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Golden Notes

Biochemistry

Rapid Revision

Handwritten Notes

HIGH-YIELD POINTS | EXAM-ORIENTED | 100+ CLINICAL CASES | MNEMONICS & TRICKS | TABLES & FLOWCHARTS

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JAYPEE

CONTENTS

CHEMISTRY OF CARBOHYDRATES

NMC Competency BC3.1
Discuss and differentiate monosaccharides, di-saccharides and polysaccharides with examples, their importance as energy fuel, structural element, and storage molecule in human body.

SAQ (Short Answer Questions)

1. Define and classify carbohydrates with suitable examples
2. Biologically important disaccharides
3. Homopolysaccharides
4. Heteropolysaccharides and GAGs *(Most Important)*
5. Dietary fiber and its role in diet
6. Glycemic index
7. Functions of dextran
8. Isomers of monosaccharides

Clinical case-1

A 2-year-old boy is brought with progressive abdominal distension, developmental delay, and recurrent respiratory infections. On examination, he has coarse facial features, short stature, hepatosplenomegaly, and corneal clouding. X-ray shows skeletal abnormalities (dysostosis multiplex). Urine analysis reveals increased glycosaminoglycans.

1. What is the most likely diagnosis?
2. Which enzyme is deficient?
3. Which substances accumulate in this condition?
4. What is the inheritance pattern?

Answers

1. Hurler syndrome (Mucopolysaccharidosis type I)
2. Deficiency of α-L-iduronidase
3. Accumulation of: Dermatan sulfate (DS) + Heparan Sulfate (HS)
4. Autosomal recessive

Clinical case-2

A 5-year-old boy is brought with progressive abdominal distension, behavioral problems, and developmental delay. On examination, he has coarse facial features, short stature, and hepatosplenomegaly. There is no corneal clouding. X-ray shows skeletal abnormalities (dysostosis multiplex). Urine analysis reveals increased glycosaminoglycans.

1. What is the most likely diagnosis?
2. Which enzyme is deficient?
3. Which substances accumulate in this condition?
4. What is the inheritance pattern?

Answers

1. Hunter syndrome (Mucopolysaccharidosis type II)
2. Deficiency of Iduronate sulfatase
3. Accumulation of: Dermatan sulfate (DS) + Heparan Sulfate (HS)
4. X-linked recessive

CARBOHYDRATES

Definition:- Polyhydroxy aldehydes or ketones or compounds which produce them on hydrolysis are known as carbohydrates

old definition - Hydrates of carbon ∈ formula $C_nH_{2n}O_n$
but lactic acid, acetic acid are not carbohydrates and deoxyribose is a carbohydrate but Not fits in to $C_nH_{2n}O_n$.

SAQ:- Define and classify carbohydrates with suitable examples.

Classification

- **Monosaccharides** [Simple sugars, single unit]
- **Disaccharides** [two sugar units, connected by glycoside bond]
- **Oligosaccharides** [3-10 sugar units] → Example ① Stachyose ② Raffinose
- **Polysaccharides** [More than 10 units]

On the basis of

No. of C-atoms	functional group	
	Aldehyde [CHO]	Ketone [C=O]
Triose 3C	Glyceraldehyde	DHA
Tetrose 4C	Erythrose	Erythrulose
Pentose 5C	Ribose	Ribulose
Hexose 6C	Glucose	fructose
Heptose 7C	Glucoheptose	Sedoheptulose

Tests for Monosaccharides (Mnemonic - MBBS)

Molisch test :- It is a group test for all carbohydrates.

Benedict's test : for reducing sugars : Bedside test for detection of sugar in urine. Semiquantitative test, gives idea about approximate amount of reducing sugar in urine (useful in DM). Green → 0.5 gm%, Yellow - 1 gm%, Orange - 1.5 gm%, Red - 2gm%, Brick Red → > 2gm%.

Barfoed test :- for monosaccharides only.

Seliwanoff's test : for ketose sugar e.g. fructose

Disaccharides

Reducing

1. Lactose : Glucose + Galactose
   β(1-4) glycosidic bond
2. Maltose : Glucose + Glucose
   α(1-4) glycosidic bond

Non-reducing

1. Sucrose
   [Glucose + Fructose]
   $\alpha_1 - \beta_2$ - glycosidic linkage

   C_1 and C_2 both are involved in the linkage formation, Not available for Osazone formation.
   * Does Not form Osazone
2. Trehalose
   [Glucose + Glucose]
   [$\alpha_1 - \alpha_1$]
   widely used to prolong food shelf life.

Osazone

carbohydrate derivatives formed when reducing sugars are reacted with phenyl hydrazine at boiling temp.

Lactosazone - Powder puff-shaped crystals.

Maltosazone - petal shaped sun flower shaped crystals.

Glucose, fructose, Mannose — Differ around C_1 & C_2 ⇒ This difference is marked when Phenylhydrazine get attached at C_1 & C_2 ⇒ Needle shaped, Broom shaped

Polysaccharide

Homopolysaccharides

[composed of same sugar units]

Heteropolysaccharides

[constituent monosaccharides are different]

HOMOPOLYSACCHARIDES

1. Starch
2. Glycogen
3. Cellulose
4. Dextrin
5. Dextran
6. Inulin
7. Chitin

> 10 Monosaccharides
Constituent monosaccharides are same
Example :- G-G-G-G-G-G-G-G-G-G

1. **Starch** :- storage form of carbohydrates in plant
Composed of **Amylose** + **Amylopection**

Amylose
① Straight chain polymer of D-glucose
② 20% of starch
③ Water soluble
④ α(1-4) glycosidic linkage betn glucose molecules.

Amylopection
① Branched chain polymer of D-glucose
② 80% of starch
③ Insoluble in water
④ α(1-4) in straight chain & α(1-6) in branch point

2. **Glycogen** :- Multibranched polysaccharide of glucose.
Main storage form of glucose in liver and skeletal muscle in animals.
Homopolysaccharide of glucose with α(1-4) glycosidic linkage in linear chain and α(1-6) at branching point.
Branching makes molecule more globular, compact and less space consuming.

3. **Cellulose** :- Polymer of glucose with β(1-4) glycosidic linkage
Can't digested by humans due to lack of enzyme
Cellulose acts as a dietary fibre
Increases bulk of stool, relieves constipation
Decreases absorption of glucose and cholesterol from intestine.

Important in management of DM and obesity

4. **Dextrin**

Partial degradation product of starch. Homopolymer of glucose.

5. **Dextran**

It is a complex, branched, homopolymer of glucose.
It is a plasma volume expander and used in the treatment of hypovolumic shock (MW > 60000 Da)
Also has antithrombotic activity (MW 40000 Da)
It is used in eyedrops as a lubricant
It provides parenteral nutrition.

6. **Inulin** - polymer of fructose
Used in assessment of glomerular filtration rate
Better marker for measurement of GFR over Urea and creatinine. *MCQ
↓
Filtered through glomerulus
Neither reabsorbed, Nor secreted by the renal tubules.
Also can act as a dietary fibre.

7. **Chitin** - present in exoskeleton of insects, cell walls in fungi
polymer of N-acetylglucosamine [amine derivative of glucose]

SAQ:- Enumerate important homopolysaccharides. Write about their functions.

HETEROPOLYSACCHARIDES

Composed of More than 10 sugar units and constituent units (monosaccharides) are different.

1. Glycosaminoglycans (GAG)/Mucopolysaccharides - Most common
2. Agar :- Made up of Glucose & Galactose, used as a bacterial culture media.
3. Agarose :- Made up of Galactose + anhydro galactopyranose used for protein and Nucleic acid electrophoresis.
4. Pectin :- Main content is galacturonic acid, used in food as gelling agent, Also it is a dietary fibre.

GLYCOSAMINOGLYCANS [GAG]

Mucopolysaccharides

Heteropolysaccharides composed of repeating units of disaccharides

⇓ Composed of

Amino sugar	+	Uronic acid
Glucosamine Galactosamine		D-Glucuronic acid L-Iduronic acid

GAGs contains Negatively charged groups [COO^-, SO_4^-, Acetyl]

⇓

Repulsion from each other

- Slippery action [Hyaluronic acid, Mucous]
- Shock absorbent in [Synorial fluid, Vitreous humor, extracellular matrix]

Examples :- Hyaluronic acid
Chondroitin sulfate
MCQ * Dermatan sulfate - N-Acetyl Galactosamine + L-Iduronic acid
Heparin
Heparan sulfate
MCQ * Keratan sulfate → NO uronic acid
N-Acetyl glucosamine + Galactose

MCQ :- HS & CS - Have role in compressibility of cartilage in weight bearing

	GAGs	Composition	Present in	Functions.
1.	Hyaluronic acid	N-Acetyl glucosamine + D-glucuronic acid No sulfate group * MCQ	Synovial fluid * vitreous humor MCQ	Lubricant Shock absorbent Cell migration Wound repair.
2.	Chondroitin sulfate	N-Acetyl galactosamine + D-glucuronic acid	Cartilage Bone Tendon	Shape and structure
3.	Dermatan sulfate	N-Acetyl galactosamine + L Iduronic acid	Skin Valves	Shape and structure
4	Heparin	Glucosamine + Glucuronic acid / Iduronic acid	Lungs mast cells Blood Skin	Anticoagulant * MCQ * causes release of LPL from capillary endothelium
5	Heparan sulfate	Glucosamine + Iduronic acid / Glucuronic acid	Skin Kidney Basement membrane Synaptic vesicles	- Negative charge on * MCQ basement membrane - Attachment of LPL to * MCQ capillary endothelium - Receptor on cell
6	Keratan sulfate	N-Acetyl glucosamine + Galactose * MCQ	Cornea Cartilage	Transparency of cornea Shape & structure

SAQ:- Enumerate different glycosaminoglycans. Add a note on their role in our body.

Proteoglycan :- Mainly contains carbohydrates (GAGs) > 95%.

Glycoproteins :- > 95% protein
< 5% carbohydrates

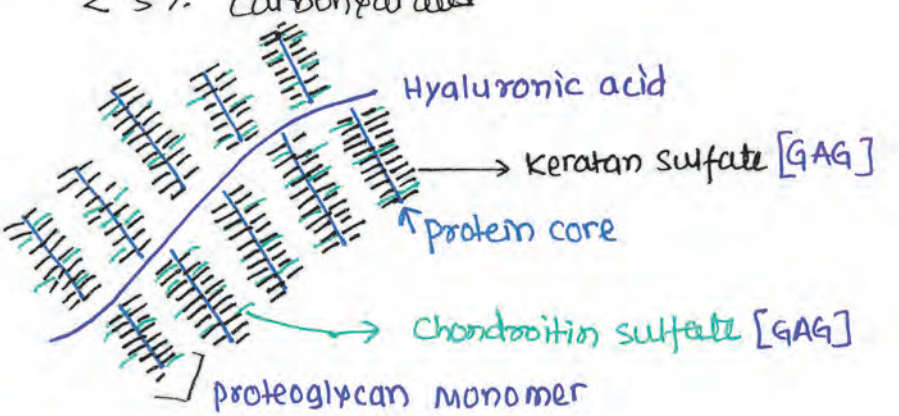


Diagram - Proteoglycan Aggregate

Match the Glycosaminoglycans (GAGs) with Their Functions

1. Hyaluronic acid	A. Provides compressibility to cartilage
2. Chondroitin sulfate	B. Acts as an anticoagulant
3. Heparin	C. Serves as a lubricant in joints
4. tan sulfate	D. Shape and structure of skin, valves, blood vessels
5. Heparan sulfate	E. Transparency of cornea
6. Dermatan sulfate	F. Charge selective GBM permeability

[NEET PG /INICET/ FMGE Examination]

Mucopolysaccharidoses (MPS) - Group of **lysosomal storage disorders** due to defective degradation of **glycosaminoglycans (GAGs)**

Enzyme deficiency → failure to degrade GAGs → Accumulation in lysosomes → **cell damage & organ dysfunction**

Common Clinical Features

- Coarse facial features
- Short stature
- Hepatosplenomegaly
- Developmental delay
- Skeletal deformities (**dysostosis multiplex**)
- ± Corneal clouding

MPS	Name	Enzyme Deficiency	Key Clinical Feature ☆	Intelligence	GAGs Accumulated
MPS I	Hurler	α-L-iduronidase ↓	Severe, coarse facies + **corneal clouding**	↓	**HS + DS**
MPS I S	Scheie	α-L-iduronidase (partial ↓)	Mild adult form	Normal	**HS + DS**
MPS II	Hunter (X-linked)	Iduronate sulfatase ↓	**No corneal clouding** + behavioral issues	↓	**HS + DS**
MPS III	Sanfilippo (A–D)	Enzymes of HS degradation ↓	**Severe CNS involvement**	↓↓↓	**HS only**
MPS IV A	Morquio A	Galactose-6-sulfatase ↓	Skeletal deformity	Normal	**KS**
MPS IV B	Morquio B	β-galactosidase ↓	Skeletal deformity	Normal	**KS**
MPS VI	Maroteaux–Lamy	Arylsulfatase B ↓	Hurler-like body + **normal IQ**	Normal	**DS**
MPS VII	Sly	β-glucuronidase ↓	**Hydrops fetalis**	↓	**HS + DS + CS**
MPS IX	Natowicz	Hyaluronidase ↓	Mild joint disease	Normal	**HA**

GLYCEMIC INDEX [GI]

Definition:- It is a value used to measure how much specific foods increases your blood sugar level. [0 to 100]
It is a system of assigning a number to carbohydrates containing foods according to how much food increases blood sugar.

Importance:-

To lose weight or to maintain a healthy weight
In the management of chronic diseases related to obesity such as DM & coronary artery diseases.

Examples

1. Low glycemic index foods :- green vegetables, Eggs
2. Medium glycemic index foods :- sweet corn, Bananas, oat
3. High glycemic index foods :- white rice, potatoes, breads, sugary foods & drinks

GI of Glucose → 100
Rice → 87
Ice cream → 51
Kidney beans → 24
Soyabeans → 16

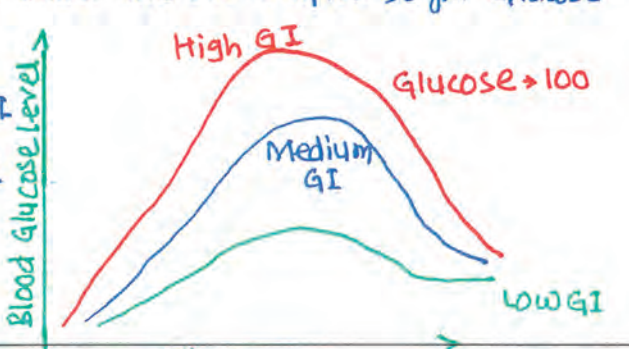
Glucose
Galactose
Lactose
Trehalose
Maltose
Isomaltose
} GI = 100

SAQ :- Define glycemic index. Mention glycemic index of some important foodstuffs.

SAQ :- Describe the importance of glycemic index.

$$\text{Glycemic index} = \frac{\text{Area under the curve after 50 gm Test meal}}{\text{Area under the curve after 50 gm Glucose}} \times 100$$

GI < 55 → Low GI
55-69 → Medium GI
70 OR More → High GI



DIETARY FIBRES

BAQ :- Enumerate four important dietary fibres.
SAQ :- What is a role of dietary fibre in human health?

Definition:- plant components that are NOT broken down by human digestive enzymes.

Names : Cellulose
Hemicellulose
Pectin
Lignin
Gums
Resistant starch
Oligosaccharides
Lactulose [Galactose + fructose] → synthetic disaccharide
Inulin etc.

Sources:- Fruits, dark green vegetables, whole grains, legumes, oats, rye, root tubers, almonds etc.

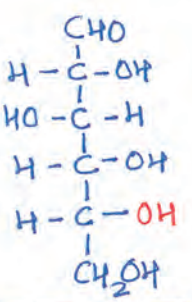
Mechanism of action → Bulking (Absorb water)
Viscosity (Thicken the content of GIT)
fermentation

Important functions

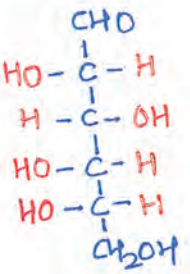
1. Increases bulk of stool and relieves constipation
2. Helpful in weight loss
3. Decreases absorption of glucose from intestine and helps in reduction of blood sugar level in DM.
4. Decreases absorption of cholesterol from intestine and help to reduce cholesterol in patient with CAD, Atherosclerosis, DM etc.
5. Reduces the risk of colon cancer and diverticulosis.
6. Helpful in gastrointestinal disorders

ISOMERS

Stereoisomers: compounds having same molecular formula but differ in orientation of groups in space.
Example: D-Glucose and L-Glucose

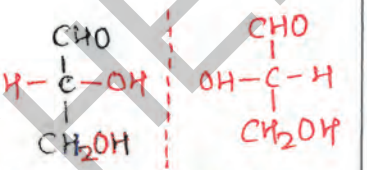


D-Glucose



L-Glucose

Orientation of -OH group to right/left around the carbon atom



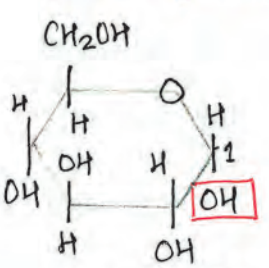
Functional Isomers: compounds having same molecular formula but different functional groups.
Example: Glucose [CHO], fructose [C=O]

Epimers: Compounds having same molecular formula but differ around single C-atom, other than anomeric carbon.

MCQ * Examples: Glucose and Galactose are epimers at 4th
Glucose and Mannose are epimers at 2nd

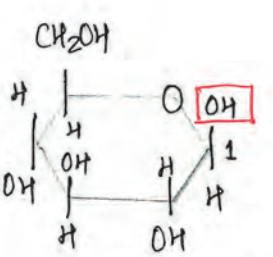
Optical isomers: optically active compounds that differ in their direction of rotation of plane polarized light
to the right (+) or d form
to the left (-) or l form

Anomers: Same molecular formula but differ in orientation of groups around anomeric C-atom. [Carbonyl carbon, 1st in glucose]



α-D Glucose

α → OH is below the plane away from Oxygen



β-D Glucose

β - OH is above the plane towards Oxygen atom

METABOLISM OF CARBOHYDRATES

NMC Competency

BC3.2 Describe the digestion, absorption and transport of carbohydrates from food along with its disorders.

BC3.3 Define and briefly describe the pathways of carbohydrate metabolism and their regulation (glycolysis, gluconeogenesis, TCA, and significance of glycogen metabolism and HMP shunt), with associated disorders.

BC3.4 Describe and discuss the regulation, functions and integration of minor carbohydrate metabolism pathways briefly along with associated diseases/disorders.

BC3.5 Discuss the mechanism and significance of blood glucose regulation (glucose homeostasis) in health and disease. Describe the types, biochemical changes, complications and laboratory investigations related to diabetes & other carbohydrate metabolism disorders.

BC3.6 Interpret the results of analytes associated with metabolism of carbohydrates and other laboratory investigations related to disorders of carbohydrate metabolism.

Important Long answer questions (LAQs)

1. Regulation of blood glucose level
2. Glycogen metabolism, regulation & disorders
3. Gluconeogenesis with regulation
4. Glycolysis with energetics and regulation
5. TCA cycle with energetics and anaplerotic reactions

Short answer questions (SAQs)

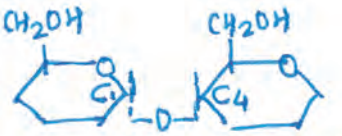
1. HMP shunt
2. Galactosemia
3. Rapoport-Leubering cycle & importance of 2,3-BPG
4. Pyruvate dehydrogenase complex
5. Glycogen storage disorders
6. Anaplerotic reactions of TCA cycle
7. Cori cycle & Cahill cycle
8. Glucose transporters
9. Fate of pyruvate

Clinical Cases

1. Diabetic Ketoacidosis (DKA)
2. Diabetes Mellitus
3. Galactosemia
4. Von Gierke Disease (GSD I)
5. Pompe Disease (GSD II)
6. McArdle Disease (GSD V)
7. G6PD Deficiency
8. Lactose Intolerance
9. Sucrose Intolerance
10. Hereditary Fructose Intolerance

Digestion and absorption of Carbohydrates

Dietary carbohydrates: Starch
Glycogen
Cellulose
Sucrose
Lactose
Fructose
Glucose etc.



α (1-4) glycosidic linkage

↑ Amylase Action

Digestion

Mouth — Partial digestion
- By the action of Salivary amylase
- Amylase breaks α(1-4) glycosidic bond

↓

Stomach (Nil)
- only hydrolysis of sucrose by HCl
- Low pH stops action of Amylase

↓

Intestine — complete digestion
- Pancreatic amylase acts on α(1-4) glycosidic linkage
- Disaccharidase acts on [sucrase, Lactase, Maltase disaccharides Isomaltase]

Intestinal amylase act on terminal α(1-4) linkage and liberates free glucose.

Lactase acts on lactose

Lactose ⟶ Glucose + Galactose

Isomaltase act on α(1-6) linkage, breaks the branching point

Maltase

Maltose ⟶ Glucose + Glucose

Sucrase

Sucrose ⟶ Glucose + Fructose

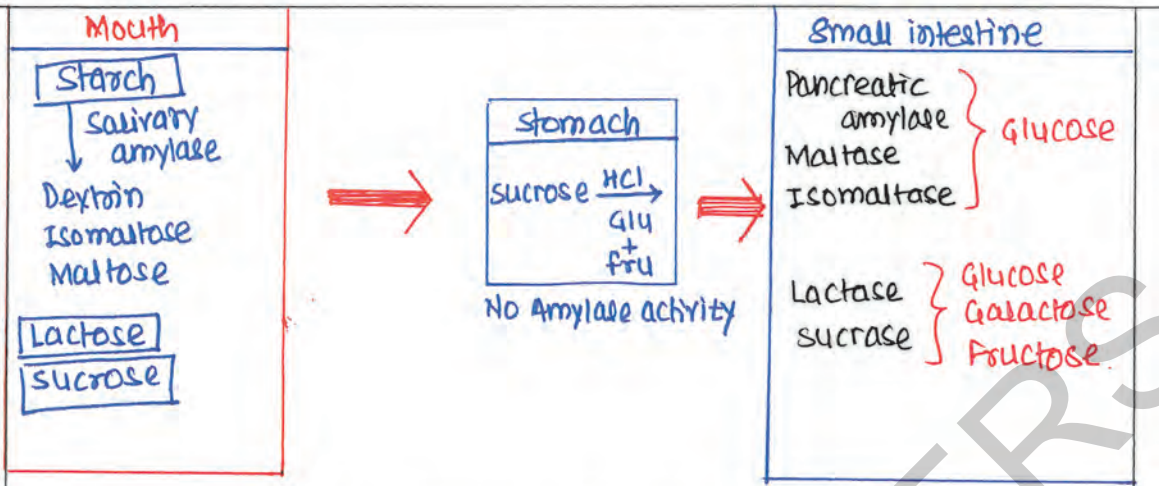


Diagram - Digestion of Carbohydrates

* MCQ

Absorption

Rate of Absorption Gal > Glu > fru

MCQ * Glucose → by secondary active transport along ē Na^+ (symport)

Fructose → by Facilitated diffusion

Pentoses → by passive diffusion

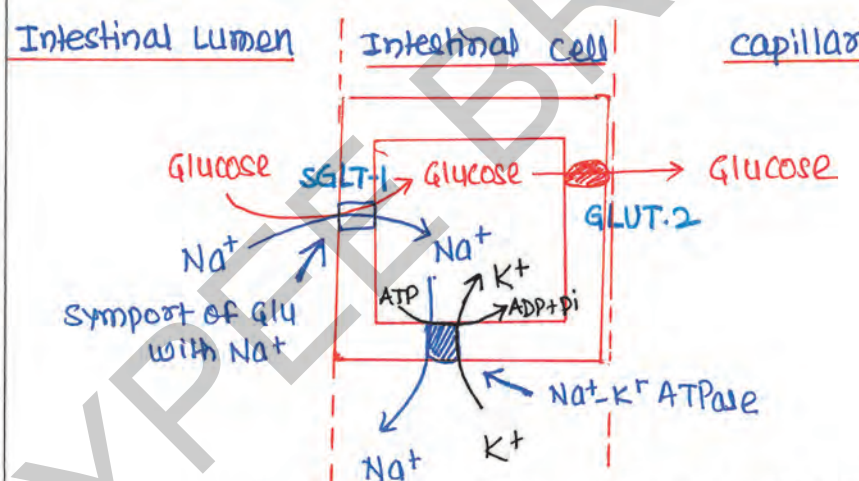


Diagram : transport of glucose across intestinal epithelium

Clinical case

A month old baby had loose stool after every breastfeed. He was crying continuously and irritable. His pediatrician put him on Lactose-free commercial preparation and miracle happened, Now baby is normal.

Probable diagnosis :- Lactose intolerance

Deficient enzyme : Lactase (intestinal brush border enzyme)

Treatment :- Lactose free diet

Commercial preparations are available in market

Symptoms of Lactose intolerance

Abdominal pain, cramps, diarrhea, flatulence etc.

Intestinal bact act on Lactose → Production of Gas: H_2, CO_2, Methane

Clinical case. 2

A 30 year old Male had a loose stool after consumption of sucrose rich diet. He had abdominal pain, cramps and flatulence. Every time when he takes such type of diet, he gets similar symptoms.

Probable diagnosis :- Sucrose intolerance

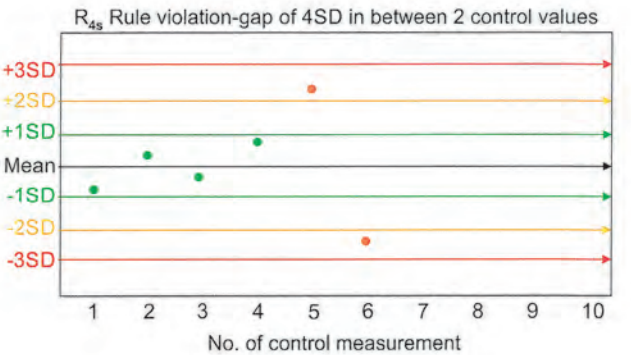
Deficient enzyme : Sucrase

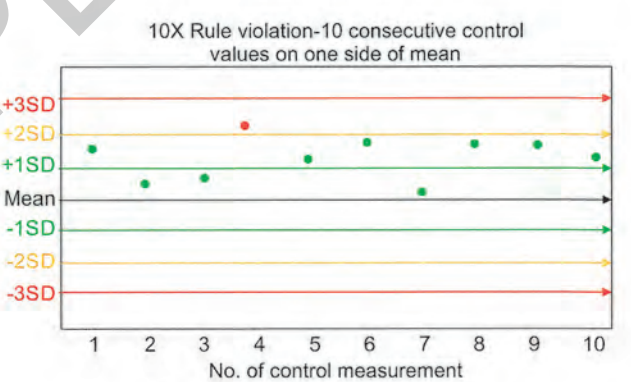
Treatment :- Avoid sucrose in diet
Sugar-free in diet.

Glucose transporters

GLUTs → 1 to 12
facilitated diffusion

Transporter	Tissue location	Function	
GLUT 1	Brain, kidney, colon, placenta, RBC's, Retina [BBB]	Basal glucose uptake	Widely distributed Abundant in RBCs Major in Brain & placenta
GLUT 2	Liver Pancreas B/L surface of intestine PCT [kidneys]	Blood glucose regulation	
GLUT 3	Neuron * placenta kidney		
GLUT 4	Skeletal Muscle Heart Adipose tissue	Insulin dependent	* GLUT 4, 8, 12 Insulin sensitive
GLUT 5	small intestine, Testis Sperm	fructose transport	
GLUT 7	Liver ER	Glucose transport in ER	





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