

LEC 16 META Q:

1. What is the primary result of fructokinase deficiency in the metabolism of fructose?

- A) Severe liver and kidney dysfunction
- B) Accumulation of fructose in the blood and tissues
- C) Fructose in the urine without significant health effects
- D) Decreased ATP production in liver cells

2. What is the main pathophysiological consequence of aldolase B deficiency in fructose metabolism?

- A) Increased production of AMP, leading to gout
- B) Hypoglycemia due to ATP depletion
- C) Excess fructose excretion in the urine
- D) Mild liver dysfunction with no long-term consequences

3. Which of the following compounds is primarily responsible for the osmotic damage seen in uncontrolled diabetes?

- A) Sorbitol
- B) Fructose
- C) Galactose
- D) Glucuronic acid

4. In the pathway from glucose to fructose via sorbitol, which enzyme directly converts sorbitol to fructose?

- A) Aldose reductase
- B) Sorbitol dehydrogenase
- C) Glucokinase
- D) Galactose-1-phosphate uridylyltransferase

5. What is the major consequence of galactose-1-phosphate accumulation in classic galactosemia?

- A) Hyperglycemia and insulin resistance
- B) Cataract formation, liver damage, and mental retardation
- C) Enhanced glycogen synthesis and muscle dysfunction
- D) Increased blood pressure and retinal damage

6. Which enzyme is primarily responsible for converting galactose to galactose-1-phosphate in galactose metabolism?

- A) Galactose-1-phosphate uridylyltransferase
- B) Galactokinase
- C) Aldose reductase
- D) UDP-glucose dehydrogenase

7. Which of the following best describes the role of UDP-galactose in metabolism?

- A) It is primarily used in ATP synthesis
- B) It is involved in the synthesis of glycoproteins, glycolipids, and glycosaminoglycans

- C) It is converted to glucose-1-phosphate in the liver for glycogen storage
- D) It enhances glycolysis in muscle cells

8. What is the key mechanism by which sorbitol contributes to cellular damage in diabetic complications?

- A) Increased sorbitol production leads to hyperglycemia
- B) Sorbitol accumulates in cells, leading to osmotic pressure and water retention
- C) Sorbitol increases the rate of glycolysis in affected cells
- D) Sorbitol inhibits aldose reductase, causing cell dysfunction

9. Which of the following is true regarding the synthesis of lactose in the mammary glands?

- A) Lactose is synthesized through the action of aldose reductase
- B) α -lactalbumin ensures that glucose is incorporated into lactose rather than other sugars
- C) Lactose is formed by the reaction of glucose and UDP-glucose
- D) The enzyme galactosyltransferase is absent in mammary glands

10. How does glucuronic acid contribute to the synthesis of glycosaminoglycans (GAGs)?

- A) It is the main energy source for GAG synthesis
- B) It is incorporated into GAGs as a sugar component
- C) It is used to synthesize glucose-6-phosphate, the main building block for GAGs
- D) It is involved in the phosphorylation of glucose for glycolysis

11. In the context of galactose metabolism, what happens when UDP-galactose is in excess?

- A) It is used for lactose synthesis in mammary glands
- B) It is converted to UDP-glucose for glycogen synthesis
- C) It is excreted in the urine to maintain metabolic balance
- D) It inhibits the formation of glycosaminoglycans

12. Which enzyme is responsible for the conversion of glucose to UDP-glucuronic acid in the glucuronic acid pathway?

- A) UDP-glucose dehydrogenase
- B) Phosphoglucomutase
- C) Galactokinase
- D) Aldolase reductase

13. What is the role of glycosaminoglycans (GAGs) in eye health?

- A) They regulate the synthesis of aqueous humor in the eye
- B) They are involved in the regulation of intraocular pressure (IOP) by controlling fluid drainage
- C) They help in the formation of cataracts in response to high glucose levels
- D) They support the development of photoreceptor cells in the retina

14. How does a deficiency of galactokinase affect the metabolism of galactose?

- A) It leads to the accumulation of galactose-1-phosphate, causing severe liver damage
- B) It activates aldose reductase, leading to the accumulation of galactitol and osmotic damage

- C) It reduces the formation of UDP-galactose, impairing glycoprotein synthesis
- D) It causes a significant increase in the synthesis of GAGs in connective tissues

15. What is the effect of elevated fructose-1-phosphate levels due to aldolase B deficiency?

- A) Decreased ATP synthesis, leading to cellular dysfunction
- B) Increased production of AMP, causing hyperuricemia
- C) Enhanced glucose metabolism and increased insulin resistance
- D) Enhanced production of glucuronic acid and GAGs

Answer Key:

- 1. C
- 2. B
- 3. A
- 4. B
- 5. B
- 6. B
- 7. B
- 8. B
- 9. B
- 10. B
- 11. B
- 12. A
- 13. B
- 14. B
- 15. A

16. What is the mechanism through which aldolase B deficiency leads to the increased degradation of ATP in liver cells?

- A) Increased fructose-1-phosphate leads to depletion of inorganic phosphate (Pi)
- B) Decreased glycolysis causes ATP depletion
- C) Increased levels of UDP-glucose directly interfere with ATP production
- D) Excess AMP inhibits oxidative phosphorylation in mitochondria

17. Which of the following is NOT a consequence of aldolase B deficiency in fructose metabolism?

- A) Increased fructose-1-phosphate accumulation
- B) Decreased AMP levels and inhibited purine degradation
- C) Hypoglycemia due to lack of ATP
- D) Hepatic failure resulting from decreased hepatic ATP

18. What is the primary reason that sorbitol accumulation is problematic in tissues during chronic hyperglycemia in diabetes?

- A) Sorbitol interferes with the function of aldose reductase
- B) Sorbitol accumulation leads to osmotic stress and cellular swelling
- C) Sorbitol inhibits the pentose phosphate pathway
- D) Sorbitol prevents the normal entry of glucose into cells

19. Which enzyme plays a crucial role in converting galactose into a form that can enter synthetic pathways for glycoproteins and glycolipids?

- A) Galactose-1-phosphate uridylyltransferase
- B) UDP-galactose 4-epimerase
- C) Hexokinase
- D) Galactokinase

20. In the context of sorbitol metabolism, which of the following tissues is most affected by excess sorbitol accumulation?

- A) Brain and kidney
- B) Liver and pancreas
- C) Retina, nerve cells, and lens of the eye
- D) Heart and muscle cells

21. What is the mechanism by which sorbitol dehydrogenase contributes to the conversion of glucose to fructose in the body?

- A) It converts glucose directly into fructose via an epimerization step
- B) It oxidizes sorbitol to fructose, generating NADH as a byproduct
- C) It reduces glucose to sorbitol, using NADP⁺ as a cofactor
- D) It promotes the conversion of sorbitol into glucose for glycolysis

22. How does the accumulation of galactose-1-phosphate in classic galactosemia cause mental retardation?

- A) By activating aldose reductase, leading to galactitol accumulation in neurons
- B) By depleting ATP levels, causing neuronal dysfunction and cell death
- C) By inhibiting protein synthesis in the brain
- D) By inducing hyperglycemia, which damages the brain's neural networks

23. In the glucuronic acid metabolism pathway, what is the significance of UDP-glucuronic acid formation?

- A) It serves as a precursor for glucose-6-phosphate in glycolysis
- B) It is used in the synthesis of glycosaminoglycans (GAGs)
- C) It is a key regulator of glucose homeostasis
- D) It is an intermediate in fructose metabolism for energy production

24. Which of the following best explains the role of α -lactalbumin in lactose synthesis in the mammary glands?

- A) It catalyzes the reaction between glucose and galactose to form lactose
- B) It modifies glucose to ensure its exclusive use in lactose synthesis
- C) It enhances the activity of galactosyltransferase in the liver
- D) It converts UDP-galactose into glucose for efficient lactation

25. In the context of galactose metabolism, what role does UDP-galactose play in the biosynthesis of glycosaminoglycans (GAGs)?

- A) UDP-galactose is converted into glucose for GAG synthesis
- B) UDP-galactose is incorporated into GAGs to maintain structural integrity
- C) UDP-galactose inhibits the production of GAGs in the ECM
- D) UDP-galactose helps in the regulation of glucose metabolism during GAG formation

26. How does increased AMP production during fructose metabolism contribute to the pathophysiology of hereditary fructose intolerance?

- A) AMP inhibits the glycolytic pathway, leading to hypoglycemia
- B) AMP accumulates and inhibits ATP production through oxidative phosphorylation
- C) AMP activates the pentose phosphate pathway, causing excessive NADPH production
- D) AMP causes hyperuricemia by increasing the rate of purine degradation

27. Why does the accumulation of galactitol lead to cell bursting in classic galactosemia?

- A) Galactitol disrupts the mitochondrial function, leading to energy depletion
- B) Galactitol alters membrane permeability, causing water influx and swelling
- C) Galactitol inhibits aldose reductase, leading to increased osmotic pressure
- D) Galactitol causes oxidative damage to the cell's lipid membranes

28. What is the relationship between sorbitol and glucose in the metabolic pathways of diabetes?

- A) Sorbitol serves as an alternative energy source when glucose is unavailable
- B) Sorbitol accumulation contributes to hyperglycemia by blocking glucose transport
- C) Sorbitol is produced in tissues with aldose reductase and leads to osmotic damage
- D) Sorbitol is converted to fructose, which increases insulin secretion in pancreatic cells

29. Which of the following metabolic consequences are shared between fructose metabolism disorders (such as fructose intolerance) and galactose metabolism disorders (such as galactosemia)?

- A) Increased production of sorbitol, leading to cell swelling
- B) Depletion of ATP, leading to hypoglycemia and organ dysfunction
- C) Accumulation of glucose-1-phosphate in tissues
- D) Increased production of UDP-glucuronic acid, leading to GAG dysfunction

30. In non-diabetic individuals, when does sorbitol production become significant?

- A) In response to high levels of dietary glucose during fasting
- B) Under conditions of chronic hyperglycemia and insulin resistance
- C) When glucose concentrations exceed normal levels, exceeding the capacity of aldose reductase
- D) Sorbitol production is insignificant in non-diabetic individuals

Answer Key:

- 16. A
- 17. B
- 18. B
- 19. A
- 20. C
- 21. B
- 22. B
- 23. B
- 24. B

- 25. B
- 26. B
- 27. B
- 28. C
- 29. B
- 30. C

31. What is the primary factor responsible for the hypoglycemia observed in hereditary fructose intolerance (HFI)?

- A) Depletion of glycogen stores due to ATP deficiency
- B) Reduced hepatic gluconeogenesis due to ATP depletion
- C) Increased glucose-6-phosphate in the liver
- D) Inhibition of insulin secretion due to excess fructose

32. In the management of hereditary fructose intolerance (HFI), why is it essential to avoid sucrose?

- A) Sucrose is converted into fructose and glucose, exacerbating fructose accumulation
- B) Sucrose directly inhibits aldolase B, worsening the deficiency
- C) Sucrose is a potent inhibitor of the pentose phosphate pathway
- D) Sucrose induces lactic acidosis, which worsens the metabolic disturbance

33. In diabetic complications, sorbitol accumulation primarily leads to which of the following cellular effects?

- A) DNA damage and mutation leading to cancerous transformations
- B) Mitochondrial dysfunction and inhibition of the electron transport chain
- C) Increased osmotic pressure and water retention, leading to cellular swelling and dysfunction
- D) Activation of the NF- κ B pathway, causing chronic inflammation

34. What is the role of UDP-galactose in the synthesis of glycoproteins and glycolipids?

- A) It is converted to UDP-glucose, which is the primary sugar in glycoproteins
- B) It is incorporated into the glycan portion of glycoproteins and glycolipids
- C) It serves as a precursor for glycolysis and ATP production
- D) It prevents the accumulation of excess glucose in glycoproteins

35. In the context of galactose metabolism, which enzyme deficiency leads to the accumulation of galactose-1-phosphate?

- A) Galactose-1-phosphate uridylyltransferase (GALT)
- B) Galactokinase
- C) UDP-glucose dehydrogenase
- D) UDP-galactose epimerase

36. Which of the following best describes the effect of increased AMP levels in individuals with hereditary fructose intolerance?

- A) Activation of ATP production through oxidative phosphorylation
- B) Inhibition of purine degradation, leading to hyperuricemia
- C) Decreased glycolysis and inhibition of glucose utilization
- D) Stimulation of the pentose phosphate pathway for enhanced NADPH production

37. How does aldose reductase contribute to diabetic complications involving sorbitol?

- A) It converts glucose to sorbitol, increasing osmotic pressure and causing cellular damage
- B) It increases the utilization of glucose in glycolysis, preventing sorbitol formation
- C) It stimulates the production of fructose, which regulates glucose homeostasis
- D) It inhibits sorbitol dehydrogenase, preventing the conversion of sorbitol to fructose

38. In classic galactosemia, the accumulation of galactose-1-phosphate is most damaging to which organs?

- A) Liver and kidney
- B) Brain and eye lens
- C) Heart and lungs
- D) Pancreas and spleen

39. Which of the following pathways is most directly impacted by a deficiency in galactose-1-phosphate uridylyltransferase (GALT)?

- A) Glucose 6-phosphate to glycolysis
- B) Conversion of glucose to glycogen via UDP-glucose
- C) Conversion of galactose-1-phosphate to UDP-galactose
- D) Conversion of sorbitol to fructose in the liver

40. Which of the following metabolic changes is most likely to occur in an individual with a deficiency of galactokinase?

- A) Excess UDP-galactose accumulation in the liver
- B) Increased conversion of galactose to galactitol, leading to osmotic damage
- C) Reduced formation of UDP-glucuronic acid for GAG synthesis
- D) Decreased synthesis of lactose in the mammary glands

41. In individuals with fructose-1,6-bisphosphatase deficiency, what is the consequence of impaired gluconeogenesis?

- A) Accumulation of glucose in the blood, causing hyperglycemia
- B) Inability to convert lactate to glucose, leading to hypoglycemia
- C) Decreased fructose-1-phosphate production, worsening hypoglycemia
- D) Reduced formation of glucose-6-phosphate, impairing ATP production

42. What is the direct consequence of increased glucose concentrations in tissues with aldose reductase activity in the context of diabetes?

- A) Increased glucose entry into the cells via insulin-mediated transport
- B) Excess glucose is converted to sorbitol, contributing to diabetic complications
- C) Glucose is metabolized through the pentose phosphate pathway, producing nucleotides
- D) Insulin resistance is alleviated, improving glucose homeostasis

43. What metabolic defect in hereditary fructose intolerance (HFI) directly causes elevated uric acid levels?

- A) Excess fructose-1-phosphate inhibits purine degradation, leading to hyperuricemia
- B) Increased AMP degradation leads to the accumulation of uric acid as a byproduct
- C) ATP depletion results in impaired uric acid excretion by the kidneys

D) Inhibition of the pentose phosphate pathway causes an accumulation of purine bases

44. What is the key mechanism by which GAGs contribute to maintaining intraocular pressure in the eye?

- A) GAGs enhance the filtration of aqueous humor by the trabecular meshwork
- B) GAGs increase the drainage of the aqueous humor through the scleral veins
- C) GAGs reduce the production of aqueous humor by ciliary processes
- D) GAGs increase the resistance to fluid outflow, preventing elevated intraocular pressure

45. How does UDP-glucose dehydrogenase facilitate the conversion of glucose to glucuronic acid?

- A) It catalyzes the reduction of UDP-glucose to UDP-glucuronic acid, utilizing NADH
- B) It oxidizes UDP-glucose to form UDP-glucuronic acid, reducing NAD⁺ to NADH
- C) It transfers glucose from UDP to galactose, generating UDP-glucose
- D) It reduces glucose-6-phosphate to glucose-1-phosphate for glycolysis

46. In the context of diabetic complications, how does the accumulation of sorbitol in the retina contribute to diabetic retinopathy?

- A) By damaging retinal endothelial cells and increasing vascular permeability
- B) By increasing the production of reactive oxygen species (ROS) in retinal cells
- C) By interfering with the function of retinal ganglion cells, leading to blindness
- D) By disrupting the blood-retina barrier through osmotic stress and cellular swelling

47. What is the biochemical rationale for avoiding fructose, sucrose, and sorbitol in the treatment of hereditary fructose intolerance?

- A) These sugars stimulate the production of fructose-1-phosphate, worsening the ATP depletion
- B) These sugars lead to excessive glycolysis, producing lactic acid and increasing lactic acidosis
- C) They enhance the conversion of fructose to glucose, leading to hyperglycemia
- D) They interfere with glycogen breakdown, causing abnormal liver function

48. In the case of galactosemia, which of the following would most likely lead to cataract formation?

- A) Accumulation of galactose-1-phosphate in the lens
- B) High levels of UDP-galactose leading to osmotic imbalance in the eye
- C) Conversion of galactose into galactitol, which causes osmotic damage in the lens
- D) Depletion of ATP in the lens cells, reducing their ability to metabolize galactose

49. How does the accumulation of fructose-1-phosphate in hereditary fructose intolerance affect ATP production in the liver?

- A) It stimulates ATP production through glycolysis
- B) It inhibits ATP production by depleting inorganic phosphate, necessary for ATP synthesis
- C) It enhances oxidative phosphorylation in mitochondria, increasing ATP levels
- D) It leads to the direct phosphorylation of glucose, bypassing ATP utilization

50. In the conversion of glucose to fructose via sorbitol, what is the role of NADPH?

- A) It is reduced during the conversion of glucose to sorbitol
- B) It is oxidized to NADP⁺ when sorbitol is converted to fructose
- C) It serves as a cofactor in the reduction of glucose to fructose directly
- D) It is used in the synthesis of UDP-glucose from glucose-1-phosphate

Answer Key:

- 31. B
- 32. A
- 33. C
- 34. B
- 35. A
- 36. B
- 37. A
- 38. B
- 39. C
- 40. B
- 41. B
- 42. B
- 43. B
- 44. A
- 45. B
- 46. A
- 47. A
- 48. C
- 49. B
- 50. B

Done By: Khaled Ghanayem