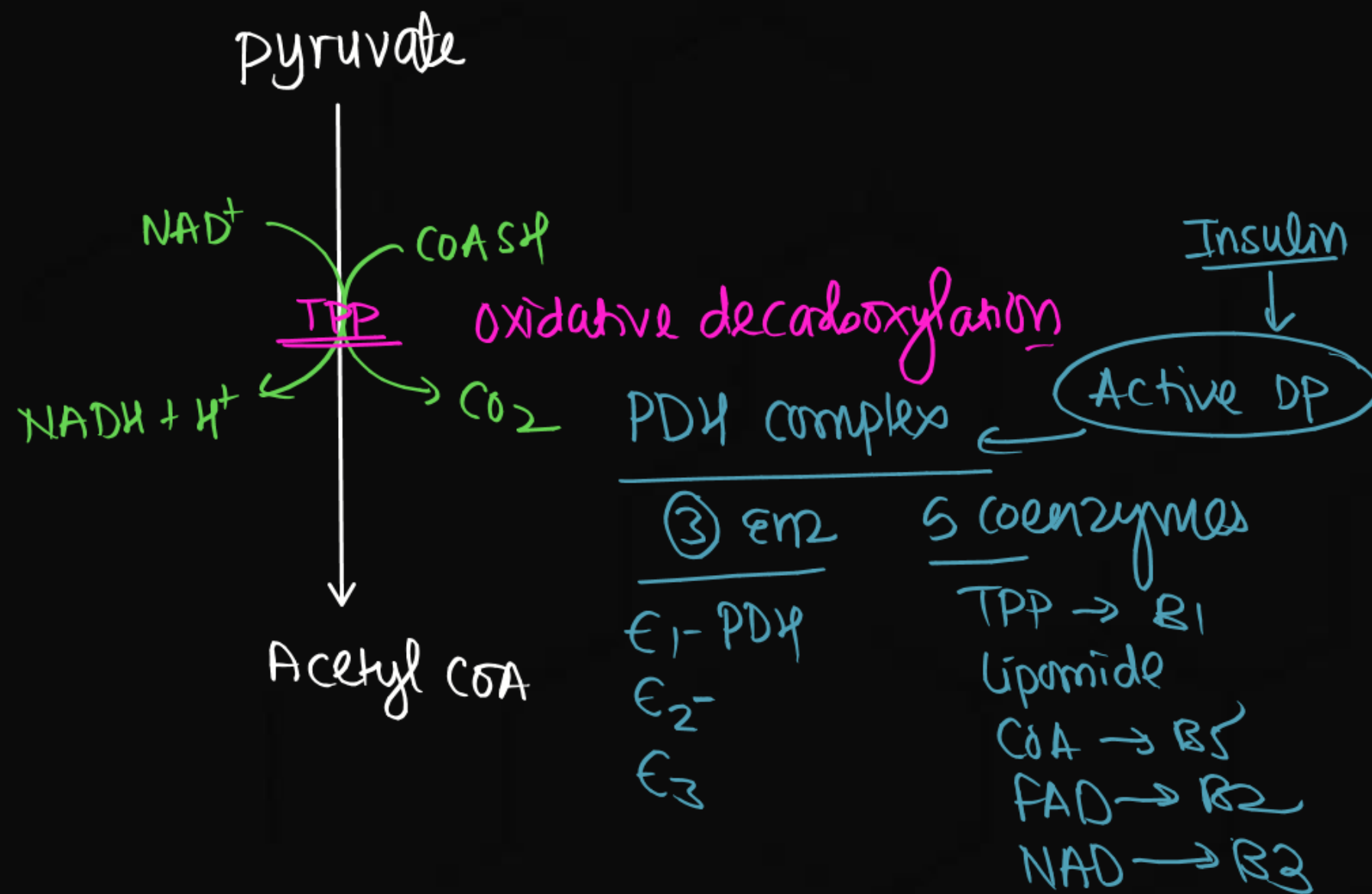


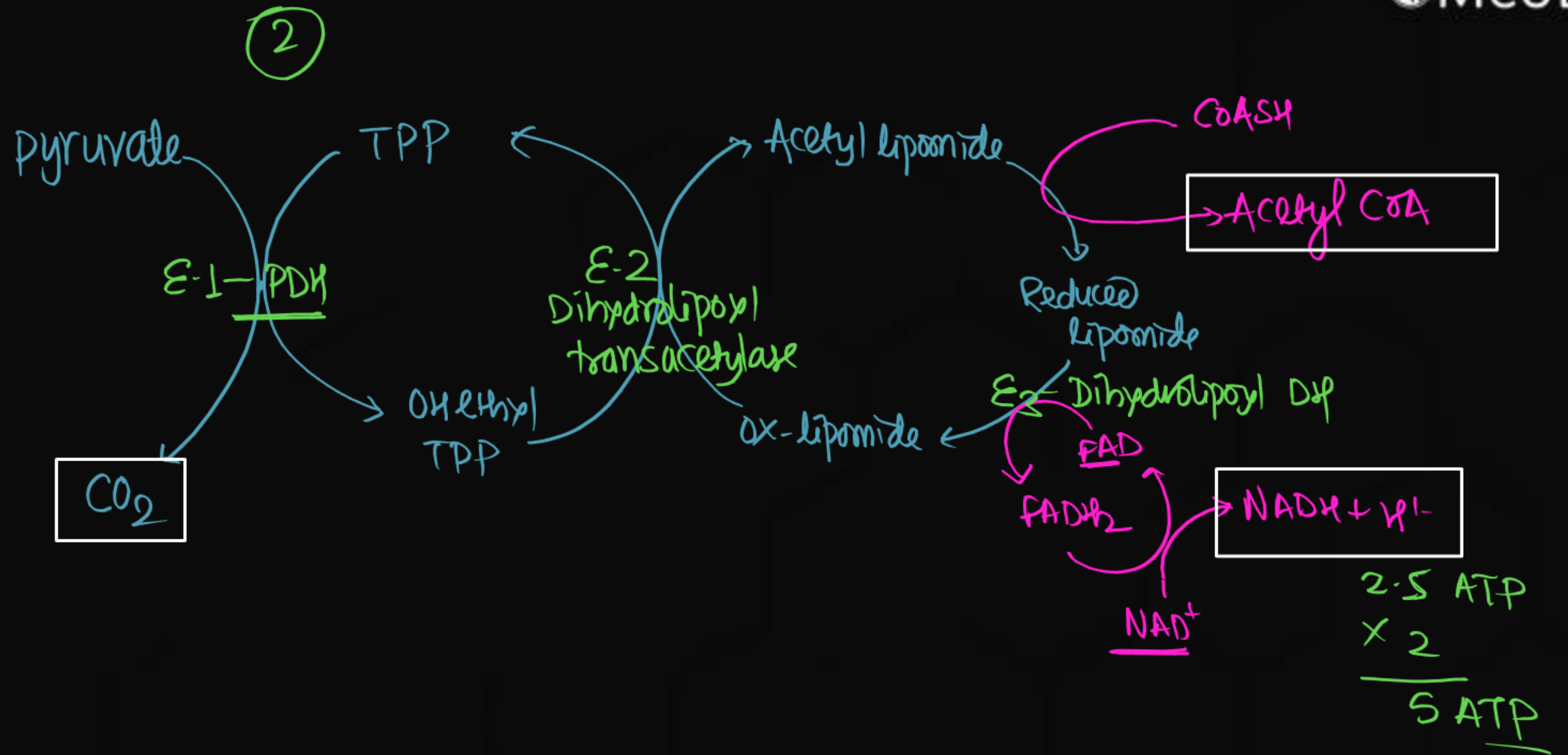




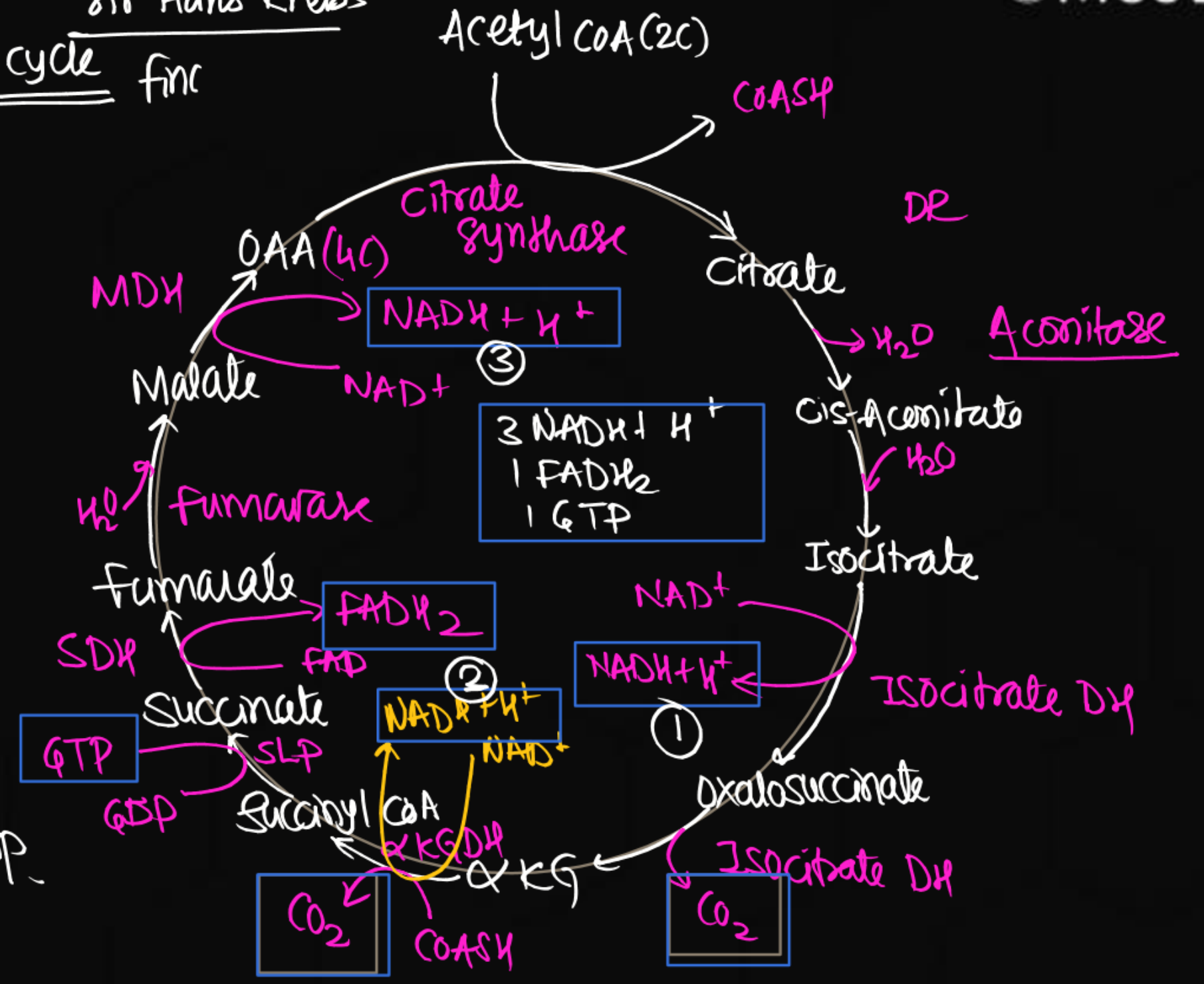
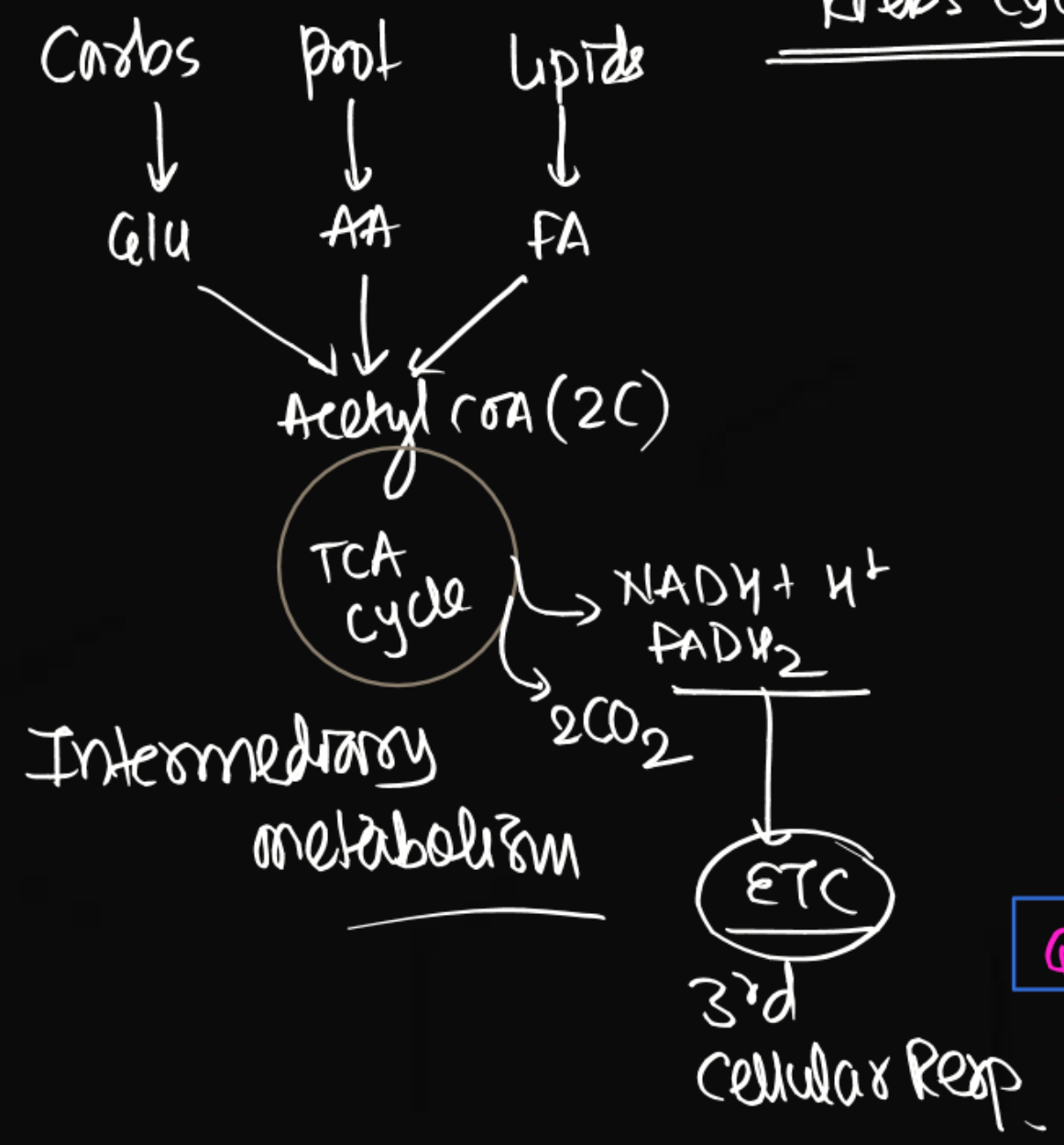


PDH complex





TCA Sir Hans Krebs
Krebs cycle fine



Energetics

$$3 \text{ NADH} + \text{H}^+ = 3 \times 2.5 = 7.5$$

$$1 \text{ FADH}_2 = 1 \times 1.5 = 1.5$$

$$1 \text{ GTP}$$

$$1 \text{ ATP} = 1$$

10 per turn TCA

from glucose 20 ATPS

Glucose

glycolysis 7

PDH 5

TCA 20

32

Regulation ③ Enz

1. Citrate synthase
2. Isocitrate DH
3. α KG DH

⊕

✓ Availability of substrate

✓ ADP

NADH + H⁺
ATPs } ⊖ TCA cycle

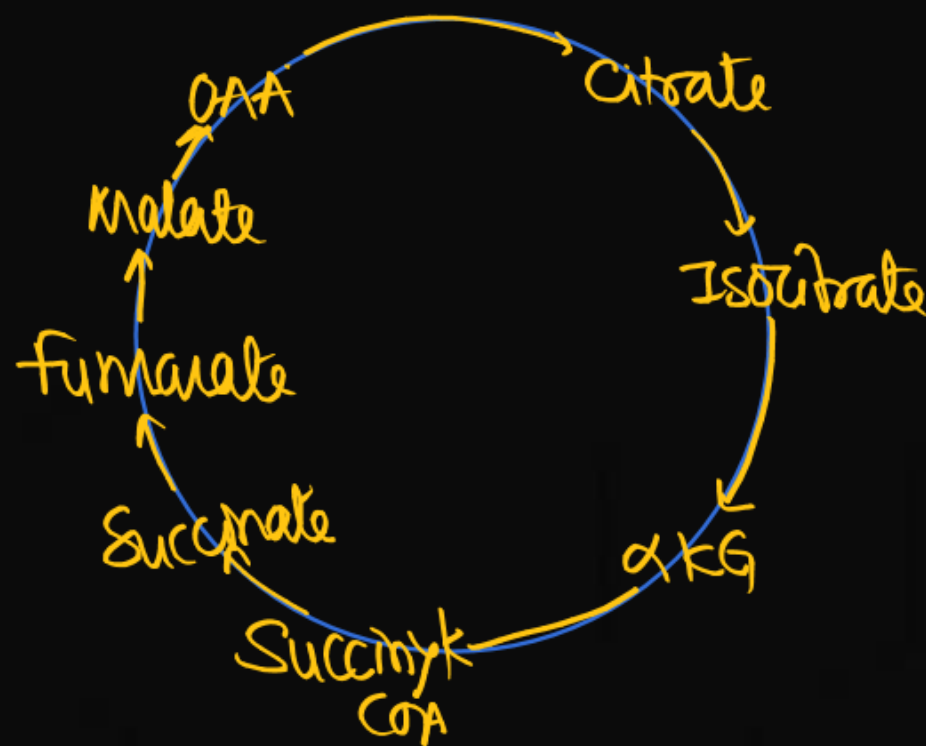
Amphibolic Role of TCA

Anabolism

Catabolism

Acetyl CoA

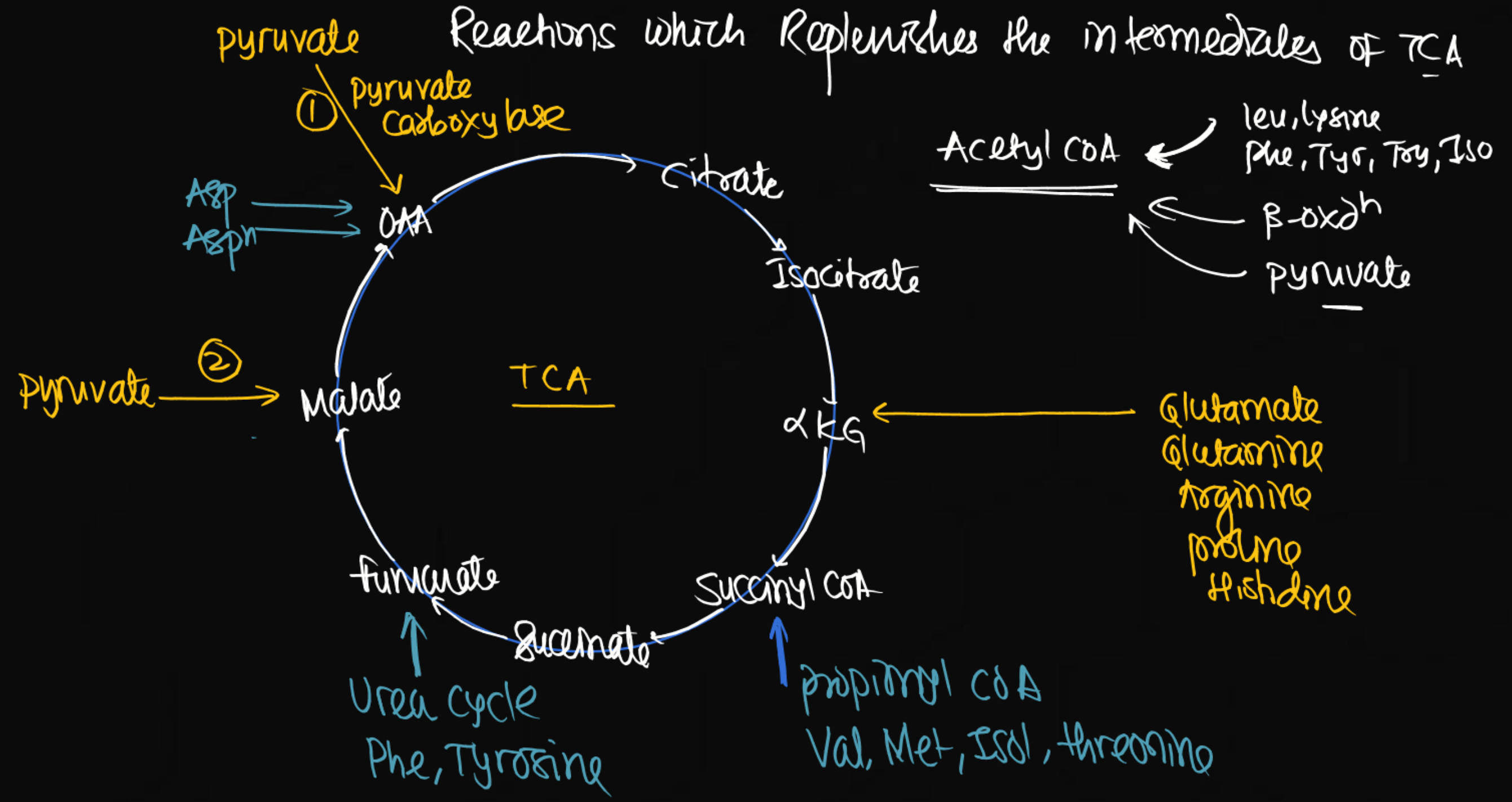
2CO₂



Anabolic Role

1. Citrate $\xrightarrow{\quad}$ Acetyl CoA $\begin{cases} \text{FA synthesis} \\ \text{cholesterol} \end{cases}$
2. α KG $\xrightarrow{\quad}$ Glutamate $\xrightarrow{\quad}$ GABA $\rightarrow$ $\ominus$ NT
Arg, Hist, proline
3. Succinyl CoA $\xrightarrow{\quad}$ Heme synthesis
Ketolysis
4. Oxaloacetate $\xrightarrow{\quad}$ Aspartate
Purine & pyrimidine NT synthesis

Anaplerotic Reactions TCA



Break

10 min

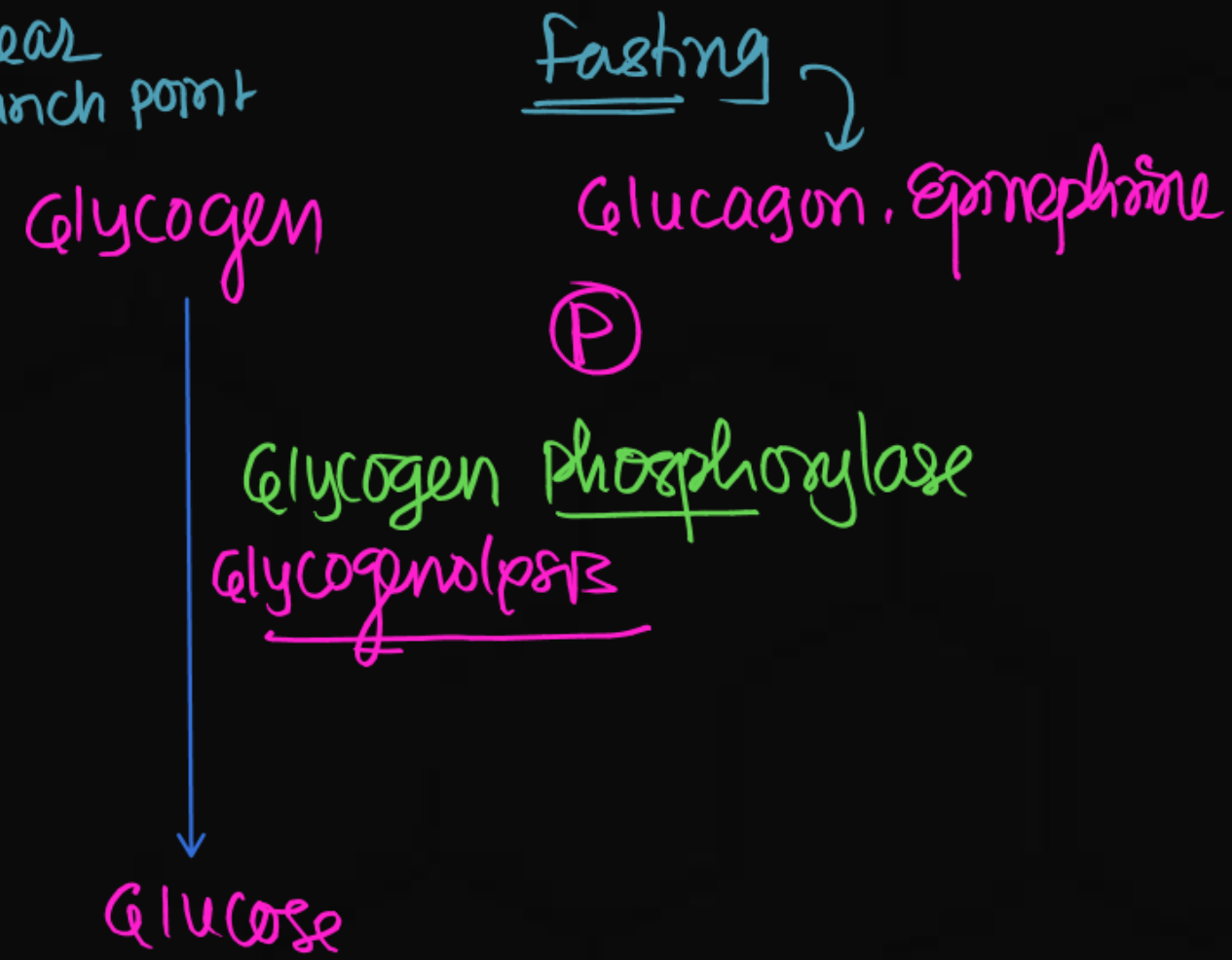
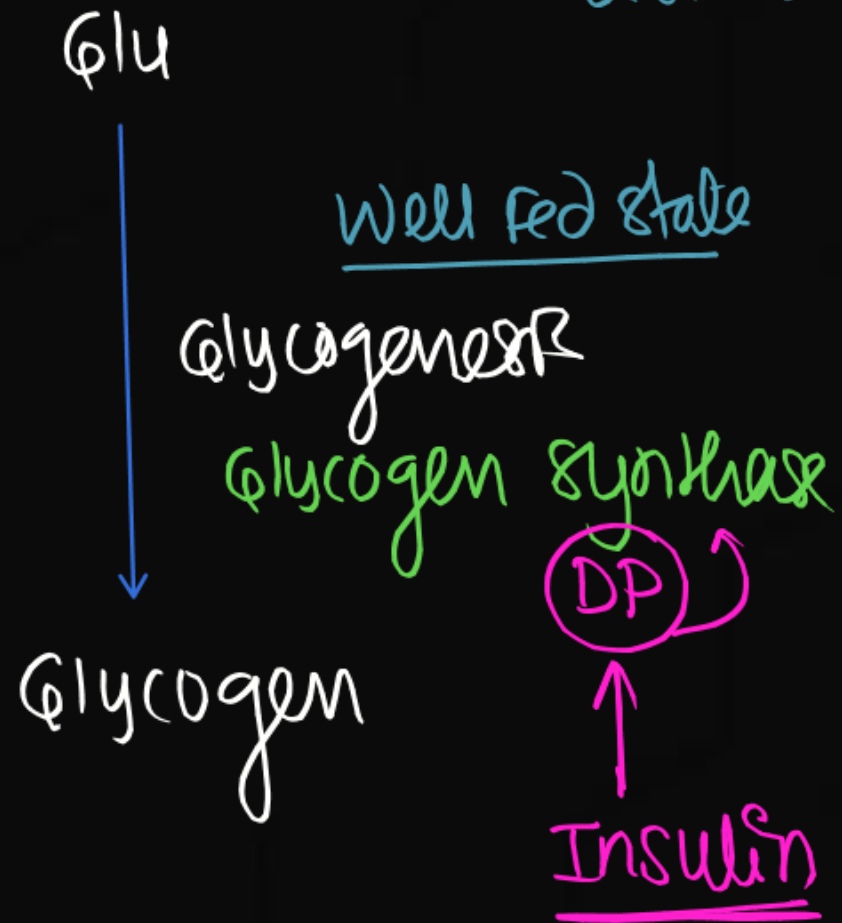
1

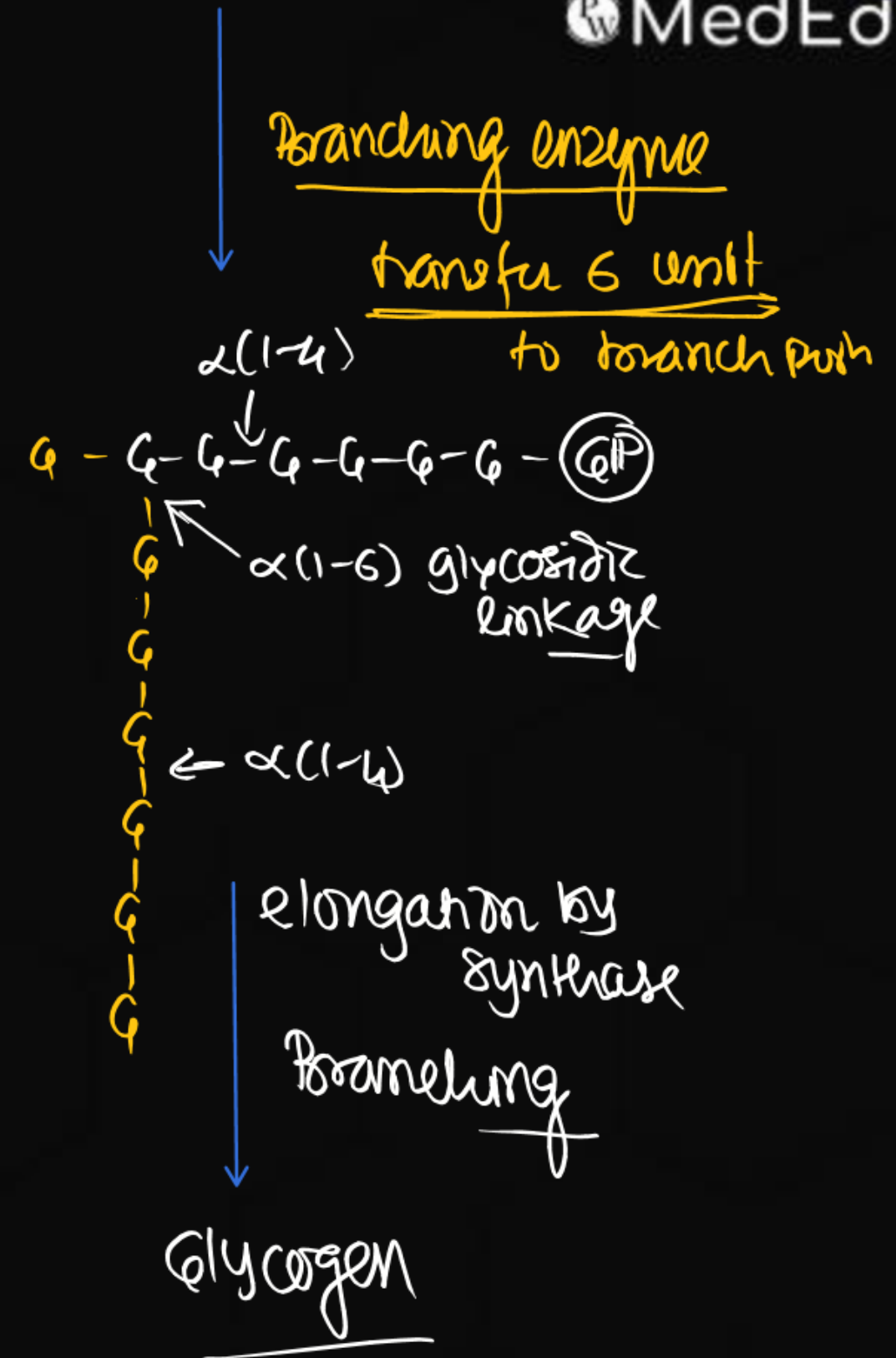
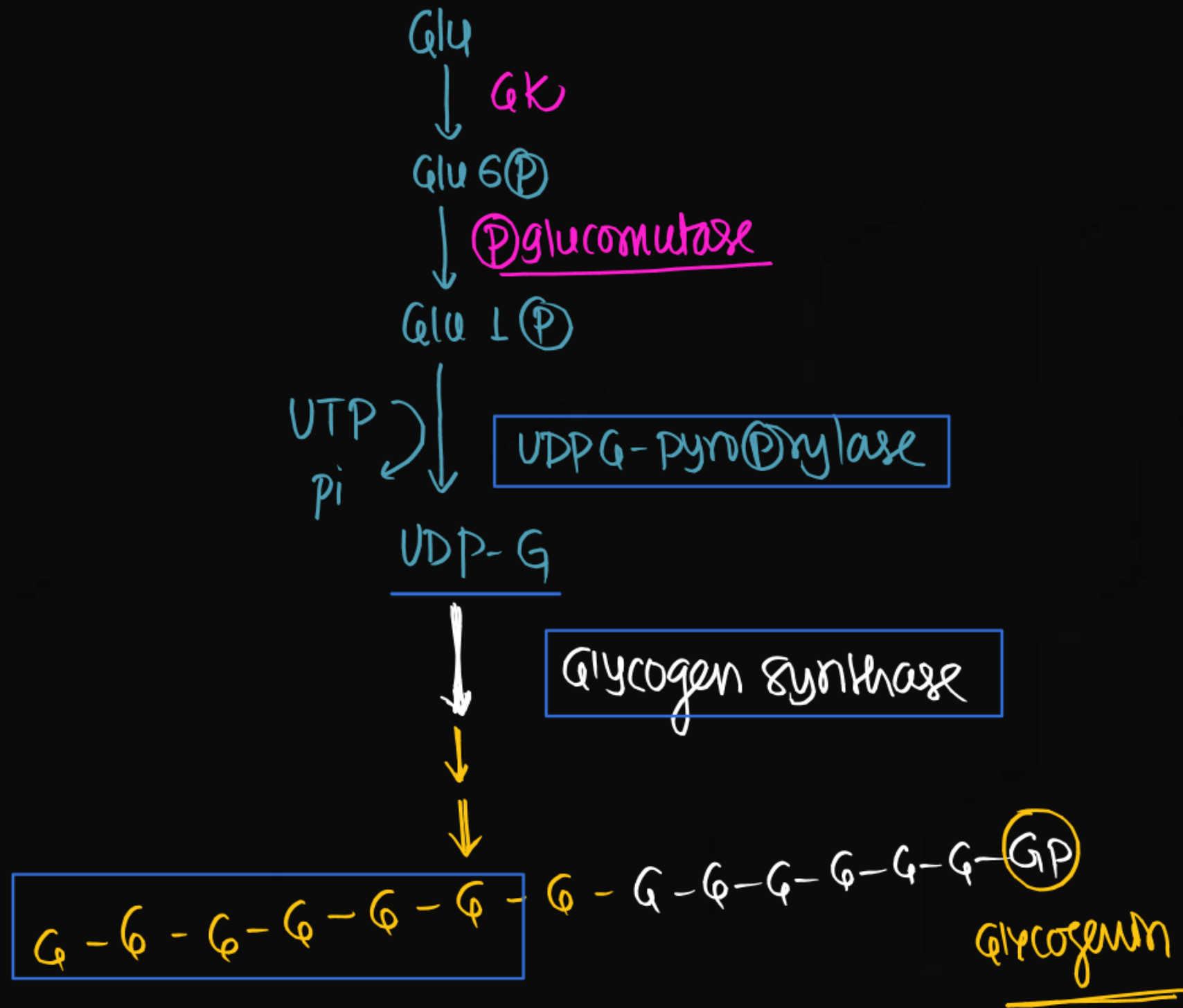
9.42

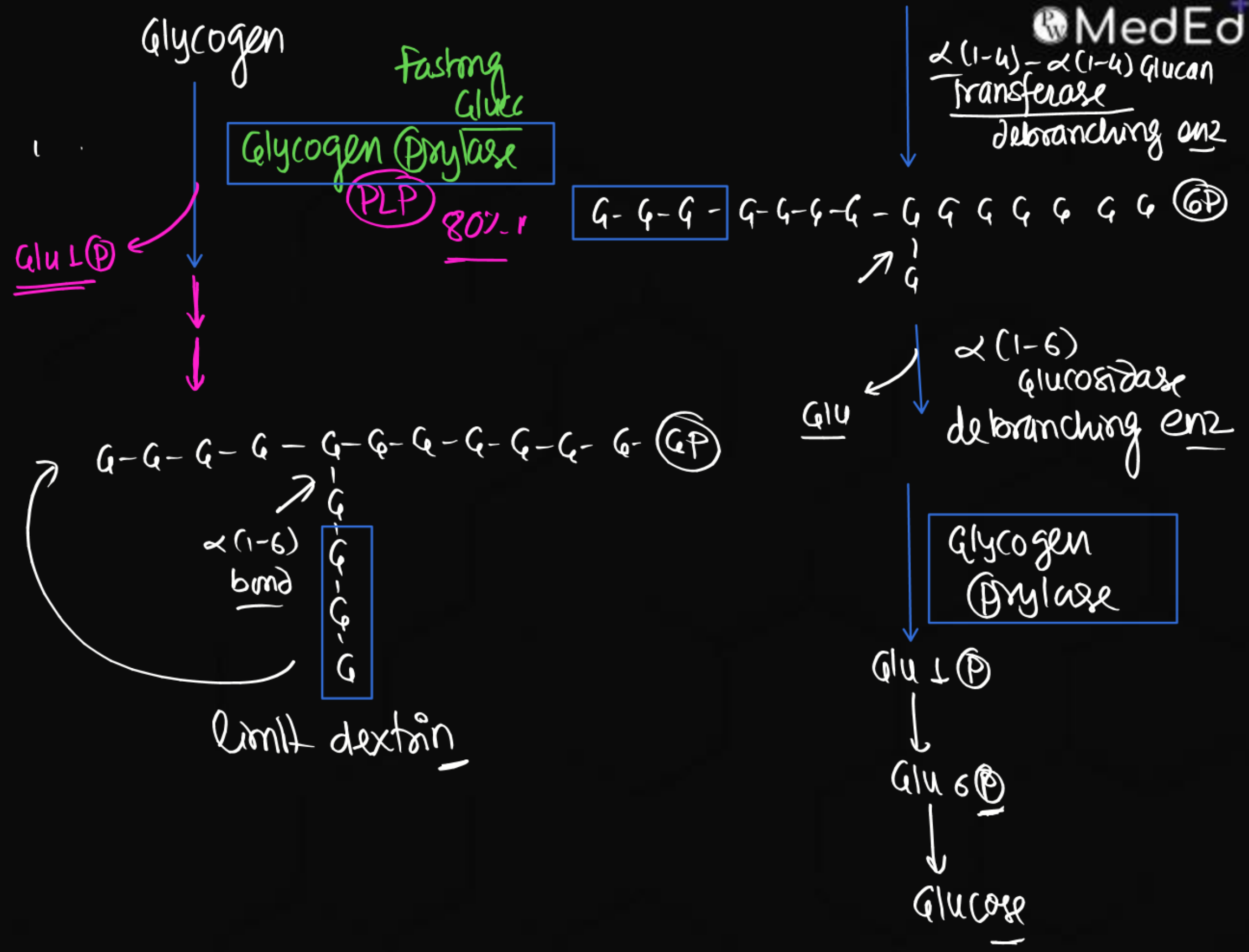
*

start

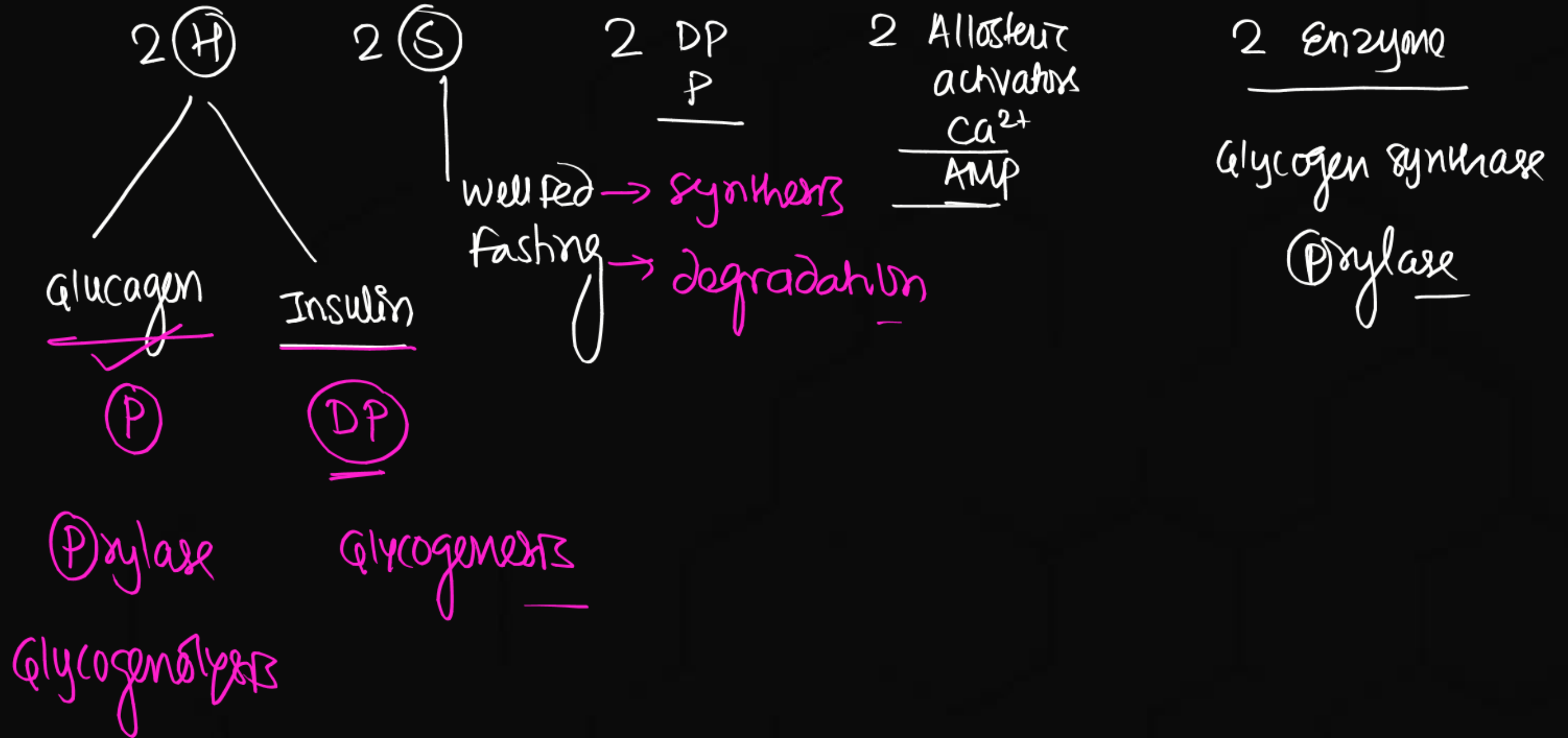
Glycogen → Homopolymer of Glu
 $\alpha(1-4)$ linear
 $\alpha(1-6)$ branch point

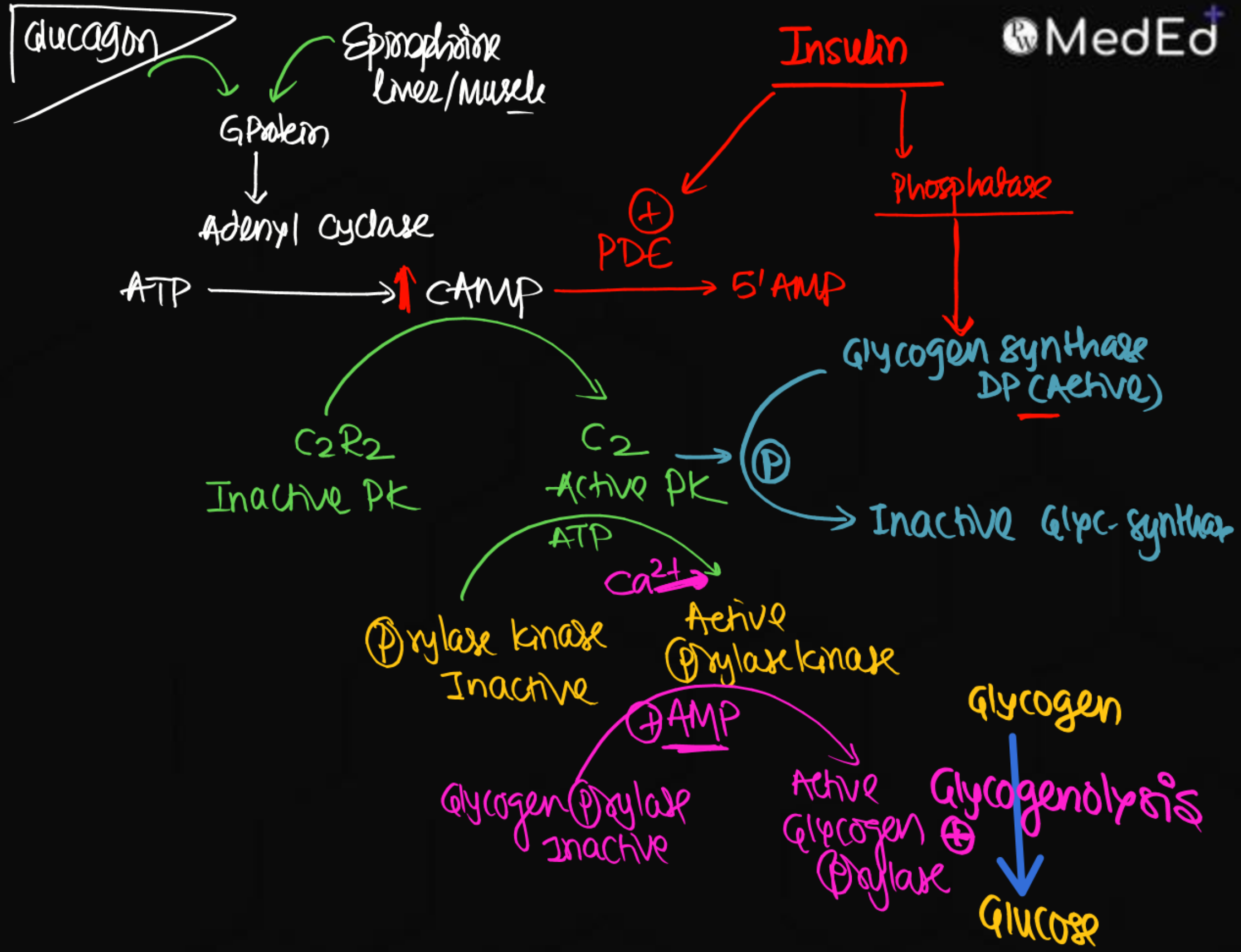






Regulation





GSD

Versatile player Con Anderson Made History
D B

Type 0 → Glycogen synthase

I → Von Gierke's ds → Glu 6 Pase

↓ BSL, ↑ UA, ↑ TG, ↑ LA
chol

II → Pompe's ds → Lysosomal Acid maltase - Heart

III → Cori's ds → Debranching enz

IV → Anderson's ds → Branching enz

V → McArdle's ds → Muscle glycogen Phylase - Exercise intolerance

VI → Hers's ds → liver Glycogen Phylase

VII → Tarui's ds → Muscle PFK

HMP shunt

(SN)

Alternate pathway for oxd^n of Glu $\rightarrow$ Co_2

Ribose $\rightarrow$ RNA & DNA

NADPH

$\rightarrow$ Reductive Biosynthesis

FA
chol
steroid

Synthesis

- Redox buffer

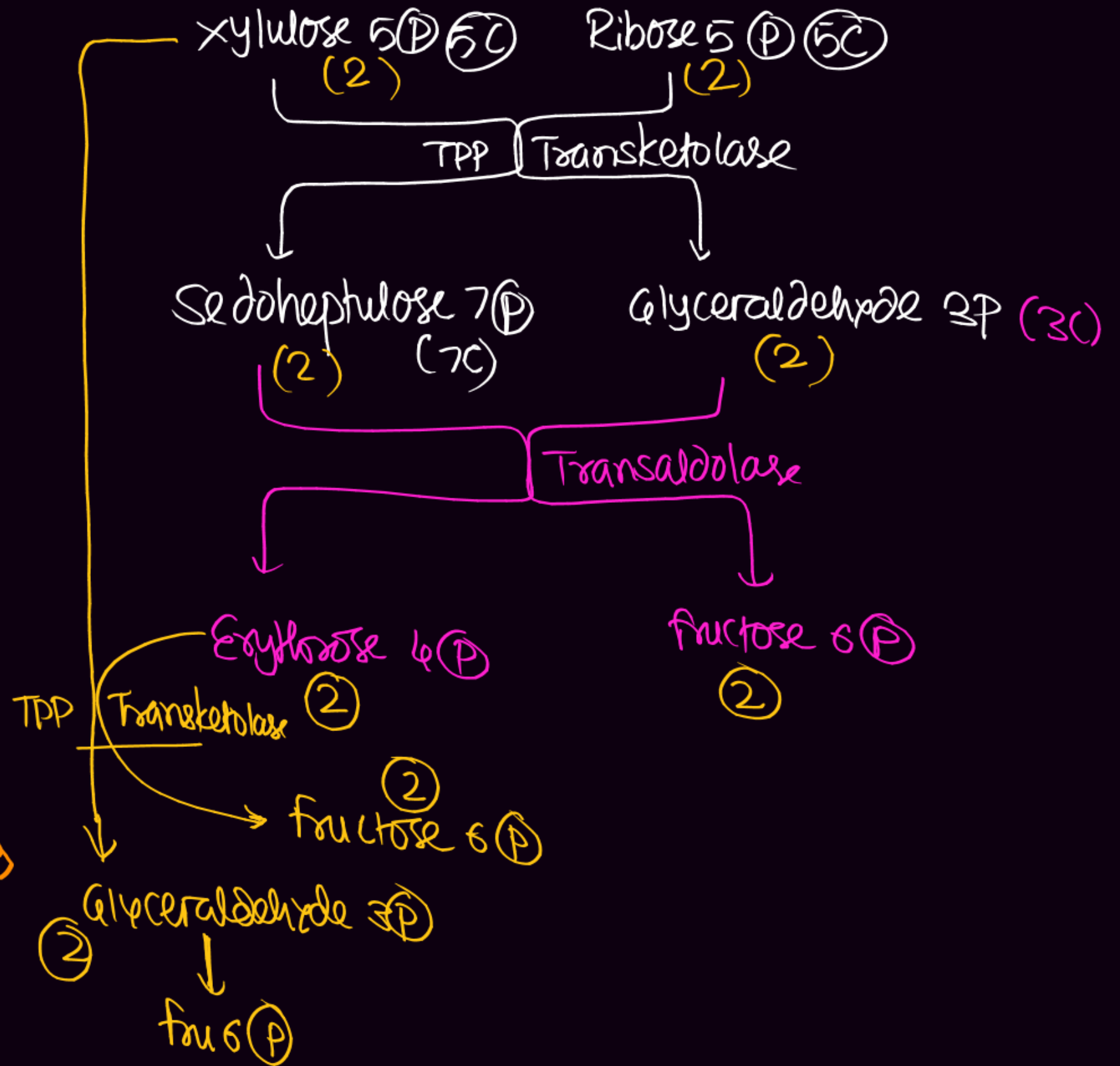
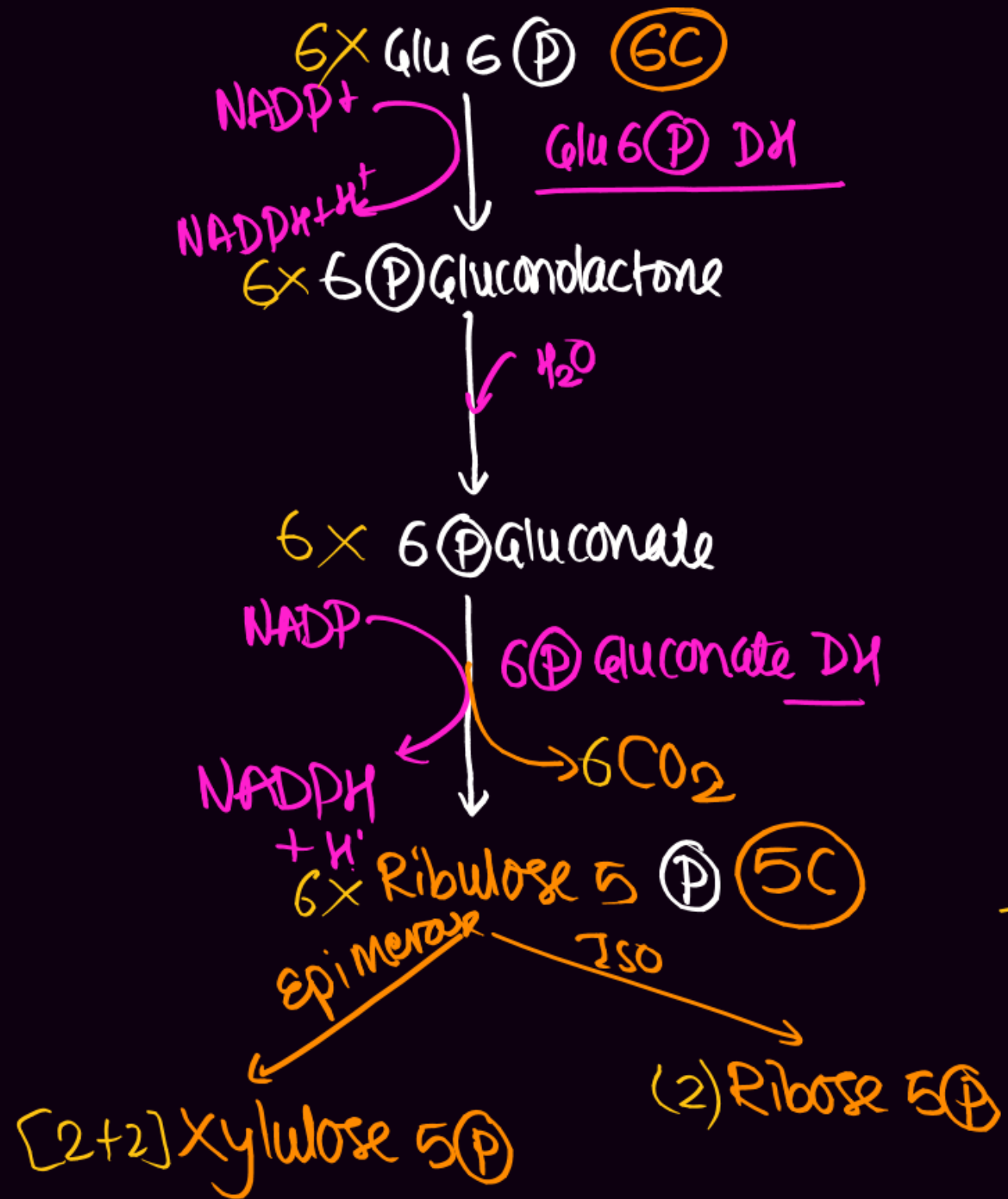
- integrity of RBCs

- prevents MetHb

- Respiratory burst

- Transparency of cornea

- Cyt P 450 enzyme-detoxification.



Primaquine

Oxidant
infⁿ

fava bean



ROS

If G6PDH ↓

↓ NADPH

Hemolysis

Jaundice

6 Glu 6 (P)

+

6 H₂O

+

12 NADP⁺



5 Fru 6 (P)

→

5 Glu 6 (P)

(30)

+

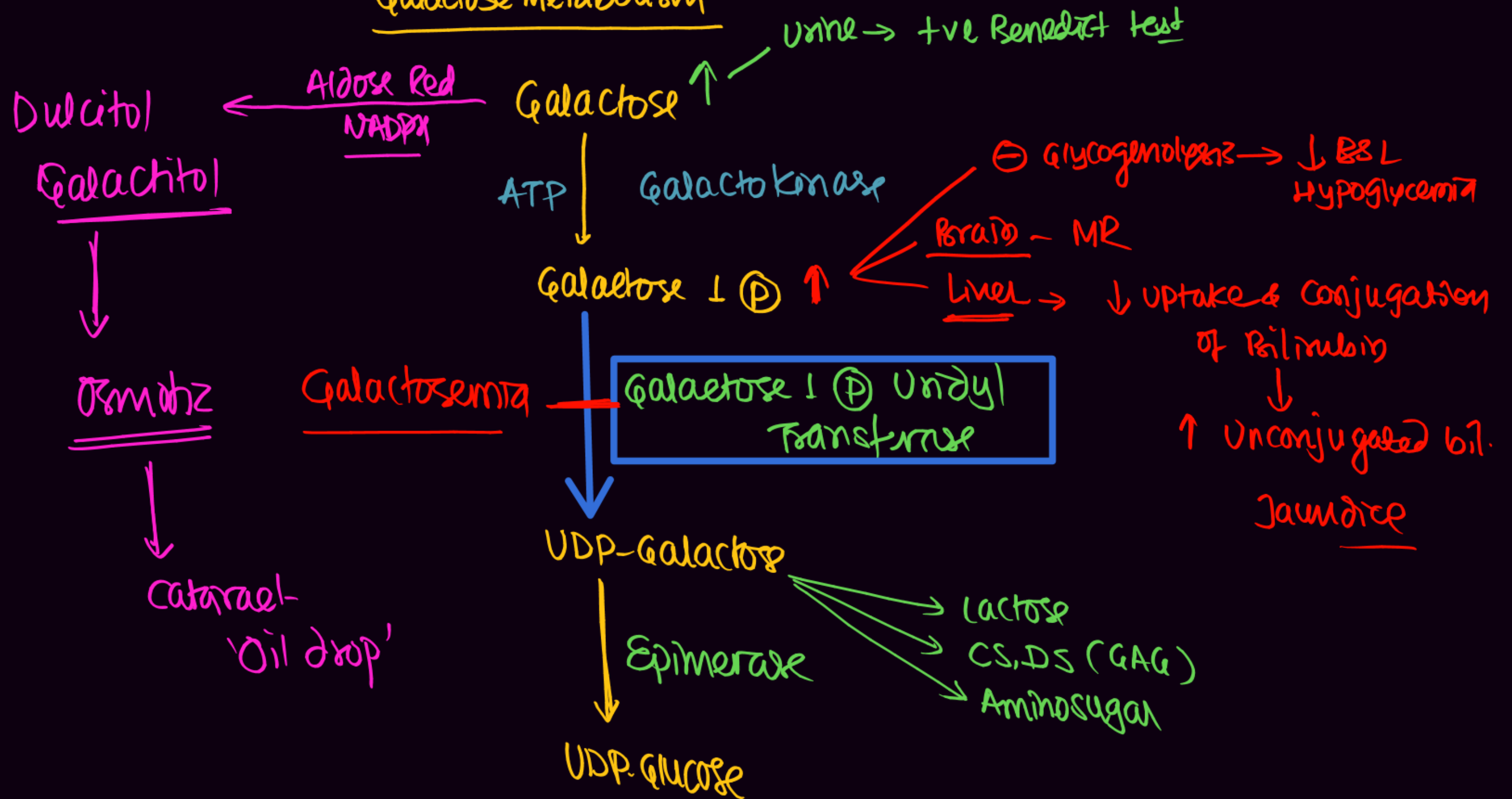
6 CO₂

+

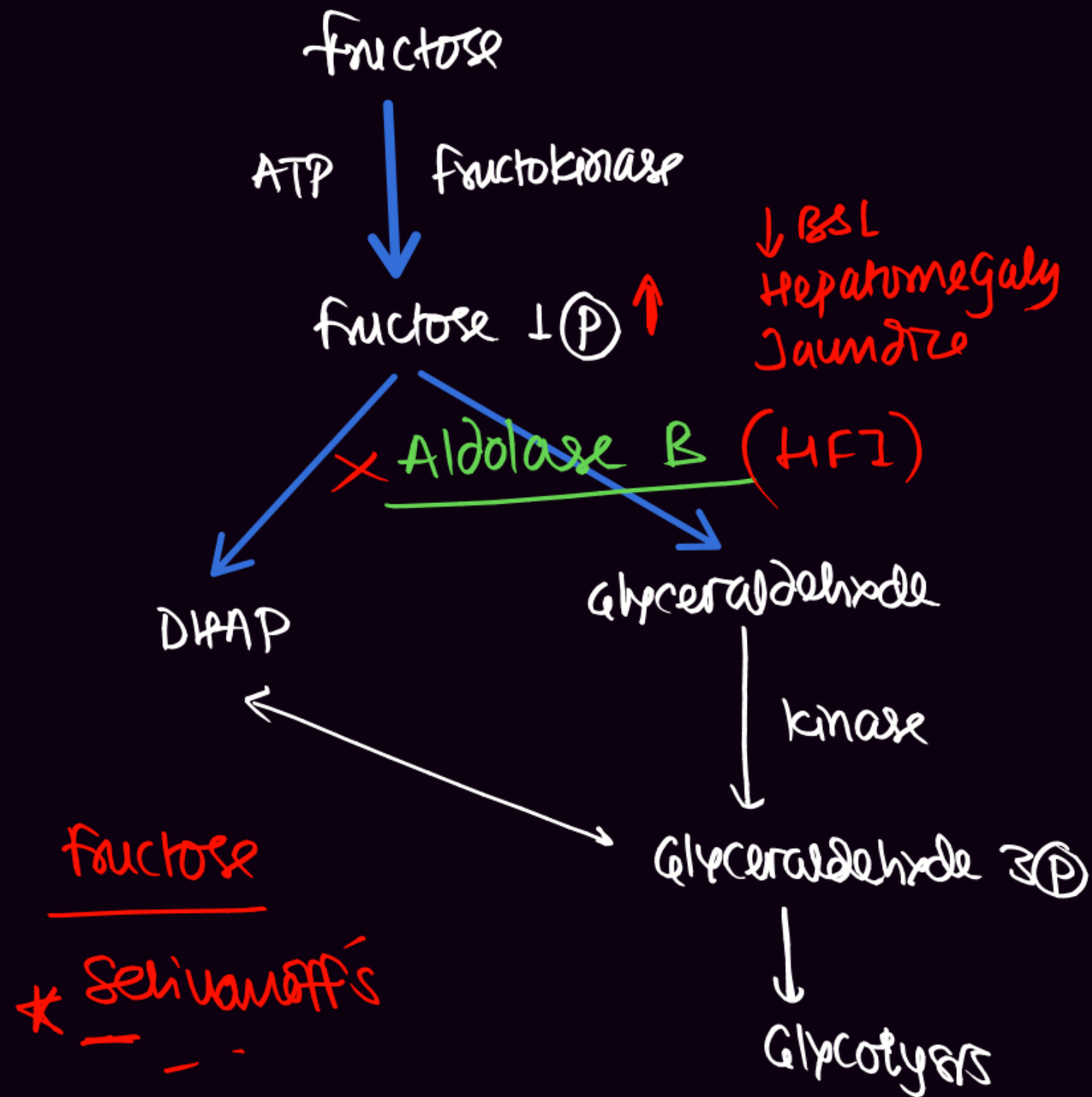
12 NADPH + H⁺

6 × 6 = 36

Galactose Metabolism



Fructose Metabolism

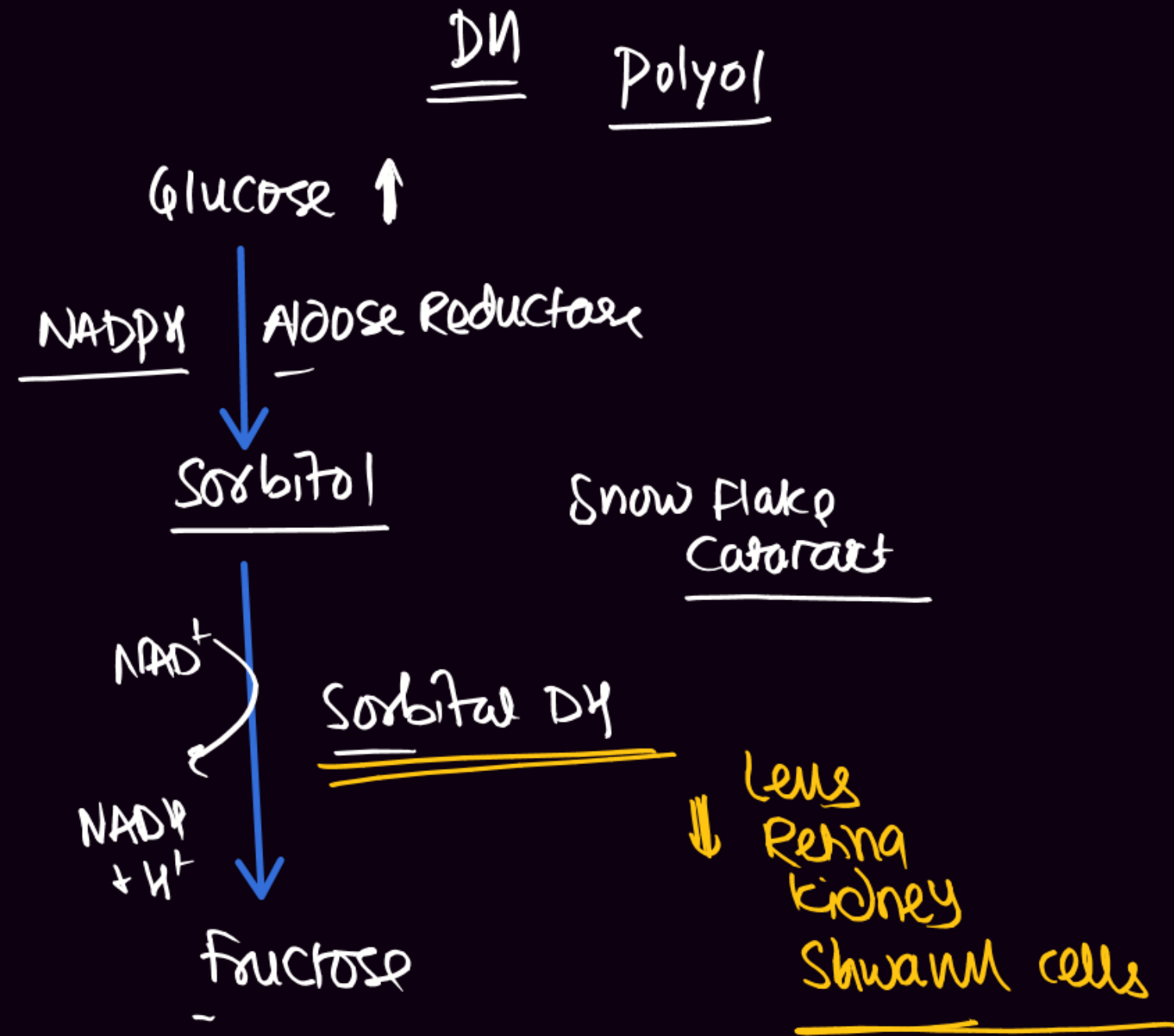


fruits
sucrose
sweets
cereals
drinks

Weaning

Hereditary fructose intolerance

- vomiting
- hypoglycemia
- Jaundice





Revision

Insulin



Glucagon



Epinephrine



GH

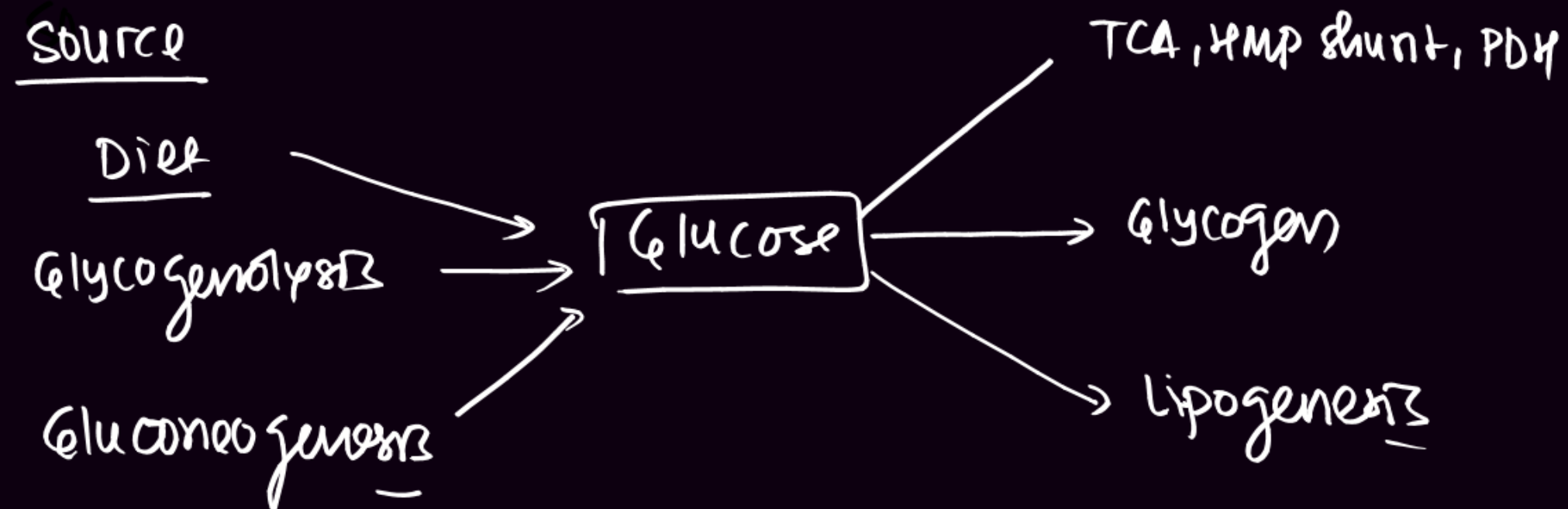


ACTH



Thyroxine





F ⇒ 70-110 mg/dl

PM ⇒ 110-140 mg/dl

Random ⇒ 70-140 mg/dl

Insulin ↑ uptake of Glu
⊕ Glycolysis
⊕ PDH
⊕ Glycogenesis

↓ Glucose

⊖
Blood Glucose

⊖ Gluconeogenesis
⊖ Glycogenesis

⊕ Glycogenolysis
⊕ Gluconeogenesis
Glucagon
Epi
GH
ACTH
Thyroxine
↑ BGL

Regulation

Glycogen metabolism
- Glycolysis
- Gluconeogenesis

Glucagon
Epi

G-protein



Adenyl cyclase



↑ CAMP



PK



Phosphorylase kinase



Glycogen Phosphorylase

A month-old baby had a loose stool after every breast feed. He was crying continuously and irritable. His pediatrician put him on Lactose free commercial preparation and a miracle happened; now baby is normal.

Probable Diagnosis? Lactose intolerance

Deficient enzyme? Lactase

Fermentation of
Lactose
by intestinal bact

Flatulence
Gases
CO₂
H₂
Methane

A 30-year-old male patient had a loose stool after consumption of Sucrose rich diet. He had abdominal cramps and flatulence. Every time he takes such type of diet, he gets similar symptoms.

Probable diagnosis? Sucrase deficiency

Deficient enzyme? Sucrase

Treatment? Avoid sucrose rich food

Clinical case

A 5-year-old child is brought to the clinic with a history of hypoglycemic episodes, developmental delay, and hepatomegaly. The child has also been experiencing poor growth and a tendency to have lactic acidosis. Laboratory tests reveal elevated serum levels of lactate, uric acid, and triglycerides. **What is the most likely diagnosis?**

- A) Pompe disease
- B) McArdle disease
- C) Von Gierke disease ✓
- D) Cori disease

Clinical case

A 6-month-old infant is brought to the clinic with a history of poor feeding, muscle weakness, and difficulty in breathing. The infant has hepatomegaly and hypotonia. Blood tests show elevated creatine kinase levels. On examination, the infant also has cardiomegaly, and an echocardiogram reveals left ventricular hypertrophy.

What is the most likely diagnosis?

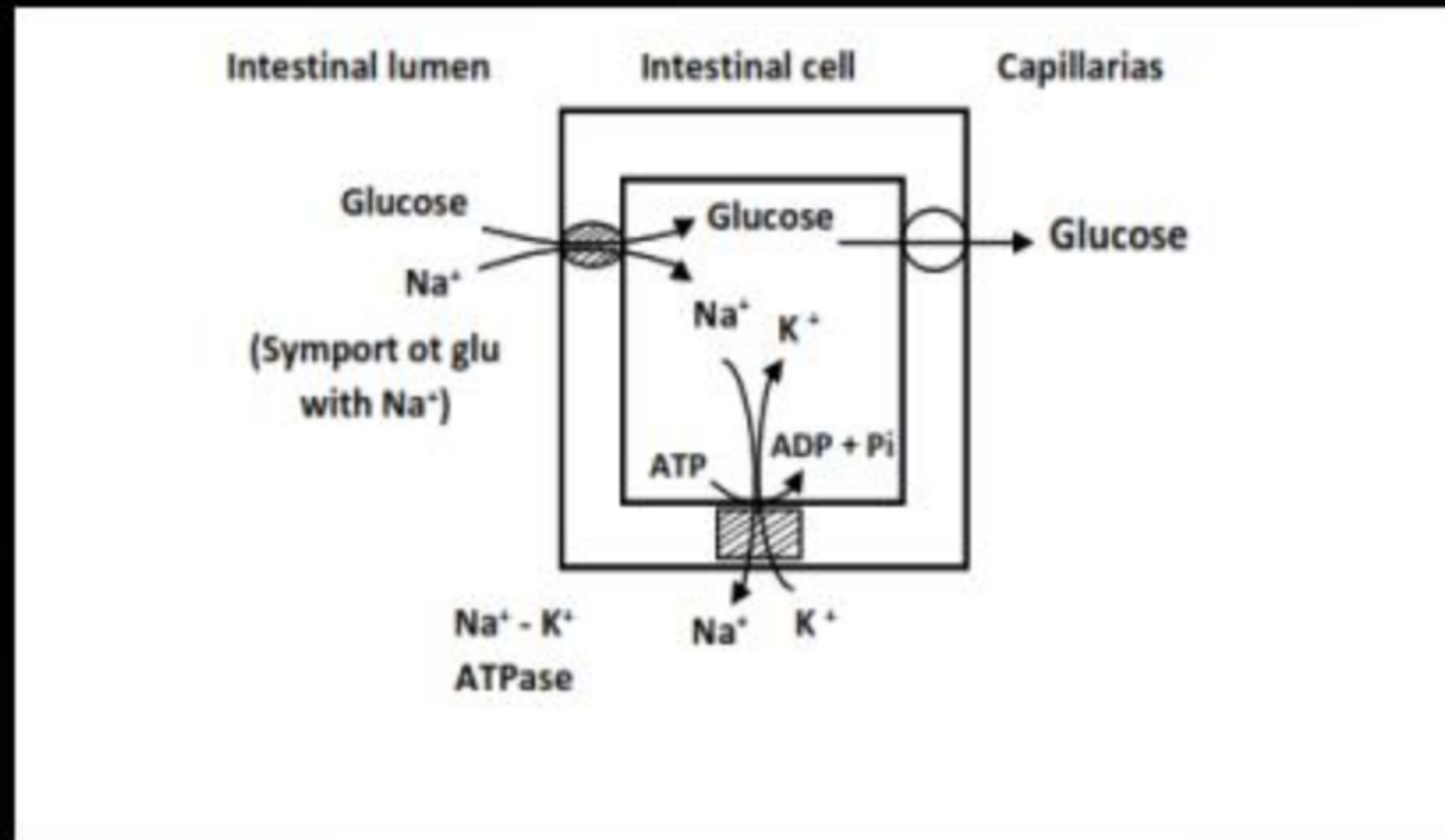
- A) Pompe disease ✓
- B) McArdle disease
- C) Von Gierke disease
- D) Cori disease

Clinical case

A 12-year-old boy is referred to the clinic due to exercise intolerance. He experiences muscle cramps and severe fatigue shortly after starting physical activities such as running or climbing stairs. However, he feels normal after resting for a few minutes. There is no history of hypoglycemia or liver enlargement. His serum creatine kinase (CK) level is significantly elevated after exercise, but his blood lactate levels are low after exercise.

What is the most likely diagnosis?

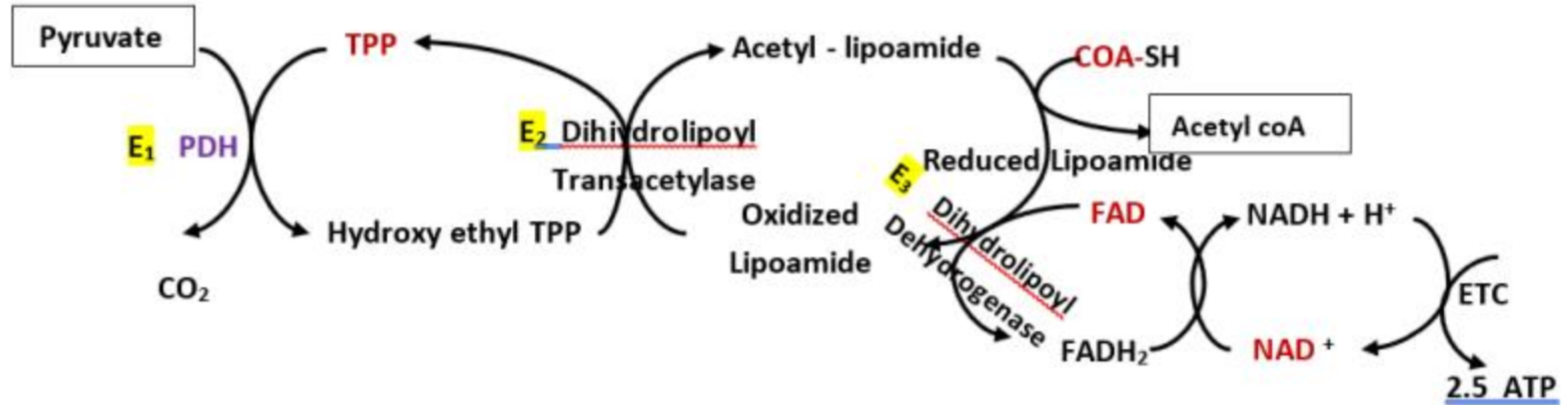
- A) Pompe disease
- B) McArdle disease ✓
- C) Von Gierke disease
- D) Cori disease



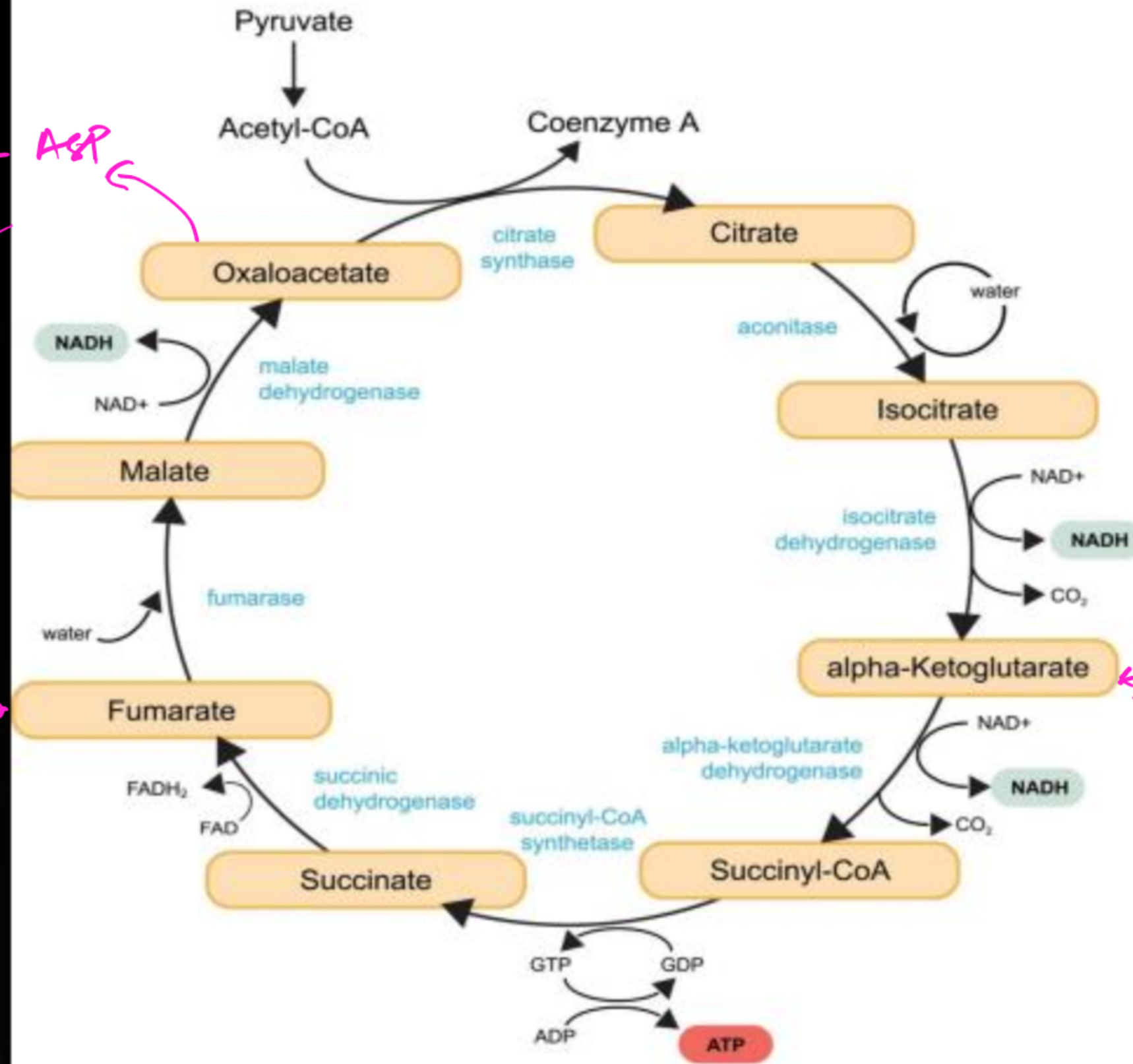
Transporter	Tissue location	Function
SGLT 1	Small intestine , Renal tubules	✓ Glucose and Galactose
SGLT 2	Renal tubules	Glucose

Transporter	Tissue location	Function	
GLUT 1	Brain, Kidney, Colon, Placenta, RBCs, Retina	Basal glucose uptake	Widely distributed, Abundant in RBCs, Major in Brain & placenta
GLUT 2	Liver, Pancreas, BL intestine, BL PCT	Regulation	
GLUT 3	Neuron, Placenta , Kidney		
GLUT 4 ✖	Skeletal muscle, Heart, Adipose tissue	Insulin dependent	Also GLUT 8,12
GLUT 5	Small intestine, Testis, sperm	Fructose transport	
GLUT 7	Liver ER	Glucose transport in ER	

PDH (3) En2
(5)



urea cycle
Asp



urea cycle

CO₂ → urea
ATP → urea
↓

NH₃ ↑
Glu
Glu

Glycogen Phosphorylase, coenzyme is

a. Pyridoxal Phosphate ✓

b. Thiamin

c. Biotin

d. Pantothenic acid

In which of the following tissues, is glycogen incapable of contributing directly to blood glucose:

a. Liver

b. Muscle

c. Both

d. None

Pompe's disease is due to deficiency of:

- a. Debranching enzyme
- b. Muscle Phosphorylase
- c. Acid Maltase ✓
- d. Branching enzyme

Type	Name of Disease	Deficient Enzyme	Clinical Features
Type 0		Glycogen synthase	No hepatomegaly, hypoglycemia in between meal
Type I	von Gierke's disease	Glucose 6 phosphatase	Fasting hypoglycemia, hyperlipidemia, hyperuricemia, lactic acidosis and Hepatomegaly
Type II	Pompe's disease	Lysosomal acid maltase	Glycogen accumulation in liver, heart and muscle, death in early age due to heart failure

Type	Name of Disease	Deficient Enzyme	Clinical Features
Type III	Cori's disease (limited dextrinosis or Forbe's Disease)	Debranching enzyme	Fasting hypoglycemia, hepatomegaly, myopathy. highly branched glycogen (Dextrin) accumulates
Type IV	Anderson's disease (Amylopectinosis)	Branching enzyme	Hepatosplenomegaly, cirrhosis of liver, death at early age
Type V	McArdle's disease	Muscle phosphorylase	Exercise intolerance, accumulation of glycogen in muscle
Type VI	Her's disease	Liver Phosphorylase	Hepatomegaly, fasting hypoglycemia

A baby refuses breast milk since the second day of birth, vomits on force-feeding but accepts glucose-water, develops diarrhea on third day, by fifth day she is jaundiced with liver enlargement and eyes show cataract. Urinary reducing sugar was positive but blood glucose estimated by glucose oxidation method was found low. The most likely cause is deficiency of:

- a. Galactose 1-phosphate uridyl transferase ✓
- b. Beta galactosidase
- c. Glucose 6-phosphate
- d. Galactokinase

A patient presents with symptoms of jaundice, vomiting, and diarrhea. Laboratory tests reveal an accumulation of galactose-1-phosphate in the blood. Which metabolic disorder does this patient most likely have?

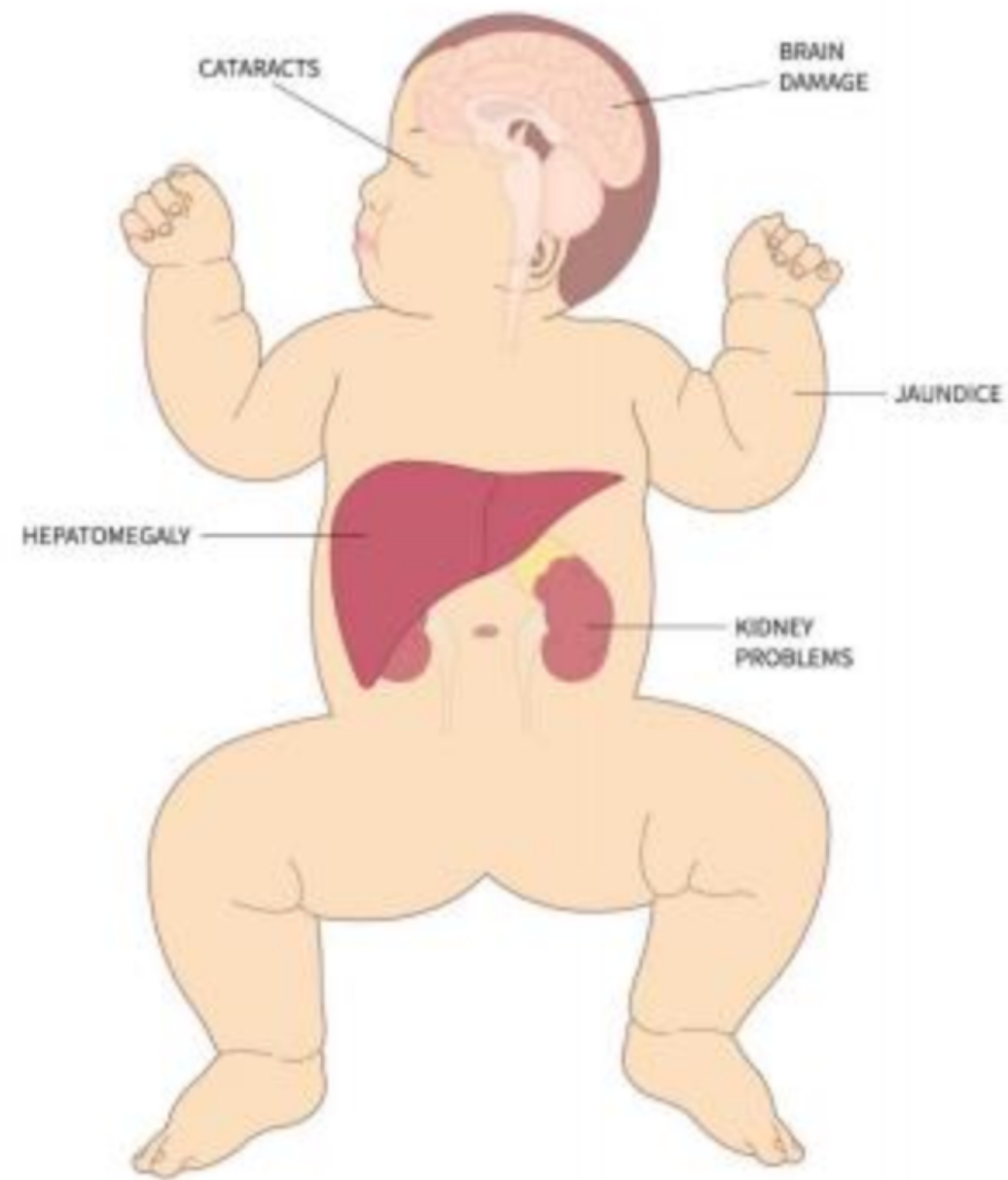
Hereditary fructose intolerance

Galactosemia ✓

Fructosemia

Glycogen storage disease type I

GALACTOSEMIA



Q. A patient from a Mediterranean country visits Africa, where he develops, malaria. He is treated with primaquine and later develops hemolytic anemia. Deficiency of an enzyme involved in which of the following pathways could be the cause?

Glycolysis

Gluconeogenesis

Hexose monophosphate (HMP) pathway ✓

Luebering-Rapoport pathway

A baby boy 10-month-old comes with vomiting, jaundice, hepatomegaly and signs of irritability when started weaning with fruit juice. Which of the following enzymes is defective?

- a. Aldolase B ✓
- b. Fructokinase
- c. Glucose 6 phosphatase
- d. Galactose 1 Phosphate Uridyl transferase

Q. A 17 years old boy diagnosed with diabetes brought to the hospital in semiconscious state and abnormal deep, rapid breathing. His plasma glucose was 673 mg/dl.

Urine shows positive Benedict's and Rothera's test.

Blood investigations reveals following results: Blood pH = 7.21, pCO₂ = 30 mmHg, bicarbonate = 16 mmol/L.

Name the acid base disorder.

22-28

↓ Metabolic

↓ 7.35 - 7.45

Acidosis

35-45

Metabolic acidosis ✓

Respiratory acidosis with compensation

Respiratory alkalosis

Metabolic alkalosis



	Type 1 DM(IDDM)	Type 2 DM(NIDDM)
Age	Childhood (up to 20 years)	> 30 years age group
Prevalence	10–20%	80–90%
Defect	Insulin deficiency due to pancreatic destruction	Decreased production or resistance to insulin
Weight	Normal or low	Obese
Genetic predisposition	Less	Very Strong
Autoantibodies	Commonly found	Rare
Complication	Ketoacidosis	Hyperosmolar state
Treatment	Only Insulin is Effective	Oral hypoglycemic drugs and insulin

WHO criteria for the diagnosis of **Diabetes Mellitus (DM)** in a table format:

④
Average BSL
8-12 wks

Test	Normal	Impaired	Diabetes
Fasting Plasma Glucose (FPG)	< 100 mg/dL (5.6 mmol/L)	100 - 125 mg/dL (5.6 - 6.9 mmol/L)	✓ ≥ 126 mg/dL (7.0 mmol/L)
Oral Glucose Tolerance Test (OGTT) (2-hour plasma glucose after 75g glucose load)	< 140 mg/dL (7.8 mmol/L)	140 - 199 mg/dL (7.8 - 11.0 mmol/L)	✓ ≥ 200 mg/dL (11.1 mmol/L)
Random Plasma Glucose (RPG)	Typically < 200 mg/dL (11.1 mmol/L)	N/A	✓ ≥ 200 mg/dL (11.1 mmol/L) with symptoms of hyperglycemia
Hemoglobin A1c (HbA1c)	< 5.7%	5.7% - 6.4%	✓ ≥ 6.5%



Homework

Pathway

Diseases



How to enrol



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Thank
You