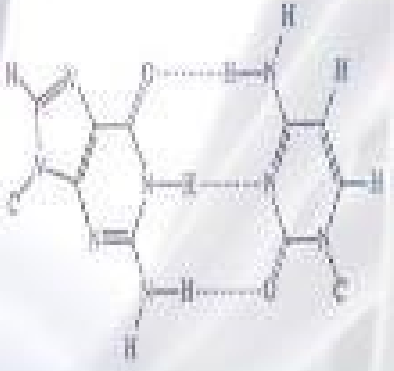
Medical Committee
The University of Jordan

SLIDE SHEET

SLIDE : 5- Glycogen Metabolism

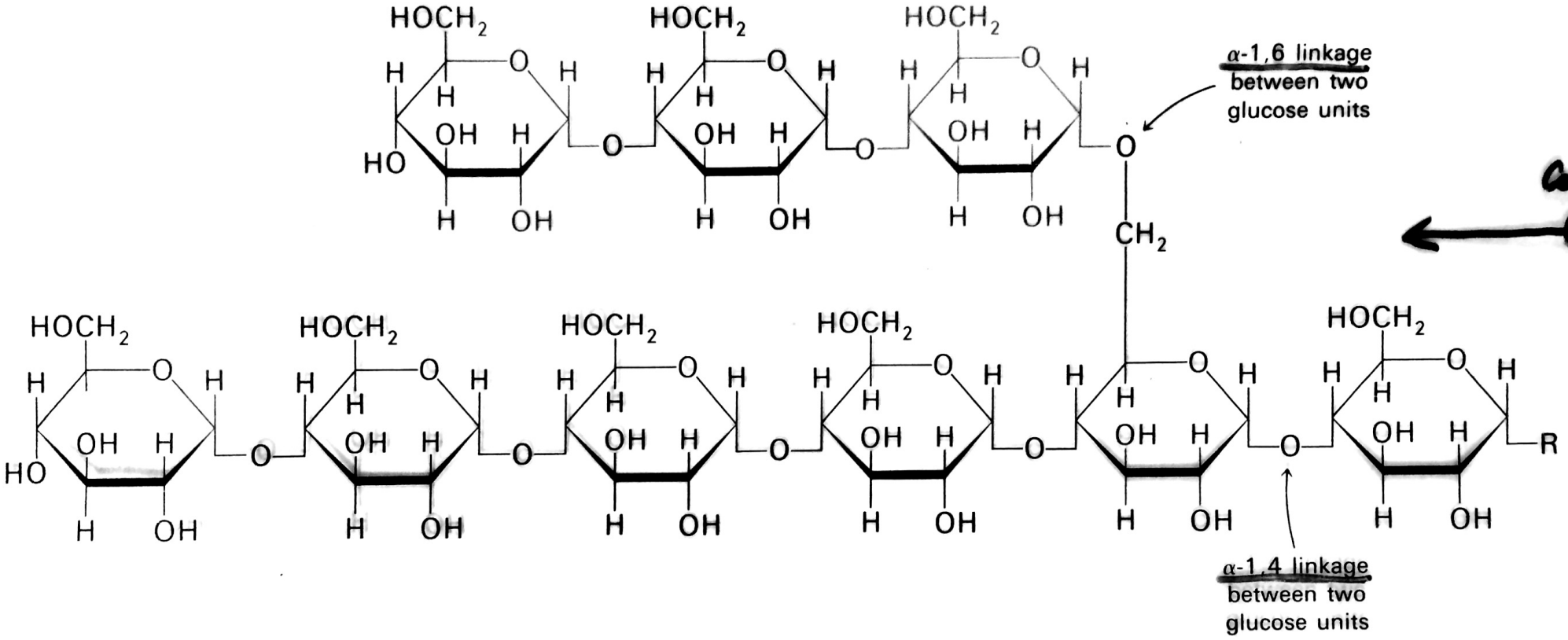
DR.NAME: Dr. Nayef

Biochemistry

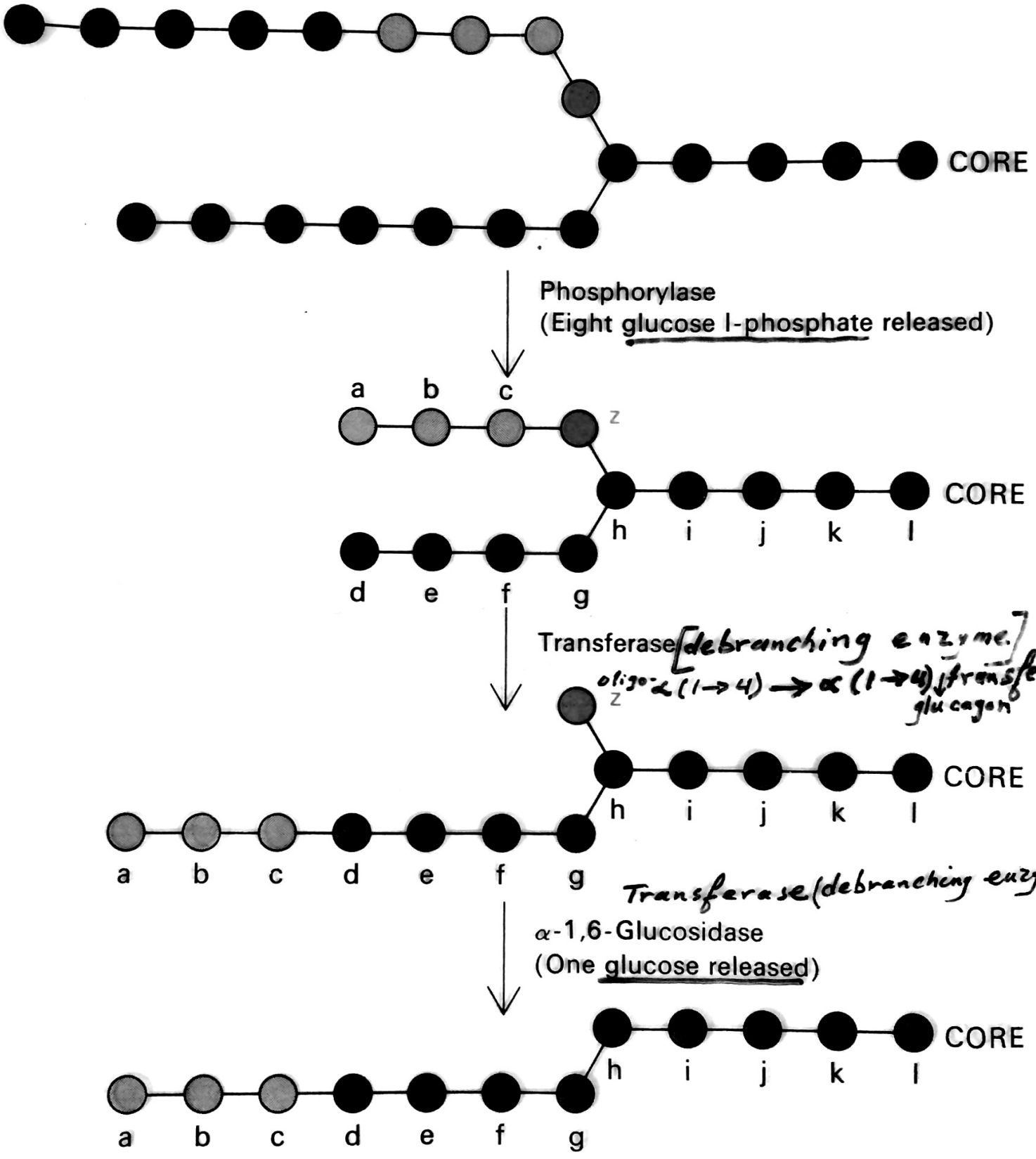


Majida Al-Foqaraa'

Glycogen structure

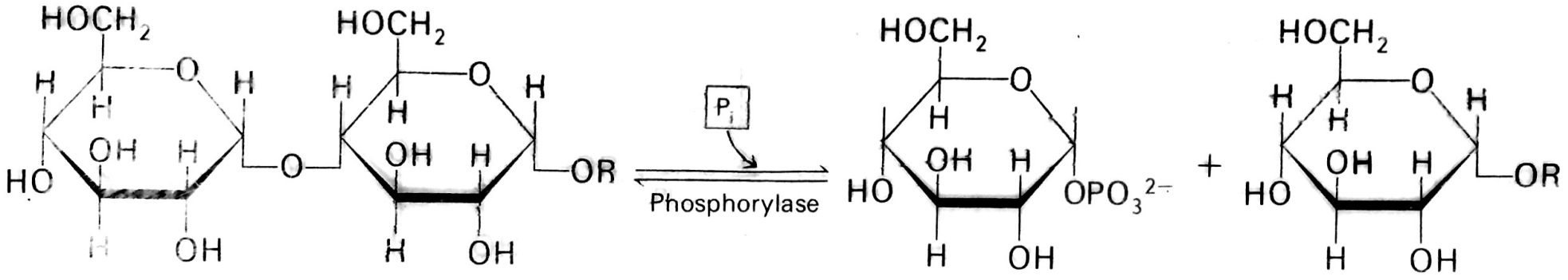


GLYCOGEN DEGRADATION



Glycogen Degradation :-

Glycogen phosphorylase



Glycogen
(*n* residues)

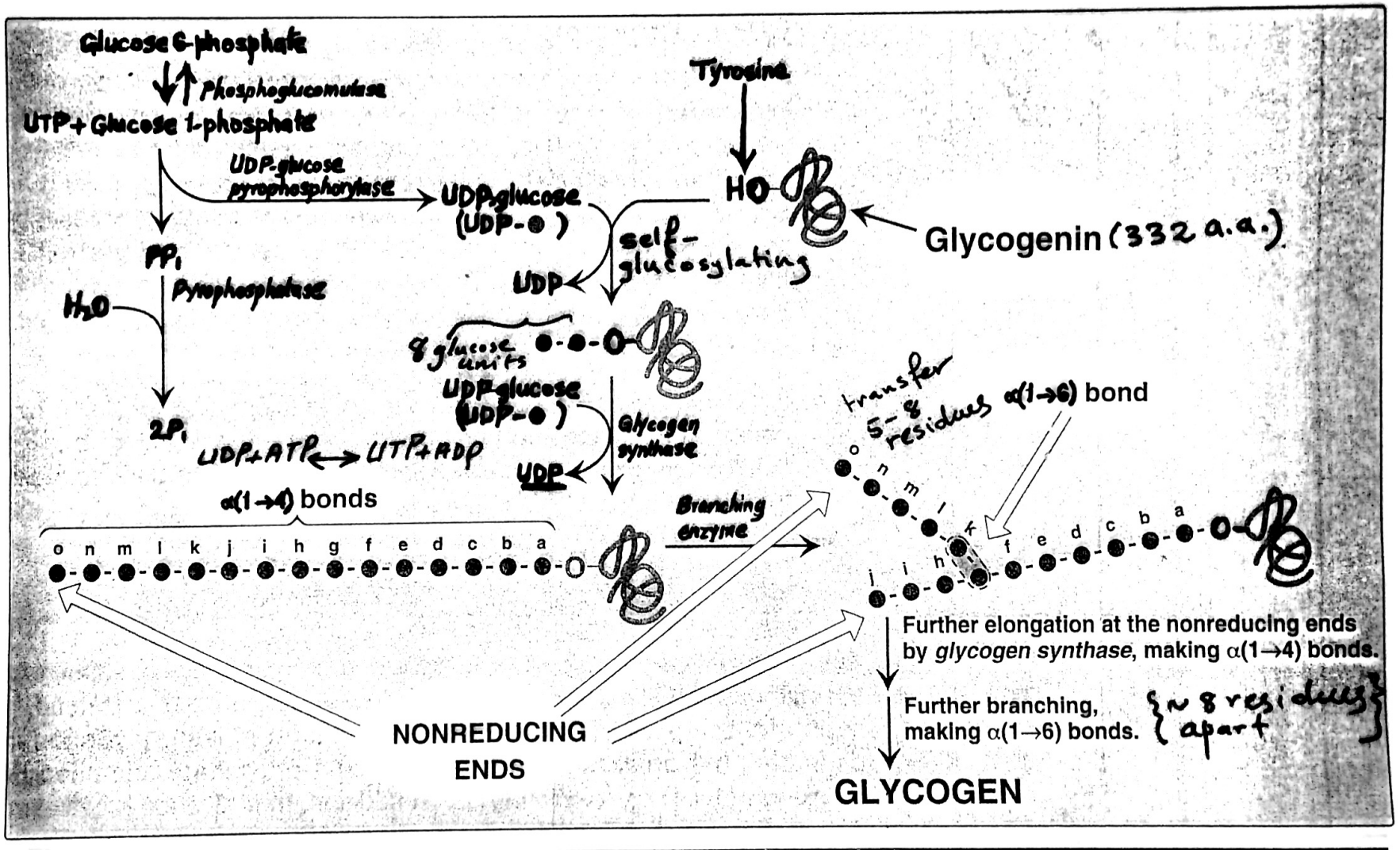
Glucose 1-phosphate

Glycogen
(*n* − 1 residues)

~ 8% of products are free Glu

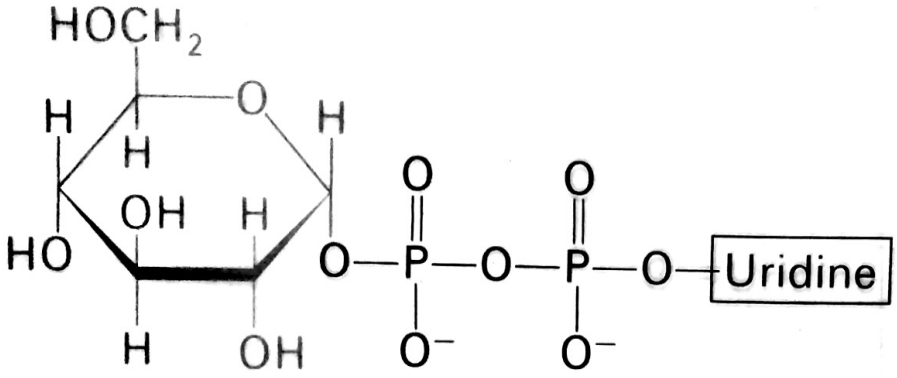
$(Glycogen)_n \longrightarrow (Glycogen)_{n-1} + G1P \longrightarrow \longrightarrow \longrightarrow$ 2 Pyruvate & NADH ? ATP

Glycogen Synthesis - branching



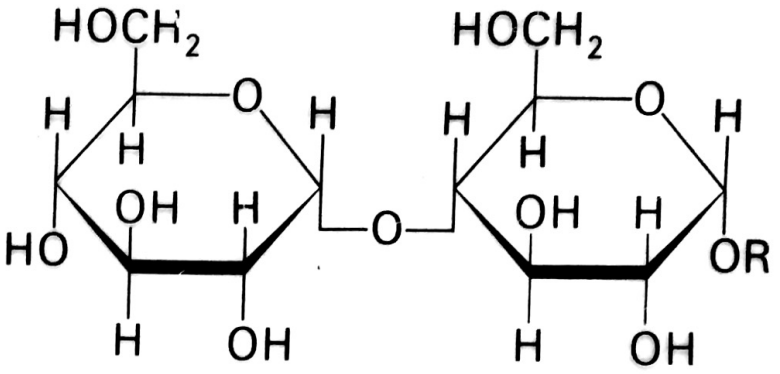
Branching :-
- far more soluble
- more non-reducing ends for synthesis and degradation

Glycogen Synthase



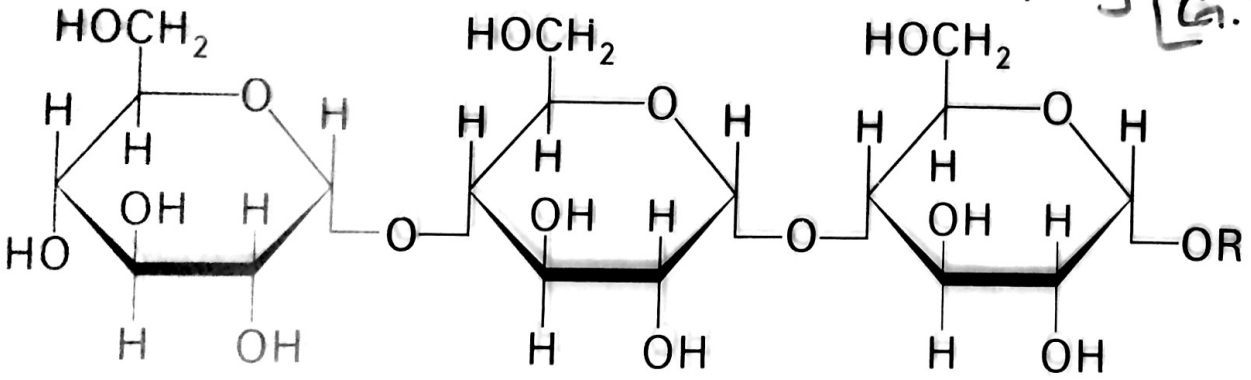
UDP-glucose

+



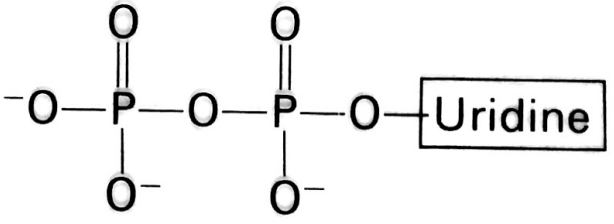
Glycogen
(*n* residues)

↓ Glycogen Synthase [G.S.] | require a primer

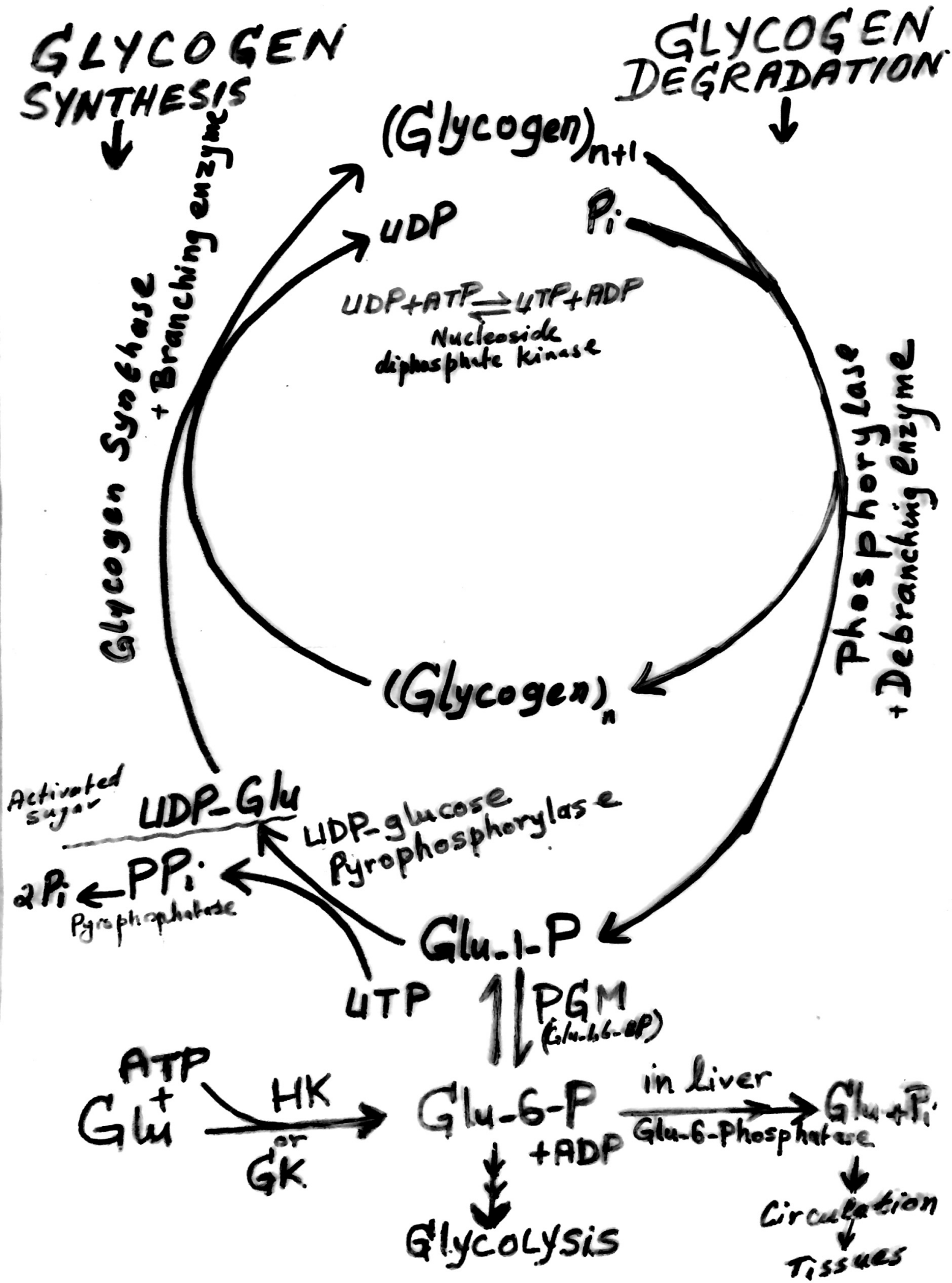


Glycogen
(*n* + 1 residues)

+



UDP


GLYCOGEN SYNTHESIS
GLYCOGEN DEGRADATION
(Glycogen)n+1
UDP
Pi
UDP+ATP ⇌ UTP+ADP
Nucleoside diphosphate kinase
Glycogen Synthase + Branching enzyme
Phosphorylase + Debranching enzyme
(Glycogen)n
Activated sugar
UDP-Glu
UDP-glucose Pyrophosphorylase
2Pi ← PPi
Pyrophosphatase
Glu-1-P
UTP
PGM (Glu-1,6-BP)
ATP
Glu
HK or GK
Glu-6-P
+ADP
in liver
Glu-6-Phosphatase
Glu + Pi
Glycolysis
Circulation
Tissues

Glycogen Storage Diseases (GSDs)

- 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 acedemia, hyperuricemia
- normal glycogen structure, increased store

II Lysosomal α(1→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

IX Liver phosphorylase kinase

X Liver cAMP-dependant Protein kinase A

→ V Mc Ardle Syndrome (Muscle Glycogen Phosphorylase)
- skeletal muscle affected; liver enzyme normal
- Weakness and cramping ↑ muscle after exercise
- no rise in blood lactate → during strenuous exercise
- high level ↑ glycogen with normal structure
- Myoglobinemia and myoglobinuria