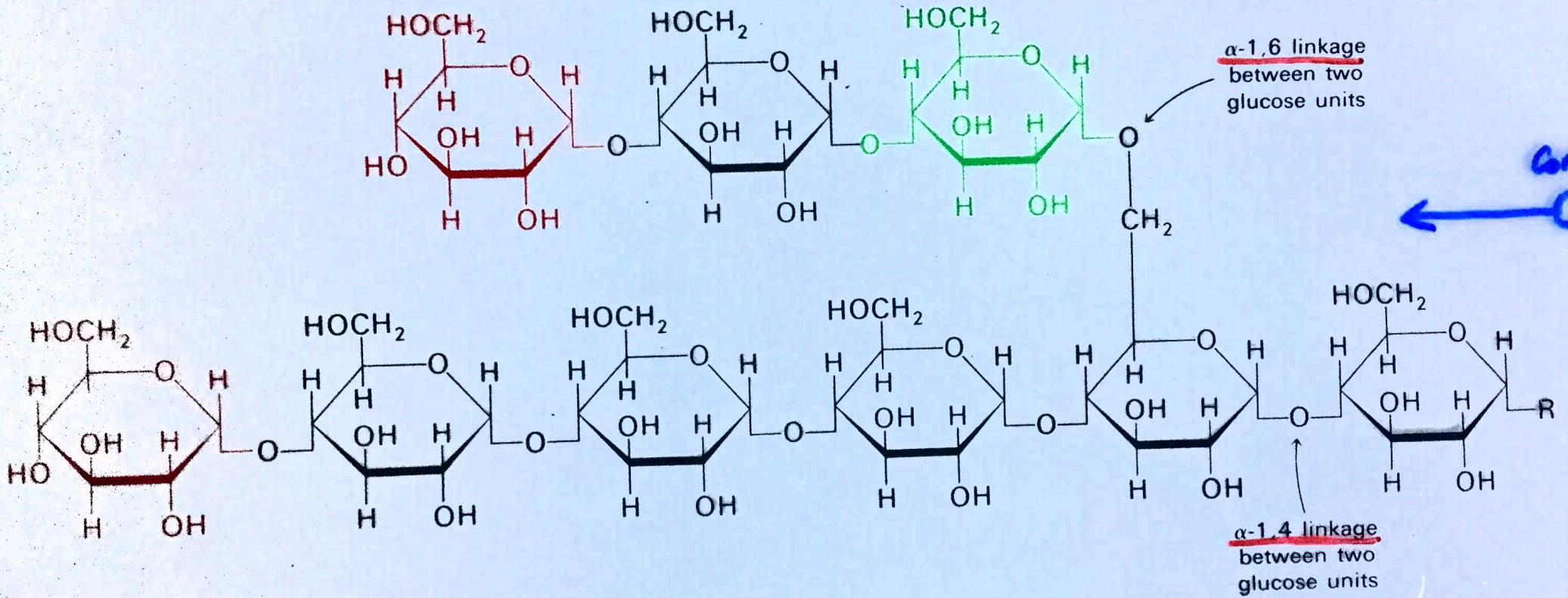
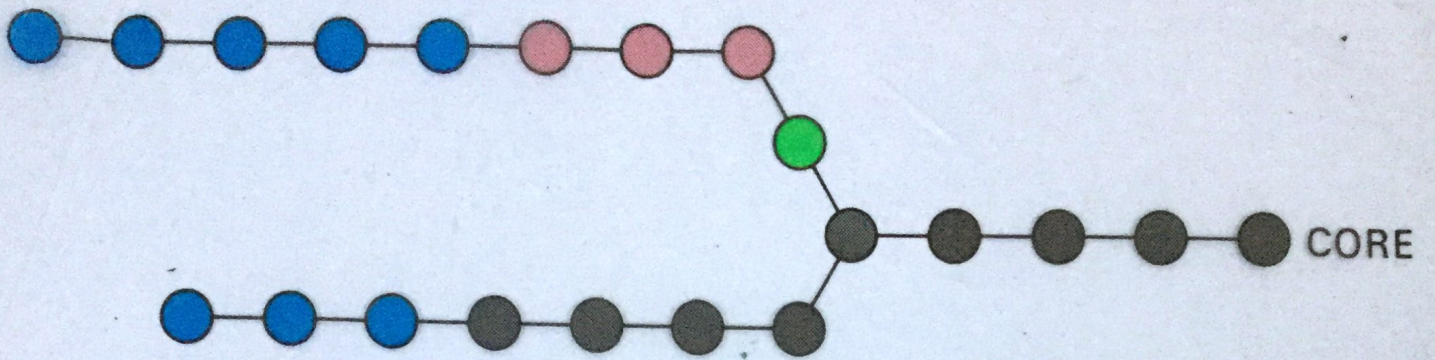


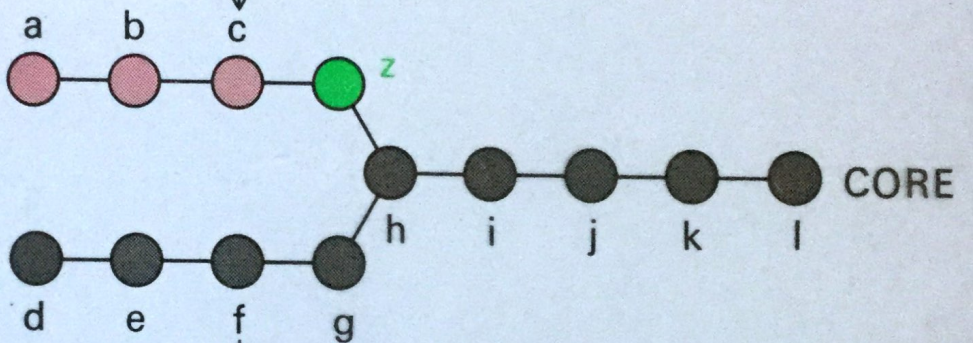
Glycogen structure



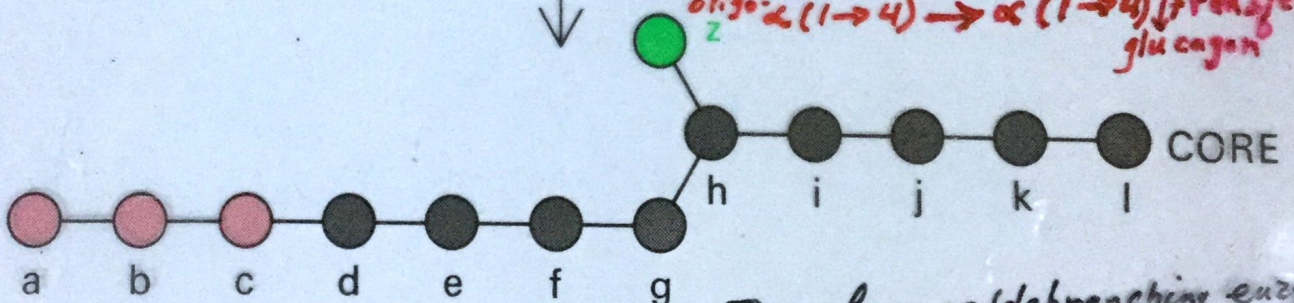
GLYCOGEN DEGRADATION



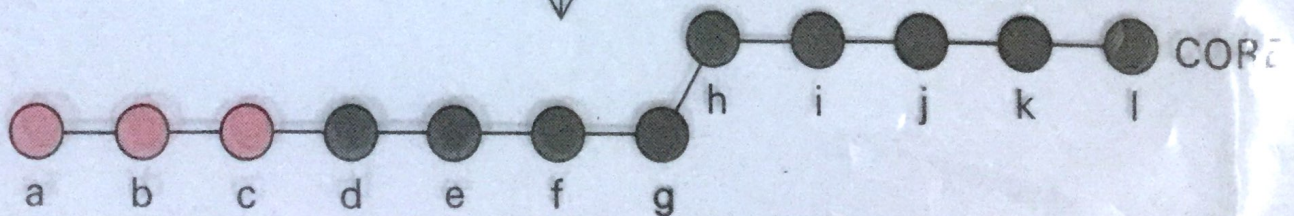
Phosphorylase
(Eight glucose 1-phosphate released)



Transferase [debranching enzyme]
 *$\alpha(1\rightarrow6) \rightarrow \alpha(1\rightarrow4)$ transferase
glucagon*

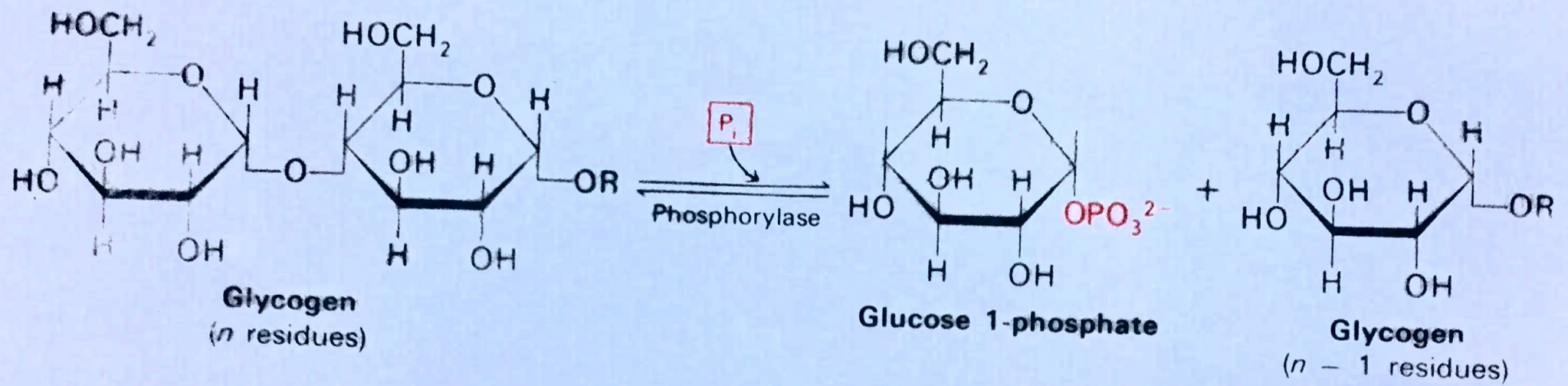


α -1,6-Glucosidase
(One glucose released)

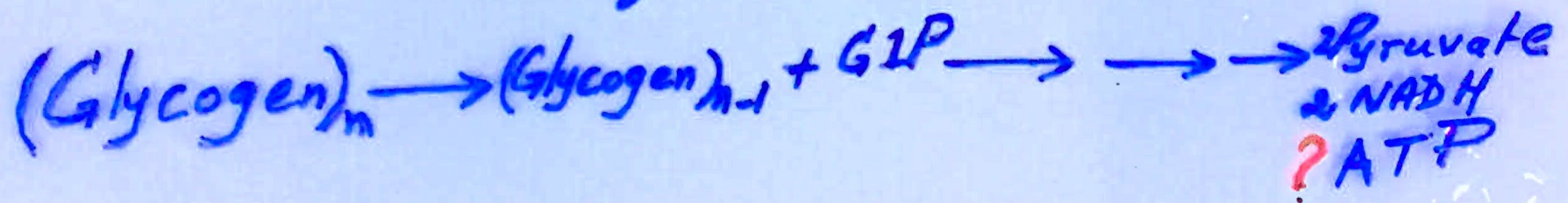


Glycogen Degradation :-

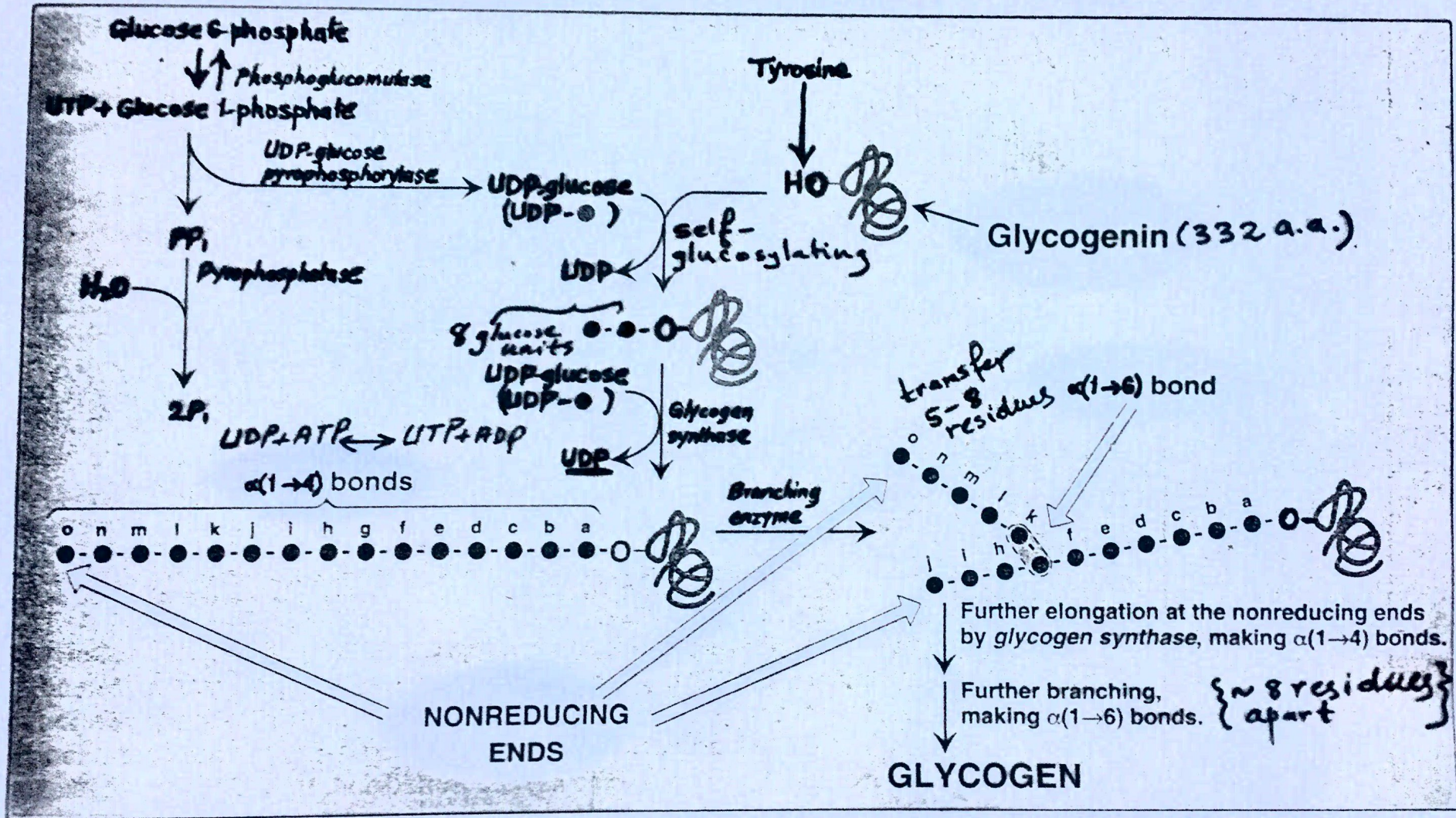
Glycogen phosphorylase



~ 8% of products are free Glu

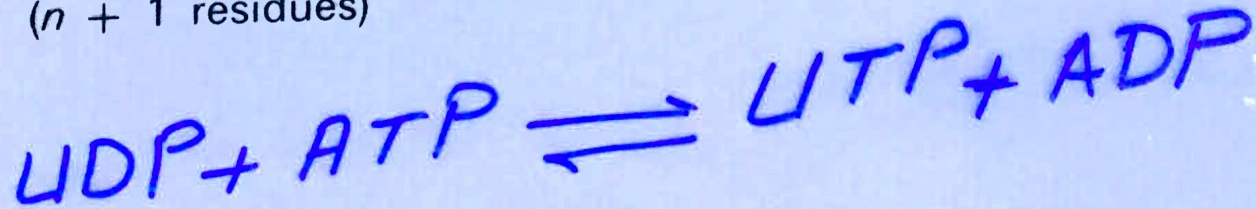
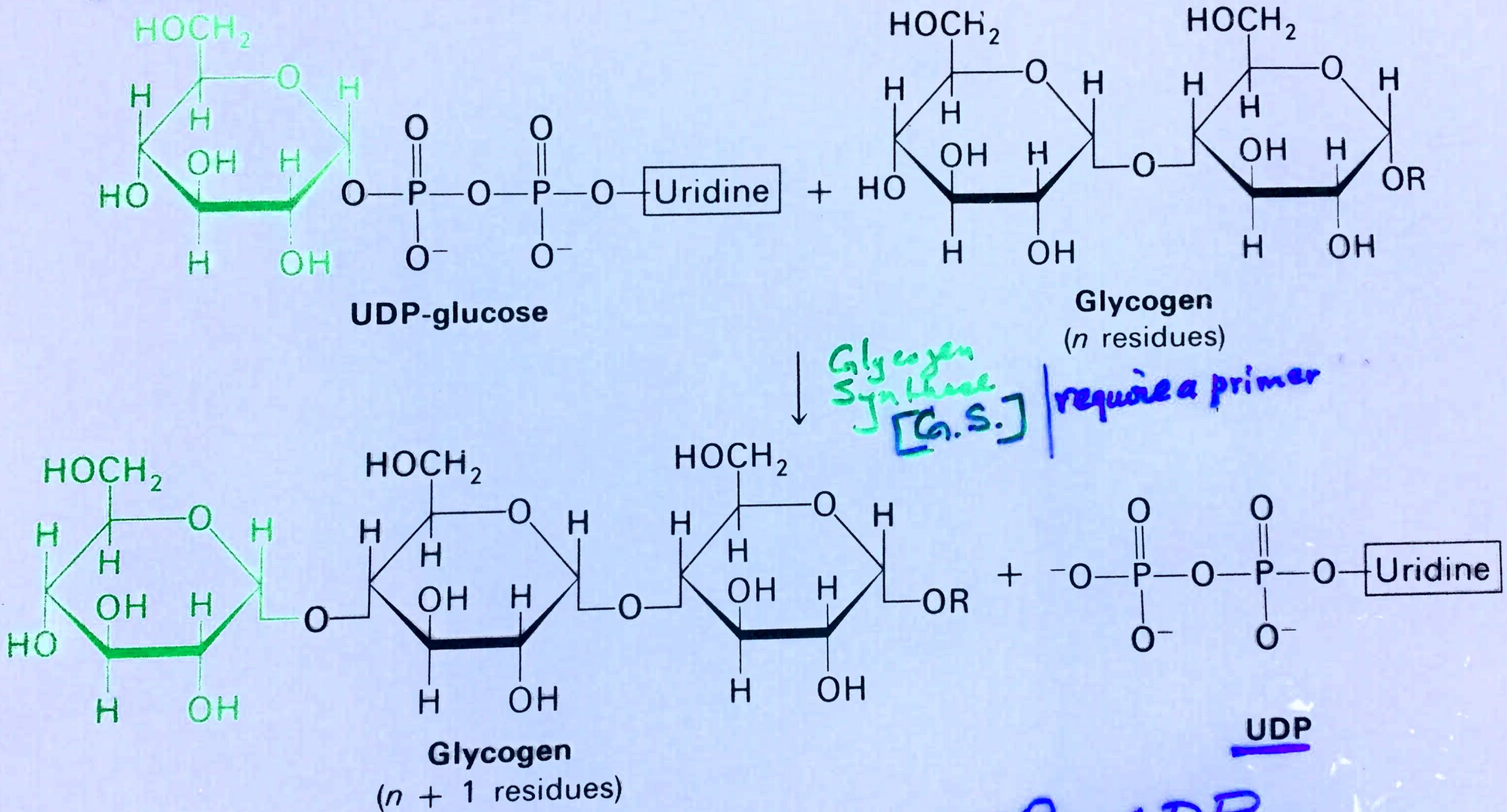


Glycogen Synthesis - branching



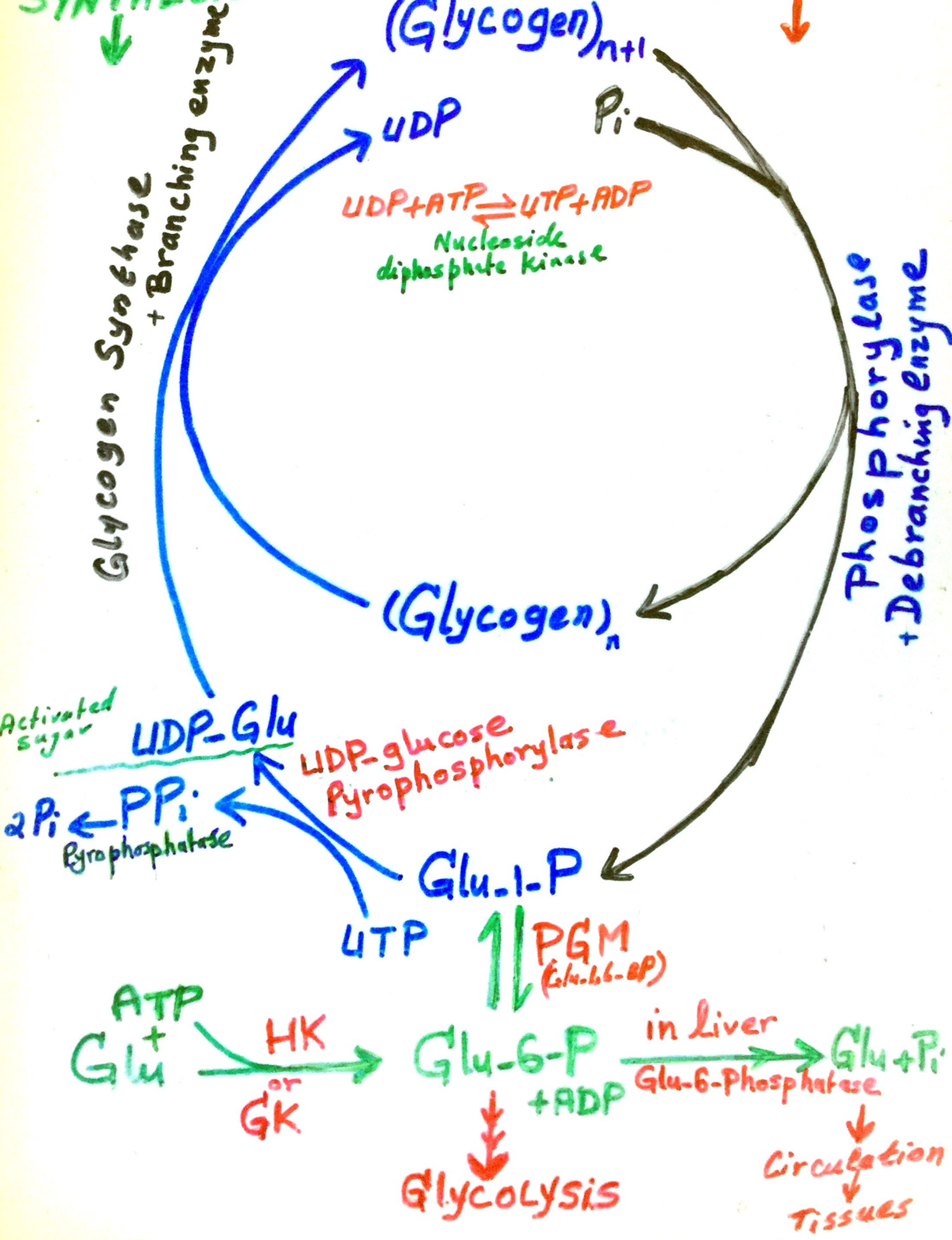
Branching :- far more soluble
 - more non-reducing ends for synthesis and degradation

Glycogen Synthase



GLYCOGEN SYNTHESIS

GLYCOGEN DEGRADATION



Glycogen Storage Diseases (GSDs) 9a

- Genetic
- defect in synthesis or degradation
- formation of glycogen with abnormal structure or excessive accumulation of normal glycogen
- Defective enzyme may be in a single tissue or more generalized
- severity of GSDs from fatal in infancy to mild disorders

0 Glycogen synthase

I Glucose-6-phosphatase (Von Gierke disease)

- Affects liver, kidney and intestine
- fasting severe hypoglycemia
- fatty liver - hepatomegaly
- progressive renal disease
- Growth retardation
- Hyperlactic acidemia, hyperuricemia
- normal glycogen structure, increased store

II Lysosomal $\alpha(1 \rightarrow 4)$ glucosidase: POMP Disease (Acid maltase)

1-3% of glycogen degraded by lysosomal glucosidase

Generalized - primarily liver, heart and muscle
Excessive glycogen conc. in abnormal vacuoles in lysosomes
Massive cardiomegaly
Normal blood sugar, normal glycogen structure
Early death from heart failure

III Debrancher

IV Brancher

V Muscle Glycogen phosphorylase McArdle syndrome

VI Liver glycogen phosphorylase

→ VII Phosphofructokinase

- as in type V plus hemolysis
- muscle and red blood cells involved

IX Liver phosphorylase kinase

X Liver cAMP-dependant Protein Kinase A

→ V **McArdle Syndrome** (Muscle Glycogen Phosphorylase)

- skeletal muscle affected, liver enzyme normal
- Weakness and cramping of muscle after exercise
- no rise in blood lactate → during strenuous exercise
- high level of glycogen with normal structure
- Myoglobinemia and myoglobinuria