LEC 14 META Q:

1. Which of the following is NOT a component of the pyruvate dehydrogenase (PDH) complex?

A) E1 (Pyruvate Decarboxylase)B) E2 (Dihydrolipoyl Transacetylase)C) E3 (Dihydrolipoyl Dehydrogenase)D) FADH2

2. In the PDH complex, what role does lipoic acid play during the conversion of pyruvate to acetyl-CoA?

A) It binds to TPP and facilitates decarboxylation.

- B) It binds the two-carbon fragment and transfers it to CoA to form acetyl-CoA.
- C) It reduces NAD⁺ to NADH.
- D) It helps recycle NADH to NAD⁺.

3. What is the effect of high NADH levels on the PDH complex?

A) It activates the complex.

- B) It inhibits the complex to prevent excessive acetyl-CoA production.
- C) It increases the activity of pyruvate dehydrogenase kinase.
- D) It stimulates the formation of pyruvate from lactate.

4. Pyruvate dehydrogenase (PDH) phosphatase is activated by which of the following?

A) High ATP levelsB) Calcium ions in muscle cellsC) NADHD) Acetyl-CoA

5. Which metabolic condition is primarily caused by a mutation in the E1 subunit of the pyruvate dehydrogenase complex?

A) Von Gierke diseaseB) Pompe diseaseC) PDH deficiency leading to congenital lactic acidosisD) McArdle syndrome

6. What is the primary effect of trivalent arsenic on metabolism?

A) It inhibits glycolysis by binding to phosphofructokinase.

B) It binds to lipoic acid, inhibiting the PDH complex.

- C) It inhibits the TCA cycle by deactivating succinate dehydrogenase.
- D) It activates the pyruvate dehydrogenase complex.

7. What is the primary energy source during prolonged fasting after glycogen stores are depleted?

A) GlycogenB) Fatty acidsC) GluconeogenesisD) Protein catabolism

8. What is the main structural feature of glycogen?

A) It consists primarily of beta-1,4 glycosidic bonds.B) It is composed of glucose residues linked by alpha-1,4 glycosidic bonds with branches formed by alpha-1,6 linkages.C) It is a homopolysaccharide made of ribose.

D) It contains primarily alpha-1,2 linkages at branching points.

9. Which enzyme is responsible for removing branches during glycogen degradation?

A) Glycogen phosphorylase

B) Debranching enzyme

C) Phosphoglucomutase

D) Glucose-6-phosphatase

10. What is the fate of glucose-6-phosphate in liver cells during glycogenolysis?

A) It is immediately converted to glucose and exported to the bloodstream.

B) It is used in glycolysis for energy production.

C) It is stored as glycogen.

D) It is converted to pyruvate for the TCA cycle.

11. Which of the following correctly describes the process of glycogenesis?

A) Glucose-1-phosphate is converted to UDP-glucose by UDP-glucose pyrophosphorylase.

B) Glycogen synthase is activated by phosphorylation during the fed state.

C) UDP-glucose is transferred to glucose-6-phosphate to form glycogen.

D) Branching enzyme adds glucose units to form linear chains.

12. In McArdle syndrome, which enzyme deficiency leads to the disease?

- A) Glycogen phosphorylase in liver cells
- B) Alpha-1,4-glucosidase in lysosomes
- C) Glycogen phosphorylase in muscle cells
- D) Glucose-6-phosphatase in liver cells

13. The most common cause of congenital lactic acidosis related to metabolic defects involves a mutation in which enzyme?

A) Pyruvate kinase

- B) Phosphofructokinase
- C) Pyruvate dehydrogenase complex (E1 subunit)
- D) Lactate dehydrogenase

14. What is the primary metabolic consequence of Pompe disease?

A) Severe hypoglycemia due to defects in gluconeogenesis

B) Accumulation of glycogen in lysosomal vacuoles

C) Reduced pyruvate dehydrogenase activity leading to lactic acidosis

D) Defective ATP production in muscle cells

15. During muscle contraction, calcium ions play a key role in glycogenolysis by:

A) Activating protein kinase A (PKA) to phosphorylate glycogen synthase

B) Stimulating glycogen phosphorylase kinase, which activates glycogen phosphorylase

C) Inactivating glycogen phosphorylase kinase

D) Promoting glucose uptake into the muscle cells

Answers:

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

15. B

16. In the context of pyruvate dehydrogenase regulation, which of the following is a direct consequence of increased levels of acetyl-CoA?

- A) Activation of pyruvate dehydrogenase phosphatase
- B) Inhibition of pyruvate dehydrogenase kinase
- C) Inhibition of the PDH complex through feedback inhibition
- D) Decreased pyruvate dehydrogenase activity in muscle cells

17. What is the primary role of NAD+ in the pyruvate dehydrogenase complex?

- A) It serves as a cofactor for the E2 subunit to transfer the acetyl group to CoA.
- B) It is involved in the regeneration of lipoic acid during the catalytic cycle.
- C) It oxidizes the lipoamide group to regenerate the active site for further reactions.
- D) It facilitates the decarboxylation of pyruvate by stabilizing the TPP enzyme.

18. In the context of metabolic disease, the buildup of lactate in the blood is typically associated with:

- A) Insufficient oxygen for oxidative phosphorylation
- B) Decreased NADH to NAD+ conversion
- C) Defects in the TCA cycle enzymes
- D) Inhibition of lactate dehydrogenase activity

19. Which of the following would be the most likely outcome of a complete loss of function in the PDH complex in a developing embryo?

- A) Decreased glucose production through gluconeogenesis
- B) Accumulation of pyruvate leading to lactic acidosis
- C) Increased pyruvate decarboxylation to acetyl-CoA
- D) Enhanced fatty acid oxidation to compensate for energy loss

20. How does calcium signaling regulate the activity of the PDH complex in muscle cells during exercise?

A) Calcium directly activates pyruvate dehydrogenase kinase, thereby phosphorylating and inactivating the PDH complex.

B) Calcium activates pyruvate dehydrogenase phosphatase, which dephosphorylates and reactivates the PDH complex.

C) Calcium inhibits the action of acetyl-CoA, increasing the PDH complex activity. D) Calcium levels modulate the NADH/NAD+ ratio, which in turn affects PDH complex activity.

21. Which of the following best describes the role of the branched-chain dehydrogenase complex in metabolism?

A) It is responsible for converting branched-chain amino acids into acetyl-CoA for entry into the TCA cycle.

B) It assists in the oxidative decarboxylation of branched-chain fatty acids into acetyl-CoA.

C) It catalyzes the conversion of pyruvate into lactate under anaerobic conditions.

D) It plays a key role in the conversion of glucose-1-phosphate to UDP-glucose.

22. Which enzyme is responsible for transferring the two-carbon acetyl group from lipoamide to CoA in the PDH complex?

- A) Pyruvate decarboxylase
- B) Dihydrolipoyl dehydrogenase
- C) Dihydrolipoyl transacetylase
- D) Lipoamide acetyltransferase

23. What is the effect of high levels of glucagon on the pyruvate dehydrogenase complex in liver cells?

A) It activates the PDH complex by promoting dephosphorylation.

B) It promotes the phosphorylation of PDH, leading to its inactivation.

C) It enhances acetyl-CoA production to support gluconeogenesis.

D) It stimulates the PDH complex through an increase in calcium ion concentration.

24. In the PDH complex, which of the following is true about the cofactor thiamine pyrophosphate (TPP)?

A) TPP directly catalyzes the transfer of the two-carbon acetyl group to CoA.

B) TPP facilitates the decarboxylation of pyruvate in the E1 subunit.

C) TPP is required by the E3 subunit to regenerate NADH from NAD+.

D) TPP is involved in the reduction of lipoamide in the E2 subunit.

25. What happens in the liver when there is a complete defect in glucose-6-phosphatase?

A) The liver cannot produce glucose for systemic use, leading to hypoglycemia.

B) The liver accumulates excess glycogen, causing hepatomegaly.

C) The TCA cycle is unable to process glucose, leading to severe metabolic acidosis.

D) The liver produces excessive lactate, causing metabolic acidosis.

26. In McArdle disease, what metabolic problem is most directly caused by a deficiency in muscle glycogen phosphorylase?

A) Inability to release glucose from glycogen during exercise, leading to muscle weakness.

B) Inability to convert pyruvate to acetyl-CoA, leading to lactic acidosis.

C) Failure to oxidize fatty acids, resulting in excessive ketone body formation.

D) Blockage of gluconeogenesis, leading to fasting hypoglycemia.

27. The debranching enzyme in glycogen metabolism has which of the following activities?

A) It transfers a glucose unit from a branch to the linear chain and hydrolyzes the α -1,6 linkage.

B) It removes glucose from the non-reducing end of the glycogen molecule to produce glucose-6-phosphate.

C) It catalyzes the formation of α -1,6 linkages during glycogen synthesis.

D) It transfers phosphate groups from glycogen to glucose-6-phosphate.

28. How does a mutation in the E2 subunit of the PDH complex affect pyruvate metabolism?

A) It causes a loss of the acetyl-CoA transfer function, preventing entry into the TCA cycle.

B) It inhibits the decarboxylation of pyruvate, leading to an accumulation of pyruvate.

C) It increases the production of NADH, causing feedback inhibition of the PDH complex.

D) It blocks the regeneration of lipoic acid, disrupting the cycle of the PDH complex.

29. What metabolic condition would most likely result from a defect in the enzyme glucose-6-phosphatase in the liver?

A) Accumulation of glucose-6-phosphate leading to hyperglycemia

B) Impaired gluconeogenesis and glycogenolysis, leading to hypoglycemia

C) Excessive glycogen breakdown, leading to hepatomegaly

D) Increased ketogenesis due to decreased glucose availability

30. The accumulation of NADH in the cell typically leads to which of the following effects on the PDH complex?

A) Activation of the complex through allosteric modulation

B) Increased phosphorylation and inactivation of the PDH complex

C) Decreased pyruvate conversion to acetyl-CoA

D) Increased levels of acetyl-CoA for fatty acid synthesis

Answers:

16. C 17. C 18. A 19. B 20. B
21. A
22. C
23. B
24. B
25. A
26. A
27. A
28. A
29. B
30. B

31. In the PDH complex, which cofactor is responsible for accepting the acetyl group from the hydroxyethyl-TPP intermediate in the E1 subunit?

A) Coenzyme A (CoA) B) Lipoamide C) NAD+ D) FAD

32. The activity of pyruvate dehydrogenase kinase is inhibited by high levels of which of the following molecules?

A) Acetyl-CoAB) ATPC) NADHD) Calcium ions

33. Which of the following best describes the role of the TCA cycle in regulating the activity of the PDH complex?

A) It directly phosphorylates and inhibits the PDH complex through feedback inhibition by NADH and acetyl-CoA.

B) It generates ATP, which activates the PDH complex to promote acetyl-CoA production.C) It provides NADH and FADH2 for electron transport, increasing the activity of PDH.D) It suppresses the PDH complex through the accumulation of citrate, an intermediate of the cycle.

34. Which metabolic pathway is directly linked to the activity of the pyruvate dehydrogenase complex, producing the high-energy intermediate acetyl-CoA?

A) Gluconeogenesis

B) Glycolysis

C) Beta-oxidation of fatty acids

D) Pentose phosphate pathway

35. In the regulation of pyruvate dehydrogenase complex, which of the following is true regarding the role of pyruvate dehydrogenase phosphatase (PDP)?

- A) It inhibits the activity of the PDH complex by phosphorylating its subunits.
- B) It activates the PDH complex by dephosphorylating and reactivating it.
- C) It is activated by high concentrations of NADH.
- D) It converts acetyl-CoA to citrate, enhancing PDH activity.

36. In a condition where there is a defect in the enzyme dihydrolipoyl dehydrogenase (E3), which of the following is most likely to occur?

- A) Accumulation of lipoamide in its reduced form
- B) Inhibition of the decarboxylation of pyruvate
- C) Impaired reduction of NAD+ to NADH
- D) Decreased production of ATP through oxidative phosphorylation

37. Which enzyme in the TCA cycle is directly responsible for converting succinate to fumarate?

- A) Succinate dehydrogenase
- B) Malate dehydrogenase
- C) Fumarase
- D) Aconitase

38. What is the effect of an elevated NADH/NAD+ ratio in the cell on the PDH complex?

- A) Activation of pyruvate dehydrogenase phosphatase
- B) Inhibition of pyruvate dehydrogenase kinase
- C) Increased inhibition of the PDH complex through phosphorylation
- D) Enhancement of the PDH complex activity for acetyl-CoA production

39. Which of the following is most likely to occur when there is a defect in the E1 subunit of the pyruvate dehydrogenase complex?

- A) Reduced activity of the TCA cycle due to insufficient acetyl-CoA production
- B) Decreased ATP production through oxidative phosphorylation
- C) Accumulation of pyruvate and lactate leading to metabolic acidosis
- D) Enhanced breakdown of fatty acids to compensate for the energy deficiency

40. What is the effect of a mutation that reduces the activity of pyruvate dehydrogenase kinase on glucose metabolism?

A) Increased conversion of glucose to lactate due to reduced PDH inhibition

- B) Decreased glucose conversion to acetyl-CoA, limiting entry into the TCA cycle
- C) Increased production of glucose through gluconeogenesis
- D) Enhanced fatty acid synthesis from acetyl-CoA

41. The breakdown of glycogen to glucose-1-phosphate is initiated by which enzyme?

A) Glycogen synthase

B) Glycogen phosphorylase

C) Debranching enzyme

D) Phosphoglucomutase

42. What is the role of the NADH produced in the pyruvate dehydrogenase complex?

A) It directly participates in the decarboxylation of pyruvate.

B) It is used in the electron transport chain for ATP production.

C) It activates pyruvate dehydrogenase kinase to inhibit the PDH complex.

D) It reduces acetyl-CoA to form citrate for the TCA cycle.

43. Which of the following is a major consequence of PDH complex deficiency in the brain during early development?

A) Decreased glucose uptake into the brain

B) Impaired fatty acid oxidation leading to a lack of ATP

C) Impaired oxidative phosphorylation and increased lactate production

D) Increased ketone body synthesis as a compensatory mechanism

44. Which of the following molecules would increase the activity of the pyruvate dehydrogenase complex?

A) High levels of acetyl-CoAB) High levels of NADHC) High levels of ADPD) High levels of glucose

45. How does pyruvate enter the mitochondria for conversion by the PDH complex?

A) Through diffusion across the inner mitochondrial membrane

- B) Via the pyruvate-proton symporter in the inner mitochondrial membrane
- C) Through active transport using ATP energy

D) Via facilitated diffusion through aquaporins

46. What would be the effect of a mutation in the enzyme pyruvate carboxylase on metabolism?

- A) Reduced gluconeogenesis, as pyruvate cannot be converted into oxaloacetate.
- B) Increased pyruvate to acetyl-CoA conversion, leading to excess ketone body production.
- C) Enhanced glucose uptake into the liver to compensate for the lack of acetyl-CoA.
- D) Decreased lactate production, as pyruvate is directed toward the TCA cycle.

47. In the presence of excess glucose, which of the following would occur in the liver to maintain homeostasis?

A) Decrease in glycolytic flux and increase in gluconeogenesis

- B) Inhibition of glycogen synthesis and increased ketone body formation
- C) Activation of the pyruvate dehydrogenase complex to enhance acetyl-CoA production
- D) Increased conversion of acetyl-CoA to fatty acids and storage in adipose tissue

48. A patient with a defect in the enzyme lipoamide dehydrogenase would most likely present with which of the following metabolic issues?

- A) A build-up of lactate due to impaired oxidative decarboxylation of pyruvate
- B) Decreased ATP production from the electron transport chain
- C) Accumulation of acetyl-CoA in tissues
- D) Impaired glycolysis due to lack of NADH regeneration

49. In mitochondrial disorders, what is a likely outcome of defective pyruvate dehydrogenase complex activity in muscle cells?

- A) Increased ATP production due to higher reliance on oxidative phosphorylation
- B) Decreased fatty acid oxidation and increased lactate production during exercise
- C) Enhanced glycolysis to compensate for impaired mitochondrial function
- D) Increased breakdown of muscle protein to supply energy for TCA cycle activity

50. Which of the following would most likely increase during periods of fasting as a compensatory mechanism for decreased glucose availability?

- A) Glycogen breakdown to produce glucose-6-phosphate
- B) Pyruvate decarboxylation to acetyl-CoA in muscle cells
- C) Ketone body production from fatty acids in the liver

D) Increased lactate production from anaerobic glycolysis

Answers:

31. B

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

51. Which of the following statements best explains the biochemical rationale for the conversion of pyruvate to acetyl-CoA by the PDH complex in relation to cellular energy needs?

A) Acetyl-CoA is primarily used for the synthesis of nucleotides and amino acids.

B) Acetyl-CoA enters the TCA cycle to generate ATP via oxidative phosphorylation.

C) Pyruvate is converted to acetyl-CoA to ensure adequate NADH production in anaerobic conditions.

D) Acetyl-CoA is used to synthesize ketone bodies during periods of excess glucose metabolism.

52. Which of the following would lead to the activation of pyruvate dehydrogenase phosphatase (PDP)?

A) High levels of NADH and acetyl-CoA

- B) High intracellular calcium concentration during muscle contraction
- C) Low levels of ATP and high levels of glucose
- D) A decrease in mitochondrial membrane potential

53. In patients with pyruvate dehydrogenase complex deficiency, what metabolic change would most likely be observed in their tissues?

- A) Decreased rate of glycolysis and excessive pyruvate conversion to lactate
- B) Increased conversion of pyruvate to oxaloacetate and impaired gluconeogenesis
- C) A shift from aerobic to anaerobic metabolism leading to increased lactate production
- D) Increased beta-oxidation of fatty acids to compensate for the lack of acetyl-CoA

54. In the regulation of the pyruvate dehydrogenase complex, which molecule is known to act as an allosteric inhibitor of the complex, signaling that the cell's energy needs are met?

A) Coenzyme A (CoA) B) Acetyl-CoA C) NAD+ D) ADP

55. During exercise, muscle cells rely on which of the following mechanisms to ensure sufficient ATP production when pyruvate dehydrogenase complex activity is limited?

A) Increased fatty acid oxidation to provide acetyl-CoA for the TCA cycle

B) Increased reliance on anaerobic glycolysis to generate ATP through lactate production

C) Enhanced pyruvate carboxylase activity to direct pyruvate to gluconeogenesis

D) Increased ketogenesis to provide ketone bodies as an alternative fuel source

56. Which of the following best describes the action of pyruvate dehydrogenase kinase (PDK) in the regulation of the PDH complex?

A) PDK activates the PDH complex by dephosphorylating its subunits.

B) PDK inactivates the PDH complex by phosphorylating its E1 subunit, reducing acetyl-CoA production.

C) PDK enhances PDH activity by increasing the supply of NADH and ATP.

D) PDK acts as a feedback inhibitor, reducing acetyl-CoA synthesis in response to high glucose levels.

57. The enzyme succinate dehydrogenase, which participates in both the TCA cycle and the electron transport chain, is located in which part of the cell?

A) Cytoplasm

B) Mitochondrial outer membrane

C) Mitochondrial matrix

D) Inner mitochondrial membrane

58. Which condition is most likely to result from a defect in the enzyme dihydrolipoamide dehydrogenase (E3), a component of the pyruvate dehydrogenase complex?

- A) Reduced production of acetyl-CoA from pyruvate, leading to decreased energy production
- B) Increased acetyl-CoA accumulation due to impaired oxidative decarboxylation
- C) Inability to synthesize ATP through oxidative phosphorylation

D) Increased formation of pyruvate as a consequence of defective pyruvate transport

59. What is the likely consequence of a mutation in the gene encoding for the TPPbinding site of the pyruvate dehydrogenase E1 subunit?

A) Reduced activity of the pyruvate dehydrogenase complex and impaired acetyl-CoA production

B) Increased production of lactate due to shunting of pyruvate to anaerobic pathways

C) Enhanced conversion of glucose to acetyl-CoA, leading to excess fatty acid synthesis

D) Accumulation of pyruvate in the cytoplasm as it cannot enter the mitochondria efficiently

60. Which of the following molecules would most likely be used to treat a metabolic disorder caused by pyruvate dehydrogenase complex deficiency, aimed at bypassing the defect?

A) Dinitrophenol to increase mitochondrial membrane permeability

B) Lipoic acid as a cofactor to stabilize E3 function

C) Ketone bodies to provide an alternative fuel source for brain and muscle cells

D) Glucose supplements to stimulate insulin secretion and promote glycolysis

61. The citric acid cycle's intermediate, oxaloacetate, can be replenished by which of the following processes when it is used up in the cycle?

A) Gluconeogenesis from lactate

B) Conversion of pyruvate to oxaloacetate via pyruvate carboxylase

C) Beta-oxidation of fatty acids to form acetyl-CoA

D) Deamination of amino acids to form alpha-ketoglutarate

62. Which of the following statements is most accurate regarding the function of the pyruvate dehydrogenase complex in mitochondria?

A) It acts as the primary site for glucose oxidation during anaerobic respiration.

B) It catalyzes the irreversible conversion of pyruvate into acetyl-CoA, linking glycolysis to the TCA cycle.

C) It is located on the inner mitochondrial membrane and generates ATP directly.

D) It provides energy for fatty acid synthesis by converting glucose into long-chain fatty acids.

63. What is the role of NADH in the electron transport chain following the activity of the pyruvate dehydrogenase complex?

A) NADH is used to reduce oxygen molecules to form water.

B) NADH provides electrons that are transferred to Complex I of the electron transport chain.

C) NADH inhibits the electron transport chain when the cell's ATP levels are high.

D) NADH is converted into NAD+ for re-entry into glycolysis and the TCA cycle.

64. How does the presence of elevated acetyl-CoA levels affect the pyruvate dehydrogenase complex?

A) Acetyl-CoA enhances the activity of the PDH complex by increasing the levels of NADH. B) High acetyl-CoA levels act as an allosteric inhibitor, signaling sufficient energy supply and downregulating the PDH complex.

C) Acetyl-CoA directly activates pyruvate carboxylase, promoting gluconeogenesis.

D) Acetyl-CoA stimulates the accumulation of citrate, which accelerates PDH activity.

65. Which of the following processes would be most affected in cells with impaired pyruvate dehydrogenase activity?

A) Fatty acid oxidation and ketogenesis

B) Glycolysis and NADH production

C) Gluconeogenesis and glucose utilization

D) TCA cycle flux and oxidative phosphorylation

66. What would likely occur in a person with a genetic defect in the gene encoding pyruvate dehydrogenase kinase?

A) The PDH complex would be continuously active, leading to excessive acetyl-CoA production.

B) The PDH complex would remain inactive due to an inability to deactivate pyruvate dehydrogenase.

C) Gluconeogenesis would be impaired, leading to low glucose production.

D) ATP synthesis would decrease due to decreased activity of the TCA cycle.

67. In the TCA cycle, which enzyme is responsible for catalyzing the conversion of malate to oxaloacetate?

A) Malate dehydrogenase

B) Fumarase

C) Succinyl-CoA synthetase

D) Citrate synthase

68. Which of the following is the most direct consequence of excessive acetyl-CoA accumulation in the liver?

- A) Increased production of glucose through gluconeogenesis
- B) Enhanced ketone body production and accumulation in the blood

C) Inhibition of fatty acid synthesis due to feedback inhibition

D) Increased oxidative phosphorylation efficiency to generate more ATP

69. In patients with pyruvate dehydrogenase complex deficiency, what would be the most likely form of energy metabolism in brain cells?

A) Increased glucose oxidation through glycolysis and the TCA cycle

- B) Increased ketone body utilization due to impaired pyruvate-to-acetyl-CoA conversion
- C) Shift to fatty acid oxidation as the main energy source
- D) Increased lactate production due to an inability to oxidize pyruvate

70. What is the most likely outcome of long-term fasting on the activity of the pyruvate dehydrogenase complex?

A) Decreased activity due to increased acetyl-CoA levels

- B) Increased activity as the body shifts to aerobic glycolysis for energy
- C) Enhanced activity of pyruvate dehydrogenase kinase to inhibit the PDH complex
- D) Inactivation of the PDH complex to conserve glucose for other tissues

Answers:

51. B 52. B 53. C 54. B 55. A 56. B 57. D 58. A 59. A 60. C 61. B 62. B 63. B 64. B 65. D 66. A 67. A 68. B 69. B 70. A

Done By: Khaled Ghanayem