

Structured Notes According to **BIOCHEMISTRY**

Revision friendly **Fully Colored Book/Structured Notes**

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(Author)

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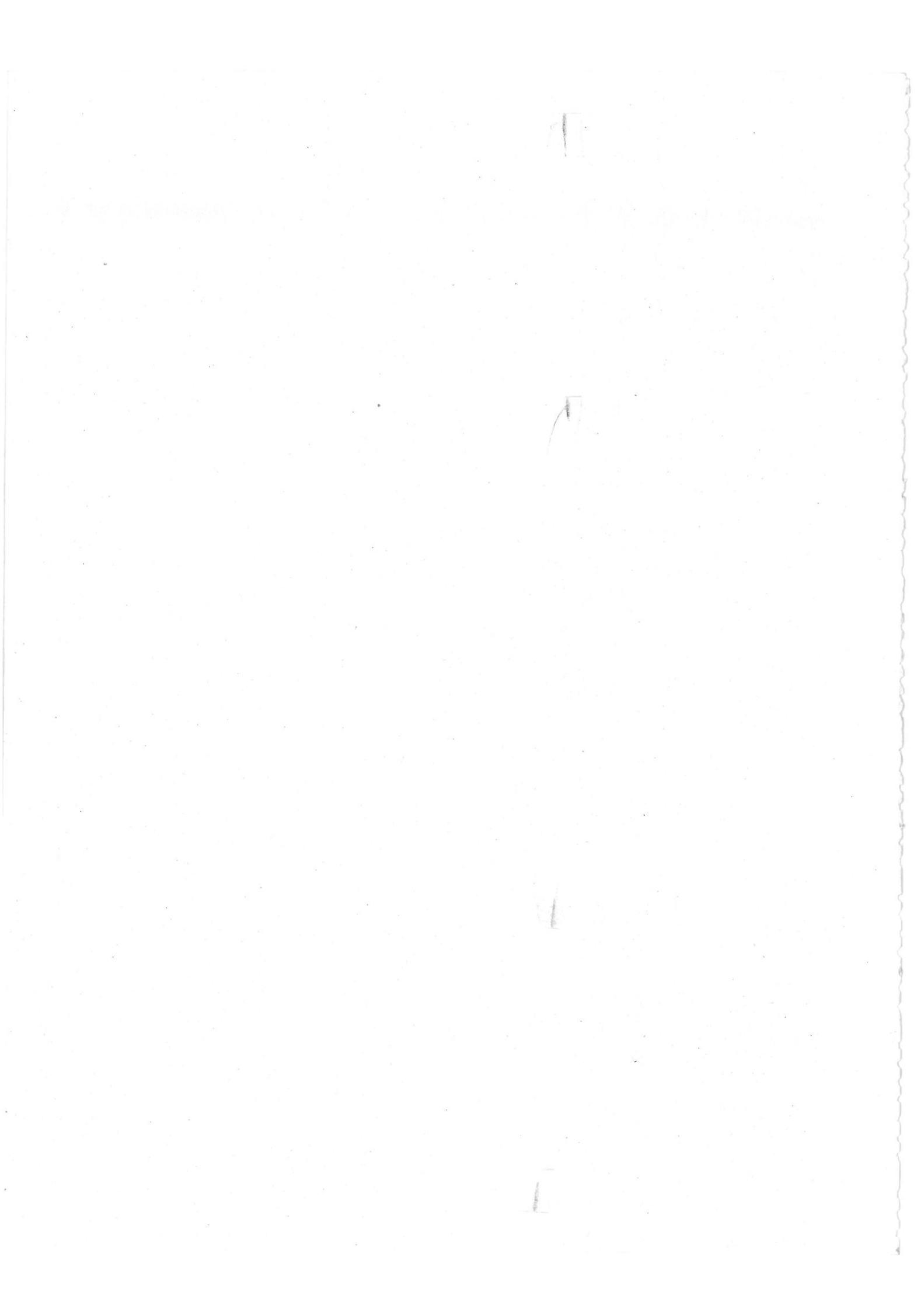
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INTRODUCTION

Cell and Integration of Metabolism

1. Marker Enzymes
2. Where are proteins Synthesized?
3. Protein Synthesis and Sorting
 - 3.1 MCQ's
4. Inclusion Cell Disease
5. Hyperoxaluria
 - 5.1 Primary Hyperoxaluria
 - 5.2 Secondary Hyperoxaluria
6. Introduction to Metabolism
 - 6.1 Catabolism
 - 6.2 Integration of Metabolism
 - 6.3 **Difference between NADPH Vs NADH**
 - 6.4 **Preferred Fuel**

Must Know

Good to Know

1 CELL AND INTEGRATION OF METABOLISM



Q. The marker enzyme of microsomes is?

- Galactosyl Transferase
- Cathepsin
- Lactate Dehydrogenase
- Glucose-6-Phosphatase**

Explanation:

- Microsome is endoplasmic reticulum; has Glucose-6-phosphatase and it performs a part of gluconeogenesis.
- **Galactosyl Transferase** is present in golgi complex; involved in glycoprotein synthesis.
- **Cathepsin** are proteases and enzyme markers of lysosomes.
- Lactate Dehydrogenase: Enzyme of Glycolysis and marker for cytoplasm.

PYQ: FMGE 2023

PYQ: FMGE 2020

Marker Enzymes

00:02:15

S no.	Organelle	Marker Enzyme
1	Nucleus	Replication-DNA polymerase Transcription- RNA polymerase
2	Endoplasmic Reticulum	Glucose-6-Phosphatase
3	Golgi Complex	Galactosyl transferase
4	<ul style="list-style-type: none"> • Mitochondria <ul style="list-style-type: none"> ○ Outer Membrane (OM) ○ Inner Membrane 	For OM - Monoamine oxidase For inner membrane - ATP synthase / Succinate dehydrogenase
5	Lysosome	Cathepsin
6	Cytoplasm	Lactate dehydrogenase
7	<ul style="list-style-type: none"> • Peroxisome: <ul style="list-style-type: none"> ○ Very long chain fatty acid oxidation, concerned with ether lipid synthesis (Plasmalogen). ○ Related to alpha oxidation of BCFA (defect leads to Refsum's Disease). 	Catalase

Q. Cytoplasmic proteins are synthesized in?

- Ribosomes**
- Nucleus
- Smooth Endoplasmic Reticulum
- Rough Endoplasmic Reticulum

- Rough Endoplasmic Reticulum: Protein synthesis.
- Smooth Endoplasmic Reticulum: Steroid synthesis.
- All protein synthesis is initiated by a free ribosome.

Where are proteins Synthesized?

00:07:00

- Free ribosomes read mRNA in a 5' to 3' direction, decoding codons one by one.
- They recruit complementary tRNA molecules carrying amino acids, enabling mRNA's nucleotide sequence to be translated into an amino acid sequence, forming polypeptide chains in a process known as translation.
- When ribosomes reach the mRNA from 5' to 3', the polypeptide chain will grow from **Amino terminal** (First amino acid) end to **Carboxyl terminal end** (last Amino acid).
- **This synthesis is done for the mitochondrial, nuclear and cytoplasmic proteins.**
- For other types of protein: ribosomes are not sufficient.

Protein Synthesis and Sorting

00:10:20

- Free ribosomes read mRNA from 5' to 3'
- They recruit tRNA with complementary anticodons, which carry specific amino acids
- This process drives translation, building the polypeptide chain from the amino terminal end to the carboxyl terminal end.
- Lysosomal protein, membrane protein or secretory protein are specific targeting proteins and targets to respective organelles.
- For the proteins to target specific organelles, need a label, e.g. proteins with label, mannose-6-phosphate targets lysosomes.
- Mannose is a carbohydrate and only ribosomes are not sufficient to synthesize it, so ER and GC helps in attaching this signal/label.
- For a protein to reach the membrane, it needs a label - lipid side chain because the membrane is lipid bilayer.
- For lysosomal, membrane and secretory proteins: targeting sequences can be attached only by endoplasmic reticulum and golgi complex and forms rough endoplasmic reticulum (RER); but formation of RER is initiated by free ribosomes.
- Again, they read mRNA from 3' end to 5' end (amino to carboxyl end) but the amino terminal end of only these three types of proteins alone have a signal recognition peptide sequence.
- Function of signal sequence:
 - Guides the ribosomes to get attached to ER to form RER.
 - Ribosomes with mRNA attaches to ER → starts translation → forms polypeptide that grows into vesicles of ER → RER formed.
 - Once RER formed, the signal is not required and removed; pre-pro proteins are converted into pro- proteins.
- Along with translation, co- translational modifications occur in RER in which proteins will be folded (1st folding in ER) → Golgi complex, pass through cis, medial and trans golgi.
- During transit through trans GC post translational modifications occur, in which proteins get packed and sorted out depending on the label present, so protein:
 - With mannose 6 residue - reaches lysosome.
 - With a lipid side chain - reaches membrane protein.
 - No lipid or Mannose- secreted as Secretory protein.
- After post translational modifications, the protein gets packed and sorted out, which depends on the label present, so protein:
 - With mannose 6 residue - reaches lysosome
 - With a lipid side chain - reaches membrane protein
 - No lipid or Mannose- secreted as Secretory protein

MCQ's

00:21:20

Q. Secretory proteins are synthesized in:

- Ribosomes
- Smooth Endoplasmic Reticulum
- Rough Endoplasmic Reticulum

d. First in Ribosomes, and then in Endoplasmic Reticulum

Q. A child is present with coarse features, HSM and mental retardation. Clinician suspected a LSD and ask the IEM lab to perform a few Lysosomal enzyme activities including Hexosaminidase. All Lysosomal enzyme activities were high. What is probably the diagnosis?

- Tay Sachs Disease
- Mucopolysaccharidosis I
- Mucopolysaccharidosis II

d. I Cell Disease

Explanation:

I Cell Disease/ Inclusion Cell Disease:

00:21:20

- Lysosomal storage Disorder caused by the defect of an enzyme in Acetylglucosamine Phosphotransferase → defective labeling of lysosomal enzymes (AKA Phosphotransferase effect).
- Function of Phosphotransferase: Form Mannose-6-Phosphate - label for a protein to be targeted to lysosome.
- Due to this defect, lysosomes are empty and are in pinocytosis (a part of cell forms an indentation, as it deepens it gets pinched out of the cell membranous endocytic vesicle, nonspecifically taking interstitial fluid into the endocytic vesicle).
- Normally, the content to be metabolised by primary lysosomes fuses with endocytic vesicles to form secondary lysosome which has the enzymes to digest the content.
- But in this child, due to defect phosphotransferase, lysosomes are not targeted properly and are empty, presents with:
 - Multiple inclusions in connective tissues → coarse facial features; in parenchymal cells → Hepatosplenomegaly.
 - Accumulation in neurons → Mental retardation.
- Since lysosomal proteins didn't reach the lysosomes → ↓ tissue lysosomal activity; they secreted into circulation as Secretory protein → ↑ plasma lysosomal activity.
- Triad of I cell disease = Clinical features resembling lysosomal storage disorder with a paradox (↑ plasma lysosomal activity + ↓ tissue lysosomal activity).

Q. A 39-year-old man came to the emergency department because of severe back pain. An ultrasound was taken and renal stones were found. Following lithotripsy, a sample of the stone was sent for biochemical analysis. Results revealed the presence of oxalate and glyoxylate. He was diagnosed with primary hyperoxaluria Type I. It is caused by?

- Protein Folding defect
- Silent Mutation
- Protein Targeting Defect**
- Acceptable Mutation

Explanation:

Hyperoxaluria

00:29:30

Primary Hyperoxaluria

- Two types: Type I & Type II; both are related to Glycine metabolism.
- **Metabolism of Glycine:**
 - Glycine reacts with a ketoacid - pyruvate (Most enzymes in the body react with keto acid).
 - Glycine gives off its amino group to Pyruvate → Alanine & Glycine → Glyoxylate (ketoacid).
 - Glyoxylate again reacts and forms Glycine.; if reverse reaction does not occur, the excess Glyoxylate will be converted to Glycolate by Glyoxylate reductase, if this reaction doesn't occur, excess Glyoxylate → Oxalic acid.
 - Site: Peroxisome; Reaction: Irreversible.
 - Enzyme: Glyoxylate alanine aminotransferase (Glycine + Pyruvate → Glyoxylate + Alanine).

Type I Primary Hyperoxaluria

- Protein targeting defect.
- Glyoxylate Alanine aminotransferase is present in mitochondria instead of peroxisome.
- Reaction is irreversible in mitochondria so, Glycine → excess Glyoxylate → Oxalic acid.

Type 2 Primary Hyperoxaluria

- Cause: Defective Glyoxalate reductase.
- Due to defective enzyme, Glyoxylate is not converted to Glycolate and excess Glyoxylate → Oxalic acid.

Secondary Hyperoxaluria

- Most common because of dietary effects: Oxalate rich food like large quantities of chocolates, beetroots, green leafy vegetables.

Enteric Hyperoxaluria

- Usually 5-10% of dietary oxalate will get absorbed and remaining will form stones with calcium → Calcium oxalate (insoluble) → excreted in feces.
- In fat malabsorption: Accumulation of fatty acids in the intestinal lumen → form complexes with calcium → excreted in feces.
- If calcium not available, free oxalates accumulate in the lumen → absorbed paracellularly → Secondary hyperoxaluria.

Introduction to Metabolism

00:35:50

- Metabolism is the process of assimilation of food we intake.
- Two parts of metabolism: Catabolism & Anabolism.

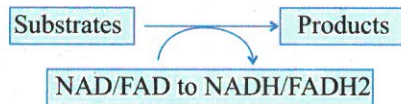
Catabolism

- Process of breaking complex molecules into simpler substances by breaking covalent linkages and energy is liberated.
- Depending upon the trapping of energy liberated in the form of ATP, it of 2 types:

1. Substrate level Phosphorylation

- Substrates are converted to products, where ADP is converted to ATP
- e.g.
 - Phosphoglycerate kinase (step of glycolysis)
 - Pyruvate kinase (step of glycolysis)
 - Succinate tyrosinase (step of citric acid cycle)
 - Creatine kinase (of muscle)

2. Oxidative Phosphorylation

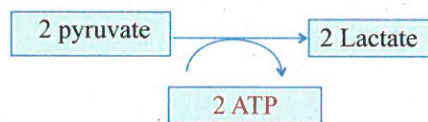


- After this, oxidation of NADH / FADH₂ → release electrons → transported to electron transport chain → liberates energy and is used for Phosphorylation of ADP to ATP.
- Oxidative phosphorylation is more common in human beings because of aerobic metabolism.

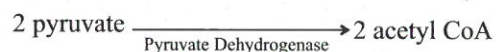
Integration of Metabolism

00:42:10

- Most of the fuels in the body: Carbohydrates (Glucose).
- Most of the glucose enter into glycolysis, one molecule of glucose will split into 2 pyruvates and 7 ATPs generated, this step is known as Aerobic Glycolysis.
- In the absence of oxygen,



- In the presence of oxygen, after the formation of Pyruvate the process continues because it's accumulation can cause metabolic acidosis.
- All fuels get finally metabolized through the citric acid cycle.
- To enter the Citric Acid cycle,



- Pyruvate dehydrogenase removes hydrogen from pyruvate, gives to coenzyme NAD → NADH (1 NADH = 2.5 ATP) and for 2 pyruvates, 5 ATP's generated.
- 2 Acetyl CoA enters Citric acid cycle and finally comes as CO₂ which can be exhaled out - Complete oxidation of glucose.
- 1 Citric acid cycle → 10 ATP's; 2 Acetyl CoA → 20 ATP's.
- Complete oxidation of glucose gives 32 ATP's.
- In High energy also, glucose enters the cell, forms 2 pyruvate → 2 acetyl CoA.
- In the presence of adequate pyruvate and acetyl CoA, feedback inhibition of glycolysis occurs and most of glucose-6-phosphate enters into glycogen synthesis while some enters into HMP shunt/ Pentose phosphate pathway.
- HMP shunt significance:
 - Acts as source of NADPH & Ribose-5-phosphate (helps in synthesis of nucleotides)

Difference between NADPH Vs NADH

00:50:30

PYQ: INICET 2021

NADH	NADPH
Can enter into ETC and give rise to 2.5 ATPs. as a part of Oxidative phosphorylation.	Necessary: <ul style="list-style-type: none"> • For reductive biosynthesis of all lipids. • For regenerating glutathione (important antioxidant mechanism of RBCs) • As a coenzyme for ribonucleotide reductase • As a coenzyme for cytochrome P450 enzymes.
Sources- all fuel oxidative pathways. <ul style="list-style-type: none"> • Glycolysis • PDH • Citric Acid Cycle • Amino acid oxidation 	Sources- <ul style="list-style-type: none"> • Major source : HMP Shunt • Minor sources: Cytoplasmic isocitrate dehydrogenase (Cyt ICDH) • Malic enzyme

- RBCs act as a major source of oxidative stress in the body because they carry oxygen; while carrying it can donate an electron converting oxygen \rightarrow Superoxide radical \rightarrow hydrogen peroxide \rightarrow peroxy radical \rightarrow lipid peroxidation.
- When H_2O_2 is generated in RBC, 2 molecules of glutathione donate one hydrogen each so that $H_2O_2 \rightarrow 2H_2O$; this reaction is catalyzed by glutathione peroxidase.
- For regeneration of glutathione: NADPH (H_2 source) and glutathione reductase (enzyme) is necessary.

Defect of HMP Shunt

PYQ: FMGE 2022

- In HMP Shunt, there is an enzyme Glucose-6-Phosphate Dehydrogenase.
 - Glucose 6 Phosphatase and Glucose-6-Phosphate Dehydrogenase are different.
 - Glucose 6 Phosphatase is an enzyme of Gluconeogenesis which increases blood glucose.
 - Glucose-6-Phosphate Dehydrogenase is an enzyme of HMP Shunt which is a source of NADPH.
- Defect in HMP shunt (defective G6PD) \rightarrow no NADPH \rightarrow no regeneration of glutathione \rightarrow RBC lysis due to oxidative stress.
- e.g. Child presenting with hemolytic anemia after intake of Fava beans or Primaquine (prophylactic drug for malaria) due to defect in HMP shunt (G6PD).

Fate of Acetyl CoA

- In low energy, all Acetyl CoA enter into the Citric Acid Cycle.
- Acetyl CoA is a building block of fatty acid and cholesterol.
 - Excess fatty acid will be stored as Triacylglycerol.
 - Excess cholesterol will be stored as the cholesterol esters.
- Carbohydrate rich diet is lipogenic; Glucose \rightarrow Pyruvate \rightarrow Acetyl CoA \rightarrow Fatty acids & cholesterol.
- To avoid hypertriglyceridemia or hypercholesterolemia: Carbohydrate intake should be restricted.

Preferred Fuel

01:03:30

- In the absence of oxygen, only glucose will be used; anaerobic glycolysis \rightarrow 2 ATP's generated.
- Fatty and amino acids can only be used in aerobic conditions.
- The choice of fuel will depend on Aerobic or anaerobic.
- If the cells are anaerobic, they have to use only glucose. These are:

PYQ: INICET 2021

- RBCs, Retinal cells and Corneal cells
- White muscle fibers (stores glucose as glycogen) without myoglobin
- Renal medulla.
- If the cells are aerobic, they can use either glucose or fatty acids, but prefer fatty acids because glucose on complete oxidation gives only 32 ATPs whereas fatty acid can give 106 ATPs; stearic acid will give 120 ATP.
- All aerobic cells choose fatty acids as preferred fuel including cardiac muscle fibers and red muscle fibers except neurons which use glucose, because fatty acids in circulation is bound to albumin which cannot cross the Blood Brain Barrier.
- RBCs use glucose anaerobically (1 glucose \rightarrow 2 ATP) while Neurons use glucose aerobically (1 glucose \rightarrow 32 ATPs + CO₂ released).



CARBOHYDRATE CHEMISTRY

Classification of Carbohydrates and Simple Carbohydrates

1. Carbohydrates
2. Classification of Carbohydrates
 - 2.1 Classification of monosaccharides
 - 2.2 Disaccharides**
 - 2.3 Classification of Polysaccharides**
3. One Liners
4. MCQs
5. Integrated Clinical Case-Based MCQS

Good to Know

Good to Know

Mucopolysaccharidosis

- 1. Definitions**
2. Previous Year Questions
- 3. Identify the Structure of Mucopolysaccharides**
4. Tips to Identify the Structure
- 5. Location of MPS**
 - 5.1 Classes of Enzymes
 - 5.2 The Functions of MPS**
6. One-liners
7. MCQs
8. Hunter's and Hurler's Syndrome
9. Image Based MCQ

Good to Know

Good to Know

Must Know

Good to Know

Complex Carbohydrates

1. Glycoproteins & Proteoglycans
 - 1.1 Paroxysmal Nocturnal Hemoglobinuria

2. MCQ

Must Know

Isomerism

1. Classification of Isomerism
2. Structure of Glucose and Fructose
3. Stereoisomerism
4. Glucose Solution
5. One-Liners
6. MCQs

Urine Investigations and Clinical Diagnosis

1. Classification of Carbohydrates Based on Reducing Property

1.1 Tests

Must Know

1.2 Clinical Cause of Reducing Sugars in Urine

1.3 Osazone Test

1.4 Algorithm/Summary

2. One Liners

3. MCQ

4. Integrated Case Based MCQs

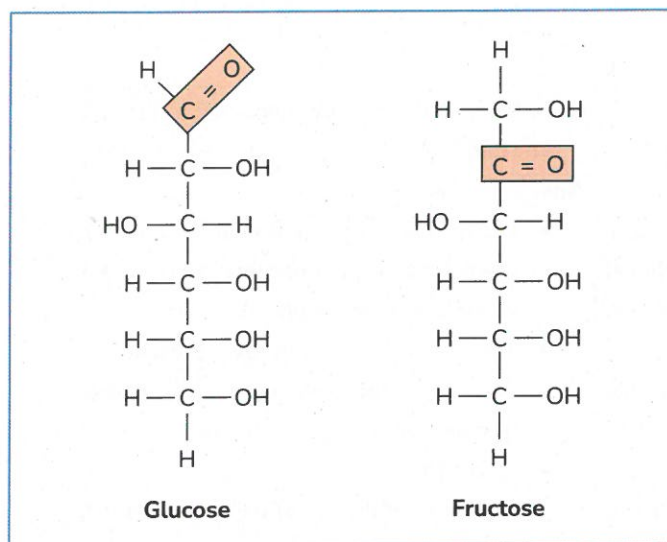
5. Alkaptonuria

Must Know

6. Image Based MCQs



Carbohydrates



- All carbohydrates are defined as polyhydroxy aldoses or ketoses.
- Having general molecular formula - $\text{C}_n\text{H}_{2n}\text{O}_n$

Classification of Carbohydrates

Simple carbohydrates	Complex carbohydrates
<ul style="list-style-type: none"> • They have carbohydrate units only 	<ul style="list-style-type: none"> • Carbohydrate attaches to protein or lipid and is of 3 types: <ul style="list-style-type: none"> ○ Glycoproteins ○ Proteoglycans ○ Glycolipids

Q. How to classify the simple carbohydrate where only carbohydrate units are present?

Ans.

- The classification of simple carbohydrates is based on the number of carbohydrate units—into three types:
 - **Monosaccharides**—single carbohydrate unit in them
 - **Oligosaccharides**—2 to 10 units of carbohydrates. E.g., **disaccharides**
 - **Polysaccharides**—these have more than 11 units of polysaccharides

Classification of monosaccharides

- Two ways to classify the monosaccharides.
 - Number of carbon atoms
 - Functional group

Based on Number of carbon atoms	Based on functional group
<ul style="list-style-type: none"> • Trioses - 3 carbon atoms. • Tetroses - 4 carbon atoms. • Pentoses - 5 carbon atoms. • Hexoses - 6 carbon atoms 	<ul style="list-style-type: none"> • Aldoses - aldehyde group • Ketoses - ketone group

Q. In glucose, how many functional groups do you find?

Ans.

- Only one functional group is present in glucose and that is monosaccharide. The functional group that presents in glucose is aldehyde. Therefore, **glucose is an Aldose.**
- **Glucose is a hexose → glucose has 6 carbon atoms.**
- **Quick tips:**
 - Based on the no. of carbon atoms – **glucose is a hexose**
 - Based on the functional group – **glucose is an aldehyde**

Q. In fructose, how many functional groups do you find?

Ans.

- In Fructose, only one functional group is present, that is monosaccharide because it got only one sugar unit.
- The functional group that presents in fructose is the Ketone group – makes fructose ketose.
- **Quick tips:**
 - Based on the no. of carbon atoms – 6 carbon atoms – **fructose is a hexose**
 - Based on the functional group – **one sugar unit makes it a ketose – ketone group.**
- Glucose and fructose share same molecular formula – $C_6H_{12}O_6$.

Facts:

- Based on a misnomer– Triose is a monosaccharide— 3 carbon atoms
 - Misnomer is **maltotriose** — **not a triose – it's a trisaccharide with 3 glucose residues → 3 x 6 = 18 carbon atoms**
- Mal in a compound name: Made up of multiple glucose residues
 - **Maltose** — **2 glucose residues**
 - **Isomaltose** — **2 glucose residues**
 - **Maltotriose** — **3 glucose residues**
- How do you name a ketose from a corresponding aldose name?

Aldose	Ketose
• Ribose	• Ribulose
• Erythrose	• Erythrulose
• Cidoheptose	• Cidoheptulose

- **The name is given by adding UL to the corresponding aldose name.**

Disaccharides

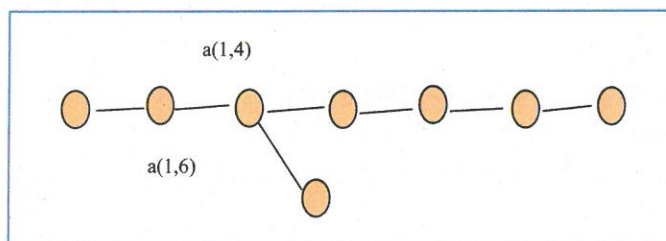
- One of the types of oligosaccharides having -Two sugar units

Disaccharide	Sugars present	Linkage
Maltose	2 glucose residues—glu+glu	$\alpha(1,4)$
Isomaltose	2 glucose residues—glu+glu	$\alpha(1,6)$
Trehalose	2 glucose residues—glu+glu	$\alpha(1,1)$
Sucrose (non-reducing - although made up of reducing agents) Also called table sugar	Glucose and fructose—glu+fru	$\alpha(1,2)$
Lactose (Present in milk)	Galactose + glucose	$\beta(1,4)$

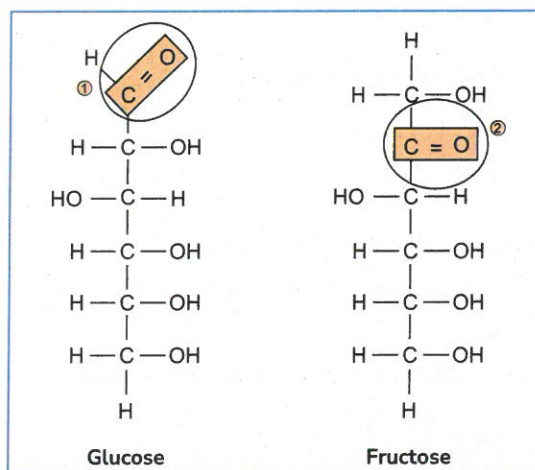
Linkages:

Basic thumb rule

- All the carbohydrates preferred to link with $\alpha(1,4)$ linkage along straight chains. At branch points, they have $\alpha(1,6)$ linkage.



Sucrose



Q. Why are glucose and fructose known as reducing sugars?

Ans.

- Because it has **reduced sugar**, both have free carbonyl groups.
- Glucose — free carbonyl group in first carbon atom
- Fructose — free carbonyl group in second carbon atom

Q. Why is sucrose a non-reducing?

Ans.

- Because sucrose is obtained by linking the 1st carbon atom of glucose & 2nd carbon atom of fructose; linkage presents in sucrose alpha (1,2)

Lactose

- Linkage in lactose is β (1,4).
- One fact to note is humans cannot digest cellulose - a plant diet comes under a fiber diet- plants act as a source of fiber- **plants have got cellulose**
- Undigested food will reach the colon - which will attract water. PYQ: NEET PG 2020
- Most human **digestive enzymes cannot attack β linkages** – linkage present in cellulose, which is made up of **multiple glucose residues** linked multiple **β (1,4) linkage**.
- **The only human digestive enzyme which can attack β linkage is lactase– lactose.**

Lactose Intolerance

00:21:00

- Lactose intolerance is a digestion defect, while other disorders—fructose intolerance and galactosemia are metabolism defects.
- It is caused by a **defect of lactase in the intestinal villi**, leading to impaired digestion of any lactose (Disaccharide) present in the diet into galactose and glucose (Monosaccharides)
- Disaccharides cannot be absorbed; only monosaccharides will be absorbed, as a result they will stay back in the lumen
- Disaccharides are osmotically active (attracting water) causing **osmotic diarrhea**.
- **Clinical presentation of lactose intolerance:**
 - Osmotic diarrhea: due to the absence of lactase in intestinal villi lactose cannot be digested, causing osmotic diarrhea.
 - Flatulence, Frothy stools, and Bloating: Micro-organisms in the colon utilize lactose, converting lactose into hydrogen and methane, causing symptoms
- **Investigation:**
 - IOC for lactose intolerance is methane breath test and hydrogen breath test.
- How are these tests done?
 - The patient needs to fast overnight.
 - Early morning breath sample is taken.
 - Amount of hydrogen or methane is measured based on laboratory preference.
 - Measured qty of lactose is given to the patient – periodic interval breath samples are collected.
 - Estimate the hydrogen or methane level in the sample.
 - **If these hydrogen or methane concentrations increase beyond the physiological level – it is diagnostic of lactose intolerance.**
- Additionally, colonic micro-organisms act on this undigested lactose to form acids – are responsible for acidic pH in stools.
- This **pH acidic in stools is responsible for perianal excoriation – features of lactose intolerance.**

Classification of Polysaccharides

- Have 11 or more numbers of units.

Homopolysaccharides	Heteropolysaccharides
<ul style="list-style-type: none">• Individual units are the same.• They are made up of repetitive units of one sugar moiety	<ul style="list-style-type: none">• Individual units are different.• Mucopolysaccharides/glycosaminoglycans GAG – best example

Homopolysaccharides

Structural Homopolysaccharides	Storage Homopolysaccharides
It acts as a structural component of the cell.	Storage forms of carbohydrates
<ul style="list-style-type: none"> • Examples: <ul style="list-style-type: none"> ○ Cellulose - is a component of plant cell wall— made up of multiple glucose residues— linked with β (1,4) linkages. ○ Inulin - used to measuring glomerular filtration rate – present in plant tubers —made up of fructose. ○ Chitin - exoskeleton scales of snake and cockroaches — made up of N-acetyl glucosamine (NAG) 	<ul style="list-style-type: none"> • Examples: <ul style="list-style-type: none"> ○ Storage form of glucose in plants - Starch ○ Storage form of glucose in animals - Glycogen

Q. Which one will be the branch among these two?

Ans.

- Storage will be **branched** – then only it will accommodate more molecules in limited space.
- Structural homopolysaccharides will be unbranched.

Starch

- Storage form of glucose in plants
- **Starch has 2 components.**
 - **Highly branched amylopectin:** it has multiple glucose residues which are linked by α (1,4) linkage along straight chain and α (1,6) at branch points.
 - Sequential linkages of α (1,4)--- Amylopectin.
 - It is very stable and has high melting temp – 50° – accounts for the solid part of starch that is amylopectin.
 - **Unbranched Amylose** – multiple glucose residues linked by α (1,4) linkages along straight chains
 - Not that stable.
 - Less melting temperature – room temp it is molten – liquid part of starch.

Glycogen

- Storage form of glucose in animals.
- Two tissues that store glycogen in the human body are Liver and Muscle.
- Glycogen is the Highly branched carbohydrate structure.
- It is a Spherical molecule – center protein called as **Glycogenin** – every glucose residue is attached directly or indirectly
 - **Straight chain** – it is directly attached to glycogenin
 - **Branch point** – indirectly attached to glycogenin
 - Straight chain has 11-13 glucose residues – linked by α (1,4) linkages and branch point α (1,6) linkage alone
 - Entire structure is arranged in 12 concentric layers to enable compactness – glucose residues in straight chain will form 1 concentric layer hence 12 layers.

 PYQ: AIIMS 2020

Question: In such a structure, where will you find a greater number of branch points Towards the center or Towards the periphery?

Answer:

- At center → more branch points, at periphery → branch points decrease
- Because glycogen synthesis starts from center and proceeds towards the periphery
- Start of synthesis, you have more no. of glucose molecules – you will introduce many possible branch points.
- Don't have many no. of Glucose molecule – stop creating branch points.

One Liners

- Glucose and fructose are functional isomers.
- Inulin is a homopolysaccharide made up of fructose.
- Chitin is made up of N-Acetyl Glucosamine.
- Non-reducing disaccharides are sucrose and trehalose.
- In starch, amylose is unbranched, and amylopectin is branched.

MCQs

Q. All the following are trioses except

- Maltotriose**
- Glycerose
- Dihydroxyacetone
- Glyceraldehyde

Q. The linkage present in lactose is:

- $\alpha(1,4)$
- $\beta(1,4)$**
- $\alpha(1,2)$
- $\beta(1,2)$

Q. The linkage present in isomaltose is:

- $\alpha(1,4)$**
- $\beta(1,4)$
- $\alpha(1,6)$
- $\beta(1,6)$

Q. All the following are aldoses except

- Glucose
- Galactose
- Ribose
- Ribulose**

Integrated Clinical Case-Based MCQS

Q. A 21-year-old man presents with diarrhea, bloating, flatulence, and frothy stools every time he consumes milk and ghee butter. The probable enzyme defect in this condition is:

- Aldolase B
- Fructokinase
- Galactose-1-phosphate uridylyl Transferase
- Lactase**